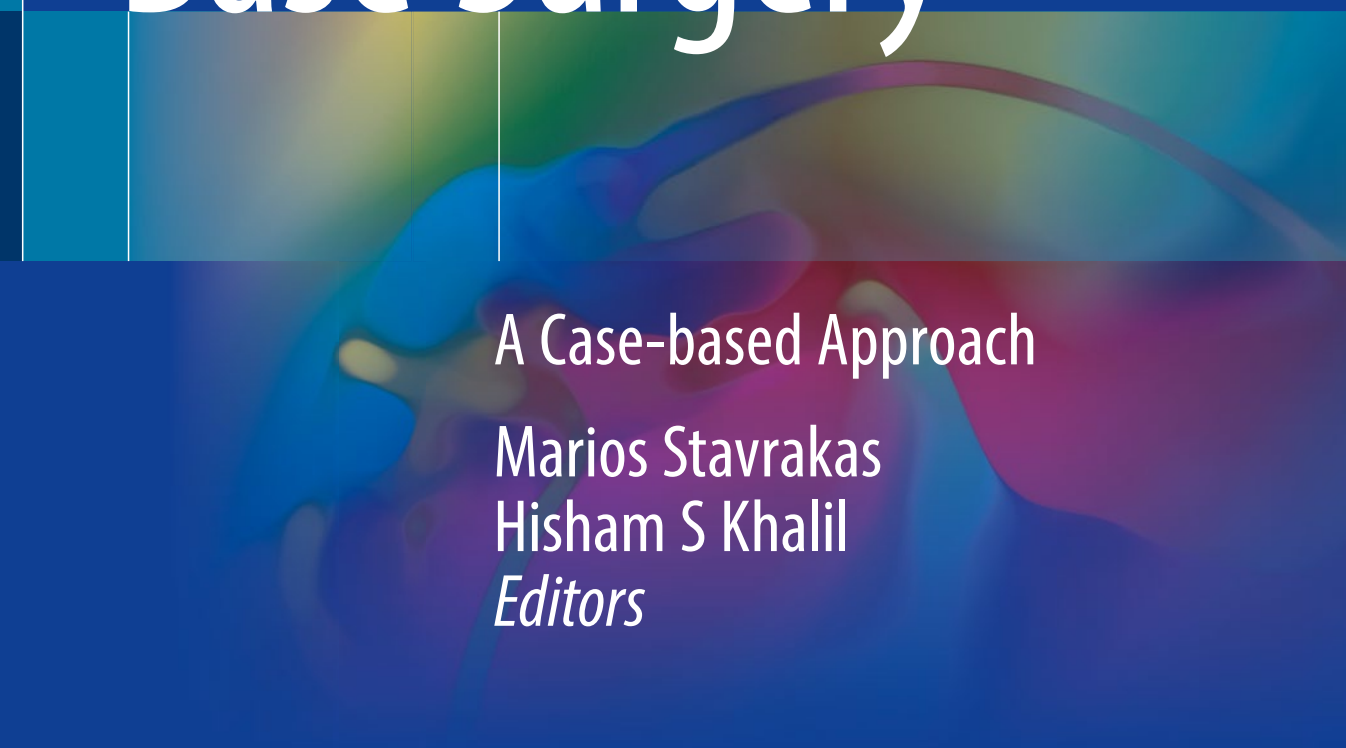
Rhinology and Anterior Skull Base Surgery



A Case-based Approach

Marios Stavrakas
Hisham S Khalil
Editors

Springer

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To my beloved parents, Dimitrios and Amalia, my constant anchors in life. Without your support, personal sacrifice, and unconditional love, I would not be where I am today. To my teachers, mentors, students, and patients in the UK, Greece, and the Netherlands, who made me a better doctor and person. My special appreciation goes to Hisham S Khalil for having believed in me and for introducing me to the fascinating world of rhinology.

—Marios Stavrakas

To my parents, Sawsan and Saleh, my wife Nehal, my daughters Sarah and Nada, and my son-in-law, Nader, thank you for your unwavering love and support in completing this project and many others. I dedicate this book to my patients who gave me the privilege of serving them. Thank you Marios for this great collaboration.

—Hisham S Khalil

Preface

Ὁ βίος βραχὺς, ἡ δὲ τέχνη μακρὴ, ὁ δὲ καιρὸς ὀξὺς, ἡ δὲ πεῖρα σφαλερὴ, ἡ δὲ κρίσις χαλεπή.
Ἱπποκράτης (c. 460 BC–377 BC)

'Life is short, and Art long; the crisis fleeting; experience perilous, and decision difficult' is a famous Hippocratic quotation from his 'aphorisms', and its meaning is of great value in our days, thousands of years after Hippocrates' time. It summarises the challenges that the clinician confronts with the expanding knowledge, innovative technologies, subspecialisation, and novel surgical techniques. Exciting as all these advances may be, doctors throughout the centuries have to employ their experiences, face similar dilemmas, and put their best efforts for the patient.

Rhinology is a rapidly expanding subspecialty of otolaryngology. It has gained popularity due to the advances in technology, the international efforts in the understanding of the pathophysiology and genetics of the various pathologies, and the close relationship with other specialties, such as ophthalmology, neurosurgery, and plastic surgery. Although advanced expertise is identified more and more in tertiary academic centres, the principles of diagnosis and management are applicable at any level of patient care.

Rhinology and Anterior Skull Base Surgery: A Case-Based Approach is a collaborative endeavour to present the wide field of rhinology, anterior skull base, and facial plastics in a concise yet simple and descriptive manner. All the authors have tried to deliver the key points for the treatment of selected cases, based on their clinical and academic experiences. Every case chapter follows a standard format, consisting of a brief case presentation, background knowledge, clinical approach, and finally, a bullet point summary with some comments from the authors. We are hoping that this effort will become a useful reference guide for all the colleagues who are involved in rhinology and otolaryngology in general.

We would like to thank all the colleagues who contributed to this effort, shared their experience, and provided feedback. In addition, we would like to

thank Elaine Leggett for her superb illustrations. We would also like to acknowledge the wonderful editorial staff at Springer for their continuous support in this project. Finally, we wish to thank our patients, who gave a unique meaning to this book.

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Part I

Emergencies in Rhinology

Epistaxis: Conservative Management

1

Konstantinos Geronatsios

1.1 Case Presentation

A 65-year-old male patient presented to the Emergency Department with severe epistaxis. The patient was well—oriented but feeling anxious at the sight of blood. His Blood Pressure was 155/93 mmHg and his heart rate 65 bpm. While applying first aid measures, we took a brief medical history, which included paroxysmal atrial fibrillation, for which he was on Rivaroxaban. The patient was cannulated, blood samples were taken (FBC, Clotting, G&S), and by using a headlight, nasal speculum and nasal suction (Fig. 1.1), we removed the blood and the blood clots from both nostrils. We found that the origin of the epistaxis was a small artery on the right nasal cavity, in the Little's area of the septum. We prepared a cotton pledget soaked with Adrenaline 1/10,000 and Xylocaine 10% and applied it on the bleeding vessel for a few minutes so that we could stop or minimize bleeding. With the use of a Silver Nitrate 75% stick, we managed to cauterize the bleeding vessel. We evaluated the laboratory studies results, which were within the normal range. After 30 min, we inspected once again the nasal cavity, which was clear of blood and clots. We examined both nostrils using a 4 mm rigid 0° nasal endoscope, without any pathological findings. The patient was discharged on the same day.

1.2 Background Knowledge

1.2.1 Anatomy: Pathophysiology

Nasal cavity receives rich vascular supply, provided by several arteries and their branches, with also many anastomoses. The main vessels are the following:

1. Branches of the **Internal Carotid Artery (ICA)** system:
   (a) Anterior ethmoidal artery
   (b) Posterior ethmoidal artery
2. Branches of the **External Carotid Artery (ECA)** system:
   (a) Sphenopalatine artery (SPA), and its branches (predominantly branches into two major vessels, the septal artery, and posterior lateral nasal artery-numerous additional branches may be present)
   (b) Greater Palatine artery
   (c) Superior labial branch of the facial artery
   (d) Infraorbital branch of the maxillary artery
   (e) Lateral nasal arteries

The most common site of anterior epistaxis is **Kiesselbach's plexus (or Little's area)**, located on the anterior cartilaginous septum. It is formed

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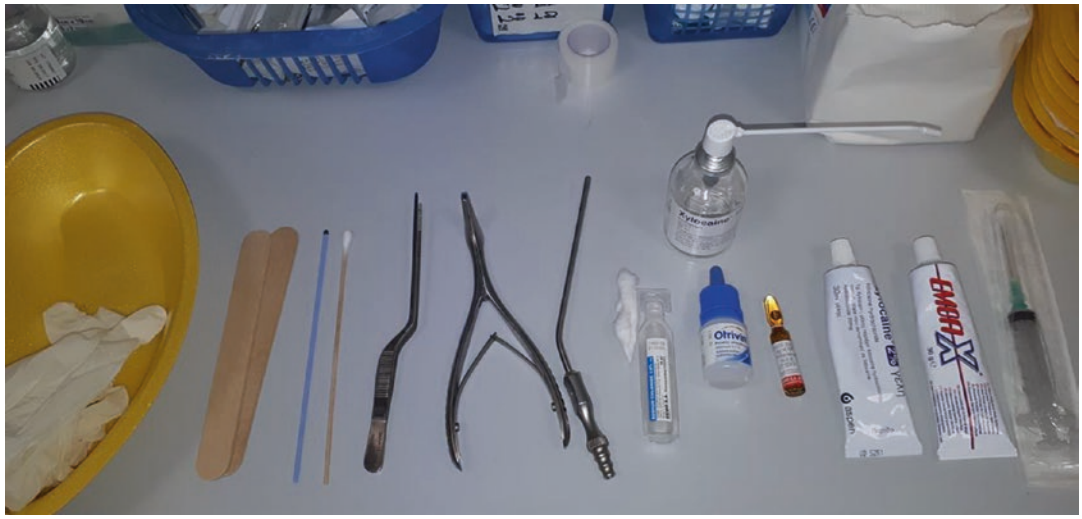

Fig. 1.1 Basic equipment for treatment of epistaxis

by anastomoses of vessels and receives blood supply from both ICA and ECA. As far as posterior epistaxis is concerned, it is usually more severe, more difficult to manage, and of arterial origin (mostly from sphenopalatine artery branches).

1.2.2 Etiology

The causes of epistaxis vary and are divided into **idiopathic**, **local** and **systemic**. Local trauma, the bad habit of nose picking, nasal infections, rhinitis, dry climate and nasal foreign bodies are the most common causes. Other possible causes are:

- *Local*: septal deviation, local inflammation, GPA, septal perforations, chemical irritants, improper use of nasal sprays, sinonasal tumors
- *Systemic:* drugs (e.g., aspirin, clopidogrel, heparin, coumarinic or other anticoagulants, cocaine use, NSAIDs), blood dyscrasias, coagulopathies, HHT, leukemia, arteriosclerosis, hypertension, hepatic failure, Hemophilia, von Willebrand disease

1.3 Clinical Approach

1.3.1 Diagnosis

The initial diagnosis of epistaxis is simple. Blood flow can be seen from the nose, mouth, or both. The most important is to identify the exact bleeding point and try to control bleeding.

A detailed history is of great importance, but sometimes it is better to consider first the possible hemodynamic instability and control significant bleeding. We should always ask about the conditions under which the nosebleed started, the frequency of the episodes, recent sinonasal trauma, history of intranasal foreign bodies, hypertension, coagulopathy—other bleeding disorders, hepatobiliary disease, systemic diseases, and of course the use of medications.

For the detailed diagnosis of epistaxis, we should always have available a suitable ENT headlight, suction, nasal speculums, nasal endoscopes, and suitable forcepses. After removing the clots and blood from both nostrils, we try to identify the origin of bleeding, a procedure that may be difficult in cases of posterior epistaxis or when anatomical disorders, such as a deviated nasal septum, are present. The use of cotton pledgets

soaked with Xylocaine spray 10% and a vasoconstrictor agent (such as Adrenaline 1/10,000, Oxymetazoline drops, cocaine) can be beneficial, as it temporarily controls bleeding. Nasal endoscopy using 0° rigid endoscope is helpful.

Laboratory studies are necessary in cases of a significant nosebleed, in older adults, in suspected coagulopathies, in frequent episodes, and in patients who take anticoagulant medication—especially coumarinic agents. These studies include a full blood count (FBC), coagulation profile (usually PT, aPTT, INR are enough), Urea, Creatinine, Electrolytes, liver function tests, Group and Save +/− crossmatch. More specialized studies should be requested on specific indications.

Imaging studies, such as a CT scan of the sinuses, are indicated mostly in cases of trauma, sinusitis, or possible foreign body. MRI scan is the gold standard in suspected sinonasal malignancy or suspected complications of sinusitis. Angiography is usually not used, except for cases of planned embolism.

1.3.2 Treatment

As mentioned above, after identifying the area of bleeding, we aim to treat the epistaxis, not just to control the hemorrhage. Also, we should apply all ATLS principles (ABCDE approach), stabilize the patient's status, administer intravenous fluids, and transfuse blood in cases of massive severe nosebleed.

Direct pressure to the cartilaginous part of both nostrils is the initial treatment in cases of anterior bleeding. This pressure should be applied continuously for about 5–10 min or more. Head should be kept elevated and slightly tilted forward. Avoid over-extended head position.

If direct pressure is not enough, we use cotton pledgets or gauzes soaked with Adrenaline 1/10,000 plus Xylocaine 10% to the side of the nosebleed, so that we can control or stop bleeding. Instead of Adrenaline, we can alternatively applicate Oxymetazoline, Cocaine 4% solution or Tranexamic acid topically. After these simple measures, we can decide our treatment plan, which can be either cauterization or nasal packing.

Fig. 1.2 A classic bipolar electrocautery device, useful in cases of epistaxis

Cauterization is considered as first-line treatment of nosebleed. Silver nitrate sticks can be useful mostly in cases of anterior epistaxis. Electrocautery (Fig. 1.2) is another choice, effective in cases of anterior and posterior epistaxis. Before the cauterization procedure, we should anesthetize both nostrils using cotton pledgets soaked with a local anesthetic. We should be careful with the method of cauterization, especially in cases of bilateral epistaxis, because it can result in septal perforation.

Nasal packing can be performed with a great variety of materials and gauzes used. It is divided into anterior and posterior nasal packing, with the second one applied in cases of severe posterior or epistaxis uncontrolled with anterior nasal packing. Nasal packing is the suggested method of treatment in cases in which it is difficult to find the origin of the nosebleed, or in cases of diffuse bleeding from the nasal mucosa. Packing should be removed in 1–4 days, depending on the severity, underlying medical conditions, and local clinical protocols. We also advise antibiotic cover if packs remain in situ for more than 48 h to prevent complications, such as Staphylococcal Toxic Shock Syndrome (STSS).

– **Anterior nasal packing**: Petroleum jelly (Vaseline) gauzes, BIPP packs, hemostatic dressings, Nasal Tampons of different sizes and materials with or without a hemostatic agent, absorbable materials with or without hemostatic, inflatable balloon devices (Figs. 1.3 and 1.4). We should perform careful and adequate

anterior nasal packing to avoid posterior dislocation of gauzes, and we always count and write down the number of gauzes we used.

– **Posterior nasal packing**: Rolled gauzes pack with or without a hemostatic agent, inflatable devices with anterior and posterior balloons and two cuffs (Fig. 1.4), Foley balloon catheters (Nr 12 or 14). Posterior nasal packing is always combined with anterior packing.

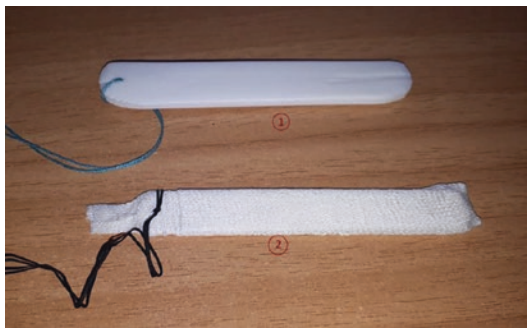

Fig. 1.3 Different types of packs for anterior nasal packing

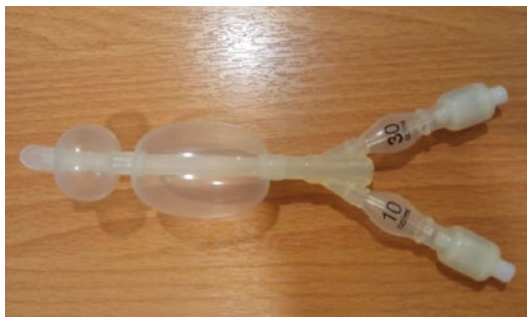

Fig. 1.4 Inflatable device with anterior and posterior balloons, for anterior-posterior nasal packing

We should be selective in which patients should undergo posterior nasal packing because it is associated with discomfort and other severe complications such as posterior dislocation (ingestion, aspiration) of the packs and packing—induced obstructive sleep apnea.

Other choices in conservative management include intravenous or local use of tranexamic acid, hemostatic matrix applied at the target site of bleeding, septal sutures, and a variety of nasal ointments.

In cases of uncontrolled epistaxis, surgical intervention or interventional radiology are preferred.

Figure 1.5 summarises our approach.

1.3.3 Follow-up

Activities such as demanding sports should be avoided for the first days after an episode of severe epistaxis (up to 2 weeks). Patients should also avoid excessive sun exposure, hot shower and facial trauma. A warm and dry environment is not recommended, as it can cause a relapse of epistaxis. Regular nasal irrigations with saline are recommended and can be combined with local application of an antibiotic (e.g., bacitracin) or petrolatum (Vaseline) ointment for moisturization.

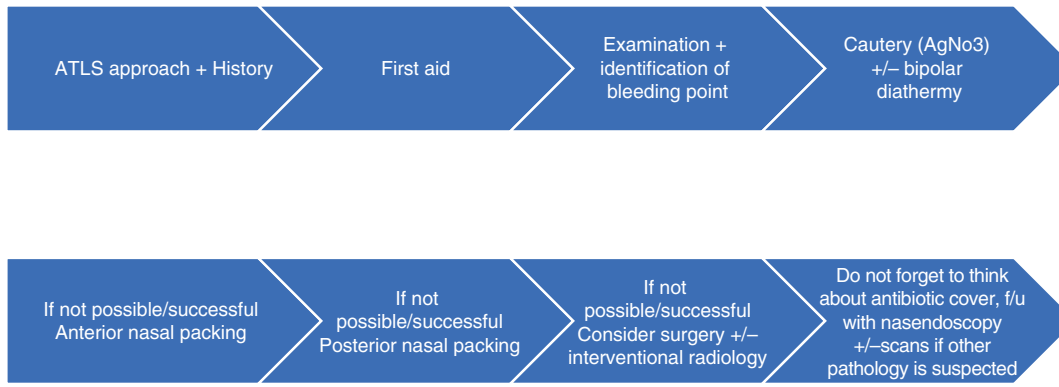

Fig. 1.5 Epistaxis management

In cases of suspected systemic disease, specific laboratory studies should be ordered combined with referral to the appropriate specialty.

If we believe that scoping the patient after treatment may trigger another nosebleed, we strongly recommend reviewing the patient in the clinic after 2 weeks. At this time, nasendoscopy should be performed to exclude malignancy or other pathologies.

Summary and Author's Comments

1. Epistaxis can be a life-threatening ENT emergency.
2. Always try to find the site of epistaxis, so you can choose an effective treatment plan.
3. Don't forget ATLS approach in every case of epistaxis and manage to have always accessible help around (e.g. trained nurse).
4. Detailed history is of great importance, including the use of medications.
5. Your aim is not just to stop bleeding, but to treat the epistaxis and its causes, and also to avoid future episodes.
6. You should always have available the indicated equipment.
7. Excessive or bilateral cautery for epistaxis can result in septal perforation.
8. Anterior and posterior nasal packing should be done carefully, so you can control bleeding and avoid serious complications.
9. Epistaxis can be the first sign of sinonasal malignancy or of a systemic disease.

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Epistaxis: Surgical Management

2

Marios Stavrakas

2.1 Case Presentation

A 26-year old male patient was admitted with severe epistaxis mainly from the right nasal cavity, after sustaining a nose injury while practicing parkour. His nose was packed with anterior packs by the ED doctors but continued bleeding. The ENT registrar inserted posterior packs, but the patient kept bleeding through the packs. He was haemodynamically stable, with a Hb of 12 g/dL. The patient was consented for EUA nose and cautery and right SPA ligation+ right anterior ethmoidal artery ligation. After the surgical treatment of epistaxis, he was kept overnight for observation and had an uneventful recovery. He was discharged the following day and no further follow up was required.

2.2 Background Knowledge

Most centres recommend a stepwise approach for the treatment of epistaxis. Refractory cases may need surgical intervention, while timing and selection of procedures are of paramount importance.

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2.3 Clinical Approach

2.3.1 Treatment

Following the stepwise principle, surgical intervention can be one of the following:

1. EUA nose + nasal cautery→ can be performed under LA or GA, depending on the severity, patient's wishes and available facilities. Bipolar cautery is used, paying attention to avoid over-coagulation, especially in a fashion that may result in septal perforation.
2. SPA ligation→ Frequently performed operation for refractory epistaxis. Essential to keep records of the bleeding side occasionally needs to be done bilaterally. The ligation strategy includes bipolar diathermy, clips, flexible laser, and depends on the accessibility and the surgeon's preference.

   In our unit, after removal of the packing material in the operating theatre, we prepare the nose with 1:10,000 Adrenaline applied on ribbon gauze or neuro patties. After elevating a mucosal flap on the lateral nasal wall, roughly 1 cm anterior to the tail of the middle turbinate, we identify the crista ethmoidalis, which represents the most constant landmark. Behind it, we search for the SPA branches, which can be more than one (Figs. 2.1 and 2.2). We coagulate with bipolar diathermy on a low setting and also cauterise the area of the

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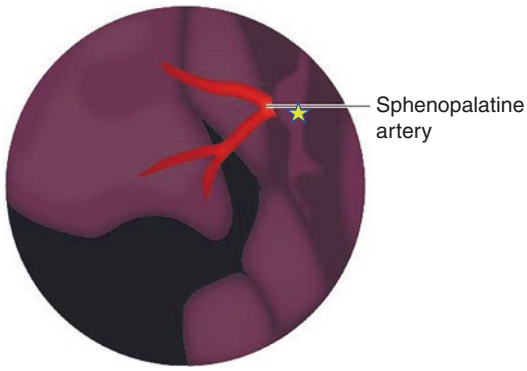

Fig. 2.1 Sphenopalatine artery and its anatomical relation with crista ethmoidalis (yellow star)

posterior septal branch, between the choana and sphenoid sinus ostium. We prefer light packing with dissolvable packing material. Occasionally this procedure can be combined with septoplasty for access or anterior ethmoidal artery ligation.

According to Swords et al. (2017), the average success rate in the literature is 88% for SPA ligation and 89% for internal maxillary artery ligation. It is essential to mention that all studies reported higher success rates for surgical ligation compared to nasal packing (87–90% vs. 48–62%).

3. Anterior ethmoidal artery ligation→ Usually required in traumatic epistaxis. It can be done by an endoscopic or an external approach (Lynch incision). During the external approach, the artery can be identified roughly 24 mm behind the lacrimal crest ('24–12-6' rule) (Fig. 2.3). The authors advocate coagulation with bipolar cautery at a low setting, aiming to avoid thermal injury to the adjacent structures.
4. Interventional radiology→ Embolization is an option for institutions with available facilities. Most studies in the literature describe a diagnostic angiogram of the internal and external carotid arteries, looking for bleeding or potentially dangerous anastomoses. Then, a micro-catheterisation of the distal internal maxillary artery takes place, followed by embolization with one or a combination of embolising materials (polyvinyl alcohol, Gelfoam, micro-coils). A review of the literature described the possible complications: TIA ~10%, stroke 1.1%, tissue necrosis 0.9%, blindness 0.3%. Success rates are reported as high as 75–92%, being comparable, or sometimes better than those for surgical or direct measures.
5. Internal maxillary artery ligation
6. External carotid artery ligation→ It is the last resort and should only be carried out if other methods have failed to control the bleeding. Regarding technical details, it is essential to identify at least two branches of the external carotid artery in the neck before tying it off, preferably with silk ties.

In cases of HHT, our current practice includes packing with dissolvable materials as a first conservative approach and definitive treatment with KTP laser ablation.

The sequence of surgical options described may vary according to the local protocols, surgical expertise and availability of interventional radiology service. The literature shows that early surgical intervention reduces the length of hospital stay and cost.

Lakhani et al. (2013) introduced a scoring tool for severe epistaxis, aiming to facilitate patient selection for SPA ligation. All their patients who underwent SPA ligation met at least one of the following criteria:

I. Persistent posterior epistaxis uncontrolled by packing
II. Hb drop >4 g/dL and/or blood transfusion required
III. Three episodes of recurrent epistaxis requiring re-packing during a single admission
IV. Repeated hospital admissions for recurrent ipsilateral epistaxis (>3 occasions in the last 3 months).

2.3.2 Follow up

In our practice, we tend to follow up postoperatively those who have undergone semi-elective surgery for refractory epistaxis. Of course, this

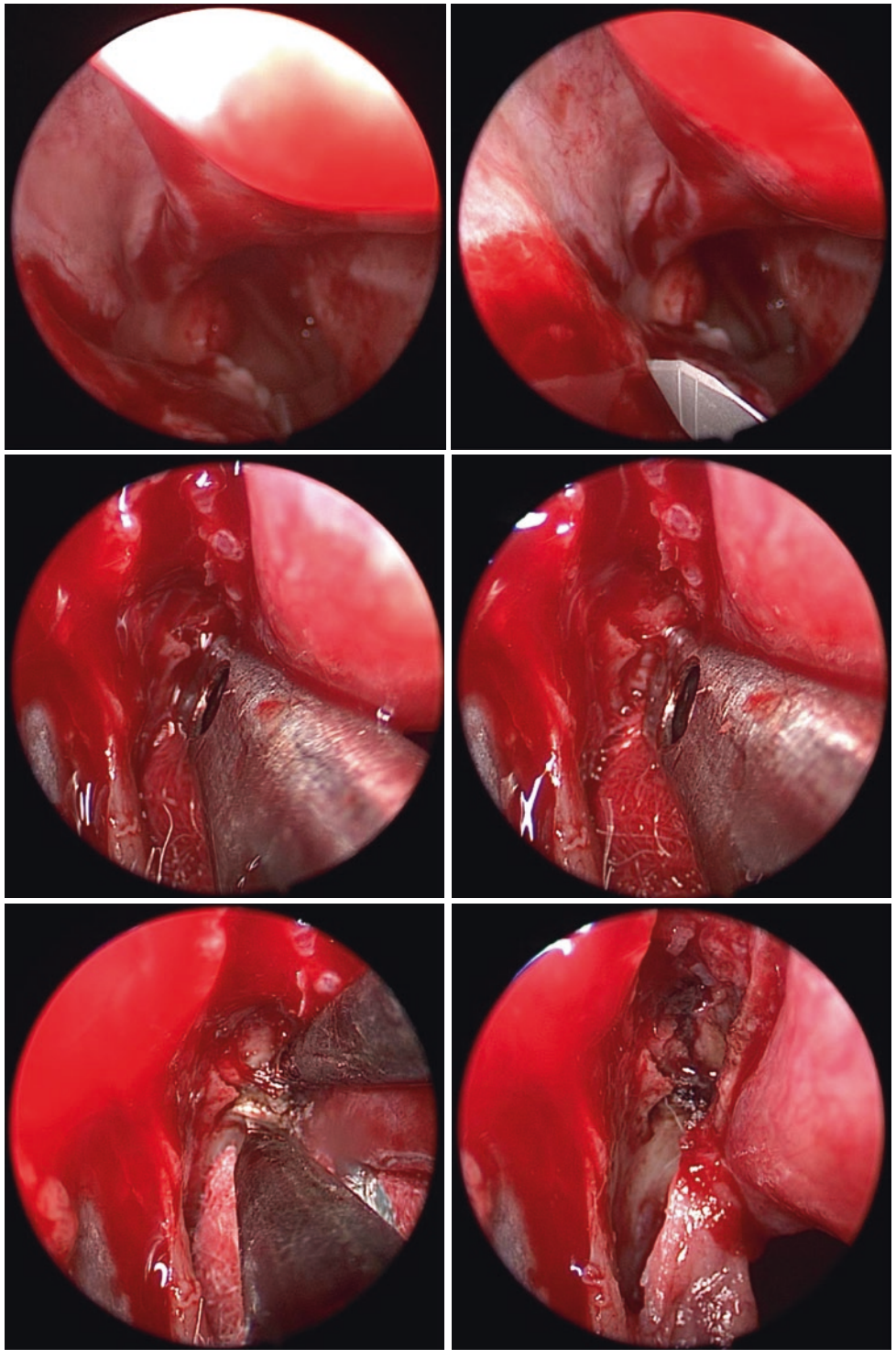

Fig. 2.2 Right SPA ligation. The incision is placed ¬1 cm anterior to the insertion of the middle turbinate, the crista ethmoidalis is identified and the SPA branches cauterised with bipolar

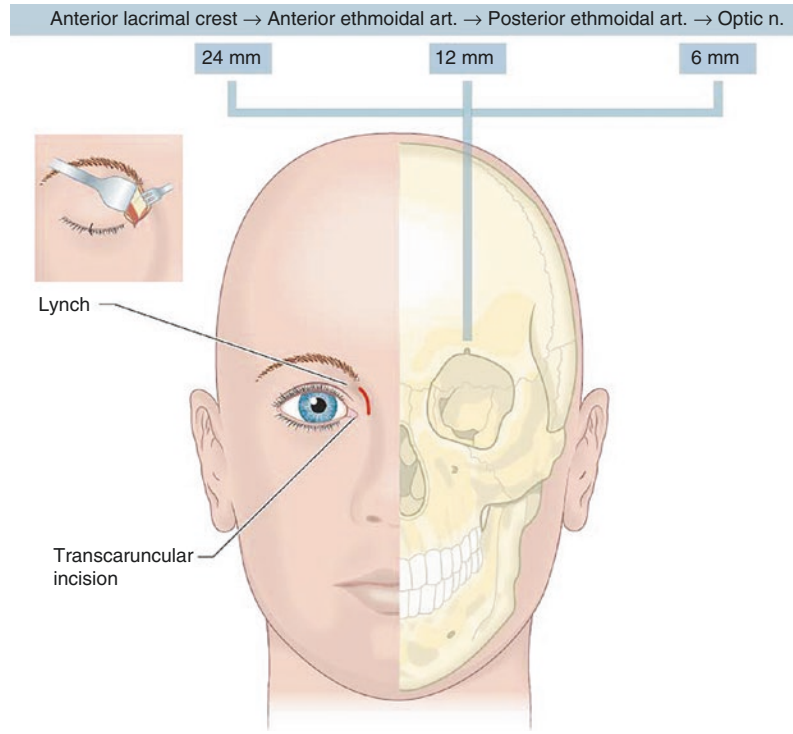

Fig. 2.3 Alternative approaches for anterior ethmoidal artery ligation

decision depends on the surgeon's preference and local protocols. Recurrent epistaxis after surgical treatment may be due to incomplete ligation of all the branches of SPA, or failure to identify the responsible vessels for the bleeding. Patients with HHT who undergo laser treatment, are under regular follow up and further treatment is warranted.

Summary and Author's Comments

1. A stepwise approach is recommended for the treatment of epistaxis.
2. EUA nose + nasal cautery → SPA ligation+/− Anterior ethmoidal artery ligation → Interventional radiology→ Internal maxillary artery ligation → External carotid artery ligation
3. Persistent cases after surgical treatment should be investigated and a Rhinologist should be involved.

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Nasal Fractures

3

Marios Stavrakas

3.1 Case Presentation

A young amateur boxer was brought to the emergency department after sustaining a nasal fracture. He reported a self-limiting episode of epistaxis, pain and change of the shape of his nose, although the oedema had started to increase. An ABCDE approach, according to ATLS principles, was adopted by the Emergency Doctor. Anterior rhinoscopy excluded septal haematoma and active bleeding. After discussion with the on-call ENT team, he was discharged with advice and plan for admission and manipulation under anaesthesia (MUA) after a week.

3.2 Background Knowledge

The nasal bones are the most commonly fractured bones of the face, most probably due to their prominence. There are several causes, such as road traffic accidents, sports accidents, assaults, falls.

3.3 Clinical Approach

3.3.1 Diagnosis

Clinical examination is key to the management of these injuries. Nasal fractures present with pain, oedema, ecchymosis and occasionally epistaxis. The examination consists of two steps:

1. External examination of the nasal pyramid (inspection + palpation): assess deviation of the nasal pyramid, degree of oedema, other facial injuries, nerve deficits
2. Examination of the nasal cavity (anterior rhinoscopy +/− endoscopy): assess epistaxis, septal haematoma, septal deviation, mucosal injury

Plain X-rays of the nasal bones are not always required as they do not add to the diagnosis or management. In the case of complex facial fractures, more detailed imaging should be pursued, mainly CT scan of the facial skeleton. This also allows for 3D reconstruction and better evaluation of the fragments and bony contours.

3.3.2 Treatment

The timing of the intervention is another important issue. It is recommended to perform manipulation of the nasal bones within 10 days for adults and 7 days for children. In general, it is believed

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that nasal fractures should be managed within 2 to 3 weeks, as soon as the oedema has settled. This is because bony callus formation takes place 4 to 5 weeks following the injury. Those patients who present after 4–5 weeks, when the nasal bones are fixed, need thorough consultation and consideration for a septorhinoplasty.

The type of intervention varies, from a simple reduction of nasal bones (closed), open reduction with ORIF for naso-orbito-ethmoid fractures and manipulation of the septum. If there is no nasal deformity, treatment on the acute phase is not required.

The incidence of post-traumatic deformity depends on the timing of diagnosis and intervention. Acute reduction failures can be attributed to traumatic oedema, pre-existing nasal deformity and occult septal injury. If the fracture is left untreated, the incidence of post-traumatic deformity ranges between 14% and 50%.

Paediatric patients have some unique anatomical characteristics that differentiate their management strategy. Babies have large cranial mass compared to their body and consequently are more susceptible to craniofacial injuries. In early childhood, fronto-orbital fractures are more common than middle third fractures, as the mid-face is protected by a prominent forehead and mandible. Moreover, the paediatric facial skeleton is more stable due to reduced pneumatization of the sinuses, a stronger maxilla and mandible from nonerupted permanent dentition. Children have less displaced fractures due to added cushioning from fat pads, more compliant sutures and increased skeletal flexibility.

3.3.3 Follow up

After manipulation under anaesthesis (MUA) of a nasal fracture, the patient needs to be reviewed in 10 days to remove the nasal splint, if its placement was deemed necessary by the surgeon. The majority of patients do not need further assessment. In cases of delayed presentation (>14 days), follow up is required for assessment and discussion about indications of rhinoplasty.

Summary and Author's Comments

1. Early diagnosis and treatment are important for the avoidance of residual deformity.
2. X-rays are not necessary for simple nasal fractures, CT is recommended for complex facial fractures.
3. Timing and type of intervention depend on patient characteristics and type of injury.
4. Special anatomical considerations should be taken into account for paediatric cases.

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4 Orbital Complications of Rhinosinusitis

Hisham S Khalil, Mihiar Atfeh, Ahmed Eweiss, and William Mukonoweshuro

4.1 Case Presentation

A 62 years old man presented to his GP with a severe right temporal headache. He was diagnosed with temporal arteritis and treated with systemic steroids. Five days later, he presented to the Emergency department of a tertiary care centre and admitted with confusion, severe headache and a "swollen" right eye. The patient was initially admitted to the stroke unit with a suspected carotico-cavernous fistula.

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4.2 Clinical Approach

History: On close questioning, the patient had a preceding upper respiratory infection. His headaches were deep-seated and referred to the right temple. He was otherwise fit and well with no significant pat history. He's headaches got worse with double vision in the preceding 2 days. He became very confused when started on dose of prednisolone 60 mg once daily.

Examination and Investigations: The patient was apyrexial and had a right ophthalmoplegia (Fig. 4.1). A full blood count revealed a slightly raised White Blood Count, C Reactive Protein and Plasma Viscosity. The patient was initially admitted to the stroke unit. A carotico-cavernous fistula was suspected, and a CT-angiogram performed (Fig. 4.2, b and c). This demonstrated an opacification of the left sphenoid sinus, a dilatation of the right superior ophthalmic vein and a filling defect in the Right cavernous sinus. There was no evidence of a carotico-cavernous fistula. A diagnosis of a right cavernous sinus thrombosis complicating a left sphenoiditis was made and the patient referred to the ENT Department.

Treatment: Emergency bilateral trans-nasal endoscopic sphenoidotomies were performed. The sphenoid sinus ostia were identified medial to the middle and superior turbinates and approximately 1.5 cm above the choanae. Mucopus was drained from the left sphenoid sinus (Fig. 4.3). The right sphenoid sinus was clear. The patient was discharged on a therapeutic dose of warfarin

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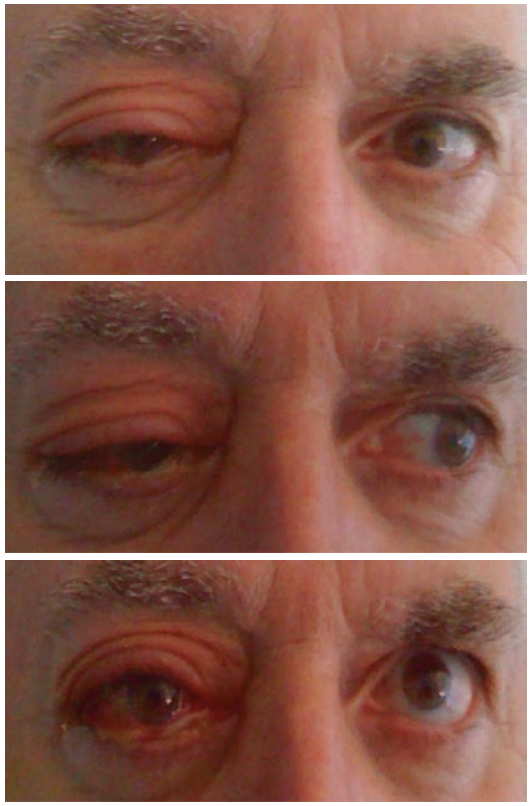

Fig. 4.1 Photograph demonstrating right ophthalmoplegia. Reproduced from University Hospitals NHS Trust guidelines on the management of orbital/periorbital infections

for 6 months. The patient made a complete recovery with resolution of his right ophthalmoplegia in the following 6 weeks.

4.3 Background Knowledge

Orbital complications of rhinosinusitis are rare and should be managed as an ENT emergency. The incidence of orbital complications is higher in the paediatric population and are encountered more in tertiary referral centres. Bacterial pathogens include Haemophilus Infleunza, Streptococcal specious and Staphylococcus Aureus. The complications are categorised according to the extent of sepsis (pre-septal/periorbital vs. post-septal/orbital) or according to the severity (cellulitis/subperiosteal abscess/intra-orbital abscess). The most severe of the orbital complications is cavernous sinus thrombosis. The most accepted classification of the orbital complications is that described by Chandler (Table 4.1).

A CT scan of the sinuses with contrast is the investigation of choice. The assessment of the orbital complications of rhinosinusitis can be challenging in children and where it is difficult to assess the eye and pupillary reflexes due to an inability to separate closed eyelids. Loss of colour vision (tested by an Ishihara Chart) is one of the early signs A detailed ophthalmological assessment is essential including the presence or absence of proptosis, limitation of ocular movement, conjunctival injection/chemosis and direct and indirect pupillary reflexes. An afferent pupillary defect should be excluded.

The care of patients with orbital complications/infections is best carried out by a multidisciplinary team comprising an otolaryngologist, an ophthalmologist, a microbiologist and a paediatrician if the patient is a child. Surgical drainage is indicated in the presence of a subperiosteal abscess as well as sinusitis associated with cavernous sinus thrombosis. The classical approach to drainage of a subperiosteal abscess is through an external approach (Lynch/Seagull incision), reflection of the periosteum over the medial orbital wall along the fronto-ethmoidal suture. The dissection is best assisted using (a) degree nasal endoscope. After drainage of the pus, a communication is established with the nasal cavity through the lamina papyracea and a drain inserted in the medial orbital compartment.

Summary and Author's Comments

Orbital complications of rhinosinusitis can result in serious morbidity including loss of vision, serious intracranial complications and mortality. Hospital admission, careful and regular assessments of the patient are essential with prompt investigations and surgical intervention are essential. Initial presentations may be misleading and mistaken for neurological conditions. Infections of the sphenoid sinuses can result in contralateral orbital complications.

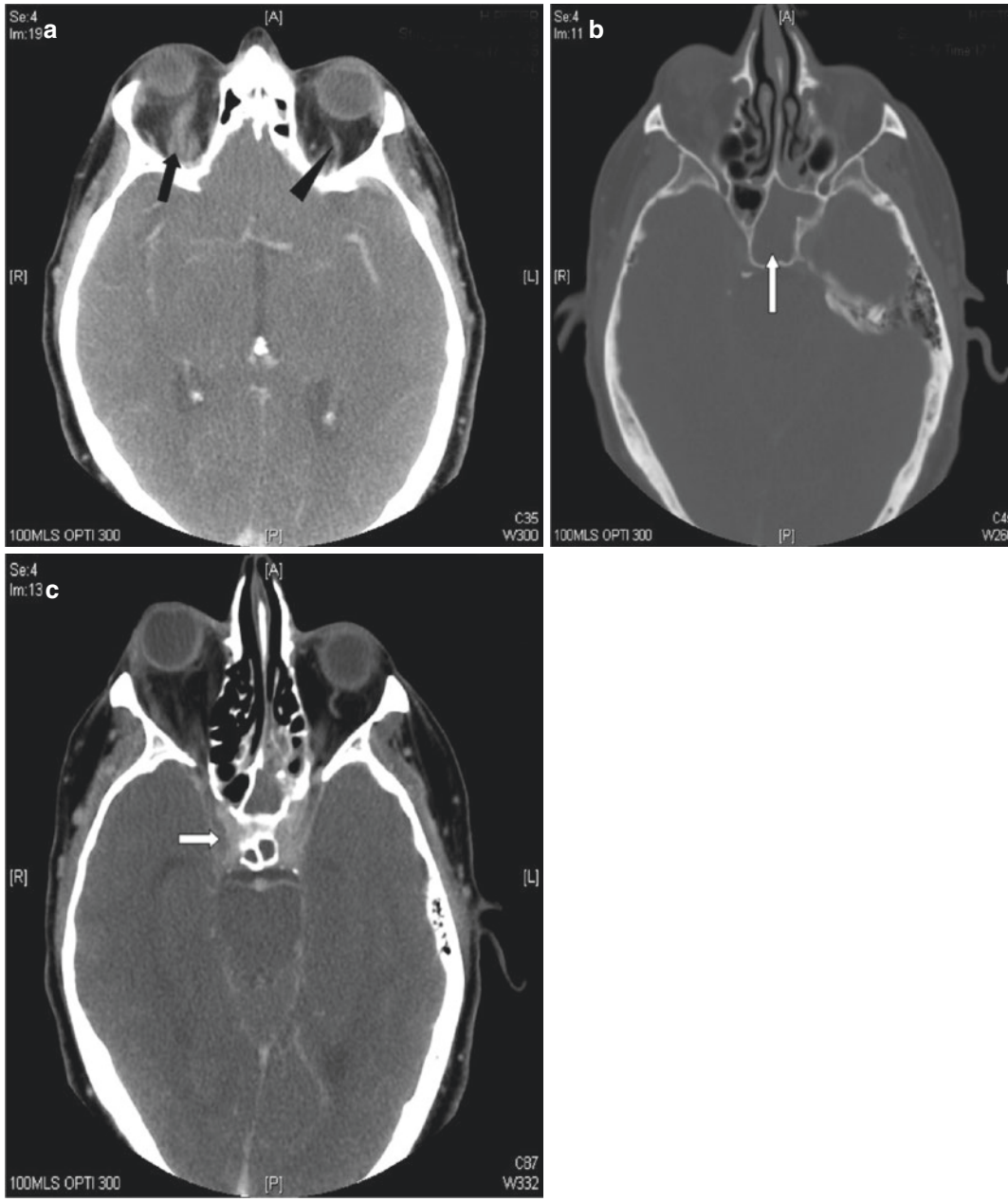

Fig. 4.2 (**a**) CT with contrast demonstrating dilated superior ophthalmic veins (Black arrows). (**b**) CT scan demonstrating opacification of a dominant left sphenoid sinus (White arrow). (**c**) CT scan with contrast demonstrating a defect in right cavernous sinus (White arrow). Reproduced from University Hospitals NHS Trust guidelines on the management of orbital/periorbital infections

Alternatively, experienced Endoscopic Sinus surgeons can drain pus in the medial orbital compartment endoscopically by performing an anterior and posterior ethmoidectomy and removing the lamina papyracea. The role of anticoagulants in the management of cavernous sinus thrombosis is controversial and will depend on the experience and philosophy of the caring team and the patient profile. A flow diagram of the multidisciplinary management of patients with orbital complications/infection in our institution (Fig. 4.4).

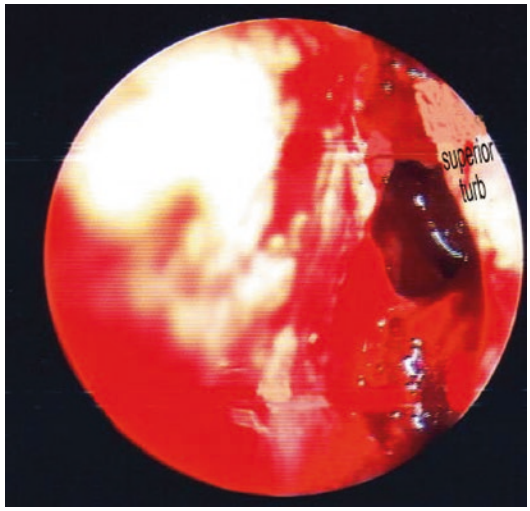

Fig. 4.3 Left sphenoidotomy. Reproduced from University Hospitals NHS Trust guidelines on the management of orbital/periorbital infections

Table 4.1 Chandler's classification of orbital infections

Chandler's stage	Clinical stage
I	Preseptal cellulitis
II	Orbital cellulitis
III	Subperiosteal abscess
IV	Orbital abscess
V	Cavernous sinus thrombosis

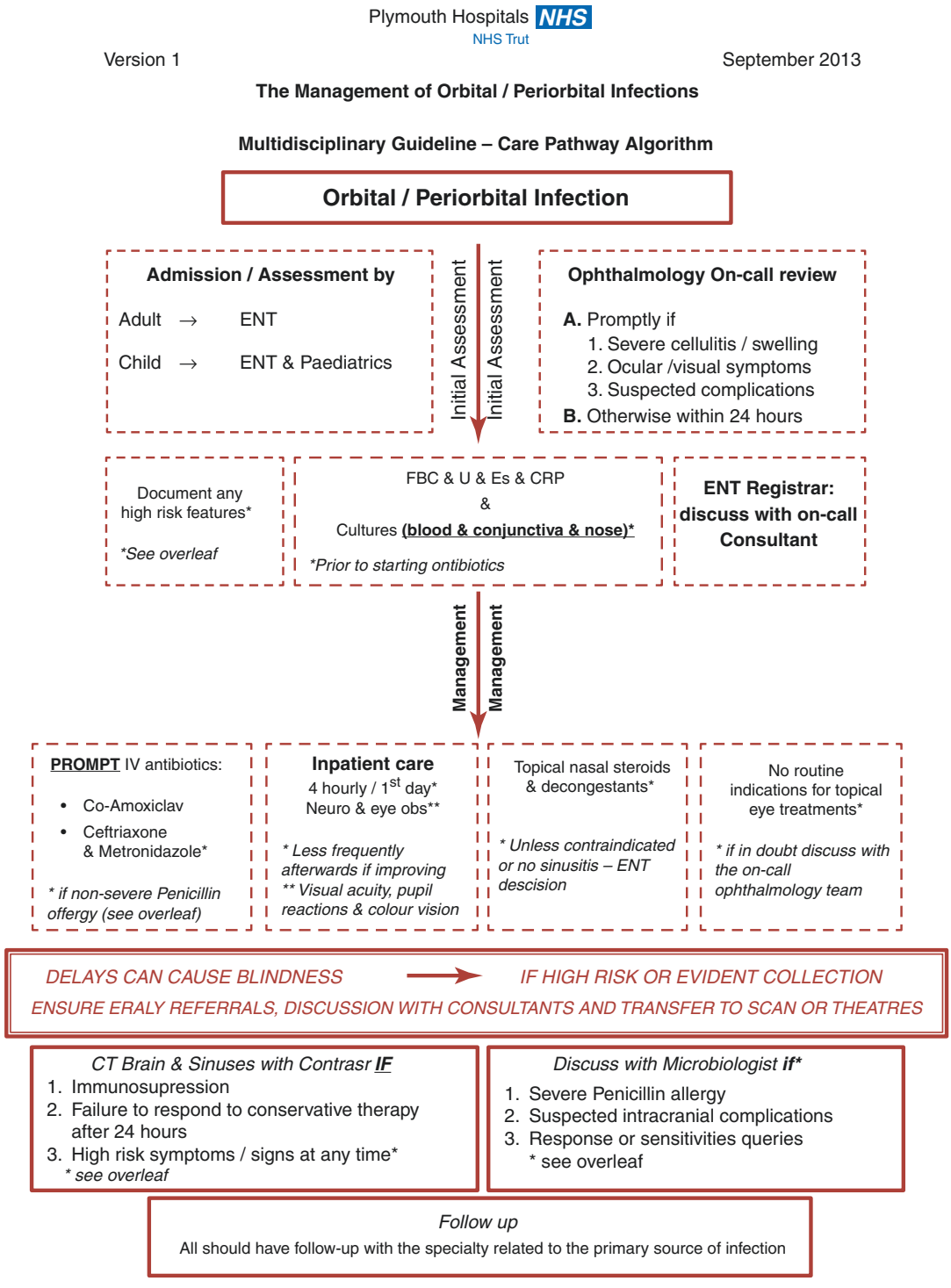

Fig. 4.4 Multidisciplinary evidence-based management. Reproduced from University Hospitals NHS Trust guidelines on the management of orbital/periorbital infections

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Acute Fungal Rhinosinusitis

5

Konstantinos Geronatsios

5.1 Case Presentation

A 39-year-old male patient with a medical history of autologous bone marrow transplantation 2 months ago due to relapse of a B-cell Hodgkin's Lymphoma, was admitted to Haematology Department with symptoms of worsening nasal congestion, purulent nasal discharge, persistent high fever, midfacial pain, and hard palate numbness. An urgent ENT assessment was requested. Nasal endoscopy revealed oedematous nasal mucosa, purulent nasal discharge, and black crust formation inside the right nostril. Multiple swabs, along with tissue samples, were collected for microbiological cultures and histopathological examination. Intraoral examination revealed a black ulcer with irregular margins over the right side of the hard palate, approximately 2×2 cm^2 in dimensions. Cranial nerve function was unremarkable. An urgent contrast-enhanced CT scan of the head and paranasal sinuses was performed, showing right maxillary sinus opacification with bone erosion (Fig. 5.1). Laboratory studies revealed elevated ESR and neutropenia. Blood cultures were also obtained. An urgent sinus MRI (Figs. 5.2 and 5.3) was performed, and the patient was taken to theatre for surgical debriment of the necrotic tissue, middle meatal antrostomies and biopsies under general anesthesia. A high dose of Amphotericin B was administered intravenously. The suspected diagnosis of mucormycosis, a subtype of Acute Invasive Fungal Sinusitis was confirmed.

5.2 Background Knowledge

Acute Invasive Fungal Rhinosinusitis is a rare and extremely aggressive disease with high morbidity and mortality (50–80%). It is related in the vast majority of cases with immunosuppression and especially malignancy, chemotherapy, uncontrolled diabetes, autoimmune disorders, and organ transplantation. There are several saprophytic fungi associated with acute invasive fungal rhinosinusitis. These fungi are inhaled and deposited in the airway, causing local or generalized inflammation to the immunocompromised patients. The most common are Mucor, Rhizopus, Rhizomucor, Absidia, Mortierella, Apophysomyces species and Aspergillus fumigatus, which is also related to the chronic form of invasive fungal sinusitis. Histopathological studies demonstrate mucosal invasion, vasculitis, arterial and venous thrombosis, and eventually, tissue necrosis. As it represents a potentially fatal and rapidly evolving disease, early diagnosis is of great importance.

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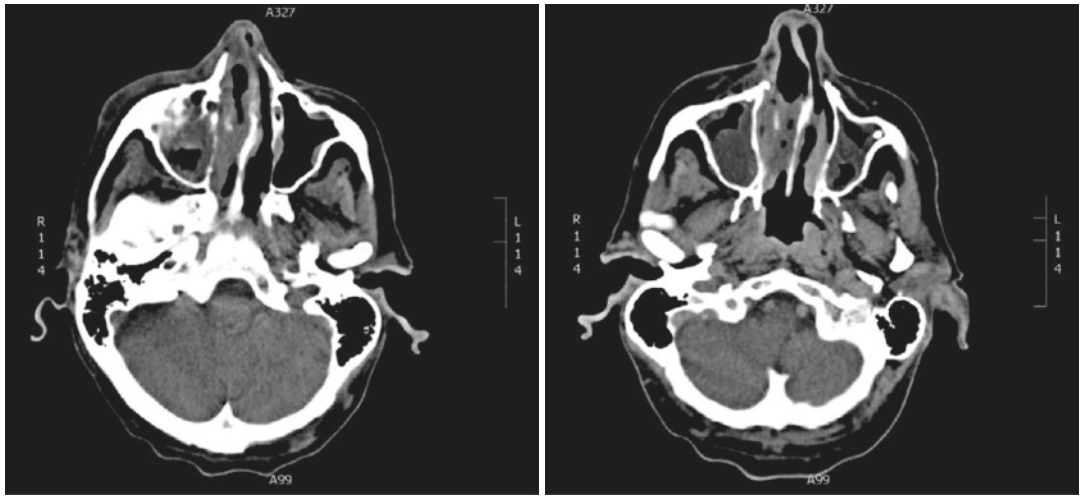

Fig. 5.1 CT scan of the paranasal sinuses

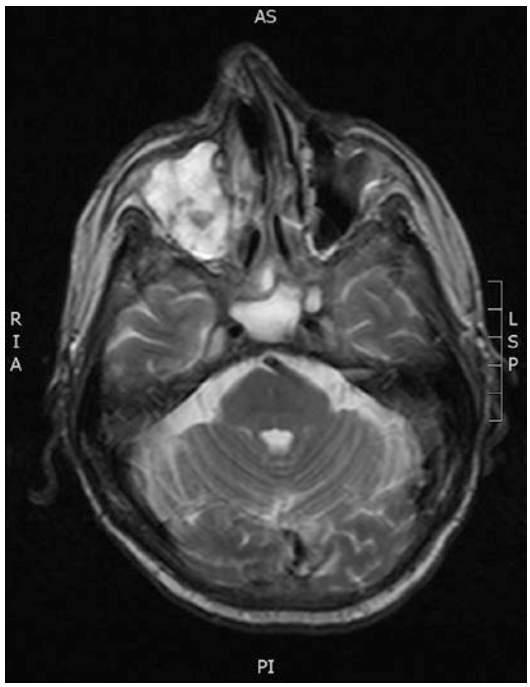

Fig. 5.2 T2-weighted MRI

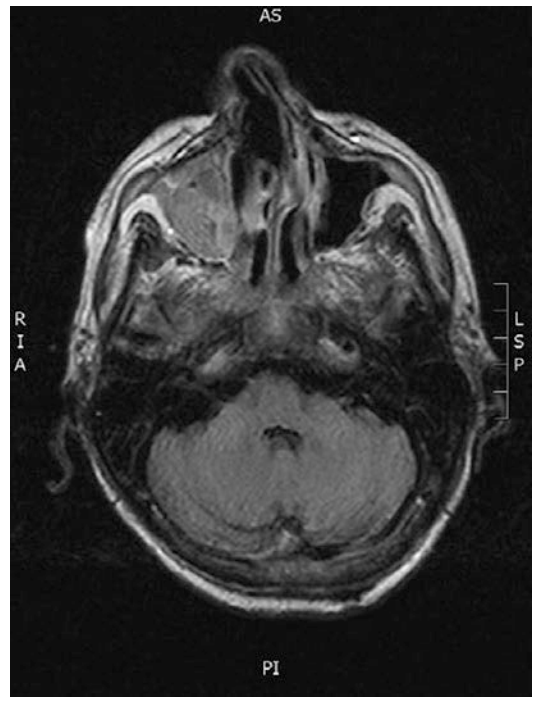

Fig. 5.3 T1-weighted MRI

5.3 Clinical Approach

5.3.1 Diagnosis

The clinician should be aware of the disease and its clinical manifestations in cases of immunosuppressed or immunocompromised patients, presenting with persistent high fever, severe headache and symptoms of rhinosinusitis. The clinical approach involves a thorough evaluation of the nasal cavities and paranasal sinuses, oral and pharyngeal mucosa, including endoscopy. Dark ulcers with eschar formation and swollen mucosa on the septum, turbinates and

palate, accompanied by nasal discharge and severe headache, are pathognomonic of the disease. Microbiology samples for bacteria and fungi should be taken. Tissue samples for culture and histopathologic examination should also be collected with great care because of the risk of bleeding, especially in patients with low platelet count. Cranial nerve function should be assessed because of the risk of intracranial invasion. Signs of ophthalmoplegia and/or exophthalmos with decreased pupillary responses indicate extension beyond paranasal sinuses to the orbit. Signs of cavernous sinus thrombosis are indicative of extensive disease. Urgent brain—sinus contrast CT scan should be performed. Bone erosions, soft tissue edema, mucosa thickening and vessel invasion may be noticed. MRI is more useful when intracranial, intraorbital or extension to the adjacent tissues is suspected.

5.3.2 Treatment

Once a rapidly evolving and possibly fatal disease, treatment should also be aggressive and effective. Urgent surgical resection with tissue debriment and complete disease removal, with simultaneous treatment of the underlying causes such as uncontrolled diabetes, neutropenia and immune system deficiency. In cases of early diagnosis with the disease isolated to the nasal cavity, without adjacent tissue invasions, surgical approach is restricted to extended endoscopic sinus surgery procedures (ethmoidectomy, medial maxillectomy, etc.). Sometimes extensive surgical procedures are required, such as transoral maxillectomy and orbital exenteration. Preoperative platelet transfusion should be considered in patients with low platelet count because of the risk of bleeding. High doses of Amphotericin B or Lipid Formulation of Amphotericin B should be administered intravenously as soon as the diagnosis of invasive fungal sinusitis is suspected.

5.3.3 Follow-up

As already mentioned, acute invasive fungal rhinosinusitis is related to high morbidity and mortality rates. Antifungal medication should be continued for an extended period after surgery. Those who remain disease-free require close monitoring due to the possibility of recurrence of the disease. Several specialists may be involved in the follow-up, such as ENT and Oral&Maxillofacial surgeons due to possible facial deformities after extensive surgeries, Hematologists in cases of hematologic malignancies, Clinical Oncologists, Immunologists, Endocrinologists in cases of diabetes etc.

Summary and Author's Comments

1. Acute Invasive Fungal Rhinosinusitis is a rare, extremely aggressive disease with high morbidity and mortality rates (50–80%).
2. Clinicians should be aware of the disease in cases of immunocompromised patients, presenting with persistent high fever, rhinosinusitis symptoms with nasal discharge, severe headache, black crust—eschar formation in the nasal cavity.
3. Intraorbital extension, opthalmoplegia, cavernous sinus thrombosis and cranial nerve involvement are signs of extensive disease.
4. Urgent wide surgical resection combined with high doses of antifungal medication and treatment of underlying conditions is the treatment of choice.

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6 Frontal Osteomyelitis-Pott's Puffy Tumour

Hisham S Khalil and Yasmine Nunwa

6.1 Case Presentation

A 36-year-old man (previously fit and healthy) presented to the Emergency Department with an 18-day history of progressively worsening pain around his forehead. This was associated with a centrally located and confined erythematous, tender forehead swelling, an intermittent fever and left- sided rhinorrhea.

6.2 Background Knowledge

Osteomyelitis of the frontal bone has become a rare occurrence since the introduction of antibiotics. The mortality rate associated with osteomyelitis of the frontal bone in the pre- antibiotic era was reported to be as high as 60%. The current mortality rate is less than 4%. The frontal bone, marrow cavity and sinus mucosa all share a venous drainage system, rich in diploic veins but devoid of valves. There are numerous diploic veins, some of which are present within the cancellous/spongy bone of the cranial bones. These are able to connect with veins and blood sinuses of the skull, thereby draining the frontal bone and the marrow cavity. The diploic veins can directly communicate with the intracranial venous sinus system. This viable, haematogenous route can allow the spread of infection within the marrow cavity of the frontal bone. The source of these infections usually originates within the frontal sinus and are subsequently carried to the frontal bone. Intracranial complications present in over 40% of patients, secondary to frontal sinus disease (Fig. 6.1).

Table 6.1 illustrates common pathogens, which have been linked to causing osteomyelitis of the frontal bone. The low oxygen concentration within the obstructed frontal sinus can cause the presence of these pathogens.

CT scans with contrast is the imaging modality of choice and is preferred to MRI scans because of its depiction of bony changes. A review of the literature suggests that a prolonged course of broad-spectrum antibiotics is warranted as an initial treatment. This should be initiated as intravenous therapy (up to 4 weeks) then continued orally. The overall antibiotic treatment should be for a minimum of 4 to 6 weeks with close monitoring of the patient.

Surgical intervention is warranted in patients who do not respond to initial treatment, patients who present with intracranial complications and

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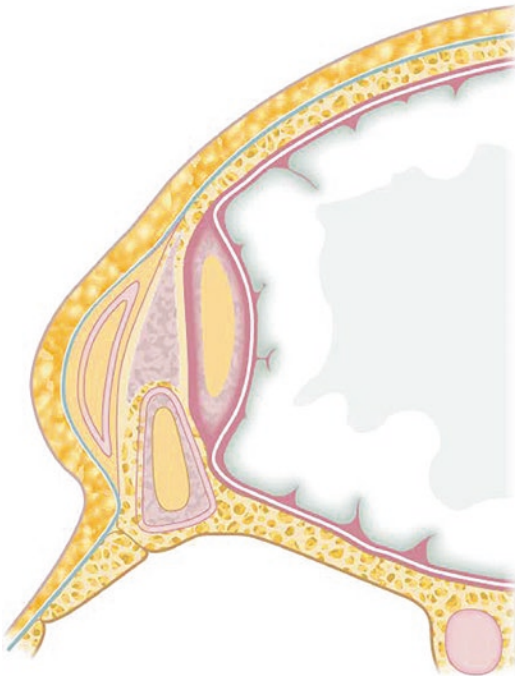

Fig. 6.1 Potential routes of spread. Intra-cranial complications present in over 40% of patients

Table 6.1 A list of the common microorganisms commonly isolated in Pott's Puffy Tumour

• Staphylococcus
• Streptococcus (alpha and beta haemolytic)
• Anaerobes
• Proteus Mirabilis
• fusobacterium
• Bacteroides
• Pseudomonas

considered in patients who have an erosion of the posterior table of the frontal sinus because of a higher incidence of intracranial complications. The traditional surgical approach has been an osteoplastic flap through a bicoronal incision. This approach is associated with multiple complications and morbidity. More recently, a mini-osteoplastic flap approach has been employed via a sub-brow incision and addressing the affected frontal sinus. A combined endoscopic and external frontal sinustomy is also a valid approach with removal of any bony sequestrum (Fig. 6.2).

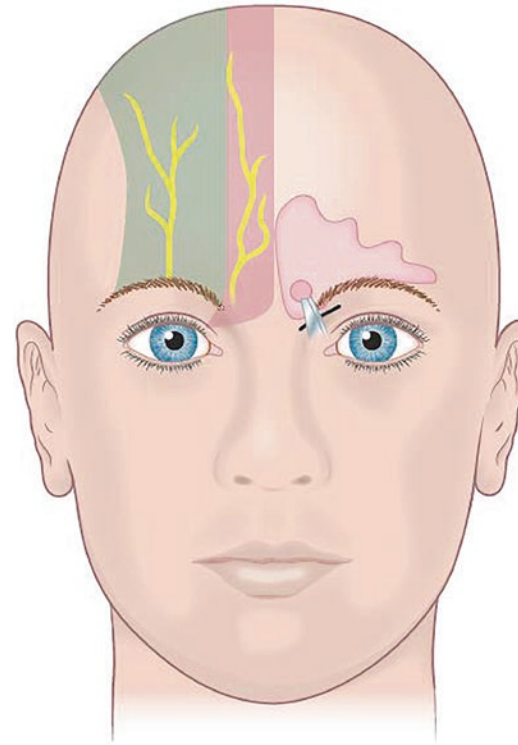

Fig. 6.2 Placement of the incision for external frontal sinusotomy, in order to avoid injury to supratrochlear and supraorbital nerves

6.3 Clinical Approach

6.3.1 History

The pain experienced was localised to the patient's forehead. The patient denied any neck stiffness, visual disturbances, photophobia, upper respiratory tract infections or recent history of preceding trauma to the head. The pain was exacerbated by bending forwards.

6.3.2 Examination

On examination, the patient looked well and was apyrexial. A 4 × 4 cm fluctuant tender, soft tissue swelling over the frontal bone region was present. In addition, the patient also had bilateral soft tissue swelling of both of his lower eyelids (Figs. 6.3 and 6.4). There was no evidence

Fig. 6.3 Sagittal view patient displaying fluctuant tender, soft tissue swelling over the frontal bone

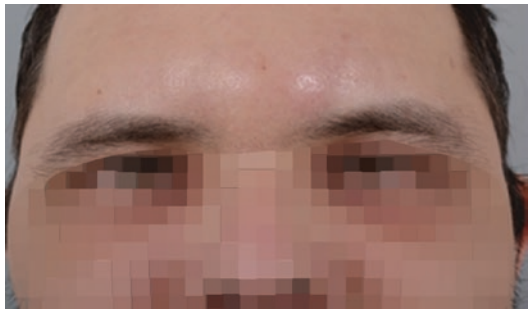

Fig. 6.4 Frontal view, 3 days of being in hospital. A notable amount of oedema is exhibited surrounding both eyes

of any focal neurological deficits or cervical lymphadenopathy.

A flexible nasal endoscopy revealed a left middle meatal discharge of which a swab was taken for microscopy, culture and sensitivity. There was no discharge present from both of his

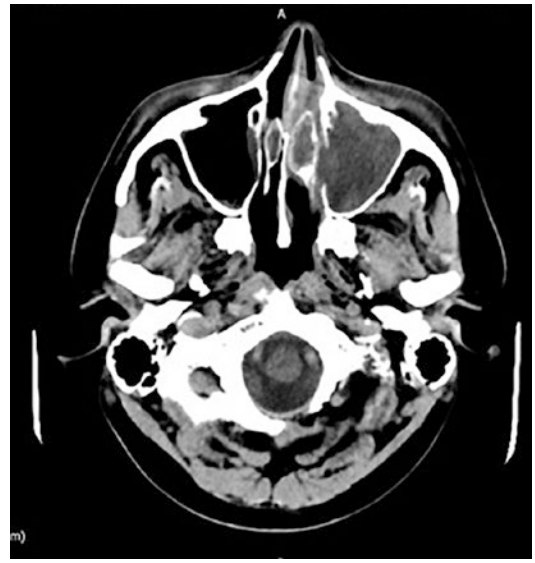

Fig. 6.5 Axial CT of the paranasal sinuses at the level of the maxillary sinuses demonstrating mucosal thickening in the left nasal cavity and bilateral conchcae bullosae

eyes and from the underling or surrounding skin. The patient gave a past history of recurrent sinusitis.

6.3.3 Investigations

A full blood count revealed a leucocytosis with neutrophilia. Computed Tomography (CT) axial scans of the patient's head and paranasal sinuses revealed a 4 × 2 × 2 cm soft tissue collection over the glabella. There was a breach of the anterior table of the left frontal sinus. The posterior table of the left frontal sinus was intact. There was no evidence of an intracranial extension or any other brain abnormality (Figs. 6.5 and 6.6).

Based on clinical and radiological findings, a provisional diagnosis of Pott's Puffy Tumour (PPT) arising from acute frontal sinusitis was made.

A nasal swab revealed: mixed growth of bacteria—Streptococcus anginosus and mixed anaerobes. Sensitivity revealed the organisms were susceptible to multiple antibiotics, in particular: penicillin, erythromycin/clarithromycin and tetracycline/doxycycline.

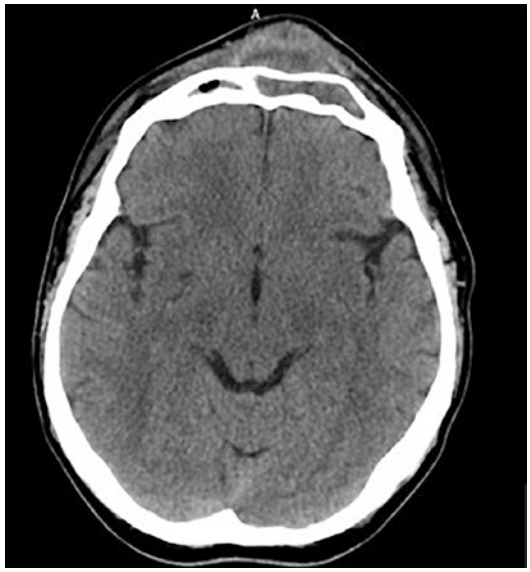

Fig. 6.6 Axial CT of the paranasal sinuses demonstrating opacification of the left frontal sinus and a left subcutaneous swelling

6.3.4 Treatment

High dose Intravenous, (IV) antibiotic therapy commenced following the patient's culture results. This included a 2-weeks course of IV co-amoxiclav, 1.2 g t.d.s followed by an additional 4-week course of oral co-amxiclav 625 mg tds. The patient was warned of the potential risk of cholestatic jaundice with a long course of co-amoxiclav. An oral course of metronidazole was also commenced for a total of 6 weeks. The patient was discharged after 2 weeks of hospitalization.

Summary and Author's Comments

1. Frontal osteomyelitis (Potts Puffy Tumour) should be treated as an ENT emergency.
2. The condition is more common in children and young adults and can be associated or result in intracranial complications.
3. The condition requires hospital admission, blood cultures if pyrexial and CT imaging of the paranasal sinuses and brain with contrast. A prolonged course of broad-spectrum antibiotics followed by oral antibiotic therapy is the recommended initial treatment.
4. Surgical intervention via a mini-osteoplastic flap or a combined external frontal sinustomy and endoscopic sinus surgery is resorted to in patients who fail to respond to medical treatment or who have associated complications.

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Septal Haematoma

7

Konstantinos Geronatsios

7.1 Case Presentation

A 35-year-old man presented to the ER Department because of nose pain following a facial trauma 2 days ago, during a sports activity. He also complained of worsening nasal congestion, without nasal discharge or epistaxis. There were no signs of external trauma, haematoma or ecchymosis. He was afebrile and vital signs were within normal limits. Using a nasal speculum, we noticed asymmetry of the septum, with reddish mucosal swelling on the left side. On palpation, the septum felt fluctuant and soft, and the whole procedure was painful for the patient. There were no signs of nasal or maxillary fractures. We also performed a nasal endoscopy with flexible nasendoscope, which was otherwise unremarkable. The diagnosis of septal haematoma, a potential serious complication of facial trauma was confirmed. Under local anesthesia, a mucosal and mucoperichondrial incision with a scalpel was performed. Blood clots were removed and a Penrose drain was placed. Bilateral nasal packing using Merocell was performed. Antibiotics were administered intravenously and the patient was discharged home 3 days later.

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7.2 Background Knowledge

7.2.1 Anatomy: Pathophysiology

The nasal septum is located in the midline and separates the two nasal cavities. It consists of the cartilaginous and bony septum, covered on both sides by a mucoperichondrial and a mucoperiosteal membrane. As the nasal septum itself is avascular, it depends on the overlying layers for its viability and for its nutrient supply. Nasal septal haematoma and nasal septal abscess are defined as a collection of blood or pus between the nasal septum and perichondrium or periosteum.

7.2.2 Etiology

Nasal trauma is very common because the nose is the most prominent part of the face. Septal haematoma is not as common as nasal bone fractures, but it is considered as a medical emergency since it is related to potentially serious complications. Haematoma can be unilateral or bilateral, involving both nostrils. Sports activities, accidents, falls, collisions with several objects are potential causes for septal haematoma. It can be found alone or combined with other injuries such as nasal bone or septal cartilage fractures. Iatrogenic septal haematoma is considered a complication of nasal surgery, such as septoplasty,

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rhinoplasty or endoscopic sinus surgery. Elderly patients receiving anticoagulant medication are prone to septal haematomas, even as a result of a low-energy trauma. Interestingly, haematomas are more prevalent in the paediatric population. This is due to the soft and immature septal cartilage, combined with a loosely adherent mucoperichondrium. Nasal septal abscess is mostly the result of bacterial colonization of a septal haematoma. It can be attributed to delayed identification of a septal haematoma. It is also considered to be a complication of septal surgery, nasal furuncle or spread from adjacent infections. Rare causes include ethmoid or frontal sinusitis, nasotracheal intubation, dentigerous cyst, dental curries and nasal insuflation.

7.3 Clinical Approach

7.3.1 Diagnosis

The clinician should be aware and suspicious of a possible nasal septal haematoma or abscess following facial trauma. It usually presents immediately or within the first 24 to 72 h after injury. Delayed septal haematoma is quite uncommon. Diagnosis is made by clinical examination and most of the time, anterior rhinoscopy using a nasal speculum or an otoscope is sufficient to demonstrate a septal heamatoma or abscess. It presents as swollen, boggy, erythematous septum, obstructing the nasal airway. In equivocal cases, it is useful to palpate the septum with our little finger or an instrument, for example a Jobson Horne probe. A haematoma or abscess feels soft, while a nasal septum deviation is firm in palpation. We also have to perform a thorough clinical examination, to exclude other injuries or fractures of the adjacent structures. Nasal endoscopy is also useful. Some patients may also present with persistent fever, facial pain, rhinorrhea, swelling, cellulitis and tenderness on palpation. Especially in children, clinicians should have in mind and investigate the possibility of non-accidental injury. A baby crying without an obvious reason, combined with difficulty breathing through the nose, should be examined for septal haematoma with anterior rhinoscopy.

Plain radiographs, CT or MRI scans are usually not necessary if other injuries or fractures are not suspected from the clinical examination.

7.3.2 Treatment

The mainstay treatment for septal haematoma and septal abscess is incision and drainage under local or general anaesthesia. Aspiration using a needle 18–20 Gauge and packing is an alternative for small collections. Our approach involves hemitransfixion incision, elevation of the mucoperichondrial–mucoperiosteal flap and evacuation of the blood clots/pus. We prefer leaving a small drain, such as Penrose or Yates drain, and placing bilateral nasal packs. Alternatives are quilting suturing or nasal (silicone) splints. Broad-spectrum antibiotics should be prescribed or administered intravenously in large haematomas or severe infections. Sometimes for the drainage general anesthesia is required, especially in children.

Untreated haematomas or abscesses can result in nasal deformity (saddle nose) secondary to septal necrosis from blood supply disruption or direct effects of the infection. Irreversible septal necrosis can occur within 72–96 h if the haematoma or the abscess are left untreated.

7.3.3 Follow-up

Patients recovering from a septal haematoma or abscess should be clinically evaluated regularly, to exclude the possibility of relapse and latent nasal deformity.

Summary and Author's Comments

1. Septal haematoma and abscess are urgent situations, and they should be suspected, in every case of facial/nasal trauma or after nose surgery.
2. Septal haematoma can be unilateral or bilateral. It usually presents immediately or within 72 h. Latent presentation is uncommon but not impossible.
3. Diagnosis is usually simple with anterior rhinoscopy, using a nasal speculum or an otoscope and palpating the septum.
4. Broad spectrum antibiotics should be prescribed or administered intravenously in all cases of septal haematoma/abscess. Treatment involves aspiration or drainage and nasal packing, sutures or nasal splints. Untreated haematomas can result in nose deformities (saddle nose).
5. In children, clinicians should have in mind and investigate the possibility of non-accidental injury.

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Nasal Foreign Bodies

8

Marios Stavrakas

8.1 Case Presentation

A 5-year-old boy was brought to the emergency department. His father gave a clear history of a nasal foreign body (plastic part of a toy) that was put while the patient was playing. Attention was paid to correct positioning and avoidance of unnecessary instrumentation which would agitate the child. There was a plastic foreign body anterior to the head of the middle turbinate. The ENT registrar removed it by passing a blunt wax hook past the foreign body and pulling it out. Minor bleeding was controlled with pressure and the patient was discharged on a short course of antibiotic nasal ointment.

8.2 Background Knowledge

Nasal foreign bodies are common among children, especially under 5 years old, or patients with mental health issues.

Some cases are witnessed, and the parents describe the nature of the foreign body. In other cases, the patient may remain asymptomatic or may present with a unilateral foul smell, nasal discharge (which may also be blood-stained), and vestibulitis. Although nasal foreign bodies may not be life-threatening, they may lead to complications from the foreign body itself or attempted removal.

Two broad categories of foreign bodies:

1. Inanimate: pebbles, beads, marbles, rubber, paper, batteries
2. Animate: myasis (fly maggot, screw warms, etc.)

Routes of foreign body entrance:

1. Through anterior nares (most common)
2. Through posterior choanae (food)
3. Through penetrating wound

Vegetable foreign bodies absorb water, swell up, and cause more discomfort. Also, they induce inflammation, and occasionally, the inflammatory reaction is sufficient to produce toxinaemia.

A foreign body could act as a nucleus if it firmly impacted or buried in granulation tissue. Eventually, it receives a coating of calcium, magnesium, phosphate, and carbonate, resulting in the formation of a rhinolith.

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8.3 Clinical Approach

8.3.1 Diagnosis

Usually, history is explanatory and adequate for diagnosis. Anterior rhinoscopy and, if tolerated, nasendoscopy can give information about the nature of the foreign body, the position and accessibility.

8.3.2 Treatment

Several methods have been described for the removal of foreign bodies. Sometimes, when the foreign body is impacted and the nasal mucosa appears oedematous, the topical application of a vasoconstricting agent may make the foreign body apparent.

- Advance a blunt wax hook beyond the foreign body and pull slowly.
- Use crocodile forceps or Hartmann nasal forceps to grasp a foreign body, if appropriate, depending on the size and shape.
- Ask the patient to occlude the unaffected nostril with their finger and blow forcibly through the affected nostril with the mouth closed.
- Advance a fine Foley (10F) or Fogarty catheter beyond the foreign body, inflate with 1–2 ml of water and withdraw slowly.

Appropriate positioning is essential. We prefer the child sitting in the parent's lap, with the legs secured between the parent's legs, one hand holding the child's arms and the other on the forehead, holding the head. This will ensure an optimal first attempt. Many attempts are not advised as they increase the patient's distress.

Common sites:

- Below the inferior turbinate
- Immediately anterior to the middle turbinate

When in doubt about the position and retrieval, the preferred method is nasendoscopy under local or general anaesthesia.

Right-sided foreign bodies are more common, as most right-handed patients tend to pick and put objects in their right nostril.

Complications: epistaxis, foul odour nasal discharge, nasal vestibulitis, mucosal irritation, local trauma due to repeated removal attempts, aspiration and need for further treatment.

Button batteries and magnets require special attention and early removal, as they may cause serious injuries to the nasal cavity. Necrosis of nasal mucosa and septal perforation are potential complications, which may require surgical debridement.

Common, non-erosive nasal foreign bodies should be usually removed within 24 h. Timing and planning (admission vs return for theatre) are regulated by local protocols. Antibiotics are not routinely required unless there are concerns about trauma of the nasal mucosa, tissue necrosis, infection or other complications.

Summary and Author's Comments

1. Detailed history is extremely important.
2. Appropriate positioning and minimal instrumentation is essential.
3. Button batteries and magnets require special attention and early removal.

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Part II

Rhinitis

Allergic Rhinitis

9

Marios Stavrakas and Hisham S Khalil

9.1 Case Presentation

A 34-year-old man was referred to the clinic with persistent rhinitis. He had marked nasal obstruction and rhinorrhea. In the past he underwent septoplasty without any significant benefit. He was keen to have revision septal surgery in the hope this would improve his symptoms. Careful history taking revealed he was on suboptimal medical treatment and there was poor compliance in taking and administering his medication.

A detailed clinical assessment revealed he was suffering from persistent allergic rhinitis and he was treated accordingly. The patient was given very clear instructions and with a request to keep a diary of his symptoms. On the next follow up appointment, he showed clinical and subjective improvement.

9.2 Background Knowledge

Rhinitis is a common phenomenon in the general population and one of the most frequent reasons for referral to the specialist ENT clinic. Chronic rhinitis is defined as symptomatic inflammation of the nasal mucosa, leading to obstruction, rhinorrhea, sneezing, or nasal/ocular itch and is estimated that it affects 30% of the general population. Rhinitis is classified as chronic if two nasal symptoms are present for at least 1 h daily for >12 weeks/year.

Chronic rhinitis includes:

- Infectious rhinitis
- Allergic rhinitis
- Nonallergic rhinitis

According to the ARIA classification (Allergic Rhinitis and its Impact on Asthma), allergic rhinitis can be Intermittent (symptoms < days/week OR <4 weeks) or Persistent (symptoms >4 days/week AND >4 weeks). The severity of symptoms and their impact on the patient's activities and quality of life may vary from mild to moderate-severe. When it comes to diagnosis, it is based on history and clinical examination, in combination with proof of IgE-mediated hypersensitivity. This can be achieved by using appropriate skin or blood tests that allow identification of the causative factor. When a diagnosis is established, it is essential to explore the patient's expectations and

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keep them realistic. The treatment strategy should be tailored to the patient's needs, and the clinician has to keep in mind patient's compliance, as there is evidence that many patients do not follow the prescriptions and advice as expected. We should not forget to monitor symptoms, and a good indicator of this is the visual analogue scale (VAS). In this direction, mobile applications that have been recently introduced seem to provide a reliable way of monitoring symptoms and treatment effectiveness.

When it comes to treatment, pharmacotherapy is the primary therapeutic strategy. If not successful, in cases that require long-term pharmacotherapy and in selected refractory cases, allergen-specific immunotherapy may be considered. Surgical management of a deviated septum or enlarged inferior turbinates is an option, again in carefully selected patients, where the nasal obstruction is a primary symptom and is not improved with medical treatment. Bousquet et al. (2020) have classified the treatment modalities for allergic rhinitis in five levels:

1. Non-sedating H_1-antihistamines
2. Intranasal corticosteroids
3. Intranasal corticosteroids + intranasal azelastine
4. Oral corticosteroid as a short course and an add-on treatment
5. Consider referral to a specialist and allergen immunotherapy.

As the treatment of allergic rhinitis is initiated and monitored to a high degree by general practitioners, it is important to assess the patient carefully, exclude other sinonasal pathology and consider referral to a specialist when this is necessary. The next generation ARIA-GRADE guidelines have reached some valuable conclusions which need to be taken into consideration when treating patients with allergic rhinitis. The combination of oral H_1-antihistamines with intranasal corticosteroids was not found to be more effective than intranasal corticosteroids alone, but the combination of intranasal H_1-antihistamines with intranasal corticosteroids was found to be more effective compared to intranasal corticosteroids alone. Finally, intranasal H_1-antihistamines are effective within minutes from the time of administration.

9.3 Clinical Approach

9.3.1 History

Detailed history taking is the basis and will give enough information for accurate diagnosis. It will also allow the clinician to characterise further the type of rhinitis and also get an idea about the patient's compliance to treatment.

9.3.2 Examination

Anterior rhinoscopy and flexible nasendoscopy aim to exclude any other causes for the patient's symptoms, such as CRS, septal deviation or other sinonasal pathology. Usually the mucosa appears swollen, pale or erythematous, there is clear rhinorrhea and the inferior turbinates appear enlarged.

9.3.3 Investigations

Skin Prick Test (SPT) or a blood test (RAST/total and specific IgE levels) are the first step to identify any allergen-specific sensitization (Fig. 9.1). In our practice, imaging is not necessary in the diagnosis of allergic rhinitis but there is sometimes an overlap between the symptoms of CRS and AR, especially in perennial sensitizations. In cases where other sinonasal pathology needs to be excluded, scanning may be an option.

9.3.4 Treatment

We adopt a stepwise approach based on medical treatment with intranasal steroids+/− antihistamines. In refractory cases, where the cardinal symptom is nasal obstruction secondary to turbinate hypertrophy, we may also consider turbinate reduction surgery such as submucosal or linear diathermy. It is important to highlight that surgical candidates must be carefully selected and the expected outcomes clearly explained.

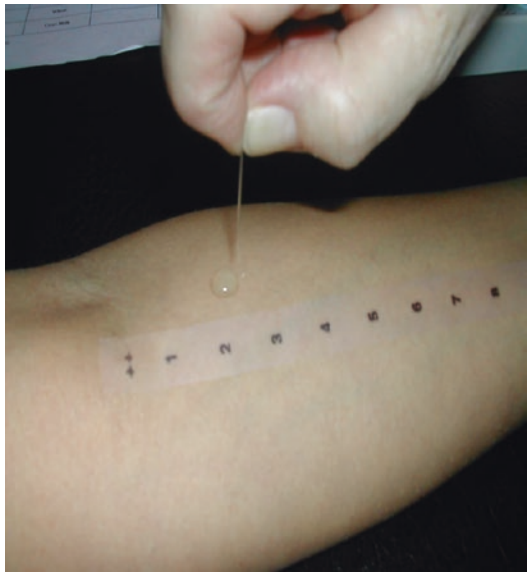

Fig. 9.1 Skin Prick Test is indicated if a type I allergy is suspected. It tests for air-borne allergens and usually takes about 15 min. A negative and a positive control are used

Summary and Author's Comments

1. Allergic rhinitis is a common clinical presentation, so General Practitioners and ENT specialists should be familiar with the current management guidelines.
2. Refractory cases may become problematic and referral to an allergy/immunology clinic should be considered.
3. Surgery is not advisable, unless there is obvious anatomical obstruction that can be corrected.
4. The primary aim of the consultation should be a detailed explanation of the condition and creation of realistic expectations. Moreover, it is useful to check whether the patient is compliant to the treatment before considering alternative management options.

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Nonallergic Rhinitis

10

Abdulaziz Abushaala, Marios Stavrakas, and Hisham S Khalil

10.1 Case Presentation

A 37-year-old man presented with a 2 year history of increasing bilateral nasal obstruction. He gave a history of using an 'Over the Counter' nasal decongestant over the same period. Over the last 6 months, he had noticed that his nasal decongestant had become less effective and he was having to use it more frequently. An ENT evaluation revealed a picture of rhinitis with a mild deviation of the nasal septum and significant hypertrophy of both inferior turbinates (Fig. 10.1). A skin prick test was negative for the tested allergens. A working diagnosis of a rhinitis medicamentosa was made. The patient was treated successfully with a gradual reduction of the nasal decongestant use with introduction of nasal steroids and bilateral diathermy of inferior turbinates.

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10.2 Background Knowledge

Nonallergic rhinitis (NAR) is a type of chronic rhinitis which does not have any signs of chronic infection or systemic signs of inflammatory allergy and affects more than 200 million in the general population. The patients who are classified in this category complain about rhinorrhea or nasal congestion, but they do not have any discoloured secretions and also, they have negative skin prick test and RAST to specific allergens.

The clinician, based on the history and the exclusion of sensitisation, should differentiate nonallergic rhinitis from other types of rhinitis (infectious, allergic, local allergic), rhinosinusitis and any other sinonasal pathology. Regarding local allergic rhinitis, it is characterised by a local allergic response of the nasal mucosa but the patient has negative skin prick test and specific IgE. Practically though, differentiating this clinical entity from nonallergic rhinitis is difficult. Most patients respond well to intranasal corticosteroids.

Table 10.1 summarises the basic characteristics of all the subtypes of nonallergic rhinitis.

Rhinitis medicamentosa is a nonallergic form of rhinitis that is associated with prolonged use of topical nasal decongestants. Topical nasal vasoconstrictive medications, such as sympathomimetic amines and imidazoline derivatives, are thought to be the main

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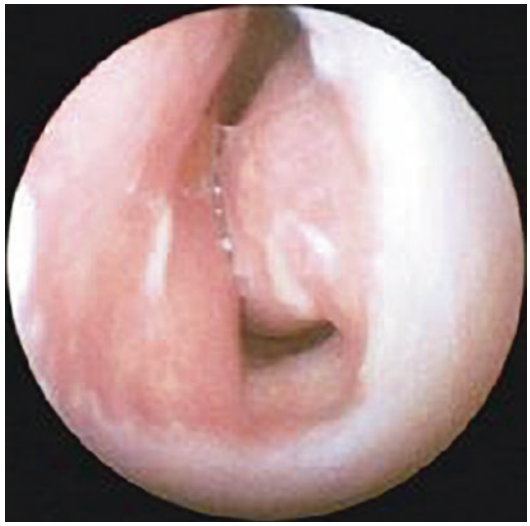

Fig. 10.1 Endoscopic view of the left nasal cavity in a patient with Rhinitis medicamentosa with hypertrophy of the inferior turbinate and a mild deviation of the nasal septum

causative agents of Rhinitis Medicamentosa. They are α-Adrenoceptor agonists. Their prolonged use results in reduction in efficacy (tachyphylaxis) and a rebound increase in nasal airway congestion and nonspecific nasal hyperreactivity. Despite its high prevalence, the pathophysiology of Rhinitis Medicamentosa remains unclear. Vasodilatation and intravascular oedema have both been implicated. Rhinitis Medicamentsa diagnosis relies mainly on the history of nasal congestion/blockage without rhinorrhea associated with prolonged topical nasal decongestant abuse. It can occur with other nasal conditions like allergic rhinitis and nasal polyps. Nasendoscopy reveals inflamed nasal mucosae, with areas of punctate bleeding. A recent systematic review by Zucker et al. (2019) suggests that the literature currently lacks compelling evidence to formally con-

Table 10.1 Basic characteristics of nonallergic rhinitis

Subtype of NAR	Characteristics	Pathophysiology	Treatment
Senile rhinitis	In patients > 65 yo. Appears as late-onset, bilateral watery nasal secretions, without mucosal or anatomical pathology	Neurogenic dysregulation, mucosal/glandular atrophy	Ipratropium bromide
Gustatory rhinitis	Watery rhinorrhea after eating hot or spicy food	Provoked by a gustatory reflex associated with a hyperactive, nonadrenergic, noncholinergic or peptidergic neural system	Avoidance, nasal capsaicin
Occupational rhinitis	Inflammation of the nasal mucosa due to exposure to a particular work environment	Neurogenic inflammation	Avoidance. Important to diagnose in order to prevent occupational asthma
Hormonal rhinitis	Associated with hormonal imbalances during menstrual cycle, puberty, pregnancy, menopause, hypothyroidism, acromegaly	Estrogens → vascular engorgement → obstruction ± hypersecretion. Beta-estradiol and progesterone → increase of histamine H1 receptor expression → eosinophil migration and/or degranulation. Testosterone has the opposite effect	Intranasal corticosteroids
Drug-induced rhinitis	1. Adverse effect of medications 2. Rhinitis medicamentosa	1. Aspirin, ibuprofen, NSAIDs, beta-blockers, sedatives, anti-depressants, oral contraceptives, drugs to treat erectile dysfunction 2. Prolonged use of decongestant sprays	Avoidance
Idiopathic rhinitis	Up to 50% of NAR do not have a clear aetiology	Neurogenic inflammation, unknown aetiology	Intranasal corticosteroids, capsaicin

struct a standardised treatment plan for RM. However, its management relies mainly on the cessation of topical decongestants, commencement of intranasal steroids and nasal saline.

Capsaicin's action is based on an initial neuronal excitation, which is followed by a prolonged period, when the previously excited neurons are not responsive to a wide range of stimuli. Its use should be carefully monitored, and depending on the institution, the desirable concentration should be produced by the hospital pharmacy. According to Fokkens et al. (2016) capsaicin is not effective in senile rhinitis or in NAR caused by smoking, while there is no evidence that capsaicin can help in allergic rhinitis. Thus, it has to be applied in carefully selected cases. Those patients who benefit from capsaicin treatment, especially those with idiopathic rhinitis, may seek re-application after a few months, when the symptoms of rhinitis re-appear.

10.3 Clinical Approach

10.3.1 History

A history of bilateral nasal obstruction and the misuse of topical and less commonly systemic decongestants should raise suspicion of a diagnosis of rhinitis medicamentosa. It is important to explore symptoms associated with other sino-nasal disorders such as allergic rhinitis with frequent sneezing, watery rhinorrhea and a history of atopy. The history taking should also exclude occupational rhinitis and rhinosinusitis.

The Otolaryngologist should spend time identifying the type of nasal or systemic decongestant, the dose and frequency of use and whether any other topical or systemic medications are being used to treat the nasal obstruction. A past medical history of hypertension should be explored as this could be a side effect of the prolonged use of decongestants.

10.3.2 Examination

Typically, patients with Rhinitis Medicamentosa have an erythematous and swollen nasal mucosa with hypertrophy of the inferior turbinates. There may be evidence of other causes of nasal obstruction such as alar collapse, a deviated nasal septum or nasal polyposis.

10.3.3 Investigations

Investigations are requested according to the patient's symptoms. It is important to exclude allergic rhinitis through a skin prick test/total and specific IgE levels. A CT scan of the paranasal sinuses should be requested in patients with a history suggestive of rhinosinusitis and where medical treatment has failed.

10.3.4 Treatment

A detailed discussion with the patient is important to highlight the need to stop the topical or systemic nasal decongestant. It is also important to highlight a working schedule for the gradual reduction of the dose and frequency of topical nasal decongestant. It is best to start with omitting the decongestants during the day whilst continuing the use at bedtime and first thing in the morning, then gradually weaning off the patient by stopping the bedtime doses.

Weaning patients off the decongestant is often helped by the use of a hypertonic Saline nasal douche and a topical steroid spray. Patients are given approximately 12 weeks to see if this approach is effective. If the approach fails or the patient symptoms are very severe at the onset, then it is appropriate to proceed with inferior turbinate reduction surgery. This will depend on the preference of the surgeon. In our institution we use a combination of out fracture together with linear diathermy of the surface of the inferior turbinates (Fig. 10.2). Other approaches include inferior turbinoplasty with or without the use of Coblation or a Microdebrider.

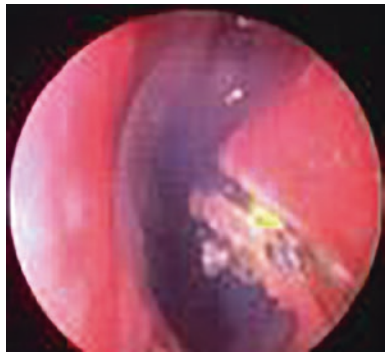

Fig. 10.2 An endoscopic view of the left nasal cavity demonstration linear diathermy of the hypertrophied inferior turbinate

Summary and Author's Comments

1. Nonallergic rhinitis is a chronic condition characterized by the absence of allergic sensitization.
2. The clinician should be familiar with the subtypes and the groups of population they affect.
3. As in allergic rhinitis, surgical treatment in the form of turbinate reduction or septoplasty, need careful consideration and the clinician must emphasize on the precise indication and the expected outcomes after surgery.

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Part III

Rhinosinusitis

Acute Rhinosinusitis

11

Marios Stavrakas

11.1 Case Presentation

A 71-year-old diabetic patient was admitted urgently with right acute rhinosinusitis and swollen upper eyelid. He had a 10-day history of coryzal symptoms, nasal discharge, facial pain and nasal obstruction which was initially treated conservatively. As his symptoms started deteriorating, his upper eyelid started swelling up. A CT scan showed opacification of the right maxillary sinus, ethmoids, frontal sinus and erosion of the posterior table. He was treated with intravenous antibiotics and topical nasal decongestants. After being reviewed by the ophthalmologists, a decision was made for surgical drainage. He underwent endoscopic middle meatal antrostomy, ethmoidectomy and opening of frontal recess. He had an uneventful recovery and after 6 weeks he had definitive treatment with revision ESS and combined approach frontal sinus surgery.

11.2 Background Knowledge

ARS is a common pathology, affecting 6–15% of the global population. It is one of the most common diagnoses in primary care and it is estimated that roughly 90% of adults have had sinusitis at some point of their life. Of all ARS cases, fewer than 2% are caused by bacterial infection and recurrent acute rhinosinusitis (RARS) affects 1 in 3000 western adults. ARS is diagnosed clinically, by the acute onset of nasal blockage, nasal discharge, facial pain and reduction of smell. According to EPOS 2020, acute bacterial rhinosinusitis must demonstrate at least three of the following five signs and symptoms: discoloured mucous, severe local pain (often unilateral), fever >38 °C, raised CRP/ESR, 'double sickening'.

Regarding predisposing factors, the clinician should take into consideration anatomical abnormalities or variations that obstruct the natural drainage pathways. Examples are infraorbital ethmoid cells, small infundibular width, pneumatization of the middle turbinate or accessory ostia. It has been suggested that anatomical variation is more likely to be of causal significance in patients with RARS. Dysfunction of the mucociliary clearance mechanism is another contributing factor. This has not only to do with congenital or syndromic conditions but is also the consequence of a viral cold, with the latter leading to progressive loss of ciliated cells, dysmorphic cilia and alterations in the mucous layer. Another predisposing factor is biofilm formation. Biofilms represent complex aggregates of extracellular matrix and independent microorganisms from multiple species, resulting in increased antibiotic resistance (up to 1000 times) when compared to free living bacteria of the same species.

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There are three key elements in the pathophysiology of ARS: narrow sinus ostia, dysfunction of the ciliary apparatus and viscous sinus secretions.

Odontogenic sinusitis is a variant with discrete pathogenesis. It has to do with the close anatomical relationship of the molars and premolars from the maxillary sinus (distance 1.97 mm). Various factors are identified in the pathogenesis of odontogenic sinusitis, such as the maxillary spread of an endodontic infection, periapical cysts, granulomas or abscess eroding the surrounding mucoperiosteum, the intrasinus displacement of endodontic materials, tooth fragments, implant or augmentation grafts, perforation of the Schneiderian membrane during tooth extractions, periodontal surgery or sinus floor elevation surgery, dental traumas or an oroantral fistula. Periapical infection spreads through the bone marrow following the path of blood vessels and the lymphatic system. Rapidly spreading acute infectious pulpal disease is much more destructive and involves the adjacent sinus within a short time. Also, we can recognise an acute and invasive phase, characterised by an initial elevation of the innate immune system (neutrophils and macrophages), and a chronic phase, in which the lesion is much more organised and characterised by an adaptive immune response.

11.3 Clinical Approach

11.3.1 Diagnosis

It is known that the 'gold standard' diagnostic test of true bacterial ARS is a positive culture from an invasive sinus puncture or meatal swab (endoscopically guided), while radiological assessment of the sinuses has less diagnostic value. Neither bacteriology nor radiology is recommended in making the clinical diagnosis of bacterial ARS or guiding management. Individual symptoms are poorly predictive, but there is limited evidence to suggest that combinations of clinical factors (while not diagnostic of bacterial infection) may alert clinicians to patients with more severe and prolonged illness- for example, lasting beyond 10 days or worsening after 5–7 days—who should be monitored and considered for more invasive treatment including antibiotics. Gluck et al. (2018) compared the diagnostic criteria for ARS in different healthcare systems. They concluded that purulent nasal discharge presenting either as an anterior or a posterior nasal drip associated with nasal obstruction were the two most common symptoms mentioned for diagnosis in both developed and developing countries. Both the US and the EPOS guidelines differentiated common cold/acute viral rhinosinusitis and postviral sinusitis from each other: acute viral rhinosinusitis is an inflammation of the nose and the paranasal sinuses caused by viral infection, which generally resolves within 10 days, whereas postviral rhinosinusitis is defined as the deterioration of the former after 5 days or extension of the disease after 10 days duration. All developed countries disregard plain X-rays as a diagnostic tool and advocate the use of CT sinuses when necessary.

11.3.2 Treatment

The management of ARS can be either watchful waiting with symptomatic relief, medical treatment mainly including oral antibiotics and finally surgery (endoscopic, open, balloon sinuplasty). The choice of treatment should be made on the grounds of evidence-based medicine, following national or international guidelines.

11.3.2.1 Medical Treatment

Fokkens et al. (2014) advocate a watchful waiting approach for uncomplicated ARS in adults, with the exception of patients with high fever, severe unilateral facial pain, purulent rhinorrhea and 'double-sickening'. They also suggest symptomatic control with non-antibiotic preparations and patient education.

Systemic reviews support the watchful waiting approach, as antibiotics do not prove to have any significant benefits over conservative treatment. Also, antibiotics may bring about adverse effects, such as gastrointestinal disorders and rash. Based on the lack of clear benefits and the

pressing global problem of antibiotic resistance, Fokkens et al. (2014) state: 'there is no place for antibiotics for the patient with clinically diagnosed, uncomplicated acute rhinosinusitis'. Another Cochrane review on antibiotics versus placebo for acute maxillary sinusitis with symptoms lasting at least 7 days found six controlled trials. There was a modest symptom resolution benefit with antibiotics, but improvement was high in both the placebo (80%) and the antibiotic treated group (90%). There was only a marginal difference in 'total care' rates between groups, with antibiotics resulting in a small reduction in relative risk of ongoing symptoms at 7–15 days. The authors conclude that the modest benefits must be weighed against the potential for adverse effects at both individual and population levels.

The decision for antibiotic administration is dictated by North American, European and UK guidelines. More specifically, the American Academy of Paediatrics has introduced a severity tool to predict acute bacterial rhinosinusitis and if the criteria are met, antibiotics should be prescribed. The Infectious Diseases Society of America suggest prescribing antibiotics based on a severity prediction tool (persistence, not improving, severe symptoms, double sickening). The vast majority of sinusitis cases are caused by viruses and will therefore resolve without antibiotics. Antibiotics are recommended only if symptoms do not resolve within 10 days. Antibiotics are specifically not recommended in those with mild/moderate disease during the first week of infection due to risk of adverse effects, antibiotic resistance or rash. Because of increasing resistance to amoxicillin, the 2012 guidance of the Infectious Diseases Society of America (IDSA) recommends amoxicillin-clavulanate as the initial treatment of choice for bacterial sinusitis. The guidelines also recommend against other commonly used antibiotics, including azithromycin, clarithromycin and trimethoprim/sulfamethoxazole, because of growing drug resistance. A short course (3–7 days) of antibiotics seems to be just as effective as the typical longer course (10–14 days) of antibiotics for those with clinically diagnosed bacterial sinusitis without other severe disease or complicating factors. IDSA guideline suggests 5–7 days of antibiotics is long enough to treat a bacterial infection without encouraging resistance.

Non-antibiotic options include information on disease course, reassurance and symptomatic treatment. Although widely used, there is no convincing evidence of clinically relevant benefits from antihistamines, steam inhalation, decongestants or saline irrigation. Topical nasal steroids have been found to have a modest effect on symptoms and speed of recovery. A Cochrane review found that symptoms of participants receiving this treatment were more likely to resolve at 2 weeks compared with those receiving placebo (73 vs. 66.4%; risk ratio 1.11, 1.04–1.18). Although this review reported no significant adverse events, possible adverse effects can include nasal irritation and epistaxis. Decongestant nasal sprays containing for example oxymetazoline may provide relief, but these medications should not be used for more than the recommended period. Longer use may cause rebound rhinitis.

11.3.2.2 Surgical Management

There is no place for surgery for the uncomplicated ARS. In cases of ARS complications, such as orbital cellulitis, when conservative treatment has failed, open or endoscopic procedures are advocated aiming to drain the abscess cavity and eradicate the disease. A study by Dewan et al. (2011) found that surgical technique selection influences duration of hospital stay. Those patients with combined endoscopic sinus surgery and subperiosteal abscess (SPA) drainage had significantly shorter length of stay, likely related to fewer abscess reaccumulation rates.

Regarding the timing of the intervention, several factors should be taken into consideration. Wan et al. (2016) studied the treatment of orbital complications following ARS in children. Endoscopic sinus surgery was performed when an improvement in the patient's condition did not occur after 48 h. But patients with orbital abscess, mobility disorders of the eyeball or decreased vision received ESS immediately within the first 24 h. The abscess is usually between the lamina papyracea and the periorbita, occasionally on the superior orbital wall. Proptosis, chemosis and

limited movement of the eyeball are common clinical manifestations. Pond and Berkowitz (1999) suggested that complete ethmoidectomy is not necessary for SPA since ethmoid disease can be cured by antibiotic therapy, but anterior ethmoidectomy is necessary to allow intranasal drainage of the SPA. Wan et al. (2016) support that complete opening of ethmoid cells is necessary for the drainage of the SPA in patients.

In the case of recurrent acute rhinosinusitis (RARS) current practice suggests limiting surgery to maxillary antrostomy and bilateral anterior ethmoidectomy. With a CT scan showing more extensive sinus disease, more surgery is recommended to address the specific areas of disease indicated on the scan.

The contribution of balloon sinuplasty in the treatment of ARS has also been studied. In the particular subgroups of critically ill and immunocompromised patients who are vulnerable to severe complications, balloon sinuplasty is a useful tool. It relies on sinus dilatation without mucosal cutting and therefore has the potential to minimise blood loss. Notably, however this procedure is unsuitable for patients suspected of invasive fungal sinusitis because a tissue biopsy is required for diagnosis.

11.3.3 Follow-Up

Cases of uncomplicated acute rhinosinusitis do not require follow up. Complicated cases and those who are predisposed for CRS may need to be reviewed. Odontogenic rhinosinusitis should be referred to a dentist or OMFS for definitive treatment of the dental source of infection.

Summary and Author's Comments

1. ARS is a common pathology, concerning both GPs and ENT specialists.
2. Diagnosis is essentially clinical, based on current guidelines.
3. Medical management is the mainstay treatment, with surgical treatment reserved for complicated cases.

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Chronic Rhinosinusitis

12

Abdulaziz Abushaala and Hisham S Khalil

12.1 Case Presentation

A 46-year old woman presented with a 18 month history of purulent nasal discharge, post nasal drip, nasal obstruction, reduced smell and intermittent facial discomfort. She had a 4 week course of Doxycycline 100 mg once daily, 12 weeks of a saline nasal douche and a steroid spray with only partial improvement of her symptoms. Skin prick tests were negative for the tested allergens. A CT scan of the sinuses was suggestive of Primary Chronic Rhinosinusitis (CRS) of the diffuse type. The patient underwent targeted 'limited' Functional Endoscopic Sinus Surgery and made a good recovery.

12.2 Background Knowledge

Chronic Rhinosinusitis (CRS) is defined as the infection or inflammation of the nose and paranasal sinuses with symptoms present for more than 12 weeks. It is a common health issue that affects 5–12% of the population. Chronic Rhinosinusitis presents in both adults and children. It has a significant impact on the quality of life as well as the requirement for several visits to the primary and secondary health care services.

The aetiology of CRS is multifactorial. Multiple environmental factors (infective, allergic and pollution) and host factors (genetics, immune, epithelial barriers and anatomical) have been implicated. The interaction of the above factors contributes to the pathophysiological complex processes (Endotypes) that lead to chronic inflammation of the nasal and sinuses mucosa which in turn lead to different clinical presentations (phenotypes).

Clinically, CRS, as recommended by EPOS 2020, in adults is defined as: inflammation of the nose and the paranasal sinuses characterised by two or more symptoms, one of which should be either nasal blockage/obstruction/congestion or nasal discharge (anterior/posterior nasal drip):

- ± facial pain/pressure
- ± reduction or loss of smell for; ≥12 weeks
- and either
  - endoscopic signs of nasal polyps, and/or mucopurulent discharge primarily from middle meatus, and/or oedema/mucosal obstruction primarily in middle meatus
  - and/or CT changes: mucosal changes within the ostiomeatal complex and/or sinuses.

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12.2.1 The Classification of Chronic Rhinosinusitis

The classification of CRS with nasal polyps (CRSwNP) and CRS without nasal polyps (CRSsNP) has been widely used and it is essentially based on the disease clinical findings. A new classification using anatomic distribution and endotype dominance has been chosen by the EPOS 2020 steering group which divides CRS into primary and secondary groups and subdivides each group into localised (allergic fungal rhinosinusitis or an isolated sinusitis) and diffuse disease (eCRS and non-eCRS).

CRS is classified as 'Primary' if no obvious sinonasal secondary pathology (i.e. Tumour, Fungal infection, odontogenic or immunodeficiency disorder). It is further divided into type 2 or non-type 2 endotype dominance.

For 'Secondary' CRS, the division is into localized or diffuse, the latter is further sub-divided depending on local pathology into mechanical, inflammatory and immunological factors.

12.2.2 Management of Chronic Rhinosinusitis

12.2.2.1 Medical Treatment

The aims of medical therapy for CRS is to reduce mucosal inflammation, improve sinus drainage, and eliminate bacterial infections. Based on the recommendation of EPOS 2020, the management of diffuse CRS include:

Appropriate medical therapy (AMT) for 6–12 weeks.

- Nasal steroid (drops/spray/rinses)
- Saline rinses
- Consider oral corticosteroids

If there is no improvement; treatment should be tailored according to clinical finding;

1. For Non type 2: AMT (± long term antibiotics) or Functional Endoscopic Sinus Surgery (FESS)
2. For Type 2: AMT (± Oral corticosteroids) or FESS

Other medications that can be considered include Anti-leukotrienes and biologicals (monoclonal antibodies). We have also found the use of nasal douching with diluted baby Shampoo useful in a selected group of patients with persistent purulent nasal discharge despite optimal medical therapy and with Staphylococcus Aureus isolated from middle meatal swabs. The management of secondary CRS will depend on the causative disease.

12.2.2.2 Surgical Treatment

FESS is indicated when maximum medical therapy fails to improve patients' symptoms. The aim of surgery is to remove polypoidal mucosa, drain and ventilate the paranasal sinuses. The extent of FESS will depend on the extent of the disease process. The surgical procedure is called "complete"/"full house" FESS if it involves bilateral maxillary antrostomies, bilateral total ethmoidectomies, bilateral sphenoidotomies, bilateral frontal sinusotomies (Draf IIa, IIb, or III). The term "targeted" surgery is used if limited FESS is performed. Complete surgery was an independent predictor of postoperative improvement of the SNOT-22 score in one study.

External approaches and combined endoscopic and external approaches are still be indicated in selected patients e.g., Frontal sinusitis or mucoceles with Type IV frontal cells.

12.3 Clinical Approach

12.3.1 History

The patient had a classic history of chronic rhinosinusitis. There was no history suggestive of atopy. It is important to ascertain that patients are compliant with the use of the medical treatment with appropriate application of the nasal medication.

12.3.2 Examination

Patients should have a complete ENT examination including a nasal endoscopy. The patient had mucopus in the left middle meatus (Fig. 12.1) and oedematous mucosa in the right middle meatus.

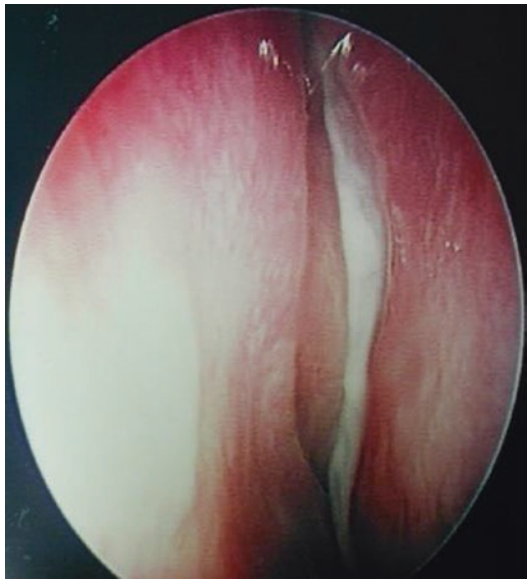

Fig. 12.1 Mucopus in the left middle meatus

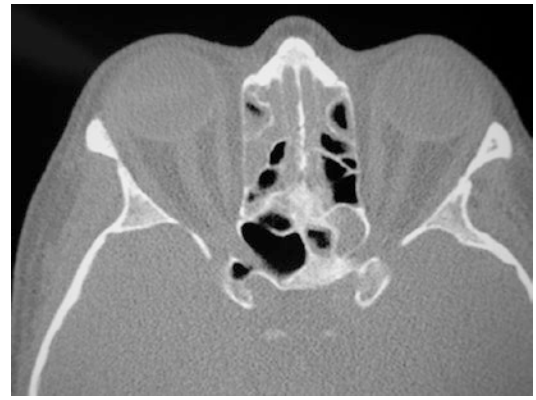

Fig. 12.2 Axial CT scan of the paranasal sinuses demonstrating mucosal thickening of the ethmoid sinuses and a dehiscent right optic nerve in an Onodi (orbito-ethmoidal) air cell

12.3.3 Investigations

Skin Prick Tests were negative for the tested allergens. CT scans should be examined carefully in the axial, coronal and sagittal planes and the imaging department alerted to acquire the images with settings suitable for computer-assisted image guidance. In addition to assessing mucosal disease and the ventilation of the sinuses, anatomical variants should also be identified. The patient's scan revealed diffuse mucosal thickening in the paranasal sinuses with bilateral disease. The axial scan revealed a dehiscent right optic nerve in a right Onodi (orbito-sphenoidal) cell (Fig. 12.2). It is crucial to identify this variant as missing it puts the optic nerve at risk during surgery. Other variants that need assessment include the depth of the cribriform plate, the presence of orbito-maxillary (Haller) cells, a Concha Bullosa (excessive pneumatisation of the middle turbinates) and paradoxically bent middle turbinates. The presence of a sphenoid inter-sinus septum that is in close relationship to the internal carotid artery(ICA) or a dehiscence of the bony canal of the ICA are also relevant when operating on the sphenoid sinus.

Fig. 12.3 Computer-assisted image guidance system

12.3.4 Treatment

A limited (targeted) bilateral FESS was carried out with avoidance of the right posterior ethmoid and right sphenoid sinuses due to the presence of a dehiscent right optic nerve. A computer-assisted image guidance system was used (Fig. 12.3). Both middle meati were packed with absorbable nasal packs.

Postoperative care includes a 2 week course of co-amoxiclav, saline nasal douching and commencing a steroid nasal spray 2 weeks after the surgery when the nasal packs will have dissolved.

Patients are seen once in the ENT outpatients approximately 6 weeks after surgery.

Summary and Author's Comments

The management of primary diffuse CRS is primarily medical. FESS is reserved for patients who have failed optimal medical treatment. It is important to ensure patient compliance with optimal medical therapy before it is deemed to have failed. The extent of surgery 'complete versus targeted' depends on the extent of the disease and the philosophy of the surgeon.

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Chronic Rhinosinusitis with Polyps (CRSwNP)-Medical Management

13

Georgios Fyrmpas

A 54-year-old male presented with recurrent bilateral nasal polyps 7 years post FESS. His main complaint was a blocked nose and a reduced sense of smell. There was no history of headaches, asthma, or ASA intolerance. Endoscopic examination revealed grade 3 polyps on the right side (Fig. 13.1a) and grade 2 polyps on the left side. He had an unsuccessful 3-month trial with intranasal fluticasone spray. Three weeks of fluticasone nasules by his GP made a marginal improvement. A CT scan was not obtained due to the patient declining any further surgery. He was then prescribed budesonide irrigations, and clear instructions on the correct application of the solution were given. Unfortunately, he could not adhere to twice-daily use of the steroid solution, and therefore he was offered a 2 week trial with oral methylprednisolone (24 mg OD in the morning) as a last resort. He reported significant improvement in nasal breathing and sense of smell without any adverse effects on follow up. Repeat endoscopy revealed a substantial reduction in polyp size. He agreed to continue with the budesonide irrigations OD for another 3 months and then mometasone steroid spray BD for maintenance. Nasal symptoms were well controlled at the 6-month follow up (Fig. 13.1b).

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13.1 Background Knowledge

Traditionally, patients with chronic rhinosinusitis (CRS) are divided into two groups: with polyps (CRSwNP) and without polyps (CRSsNP). According to EPOS 2020 guidelines, there is further characterization of CRS to primary and secondary, based on the aetiopathogenesis of the chronic inflammation. In addition to this, CRS can also be sub-classified in diffuse or localized, and also the clinician should take into consideration the various endotypes and phenotypes that may be involved.

The core concept in the pathophysiology of CRSwNP is the faulty interaction between the host immune system and several exogenous factors (bacteria, viruses, fungi, toxins), which initiates and maintains an inflammatory mucosal state. The exact interplay between these factors is not well understood, but several phenomena have been implicated: (a) an epithelial integrity breakdown that permits recognition of pathogens from the active immune cells of the submucosal space (b) stimulation of epithelial cells to promote inflammation (c) reduced mucociliary clearance that allows bacterial colonisation (d) the presence of S. Aureus in the interstitial space and intracellularly along with its enterotoxin e) the loss of diversity of bacterial communities in the nasal mucosa with reduction of commensal and dominance of pathogenic taxa (f) the presence of biofilms and (g) increased numbers of fungi. Based

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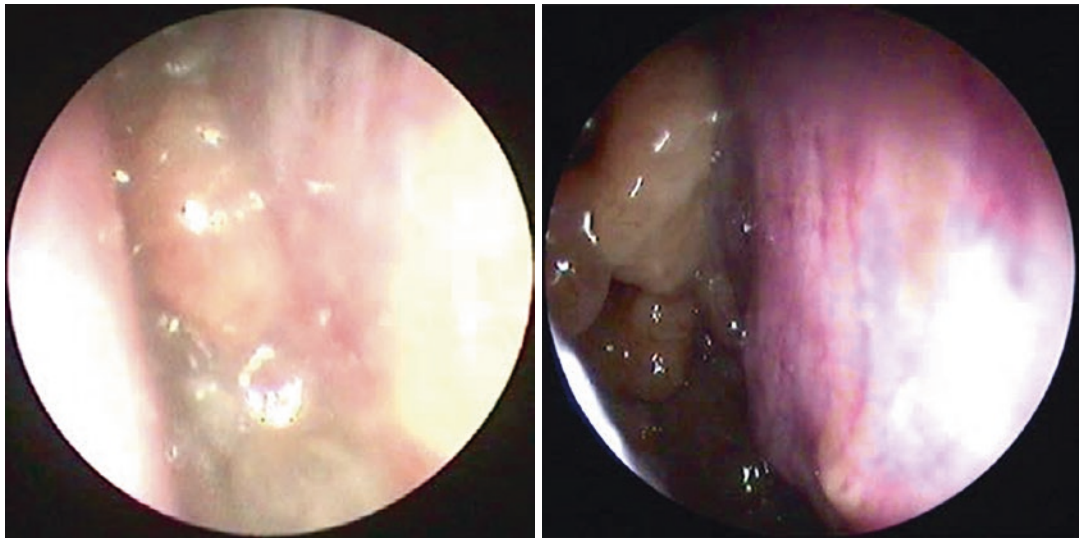

Fig. 13.1 Polyps in the right nasal cavity. (**a**) Before treatment with steroids. (**b**) At 6 months follow up

on the above observations, several theories have been developed. The single pathogen theory has focused on S. Aureus as the primary bacterial factor responsible for the initiation of the inflammatory process (through the secretion of exotoxins). Fungi have also been implicated, but the data are contradictory. The biofilm theory focuses on the formation of complexes with clusters of pathogens such as S. Aureus and P. Aeruginosa, which are protected from the host environment. Although patients with biofilms have a worse prognosis despite any mode of treatment, a definite causative link is still missing. With the application of modern molecular techniques, diverse microbial communities have been detected in the sinonasal mucosa. An imbalance between the different taxa is thought to be the factor initiating or perpetuating the inflammatory process (the dysbiosis theory).

The clinical division into CRSsNP and CRSwNP is artificial and based mainly on the endoscopic picture (Table 13.1). Both presentations share common inflammatory mediators and cells and may have similar outcomes following treatment. Conversely, patients within the same class may have a completely different course of their disease. The inability to correctly identify patient cohorts with similar natural history on clinical grounds solely has led to the new concept of endotypes. Patients are classified based on their inflammatory signature, a specific mix of inflammatory markers (cytokines), and cells (neutrophils and eosinophils). Analysis of inflammatory biomarkers from polyp tissue biopsies has identified 3–10 different disease clusters (Table 13.2). Patients with recalcitrant or steroid-resistant disease may be identified early so that the treatment can be tailored accordingly. To that end, there are a few studies exploring practical ways to identify different endotypes in the clinical setting. Turner et al. have analysed the inflammatory mediators in the nasal mucous and identified six disease clusters.

13.2 Clinical Approach

13.2.1 History

History taking should focus on: the severity and duration of the disease, exclusion of sinister or systemic causes and identification of relevant comorbidities, prior therapies and patient expectations.

Nasal obstruction is the main presenting symptom which responds well to medical and/or surgical treatment. Reduced olfaction may pose a significant burden on quality of life, including

Table 13.1 Phenotypes of nasal polyps

Nasal polyp presentation	Disease phenotypes	Treatment
Unilateral localised	Chronic sinusitis (e.g. odontogenic)	Antibiotics, intranasal steroids, surgery
	Neoplasia (inverted papilloma, carcinoma)	Surgery
Bilateral localised	Ostiomeatal complex inflammation (without allergy)	Intranasal steroids, antibiotics, surgery
	Allergy (central compartment allergic disease)	Intranasal steroids, antihistamines, immunotherapy, surgery
Bilateral diffuse	Eosinophilia (with/without asthma)	Intranasal/oral steroids, doxycycline, surgery, biologics
	Eosinophilia with asthma and ASA intolerance (Samter's triad)	Intranasal/oral steroids, doxycycline, montelucast, aspirin desensitisation, surgery, biologics
	Allergic fungal rhinosinusitis	Intranasal/oral steroids, surgery, fungal desensitisation
	Neutrophilia	Macrolides, surgery, intranasal steroids (usually steroid resistant)
	Cystic fibrosis	Management of local infections, surgery for drainage/irrigations
	Churg-Strauss syndrome	Oral steroids, monoclonal antibodies, cyclophosphamide

Table 13.2 CRS endotypes

Th2 related markers (IL-5, IgE) and eosinophils
Th1 related markers (IL1β, IL-6, IL-8) and neutrophils
Th17/Th22 (IL-17A, IL-22)
Th1 with IFN-γ

From Tomassen et al.

professional life (wine tasters, chefs). Long-standing hyposmia is less likely to improve with any treatment, and patients should maintain realistic expectations. Headaches are unusual, but if present, they could be a sign of a blocked frontal sinus that is inaccessible to steroid sprays and may dictate a short course of oral steroids. Nosebleeds are often the result of an inappropriate application of a nasal spray. It may alert the physician to the presence of a nasal tumour. Severe and challenging to control pulmonary symptoms may raise suspicion of a systematic vasculitis (Churg-Strauss syndrome).

Comorbidities such as allergy, asthma, and intolerance to non-steroidal anti-inflammatory medications (NSAID) or aspirin (ASA) may imply a difficult-to-treat patient and escalation of treatment should not be delayed. Patients with undiagnosed ASA/NSAID intolerance may not report an association of the medications mentioned above with nasal symptoms. However, they may report an exacerbation of nasal congestion on drinking alcohol or eating certain foods that contain salicylates (mushrooms, tomatoes).

The necessity to investigate children and adolescents with bilateral nasal polyps for cystic fibrosis should not be overemphasized. Elderly patients may also present a different subset of patients with reduced immunity to bacteria and a neutrophilic mucosal inflammation that responds poorly to steroids.

In a multicultural environment, the ethnic background of patients has its own relevance. Europeans and Americans often present with a Th2 eosinophilic inflammation; the Chinese predominantly show a neutrophilic inflammatory profile and the Japanese a Th17/Th22 profile.

13.2.2 Clinical Examination

The single most important exam is nasal endoscopy. The presence of polyps confirms the cause of the nasal obstruction. Bilateral, reddish, or oedematous polyps arising from the middle meatus is the most typical scenario. Unilateral, bleeding or white polyps should raise suspicion of a tumour or meningocele, respectively. Bilaterality or inflammatory appearance of the polyps does not exclude a benign tumour such as the inverted papilloma. The presence of brown, thick and sticky secretions (mucin) can point to allergic fungal rhinosinusitis (AFRS) and purulent secretions to the co-existence of a bacte-

rial infection. Limited polyps in the middle meatus area and the posterior septum could be a sign of polyposis with significant allergic contribution.

13.2.3 Imaging

At the initial presentation, imaging is not necessary unless there is a diagnostic challenge. Suspicion of a tumour, meningocele, complication (e.g., meningitis, diplopia), allergic fungal rhinosinusitis, or odontogenic sinusitis will require an initial high resolution computed tomography scan (HRCT). If surgery becomes an option, an HRCT scan is imperative to identify the relevant anatomy of the sinuses (including the frontal sinus drainage pathway) and secondarily the disease extent. A hanging anterior ethmoidal artery, an atelectatic uncinate process, a dehiscent lamina papyracea or skull base, an Onodi cell and an exposed carotid artery in the sphenoid sinus are relevant radiological information to prevent surgical complications. In revision cases, a CT scan will additionally identify any remaining bony septa that need removal or bony dehiscences. Magnetic resonance imaging is only indicated as complementary to HRCT in neoplastic polyps, suspected meningoceles or orbital/central nervous system complications.

13.2.4 Other Tests

Skin prick tests or radioimmunoassay tests for allergens will reveal a concomitant allergy or sensitisation to fungal elements, which is one of the diagnostic criteria of AFRS. A nasal provocation test will identify patients with ASA intolerance who may benefit from desensitisation. Blood tests would reveal serum eosinophilia, suggesting a Th2 inflammation or Churg-Strauss syndrome (Table 13.3). An elevated IgE is characteristic of allergic CRS, recently termed Central Compartment Allergic Disease. Tissue biopsy or nasal mucous analysis to assign patients to certain endotypes has not yet been incorporated into clinical practice.

Table 13.3 Laboratory tests in CRSwNP

Test	Significance
Absolute eosinophil count	>240/μL predicts high tissue eosinophilia
	>1500/μL indicates a hypereosinophilia syndrome
Eosinophil to total WCC in serum	>4% predicts high tissue eosinophilia
	>10% indicates a hypereosinophilia syndrome
Total IgE	>1000 iu/L signifies severe allergy
ESR	high value points to an autoimmune disease
cANCA/pANCA/ENA	high values point to Polyangitis
IgG, IgM, IgA	low values indicate immunodeficiency (e.g. CVID)

ESR erythrocyte sedimentation rate, *ENA* extracted nuclear antibodies, *WCC* white cell count

13.2.5 Medical Treatment

Steroids, in any form, are the mainstay of conservative treatment of inflammatory nasal polyps.

The main goal is to block the inflammatory process and control symptoms. The effectiveness of steroids depends on the type of sinonasal inflammation and mode of delivery. Eosinophilic polyps respond rapidly to oral steroids, whereas neutrophilic polyps do not. The highest tissue concentration of steroids is achieved by oral administration. Polyps are maximally reduced at 3 weeks and then gradually grow almost to their initial size at 2–3 months unless another treatment is given. Therefore short courses of oral steroids are indicated (a) as an initial treatment in severely affected patients to increase patency for intranasal sprays and irrigations (b) as a trial, to check steroid responsiveness of nasal polyps (c) preoperatively, to reduce congestion and provide a better surgical field (d) postoperatively, as an adjunct to prevent stenosis of the frontal recess. Long term low dose prednisolone (2.5 mg/day) has also been used to suppress polyp regrowth in refractory cases. Systemic steroid administration has its consequences. High doses for long term ("high dose" is the supraphysiological dose of >7.5 mg/day in long term treatment, >40–60 mg/day in short term treatment, and long term equals

Table 13.4 Risk factors for adrenal suppression

Oral steroids
>2 weeks of continuous use
>3 cumulative weeks in last 6 months
multiple doses per day

>3 weeks) can lead to adrenal suppression (Table 13.4). Long systemic steroid treatment may also lead to glucose intolerance, osteoporosis, skin fragility, cataract, hypertension, and weight gain. Patients should also be warned for reactions that may occur even after short courses of steroids, mainly mood changes, insomnia, and psychotic episodes without a prior history of psychiatric illness. The risk of avascular hip necrosis is increased with a high total cumulative dose and comorbidities such as excess alcohol intake, hyperlipidemia and cancer. A cumulative dose of 290 mg is considered safe.

Intranasal steroid administration results in less systematic bioavailability and reduced undesired bioactivity. Potency, particle size, lipophilicity and delivered volume of the intranasal steroid agent will define the effect on the nasal mucosa. The highest delivery is achieved, in increasing order, by sprays >> drops >> high volume irrigation. The correct application of sprays and drops also plays a crucial role in drug deposition on the lateral nasal wall. High volume irrigation results in only 5% of the initial steroid dose reaching the nasal mucosa. Most of the intranasal corticosteroid, whatever the form of delivery, is swallowed and inactivated by the first pass metabolism. Systemic absorption occurs through the nasal mucosa. The older corticosteroids (e.g., budesonide) result in higher bioavailability in plasma but also have a short half-life. The newer corticosteroids (mometasone, fluticasone) are highly lipophilic. Although they have a very low plasma concentration, they accumulate in the tissue compartment and are slowly released over a long period. The clinical implication is that isolated intranasal administration at recommended doses may affect plasma and urine cortisol levels but does not have a significant clinical impact on growth and adrenal suppression. The concurrent use of inhaled corticosteroids for asthma increases the total steroid burden and thus, the likelihood of clinically significant systemic side effects. The lowest effective dose of the intranasal spray should be prescribed. If steroid irrigations are planned long term, patients should be monitored for asymptomatic adrenal suppression. Other complications of topical treatment with intranasal steroids are epistaxis due to local trauma and dryness from incorrect use. Spray preservatives such as benzalkonium chloride have not proved deleterious to the nasal mucosa but can cause nasal irritation masquerading as nasal allergy. Prolonged treatment with any form of intranasal steroid has not led to mucosal atrophy.

Alternative medical therapies with probable benefit in CRSwNP are doxycycline, macrolides, leukotriene antagonists and biological factors.

Doxycycline (200 mg/first day followed by 100 mg/day for 3–12 weeks) has been shown to reduce the size of nasal polyps and improve quality of life scores but the treatment effect is much smaller compared to oral methylprednisolone. Patients without asthma or aspirin-exacerbated respiratory disease tend to respond better to this treatment. Although doxycycline has known antibacterial, anti-inflammatory effects and the ability to reach intracellular Staph. Aureus, the exact mode of action is unclear and more studies are necessary to assess its benefit in CRSwNP.

Macrolide anti-inflammatory effect was initially observed in panbronchiolitis patients. Macrolides disrupt the neutrophil functions at several stages, such as cell migration and adhesion and currently form a standard treatment for neutrophilic upper and lower airway inflammation. Long term (12–24 weeks) low dose (250 mg/day) macrolide treatment is effective in CRS without polyps but to date, no clear benefit has been observed in CRSwNP. There is little evidence to suggest a better polyp score with clarithromycin and therefore, macrolide treatment can not yet be justified in eosinophilic CRSwNP.

Cysteinyl-leukotriene receptors participate in the inflammatory process and have been detected in nasal polyps. Montelukast, a cysteinyl-leukotriene antagonist and zileuton, a 5-cyclooxygenase inhibitor that blocks the formation of these receptors, have proved beneficial

in the management of allergy and asthma. There is some evidence that they may also be effective in CRSwNP.

Biologic factors are monoclonal antibodies that block certain targets of the Th2 inflammatory pathway (IgE, IL-4, IL-5, IL-13) and thus improve the local control of nasal polyps. These factors were initially used with success in patients with eosinophilic asthma and urticaria. The idea is that CRSwNP and asthma share a common inflammatory pathway and one systematic treatment can manage both conditions. An emerging indication is uncontrolled nasal polyposis after surgery. According to the EPOS 2020 group, candidates for biologics are those patients who have bilateral nasal polyps, have undergone endoscopic sinus surgery and fulfill at least three of the following criteria: tissue eosinophilia/Th2 markers (evidence of type 2 inflammation), significant decrease in quality of life, need for systemic corticosteroids or contraindication to systemic steroids, significant loss of sense of smell, diagnosis of asthma. Biologics, as an alternative to surgery, have been proposed on the grounds that many patients will receive them for uncontrolled asthma. Treatment response, though, has been modest and many patients will eventually need surgery. The high cost, the risk of anaphylaxis, the need for an injection and the lack of long term disease control should be born in mind before prescribing this novel treatment.

Summary and Author's Conclusions

1. Nasal polyps represent the common end-stage of different inflammatory processes.
2. There are distinct CRSwNP phenotypes with different prognosis and response to medical treatment.
3. Steroids are the mainstay of medical treatment.
4. Patients who receive multiple courses of oral or intranasal plus inhaled steroids should be monitored for short and long term side effects.

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14 Non-invasive Fungal Rhinosinusitis

Nikolaos Tsetsos, Marios Stavrakas, and Petros D. Karkos

14.1 Case Presentation

A generally fit and well 63-year-old lady presented to her GP complaining about retro-orbital headache and mucopurulent discharge. She was initially treated as having a common cold. Later on, the patient developed double vision and ptosis on the left eye. She was subsequently referred to our department for further investigations and definite treatment. On clinical examination, she complained about persistent nasal congestion and mucopurulent discharge. Headache and opthalmological symptoms deteriorated over the time. Flexible nasendoscopy revealed mucopurulent discharge coming from the sphenoethmoidal recess. Ophthalmology review confirmed left oculomotor nerve palsy while CT scan showed complete opacification of the left sphenoid sinus and thickening of the sinus walls (Fig. 14.1). She was started empirically on intravenous broad spectrum antibiotics with no significant clinical improvement. The patient underwent endoscopic sinus surgery, during which the surgeon identified debris coming out of the widened sphenoid ostium (Fig. 14.2). Histological evaluation of the specimen showed nonseptate fungal colonies accompanied by cellular debri and numerous neutrophils. The patient was under regular follow up for 6 months, and no recurrence was noted. Endoscopic evaluation of the nose was unremarkable and the ocular symptoms eventually improved.

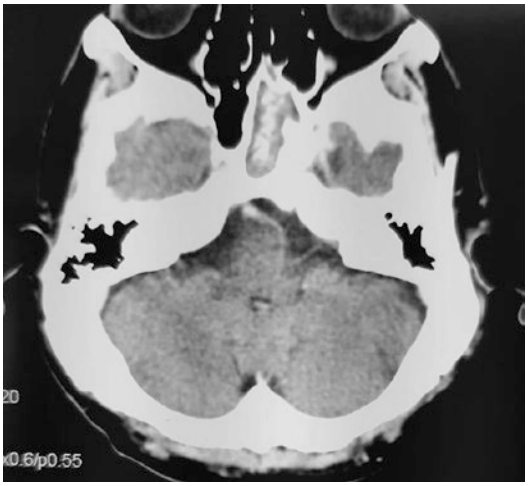

Fig. 14.1 CT sinuses demonstrating complete opacification of the left sphenoid sinus

14.2 Background Knowledge

Fungal Rhinosinusitis (FRS) is commonly classified into two categories based on histopathological evidence of mucosal layer invasion by fungi:

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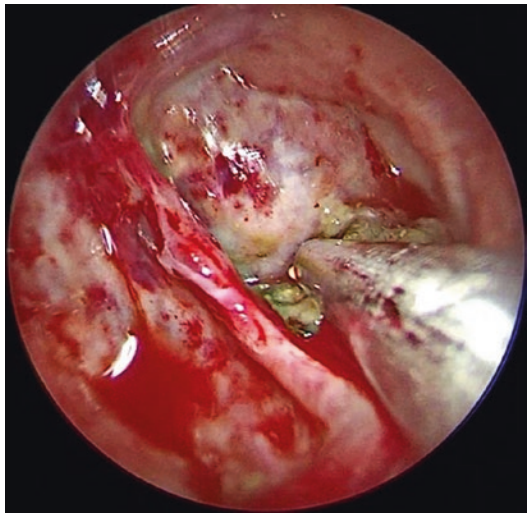

Fig. 14.2 Intraoperative photograph. After wide sphenoidotomy, the fungal ball is debrided and removed

invasive and non-invasive. The invasive diseases include (1) acute invasive (fulminant) FRS, (2) granulomatous invasive FRS, and (3) chronic invasive FRS. The non-invasive forms of FRS include (1) saprophytic fungal infestation, (2) fungal ball, and (3) allergic fungal rhinosinusitis (AFRS).

14.2.1 Saprophytic Fungal Infestation

Saprophytic fungal infestation refers to fungal colonization of the secretions of the sinonasal mucosa. Often seen in patients with a history of previous sinus surgery leaving an inflamed or crusted nasal mucosa with impaired mucociliary clearance, which subsequently gets infected with inhaled fungal spores. It is not invasive and is confined to inflamed and ulcerated/crusted sinonasal mucosa. It is usually diagnosed by endoscopy in an asymptomatic patient; however, it can present with a foul smell in the nasal cavity. This form of FRS is speculated to precede the development of a "fungus" ball. Treatment consists of mechanical removal on endoscopy and nasal douching to avoid recurrence. No formal surgical intervention is advised.

14.2.2 Fungus Ball

A fungal ball is characterized by a non-invasive dense accumulation of fungal hyphae within the mucosa of the paranasal sinuses. According to recent recommendations, fungal ball is considered the most accurate term for this entity as opposed to old non-specific terms, such as "mycetoma" and "aspergilloma".

Fungus ball of the paranasal sinuses is mostly presented in middle-aged to elderly immunocompetent persons with the mean age being in the seventh decade. In most series, it is more prevalent in the female population (2:1). These fungal balls are more frequently identified unilaterally, with the most common site being the maxillary sinus, followed by the sphenoid sinus.

The pathogenesis of the disease is not entirely understood, and no apparent risk factors for the formation of the fungal ball have been described. It has been speculated that certain components of sealers, such as zinc oxide, may play a role. However, the presence of fungus ball in patients without a previous history of dental treatment combined with the fact that fungus ball may be created in the frontal or sphenoid sinus suggests that unknown factors may be involved.

14.2.2.1 Clinical Approach

Diagnosis

The clinical picture of a fungal ball is usually non-specific and nasal discharge, nasal obstruction, headache, and facial pain have all been described. However, it can be recognized as an incidental radiological finding in an asymptomatic patient. It is not uncommon to be encountered as a part of the investigation and treatment for chronic rhinosinusitis (CRS) resistant to traditional medical treatment. The suspicion of a fungus ball is usually raised radiologically by a CT scan. CT scans generally show heterogeneous, hyper-dense sinus opacity, with microcalcifications or metallic appearance, partial or total.

The diagnosis is generally confirmed intraoperatively when 'cheesy' and 'clay-like' inspissated mucous is found within the sinus. The

aggregation of fungal hyphae within one or more sinuses with no evidence of invasion on histopathology, a predominance of eosinophils, granuloma, or allergic mucin, establishes the diagnosis. Culture for fungus is positive in only 30% of the cases, although fungal hyphae are identified on histopathologic examination in more than 90% of the cases.

Fungal balls are not invasive, therefore systemic or topical antifungal medications are not recommended. Treatment consists of endoscopic sinus surgery based on the location of the fungus ball (middle antrostomy, sphenoidotomy, and ethmoidectomy) and management of any contributing factors (i.e., oroantral fistula or retained dental amalgam). Any fungal material should be removed, and the sinus should be irrigated. It is also crucial to biopsy surrounding the mucosa to rule out any microscopic invasion by fungi. The success rate is high with a recurrence rate reported to be as low as 1% after surgical removal.

Topical or systemic antifungal medications should be considered only in case of a fungus ball presenting in an immunocompromised patient. Therefore, close observation of these patients to exclude invasive disease has been recommended instead of prescribing expensive medications with questionable evidence and potential side effects.

14.2.3 Allergic Fungal Rhinosinusitis

This is the most common form of fungal sinus disease and the third form of non-invasive fungal sinusitis. Allergic FRS is usually presented in immunocompetent, atopic patients complaining about symptoms of chronic rhinosinusitis (CRS) resistant to standard conservative medical therapy. Histopathology, AFRS shares similarities with allergic bronchopulmonary aspergillosis. Fungi associated with this disease are diverse, and cultures most commonly grow dematiaceous species, such as Bipolaris, Curvularia, and Alternaria.

The physiopathology of AFRS is controversial and not completely understood. The IgE-Mediated Type I Hypersensitivity and, possibly, IgG-mediated antigen-antibody complex formation (type III hypersensitivity), according to Gell and Coombs classification, are the most commonly cited mechanisms in the development of AFRS. Sinonasal eosinophilia and inflammation initiate a cycle of chronic edema, mucociliary dysfunction, stasis of secretions, combined with viscid allergic mucin, easily obstructs the normal drainage pathway. The fungi that provoke the hypersensitivity live and grow in the mucin stimulating the hypersensitivity reaction continuously.

The typical patient will suffer from chronic rhinosinusitis with nasal polyps refractory to conventional medical and even surgical therapy. Patients may have used several courses of antibiotics and topical nasal medications without resolution of their symptoms. Some clinical characteristics showing an alert sign for the clinician include a young (average age of 22 years), immunocompetent patient with unilateral or asymmetric paranasal sinuses findings, history of atopy, nasal polyposis, and relatively pain-free. In more severe cases, patients will present with proptosis, telecanthus, or gross facial dysmorphia. Patients may discharge a semisolid, thick, viscous consistency of yellow-green, white-brown, gray, brown, or black colour, described by some as a peanut butter appearance and consistency, called allergic/eosinophilic mucin. This mucus is composed of degranulating eosinophils in a background of mucin and fungal hyphae. The histological examination is of primary importance and with show eosinophilia, Charcot-Leyden crystals (a breakdown product of degranulating eosinophils) and non-invasive fungal hyphae. In addition to eosinophils, inflammatory infiltration with lymphocytes and plasma cells is also encountered.

CT images frequently present a dense, unilateral or asymmetric involvement of one or more paranasal sinuses, most commonly of the ethmoidal and maxillary sinuses. The most classical finding of AFRS on CT is the ‘double density’ sign. This is caused by metallic densities of fungal hyphae within the eosinophilic mucin surrounded by hyperplastic mucosa. Bony erosion may allow the disease to invade the neighboring

Table 14.1 Bent and Kuhn criteria

Major	Minor
Type I hypersensitivity	Asthma
Nasal polyposis	Unilateral disease
Characteristic CT findings	Bone erosion
Eosinophilic mucin without invasion	Fungal cultures
Positive fungal stain	Charcot-Leyden crystals
	Serum eosinophilia

CT computed tomography

tissues, affecting vital organs such as the brain, orbit, and large vessels. Peripheral enhancement caused by the inflamed mucosa and a central low signal on both T1 and T2 is the characteristic findings on MRI.

The most widely accepted diagnostic criteria were published by Bent and Kuhn, based on the histologic, radiographic, and immunologic characteristics of the disease (Table 14.1).

14.2.3.1 Treatment

The ideal treatment of patients with AFRS is still elusive and controversial. The management aims to remove as much antigenic and inflammatory load as possible and restore ventilation and drainage of the sinuses something that is achieved by functional endoscopic sinus surgery (FESS). Complete ventilation of the paranasal sinuses and removal of all nasal secretions, fungal mucin, fungal debris, nasal polyps with respect of the underlying mucosa not only decreases the fungal antigens but also allows access for post-operative treatment.

As for the post-operative treatment, it includes either immunomodulation (immunotherapy and/or corticosteroids) or antimicrobial and antifungal medications. Post-operative treatment should start instantly after surgery by nasal saline irrigation if a good long term outcome is to be achieved. Systemic corticosteroids are valuable in the post-operative period and coupled with nasal sprays are the most effective agents in preventing recurrences. Oral antifungals have been used in the post-operative management of AFRS to reduce the fungal load and, as a result, the immune response to it. However, given their disputed benefit and their side effects, they should be used as a last resort in patients not adequately responding to steroid therapy. The evidence for the use of topical antifungals and leukotriene modulators (i.e., Montelukast) is inadequate, and no recommendations have been made for their routine use.

Immunotherapy has also been shown to be very efficient, and when combined with surgery and medical treatment with corticosteroids, it prevents recurrences and reduces the need for corticosteroids in the future. Lastly, monoclonal antibodies constitute a novel therapy in patients suffering from CRS with nasal polyps and concomitant asthma. By targeting IgE, IL-5 cytokine pathways, omalizumab, and mepolizumab have shown encouraging results, and biologic therapy seems to be safe and well-tolerated. However, high-quality trials designed to assess these therapeutic alternatives for this specific subpopulation of patients with AFRS are called before recommendations can be made on their use.

Summary and Author's Comments

1. It is speculated that fungi play a developmental role in CRS, but their exact mechanisms remain unclear.
2. The wide variety of clinical manifestations in fungal sinus disease makes it a challenging entity to be diagnosed and treated effectively.
3. High-quality randomized controlled trials are required to determine the real benefit of immunotherapy, antifungal therapy and monoclonal antibodies in the treatment of AFRS.
4. AFRS is considered a chronic disease that may recur even after radical surgical and aggressive medical therapy and appropriate follow-up.

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Balloon Sinuplasty

15

Mohammed Salem, Marios Stavrakas, and Hisham S Khalil

15.1 Case Presentation

A 36-year-old man was referred to our rhinology service from the maxillo-facial team, complaining of persistent left maxillary discomfort and upper jaw pain for the last 13 months. There was no local dental or temporomandibular joint cause to explain the unusual discomfort. He denied nasal symptoms, including nasal infections, trauma, previous surgery or signs of allergic rhinitis. Nasendoscopy revealed normal mucosa, with mild ipsilateral congestion of middle meatus. The CT scan showed slight mucosa thickness of left maxillary sinus with ipsilateral opacification of the osteomeatal complex. After the failure of medical treatment, a discussion took place regarding the surgical options. Since the disease was isolated and confined to one sinus, we offered FESS or balloon sinuplasty and the patient opted for the second approach. The balloon sinuplasty was performed under sedation; the patient was discharged on the same day and had an uneventful postoperative recovery.

15.2 Background Knowledge

Balloon sinuplasty (BSP) is a minimally invasive technique which was approved by the Federal Drug Administration (FDA) in 2005. It became more popular for the treatment of chronic rhinosinusitis (CRS) and rarely acute sinusitis. It shares a similar concept with the Seldinger technique used by cardiologists and basically it aims to dilate the ostia and leave the mucosa intact, maintaining physiological function at the same time.

According to Hopkins et al. (2011), balloon sinuplasty carries several advantages. It can be useful in acute sinusitis because of its minimally invasive nature. Also, it can be performed under local anaesthesia, which makes sinus ostia dilatation and washouts easier, even in a non-theatre environment. Obviously, adequately trained staff is a prerequisite.

BSP complications included device failure, CSF leak in the ethmoid roof and sphenoid and intraoperative cardiac arrest. Equally serious complications that have been recorded are orbital and skull base injuries, due to false passage into the orbit or cranium.

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According to the literature, ostia patency was 80.5%, 85.1% and 91.6% at 6, 12 and 24 months, respectively. Hopkins et al. (2011) stated that success rate post ostia dilation was 98% but definitive improve was noted in only 62% of patients.

15.3 Clinical Approach

15.3.1 History

We follow the same history taking principles as in patients with acute, recurrent acute or chronic rhinosinusitis. It is essential to take into consideration the presence of polypoidal nasal disease, previous treatment, and most importantly, previous surgeries that may have altered the anatomy. A complex medical history, age and patient's expectations are factors that are worth exploring, as they may favour a minimally invasive intervention.

15.3.2 Examination

Head and neck examination, including nasendoscopy, is the first step. Patients with nasal polyposis are not good candidates for balloon sinuplasty only, and FESS polypectomy should be considered.

15.3.3 Investigations

CT scan of the sinuses is essential and as in endoscopic sinus cases, we do not proceed without any radiological information. We want to assess the extent of the disease, the number of sinuses involved and the Lund-Mackay score.

15.3.4 Treatment

The type of anaesthetic depends on the patient's medical status and wishes. Our patients have either a general anaesthetic or sedation + local anaesthetic. Recently we started using image-guided balloons, which makes the placement easier, allows closer supervision when a trainee performs the surgery and also has the potential of reducing any complications. After identifying the ostium in this case, the left maxillary antrum sinus was dilated successfully up to 12 atmospheres for 10 s. The dilated ostium is inspected with a 30° scope. If packing is required, we prefer dissolvable packs. These are day cases and the patients are advised to use steroid nasal sprays/irrigations and saline douches.

Summary and Author's Comments

1. Balloon sinuplasty can be considered for selected cases of CRS and ARS.
2. It represents a good option for opening of the sinus ostia under local anaesthetic or in a non-theatre environment.
3. We can achieve good ostia patency but improvement of the patient's symptoms varies.

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Mucoceles of the Paranasal Sinuses

16

Hisham S Khalil and Abdulaziz Abushaala

16.1 Case Presentation

A 46 year old man presented with a 6 week history of a frontal swelling and right frontal pain. His symptoms had not improved with amoxicillin prescribed by his GP. A CT scan of the paranasal sinuses and a subsequent MRI scan confirmed a frontal sinus mucocele. This was drained through a combined external frontal sinustomy and functional endoscopic sinus surgery. The patient made an uncomplicated recovery.

16.2 Background Knowledge

Mucoceles are benign, cystic and slow growing lesions found in the paranasal sinuses, that are locally expansile, causing bony resorption and displacement of adjacent structures.

Histologically they are mucus-containing cystic lesions that are covered by pseudostratified columnar epithelium. Mucoceles mainly affect the frontoethmoidal region (60–89%) and less commonly in maxillary and sphenoid sinuses. They are usually unilateral. The exact pathophysiology is not clear; however, sinus ostium obstruction and inflammation were found to be contributing factors. It is postulated that a combination of ostium obstruction and infection results in cytokines release from lymphocytes and monocytes, subsequent release of collagenase and prostaglandins with increased bone resorption.

Mucoceles are of a primary type if appearing de novo or secondary if following trauma or sinus surgery. They can be found in association with chronic rhinosinusitis. They may also be associated with fibrous dysplasia and sino-nasal malignancy.

The clinical presentation depends on the site and size of the mucocele. The symptoms are mainly due to involvement of neighbouring structures mainly the orbit (proptosis, visual disturbances, diplopia) and intracranial structures (Headache and cranial nerve palsies) in addition to nasal blockage and facial pain. It was reported that over 80% of patients with mucoceles present with proptosis, and 70% of patients presented initially to Ophthalmologists.

A pyocele, is an infected mucocele which requires urgent medical attention as it may result in serious complications including orbital abscess, meningitis or intracranial abscess Mucoceles' are best investigated by CT and MRI

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scans. The treatment of mucocele is surgical in the form of Endoscopic marsupialisation and drainage of the affected sinus. Combined endoscopic and external approach may be utilised. External approaches include an osteoplastic flap (less commonly used), a mini-osteoplastic flap centred around one rather than both frontal sinuses and a frontal sinus trephine/sinustomy. The endoscopic approaches aim to marsipulise the mucocele by opening the frontal recess and possible widening of the frontal sinus ostium via a Draf I–III procedure. An exclusively endoscopic approach is unsuitable for laterally-placed mucoceles of the frontal sinus or where there is a complete inter-sinus septum.

16.3 Clinical Approach

16.3.1 History

The patient had a long history of nasal obstruction, nasal and postnasal discharge, reduced smell and facial discomfort. He had been using a steroid nasal spray intermittently. Six weeks prior to presentation he noticed a swelling over his forehead with increasing facial discomfort. His GP started him on a 7 day course of amoxicillin but he failed to improve.

16.3.2 Examination

The patient presented with an obvious frontal swelling (Fig. 16.1). An ophthalmological assessment was normal. A nasal endoscopy revealed bilateral grade II inflammatory looking polyps.

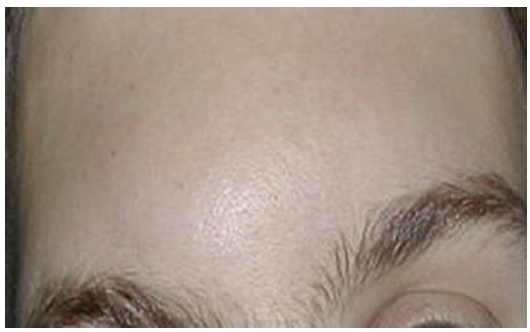

Fig. 16.1 Patient presenting with a frontal swelling

16.3.3 Investigations

A combination of CT and MRI scans are required to confirm the diagnosis. The T2-weighted MRI scan revealed a high signal within the frontal swelling. A CT scan of the paranasal sinuses revealed the mucocele to be within a Type IV Kuhn cell (Fig. 16.2). There was evidence of erosion of the posterior table of the right frontal sinus (Fig. 16.3).

The state of the anterior and posterior tables of the frontal sinuses, involvement of fronto-ethmoidal cells, the presence of inter-sinus septa and associated pathology are all important areas to consider when reviewing the imaging.

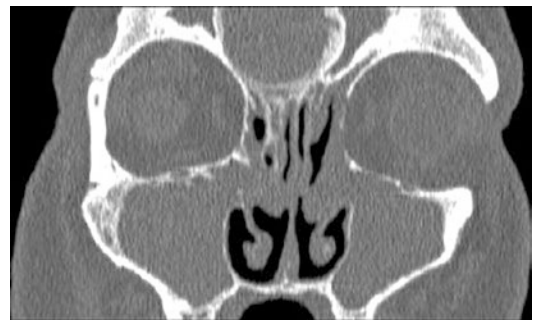

Fig. 16.2 Coronal CT of the paranasal sinuses demonstrating a large mucocele in a Type IV Kuhn cell

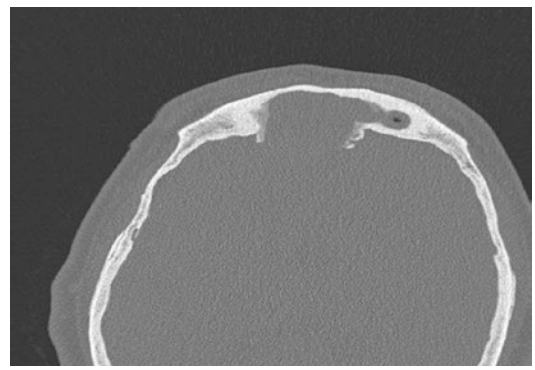

Fig. 16.3 Axial CT of the paranasal sinuses demonstrating the frontal sinus mucocele with erosion of the posterior table of the frontal sinus

16.3.4 Treatment

The treatment of mucoceles is surgical with medical treatment also required for pyoceles/infected mucoceles. The decision on the surgical approach will depend on the site and size of the mucocele, presence or absence of erosion of the anterior and posterior tables, associated pathology as well as the functional status of the patient.

This patient had two relevant factors to consider; the mucocele within a Type IV Kuhn cell with potential difficulties draining this through an endoscopic approach only, as well as erosion of the posterior table with a potentially increased risk of intracranial complications. The presence of a defect in the posterior table of the frontal sinus is also a relative contraindication to an osteoplastic flap. Given, these two factors, it was decided to drain the mucocele through a combined external frontal sinustomy and endoscopic sinus surgery.

The external frontal sinustomy was performed through an incision just below the eyebrow. Care should be taken when making the incision and deepening it, to avoid injury of the supraorbital and supratrochlear nerves and trochlea in the supero-medial compartment of the orbit. The trephine is best made in the inferior aspect of the frontal sinus except if there is an existing breach of the anterior table (Fig. 16.4). The sinustomy allowed full inspection of the drained mucocele cavity, defect in the posterior table and removal of the walls of the Type IV Kuhn cell. The patient had simultaneous bilateral functional endoscopic sinus surgery with an extended frontal recess approach (Draf I). The patient was followed up for 12 months with no recurrence.

Summary and Author's Comments

The state of anterior and posterior tables of frontal sinus, type of Kuhn cell and presence of a complete intrasinus septum all influence the choice of surgical approach used. The general condition of the patient and the presence of associated pathology should also be considered when treating patients with frontal sinus mucoceles.

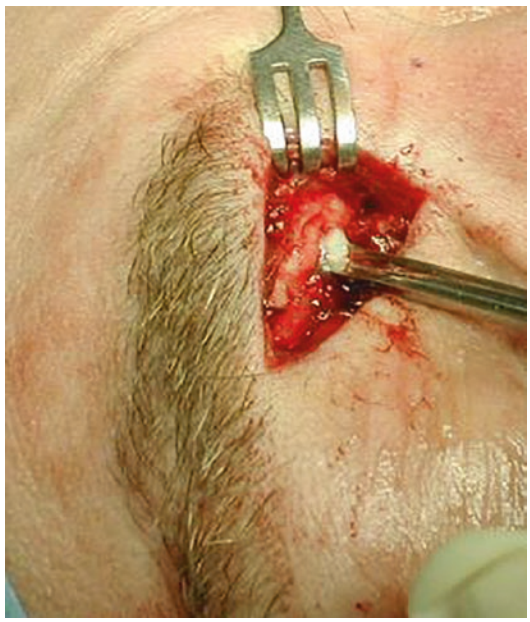

Fig. 16.4 Incision of an external frontal sinustomy below the right eyebrow with a trephine in the floor of the right frontal sinus

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Concha Bullosa

17

Marios Stavrakas and Petros D. Karkos

17.1 Case Presentation

A patient was referred to the ENT clinic with symptoms of chronic sinusitis, particularly facial pressure, especially on the right side of his face. His symptoms failed to improve despite a long course of intranasal steroid spray. A CT scan showed septal deviation to the right and a large concha bullosa, obstructing the right osteomeatal complex. The maxillary sinus appeared completely opacified. Surgical intervention was planned in terms of septoplasty, concha bullosa reduction and right middle meatal antrostomy. The patient recovered fully and his nasal obstruction improved significantly. He was put on long term intranasal steroid spray.

17.2 Background Knowledge

Concha bullosa is one of the most common variations of the middle turbinate, and its frequency varies between 14% and 53%. The term "concha bullosa" (CB) is used to describe aeration in the horizontal part of the turbinate, with or without the involvement of the vertical component.

Bolger et al. (1991) categorized the pneumatised middle turbinate:

1. Lamellar concha bullosa=pneumatization of vertical lamella
2. Bulbous concha bullosa=pneumatization of inferior segment
3. Extensive concha bullosa=pneumatization of both parts.

Like other aerated cells, the CB possesses a mucociliary transport system with the ostium connecting the airy cell lumen to the frontal recess. Obstruction of this ostium by trauma, polyps, surgery, or tumours can lead to a mucocele. The formation of a mucocele or a mucopyocele represents potential complications of a concha bullosa. A mucocele occurs when an epithelium - lined cavity is filled with mucus. In case of infection of the content, the resultant pathology is a pyocele (Fig. 17.1). A mucocele is formed when there is a poor function of the mucociliary transport system, mainly due to obstruction. Also, concha bullosa may result in osteomeatal complex obstruction and may predispose to sinus disease (CRS).

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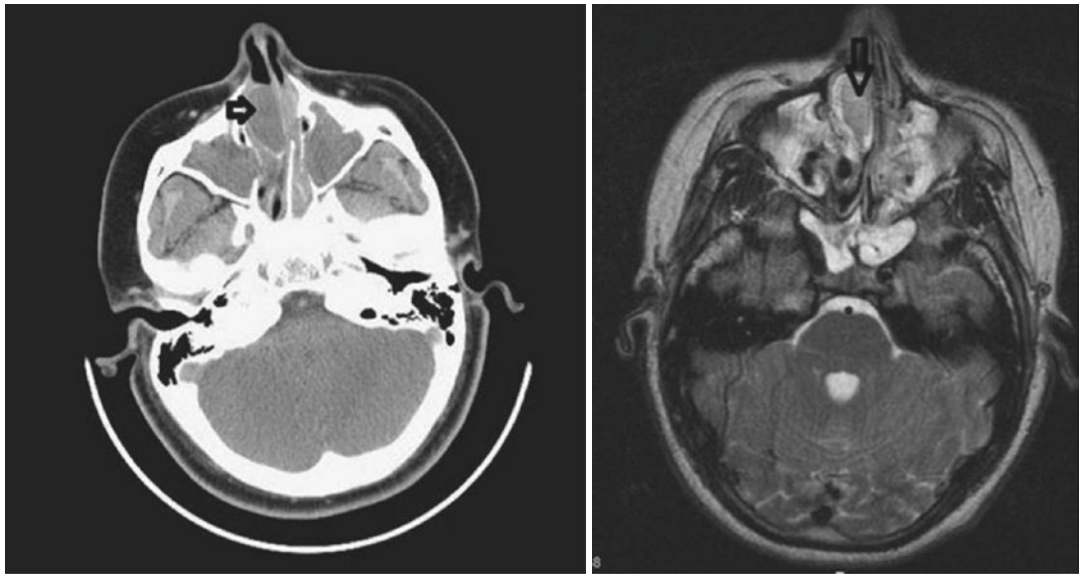

Fig. 17.1 A case of concha bullosa pyocele. CT and MRI scans have revealed a right concha bullosa, with signs of opacification consistent with concha bullosa mucocele/pyocele (arrows). (Karkos PD, Stavrakas M, Triaridis S, Markou K, Tsalighopoulos M. Bardet-Biedl syndrome and a large concha bullosa pyocele. Int J Pediatr Otorhinolaryngol. 2014;78(12):2316–2318. https://doi.org/10.1016/j.ijporl.2014.10.012)

17.3 Clinical Approach

17.3.1 Diagnosis

CB alone is usually asymptomatic and is, most of the time, an incidental finding on CT scans. Headache, orbital pain, nasal discharge, postnasal drip, nasal obstruction, and loss of the sense of smell are all possible symptoms of a patient with CB pyocele.

Anterior rhinoscopy is inadequate for diagnosing either conchal pneumatization or a pyocele/mucocele, as it can only provide information about a conchal mass and its obstructive effect on the nasal passage. CT gives valuable information about nasal and paranasal structures and can reveal conchal pathology and coronal plane CT of the paranasal sinuses is routinely applied to evaluate patients with sinus-like complaints. Magnetic resonance imaging (MRI), however, is considered superior to CT in diagnosing the intracranial and intra-orbital complications of inflammatory paranasal sinus disease.

17.3.2 Treatment

When it comes to the treatment of mucoceles and pyoceles, endoscopic surgery is the method of choice. The methods described for the treatment of concha bullosa and its complications (mucocele/pyocele) are lateral marsupialisation, medial marsupialisation, crushing, and transverse excision. Also, occasionally it is required to use one of the above methods to reduce a concha bullosa to improve access to middle meatus when performing ESS and correct osteometal complex obstruction.

Our preferred treatment method is lateral marsupialisation.

- Infiltration with LA
- Incision on the inferior border of turbinate until reaching a hollow cavity
- Resection of the lateral part of turbinate using microdebrider or cold instruments
- The attachment of the turbinate and the medial portion are preserved
- Packing is applied if deemed necessary

17.3.3 Follow-up

As this condition is most frequently treated in the context of endoscopic sinus surgery, follow-up is planned as per FESS follow-up protocols.

Summary and Author's Comments

1. Concha bullosa (pneumatized turbinate) is one of the most common variations of the middle turbinate, and its frequency varies between 14% and 53%.
2. CB alone is usually asymptomatic and is, most of the time, an incidental finding.
3. Endoscopic surgery is the treatment of choice.

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Silent Sinus Syndrome

18

Nikolaos Tsetsos and Marios Stavrakas

18.1 Case Presentation

A 56-year-old male patient was referred by the Opthalmology clinic with silent sinus syndrome. He initially presented with enotpthalmos and hypoglobus which deteriorated gradually over the years. He had a computed tomography (CT) scan of his orbits and sinuses which confirmed the diagnosis. After discussion about the potential treatment modalities, the patient underwent endoscopic sinus surgery-middle meatal antrostomy. During follow-up, there was no further deterioration or diplopia and further surgical intervention was not required.

18.2 Background Knowledge

Silent sinus syndrome is a clinical entity that refers to progressive enophthalmos and hypoglobus due to gradual collapse of the orbital floor with opacification of the maxillary sinus, associated with subclinical maxillary sinusitis. This condition was first described by Montgomery in 1964 in two patients with maxillary sinus mucocele but the term "Silent Sinus Syndrome", was introduced in 1994 by Soparker et al.

Although, the exact pathophysiology of silent sinus syndrome is still elusive, several possible mechanisms have been described. Hypoventilation of the maxillary sinus secondary to ostial obstruction seems to be the most widely accepted theory. Subsequently, the hypoventilation leads to resorption of gases into the capillaries of the maxillary sinus, which creates a vacuum. This results in an accumulation of secretions with chronic subclinical inflammation, leading to maxillary atelectasis and wall collapse; a process similar to that presenting in the middle ear of a patient with eustachian tube dysfunction. The retention of secretions and the chronic inflammation may result in osteolytic activity and osteopenia rendering the orbital floor thin. The thinning of the orbital floor combined with the negative pressure results in herniation of orbital tissue into the maxillary sinus, a mechanism that seems to be responsible for the enophthalmos and the hypoglobus in the silent sinus syndrome. Congenitally hypoplastic maxillary sinus has also been suggested as a predisposed condition responsible for the development of the disease. However, the syndrome is also reposted to occur in patients with normal sized maxillary sinuses based on radiographic imaging having been performed prior to disease onset suggesting that it is an acquired condition and not congenital hypoplasia.

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18.3 Clinical Approach

18.3.1 History

Silent sinus syndrome usually presents during the third to fifth decade of life, with no gender predisposition. It almost exclusively occurs unilaterally in the maxillary sinus. The main clinical features include ophthalmologic complaints such as spontaneous, gradual and progressive enophthalmos over the course of several weeks to several months and hypoglobus. Additionally, patients may present orbital asymmetry, a deepening of the superior orbital sulcus, or eyelid retraction and ptosis of the affected eye. Visual acuity, intraocular pressure and extraocular movements are usually normal. Only a few patients may complain of diplopia and others may seek ophthalmologic evaluation for the protrusion of the contralateral eye due to misinterpretation of the retraction of the affected eye. Symptoms related to rhinitis or sinusitis such as nasal congestion, nasal discharge and headache are quiet rare.

18.3.2 Clinical Examination

The diagnosis of silent sinus syndrome is based on the characteristic clinical features of gradual, progressive enophthalmos and/or hypoglobus, in the absence of orbital trauma (including surgery) or sinus disease, nasal endoscopy and CT scans of the nose and paranasal sinuses. Nasal endoscopy will invariably show normal or mildly inflamed nasal mucosal lining, enlarged middle meatus, uncinate process completely retracted laterally obstructing maxillary natural ostium and no pathologic secretion in the ethmoido-maxillary drainage pathway.

18.3.3 Investigations

Computed tomography (CT) is the best radiographic modality for the assessment not only of the paranasal sinus diseases but of the silent sinus syndrome as well. Magnetic resonance imaging (MRI) may also be a useful tool for the evaluation of orbital and paranasal sinus soft tissue, but has the disadvantage of poorer results of bone assessment as compared to CT. The images usually demonstrate complete or near complete unilateral opacification of the maxillary sinus and obstruction of the ostomeatal complex. Lateralization of the uncinate process is a common radiographic finding resulting in enlargement of the middle meatus when compared to the contralateral side. The most common imaging finding is the inward retraction of the medial wall, posterolateral wall, and orbital floor associated decrease in sinus volume. The reduction in maxillary sinus volume results in orbital volume enlargement. Most commonly the orbital floor is thinned due to osteopenia or may be completely absent from complete resorption, however, thickening of the bony walls may be encountered as well. Most commonly, the normal upward convexity of the orbital floor is lost with inferior orbital wall down-ward bowing with resulting concavity of the superior maxillary wall. The globe is typically displaced inferiorly and the orbital contents accompany the globe into the maxillary sinus, resulting in hypoglobus creating the eye or facial asymmetry identified by the patient. The inferior rectus is most often affected.

Most possibly, the histopathologic findings show thickening and oedema of the maxillary sinus mucosa with chronic nonspecific inflammatory cell infiltration with occasional reparative bone changes. Bacterial cultures are usually negative.

Silent sinus syndrome is a rare clinical entity, therefore, it is essential to be differentiated from a wide range of diseases that are presented with hypoglobus and enophthalmos. The most common diseases include orbital trauma, chronic sinusitis, osteomyelitis, malignant infiltration, congenital facial asymmetry, diffuse facial lipodystrophy, Parry-Romberg Syndrome, linear scleroderma and pseudo-enophthalmos.

18.3.4 Treatment

Treatment of silent sinus syndrome involves two steps: (1) clearing of the sinus and restoration of normal maxillary sinus aeration, and (2) re-

establishment of normal orbital anatomy. The desirable outcomes can be achieved either in single or two-stage surgery.

The Caldwell-Luc approach was most commonly used in the past but it has been replaced by Functinal Endoscopic Sinus Surgery which is nowadays the "gold standard" for addressing the maxillary sinus pathology. If a deviated septum contributes to the formation of the disease and impedes access, a septoplasty should be the first step of the operation. The recommended method is an endoscopic uncinectomy and opening of the maxillary sinus ostium. The surgeon must be mindful of the abnormally low position of the globe and orbital floor. The chance of causing damage to the orbital contents is significantly higher in these patients than those undergoing endoscopic sinus surgery for others conditions. Image-guided technology can be practical but does not replace a thorough examination and understanding of the nasal and orbital anatomy. A wide antrostomy is the goal. A balloon sinuplasty has recently been proposed as a less invasive, alternative option to successfully treat silent sinus syndrome, however, it is not yet clear if this technique can be applied in all cases of maxillary sinus atelectasis. Prospective studies are needed to establish whether the use of balloon sinuplasty is safe and effective for the treatment of this modality.

The second stage in the treatment is a surgical procedure done to restore the orbital volume and symmetry, typically through placement of an orbital implant. There is a wide variety of diferrent implants for floor reconstruction, including both alloplastic materials and autogenous grafts. Transconjunctival and subciliary approaches to the orbit are most commonly preferred.

The optimal time for addressing the orbital floor is still under discussion. Multiple studies suggested that orbital floor restoration should be performed simultaneously with the sinus treatment. The main advantage is avoiding the morbidity of a second hospital stay and a second anesthesia event. There is evidence of spontaneous resolution of the enophthalmos and hypoglobus following only restoration of the maxillary sinus aeration without reconstructing the orbital floor. The two-stage approach to orbital repair is suggested today, and orbit should be addressed at least 6 months after the first surgery. The orbital floor repair is absolutely advised when despite the endoscopic sinus surgery, the enophthalmos or hypoglobus persist 6 months after the first stage. A two-stage approach has the edge of preventing placing an orbital implant in an infected orbit and preventing overcorrecting the globe position.

Summary and Author's Comments

1. Silent sinus syndrome is a rare entity of spontaneous, gradual, progressive asymptomatic collapse of the maxillary sinus.
2. The diagnosis is based on the gradual onset of enophthalmos and/or hypoglobus, in the absence of orbital trauma (including surgery) or prior symptoms of sinus disease.
3. Treatment is surgical, functional endoscopic sinus surgery to address the maxillary sinus atelectasis as the first stage, and orbital floor repairing if significant symptoms persist 6 months later, as the second stage.
4. Endoscopic maxillary antrostomy needs to be done with care to avoid injury to the orbital contents due to lateral position of the uncinate process and low position of the globe and orbital floor.
5. Silent sinus syndrome is presented with a combination of ocular and sinonasal findings, so both otorhinolaryngologists and ophtalmologists should be familiar with it.

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Part IV

Facial Plastics

Functional Septorhinoplasty for Nasal Obstruction

19

Hisham S Khalil and Marios Stavrakas

19.1 Case Presentation

A 24-year old woman presented with a long history of bilateral persistent nasal obstruction and concerns regarding the shape of her nose. She underwent an external septorhinoplasty with improvement of her nasal breathing and nasal appearance.

19.2 Background Knowledge

Rhinoplasty is a vast field with various approaches, indications, techniques, pre- and post-operative evaluation strategies. This short chapter aims to outline the principles of pre-operative evaluation and the basic surgical approach.

Functional rhinoplasty is a term commonly used to describe any technique aiming to address nasal obstruction in the nasal valve area.

19.3 Diagnosis

1. Diagnosis and pre-operative evaluation
   - History
   - Nasal obstruction, reduced nasal airflow
   - Nasal trauma
   - Cleft lip/palate +/− previous surgery
   - Previous septal surgery and/or rhinoplasty
   - Sinonasal pathology
   - Rhinitis (allergic, non-allergic)
2. Examination
   - Inspection: thorough assessment of nasal pyramid, aesthetic considerations, other facial features, e.g., chin, ethnic nose
   - Palpation: tip support, skin, symmetry, tenderness
   - Cottle's/modified Cottle's manoeuvres for the assessment of nasal valve
   - Endoscopic evaluation: dynamic vs static valve collapse, posterior vs anterior septal deviation, other sinonasal pathology (e.g., nasal polyposis)
   - Anterior rhinoscopy
3. Objective assessment
   - Peak Nasal Inspiratory Airflow Flow
   - Rhinomanometry
   - Acoustic rhinometry
   - Nasal Obstruction and Septoplasty Effectiveness (NOSE) score, Visual Analogue Score(VAS)
   - Body Dysmorphic Disorder assessment-BDDQ score

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4. Photography
   - Nasal series: Necessary visual information in the events of rhinoplasty patients are the anterior, profile (left and right), basal, oblique and cephalic views. Standardisation of photographic documentation in rhinoplasty surgery is widely recognised as being critical in enabling accurate evaluation of individual patient outcome and also the comparison between different patients. Standardisation reduces the variables such as distance to subject, patient positioning, background, lens distortion, exposure, and lighting that can alter our perception of anatomical detail and give false postoperative surgical appearances. Frankfurt plane: a plane passing through the inferior orbital margin and the upper margin of each ear canal or external auditory meatus. This plane is chosen because the inferior orbital rim is an easily palpable bony landmark that can be easily identified. Also, appropriate illumination, selection of camera, and background are essential.
   - Computer imaging and morphing

Basic aesthetic considerations

- Horizontal 3rds
- Vertical 5ths
- Angles: Nasolabial (M 90°–95°, F 95°–110°), nasofrontal (115°–135°), nasofacial (30°–40°)
- Projection: Goode's Method → alar grove to tip: nasion to tip = 0.55–0.6
- Tripod theory: tip height and rotation
- Other facial features: chin/retrognathia, facial asymmetry

19.4 Treatment

Rhinoplasty or septorhinoplasty when combined with septal surgery can be carried out for aesthetic or functional reasons. Aesthetic principles should be observed at all times irrespective of the intention of the surgery. The choice of an endonasal or an external approach will depend on many factors including:

- The skills and preference of the surgeon
- A history of previous rhinoplasty
- The need for tip work

19.5 Basic Rhinoplasty Techniques

1. Incisions
   - Intercartilaginous: between the upper and lower alar cartilages
   - Transcartilaginous: through the lower alar cartilage at the location of the cephalic resection
   - Infracartilaginous/marginal: along the caudal border of the alar cartilage with extension along the columella caudal to the medial crura
   - Columellar: usually inverted V or stairstep-shaped, for open rhinoplasty approach
   - Transfixion: incision through both sides of the membranous septum
2. Septoplasty
   - Follow the basic principles of septoplasty with preservation of caudal and distal struts. In cases of crooked septum, inadequate tip support, need for extensive grafting, an extracorporeal approach may be considered, using PDS plates and various graft materials.
3. The Nasal Hump
   - Removal of osteocartilagenous hump
   - Reduce caudal septum
   - Reduce lateral crura of lower lateral cartilages
   - Shield graft
4. Mid-third and osteotomies
   - Medial osteotomies
   - Lateral osteotomies (endonasal or external)
   - Superior/horizontal osteotomies
   - Intermediate osteotomies (with a crooked nose)

- Spreader grafts (with a narrow middle third of the nose)

5. Strategies for the 'Crooked' Nose
   - Septoplasty
   - Separation of upper lateral cartilages from the nasal septum
   - Osteotomies
   - Digital manipulation of central complex
   - Camouflage grafts
6. Increasing Nasal Tip Projection
   - Dome or shield graft
   - Collumelar strut
7. Decreasing Nasal Tip Rotation
   - Full septal transfixion incision
   - Lower septal angle
   - Shorten crura of lower lateral cartilage
8. Decreasing Nasal Tip Projection
   - Reduce caudal septum
   - Augment the nasal dorsum
9. Increasing Nasal Tip Definition
   - Intradomal sutures
   - Transdomal sutures
10. Augmentation techniques
    - Septal cartilage
    - Auricular cartilage
    - Rib cartilage

19.6 Clinical Approach

19.6.1 History

The patient complained of bilateral, alternating nasal obstruction that failed to improve with a combination of a saline nasal douche and a steroid nasal spray. She had a number of concerns regarding the shape of her nose, including a dropping tip, prominence of the bony and cartilaginous framework and a somewhat 'twisted nose'. The patient did not have a history of facial trauma or previous nasal surgery. There was no history suggestive of a Body Dysmorphic Disorder (BDD). This is important to exclude before any nasal framework surgery. It is equally important to seek the opinion of a clinical psychologist if the surgeon suspects a BDD or mental health problems that may result in patient dis-satisfaction postoperatively.

There was no history of a bleeding tendency and the patient was not on any medication that increased the risk of postoperative bleeding.

19.6.2 Examination

The patient had bilateral restriction of the nasal airflow but no evidence of alar collapse on inspiration. The Cottle's test was positive bilaterally. A nasal endoscopy revealed an S-shaped deviation of the nasal septum compromising both nasal valve areas.

The patient had a 'drooping' tip accentuated by the presence of an osteo-cartilaginous nasal hump.

19.6.3 Investigations

Preoperative photographs are required before surgery (Fig. 19.1). Some surgeons prefer to counsel patients pre-operatively with 'profiling' using a photo editing software. Other investigations are requested depending on the patient's clinical history.

19.6.4 Treatment

An external septorhinoplasty approach was chosen due to the presence of a tip ptosis and the ease of bilateral cephalic trims of the lower lateral cartilages through this approach.

The surgical principles adopted to address the patient's functional and aesthetic concerns were as follows:

- An endonasal septoplasty to straighten the septum and the nasal framework. This included separating the upper lateral cartilages from the nasal septum to help straighten the cartilagenous pyramid.
- Medial, external lateral and superior (horizontal) osteotomies to straighten the bony pyramid.
- Removal of the osseocartilagenous hump and bilateral cephalic trimming of both lower lat-

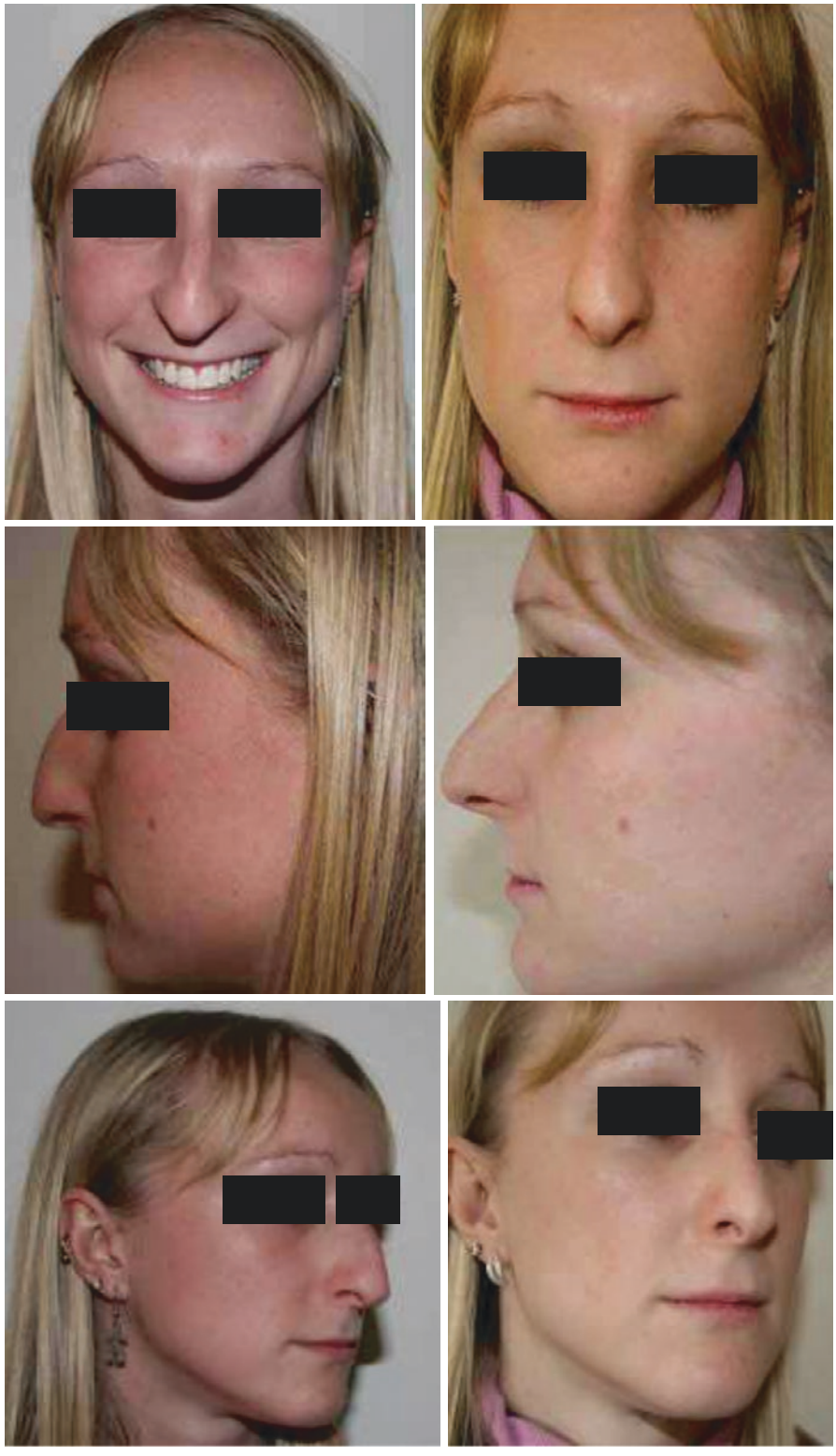

Fig. 19.1 Preoperative and postoperative photographs demonstrating improvement in aesthetic appearance

eral cartilages. Both these steps served to address the nasal tip ptosis. There was no need to use a cartilaginous nasal 'strut' between the medial crura of the lower lateral (alar) cartilages.

Postoperative care includes regular saline douching, analgesia, removal of nasal splints and nasal tip sutures at 7 days. The patient is then followed up at the end of 3, 6 and 12 months after surgery.

Summary and Author's Comments

Septorhinoplasty is a surgical procedure that requires very careful patient assessment and counselling. The success of the surgery depends on this and on having clear understanding of the operative strategies required to address the patient's complaints.

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Nasal Valve Surgery

20

Marios Stavrakas, Mihiar Atfeh, and Hisham S Khalil

20.1 Case Presentation

A 58-year-old man was referred to the Rhinology clinic after having a septoplasty, which failed to improve his nasal obstruction. On examination, there was no significant residual septal deviation, but on dynamic assessment, there was a marked collapse of the lateral nasal wall on inspiration. The patient consented for alar batten surgery and inferior turbinate reduction. It was made clear during the consenting process that his nose may look different, because of the insertion of the grafts. The patient had an uneventful recovery and was pleased with the outcome.

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Table 20.1 Anatomical boundaries of internal and external valve

Internal nasal valve (angle: 10–15°)	External nasal valve
Caudal border of upper lateral cartilages	Medial crurae of alar cartilages
Nasal septum	Caudal septum
Head of inferior turbinate	Alar rim
Nasal floor	Nasal sill

20.2 Background Knowledge

The nasal valve is an important anatomical complex, contributing to normal breathing and, consequently, to nasal obstruction (Table 20.1). Nasal valve dysfunction is responsible for up to 13% of chronic nasal obstruction and up to 95% for persistent nasal obstruction after septoplasty.

It is a commonly missed condition, so suspicion and detailed clinical examination are essential. Causes of nasal valve dysfunction can be divided into mucosal and structural, with the latter including trauma, idiopathic, iatrogenic (rhinoplasty-failure to reconstruct anatomical units) and ageing.

Static obstruction: an anatomically narrowed portion of the nasal valve (Poiseuille's law) Causes: trauma, previous rhinoplasty.

Dynamic obstruction: the collapse of the nasal sidewall during inspiration (Bernoulli principle) Causes: over projected septal cartilage, e.g., tension nose.

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External valve collapse is less frequent and is associated with previous nasal surgery, while internal nasal valve collapse has an anatomical background and may present with ageing.

20.3 Clinical Approach

20.3.1 History

This includes a history of prolonged nasal trauma or surgery e.g. rhinoplasty or tumour resection.

20.3.2 Examination

1. Inspection: Examine during inspiration and natural state. A pinched middle third may be associated with valve dysfunction. Inspect for previous rhinoplasty scars/incisions.
2. Palpation: Will give information on the strength of nasal cartilages, alar rim strength and nasal tip support. Evaluate skin thickness.
3. Internal inspection: Evaluate septum and potential septal deviation, inferior turbinate hypertrophy, lateral wall nasal collapse and valve narrowing. Nasendoscopy may also be useful.

The patient had left alar collapse on inspiration (Fig. 20.1).

The Cottle manoeuvre (Fig. 20.2) is a popular test when examining for nasal function dysfunction. Still, it is a non-specific test, in the sense that it improves nasal airflow even in patients without nasal valve dysfunction. The modified Cottle's manoeuvre has been proposed as an alternative, aiming to predict surgical outcomes. This is not supported by the literature, as Barrett et al. (2016) have identified only one study that has proven this.

Objective measures for nasal valve dysfunction include acoustic rhinometry, rhinomanometry, peak nasal inspiratory flow, CT imaging.

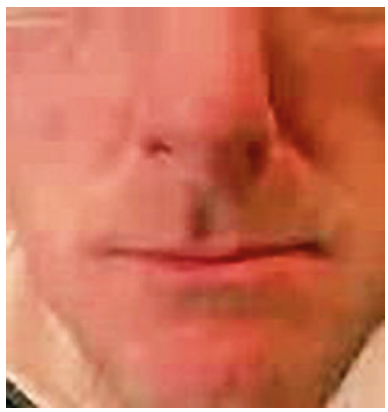

Fig. 20.1 Left alar collapse on inspiration

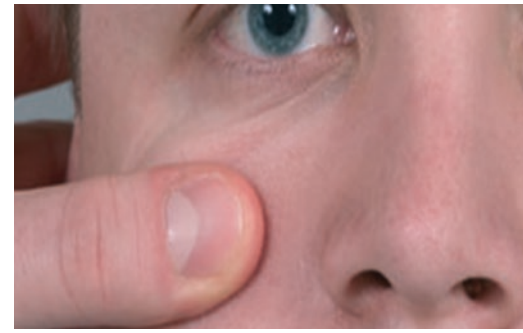

Fig. 20.2 The Cottle manoeuvre for the right nasal cavity

20.3.3 Treatment

The patient underwent bilateral alar battens surgery although his alar collapse was mainly on the left side. This is to achieve symmetry postoperatively and minimise the aesthetic impact of the surgery.

Alar battens surgery involves the placement of a cartilage strip (septal, auricular) in a subcutaneous pocket created at the level of the scroll area via an intercartilaginous incision (Figs. 20.3, 20.4). Suturing the graft in place with transcutaneous sutures is not necessary unless the pocket is too large. The intercartilaginous incisions are closed with absorbable suturing material and we tend to support the area with dissolvable nasal packing and external taping with Steri-Strips. Commonly, alar batten grafting is combined with septoplasty and/or inferior turbinate reduction. Potential complications include bleeding, infection, failure to improve symptoms, graft migration, widening of the mid-third of the nose. The last risk is something we explain to our patients very clearly, as inevitably, there will be an altera-

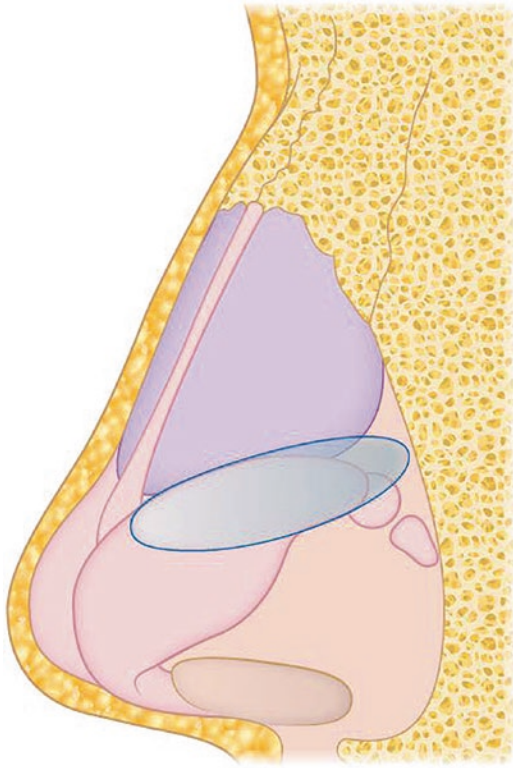

Fig. 20.3 Illustration depicting the placement of alar batten graft

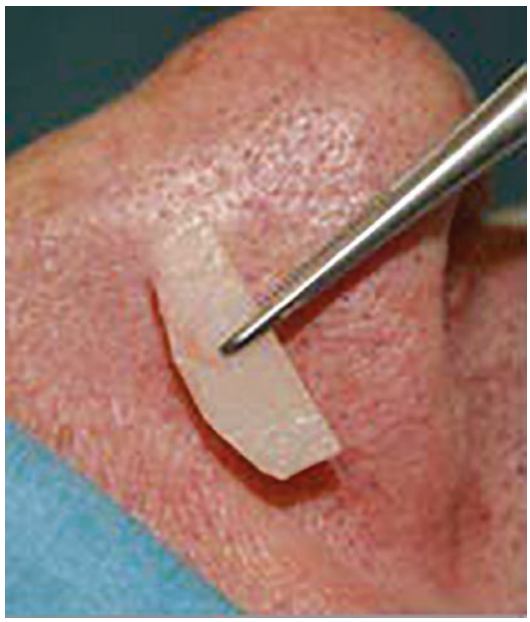

Fig. 20.4 Right alar batten

tion to the shape of the nose, which may not agree with one's aesthetic expectations.

- Various treatment methods of nasal valve dysfunction

1. Non-surgical
   - 'Breathe right' strips
   - Nasal splints
2. Surgical-internal nasal valve
   - Spreader grafts (INV-static)
   - Spreader flaps (INV-static)
   - Flaring sutures (INV-static/dynamic)
   - Butterfly graft (INV/ENV-static/dynamic)
   - Lateral nasal valve suspension (INV-static/dynamic)
3. Surgical-external nasal valve
   - Alar batten graft (INV/ENV-dynamic)
   - Alar rim graft (ENV-static/dynamic)
   - Lateral crura underlay strut graft (ENV-static/dynamic)
   - Lateral crura turn-in (ENV-static/dynamic)
4. Newer methods
   - Radiofrequency-induced thermotherapy (INV/ENV)
   - Upper lateral strut graft (INV)
   - Stairstep graft (ENV)
   - Nasal valve lift and nasal tip lift (INV/ENV)

Summary and Author's Comments

1. Nasal valve dysfunction is responsible for up to 13% of chronic nasal obstruction and up to 95% for persistent nasal obstruction after septoplasty.
2. We should assess the nasal valve carefully before deciding for the surgical treatment of nasal obstruction.
3. The treatment modality should be selected based on the patient's characteristics and the surgeon's expertise.

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Prominent Ears-Pinnaplasty

21

Marios Stavrakas

21.1 Case Presentation

A 6-year-old boy was seen in the clinic, accompanied by his parents. The ear deformity was evident, and the parents reported incidents of bullying. They did not use any non-invasive methods such as splinting and they are considering correction. The main anatomical abnormality was absent antihelix; therefore, we opted for a Mustarde technique. We advocate a detailed consenting process and teach-back technique for all our aesthetic interventions.

21.2 Background Knowledge

It is a commonly seen ear deformity, with a prevalence of 6% among the UK population.

Characteristics of the normal ear

1. Distance between the helical rim and mastoid → 17–21 mm
2. Auriculomastoid angle →20°–30° (Figs. 21.1, 21.2, 21.3)

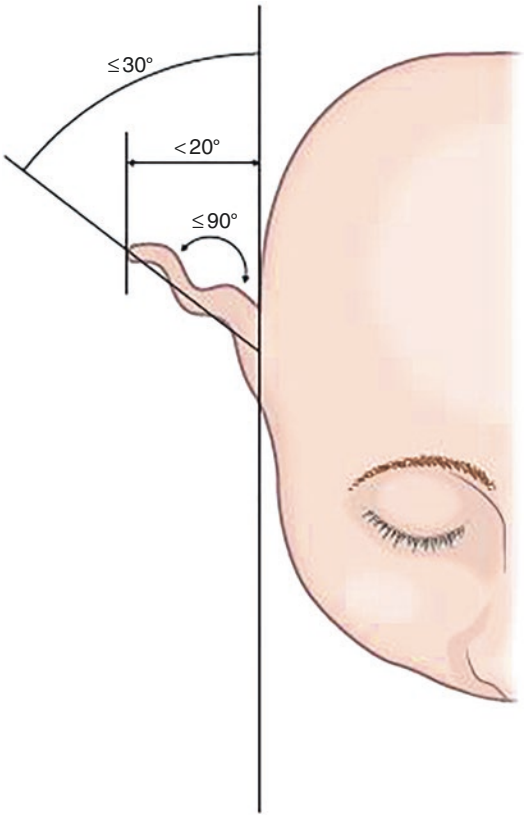

Fig. 21.1 Normal auriculomatoid angle

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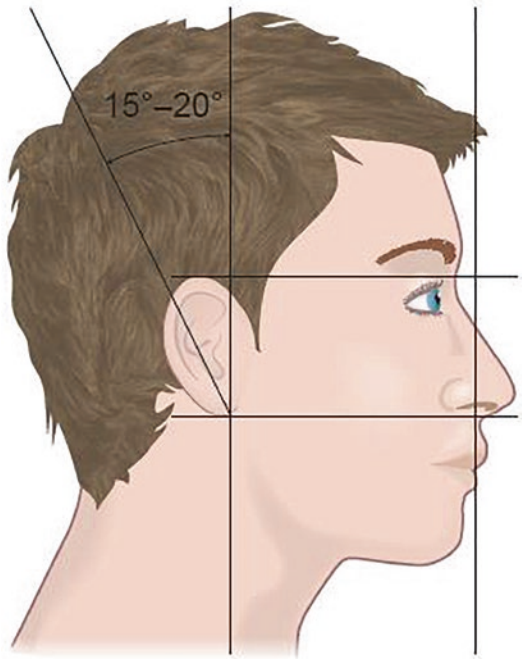

Fig. 21.2 Normal position of the pinna

Common anatomical abnormalities

1. Absent/under-developed antihelix
2. Conchal hypertrophy
3. Combination of the two
4. Prominent earlobes

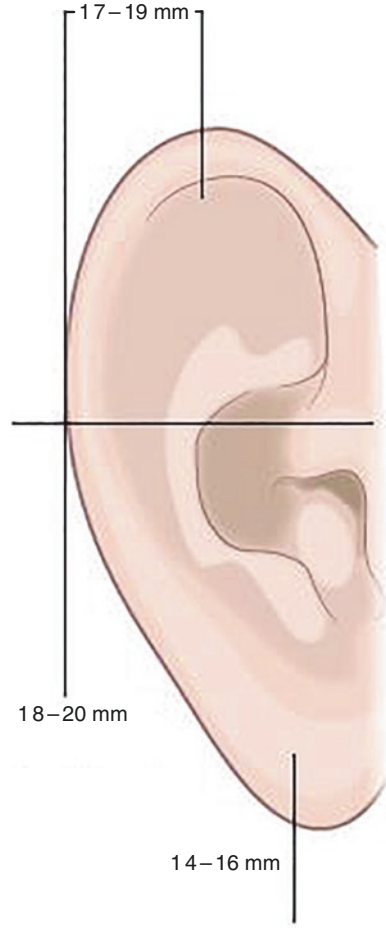

Fig. 21.3 Various distances between parts of the pinna and mastoid

21.3 Clinical Approach

21.3.1 Treatment

Pre-operative planning

1. Pre-operative photography
2. Measurement of projection of upper, middle and lower thirds of the ear

The cartilage of the newborn ear is incredibly pliable due to the influence of circulating maternal oestrogen. 'Ear buddies' have been used to improve the shape of the ears in the first weeks of life. Early detection and awareness can allow non-surgical correction of prominent ears and other deformities such as lop ear, Stahl's bar, rim kink and cup deformity. 'Ear well' system has been found to be equally effective in avoiding surgery in less severe abnormalities.

Techniques

1. Cartilage sparing
   - Mustarde
   - Furnas conchal setback
2. Cartilage scoring, e.g. Chongchet
3. Combination of the two
4. Earlobe setback

The authors prefer cartilage sparing techniques alone or in combination if indicated (Fig. 21.4). The surgical steps we follow are:

1. Pre-operative measurements

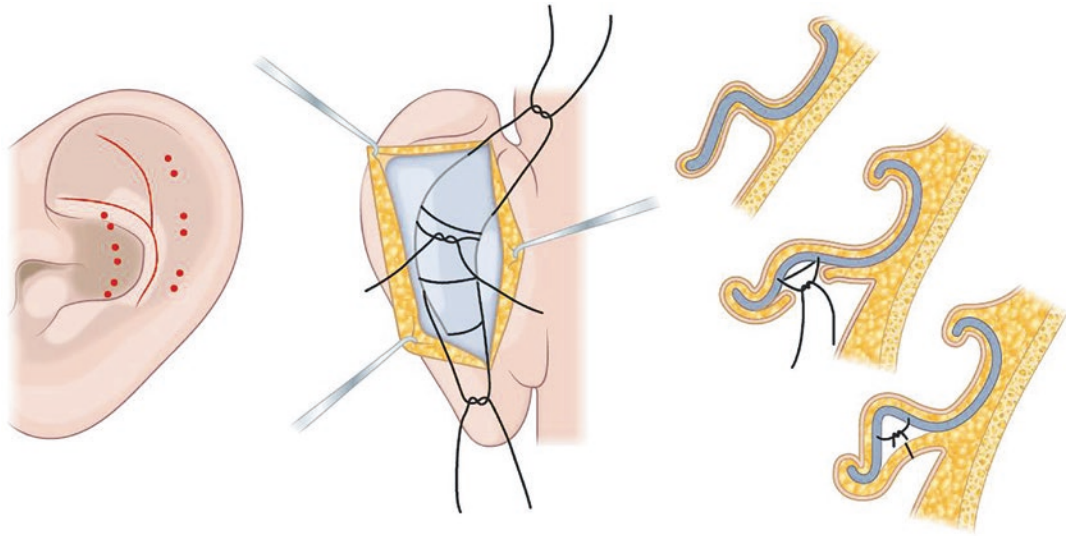

Fig. 21.4 Surgical steps-Mustarde cartilage sparing technique

2. Marking of the expected position of the antihelix with methylene blue (3 pairs of dots)
3. Infiltration with LA and incision on the posterior surface of the pinna (a wedge of skin excised)
4. Horizontal mattress sutures (undyed Silk, Mersilene)—leave sutures long and tighten at the end
5. If concha setback is required, sutures between the concha and the mastoid periosteum are placed
6. Close incision and record post-operative measurements
7. Head bandage

21.3.2 Complications

- Haematoma
- Infection/perichondritis
- Skin/wound healing problems
- Suture-related problems
- Pain and itching
- Poor aesthetic outcome and need for revision
- Asymmetry/telephone ears

The timing of surgery varies and depends on the surgeon's experience and practice as long as the patient's and parent's preferences. Some surgeons believe that early correction is advisable, as 85% of the pinna is formed by the age of 3 years. Others prefer to operate at an older age when the child can participate in the consultation and express the desire for correction. Our standard practice is to operate before the child goes to school, as this may have a psychological impact when socialising with other children.

21.3.3 Follow Up

Post-operative care

- Pressure dressing is applied, usually for 1 week.
- Following that, the patient is advised to use an elastic headband, especially during sleep (3–6 weeks).

According to Sadhra et al. (2017), cartilage sparing techniques have a higher rate of recurrence and revision surgery. Cartilage scoring techniques have a higher rate of skin/wound healing problems.

The significance of psychological distress caused by prominent ears is widely known, and it is clear that surgical correction can alleviate psychological impact.

Summary and Author's Comments

1. Detailed discussion during the pre-op visit, involving teach-bach techniques is advisable.
2. Non-invasive methods should be considered early after birth.
3. The timing of the surgery and the surgical method need to be discussed.

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Reconstruction of the Nose

22

Marco Malahias and Platon Trigkatzis

22.1 Embryology & Anatomy & Aesthetic Relationship

In utero, the nose is developed in the foetus from the third week gestation onwards; during weeks three and four the frontonasal prominence of the forebrain differentiates into the nasal and olfactory placodes that further develop into the medial and lateral processes. The medial nasal process will form the premaxilla, philtrum, columella and the nasal tip.

The lateral nasal process will form the ala nasi. These different parts will later on represent the various aesthetic subunits of the nose. During weeks five and six the nasal processes fuse, after enlarging and migrating centrally.

- The arterial supply is derived from the *Angular Artery* (terminal branch from the Facial Artery) which supplies the caudal end of the nose and the lateral walls; *Superior Labial Artery* supplies the nasal sill, columella and nasal septum; the *Infraorbital Artery* (terminal branch of the maxillary Artery) supplies the dorsum of the nose and lateral nasal walls. The venous drainage mirrors the arterial blood flow.
- The nerve supply includes *motor* supply from the Facial Nerve to Procerus, Nasalis and the Depressor Septi Nasi muscles and *sensory* supply via the Trigeminal nerve. The Ophthalmic division (V1) of Trigeminal nerve innervates the rhinion, radix, dorsum of nose and tip of nose; the Maxillary division (V2) of Trigeminal nerve supplies the columella, lateral vestibule and lateral lower half of the nose.

The nose is formed of nine aesthetic subunits, including one of: dorsum, tip, columella and two of sidewalls, alae, soft triangles.

Numerous types of reconstruction following resection have been described such as the Banner Flap, Bilobed Flap and Glabellar flap, to name but a few.

The author presents the following options of reconstruction after resection:

(1) Nasolabial Flap with ipsilateral Conchal cartilage graft.

- The lesion to be excised is marked
- A safety margin between 3–6 mm is marked around the lesion
- The lesion is excised with the underlying cartilage, as the lesion invades it
- Ipsilateral conchal bowl cartilage is harvested via an anterior auricular approach
- Cartilage is placed into the defect
- An ipsilateral nasolabial flap is raised and the tip of the flap is defatted and rounded off
- Wound closure is achieved with Prolene 5/0

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https://doi.org/10.1007/978-3-030-66865-5_22

- Sutures are removed at 10 days
- NB when consenting remember to include ipsilateral nasal occlusion until post-operative oedema settles

Images for example case of Nasolabial Flap with ipsilateral Conchal cartilage graft (Figs. 22.1, 22.2, 22.3, 22.4).

(2) Glabellar Flap.

- The lesion to be excised is marked
- A safety margin between 3–6 mm is marked around the lesion
- Skin lesion is resected
- The glabellar skin between frown lines is elevated as a full thickness triangle, leaving the base attached
- The flap is rotated into the defect and the triangular flap is customised to the defect
- Monocryl 4/0 anchoring suture is placed
- Interrupted Prolene 5/0 sutures are placed and removed at 10 days

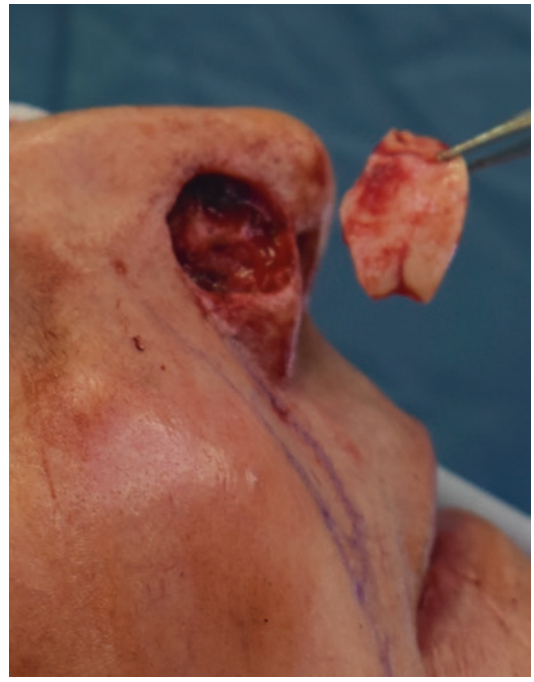

Fig. 22.2 Free choncal cartilage graft

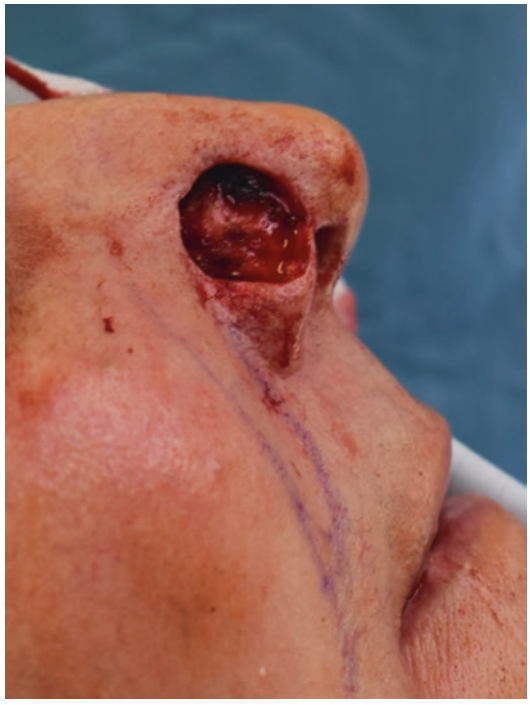

Fig. 22.1 Alar defect, including resected nasal cartilage

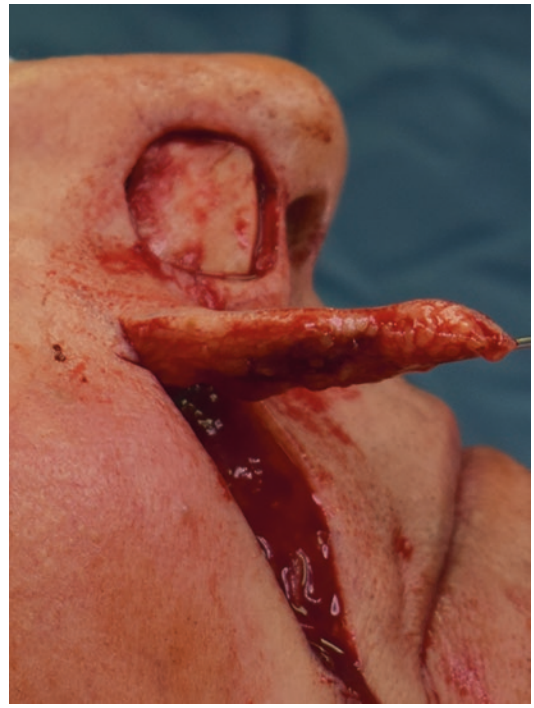

Fig. 22.3 Nasolabial flap raised

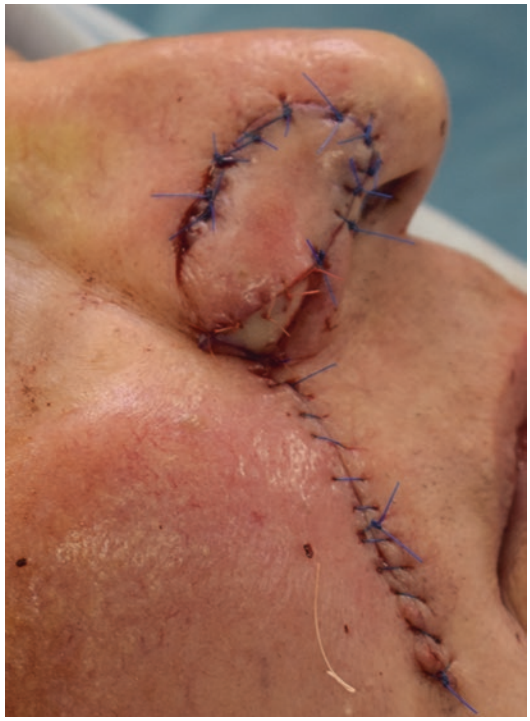

Fig. 22.4 Flap and donor site closure

Images for example case of Glabellar Flap (Figs. 22.5, 22.6, 22.7, 22.8).

(3) Rieger Flap.

- The lesion to be excised is marked
- A safety margin between 3–6 mm is marked around the lesion
- Skin lesion is resected down to bone/cartilage
- The entire dorsum of the nose and the glabellar skin is degloved in a the plane superficial to the perichondrium
- Undermining of the skin surrounding the flap, will facilitate ease of rotation of the flap into the defect
- Monocryl 4/0 anchoring sutures are placed in the skin edges
- Interrupted or continuous Prolene 5/0 sutures are placed and removed at 10 days

Images for example case of Rieger Flap (Figs. 22.9, 22.10, 22.11, 22.12, 22.13).

(4) Hatchet Flap.

- The lesion to be excised is marked

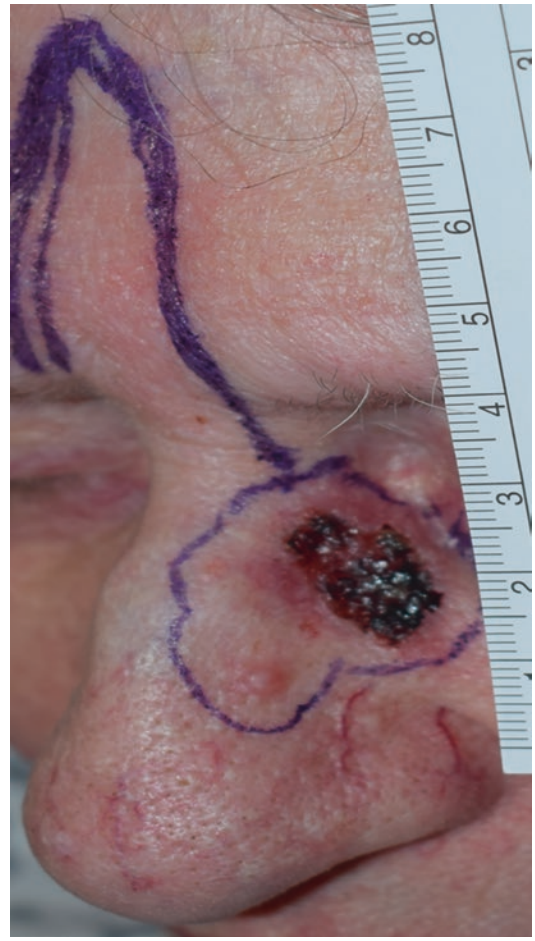

Fig. 22.5 Lesion and Flap marked

- A safety margin between 3–6 mm is marked around the lesion
- Skin lesion is resected
- The Hatchet flap on the side of the nose is elevated, leaving the base attached to the Levator Labii Sperioris
- Alaeque Nasi Muscle and the Transverse Nasalis Muscle, thus protecting the nasal branches from the angular artery
- The flap is advanced into the defect and the proximal part of the donor site is closed in a V–Y pattern
- Monocryl 4/0 anchoring suture is placed
- Interrupted Prolene 5/0 sutures are placed and removed at 10 days

Images for example case of Hatchet Flap (Figs. 22.14, 22.15, 22.16, 22.17, 22.18).

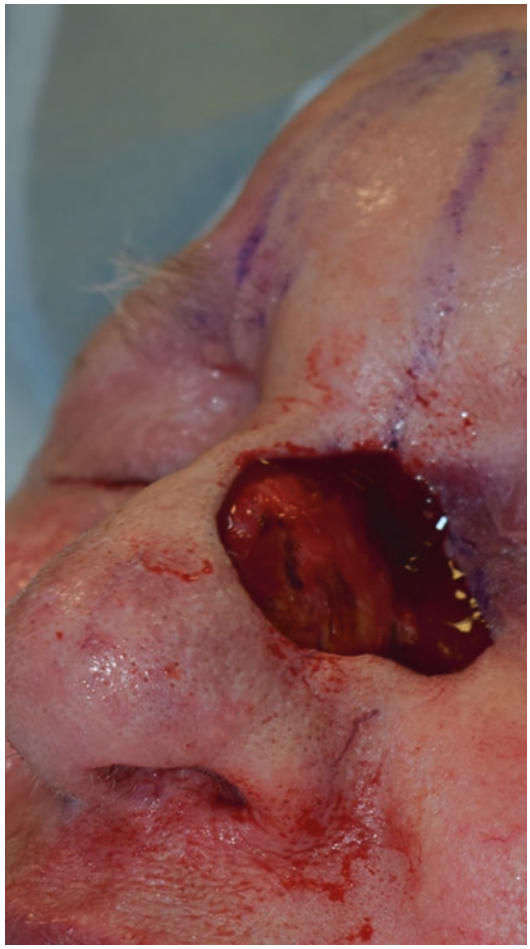

Fig. 22.6 Defect to left side of nose

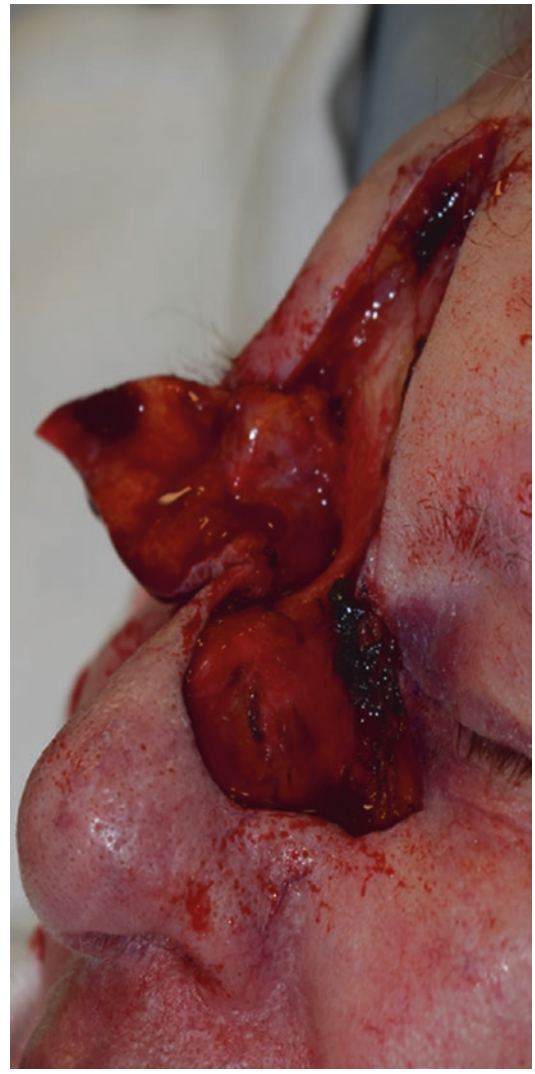

Fig. 22.7 Flap mobilised

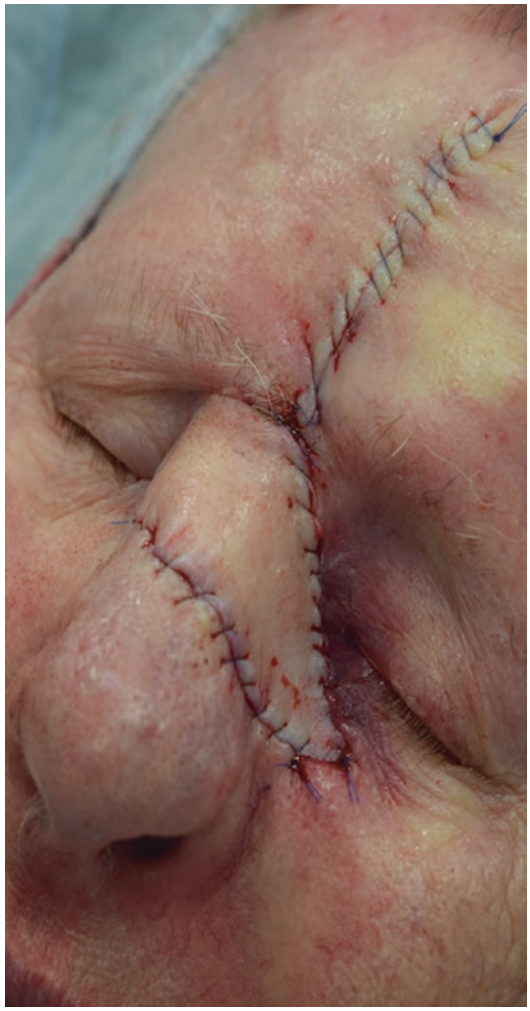

Fig. 22.8 Flap and donor site closure in two layers

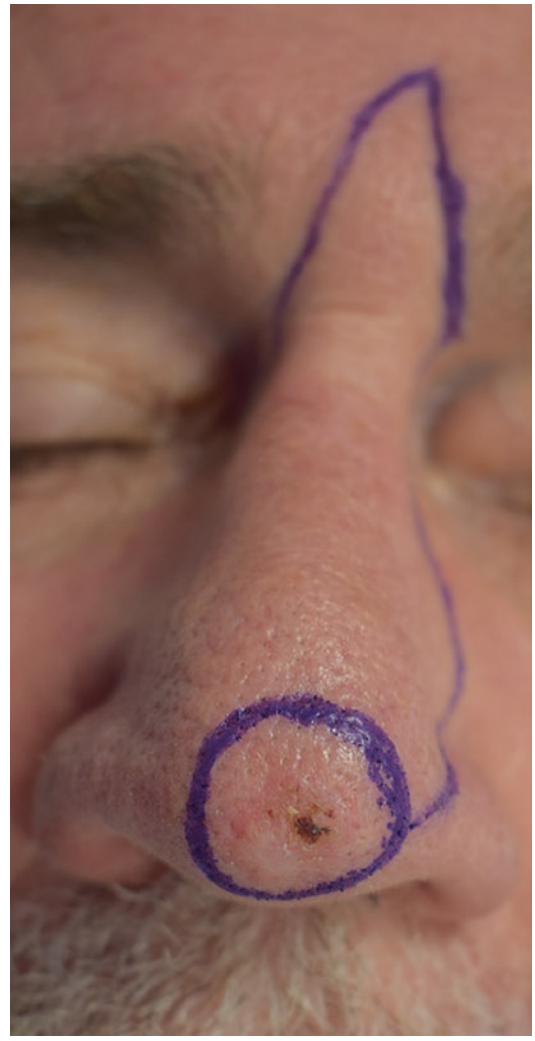

Fig. 22.9 Lesion and flap outlined

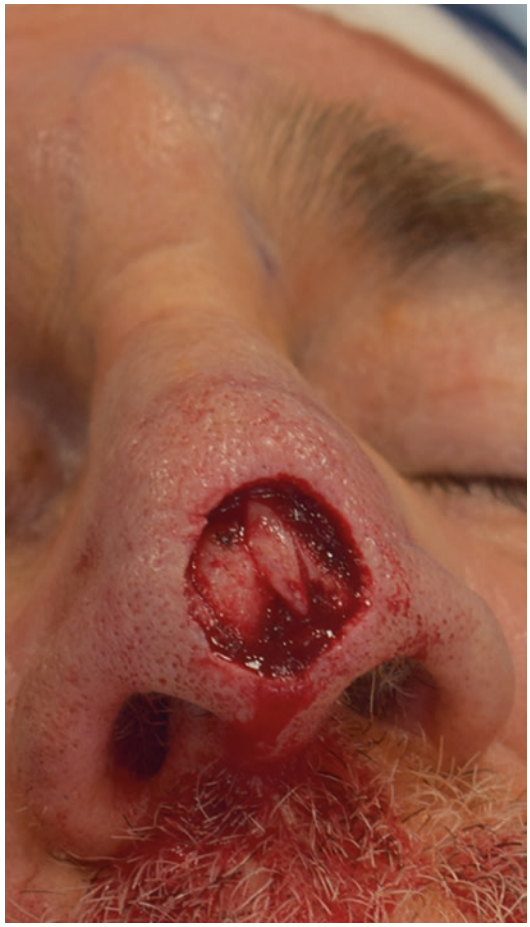

Fig. 22.10 Lesion excised

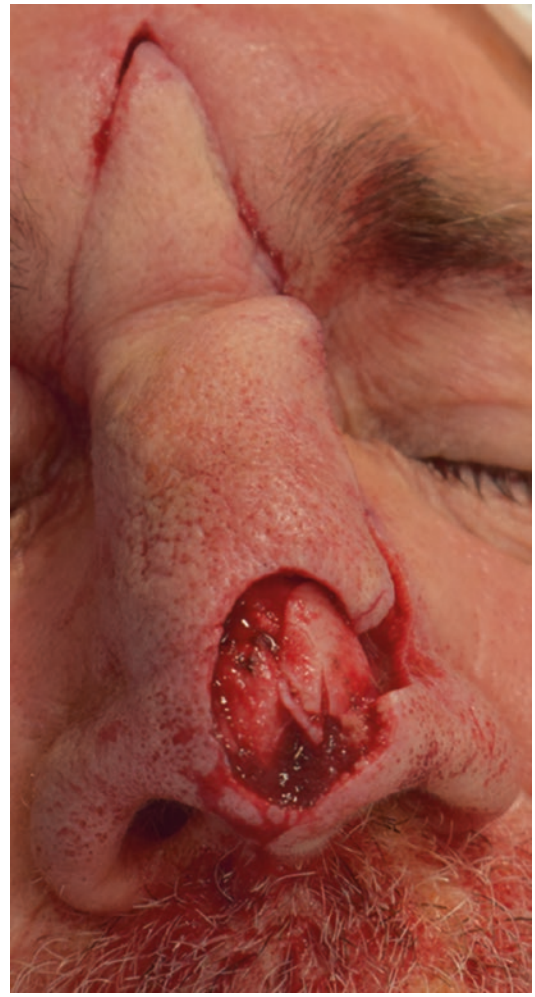

Fig. 22.11 Flap elevated

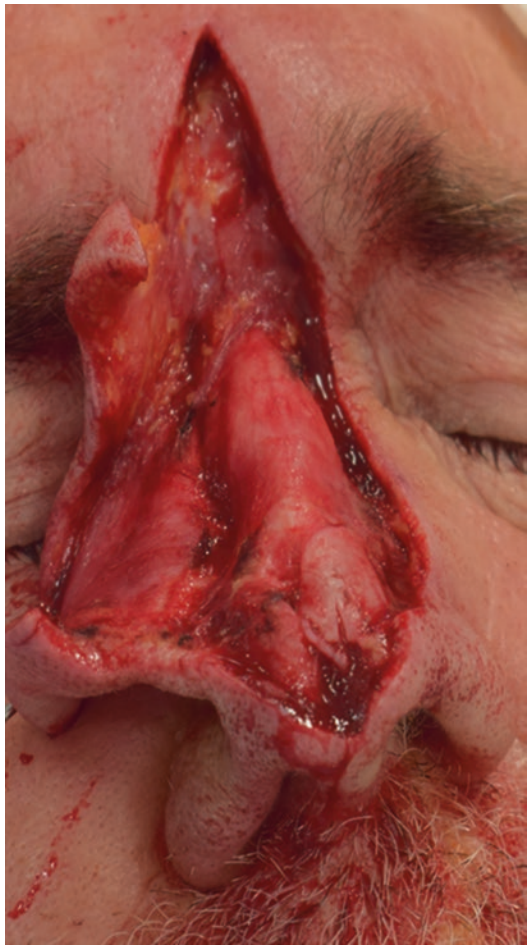

Fig. 22.12 Undermining of flap to increase mobility

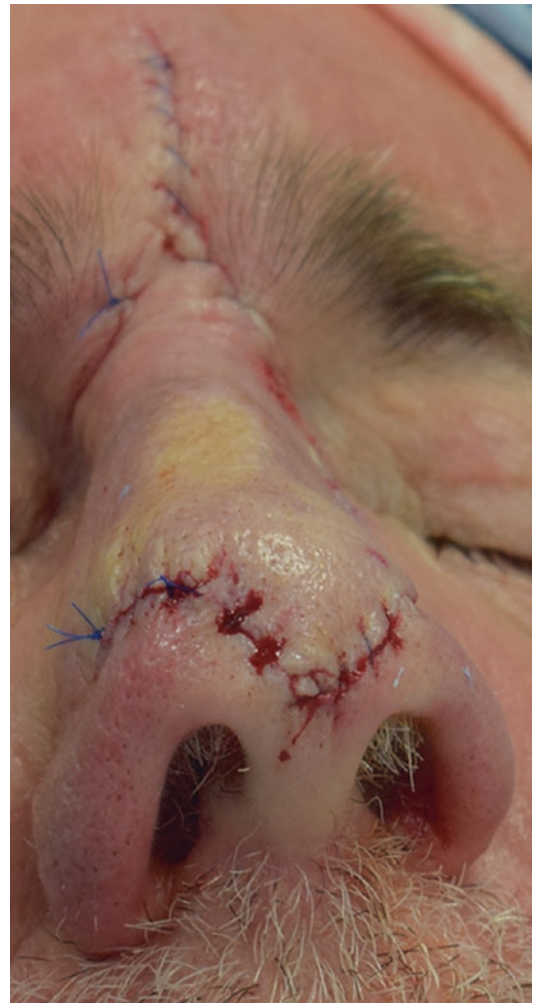

Fig. 22.13 Closure of flap and donor site in two layers

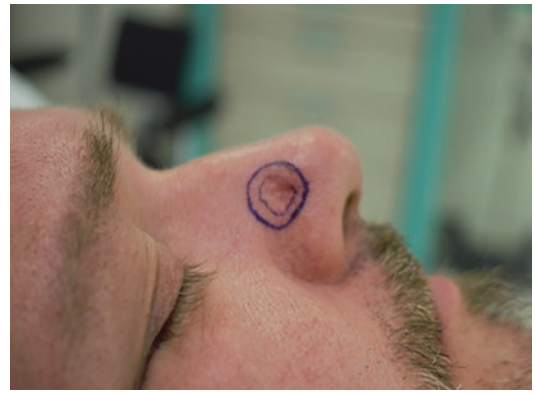

Fig. 22.14 Lesion with margin marked

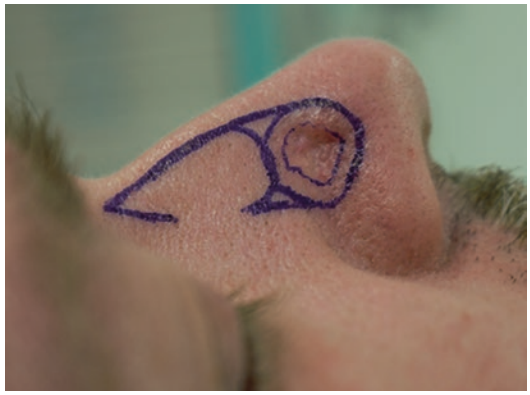

Fig. 22.15 Flap designed

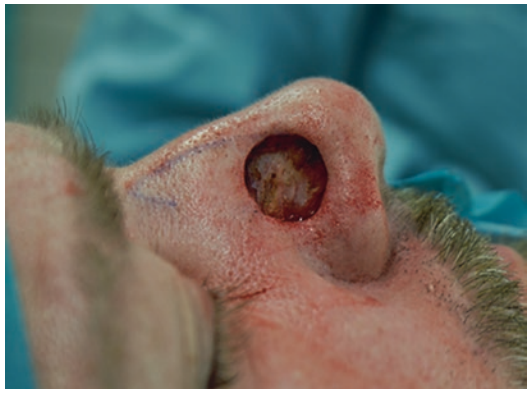

Fig. 22.16 Lesion excised

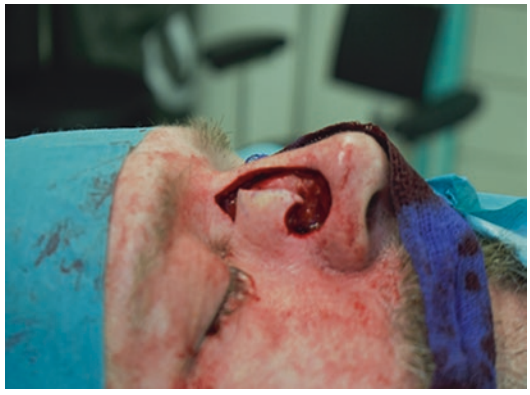

Fig. 22.17 Flap elevated and undermined

Fig. 22.18 Two weeks after wound closure

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Reconstruction of the Ear

23

Marco Malahias and Platon Trigkatzis

23.1 Embryology & Anatomy & Aesthetic Relationship

In utero, the ear is developed in the foetus during the sixth week gestation and is derived from the first branchial arch (known as the "Mandibular Arch") and second branchial arch (known as the "Hyoid Arch"), by forming six buds of mesenchyme between them, commonly known as the *hillocks of Hiss*. The 1st–3rd buds/hillocks are derived from the 1st branchial arch and the 4th–6th buds/hillocks from 2nd branchial arch.

The 1st, 2nd and 3rd buds/hillocks (of the 1st branchial arch) form the tragus, root of helix and superior helix respectively, whilst the 4th, 5th and 6th buds/hillocks (of the 2nd branchial arch) form the antihelix, antitragus and ear lobule respectively.

This explains why the blood supply, nerve supply and lymphatic drainage differ from the various parts of the ear!

1. The arterial supply is derived from the Posterior Auricular Artery (dominant), Superficial Temporal Artery, and Occipital Artery. The venous drainage however is into the Posterior Auricular Vein, Superficial Temporal Vein and Retromandibular Vein.
2. The nerve supply includes *Great Auricular Nerve* (C2, C3) to the lower half of the ear; *Lesser Occipital Nerve* (C2, C3) to the superior cranial part of the ear; *Auriculotemporal nerve* (V3) to the superior lateral part of the ear as well as anterior wall of external auditory; *Arnold's* branch of Vagus (Cranial Nerve X, including contributions from Cranial Nerve VII (Facial) and Cranial Nerve IX (Glossopharyngeal)), which innervates the posterior wall of external auditory meatus and conchal bowl.
3. The Lymphatic drainage corresponds to the six hillocks of Hiss and is subdivided as follows: tragus—1st hillock, root of helix—2nd hillock and superior helix—3rd hillock (anterior hillocks) all drain into the parotid lymph nodes; the antihelix—4th hillock, antitragus—5th hillock and lobule—6th hillock (posterior hillocks) drain into the cervical lymph nodes.

The adult ear measures between 5.5 and 6.5 cm, which is corresponding to the distance between the angle of the mouth and lateral canthus of the eye; its width is approximately 55% of its height. The external ear protrudes 1–2 cm laterally from the scalp and has an average incline ranging between 21–25 degrees away from the scalp.

Numerous types of reconstruction following resection have been described such as Antia Buch

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https://doi.org/10.1007/978-3-030-66865-5_23

Flap, Tanzer's procedure, the Flip Flop flap as well as the Dieffenbach's intervention, to name but a few.

The author presents the following options of reconstruction after resection:

(1) Wedge resection.

1. The lesion to be excised is marked
2. A safety margin between 3–6 mm is marked around the lesion
3. The circle is converted in a triangle tangentially to the circumference of the circle
4. Full thickness wedge is resected
5. Monocryl 4/0 anchoring sutures are placed in the substance of the ear
6. Interrupted Prolene 5/0 sutures are placed and removed at 10 days
7. NB. During consenting it is important to mention permanent numbness, skin necrosis, dehiscence and cauliflower deformity, in addition to the standard risks and complications routinely mentioned for any other skin surgery

Images for example case of Wedge resection (Figs. 23.1, 23.2, 23.3, 23.4).

(2) Reverse Antia Buch flap.

8. The lesion to be excised is marked
9. A safety margin between 3-6 mm is marked around the lesion
10. Full thickness rectangle is resected
11. The ear is split longitudinally along the Antihelical fold, into an anterior dermocartilaginous and a posterior cutaneous flap
12. A partial thickness ellipse of skin is resected from the ear lobe in line with the longitudinal split
13. The caudal part of the ear is mobilised in a cephalic direction, closing the defect, without any tension
14. Single Monocryl 4/0 anchoring suture is placed in the substance of the ear
15. Interrupted Prolene 5/0 sutures are placed and removed at 10 days

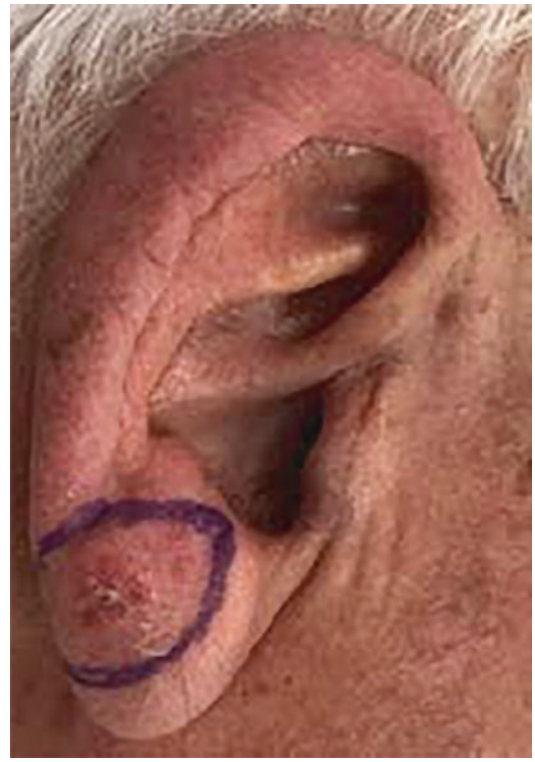

Fig. 23.1 Lesion marked

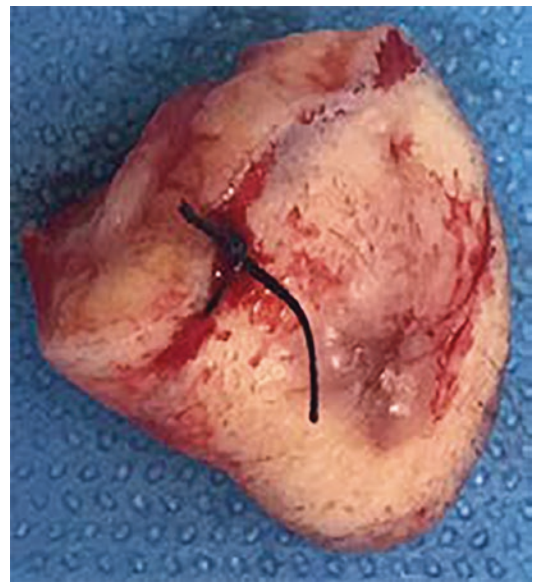

Fig. 23.2 Wedge excised

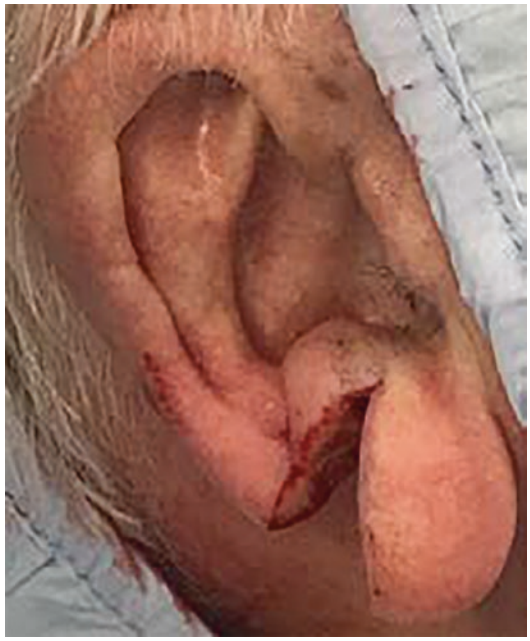

Fig. 23.3 Full thickness wedge

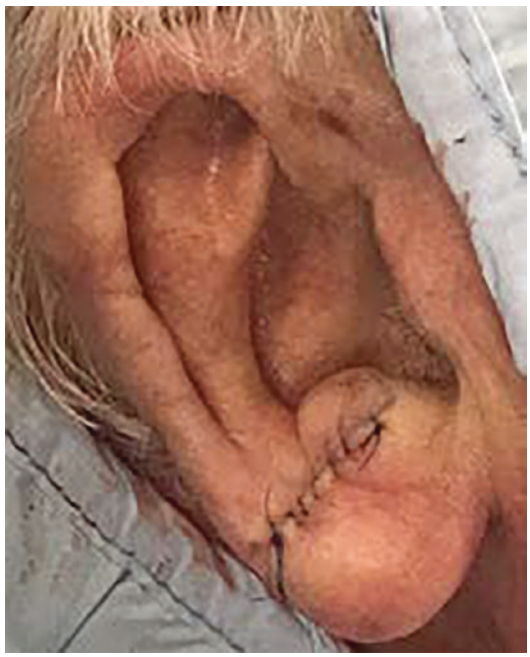

Fig. 23.4 Two layered closure

16. Jelonet® 10 × 10 cm piece is moulded into the Antihelical fold and a bandage is applied, so as to prevent haematoma formation

17. NB. During consenting it is important to mention permanent numbness, skin necrosis, dehiscence and cauliflower deformity, in addition to the standard risks and complications routinely mentioned for any other skin surgery

Images for example case of Reverse Antia Buch (Figs. 23.5, 23.6, 23.7, 23.8).

(3) Rotation flap.

18. The lesion to be excised is marked

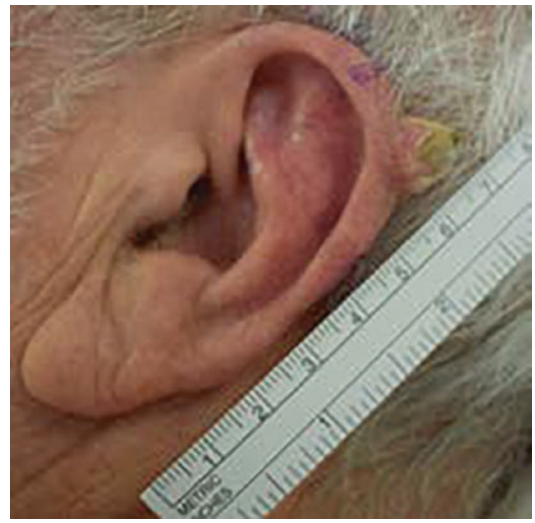

Fig. 23.5 Lesion to superior pole of left ear

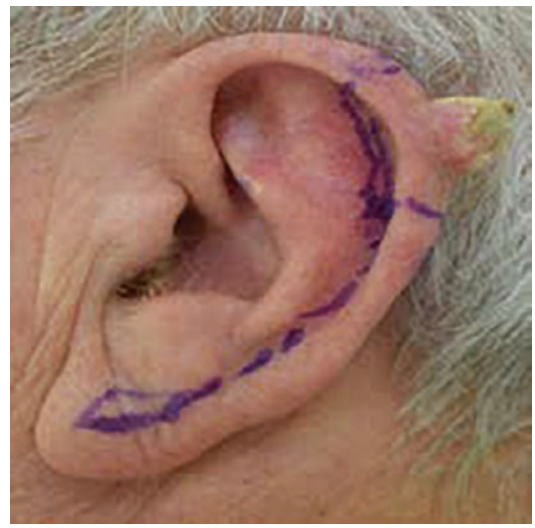

Fig. 23.6 Markings of resection and skin incision

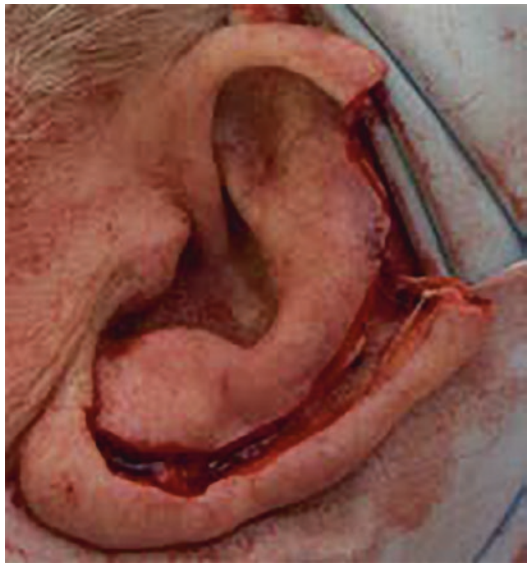

Fig. 23.7 Lesion resected, triangle from ear lobe removed and ear split

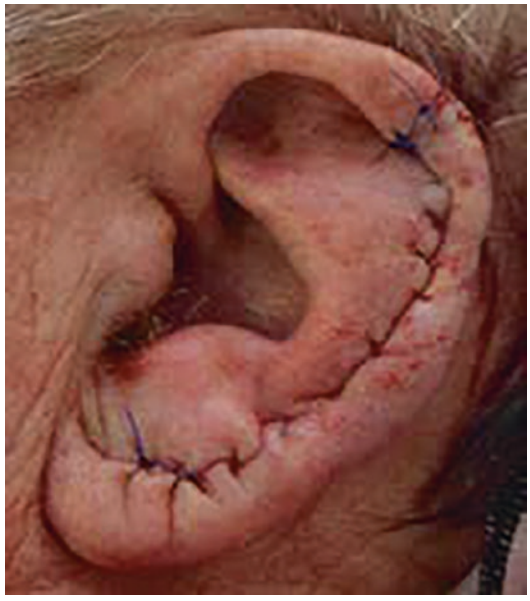

Fig. 23.8 Wound closure in two layers

19. A safety margin between 3–6 mm is marked around the lesion
20. Skin lesion with the involved, underlying cartilage is resected
21. A full thickness skin flap is raised; the curvature of the free edge of the flap should be in line with the cephalic edge of the defect
22. Undermining of the skin surrounding the flap will facilitate ease of rotation of the flap and direct closure of the donor site
23. Monocryl 4/0 anchoring sutures are placed in the skin edges of the donor and recipient sites
24. Interrupted or continuous Prolene 5/0 sutures are placed and removed at 10 days
25. Jelonet® 10 × 10 cm piece is moulded into the Antihelical fold and a bandage is applied, so as to prevent haematoma formation
26. NB. During consenting it is important to mention permanent numbness, skin necrosis, dehiscence and cauliflower deformity, in addition to the standard risks and complications routinely mentioned for any other skin surgery

Images for example case of Rotation flap (Figs. 23.9, 23.10, 23.11, 23.12, 23.13).

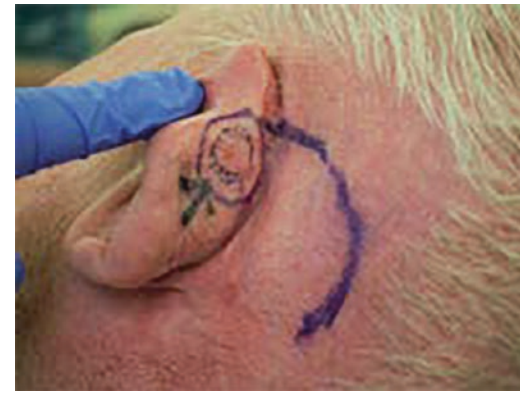

Fig. 23.9 Lesion to posterior aspect of left ear and rotation flap outline

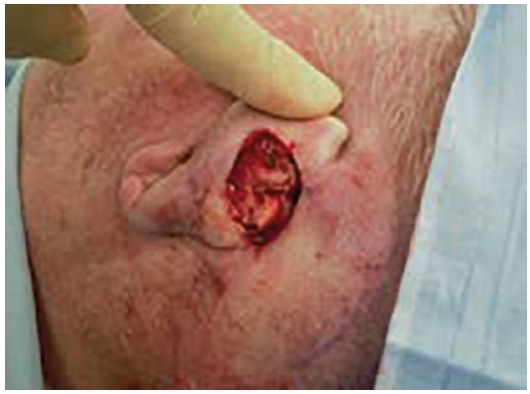

Fig. 23.10 Skin & cartilage resected

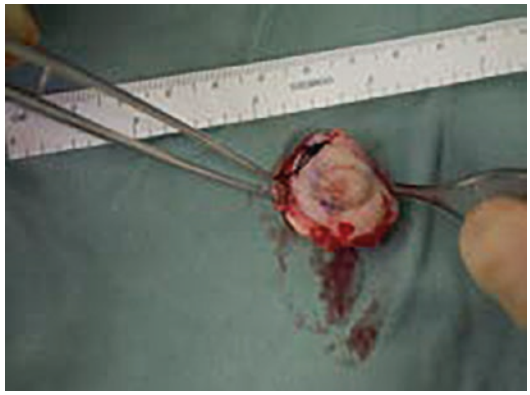

Fig. 23.11 Resected lesion

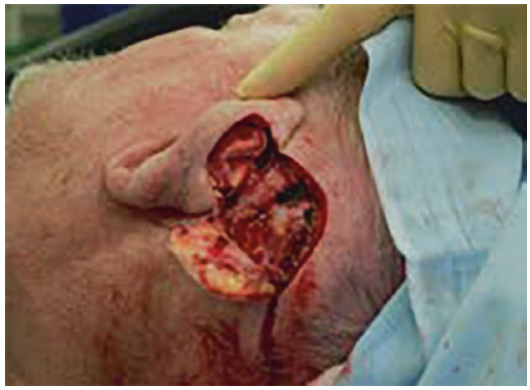

Fig. 23.12 Mastoid skin flap elevated

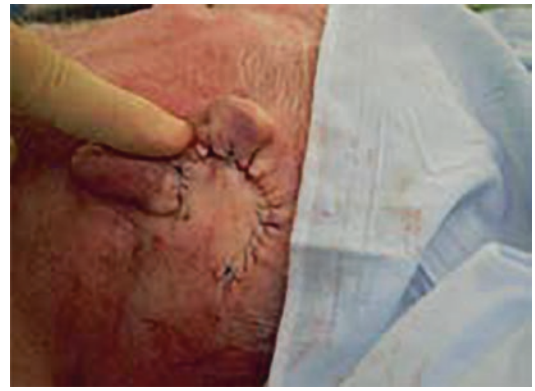

Fig. 23.13 Skin closure

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Reconstruction of the Lip

24

Marco Malahias and Platon Trigkatzis

24.1 Embryology & Anatomy & Aesthetic Relationship

In utero, the lip is developed in the foetus during the fourth and seventh week gestation and is formed from five facial *primordia* including the frontonasal prominence, bilateral maxillary prominences and bilateral mandibular prominences.

Failure of the medial nasal process to connect to the maxillary prominences will result in a cleft lip.

The arterial supply is derived from the bilateral Superior and Inferior Labial arteries, branches of the Facial artery artery. The venous drainage into the corresponding Superior and Inferior Labial Veins, that drain in the corresponding Facial veins.

The nerve supply includes *Trigeminal nerve Cr V* (sensory) and the *Facial Nerve Cr VII* (motor).

The Lymphatic drainage is into the Submental and Submandibular cervical lymph nodes.

The layers of the lip include: Skin, subcutaneous fat, muscles (intrinsic: Orbicularis Oris and extrinsic: Buccinator, Depressor Anguli Oris, Depressor Labii Inferioris, Levator Anguli Oris, Levator Labii Superioris, Levator Labii Superioris Alaeque Nasi, Mentalis, Platysma, Risorius, Zygomaticus Major and Zygomaticus Minor) and mucosa.

External landmarks include the Commisure, Cupid's bow, Philtral columns (ridges formed by the decussating Orbicularis Oris), Philtral groove, Tubercle, White roll (ridge formed by Orbicularis Oris) and the Red line (wet–dry vermilion line).

The aesthetic subunits of the lip include one medial (philtral) and two lateral subunits forming the upper lip, and the lower lip, which is considered a single unit.

Numerous types of reconstruction following resection have been described such as Abbe Lip Switch, Estlander procedure.

As well as Karapandzic flap intervention, to name but a few.

The author presents the following options of reconstruction, after resection:

(1) Wedge resection

1. The lesion to be excised is marked with a safety margin between 3-6 mm is marked around the lesion
2. Full thickness wedge is resected, extending the incision along the lateral border of the mental prominence (in large cases)
3. Monocryl 4/0 anchoring sutures, placed in the substance of the lip to approximate the divided Orbicularis Oris

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4. Interrupted Prolene 5/0 sutures are placed on the skin side and removed at 10 days
5. Interrupted Vicryl Rapide 4/0 sutures are placed on the mucosa and Vermilion

Images for example case of Wedge resection (Figs. 24.1, 24.2, 24.3).

(2) Estlander flap.

6. Suitable for large lateral lip defects
7. The lesion to be excised is marked with a safety margin between 3–6 mm, marked around the lesion

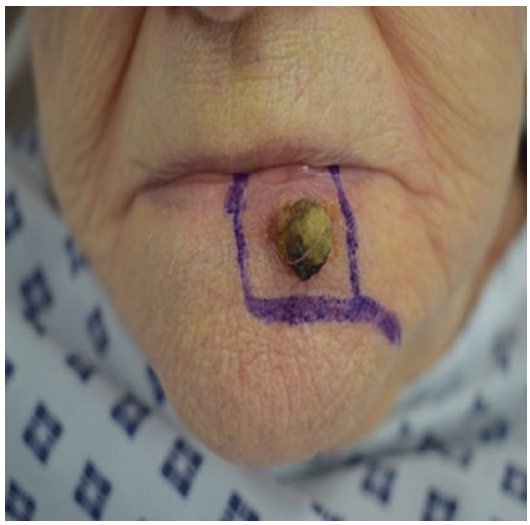

Fig. 24.1 Lesion including skin incision margins

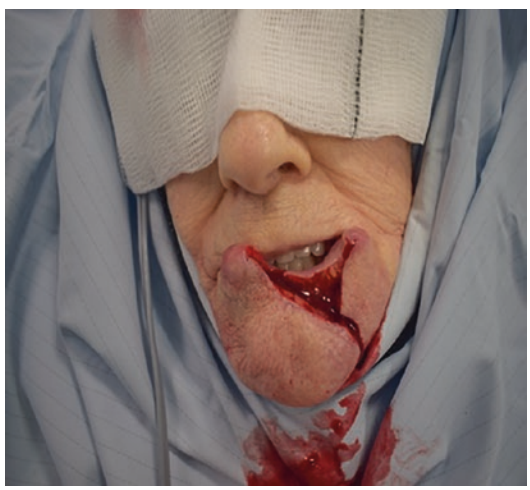

Fig. 24.2 Full thickness wedge excision

Fig. 24.3 Direct closure in three layers

8. Full thickness triangular wedge is resected
9. A full thickness Triangle from the opposing lip is dissected, preserving the Labial Artery which runs superficially in the substance of the lip
10. Hilton technique allows sceletonisation of the labial vessels without damaging them
11. Monocryl 4/0 anchoring sutures are placed in the substance of the lip
12. Interrupted Prolene 5/0 sutures are placed on the skin side and removed at 10 days
13. Interrupted Vicryl Rapide 4/0 sutures are placed on the mucosa and Vermilion
14. NB. During consenting it is important to mention that the angle of the mouth may be rounded, requiring commisuroplasty after three months

Images for example case of Estlander Flap (Figs. 24.4, 24.5, 24.6, 24.7, 24.8, 24.9).

(3) Modified Karapandzic flap.

15. This is suitable for central (philtral/medial) defects of the upper lip
16. The lesion to be excised is marked with a safety margin between 3–6 mm is marked around the lesion
17. Skin lesion with full thickness lip is resected
18. A partial thickness Burrow's triangle is resected from either side of the nose to facilitate skin approximation and closure

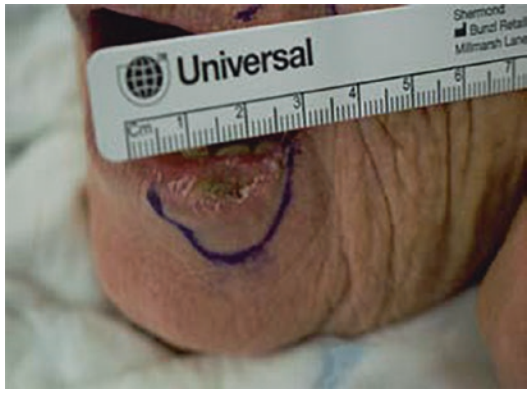

Fig. 24.4 Lesion marked

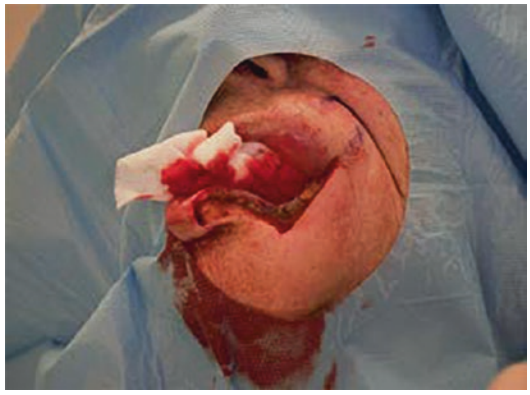

Fig. 24.5 Full thickness excision

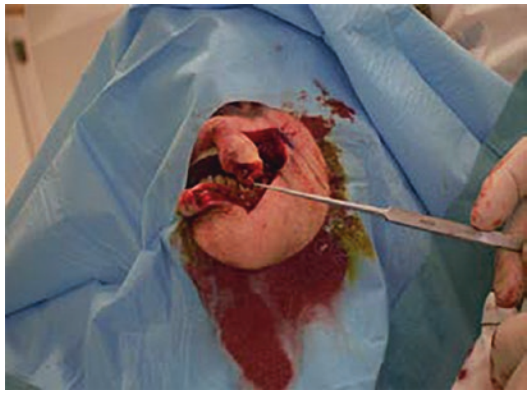

Fig. 24.6 Flap from upper lip prepared

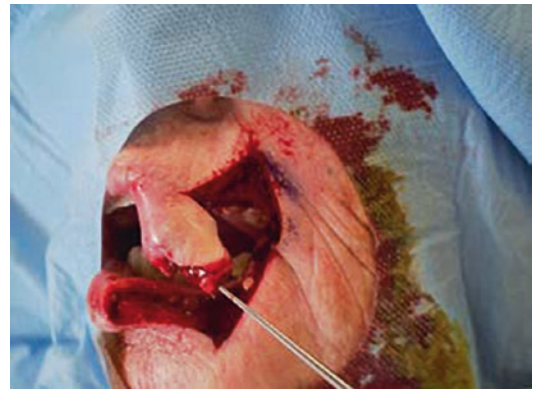

Fig. 24.7 Flap turned down

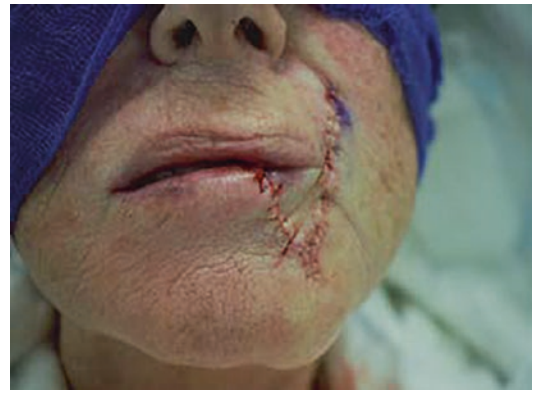

Fig. 24.8 Closure of wound and donor site

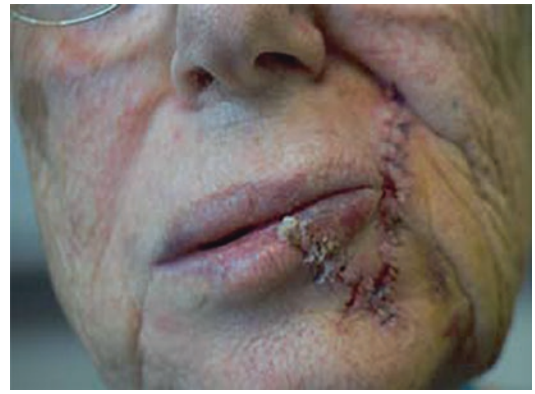

Fig. 24.9 Three days after operation

19. Undermining of the skin surrounding the flap will facilitate ease of rotation of the flap and direct closure of the donor site
20. Monocryl 4/0 anchoring sutures are placed in the skin edges of the donor and recipient sites
21. Interrupted or continuous Prolene 5/0 sutures are placed and removed at 10 days
22. Interrupted Vicryl Rapide 4/0 sutures are placed on the mucosa and Vermilion

Images for Modified Karapandzic flap with Burrow's triangle on either side of nose (Figs. 24.10, 24.11, 24.12, 24.13, 24.14).

(4) Labio-mandibular flap

23. The lesion to be excised is marked with a safety margin between 3–6 mm is marked around the lesion
24. Partial thickness wedge is resected
25. Labio-mandibular flap is raised in supra muscular plane, with the skin incision placed in the caudal end of nasolabial crease
26. Single Monocryl 4/0 anchoring suture is placed to secure the flap into the wound

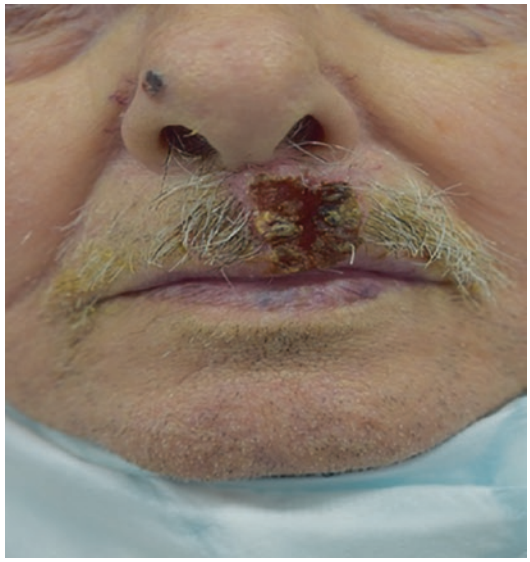

Fig. 24.10 Central upper lip lesion

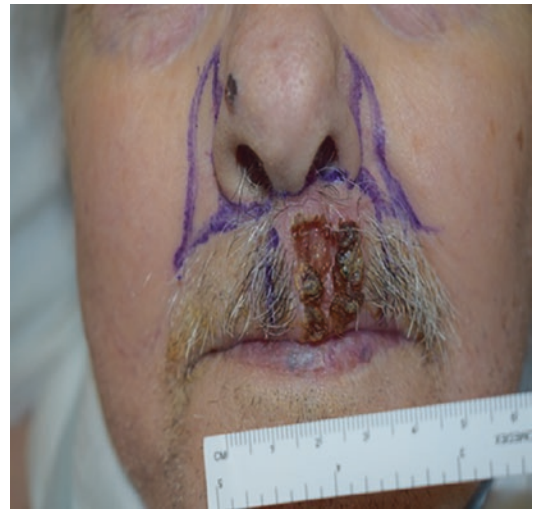

Fig. 24.11 Burrows triangles to sides of nose

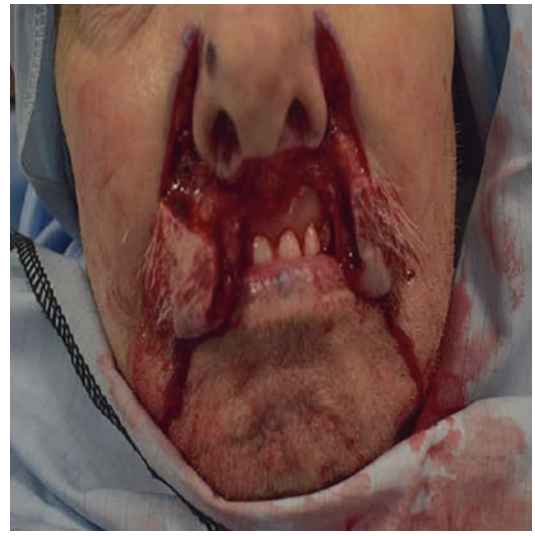

Fig. 24.12 Full thickness central excision, with resection

27. Interrupted Prolene 5/0 sutures are placed on the skin side and removed at 10 days

Images for example case of Labio-mandibular resection (Figs. 24.15, 24.16, 24.17, 24.18).

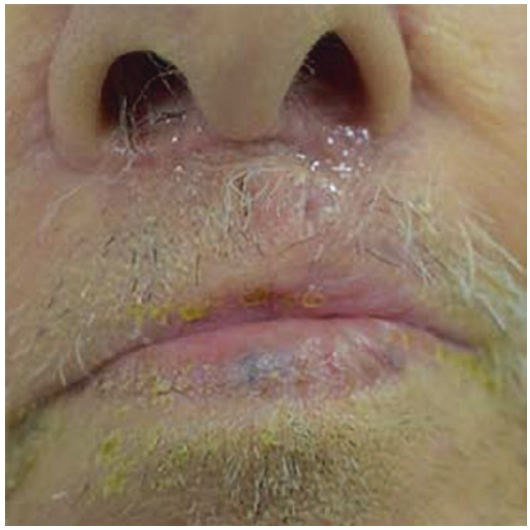

Fig. 24.13 Closed mouth 6 weeks post op

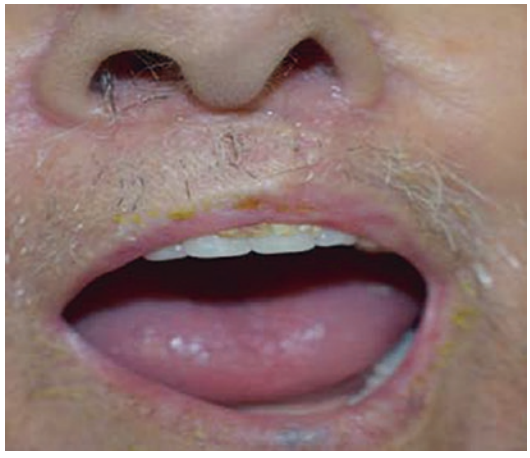

Fig. 24.14 Open mouth 6 weeks post op

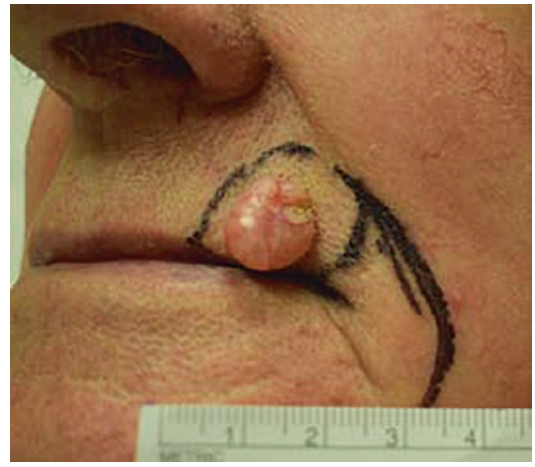

Fig. 24.15 Lesion and flap design

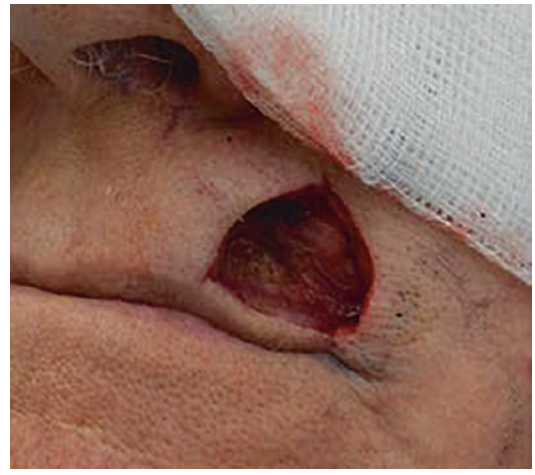

Fig. 24.16 Lesion excised

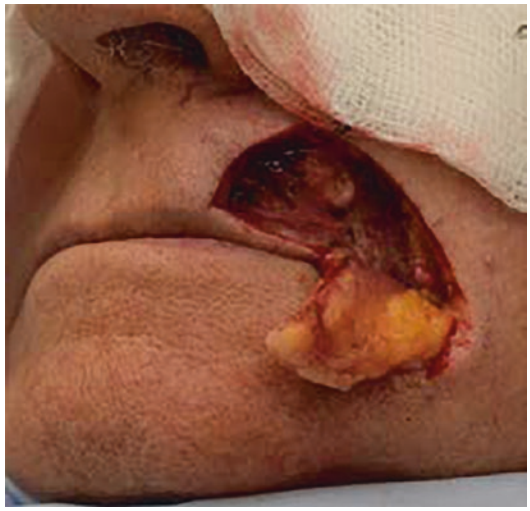

Fig. 24.17 Flap elevated

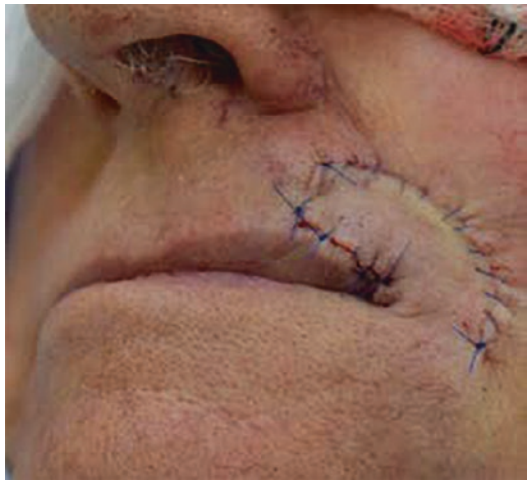

Fig. 24.18 Two layered closure

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Common Local Flaps

25

Marios Stavrakas and Platon Trigkatzis

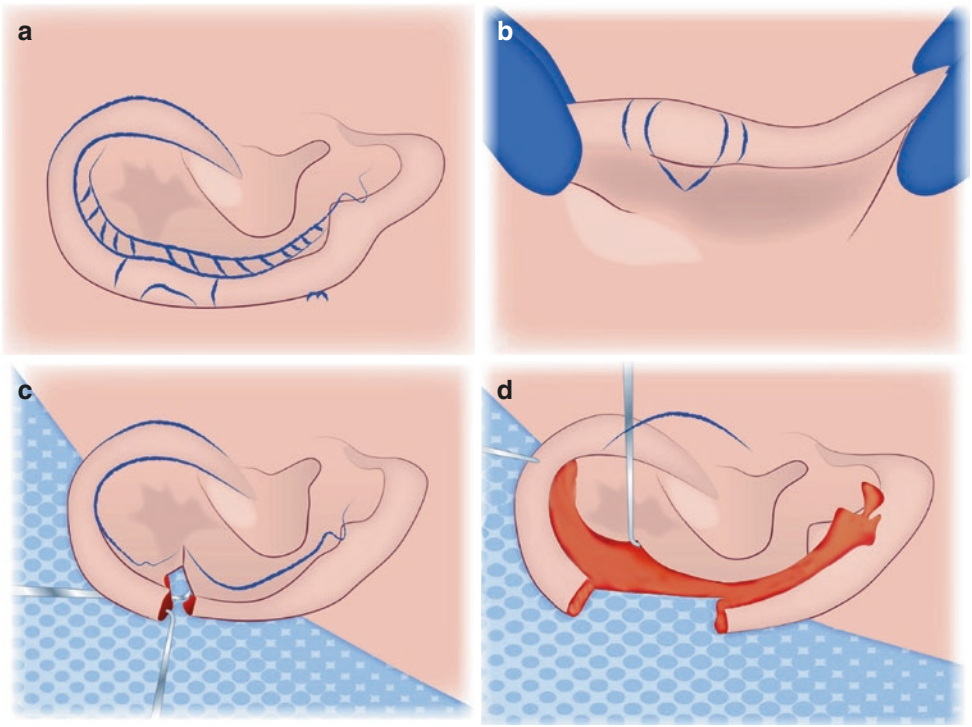

Fig. 25.1 Antia-Buch flap (type of advancement flap). (**a**, **b**) Marking of excision margins of lesion. (**c**) Excision of skin lesion from helix. (**d**) Incision inside helical rim (through anterior skin, through cartilage, leaving posterior skin intact). Dissection of chondrocutaneous flaps (detachment of anterior skin and cartilage from posterior skin). Advancement and alignment of helix. Closure

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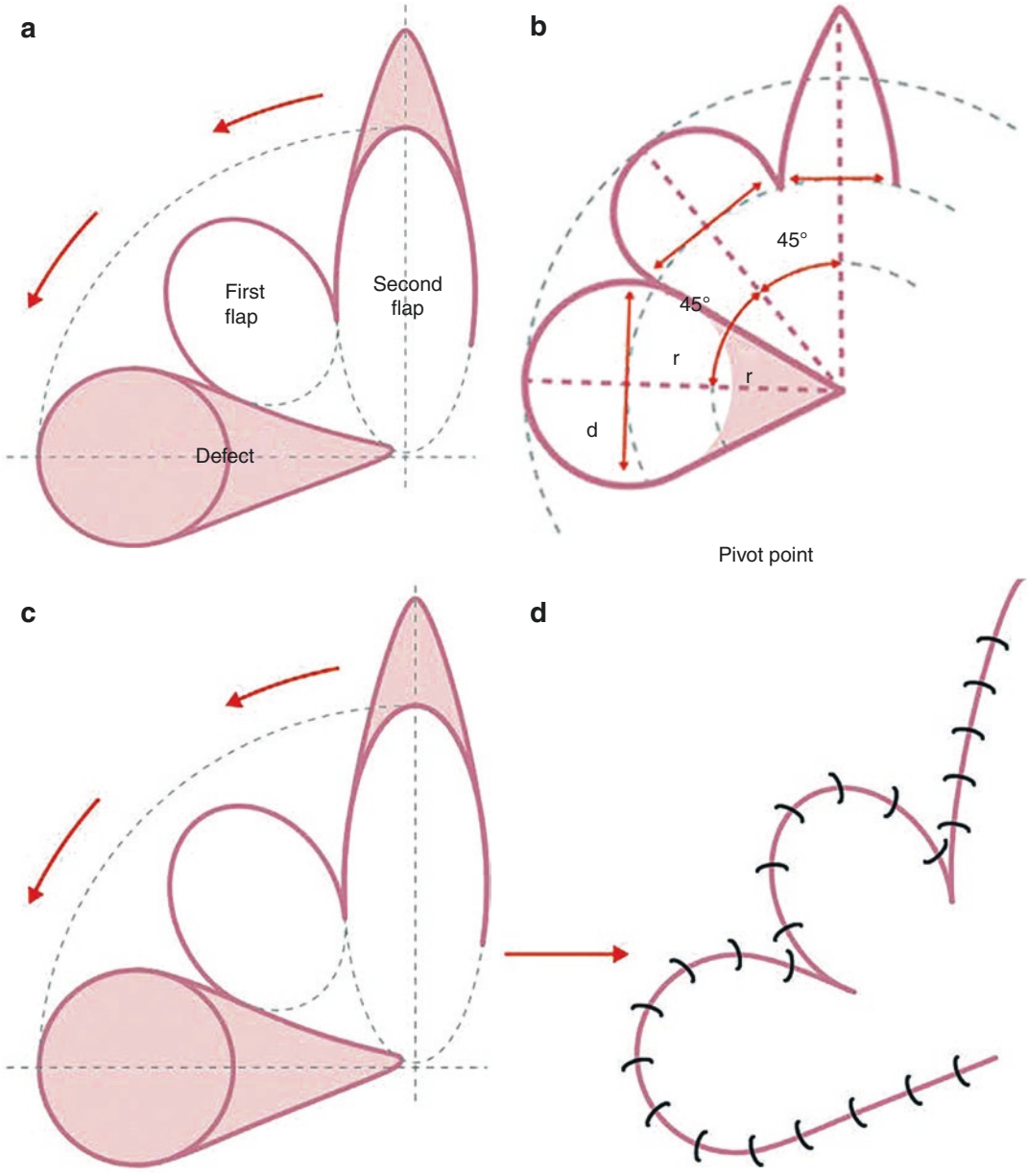

Fig. 25.2 Bilobed flap (type of transposition flaps). (**a**) The first flap covers the defect. The second flap covers the secondary defect. (**b**, **c**) The radius of the defect is measured and the pivot point is mark in one "r" distance from the defect. Total pivot arc = 90° • Pivotal arc between each flap = 45° • The first flap is equal or slightly smaller than the defect • The second flap is smaller, and can be up to 1/3 of the size of the first flap. (**d**) Outcome after closure

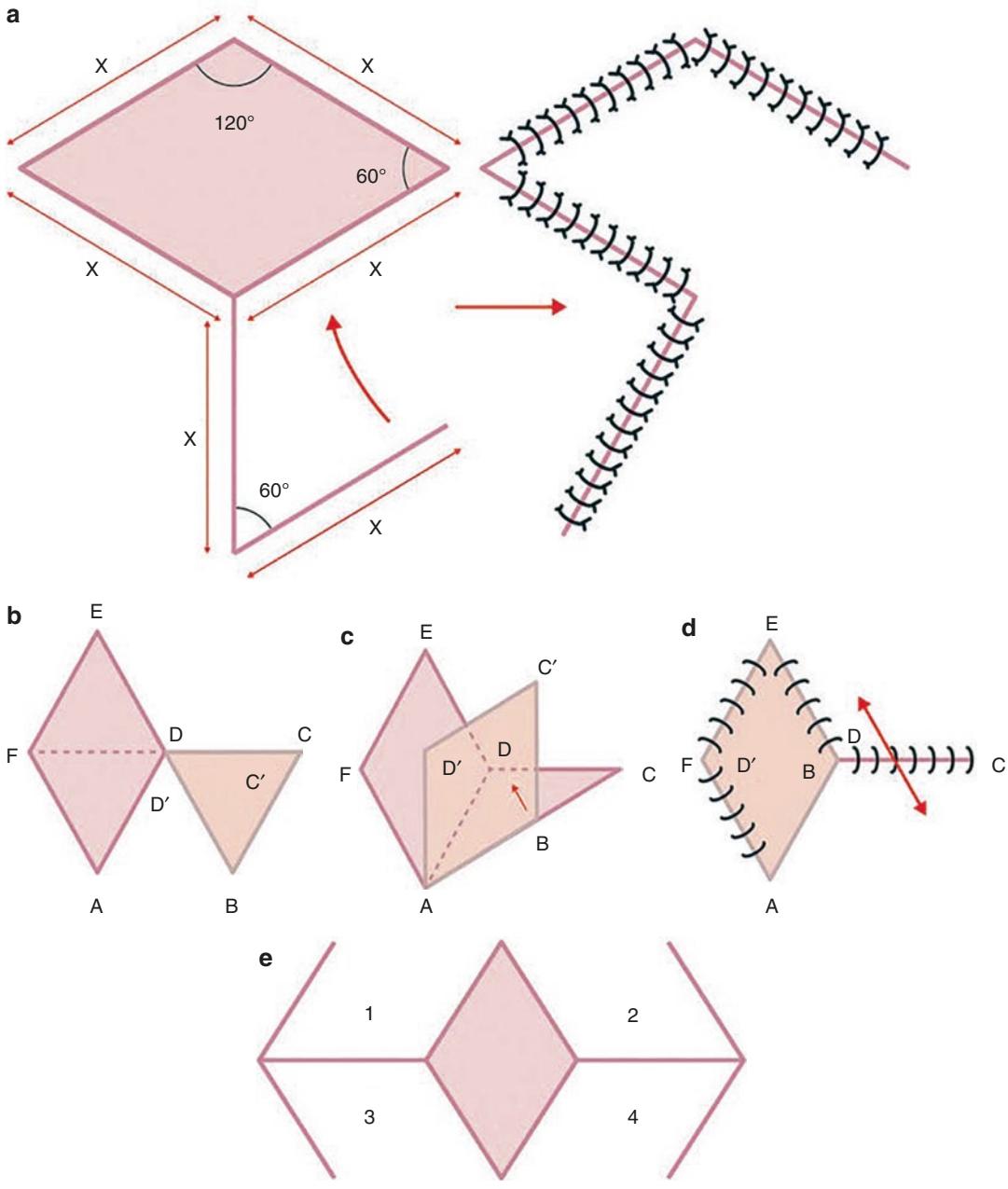

Fig. 25.3 Rhomboid (Limberg) flap (type of transposition flap). (**a**) The excision has to be marked in a rhomboid shape with 60° and 120° angle. Outcome after closure. (**b**–**d**) Planning of rhomboid flap. Transposition movement of the flap. Closure. (**e**) For any rhomboid defect, four different rhomboid flaps can be designed

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Basal Cell Carcinoma (BCC)

26

Platon Trigkatzis and Marco Malahias

26.1 Introduction

- The most common human malignancy worldwide.
- BCC is a slow growing, locally invasive malignant epidermal tumour, arising from pluripotential epithelial cells of epidermis (Table 26.1) and hair follicles (from the basal layer). Predominantly affects Caucasians.
- Metastasis is extremely rare; varies from 0.0028% to 0.55%.

26.2 Demographics

- 80% of all skin cancers.
- Lifetime risk: Approx. 20–30%
- 80% of all BCCs appear on head and neck region.

26.3 Aetiology

- Genetic predisposition.
- Ultraviolet radiation.

26.4 Risk Factors

- Skin type I, II (Table 26.2).
- Ultraviolet radiation/sunlight.
- Advanced age.
- Immunosuppression
- Carcinogen exposure: UV/arsenic
- Premalignant lesions:
  - Naevus sebaceous of Jadassohn
  - Nevoid basal cell syndrome (Gorlin's syndrome)

Table 26.1 Layers of skin and epidermis

Layers of skin	Layers of epidermis
Epidermis	Stratum corneum
Dermis	Stratum lucidum
Hypodermis (subcutaneous tissue)	Stratum granulosum
	Stratum spinosum
	Stratum basale

Table 26.2 Fitzpatrick skin types

Fitzpatrick skin types
I. Pale white skin: Always burns, never tans.
II. White skin: Burns easily, tuns minimally.
III. Light brown skin: Sometimes burns, gradually tans.
IV. Moderate brown skin: Rarely burns, always tans.
V. Dark brown skin: Rarely burns, tans well and darkens.
VI. Deep dark brown to black skin: Never burns.

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https://doi.org/10.1007/978-3-030-66865-5_26

26.5 Characteristics of BCC

Usually:

- Raised borders
- Pearly central area
- Telangiectasias
- May appear scaly with areas of atrophy or scarring from chronic inflammation

26.6 Subtypes

Infiltrative + Morpheaform = Most Aggressive

Nodular + Superficial = Least aggressive

- Nodular 60% of all BCC'c
- Micronodular 15%
- Superficial 10–15%
- Pigmented
- Cystic
- Infiltrative
- Morpheaform

26.7 Risk of Recurrence and New Primaries

- Recurrence after surgical excision <2% in 5 years.
- New Primary BCC:
  - 35% in 3 years after appearance of first BCC
  - 50% in 5 years
- Metastasis <1% (LN's, Lungs, Bones)

26.8 Treatment

- **Destructive Methods**
  - *Curettage and Cautery (C&C):* Curette removes visible tumour, cautery (electrodesiccation) removes residual tumour cells. Healing by secondary intention.
  - *Cryosurgery:* Suitable for small to large BCC's. Prolonged oedema (4–6 weeks). Permanent pigment loss.
  - *Laser phototherapy (CO_2 laser):* Inability to evaluate surgical margins. Good option for superficial BCCs.
  - *Photodynamic therapy treatment:* Light activated photosensitising drug. Following this, the drug creates free oxygen radicals that destroy tumour.
- **Surgical Methods**
  - *Mohs surgery:* HORIZONTAL sequential excision of lesion and repeat until all margins are free (requires specimen histological examination under microscope at the same time). Most effective treatment. 99% curative
  - *Surgical Excision* (Table 26.3):

Authors recommendation:

- 4 mm for low risk and well-defined BCCs with a layer of underline fat
- 5 mm to 1 cm for high risk and ill-defined BCCs down to underlying fascia (Table 26.4)

Table 26.3 Comparison of excision margins guidelines for BCC

	Low risk BCC		High risk BCC
BAD (British Association of Dermatologists)	Peripheral	4–5 mm (96% curative rate)	5 mm to 1 cm For morphea form 15 mm
	Deep	Through subcutaneous fat	Through subcutaneous fat
NCCN (National Comprehensive Cancer Network-USA)	Peripheral	4 mm	>4 mm
	Deep	Not specified	Not specified
EDF (European Dermatology Forum)	Peripheral	4 mm	>5 mm
	Deep		Level of underlying fascia, or perichondrium (ex on ear) or periosteum (ex on scalp)

26.9 Follow up (F/U)

There is no clear guideline for BCC F/U.

According to the author's experience, patients can be discharged after surgical excision with clear peripheral and deep margins, unless:

- BCC has perineural or perivascular invasion. Case has to be discussed at skin MDT and most often Radiotherapy is offered.
- BCCs excised completely but with narrow margin can be F/U 6 monthly for 1 year
- Patients with recurrent BCCs, multiple BCCs, high risk BCCs (ex morpheaphorm) can be F/U 6 monthly for 3 years (there is no common practice within the British Health care system and cases have to be individualised).
- High risk patients (e.g. Basal Cell naevus syndrome, renal transplant patients) can be F/U for life.

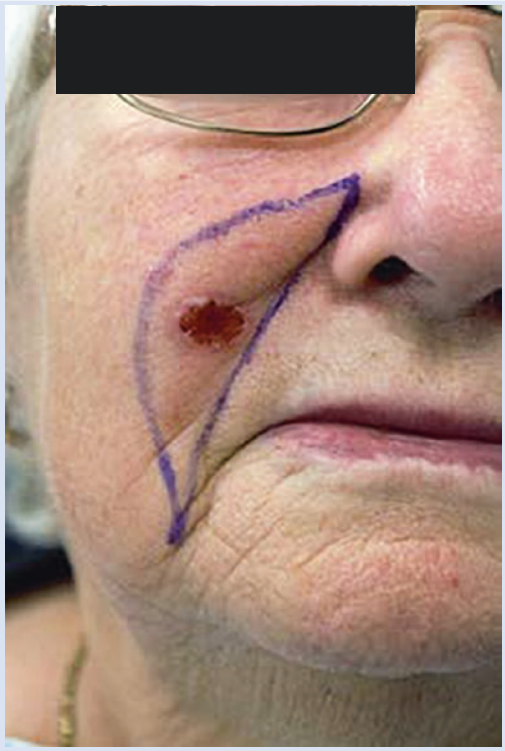
Table 26.4 Risk assessment of BCC

Low risk	High risk
<2 cm in size (on face <6 mm)	>2 cm
Well-defined	Poorly defined
Primary	Recurrent
Nodular, superficial	Morpheaform, infiltrative

26.10 Example of Surgical Treatment of Typical BBC (Table 26.5)

- First mark lesion under bright light and ideally with loupes.
- Mark excision margins.
- Orient scar parallel to wrinkles or natural creases/folds if possible.
- Deep margin: fat or underlying fascia.
- Marker stitch (5-0 silk preferably) to orient specimen (author prefers at 12 o/c).

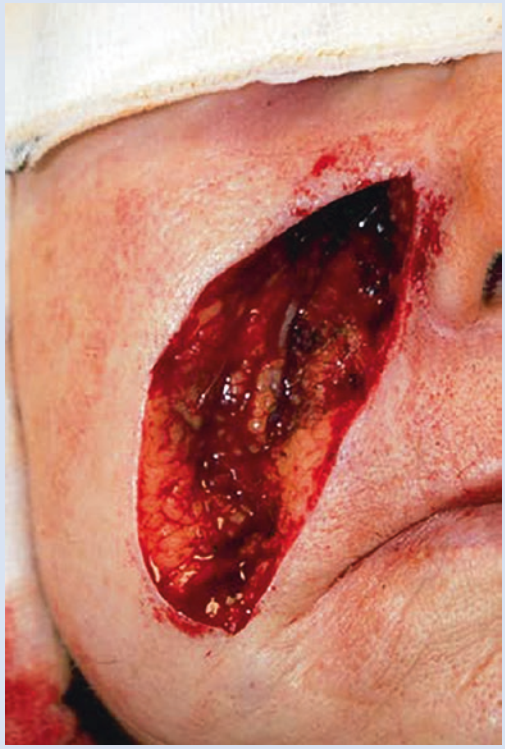
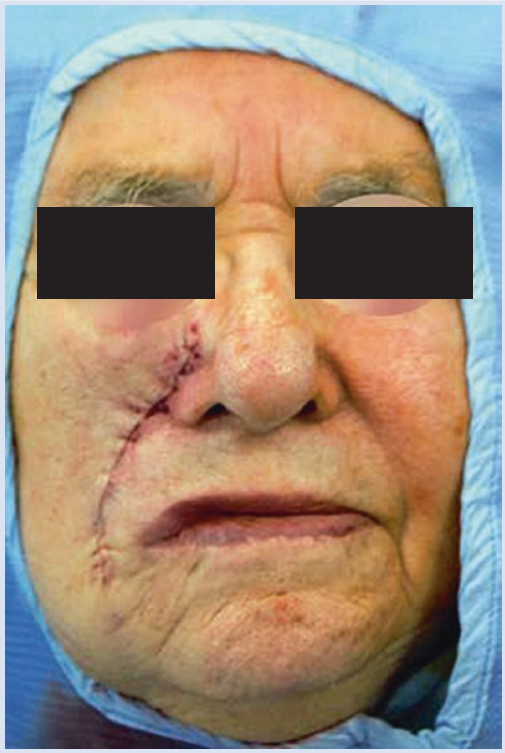
Table 26.5 Example of surgical treatment of typical BBC on the right cheek



BCC of the right medial cheek (nasolabial fold). Marked for excision with a 6 mm margin.

(continued)

Table 26.5 (continued)

	Defect after the excision.
	Direct closure after small undermining of the cheek flap.

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Squamous Cell Carcinoma (SCC)

27

Platon Trigkatzis and Marco Malahias

27.1 Definition

- SCC is a malignant tumour that arises from the keritinising cells of the epidermis and its appendages.
- SCC is the second most common skin cancer after BCC (approx. 20% of all new skin cancers per year).

27.2 Risk Factors

- Usually related to chronic ultra violet light exposure.
- Fair skin: Fitzpatrick I, II.
- Sun exposure.
- Age >50
- Male gender (Male to Female 1.7:1).
- Immunosuppression (ex organ transplant patient, HIV, Leukaemia).
- History of non-melanoma skin cancers.
- Human papilloma virus.
- Carcinogens: Arsenic, organic hydrocarbons.
- Chronic wounds or scars from burns, fistulae.
- Albinism, xeroderma pigmentosum.
- Precancerous conditions: Actinic keratosis (AK), Leukoplakia, Bowen's disease (in situ SCC).

27.3 Rates of Local Recurrence and Metastasis

- 96% of all local recurrences occur within 2 years after excision
- Perineural invasion is a strong predictor of local recurrence and lymph node metastasis
- 5 years local recurrence is about 7%.
- Metastatic rate of SCC is about 5% on the trunk, 10–20% on the head & neck and extremities.
- Regional nodal metastasis is the most common site for metastatic disease. Less frequently can metastasise to bones, brain, lungs.
- Most commonly involved lymph nodes are the neck lymph nodes (41%), followed by the axillary lymph nodes (28%), parotid lymph nodes (22%), inguinal lymph nodes (3%).
- SCC metastasis has generally poor prognosis with a 3-year disease free survival rate of 56%.

27.4 Clinical Presentation-Most Common Characteristics

- Firm nodular plaque or crusted lesion on erythematous base.
- Raised borders.
- Central ulceration.
- Increases in size over weeks to months.

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27.5 Clinical Types

- Verrucous: well-differentiated SCC, exophytic, slow growing, rarely gives local metastasis.
- Ulcerative: Aggressive SCC with raised borders and central ulceration. Can metastasise to regional Lymph nodes.
- Marjolin' s ulcer: aggressive ulcerating SCC arising in an area previously traumatised, chronically inflamed or scarred skin. High (>30%) metastatic rates and highly aggressive recurrences are reported in Marjolin's ulcer.
- Subungual: SCC that involves the nail bed. Erythema, swelling, localized pain, and nail dystrophy, proceeding to nodularity, ulceration, and bleeding. Tumour growth is very slow, and the lesions are frequently misdiagnosed as warts, paronychia, or pyogenic granulomas.

27.6 Predictors of Local Recurrence and Metastasis

- SITE in order of increasing metastatic potential
  - SCC arising at sun exposed areas (excluding lip and ear)
  - SCC of the lip
  - SCC of the ear
  - SCC in non-sun-exposed sites (ex. sole of foot, perineum)
  - SCC arising in scar and chronic wounds
- Size >2 cm
- Depth and Level of Invasion: Tumours >4 mm in depth or extending into or beyond subcutaneous tissue (Clark level V)
- Degree of cellular differentiation (poorly and moderately differentiated have higher recurrence rate than well differentiated).
- Perineural, Lymphatic or Vascular invasion.
- Subtype: Acantholytic, Spindle and Desmoplastic have poorer prognosis.
- Host Immunosuppression.
- Previous Treatment.

27.7 Staging of SCC (Table 27.1)

British Association of Dermatologists and the Royal College of Pathologists follow the 8th edition of TNM staging published by the Union of International Cancer Control (UICC-TNM 8).

Table 27.1 Staging of SCC

T: Primary Tumour. Every clinician has to measure SCC's maximum clinical dimension to establish T. If the clinical dimension is not available, then the histopathological measurement can be used.			
Tis	In situ SCC-no invasion (Bowen's disease)		
T1	≤2 cm in greatest dimension		
T2	2–4 cm in greatest dimension		
T3	>4 cm in greatest dimension		
T4a	Invades cortical bone/marrow invasion		
T4b	Invades skull base, axial skeleton, vertebral foramen to epidural space		
N: For the Head and Neck SCCs, the cervical node N classification is used			
N0	No regional LN		
N1	Single ipsilateral LN ≤ 3 cm in greatest dimension		
N2a	Single ipsilateral LN > 3 cm but not more than 6 cm		
N2b	In multiple ipsilateral LNs, none more than 6 cm		
N2c	Bilateral or contralateral LNs, none more than 6 cm		
N3	LN more than 6 cm		
M: distant metastasis			
M0	No distant metastasis		
M1	Distant metastasis		
Stage grouping			
Stage	**T**	**N**	**M**
0	Tis	N0	0
I	T1	N0	0
II	T2	N0	0
III	T3	N0	0
	T1,2,3	N1	
IVA	T1,2,3	N2,3	0
	T4	Any N	
IVB	Any T	Any N	M1

27.8 Treatment

- *Medical*
  - Radiation: reserved for poor surgical candidates, as adjuvant therapy and recurrent tumours that require multimodal therapy.
  - Topical: 5 Fluorouracil (5 FU): only for premalignant lesions (e.g. Actinic Keratosis).
  - Photodynamic therapy: for premalignant lesions.
- Destructive methods
  - Curettage and cautery and cryosurgery can be used in small superficial lesions but they don't produce a specimen for histology and margin analysis.
- *Surgical excision*
  - **This is the treatment of choice** for the majority of SCCs.
  - For low risk SCCs, well defined, smaller than 2 cm in diameter:
    - 4 mm peripheral surgical margin completely removes tumour in 95% of cases
- For high risk SCCs, more than 2 cm, ill-defined, moderately or poorly differentiated, and those on ear, lip, scalp, eyelids, nose:
- ≥6 mm peripheral surgical margin is recommended or Mohs Surgery
- Deep margin: in either low or high-risk SCC, excision down to the underlying fascia/anatomical plane is recommended by the author.
- Mohs Surgery has the same indications as in BCC's. Has a great success rate in completely removing the tumour (>95%). Can be used in aesthetic units that are difficult to be reconstructed (ex eyelids, nose).
- There are no prospective randomised studies comparing outcomes between conventional surgical excision and Mohs surgery.

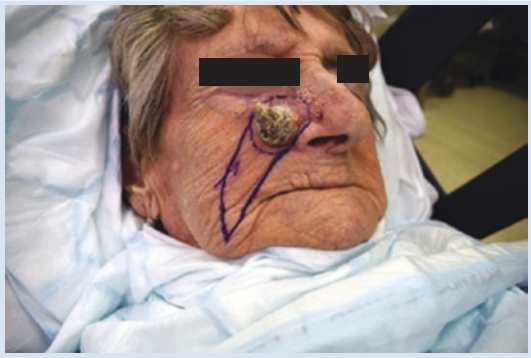
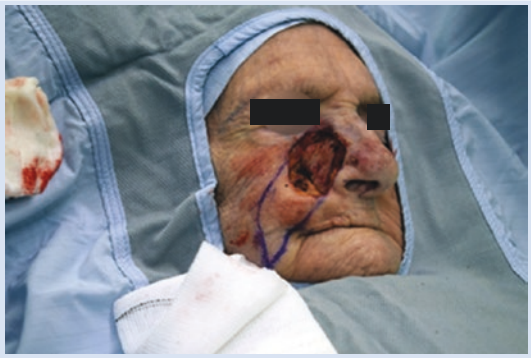
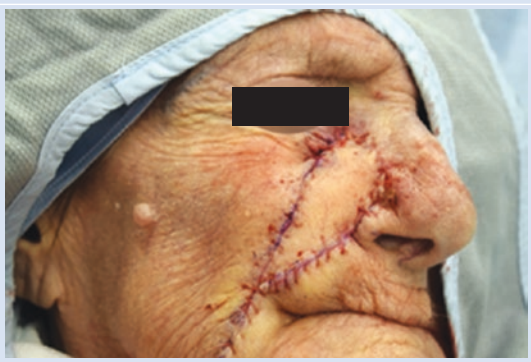
27.9 Follow up (F/U)

- All SCC cases should be discussed at the Skin Cancer MDT regarding either further treatment or follow up.
- Usually:
  - All patients should be given clear instruction in self-examination of the surgical scar site, local skin and lymph nodes.
  - For low risk SCC there is no need for F/U after excision with clear deep and peripheral margin unless advised otherwise by MDT.
  - For high risk SCC (e.g. poorly differentiated) F/U 3 monthly for the first 2 years and 6 monthly for the following 3 years (5 years in total).
  - Some high-risk patients (e.g. renal transplant patient) may need F/U for life.

27.10 Case Presentation

(Table 27.2)

Table 27.2 Excision of SCC from right cheek and reconstruction with V-Y flap

	SCC of the right medial cheek and right side of nose 6 mm margin is marked around the SCC V-Y advancement pedicled flap is designed
	SCC has been excised with 6 mm margin, down to the underlying anatomical plane.
	Defect has been reconstructed with V-Y advancement pedicled flap.

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Malignant Melanoma (MM)

28

Platon Trigkatzis and Marco Malahias

28.1 Definition

- Is a malignant tumour that arises from melanocytes (melanin producing cells) mainly found on basal layer of epidermis.
- Can also arise from:
  - mucosa (e.g.: oropharynx, nasopharynx, oesophagus, anorectum, genitalia)
  - eyes: retina, uvea
  - meninges
- MM is the third most common skin cancer in the UK.

28.2 Risk Factors

- Fitzpatrick skin type I or II (easy to burn, no tan or difficult to tan).
- Excessive exposure to sunlight (UVA).
- Sunburn especially intermittent.
- Sunbeds (UVB).
- Genetic predisposition.
- Pre-existing Dysplastic naevi.
- Multiple naevi >50.
- Congenital giant naevus.
- History of melanoma or other skin cancers.
- Immunosuppression
- Family history.

The majority of melanomas can appear as new mole—de novo—or can develop slowly in or near an existing mole (20–30% of melanomas).

28.3 Clinical Examination and Evaluation of Suspicious Lesions for Melanoma

- AMERICAN SYSTEM

Asymmetry	If you "split" the pigmented lesion in quarters, they don't match.
Border	Irregular, Blurred, Notched
Colour	Not the same all over, areas of darker pigmentation, and multiple pigmentation,
Diameter	>6 mm
Evolving	Changes in Size, Shape, Colour

- WEIGHTED 7-POINT CHECKLIST

Recommended by NICE for routine use in the UK General Practise for referral. All suspicious pigmented lesions scoring *3 points or more* should be referred as a 2 week wait (2 w/w).

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Major features (2 points each):

- Change in size
- Irregular shape
- Irregular colour

Minor signs (1 point each):

- Largest diameter 7 mm or more
- Inflammation
- Oozing
- Change in sensation

When examining a patient with multiple pigmented lesions as a rule of thumb is to excise or refer for excision of the mole that sticks out and looks different form the others. This is called the "Ugly duckling sign" but this method has limitations.

The most accurate evaluation of pigmented lesions can be performed with dermoscopy. Dermoscopy is a non-invasive, in vivo evaluation of the pigmentations and microstructures of epidermis, dermo-epidermal junction and papillary dermis that are not visible to naked eye.

28.4 Types of Melanoma

- Superficial Spreading Melanoma: Most common type. About 70% of all cases. Usually arises from pre-existing lesions as a flat irregular pigmentation with prolonged (can be years) horizontal growth pattern (initial radial growth phase within the epidermis and sometimes within the papillary dermis). Good prognosis.
- Nodular Melanoma: 15–20% of all cases. More often arises de novo. Aggressive. In vertical growth pattern (deeper extension).
- Lentigo Maligna Melanoma: Approximately 5% of cases. Clearly related to sun exposure. Typically in elderly people on the face in precursor lesions/freckles (Hutchison freckle) that slowly become irregular, with multicolouration. Best prognosis among melanomas.
- Acral Lentiginous Melanoma: Usually on palms, soles of feet, subungual. 2–3% of all melanomas but most common type in Afro-Caribbean and dark skin papulation
- Amelanotic Melanoma: Rare, challenging diagnosis due to lack of pigmentation. Typically presents as an asymmetrical macule pinkish or reddish or can have some light pigmentation at the periphery.
- Desmoplastic melanoma 1% of all melanomas. Mainly on head and neck of elderly people, usually lacks typical melanoma features, can mimic scar, propensity for perineural invasion (also called neurotropic melanoma), high rate of spread to regional lymph nodes.

28.5 Histologic Features

- The most important prognostic factor is BRESLOW THICKNESS: Tumour thickness measured from the granular layer of overlying epidermis (or base of ulceration in ulcerative melanomas) to deepest malignant cells involving dermis.

<1 mm	1–4 mm	>4 mm
Breslow thickness	Breslow thickness	Breslow thickness
thin melanomas	intermediate melanomas	thick melanomas

- Essential Histologic Features
  - Size
  - Breslow thickness
  - Ulceration
  - Mitotic Rate
  - Peripheral and deep margin status
  - Microsatellitosis

- Optional Histologic Features
  - Gross description
  - Lymphovascular invasion
  - Neurotropism/perineural invasion
  - Regression
  - Clark Level: Anatomic level of invasion through skin layers, five levels as follows:
    Only on epidermis, no invasion: Melanoma in situ
    Invades papillary dermis
    Invades papillary-reticular interface

Invades reticular dermis
Invades subcutaneous fat
- Horizontal/Vertical growth
- Histologic subtype
- Tumour infiltrating lymphocyte (TIL)

28.6 Metastatic Melanoma

- Melanoma can metastasise through lymphatics or blood vessels to adjacent skin, distant skin, lymph nodes or other organs (mainly lungs, liver, brain and bones).

Non-nodal Regional Metastasis:

- In-transit metastasis: are melanoma deposits within the regional dermal and subdermal lymphatics lymphatic vessels of the skin, located more than 2 cm away from the site of primary melanoma and before the regional lymph nodes.
- Satellite metastasis: lesions occurring within 2 cm of the primary tumour.
- Microsatellite: Histopathologically determined on the excised specimen.

Nodal metastasis:

- The first lymph nodes to be involved are the regional lymph nodes.
- Sentinel lymph node biopsy (SLNB): The removal and examination of the first lymph node to which cancer cells are likely to spread from the primary tumour.
- Can be detected using lymphoscintigraphy prior to surgery and/or injection of blue dye intraoperatively.
- Indications for SLNB:
  - Melanomas with Breslow >0.8 mm (in some Trusts still perform SLNB when >1 mm).

Consideration of SLNB in:

- Melanomas with Breslow <0.8 mm with ulceration, high mitotic rate, Clark IV or V, lymphovascular invasion
- Melanomas with Breslow >4 mm and clinically negative LNs.

28.7 Management of Patient with Primary Cutaneous Melanoma

- E.g. Patient is referred to the Clinic with suspicious pigmented lesion.
  - History, examination (including LNs), dermoscopy of lesion if possible.
- If lesion is clinically suspicious for MM, we perform excision biopsy i.e. excision with 2 mm margin and small amount of underlying fat.
- NB. Do not give unnecessary long scars and always think the orientation of the scar in order to facilitate Wide Local Excision (WLE) on a later stage if required.
- Big margins on initial biopsy a) will make WLE more difficult, b) will disrupt microlymphatic vessels and might alter or make SLNB non feasible if required.
- If the histology report confirms the excised lesion as MM, then the case has to be discussed at the skin cancer MDT.

Wide local WLE of the melanoma scar ± SLNB will be recommended

WLE reduces the risk of local recurrence

The margin of the WLE depends on the Breslow thickness of melanoma (Table 28.1).

- These margins cannot always be achieved in difficult anatomical areas.
- Deep margins: There are no randomised control trials for the depth of the WLE. However, most common practice is to excise:
  - Deep to the underlying fascia (leaving fascia intact).
  - On extremities down to muscular fascia.
  - On the neck down to platysma muscle fascia.

Table 28.1 Guidelines for WLE

Thickness	Excision margin
In situ	5 mm
<1 mm	1 cm
1–2 mm	1–2 cm
2–4 mm	2 cm
>4 mm	2 cm

– On the head down to periosteum (galea should be excised).
– On the nose down to perichondrium or periosteum.
– On the ears is preferable to excise the cartilage as well.

28.8 Staging of Melanoma

There is a clinical TNM classification (cTNM) and a pathological TNM (pTNM) classification that uses information gained from primary melanoma, wide local excision, SLN biopsy and/or regional lymphadenectomy.

British Association of Dermatologists and Pathologists follow the Union for International Cancer Control (UICC-TNM 8th edition) classification to stage melanoma in 0-IV stages. UICC-TNM 8 is now identical to the American classification of melanoma AJCC-TNM 8th edition. This is the latest update of melanoma classification introduced in 2018

- **T:** Related to primary tumour.
- **N**: Determined by lymph node involvement/status.
- **M**: Metastasis to other organs or parts of the body (including distant skin sites and distant lymph nodes).
- **T category (Stage I, II)** defines the primary tumour as summarised in Table 28.2.

Breslow thickness and ulceration are the main parameters. Mitotic rate and Clark level have been removed on the updated AJCC-TNM8 classification.

- **N category (Stage III)** includes information gained from SLNB, core biopsy, clinically detected LNs and lymph node dissection. Table 28.3 is a simplified summary of the Nodal status.

Suffix a (Na) indicates clinically occult lymph nodes (i.e. SLNB detected).

Suffix b (Nb) indicates clinically detected lymph nodes.

Suffix c (Nc) indicates in-transit, satellite or microsatellite metastasis.

- **M category (Stage IV)** defines the distant metastasis (Table 28.4).

Suffix 0 indicates non-elevated LDH. e.g. M1a(0)

Suffix 1 indicates elevated LDH. e.g. M1c(1) (Table 28.4)

Table 28.2 Primary Tumour (T) with no lymph node involvement (N0) or metastasis (M0)

Stage	T		N	M
0	Tis	In situ	0	0
IA	T1a	<0.8 mm, without ulceration	0	0
IA	T1b	<0.8 mm, with ulceration 0.8–1 mm, with or without ulceration	0	0
IB	T2a	>1–2 mm, without ulceration	0	0
IIA	T2b	>1–2 mm, with ulceration	0	0
IIA	T3a	>2–4 mm, without ulceration	0	0
IIB	T3b	>2–4 mm, with ulceration	0	0
IIB	T4a	>4 mm, without ulceration	0	0
IIC	T4b	>4 mm, with ulceration	0	0

Table 28.3 Lymph node status without metastasis (M0)

Stage	N	M
IIIA–IIID	N1a: 1 clinically occult LN	0
	N1b: 1 clinically detected LN	0
	N1c: 0 LN, presence of satellite, microsatellite, in-transit	0
	N2a: 2–3 clinically occult LNs	0
	N2b: 2–3 clinically detected LNs	0
	N2c: 1 LN, presence of satellite, microsatellite, in-transit	0
	N3a: ≥4 lymph nodes, at least one of them clinically occult	0
	N3b: ≥4 lymph nodes, at least one of them clinically detected	0
	N3c: ≥2 LNs, presence of satellite, microsatellite, in-transit	0

Table 28.4 Metastatic diseases

Stage	M
IV	M1a: Metastasis to distant skin and/or non-regional LN.
	M1b: Metastasis to lung.
	M1c: Metastasis to non-CNS organs.
	M1d: Metastasis to CNS.

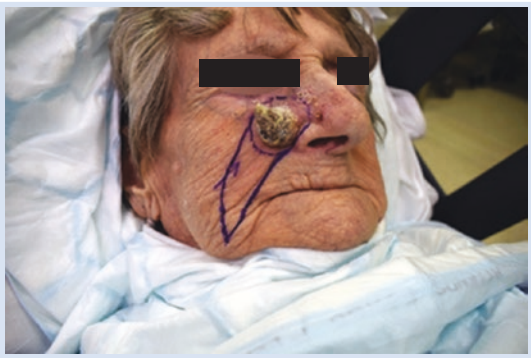
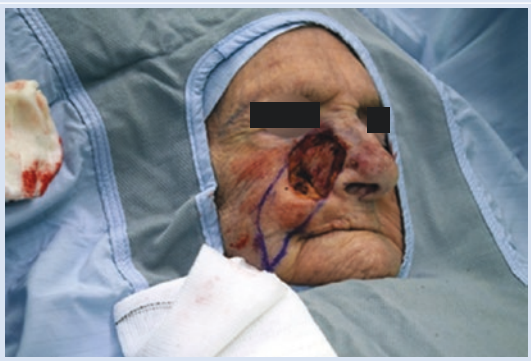
28.9 Follow up (F/U)

All patients should be taught to self-examine once per month.

- Melanoma in situ: No need for F/U after WLE.
- Stage IA: F/U for 1 year (2–4 visits).
- Stage IB-IIC: F/U for 5 years (3 monthly first 3 years, 6 monthly the following 2).
- Stage III-IV: F/U for 10 years (3 monthly first 3 years, 6 monthly the following 2, yearly from the fifth up to the tenth year).
  - Investigations may be needed following MDT recommendations, including full body CT surveillance and blood tests including Serum Lactate Dehydrogenase Level (LDH).
  - High LDH levels is a poor prognostic factor in patients with distant metastases, especially in the lung and liver.

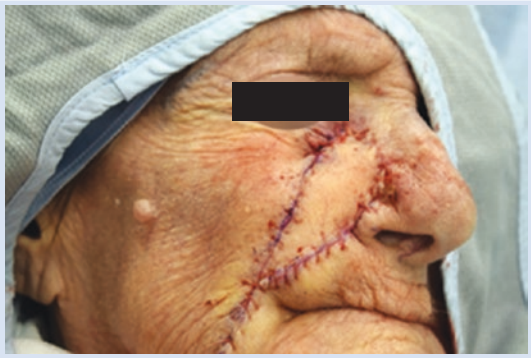
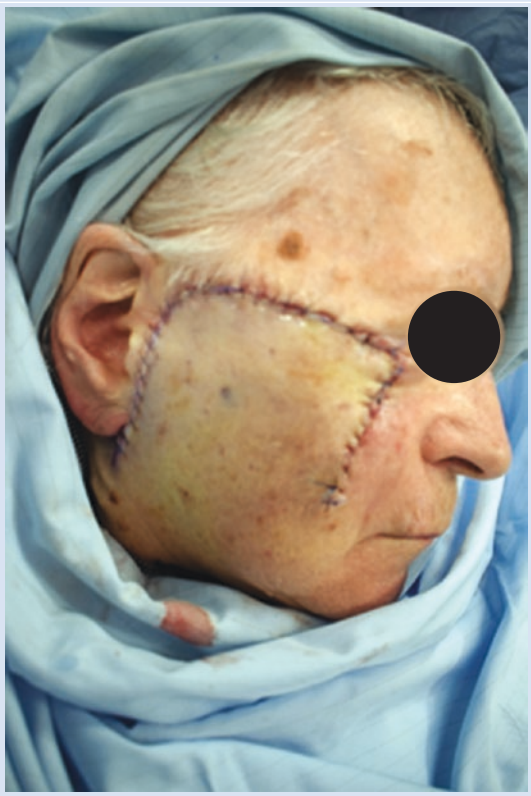
28.10 Case Presentation (Table 28.5)

Table 28.5 Wide Local Excision of Lentigo Maligna Melanoma of Right cheek and reconstruction with cheek rotational flap

	Lentigo Maligna of right cheek marked to be excised with a 5 mm margin after MDT recommendation. Cheek rotation flap designed to resurface the defect.
	Defect after surgical excision.

(continued)

Table 28.5 (continued)

	Flap raised on the underlying anatomical plane (SMAS).
	Closure of the wound with 5-0 prolene suture.

[a]Superficial Muscular Aponeurotic System

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Rhinophyma

29

Hisham S Khalil and Marios Stavrakas

29.1 Case Presentation

A 61-year-old patient was referred to the Rhinology clinic to discuss about treatment options for his rhinophyma. He did not receive any previous therapy and the deformity of the nose has progressively deteriorated, having a negative impact on his confidence and social life. He did not have any other dermatological problems and was on Warfarin for Atrial Fibrilation. After discussion about the surgical treatment, pre-operative photographs were taken (Fig. 29.1) and warfarin was switched to LMWH, during the perioperative period during the local protocol. He underwent a microdebrider-assisted rhinophyma excision and biopsies were taken to exclude any skin malignancy (Fig. 29.2). The wound was left to heal by secondary intention and was covered with non-adhesive dressings and antibiotic ointment. He was reviewed regularly (weekly for the first 4 weeks) in order to assess the wound healing and exclude any wound infection. The cosmetic result was satisfactory, and no more revision surgeries were required.

29.2 Background Knowledge

Rhinophyma is a rare condition, affecting predominantly Caucasian males, with a male-to-female ratio of 12–30:1. Its exact pathogenesis remains unclear. Rhinophyma is closely associated with acne rosacea and, more specifically, Type III of acne rosacea. Another possible aetiologic factor is the colonization of the sebaceous glands with Demodex Folliculorum.

29.3 Clinical Approach

29.3.1 Diagnosis

The diagnosis is based on clinical examination and basically inspection. Differential diagnosis includes Basal Cell Carcinoma (BCC), which has been reported to be between 5% and 10% in the rhinophymatous tissue, Squamous Cell Carcinoma (SCC), sebaceous carcinoma, angiosarcoma, metastatic disease, granuloma eosinophiliacum, sarcoidosis and lymphoma.

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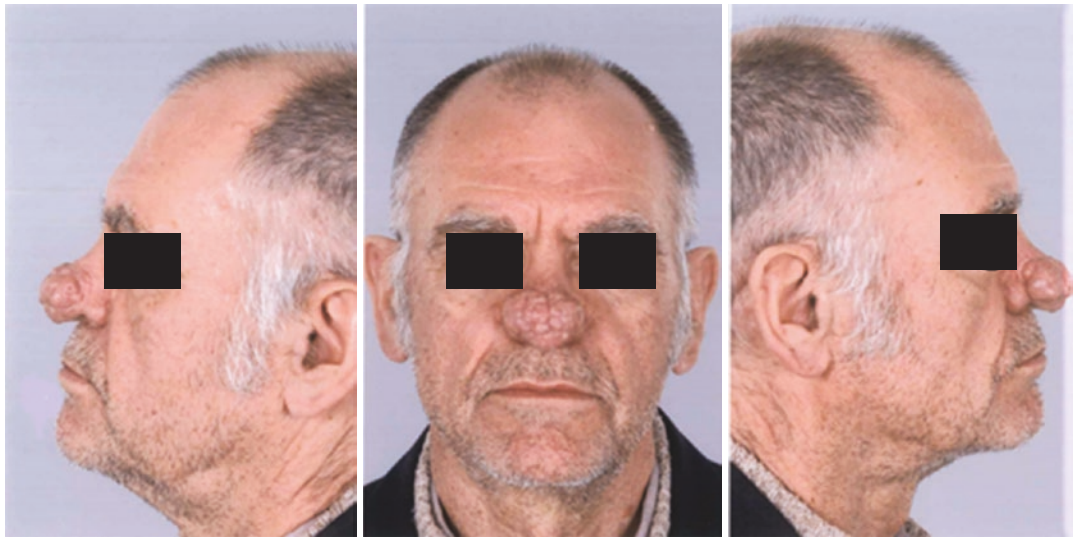

Fig. 29.1 Preoperative photographs

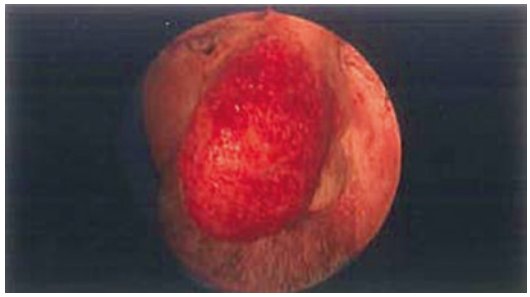

Fig. 29.2 Intraoperative photograph after the final sculpturing using the macrodebrider

29.3.2 Treatment

Medical treatment (early stages) especially in the early stages of rhinophyma, although here is no solid evidence in the literature confirming that these treatments are successful.

- Low-dose isotretinoin → to slow down the disease progression.
- Tamoxifen → to reduce the production and secretion of TGFb2 by rhinophyma-associated fibroblasts.

The mainstay in the treatment of rhinophyma is surgery.

- Cryosurgery: The main advantages are minimal bleeding, little pain and no destruction of the underlying nasal cartilages if used appropriately. On the other hand, it can result is dyschromia and scarring.
- Blade excision: It is still widely used. A modification is the Shaw heated scalpel, which cuts tissue and coagulates blood vessels at the same time. Disadvantages of this method are postoperative pain, mild scarring and slight nasal alar collapse.
- Coblation: It raises the temperature to 60–70 °C, compared to the Shaw blade, which reaches temperatures from 150 °C to 200 °C. In that way, Coblation ensures a bloodless field and minimal pain. The disadvantages of the method are hypopigmentation and prolonged erythema.
- Grafting techniques and the subunit method: The latter aims to address the hypertrophic sebaceous tissues, the excess skin problem and also the destruction of support.
- Other surgical approaches: harmonic ultrasound scalpel, dermabrasion, electrosurgery, Versajet Hydrosurgery system. Also, laser-assisted treatments have been described, including CO_2 laser, Erbium: YAG laser and the diode laser.
- Microdebrider-assisted rhinophyma excision is not a new method in the literature, although few cases have been presented up to date. We follow a two-stage approach in our cases.

Scalpel blade excision allows quick removal of the main tissue bulk and obtaining tissue samples for histology. At the same time, the use of the microdebrider is beneficial for quick contouring and shaping of the nasal subunits, reducing the risk of damage to the underlying cartilaginous structures. We believe that a straight microdebrider handpiece is appropriate for tip refinement and work around the nasal alae. The preservation of deeper skin layers allows re-epithelialisation with less scarring and also the tactile feedback that the surgeon takes from the instrument. As it is a relatively quick method, it reduces intraoperative blood loss and the risk of infections.

29.3.3 Follow up

In our practice, we leave the wound heal by secondary intention after microdebrider-assisted rhinophyma excision. It is important to monitor the patients closely during the post-operative period, prevent any infections and observe the wound healing process. The use of non-adhesive dressings and antibiotic ointments is also adviced.

Summary and Author's Comments

1. Rhinophyma is a rare condition with unclear pathogenesis.
2. Differential diagnosis should include skin malignancies, such as BCC (5–10%) and SCC.
3. The mainstay in the treatment is surgical excision. The authors prefer microdebrider-assisted rhinophyma excision.

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Part V

Anterior Skull Base Disorders

Cerebrospinal Fluid Rhinorrhea

30

Hisham S Khalil

30.1 Case Presentation

A 42 year old man presented with a history of recurrent watery rhinorrhea and a salty taste in his mouth. He was previously diagnosed with a traumatic CSF leak and underwent a trans-nasal trans-sphenoidal repair under the care of the neurosurgeons. He had a history of hydrocephalus, learning difficulties and a previous ventriculo-peritonea (VP)l shunt. A CT Cisternogram demonstrated a defect and leak in the left sphenoid sinus. A new venticulo-peritoneal shunt was inserted after removal of the old one, followed by a trans-nasal endoscopic repair of the defect in the sphenoid using fascia lata and a fat graft.

30.2 Background Knowledge

CSF rhinorrhea occurs due to traumatic and non-traumatic causes with the former being the most common. Traumatic causes could be iatrogenic or accidental with head injuries.

The treatment of CSF Rhinorrhea will depend on the underlying cause, the site and volume of the CSF leak. For traumatic causes, a conservative approach including bed rest with head elevation, avoidance of straining and possibly a lumbar puncture for 3–5 days usually suffices in the vast majority of patients. The treatment of iatrogenic CSF leaks is best carried out at the time of surgery. For patients with CSF rhinorrhea detected postoperatively, imaging is important in detecting the site of the leak as well as measures to prevent/treat infections including meningitis. Current practice favours a trans-nasal endoscopic approach, though a craniotomy may have to be resorted to in patients with recurrent and persistent CSF rhinorrhea and where the endoscopic approach has failed.

The treatment of patients with recurrent CSF rhinorrhea in the presence of persistent intracranial pathology is challenging. It is important to adopt a multidisciplinary approach in assessing and treating these patients with input from Otolaryngologists, Neurosurgeons and Maxillofacial surgeons. Identification and treatment of causes of a raised intracranial pressure is important. These include benign intracranial hypertension, hydrocephalus and irresectable intracranial tumours. Patients with skull base tumours may develop CSF rhinorrhea following chemoradiotherapy and due to shrinkage of tumour tissue in the presence of an underlying skull base defect. It is important to have a low threshold for suspecting a CSF leak in such patients who develop a persistent watery rhinorrhea. It is equally important to treat such leaks promptly due to the high risk of infection.

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30.3 Clinical Approach

30.3.1 History

A history of head or facial trauma, a Neurosurgical/Nose/Ear surgery is relevant. In this patient, there was a history of trauma and a previous neurosurgical procedure with an associated hydrocephalus. His existing VP shunt had became infected and stopped working. Other examples of associated intracranial pathology include benign intracranial hypertension and inoperable tumours. It is important to explore the possibility of a CSF rhinorrhea in patients with a clear nasal discharge following sino-nasal, anterior skull base or nasal framework surgery. CSF oto-rhinorrhea could also occur after ear and lateral skull base surgery. In addition to a watery rhinorrhea, the patient often experiences a salty taste. Patients may also complain of non-specific headaches, visual disturbances or seizures depending on the underlying cause. Symptoms of meningitis should also be actively explored in the history.

30.3.2 Examination

The assessment of a patient with a suspected CSF rhinorrhea includes a nasal endoscopy where there may be evidence of fluid in the nasal cavity (Fig. 30.1) or nasal pathology. This may be an associated meningocele or meningoencephalocele. The nasal endoscopy could also identify granulations around the area of CSF leak in the skull base. An ear examination is important in patients with a history of ear surgery and fractures of the petrous temporal bone. Patients with a CSF rhinorrhea due to benign intracranial hypertension may have evidence of bilateral papilloedema.

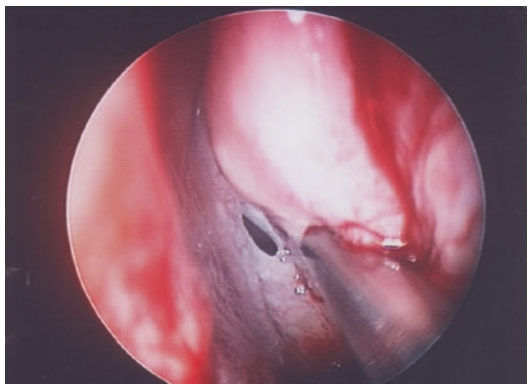

Fig. 30.1 CSF rhinorrhea emanating from the left sphenoid sinus ostium

30.3.3 Investigations

Biochemical Assay Collection of nasal fluid in a universal sterile container (aided by the patient's head in a dependent position) is important for *B*2 Transferrin (TAU protein) testing. This usually requires about 3–5 ml of fluid to complete the test. Where available, a *B*2 trace protein test allows confirmation of the presence of a CSF leak with much smaller amounts of collected fluid. Patients are often given a container to collect nasal fluid at home. The *B*2 transferrin protein remains stable in extracorporeal CSF at room temperature for up to 7 days. It is prudent to highlight to patients the need to return collected fluid to their General Practitioner practice or local hospital as soon as is feasible to allow the sample to reach the laboratory before 7 days have passed since its collection to avoid false negative results. The sensitivity and specificity of B2 transferrin are both very high ranging from 90 to 100%.

Imaging The imaging work-up includes MRI scans of the Head and Sinuses as well as a CT scan of the Sinuses. These serve to detect intracranial pathology and a skull base bony defect respectively. A CT Cisternogram should be requested with a leak that occurs with a regular frequency and serves to localise its site. This involves the intrathecal injection of contrast (Fig. 30.2). An MRI Cisternogram can also be requested.

30.3.4 Treatment

The patient was discussed in the Skill Base MDT and a new VP shunt was inserted after removal of the old one by the neurosurgical team prior to

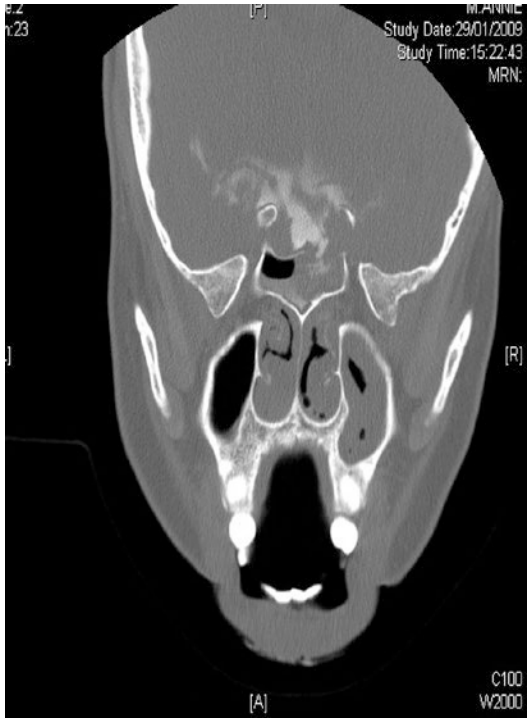

Fig. 30.2 A CT Cisternogram demonstrating a CSF leak in the roof of the left sphenoid sinus with contrast in the sinus

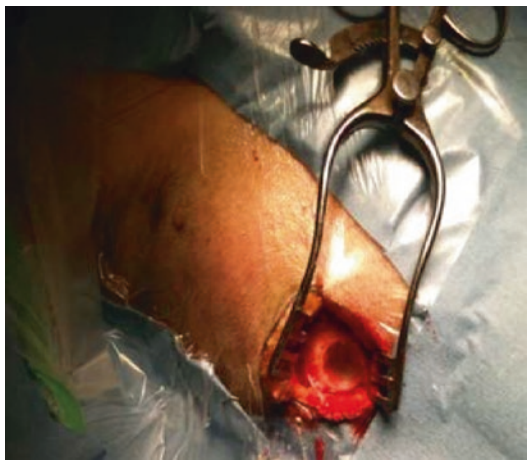

Fig. 30.3 Insertion of VP shunt

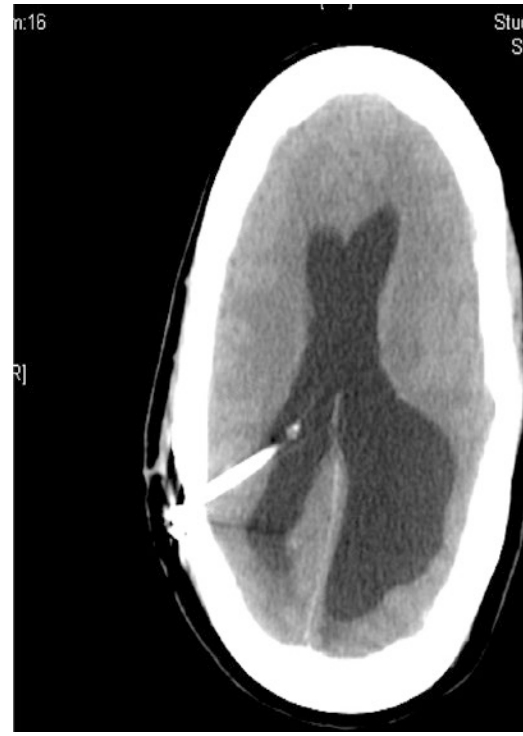

Fig. 30.4 VP shunt in situ on a CT scan

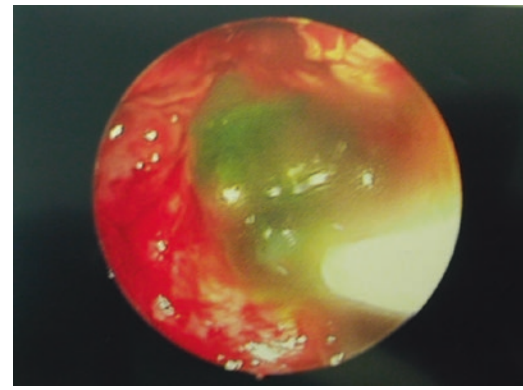

Fig. 30.5 Green fluorescence at site of leak after topical application of fluorescein

endoscopic repair (Figs. 30.3 and 30.4). The patient then had an endoscopic trans-nasal, trans-sphenoidal repair of the CSF leak using fat and fascia lata grafts. The site of the leak in the roof of the sphenoid sinus involved the planum sphenoidal at its junction with the Sella. The site of the leak was detected intraoperatively by applying neurosurgical patties soaked with topical 5% fluorescein for intravenous injection. This negated the need for an intraoperative intrathecal diluted fluorescein injection with its inherent risk of neurotoxicity. There was green fluorescence at the site of contact of CSF with the fluorescein (Fig. 30.5).

The type of graft material used depends on the site of the leak. Fat grafts work well for leaks in the sphenoid sinus. They could be used as a 'dumbbell' graft with intracranial and extracranial components. Alternatively, a Fascia Lata graft could be used as an underlay graft (Fig. 30.6). In other areas of the skull base, Fascia Lata grafts, turbinate mucosal grafts and Denatured Bovine Pericardium patch could be used. In patients who have a sizeable defect of the skull base, a multi-layered repair is resorted to with the use of an underlay Fascia Lata graft, a septal cartilage graft to cover the bony defect and an overlay Fascia Lata graft over the cartilage to complete the repair. The grafts are glued in place using tissue glue, either Tisseel Fibrin glue or Duraseal. It is best to avoid insertion of lumbar drains where possible postoperatively. This reduces the risk of infection and pneumocephalus. In patients with persistent raised ICP, a ventriculo-peritoneal shunt is advocated. Figure 30.7 is a postoperative CT Cisternogram 3 years later when the patient had a suspected recurrence of the CSF leak. The CT Cisternogram demonstrated that the repair was intact.

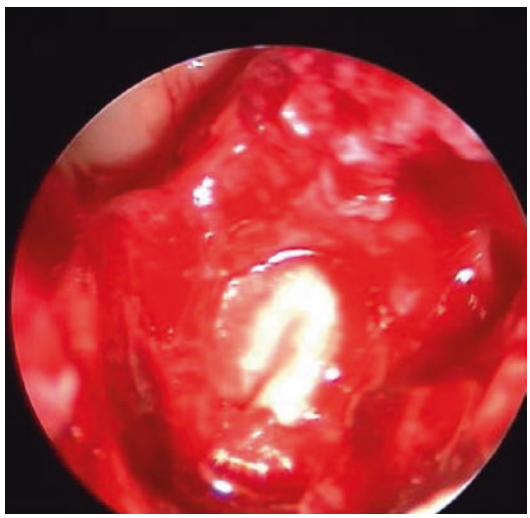

Fig. 30.6 Underlay placement of fascia lata graft to repair the defect, abdominal fat was used as a second layer in the sphenoid sinus

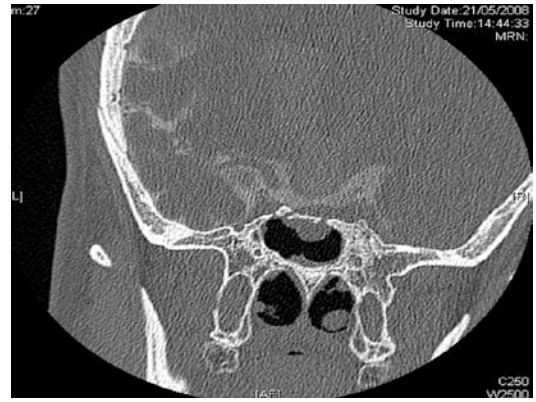

Fig. 30.7 Postoperative CT Cisternogram 3 years after the repair

Summary and Author's Comments

1. CSF rhinorrhea is mostly traumatic in origin. A conservative approach is effective in most of these patients.
2. The management of patients with CSF rhinorrhea and persistent intracranial pathology is challenging and requires a multidisciplinary approach to management.
3. Addressing the cause of raised intracranial pressure is important if a recurrence of the leak is to be avoided.

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31 Non-functioning Pituitary Adenoma

Ellie Edlmann and Samiul Muquit

31.1 Case Presentation

A 71 year old lady presented to her general practitioner reporting a 1 year history of increasing blurred vision in the right eye and was referred to Ophthalmology. Subsequent assessment revealed reduced visual acuity in the right eye with 20/70 vision and normal vision (20/20) in the left eye. In addition to this a bitemporal visual field deficit was detected, worse on the right side (see Fig. 31.1).

The Ophthalmology team referred the patient for imaging of the brain and orbits to assess further (see Fig. 31.2). This revealed a large lesion extending from the sella, measuring 36 × 21 × 26 mm and invading the cavernous sinuses. The lesion was causing compression and elevation of the optic chiasm, worse on the right side and was heterogeneously enhancing with a cystic region.

On further history no endocrine symptoms were reported and a full endocrinology blood panel was performed showing no abnormalities. This case was discussed at the neuro-endocrine multi-disciplinary meeting and determined to be a non-functioning pituitary adenoma (NFPA). Due to the compression on the optic chiasm and risk to further visual deterioration the patient was offered early surgical resection.

31.2 Background

31.2.1 Anatomy

The pituitary gland is nestled within the pituitary fossa, or *sella turcia*, a depression within the sphenoid bone in the middle cranial fossa. Just below this sits the sphenoid air sinus, which provides a good access route for surgery via the nasal cavity, as the *transsphenoidal* approach. The optic chiasm is immediately superior to the pituitary gland, leaving it vulnerable to compression, and hence visual decline, with expanding pituitary tumours (see Fig. 31.3). Laterally adjacent to the pituitary gland are the cavernous sinuses, containing the internal carotid artery and upper cranial nerves (III–VI). Pituitary tumours may extend into the cavernous sinuses. The Knosp classification is commonly used to grade the extent and the site of invasion.

31.2.2 Pathophysiology

A recent publication from the United States reported that 16.6% of brain tumour arose in the pituitary region. Pituitary tumours are most

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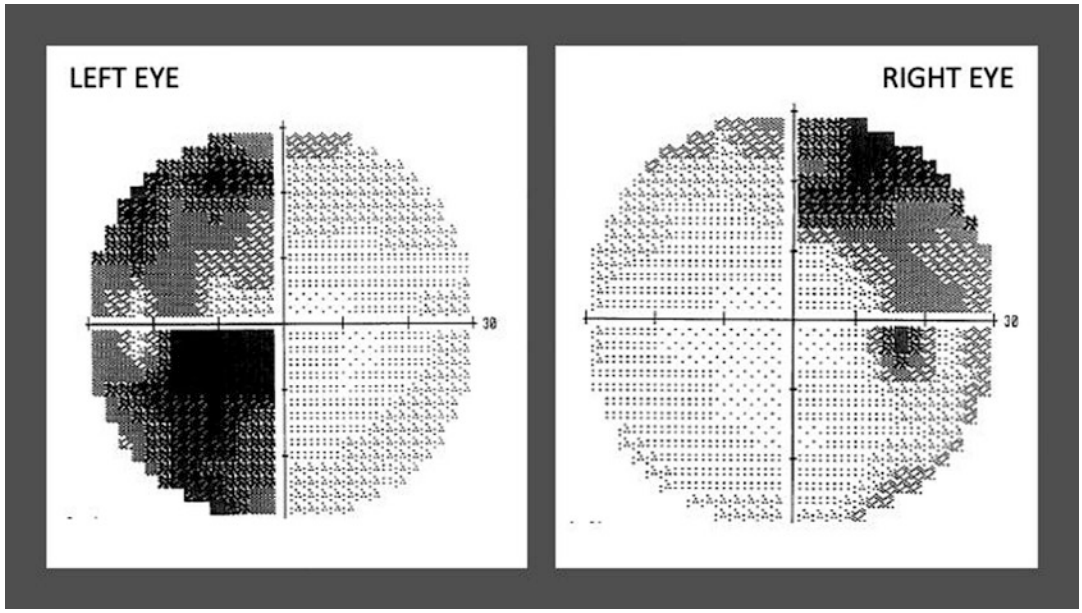

Fig. 31.1 Visual field assessment at baseline

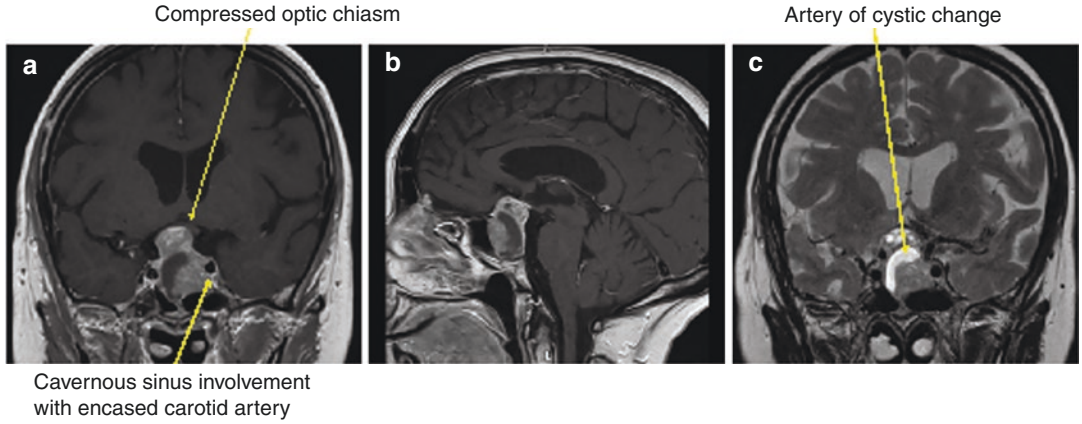

Fig. 31.2 (**a**) Coronal contrast-enhanced T1 highlighting compression of optic chiasm and cavernous sinus involvement, (**b**) sagittal contrast-enhanced T1, (**c**) coronal T2 highlighting cystic area

commonly adenomas, and those that are clinically relevant have been shown in cross-sectional epidemiological studies to be prevalent in around 78–94 per 100,000 population. The most common subtype of adenoma in 57–66% is a prolactinoma, followed by a non-functioning pituitary adenoma (NFPA) in 15–28%. NFPA, or *silent* adenomas, are so-called because they do not result in hypersecretion of pituitary hormones. Due to their non-functional nature, they are usually discovered either incidentally or once they are large enough to cause symptoms from local mass effect. Large series of treated NFPAs have shown that 92% are macroadenomas (>10 mm) at presentation, with a mean size of 23–25 mm. Because of being diagnosed later, patients also tend to be older (mean age 65) than for other pituitary tumours such as prolactinomas (mean age 49).

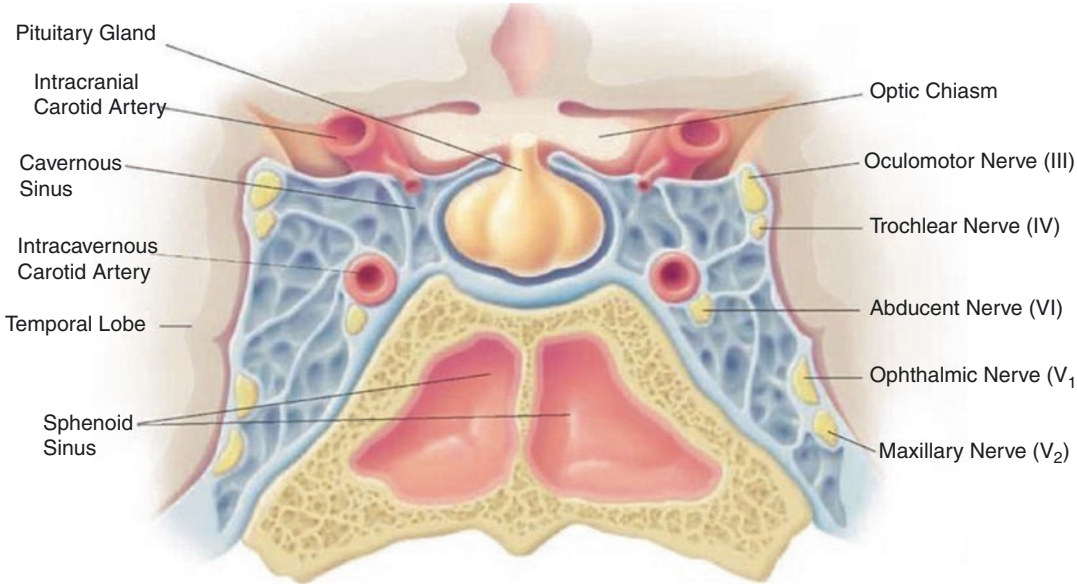

Fig. 31.3 Diagrammatic representation of pituitary gland with superior optic chiasm and inferior sphenoid bone and sinus (ref http://what-when-how.com/acp-medicine/pituitary-part-1/)

31.2.3 Histology

Pituitary tumours are categorised by the World Health Organisation (WHO) according to their cell lineage, which is determined on immunohistochemistry by pituitary transcription factors and adenohypophyseal hormone expression including; prolactin (PRL), growth hormone (GH), follicle-stimulating hormone (FSH), luteinizing hormone (LH), thyroid-stimulating hormone (TSH) and adrenocorticotroph hormone (ACTH). Very few NFPAs contain no hormone staining and are determined as null-cell tumours (2%), all other NFPAs are "silent", with hormone staining despite no hypersecretion. Most common, around 80%, are gondotrophs (FSH, LH) followed by corticotrophs (ACTH) in 15%, with the latter behaving more aggressively with invasion and recurrence. The remaining hormones are rare and occasionally hormones can occur in combination, further to this staining for Ki-67 is important to help assess proliferation, with high levels associated with tumour recurrence.

31.3 Clinical Approach

31.3.1 Diagnosis

Presenting symptoms most commonly include visual disturbance and/or headache, followed by symptoms of reduced pituitary function (low libido, irregular menses, galactorrhoea and impotence) and more rarely ophthalmoplegia and pituitary apoplexy. Formal visual assessment with acuity and Goldman visual fields is essential, as field deficits can occur in up to 75% and acuity in 55%, but this may not be reported by the patient. Visual deficits are common in NFPAs both due to the size of the tumour at presentation and because of a preferential growth pattern in the suprasellar direction, rather than infrasellar growth seen with some other pituitary tumours.

Baseline endocrine testing is also essential as anterior pituitary deficiencies are found in 41–61% of NFPA patients. This includes panhypopituitarism in 7–20% or most commonly

gonadotropin or thyrotropin deficits. Mildly raised prolactin may also be present due to mass effect on the pituitary stalk, which reduces following surgery.

31.3.2 Treatment Algorithm

Firstline treatment for symptomatic or growing NFPAs is transsphenoidal surgery, or observation for microadenomas and incidental lesions, as only 10% of these ever grow. Medical treatment is often employed for functioning pituitary adenomas, particularly prolactinomas which are highly responsive to dopamine agonists and for some GH secreting adenomas which can respond to somatostatin analogues. However, for NFPA, hormone hypersecretion is not a concern and medical treatment is not recommended. Radiotherapy is only used as a primary treatment for patients who cannot tolerate surgery, but can be used as an adjunct to surgery with the aim of reducing tumour recurrence. This may be better targeted at more aggressive NFPAs, such as those that are larger, have cavernous sinus invasion or have concerning MRI characteristics such as low apparent diffusion coefficient value/ratio.

Delayed secondary treatment can also be used following surgery if there is incomplete excision or recurrent growth. Retrospective studies on NFPAs report good rates of resection, with around 20% of patients with residual tumour and a median tumour reduction of 80%. Delayed secondary surgery or radiotherapy is usually required for tumours that were larger at presentation with suprasellar involvement and those with a post-operative remnant that shows early growth on serial imaging. Reported rates of secondary treatment are between 22 and 24% of patients, either with irradiation (13–16%) or repeat surgery (6–9%). The introduction of stereotactic radiosurgery may be more appealing than conventional radiotherapy as equivalent tumour growth control can be obtained, with a potential lower side-effect profile.

31.3.3 Surgical Treatment and Post-operative Care

Transsphenoidal surgery can be performed using a microscope or endoscope, with a transition to the latter technique becoming more popular. In 2017 a meta-analysis of 23 studies summarised that endoscopic surgery resulted in higher rates of tumour resection with no increased complications, likely due to better lateral and suprasellar visualization. A recent trial has also shown that even less experienced surgeons using an endoscope for NFPAs had similar tumour resection rates than for microscopic surgery, with fewer complications, supporting a transition to this technique.

This patient had an endoscopic transsphenoidal (ETS) resection of the pituitary adenoma using a rigid 0° scope and a binostril approach. A pre-operative CT (see Fig. 31.4) allows assessment of pneumatization of the sphenoid air sinus, which can be conchal (2%), presellar (21%), sellar (55%) or postsellar (22%). Assessment of the septations within the sphenoid sinus also aids surgical planning. The CT is used for intra-operative neuronavigation and confirmation of landmarks.

The operative technique involves lateralization of the middle turbinates and removal of the posterior part of the septum (perpendicular plate)

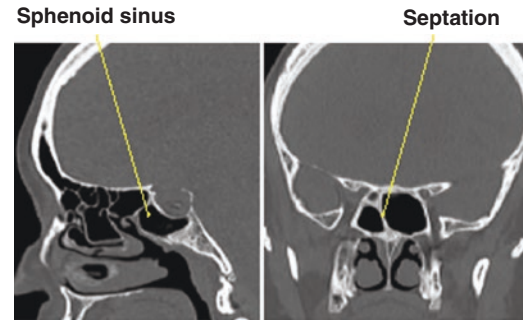

Fig. 31.4 Pre-operative CT showing sellar pneumatization of the sphenoid air sinus (left) and a single sphenoid septum (right)

to allow access to the sphenoid ostia. A rescue flap is always created so that this can be extended into a nasoseptal flap if necessary. An anterior sphenoidotomy is then performed with a high-speed drill. Bony septations within the sphenoid sinus are also removed with a high speed drill if necessary. Care is taken not to fracture these as they frequently extend towards one or both carotid arteries. The anterior wall of the sella turcica and surrounding anatomical landmarks are then confirmed with neuronavigation prior to drilling a small opening to access the pituitary fossa. Following dural opening the pituitary tumour is removed piecemeal with curettes and microdissectors until the arachnoid comes into view. A 30° scope may be used to inspect and resect tumour in the lateral recesses and suprasellar region. There are many closure techniques, the authors prefer an onlay synthetic patch (Lyoplant®) and a fibrin glue (DuraSeal®), unless there is a CSF leak in which case a fat graft is used ± nasoseptal flap. Other options include fascia lata graft. There are reports of using a titanium mesh to rebuild the sellar floor which may potentially reduce the rate of post-operative CSF leak. The sphenoid sinus and posterior nasal space are filled with absorbable nasal packs and the choana evacuated of blood. Finally the middle turbinates are moved back to their anatomical position.

Post-operative complications can be classified as:

- **Visual**: worsening or failure to improve visual fields or acuity, diplopia
- **Endocrine**: new onset diabetes insipidus (DI) or hyponatraemia, failure to improve pre-operative dysfunction or new panhypopituitarism
- **Operative**: CSF leak, meningitis, vascular complications and prolonged hyposmia/anosmia.
- **General**: those that can occur with any surgery (e.g. deep vein thrombosis)

Visual complications after ETS for NFPA are rare with only 3% of patients in a series of 300 reporting permanent worsening of vision, often related to post-operative haematoma. In patients with pre-operative deficits, 42% had complete recovery and 45% partial recovery. Visual field defects have been reported to be more likely to improve (44% return to normal) than loss of acuity (27% return to normal) and both should be tested routinely post-operatively.

There is wide variation in reported improvements in endocrine function post-operatively, with many surgeons anticipating no improvement in patients with pre-operative dysfunction. However, some cases have reported between 32 and 55% of patients showing some improved function over months to years. Up to 23% of patients with previously normal function will have new post-operative dysfunction. Most critically, hydrocortisone must be continued in the early post-operative period until cortisol response is fully tested. Our routine practice is 100 mg at induction of anaesthesia, 20 mg, 10 mg, 10 mg on post operative day 1, reduced to 10 mg, 5 mg, 5 mg thereafter. Morning cortisol levels can be checked on day 3 post-operatively to determine need for on-going treatment. Daily urine output, osmolality and serum sodium must also be monitored, as there is a risk of hyponatraemia (usually due to Syndrome of Inappropriate ADH) or DI within the early post-operative period. We also recommend a repeat sodium after 1 week, as hyponatraemia is the most common cause of re-admission after surgery. Routine fluid restriction may reduce the risk of this.

Intra-operative CSF leaks can occur commonly (up to 35% in one series of 300 ETS), but sound repair means they rarely occur post-operatively (only 3% in the same series). The latter is related to a risk of meningitis, which can be fatal if not promptly and adequately treated.

31.3.4 Follow-up

Post-operative MRI imaging is undertaken to assess the extent of resection, unless there is clinical concern this is delayed 3–4 months to allow

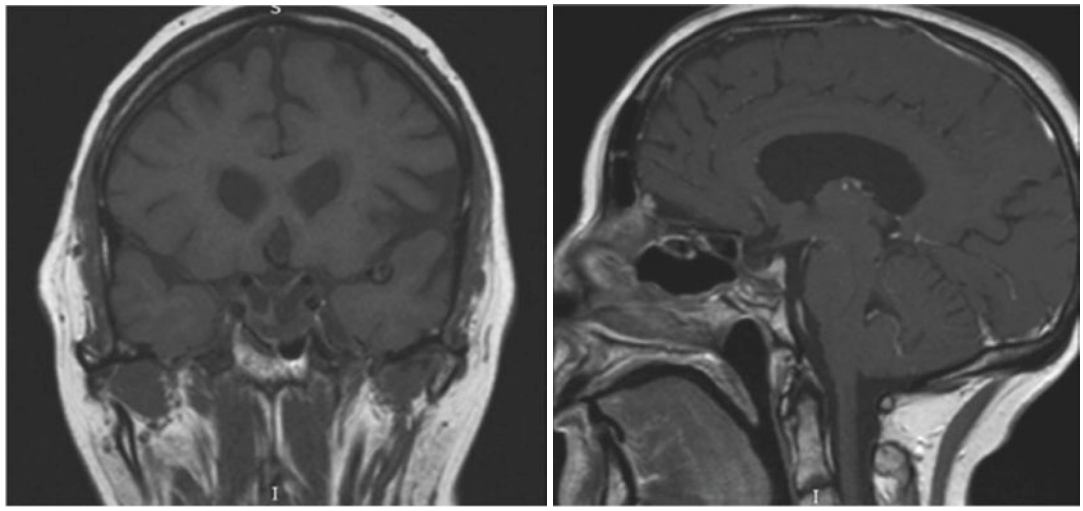

Fig. 31.5 Post-operative MRI

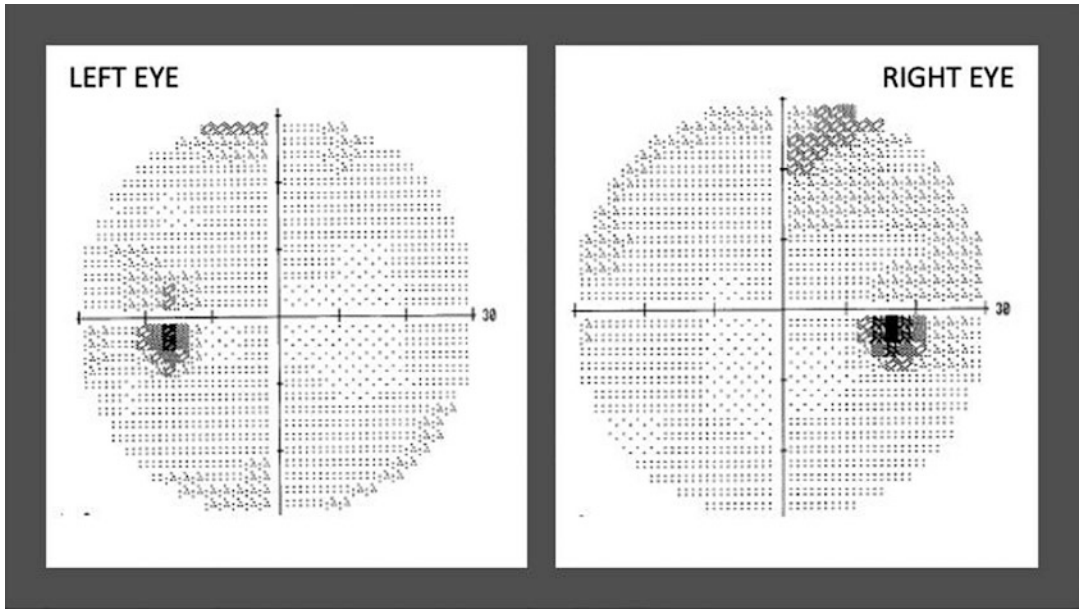

Fig. 31.6 Post-operative visual fields

early post-operative changes (blood/debris/packing) to resolve. In this case good resection of the NFPA with complete decompression of the optic chiasm (Fig. 31.5) and improvement in the vision was achieved (Fig. 31.6). Endocrine function was normal post-operatively. Optical coherence tomography can be helpful in predicting recovery

Summary and Author's Comments

1. NFPAs often present late as macroadenomas (>10 mm) with visual deterioration, which may not be recognised by the patient but must be clinically assessed.

2. Many NFPA patients often have endocrine dysfunction due to undersecretion of hormones, particularly gondaotropins (FSH/LH) and thyrotropin (TSH).
3. First line treatment for symptomatic NFPA is surgical resection with the endoscopic transsphenoidal approach. It is essential that a prolactinoma has been excluded first.
4. Routine prescription of cortisol and assessment of fluid balance and sodium are essential in the early post-operative period to avoid complications from endocrine dysfunction.

of vision. This patient will continue to be followed up by the neuroendocrinology MDT with annual surveillance imaging to assess for tumour re-growth.

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Endoscopic Resection of Clival Chordoma

32

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32.1 Case Presentation

This patient presented in their fifth decade with a history of non-specific headaches, followed by diplopia. Local scans were organized which demonstrated an expansile clival mass abutting the brainstem (Fig. 32.1). The patient was referred for specialist assessment and treatment. After discussion in the skull base multidisciplinary team meeting, the patient consented for endoscopic resection (Fig. 32.2), with histology confirming the diagnosis of clival chordoma. Post-operatively the patient underwent proton beam radiotherapy.

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32.2 Background Knowledge

Chordomas are rare tumours of notochordal origin with dural epithelial/mesenchymal differentiation that usually arise from within bone along the craniospinal axis. They can be known to have slow growing indolent behavior but are locally aggressive and can metastasize. Management is challenging due to recurrence and best treatment is still debated. A third of chordomas arise from the clivus of the skull base with other regions including sacrum (50%) and rarely cervical spine (3–7%). The incidence of chordomas is 0.08 per 100,000 with a male preponderance. They can occur at any age but patients primarily present in the 4th–6th decade of life.

32.2.1 Anatomy

The clivus is the sloping midline surface consisting of the ventral part of the occipital bone, separating the nasopharynx from the posterior cranial fossa. Located at the centre of the skull base, it lies anterior to the foramen magnum and posterior to the dorsum sellae. Craniocaudally the clivus was classified by Rhoton into thirds:

M. Stavrakas, H. S Khalil (eds.), *Rhinology and Anterior Skull Base Surgery*,
https://doi.org/10.1007/978-3-030-66865-5_32

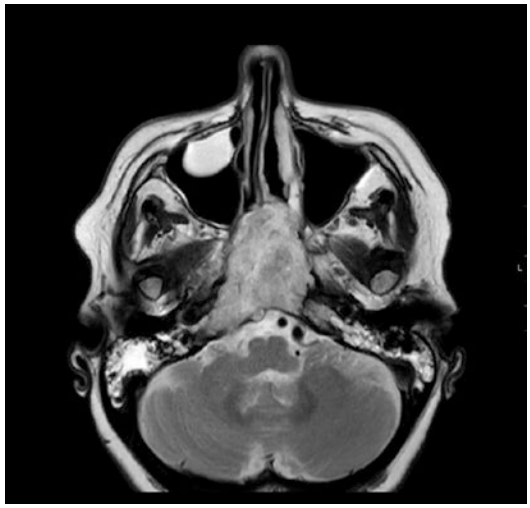

Fig. 32.1 Pre-operative axial T2 MRI scan demonstrating significant expansile clival mass in keeping with chordoma

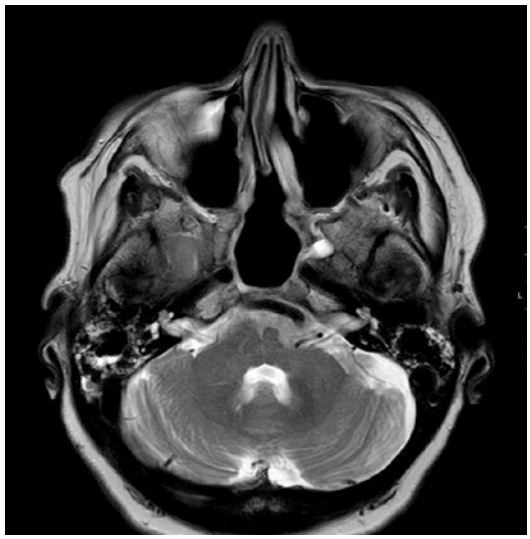

Fig. 32.2 Post-operative axial T2 MRI scan after endoscopic resection. The patient went on to undergo proton beam radiotherapy

- Upper—Posterior clinoid and dorsum sellae to sellar floor
- Middle—Upper boundary to the level of the floor of the sphenoid sinus
- Lower—Middle boundary to foramen magnum.

Lesions associated with the clivus develop adjacent to critical structures including the pons, basilar artery and cranial nerves III to XII. Such a location provides challenges in the treatment of chordomas, with existing treatments of surgery and radiotherapy carrying significant risk of damage to surrounding critical neurovascular structures.

32.2.2 Pathophysiology

Chordomas derive from the notochord which embryologically differentiate to form the vertebral column and skull base. As a result of this relationship between chordoma formation and the embryonic notochord, these tumours can arise anywhere along the axial skeleton. Most commonly, chordomas arise from the sacrum, clivus, and cervical vertebrae. Clival chordomas represent an important subtype with unique surgical challenges due to their location.

32.2.3 Histology

Chordomas have a characteristic appearance predictably similar to fetal notochordal tissue, with fibroblast-like cells encapsulating epithelioid tumour cells. Histological diagnosis is often made with a combination of microscopic and immunohistochemical features. Under the current World Health Organization (WHO) classification, chordomas can be characterized as either:

- Classic/conventional—most frequent, islands and cords of eosinophilic and clear vacuolated cells basophilic myxoid/mucoid background;
- Chondroid—associated with a better prognosis, identified by the presence of chondroid differentiation;
- Dedifferentiated—rare and associated with a poor prognosis, identified by a prominent mesenchymal component, have a malignant and mesenchymal component;
- Sarcomatoid—epithelioid cells commonly associated with chordomas replaced with spindle cells.

The identification of brachyury transcription factor expression in chordomas has resulted in the development of brachyury immunohistochemistry in clinical practice confirms the diag-

nosis of chordoma. Of note however, brachyury expression is not found in dedifferentiated chordoma.

32.3 Clinical Approach

32.3.1 Diagnosis

Given that it is thought chordomas are often slow growing lesions, often patients report a prolonged history of non-specific non-localizing symptoms when finally presenting with neurological signs such as abducens nerve palsy with diplopia. Subsequent imaging features of clival chordomas include isointense morphology on T1-weighted MRI that enhance with gadolinium hyperintensity on T2-weighted MRI imaging. In addition to the characteristically well demarcated tumour, there can be surrounding lytic bone involvement with erosion.

32.3.2 Medical Treatment

There are currently no licensed medical treatments that can be given with a curative intent in the treatment of chordoma. A systematic review of molecular targeted therapies for chordoma included 33 studies testing the use of PDGFR inhibitors, EGFR inhibitors, VEGFR inhibitors, and mTOR inhibitors. Studies investigating the role of imatinib made up the majority, demonstrating mixed results from predominantly case reports, case series and phase I–II studies. Other treatment modalities included a brachyury vaccine with the results of a phase II study currently awaited. More recently, based on the identification of defective homologous recombination DNA repair genes as a driver of chordoma, the PARP inhibitor olaparib was given to a patient with inoperable sacrococcygeal chordoma.

32.3.3 Indications for Surgery

Treatment is based on primary surgery and adjunctive radiation therapy. Multidisciplinary planning for management of patients with clival chordoma is imperative, ideally in a specialized skull base unit. Full consideration of all treatment modalities must be considered prior to proceeding with surgical excision. Particularly in the planning of extended endonasal approaches, craniocervical stability must be considered where peritumoral bony involvement extends to the occipital condyles.

32.3.4 Surgical Treatment and Post-op Care

The International consensus statement on endoscopic skull base surgery included management of clival chordomas. It recommended endoscopic approach can be utilized for midline clival tumours in centres with sufficient experience in a multidisciplinary setting. It can be used in conjunction with an open approach for certain cases with lateral and lower clivus extension components (Figs. 32.3–32.6). There is modest potential for significant neurovascular complications in endoscopic approaches, but no direct comparisons compared with open approaches. There is decreased surgical morbidity associated with an endoscopic approach as demonstrated in multiple case series.

Care is delivered in specialized units, normally utilizing intraoperative image guidance

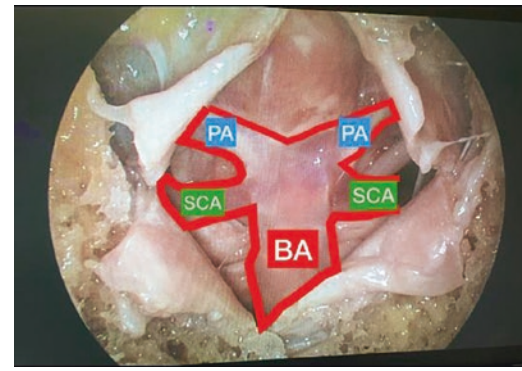

Fig. 32.3 Transclival cadaveric dissection with of basilar artery and branches exposed. PA—Posterior cerebral artery, SCA—Superior cerebellar artery, BA—Basilar artery

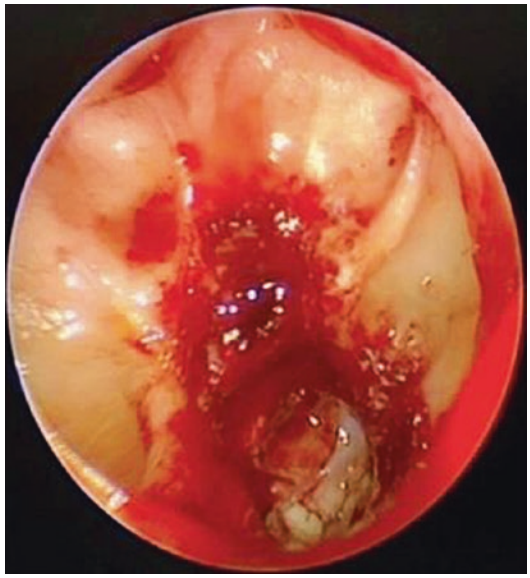

Fig. 32.4 Zoomed out initial endonasal view of chordoma bulk. Note bilateral sphenoidectomy with ample rostrectomy and posterior septectomy

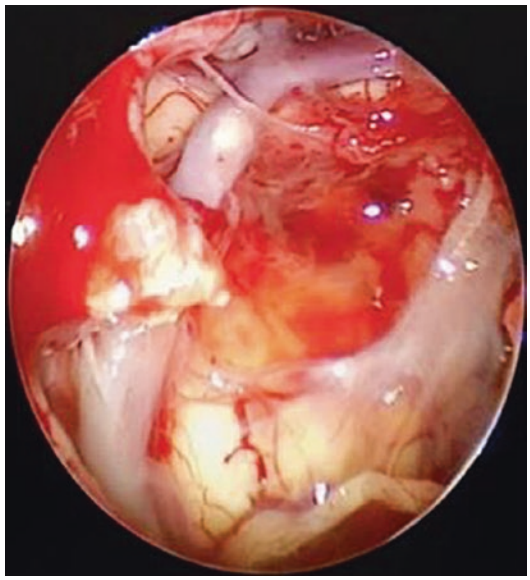

Fig. 32.5 Intraoperative chordoma resection revealing first views of basilar artery

based on preoperative CT and MRI scans. In some institutions, where available, intraoperative MRI scanning can be utilized to assess tumour resection in real time. Surgery is divided into three important phases: Tumour exposure, Tumour resection and Skull Base reconstruction. When employing skull base reconstruction it is

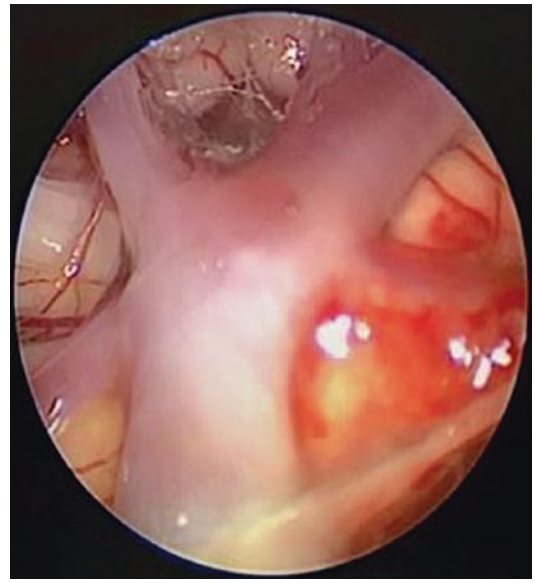

Fig. 32.6 Intraoperative view of basilar artery and terminal branches

vital to appreciate that this is a high flow CSF leak site and multi layer closure is advised. Many graft materials can be used to achieve this, however the most critical factor is surgical technique. Graft options include autografts (e.g. abdominal fat, tensor fascia lata), allografts (e.g. Alloderm®), xenografts (e.g. Biodesign®) as well as a number of commercial products (such as Duragen®). The use of vascularised pedicled flaps, such as the nasoseptal have markedly reduced post operative CSF leak rates. An exemplary and fastidious repair must be employed and we suggest on table valsava manouvre to confirm any residual leak post repair, which can be addressed intraoperatively. The authors' preference is for mutli layer reconstruction utilizing fat, fascia lata and nasoseptal flap repair where possible.

Endoscopic Images: Cadaveric and Intraoperative

A surgical classification of chordomas by Al-Mefty and Borba in 1997 includes:

- Type 1—Tumours limited to one compartment
- Type 2—Tumour involving two or more continuous areas yet requiring only one surgical procedure
- Type 3—Tumour extending into several components requiring more than one surgical procedure.

There may be a role for intraoperative MRI to aid resection. Metwali et al (2019) showed that it aided intraoperative assessment in volume, location and degree of resection. Future surgical advancements may include further minimally invasive techniques. A transpalatal-transoral robotic approach has been described by Henry et al (2019) but this is by no means common/standard practice.

32.3.5 Follow-Up

Tumour recurrence can be high despite total surgical resection due to the tumour location and the nature of chordomas. Post-operative radiotherapy is therefore advantageous to support long term tumour control. Combined treatment with surgery compared to radiotherapy alone is beneficial as it allows decompression of adjacent structures reducing the radiotherapy dose to normal tissues and therefore reducing morbidity. Consequently, global consensus is that radiotherapy should be considered for all clival chordoma if margins of less than 1mm are identified in the macroscopic tumour. Radiotherapy can be given in the form of Intensity-modulated radiation therapy (IMRT) but preferably proton beam radiotherapy which gives a tighter dose delivery.

Summary and Author's Comments

1. Clival chordomas are rare tumours that are challenging to treat despite their slow-growing nature due to local bone invasion and location near to critical neurovascular structures
2. Currently there are no medical treatments licensed for the treatment of clival chordoma; primary treatment is surgical resection
3. Endoscopic endonasal approaches are gaining favor due to the ability to avoid brain retraction while achieving excellent visualization of the tumour at close proximity
4. Following surgery, patients can be considered for adjuvant radiotherapy or proton beam therapy, with a favourable prognosis if a complete resection is achieved.

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Chondrosarcoma of the Skull Base

33

Hisham S Khalil and Mihiar Atfeh

33.1 Case Presentation

A 57 year old woman presented with a few months history of predominantly left-sided facial pain, deep-seated headaches and left tinnitus. She was seen in the ENT Outpatients and her initial evaluation was unremarkable. An MRI scan of the head was requested. This revealed a large destructive mass arising from the clivus. A further CT san of the Skull base and an endoscopic biopsy confirmed the lesion to be a Grade II Chondrosarcoma. This was treatd with endoscopic excision and postoperative proton beam therapy. The patient remains free of disease 7 years after completion of the treatment.

33.1.1 History

The patient had developed a gradual onset of deep-seated headaches which increased progressively in severity to the degree that standard analgesia was ineffective. The facial pain was a sense of pressure and more localised on the left side. There was no history of nasal discharge, nasal obstruction, abnormal smell or epistaxis. There were no visual symptoms. The patient also had an intermittent left tinnitus but no hearing loss or balance problems. She had been fit and well with no significant past medical history. She was an ex-smoker.

33.1.2 Examination

The patient looked well. A flexible nasal endoscopy was normal. Cranial nerves were intact with no orbital signs. Examinations of the ears, throat and neck were unremarkable. A pure tone audiogram revealed mild symmetrical presbyacusis.

33.1.3 Investigations

An initial MRI of the head revealed a significant abnormality of the skull base and a subsequent CT angiogram was requested. Both scans (Figs. 33.1, 33.2, 33.3 and 33.4) revealed a Left petrous apex mass extending across petro-occipital fissure to the clivus and causing bony erosion. The mass encased and displaced the Internal Carotid Artery without compression.

A trans-nasal endoscopic biopsy was carried out using CT image guidance. A biopsy was obtained through the right sphenoid sinus and the

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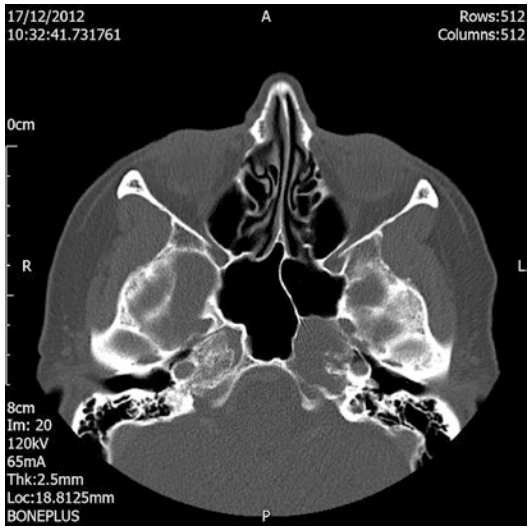

Fig. 33.1 Axial CT with contrast revealing a destructive lesion involving the left petrous apex

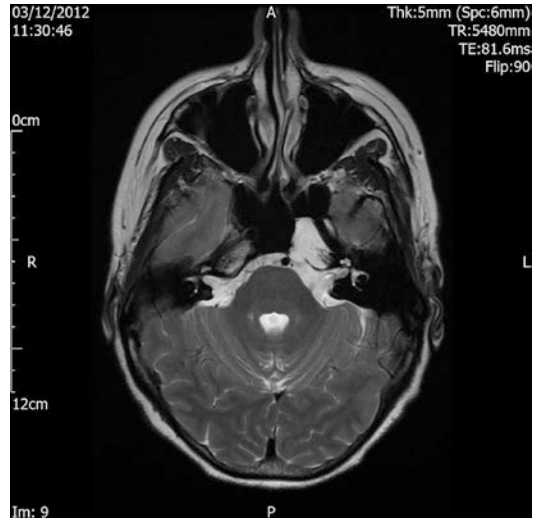

Fig. 33.3 Axial MRI scan with an enhancing lesion in the left petrous apex and left sphenoid sinus and separate from the left vestibulo-cochlear nerve

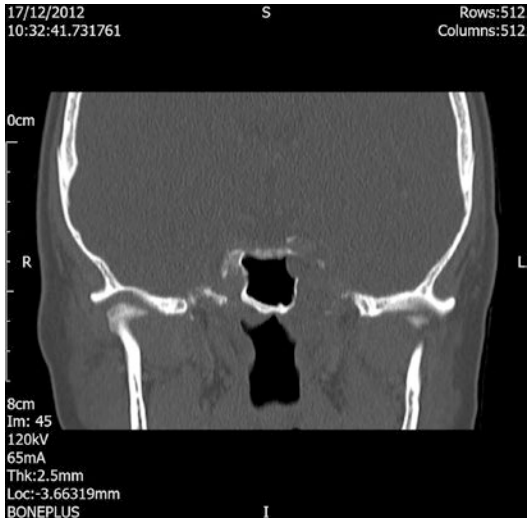

Fig. 33.2 Coronal CT showing a destructive mass involving the left sphenoid sinus and breaching the inter-sinus septum into the right sphenoid sinus

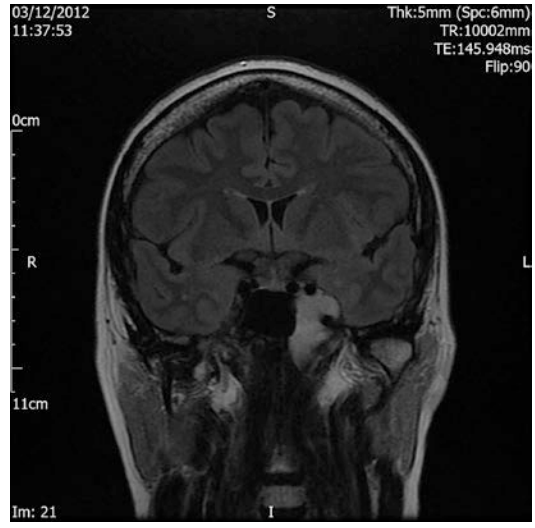

Fig. 33.4 Coronal T1 weighted MRI scan demonstrating a lesion in the left sphenoid sinus wrapped around the left Internal-carotid artery

inter-sinus septum to avoid the left internal carotid artery (Fig. 33.5). The histopathology revealed a Trojani grade II chondrosarcoma.

33.1.4 Treatment

After discussion in the MDT, endoscopic debulking of the tumour was carried out and the patient was referred for Proton Beam Therapy for the residual tumour.

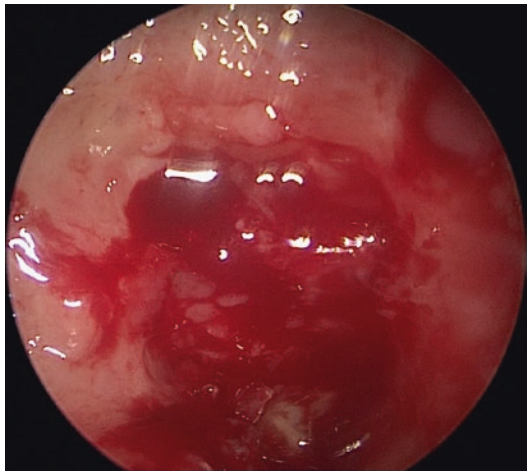

Fig. 33.5 Mass presenting in the right sphenoid sinus through the inter-sinus sphenoid septum

The Proton beam Therapy was delivered as 41 sessions in the USA—5 sessions weekly—45-minute each. Short-term side effects included malaise, skin burns and rhino-sinusitis. Long-term side effects included hair thinning, post nasal drip, crusting and facial pain. The patient continued to have yearly surveillance MRI scans post-treatment. The patient remains free of disease 7 year post-treatment (Fig. 33.6).

33.2 Background Knowledge

Chondrosarcomas (CS) are skeletal tumours that originate from enchondral tissues. They account for approximately 6% of skull base tumours. Common sites include the temporo-occipital junction, parasellar, spheno-ethmoid complex and clivus. They are usually slowly progressive and asymptomatic resulting in a delayed diagnosis.

The delayed diagnosis, extensive local growth, critical tumour location, and proximity to critical structures necessitate complex management and a multidisciplinary approach.

There is a wide range of described treatment strategies in the literature.

Surgical maximum safe excision and neurovascular decompression followed by Proton

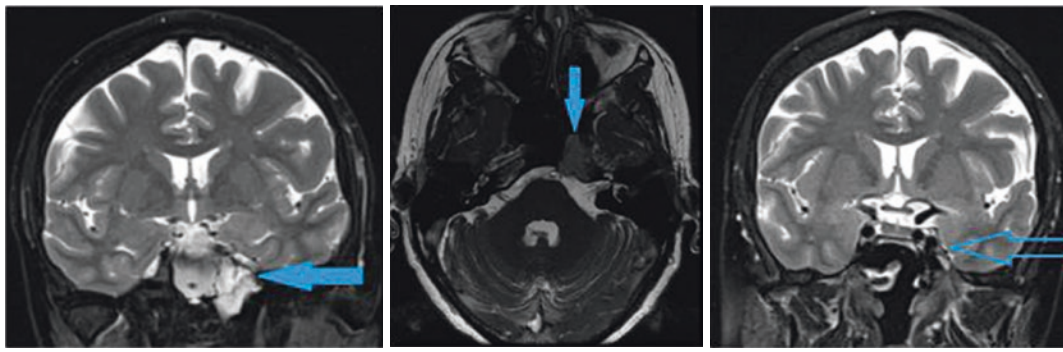

Fig. 33.6 Pre-treatment MRI scan(solid green arrow) and post-treatment MRI scan (hollow green arrow)

Therapy (PT) is considered to be the "gold standard". Complete surgical excision is usually compromised by microscopic residual disease and/or the likely proximity to vital structures.

Proton Beam therapy has become the standard adjunct to surgery in the treatment of CS of skull base due to the greater ability to deliver higher radiation doses to the target tissue while sparing adjacent normal tissues, and therefore avoiding an increasing toxicity.

It also allows improved sparing of adjacent critical structures because of its minimal exit dose after energy deposition in the target and its sharp margins. Proton Beam Therapy has a slightly greater biological effective dose.

Summary and Author's Comments

1. Skull base Chondrosarcomas can vary in their clinical presentation. Presenting symptoms can be misleading and non-specific requiring a low index of suspicion by the clinician if an early diagnosis is to be made.
2. The diagnosis and management planning require thorough assessment, imaging evaluation and a multidisciplinary approach.
3. Safe surgical excision followed by Proton Therapy is the gold standard treatment.

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Anterior Skull Base Meningioma

34

Jason Yuen, Arif Janjua, Peter Gooderham,
Vinay Varadarajan, and Samiul Muquit

34.1 Case Presentation

A 62 year old female was referred with a 4 year history of multiple intracranial meningiomas. She had undergone multiple previous open resections (for lateral sphenoid wing and anterior falcine meningiomas). Her serial MRI scan follow up demonstrated rapid enlargement of her planum sphenoidale lesion (Figs. 34.1 and 34.2).

Due to the rapid enlargement of this lesion, the patient was consented for resection. This was deemed highly suitable for an endonasal endoscopic approach.

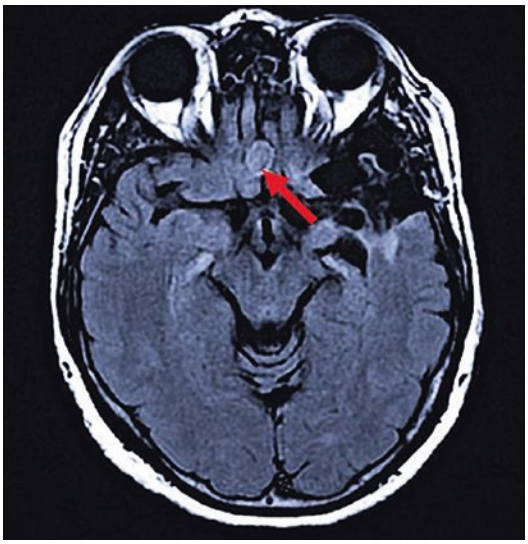

Fig. 34.1 T1 axial MRI scan, demonstrating anatomic approximation of lesion relative to frontal lobes

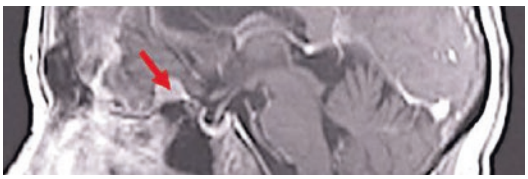

Fig. 34.2 MRI scan (T1 sequence) with contrast—mid-sagittal view. Red arrow denotes lesion

34.2 Background Knowledge

34.2.1 Anatomy

Meningiomas are most commonly found in the convexity (19–34%) and parasagittal locations

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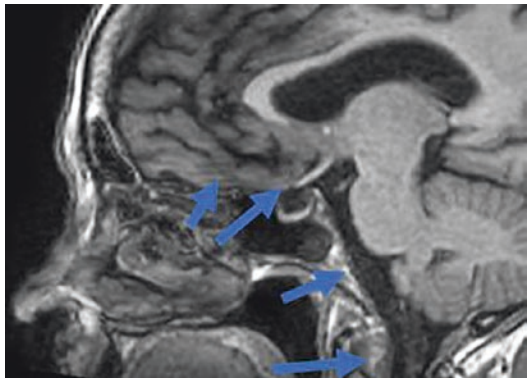

Fig. 34.3 Extended endoscopic approaches (arrows). From left to right: Trans-cribriform; transplanum; trans-clival; trans-odontoid

(18–25%), followed by sphenoid wing and middle cranial fossa (17–25%), anterior skull base (10%), posterior fossa (9–15%), cerebellum (5%) and clivus (<1%).

The most common midline anterior skull base dural attachment of these lesions is the tuberculum sellae (3.6%) followed by the olfactory groove (3.1%). They tend to derive the majority of their vascular supply from the base of the cranium, deep to critical neurovascular structures. Figure 34.3 shows some of the surgical approaches.

Skull base meningiomas can be challenging to treat due to their intimate relationship with important neurovascular structures and relative difficulty in approaching them. Structures such as the frontal lobes and optic apparatus can be vulnerable.

Tuberculum sellae lesions can displace the optic nerves and chiasm with the direction of displacement depending on whether the optic chiasm lies anterior to the chiasmatic sulcus (pre-fixed chiasm, superior displacement) or posterior to the chiasmatic sulcus (post-fixed chiasm, posterior and lateral displacement).

Therefore detailed neuropthalmic examinations as well pre-operative radiological investigations are critical in the assessment and diagnostic period.

34.2.2 Pathophysiology

Meningiomas are thought to arise from arachnoid cap cells, although intra-osseous and intraventricular variants are also known. Risk factors include female gender, older age, history of ionizing radiation exposure, genetic predisposition e.g. neurofibromatosis type 2 and possibly a history of head injury.

34.2.3 Histology

According to a recent report, meningiomas are the commonest primary central nervous system (CNS) tumours (37%) in the USA with an annual incidence of 8.33 per 100,000 population. They are mostly benign tumours (World Health Organization, WHO Grade I; approximately 95%) but there are variants with higher grades (WHO Grades II and III). Skull base meningiomas are less likely to have WHO Grade II and III histology compared to non-skull base ones.

34.3 Clinical Approach

34.3.1 Diagnosis

Patients with anterior skull base meningiomas can present with headache, visual loss (caused by compression of visual apparatus), ophthalmoplegia, mental status changes, endocrine dysfunction (caused by impairment of pituitary gland or stalk), seizure, anosmia, sinusitis, or as an incidental radiological finding. Compared to non-skull base meningiomas, meningiomas involving the skull base are more likely to present with neurological deficits but less often with seizures. Occasionally, they may also present with Foster-Kennedy syndrome, composing of unilateral optic atrophy, unilateral anosmia and contralateral papilloedema. Neuropthalmology and endocrine assessment prior to treatment is essential.

CT scanning often reveals an isodense lesion, which enhances uniformly with contrast, with or without regions of calcifications (hyperdensities). Meningiomas are isointense on T1 MRI scan and enhance uniformly with gadolinium. A "dural tail", enhancement of the dural perimeter surrounding the dural attachment may be seen. It is important to carefully study the relationship of the critical neurovascular structures (such as optic nerves, anterior cerebral arteries and internal carotid artery) to the tumour capsule, which

may be facilitated by the use of magnetic resonance angiography.

34.3.2 Non-Surgical Treatment

Non-surgical treatment may involve short-term steroids treatment in cases with significant mass effect, anti-epileptic medications (prophylactic or secondary), as well as symptomatic treatments such as anti-emetic medications. Serial imaging may be considered in those who are not fit for surgery, for stable or slow growing lesions and in those with a highly calcified meningioma.

Stereotactic radiosurgery (SRS), using either a linear accelerator-based system or the Gamma knife, is considered an effective alternative to surgical removal of small to medium benign meningiomas as well as an adjuvant modality to reduce the risk of tumour progression after subtotal resection. The incidence of cranial nerve deficit is generally low, especially in those with smaller size lesions and those without pre-treatment deficits. Seizure risks vary from 1 to 2%. However, there are case reports of tumour progression in the long-term. In cases where preservation of sense of smell and taste is important, such as for those with particular occupations, SRS may be preferable. Tumour control rate (decreased in volume or remaining the same) is 84–93%. One study showed that tumours with volume < 14 cm^3 were more likely to have effective tumour control.

34.3.3 Indications for Surgery

The indications for surgery are: (1) progressive enlargement with serial imaging, (2) symptoms from mass effect and (3) visual loss.

34.3.4 Surgical Treatment and Post-Op Care

The rate of recurrence of meningioma directly correlates with the extent of resection of the tumour, its dural attachment and if applicable, any affected pathological bone. Hyperostosis is a controversial radiographic finding, with some studies suggesting that histologically, hyperostosis in a skull base tumour is associated with tumour invasion, ranging from 23% to 100% and such involvement is associated with higher recurrence rate. However, such invasion does not represent malignancy as most skull base meningiomas are benign, with some suggestions that the hyperostosic element behaves like a slowly growing tumour.

Pre-operative embolization is sometimes employed as an adjunct in highly vascularized tumours in order to limit surgical blood loss, shorten operative times and aid intra-operative resection. According to a systemic review, the complication rate directly related to tumour embolization was 4.6%. This includes infection, hemiparesis, facial palsy, disseminated intravascular coagulation, glaucoma, tumour swelling, transient SIADH (syndrome of inappropriate antidiuretic hormone secretion), dysphagia, and cranial nerve deficit, and 14% of these are "major" or fatal.

There are generally two approaches to resect anterior skull base meningiomas—transcranial and endoscopic endonasal. Open transcranial approaches for olfactory groove, tuberculum sellae, planum sphenoidale meningiomas have been reported to have higher rates of total resection and lower post-operative CSF leak rates by some authors. However, in certain cases, with careful patient selection and good multi-layer closure technique, endoscopic endonasal approach may be suitable without the morbidities associated with open craniotomies.

Visual improvement may be higher via the endonasal approach than the transcranial approach for tuberculum sellae meningioma (TSM) but not for olfactory groove meningiomas (OGM), although the authors of the systematic review felt that heterogeneity of studies may be a factor. CSF leak was also deemed higher among endonasal patients for both types of meningiomas (25.1% vs. 10.5% for OGM and 19.3% vs. 5.81% for TSM). It is important to appreciate that CSF rates may vary from centre to centre and also on the techniques employed. Mortality was not significantly different between endonasal and transcranial patients.

Some may recommend that an endonasal approach may be more suitable for smaller lesions located primarily in the midline without encasement of critical neurovascular structures. However, again this depends upon the institution and their experience and skills in performing endoscopic endonasal surgery.

34.3.5 Transcranial Approach

Several transcranial surgical approaches exist, such as transbasal and orbitozygomatic (OZ) approaches. Factors influencing which approach to use include the extent of tumour size and the neurovascular structures that are in close approximation. It is important with these approaches is to minimise brain retraction by maximizing removal of relevant bony anatomy.

Endoscopes may be used to improve visualization in open transcranial approaches in the form of "Endoscopic Assisted" open procedures.

34.3.6 Endonasal Approach

The Endonasal approach avoids excessive manipulation of critical neurovascular structures and brain retraction which often accompany transcranial approaches. It is also more cosmetically appealing and is perceived by patients as being less invasive than the transcranial approach. Case series have demonstrated that a purely endoscopic endonasal approach can be undertaken for a wide variety of meningiomas with acceptable morbidity and mortality, as well as a high likelihood of visual improvement. Good command of the neurovascular anatomy viewed from the transnasal endoscopic route is essential with experience normally built within a multi specialty "Endoscopic Skull Base Surgery Team".

The surgical approach is tailored to lesional extent and location. With surgery being compartmentalised into three stages—tumour exposure (Fig. 34.4), tumour resection (Figs. 34.5 and 34.6) and skull base reconstruction (Figs. 34.7, 34.8 and 34.9). Traditionally, this is achieved by a binostril, 2 surgeon, 4-hand technique routinely involving a Rhinologist and a Neurosurgeon.

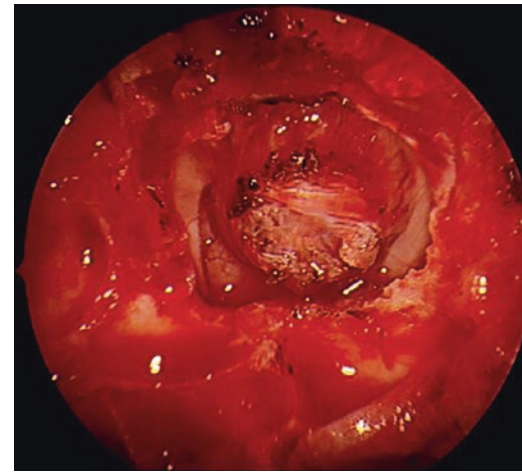

Fig. 34.4 Intra-operative image of trans-planum approach with meningioma clearly seen

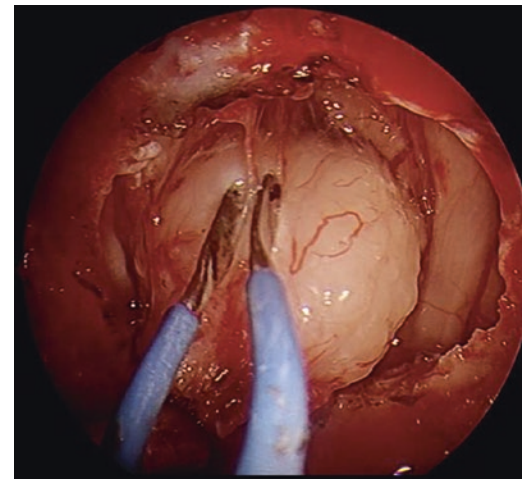

Fig. 34.5 Judicious use of bipolar diathermy during resection

Principles of meningioma resection include early devascularization of the tumour when indicated (including consideration of the anterior and posterior ethmoidal arteries), focused tumour debulking and subsequent detachment of all tumour elements from surrounding structures using careful dissection strategies. Furthermore, angled endoscopes can be useful in tumour dissection as well as ensuring complete clearance of tumour margins. Outcomes are broadly based on case selection and institutional experience; whereas radiological findings such as cortical cuff or brain oedema are less important.

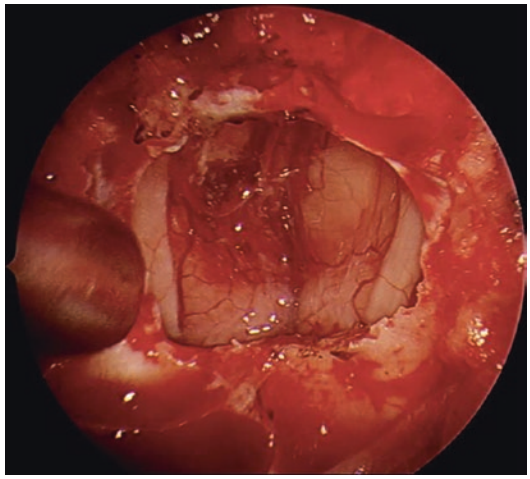

Fig. 34.6 Endoscopic view of frontal lobes after meningioma resection. Note the olfactory nerves are clearly visible on both sides

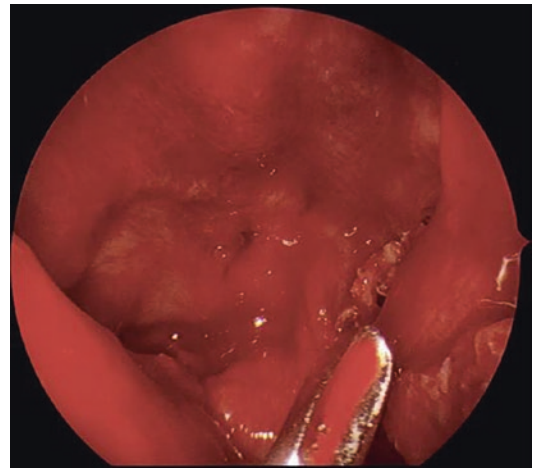

Fig. 34.8 The nasoseptal flap is placed over the intradural inlay and extradural onlay Duragen® grafts

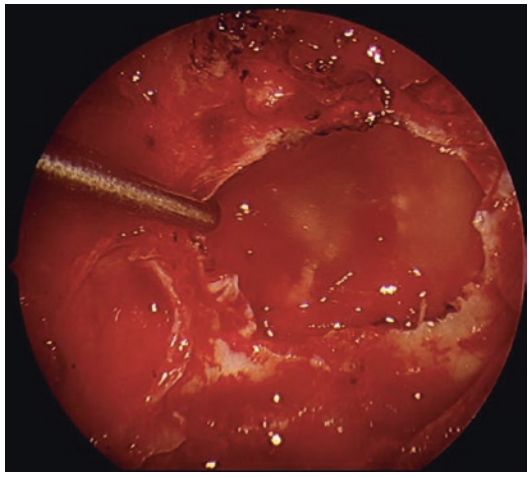

Fig. 34.7 First layer of skull base reconstruction. Intradural Duragen® inlay graft

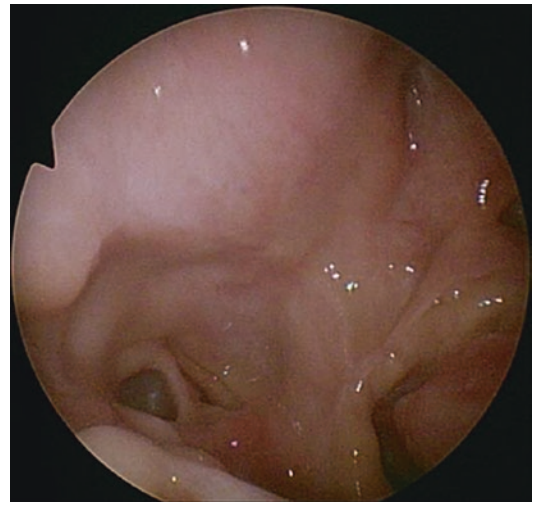

Fig. 34.9 Three months post operative view. Healed nasoseptal flap and skull base repair. *Note: Intraoperative photos supplied courtesy of Dr Arif Janjua and Dr Peter Goodherham, Vancouver General Hospital*

After tumour resection, the skull base is repaired in a multi-layer fashion to reduce the risk of CSF leak. The materials used to achieve this are much less important than the surgical technique which must be fastidious and impeccable. Graft options include autografts (eg abdominal fat, tensor fascia lata), allografts (eg Alloderm®), xenografts (eg Biodesign®) as well as a number of commercial products (such as Duragen®) It is good practice to perform an intraoperative Valsalva manouvre to check for CSF leak and skull base integrity after reconstruction.

Generally reconstructions can be protected and supported using silastic sheeting and a combination of absorbable and non-absorbable nasal packing. Although not essential, some advocate the use of an intranasal balloon catheter (to provide counterpressure) in the early phase of healing, and a lumbar spinal drain to reduce the risk of a subsequent CSF leak.

The use of vascularized pedicled flaps (eg nasoseptal flap) have revolutionized skull base reconstruction and are used routinely (when possible) in skull base reconstruction. It is important that any recipient bone that the flap sits on is fully demucosalised to reduce the CSF leak rate as well as the possibility for future mucocele formation. In this particular case, skull base reconstruction was achieved utilizing an intradural Duragen® inlay graft, followed by a extradural Duragen® onlay graft, followed by a nasoseptal flap. This was secured at the edges utilizing Tisseel® surgical glue, and reinforced using slistastic stenting and Merocel® nasal packing.

34.3.7 Follow Up

Post-operatively, patients are managed with prophylactic antibiotics (as per institutional protocol) and regular monitoring of fluid balance and electrolytes. Furthermore, patients must be regularly assessed for signs of infection and CSF leak. A post-operative scan is obtained to assess the extent of resection and nasal packing is generally removed between Day 3 and Day 5 postoperatively. For difficult residual tumours, or in cases of high-grade histological diagnosis, radiosurgery may be considered after discussion at the Skull Base multidisciplinary team meeting.

Summary and Author's Comments

1. Anterior skull base meningiomas may present with cranial nerve deficits, seizures, mental status changes, or incidentally on radiological studies.
2. Management modalities generally consist of serial surveillance, radiosurgery and surgery.
3. Surgical treatment can be divided into endonasal endoscopic approaches, endoscopic assisted open approaches and purely transcranial approaches.
4. The endoscopic endonasal approach is safe and effective in appropriately selected cases.

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35 Pre-Operative Embolization with Direct Onyx Tumoral Puncture Prior to Endoscopic Resection of Massive Juvenile Nasopharyngeal Angiofibroma

Vinay Varadarajan, Arif Janjua, and Manraj K. S. Heran

35.1 Case Presentation

A 19 year old male college student was referred to our unit with a known large left sided endonasal mass. He was symptomatic for approximately one year with symptoms of clear unilateral nasal discharge, worsening nasal obstruction, weight loss, and epistaxis. On examination, he had a large invasive tumor centered in the left nasal cavity. On imaging, the tumor demonstrated mass effect with extension through the posterior wall of the maxillary sinus and bony destruction of the pterygoid wedge, floor of the sphenoid sinus, and medial floor of the middle fossa bony plate (Fig. 35.1). MRI scanning demonstrated middle

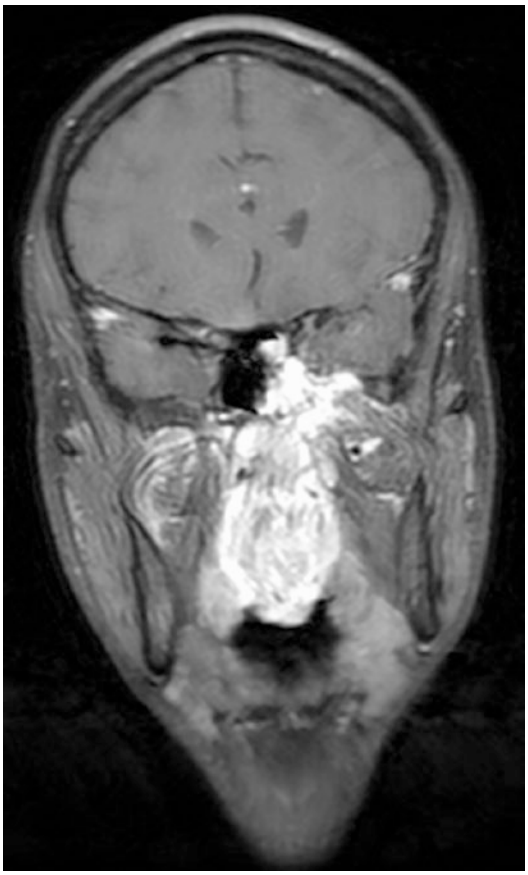

Fig. 35.1 Pre-operative Coronal contrast-enhanced T1 MRI of JNA involving left pterygoid wedge, sphenoid floor, and middle cranial fossa tegmen plate

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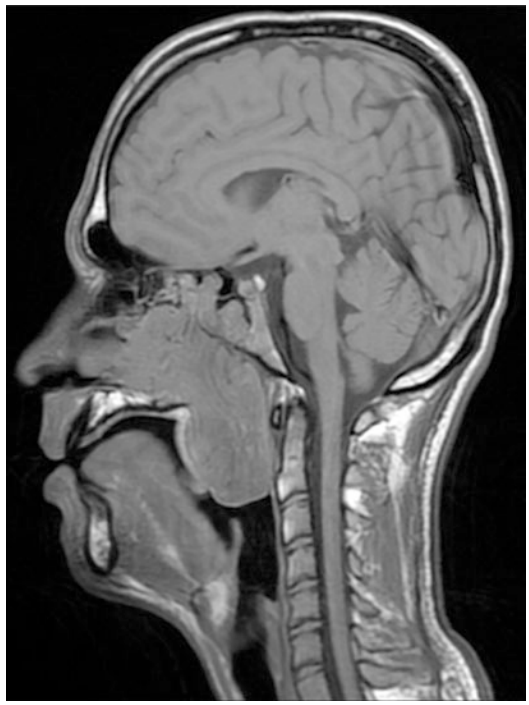

Fig. 35.2 Pre-operative sagittal unenhanced T1 MRI scan demonstrating tumor extent

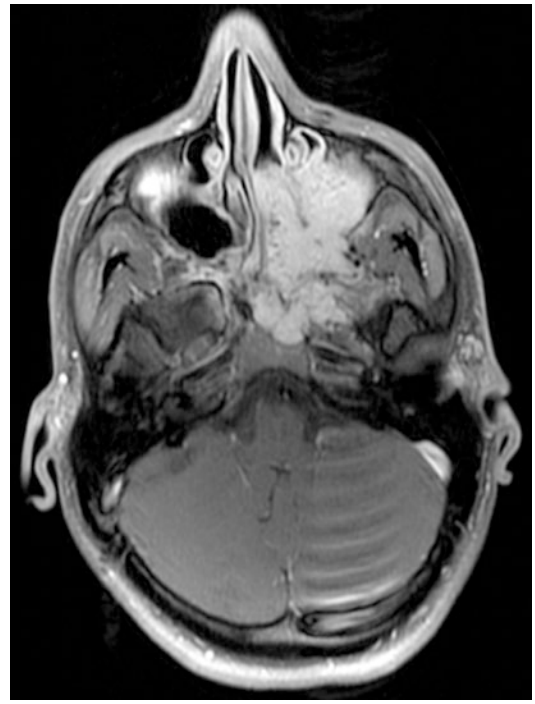

Fig. 35.3 Pre-operative axial contrast-enhanced T1 MRI scan demonstrating tumor extent

cranial fossa involvement, with the tumor abutting the temporal lobe, though no dural involvement was seen. Figures 35.2 and 35.3 are MRI sagittal and axial MRI slices demonstrating tumor extent.

He underwent successful pre-operative embolization followed by complete endoscopic transnasal resection of his tumor. Intraoperative blood loss was less than 300mls.

35.2 Background Knowledge

Juvenile nasopharyngeal angiofibroma (JNA) is a hypervascularized tumor that is locally invasive, benign, and occurs almost exclusively in teenage males. It is approximated to account for 0.5% of all head and neck tumors. Originating from the sphenopalatine foramen, if left untreated, JNA can be locally destructive and invade into the sphenoid sinus, pterygoid bone, infratemporal fossa and middle cranial fossa. The treatment of choice for such lesions is total surgical resection. Resection can either be performed via traditional open approach surgery or also via a fully endoscopic route. In some instances, a combined surgical approach can be helpful or necessary. One critical intraoperative factor in treatment success is removal of all aspects of the tumor to ensure no residuum is left behind. This is largely aided by optimal intraoperative visualization through limiting blood obscuring the surgical field. As would be expected, tumor recurrence rates have been shown to be reduced with reduced intraoperative blood loss.

Pre-operative embolization of these vascular tumors has been proven to reduce blood loss by approximately 45% to 70%. This allows for superior intraoperative visualization of the tumor and therefore a better chance of total resection, as well as reducing the potential need for blood transfusions. Vascular supply to these tumors can be complex. On systematic review, it has been suggested that 35.6% of tumors receive some of their blood supply from the ipsilateral internal

carotid artery and 30.8% of tumors receive bilateral vascular supply. This is mirrored in our own experience with these tumors.

We find that in larger tumors, as well as with those with obvious skull base involvement, there is often direct tumoral supply arising from the internal carotid, on the lesional side. This is typically by the artery of the foramen rotundum and/or the vidian artery. In addition, other unnamed collateral branches may exist. The traditional technique of pre-operative embolization is that of transarterial particle embolization of the internal maxillary artery on the lesional side. However, embolization of this (and other external carotid artery branches supplying the tumour) leaves behind vascularized tumor segments supplied by the internal carotid that can bleed significantly during surgical resection.

We describe the combined use of direct tumoral puncture and intra-lesional embolization with the Onyx ethylene vinyl alcohol copolymer (Medtronic, California) after traditional particle embolization of ECA tumour-supplying branches. This allows for excellent devascularisation of all tumor segments, including the critical parts supplied by the internal carotid artery. Use of this technique has greatly enhanced our ability to perform pre-operative embolization of large JNAs and has made resection of tumor segments near the pterygoid wedge and skull base markedly easier.

35.3 Clinical Approach

MRI enhanced gadolinium sequences, along with patient background factors confirmed the diagnosis of a JNA. It must be noted that in cases like these, radiological diagnosis is sufficient and biopsy for definitive histology is not recommended due to the risk of hemorrhage.

A 6 French guiding catheter was used to perform diagnostic angiography after selective internal carotid and external carotid arterial catheterization bilaterally. The guiding catheter was then positioned in the proximal external carotid artery (ECA), beginning on the lesional side. Angiography was performed to assess the different branches potentially supplying the tumor, as well as to assess for any obvious ECA to ICA anastomoses. Under roadmap technique, we introduced a Penumbra PX Slim microcatheter (Penumbra Inc., Alameda, California) into the internal maxillary artery. Particle embolization of the internal maxillary artery was then performed utilizing 300–500 micron particles (Beadblock), with the microcatheter positioned distal to the origin of the middle meningeal artery. Following this, coil embolization was performed of the ipsilateral side IMA for minimizing arterial recanalization and to aid in vessel sacrifice at the time of endoscopic surgical resection. To prevent the potential head and neck complications of permanent bilateral IMA arterial embolization, initial particle embolization of the sphenopalatine branches of the contralateral IMA was followed by gelatine sponge embolization of the parent artery. This allowed for "temporary" embolization of the contralateral IMA. Periodic ECA angiograms were performed throughout the case to assess tumor vascularity, as well as the presence of other tumoral branches. In addition to the above-described ECA branches, transarterial embolization was also performed of additional ipsilateral vessels, including the anterior division of the ascending pharyngeal artery, a hypertrophied accessory meningeal branch of the middle meningeal artery, as well as a prominent branch arising from the facial artery.

The above transarterial embolization was further aided by trans arterial Onyx embolization in the lesional side sphenopalatine branches, as well as in a hypertrophied branch arising from the middle meningeal artery.

Common, ECA and ICA angiograms were obtained to identify residual tumoural supply, as well as to assess the location and degree of tumor blush for any residual hypervascular areas of the tumor, with particular attention paid to the supply derived from the ICA. Direct tumoral supply arising from the internal carotid artery on the lesional side was identified from the artery of the Foramen Rotundum as well as the Vidian artery. The tumor segments supplied by these ICA vessels were identified, and a trans nasal approach was used to percutaneously access these areas with a 22-guage

Fig. 35.4 Trans nasal approach for Direct Tumor Puncture utilizing a 22-gauge 3–1/2″ spinal needle

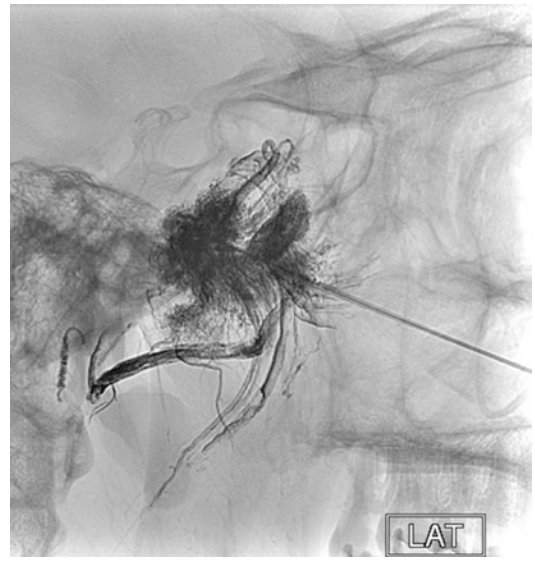

Fig. 35.5 Needle in position for percutaneous intratumoral Onyx embolization. Note the significant amount of tumor vascularization/tumor blush despite prior transarterial embolization of the ECA supply

3–1/2″ spinal needle using a combination of needle-guidance technology on the angiography unit, and roadmapping fluoroscopy (Fig. 35.4). Needle position was confirmed upon intralesional contrast injection performed under biplane angiography. DMSO was used to prime the dead space of the needle and Onyx was then administered intralesionally for embolization of the target areas (Fig. 35.5). Monitoring angiograms were used constantly to insure a margin of safety between the region being embolized and the parent ICA, in order to avoid non-target migration of embolic material. Slow and intermittent injection allowed for the necessary control to prevent the Onyx liquid embolic from migrating into the ICA. Post-embolization angiography confirmed near-complete tumor devascularization (Fig. 35.6). Surgical resection of the tumour was then performed (Figs. 35.7 and 35.8).

Complete tumour excision was achieved endoscopically, highly facilitated by the excellent pre-operative Onyx embolization. The tumour was able to be resected with utilisation of the microdebrider along with cold steel endoscopic instrumentation, with minimal use of endoscopic drilling. In general, massive tumors such as this,

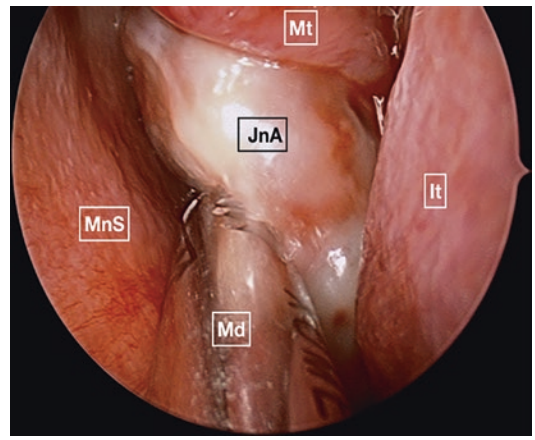

Fig. 35.6 Intraoperative image of left nasal cavity. The tumor has been so well embolized that we are able to perform resection with a microdebrider. MnS *Mid nasal septum, JnA* Juvenile nasal angiofibroma, *Md* Microdebrider, *It* Inferior turbinate

have already performed a large amount of "approach work" due to tissue destruction caused by tumour growth. Oftentimes there may be multiple attachment sites which must be addressed. If not addressed tumor recurrence may occur, one common site is at the vidian canal. In this case the tumour had invaded the pterygoid wedge with

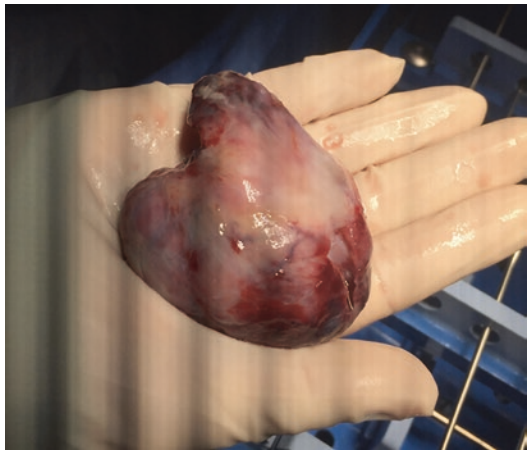

Fig. 35.7 The nasopharyngeal and oropharyngeal component of the tumor has been excised and delivered via the oropharynx and oral cavity

Summary and Author's Comments

1. Large JNAs involving the skull base often are supplied by multiple vessels including those of the internal carotid artery
2. These cases can be resected endoscopically, however excellent pre-operative embolization has been shown to reduce intraoperative bleeding, helping to improve surgical visualization
3. Use of direct tumoral puncture with Onyx aids with devascularizing tumor segments supplied by the internal carotid artery and therefore can be indispensible in cases such as this

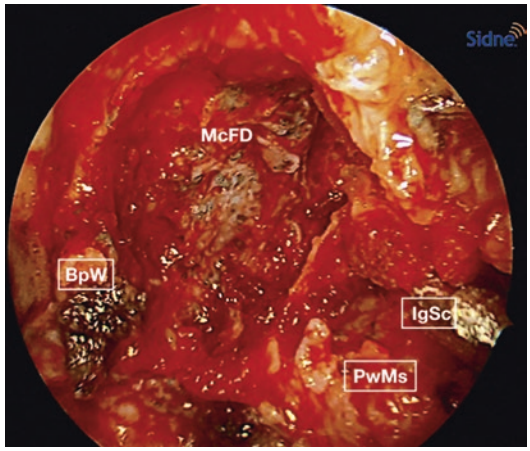

Fig. 35.8 Intraoperative image of left nasal cavity. The tumor has been fully resected endoscopically with excellent hemostasis provided by the preoperative embolization. Note the small dark flecks of Onyx embolic material abutting the middle cranial fossa dura but not penetrating it. This marks excellent pre-operative tumor devascularization. *BpW* Bony wedge of pterygoid, *IgSc* image guidance suction catheter, *McFD* Middle cranial fossa dura, *PwMs* Posterior wall of maxillary sinus

infiltraction of the infratemporal fossa, therefore a coronal plane approach had to be instituted. In addition, in large tumors such as this the tumour can be resected in "segments". In this case the first segment of tumour was resected and delivered via the oropharynx (Figs. 35.7 and 35.8).

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Part VI

Sinonasal Tumours

Inverted Papilloma

36

Mohammed Salem, Marios Stavrakas, and Hisham S Khalil

36.1 Case Presentation

A 51-year-old male patient was referred to the Rhinology clinic with an eight-week history of left-sided recurrent epistaxis and progressive nasal obstruction. He did not complain of hyposmia, mucopurulent rhinorrhea, headache, or any visual impairment. The patient was previously treated with intranasal steroid sprays, which did not improve his symptomatology. He was otherwise fit and well. After full head and neck examination, including flexible nasendoscopy, the patient underwent cross-sectional imaging with CT (Fig. 36.1) and MRI scans. A biopsy was taken and revealed inverted papilloma (IP). Eventually, he underwent endoscopic sinus surgery in the form of an endoscopic medial maxillectomy and is now 5 years disease-free.

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36.2 Background Knowledge

Inverted papilloma is a benign epithelial tumour extending into the underlying stroma of the nasal cavity and paranasal sinus, and originates from the Schneiderian respiratory membrane. It accounts for 0.5–4% of primary sinonasal tumours. Etiologically, it is most likely related to HPV infection, as studies have found HPV DNA in both the IP and cells of adjoining mucosa with normal appearance. Therefore, removal of the adjoining predisposed mucosa, with a normal appearance at the attachment site, may reduce the recurrence rate. IP has a high rate of recurrence, especially when the patient is undergoing revision surgery (14–78%). It also has the potential for malignant transformation, with studies describing a risk up to 11%.

The typical presentation is unilateral nasal obstruction. Less common symptoms may include epistaxis, bloody nasal discharge, headache, facial pain, smell disorders and epiphora. IP pre-surgical workup includes head and neck examination, nasendoscopy, CT and MRI scans and biopsy. There are various classification systems but Krouse classification is most commonly used in the literature (Table 36.1).

The mainstay treatment is surgical resection. Nowadays, less invasive endoscopic approaches have replaced the aggressive external approaches e.g. lateral rhinotomy, mid-facial degloving or medial maxillectomy. Imaging and clinical

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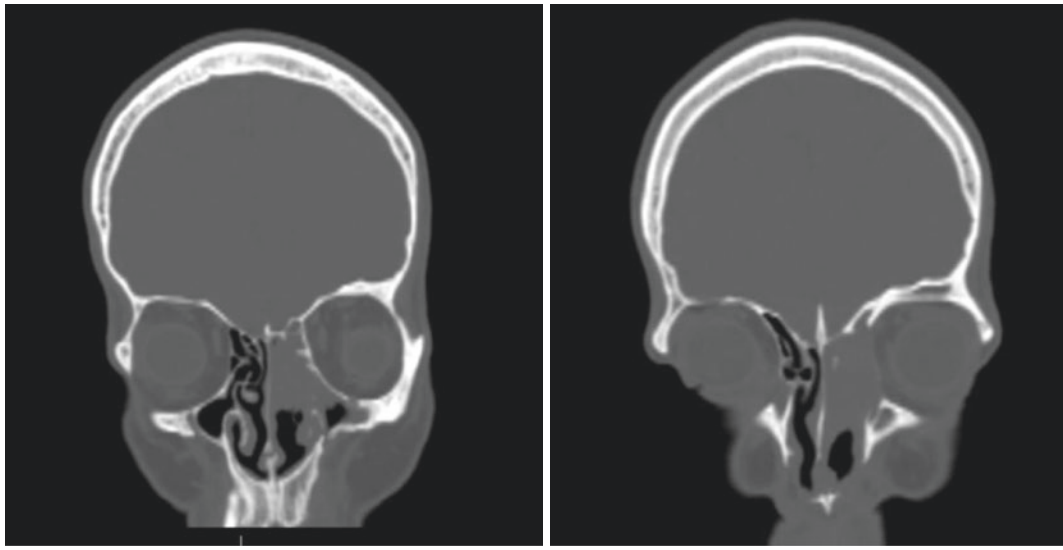

Fig. 36.1 Coronal CT scans of the patient's sinuses

Table 36.1 Krouse classification system for inverted papillomas

Stage T1	Confined to the nasal cavity
Stage T2	Involves ostiomeatal complex region, ethmoid, or medial wall of the maxillary sinus
Stage T3	Involves any wall of the maxillary sinus but medial, frontal sinus, or sphenoid sinus
Stage T4	Any extranasal or extrasinus extension or presence of a malignant neoplasm

assessment allow for estimation of the extent of the disease and selection of the most appropriate approach (e.g. medial maxillectomy, pre-lacrimal approach, endoscopic Denker's procedure, Draf II/III, combined approach to the frontal sinus). Radiation therapy may be considered in cases of associated carcinoma or inoperable cases.

Some of the challenges that the surgeon may encounter have to do with recurrent disease and difficulties in addressing the site of origin. It is accepted that recurrence may have to do with incomplete resection of the tumour, with healthy margins, during primary surgery. Also, recurrence rates may be associated with the stage of the primary disease. Lisan et al. (2017) conducted a meta-analysis and concluded that T3 disease was at significantly higher risk of recurrence when compared with stage T2. No differences in recurrence rates were found between stages T1 and T2 disease or between stages T3 and T4. Adriaensen et al. (2015) advocate that when the size and extent of the tumour permits, en-block resection with a cuff of macroscopically healthy mucosa is adviceable. The bone underlying the attachment can be drilled using a diamond drill. In areas where the bone was too thin, such as the lateral lamella or the cribriform plate, gentle coagulation is an option. The same study has evaluated the topical use of 5-fluorouracil (5-FU) in the postoperative management of inverted papilloma. The authors concluded that there is place for the topical use of 5-FU as adjuvant therapy, especially in recurrent cases.

Close follow up is mandatory, especially in the first years when most recurrences occur. As there is evidence of recurrence or malignant transformation beyond 5 years, many centres, including our unit, have changed their practice and prefer lifelong follow up. In our unit, we see the patients every 2 months initially for clinical examination and nasendoscopy, then the follow up intervals are longer and eventually the patients have yearly follow up. MRI scan has also a place in postoperative monitoring.

36.2.1 Clinical Approach

36.2.1.1 History

It is essential to approach such patients with persistent unilateral symptoms with a high index of suspicion. The clinician's primary aim should be the exclusion of malignancy. Red flag symptoms such as unilateral recurrent epistaxis, nasal obstruction, anosmia, epiphora, headaches, visual disturbance, numbness at the distribution of V2, warrant detailed head and neck examination and further diagnostic workup. Also, we should always keep in mind the patient's occupation (exposure to organic solvents or welding fumes), history of previous sinonasal surgery and other risk factors.

36.2.1.2 Examination

Initial ENT assessment, including fiberoptic nasendoscopy, revealed a large firm polypoid mass filling the left nasal cavity associated with a lobulated and polypoid tumour, extending into the nasopharynx. There was no associated numbness, opthalmoplegia or alteration of the patient's visual acuity.

36.2.1.3 Investigations

Computerized tomography (CT) and magnetic resonance imaging (MRI) confirmed the presence of a large hyperdense left-sided sinonasal avascular mass extending backwards to the nasopharynx. The irregular surface was noted along the lateral wall of medial orbital wall with enlarging the ostium-meatal complex. There were no signs of bone erosion. Inverted papillomas may have a cerebriform pattern on the MRI scan. CT scan will provide valuable information regarding bony erosion, extension to the orbit or the skull base and also in primary cases can show a hyperostotic nidus at the area of the origin of the pathology.

36.2.1.4 Treatment

After detailed discussion about the nature of the pathology and the potential risk of malignant transformation, the patient opted for surgery. A trans-nasal endoscopic medial maxillectomy was planned and the operative findings showed the tumour to be originating from a bony prominence over the lateral wall of the maxillary antrum which was drilled down at the end of the procedure to reduce the risk of recurrence. Full macroscopic clearance of the tumour was achieved. The diagnosis of IP was subsequently confirmed on paraffin-embedded histological assessment. The patient was followed up every 2 months in the first 6 months after surgery through the nasal endoscopic exam that showed a re-epithelized nasal cavity with no signs of disease then every year for 5 years and no disease recurrence noted during this time.

Summary and Author's Comments

1. Inverted papilloma is a benign sinonasal tumour with local aggressive behaviour and potential for malignant transformation.
2. Careful preoperative evaluation and scanning will help the surgeon select the indicated approach and management of the site of origin, aiming for total resection, which reduces the recurrence rates.
3. Wide surgical resection, preferably via an endoscopic approach is the main treatment modality and carries good outcomes.
4. Long follow up is important, some centres are in favour of lifelong monitoring.

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Sino-Nasal Squamous Cell Carcinoma

37

Hisham S Khalil

37.1 Case Presentation

A 54 year old man presented with a one year history of increasing right-sided nasal obstruction and recurrent right epistaxis. Over the previous 2 months he had noticed increasing frontal headaches and discomfort in his right eye. Imaging revealed a destructive sino-nasal lesion involving the right sino-nasal tract, extending to the left nasal cavity, the right orbit, skull base and brain. A biopsy revealed a squamous cell carcinoma (SCC). The patient was treated with endoscopic de-bulking of the lesion followed by chemoradiotherapy. The patient remains free of disease 3 years after completion of treatment.

37.2 Background Knowledge

Sinonasal malignancies are rare tumours of the nasal cavity and paranasal sinuses, accounting for less than 1% of all malignancies and about 3–5% of head and neck malignancies. The vast majority of sinonasal cancers are histologically squamous cell carcinomas, with adenocarcinomas being the second commonest histology subtype. The primary risk factor for sinonasal carcinoma is occupational exposure to wood dust, with other occupational risk factors including leather dust, nickel and radium5. Within the United Kingdom, sinonasal cancer is the second most common cancer site for occupational attribution. Tobacco smoking is a well-known risk factor. There has been a correlation between EBV and nasal Lymphoma. There is also an increasing association between HPV and sino-nasal SCC.

Mahalingappa and Khalil reported five-year results of treating 30 patients with sino-nasal cancer. The nasal cavity was the most common site for presentation, followed by the maxillary sinuses. Fifty percent of patients had a squamous cell carcinoma and 27% had a malignant melanoma. Half of the patients presented at stage IV of the cancer and 20% at stage III. Thirty-seven percent of patients underwent surgical management. The mortality in their series was 30% over the studied period. They concluded that late-stage presentation of sinonasal malignancy has resulted in increased patient mortality in their case series.

Histological subtypes of SCC include keratinising SCC, Non-keratinising SCC. Basiloid, Papillary, Adenosquamous and Spindle cell varieties. Given the rarity of these tumours, pathologists may experience difficulty in differentiating these tumours from other histological types of sino-nasal cancer.

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Complete surgical resection with clear margins remains the gold standard treatment. However, this is often not achievable due to the late stage at presentation. The available evidence does not suggest that more conservative endoscopic resections have inferior outcomes compared to more extensive traditional approaches such as a total maxillectomy or craniofacial resection. The overall prognosis of SCC of the sino-nasal tract remains poor like other types of sino-nasal cancer with 5-year survival rates ranging from 50 to 60% on average. There has been an insignificant trend of improvement of survival rates over the last decade. More recently, there has been increased attention to HPV related SCC with evidence of over-expression of p16 and presence of HPV DNA. There are some studies that suggest better prognosis in HPV related SCC. There is a need for large multi-centre trials comparing different treatment modalities and outcomes in the various histological types and sub-types.

37.3 Clinical Approach

37.3.1 History

The symptoms of sino-nasal cancer in general including squamous cell carcinoma (SCC) are generally non-specific, resulting in late presentation of patients to their General Practitioner/Family Doctor. There may also be a delay in referral to an ENT specialist for the same reason. In a 10-year audit of sino-nasal cancer patients referred to our department, we found a combination of unilateral nasal obstruction and persistent unilateral epistaxis to be the most sensitive predictor of malignancy from a symptoms perspective.

A focused history exploring the presence of visual symptoms, headaches, behavioural changes, general medical history and functional status is important as part of the overall assessment. It is equally important to inquire about risk factors including smoking and chemical exposure. This patient was a non-smoker and had not had any previous chemical exposure.

37.3.2 Examination

A presumed cause for the delayed diagnosis is the lack of examination equipment in primary care. Primary care physicians and dentists should be encouraged to use an Otoscope to examine the nose in patients with sino-nasal symptoms. A patient with a unilateral nasal mass should warrant a 2 week fast-track referral.

The evaluation of patients with sino-nasal cancer including SCC, should include a detailed Ophthalmological examination, including Visual Acuit, Visual Field Assessment, Fundus examination, intro-ocular pressure and in the presence of diplopia, an evaluation by an Orthoptist. In this patient, there was evidence of a subtle displacement of the right eye globe.

37.3.3 Investigations

37.3.3.1 Imaging

The patient had an initial CT scan of the paranasal sinus, followed by an MRI scan of the Head and the Paranasal Sinuses. These demonstrated a large destructive lesion in the right nasal cavity, right maxillary, ethmoid, frontal and sphenoid sinuses. The lesion extended in the tight orbit, invaded the right skull base with brain involvement. The lesion also traversed the nasal septum to involve the left nasal cavity (Figs. 37.1, 37.2 and 37.3). Once the histopathology was confirmed, the patient had a staging MRI scan of the neck and CT scan of the Thorax and upper abdominal cavity. The patient's staging was T4B, N0M0.

37.3.3.2 Histopathology

The biopsy demonstrated a moderately-differentiated SCC with areas of severe dysplasia.

37.3.4 Treatment

After discussion in the Head and Neck MDT and with the patient, it was decided to offer him endoscopic debulking followed by chemoradiotherapy

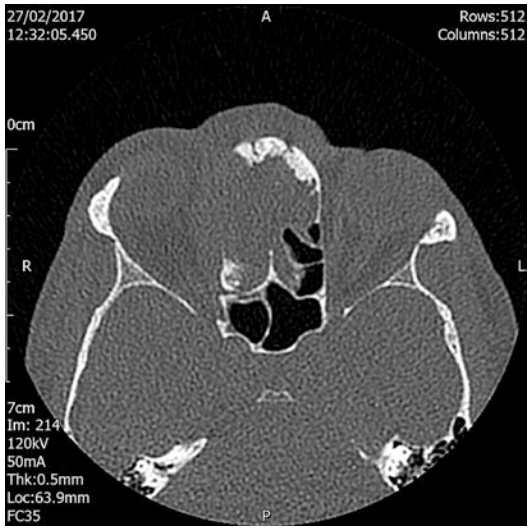

Fig. 37.1 Axial CT scan of the paranasal sinuses demonstrating a destructive lesion involving both ethmoid and frontal sinuses and the right orbit

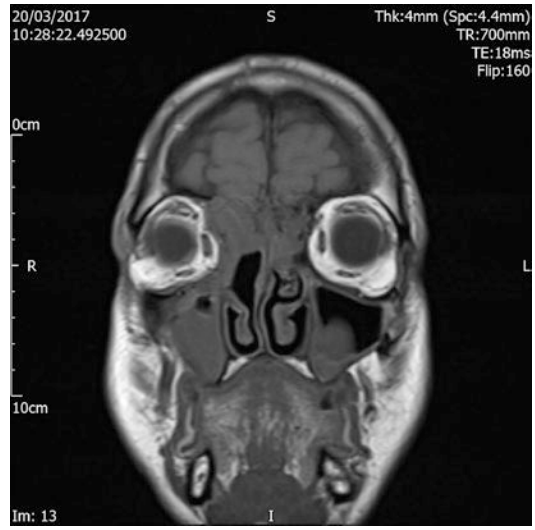

Fig. 37.3 Coronal MRI scan of the head and paranasal sinuses demonstrating a large sino-nasal lesion with intracranial extension

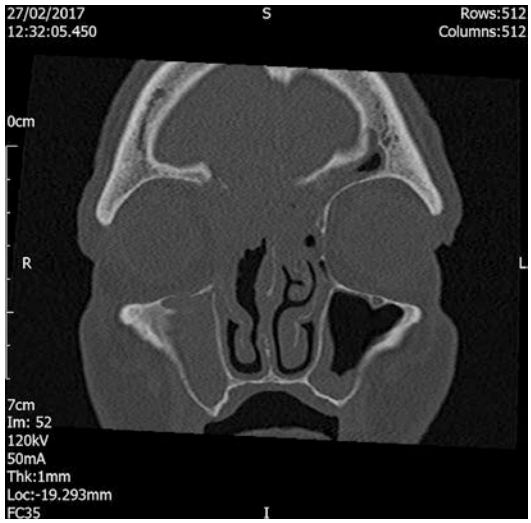

Fig. 37.2 Coronal CT of the paranasal sinuses demonstrating a destructive sino-nasal lesion with involvement of the skull base and right orbit

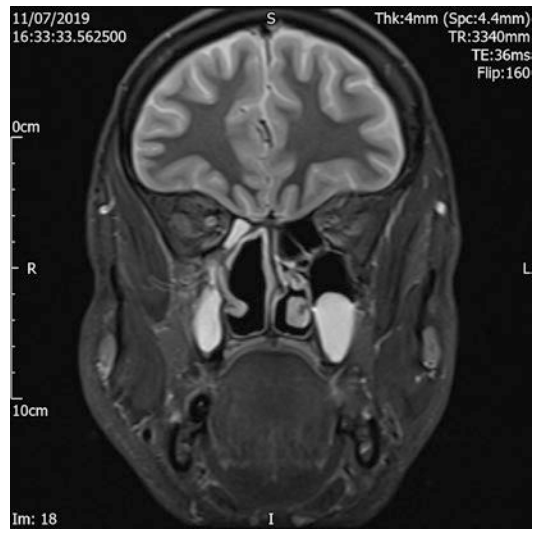

Fig. 37.4 A 2 year post-treatment coronal T2-weighted MRI scan demonstrating complete resolution of the sino-nasal cancer with inflammatory changes in both maxillary sinuses and right ethmoids

with a palliative intent given the presence of a number of poor prognostic factors, including invasion of the brain and orbit. The patient had induction chemotherapy with Carboplatin followed by 60 Gy of External Beam Radiation. The patient developed significant nasal crusting and a right epiphora following the treatment but retained his vision in both eyes. A small area of occipital alopecia resolved. A baseline MRI scan of the paranasal sinuses was requested at 3 months post-treatment. Further surveillance was achieved through 2 monthly diagnostic nasal endoscopies in the Outpatients for the first 2 years then 3 monthly in the third year. The patient also had 6 monthly MRI scans staggered with 12

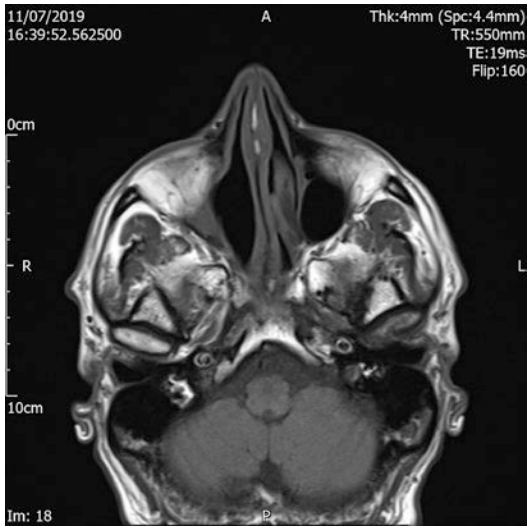

Fig. 37.5 A 2 year post-treatment axial T2-weighted MRI scan demonstrating complete resolution of the sino-nasal cancer in both nasal cavities

monthly diagnostic nasal endoscopies and check biopsies under general anaesthesia. Post-treatment MRI scans demonstrated complete resolution of the sino-nasal cancer including the orbital and intracranial components (Figs. 37.4 and 37.5).

The patient remains free of disease 3 years after completion of the treatment. The only concern the patient has is the right epiphora which persists despite demonstrating patent right naso-lacrimal drainage.

Summary and Author's Comments

1. SCC of the sino-nasal tract are rare but constitute the most common type of sino-nasal cancer in most reported series. They have various histological sub-types. HPV induced sino-nasal SCC are thought to have a better prognosis. Late presentation is the norm with overall poor 5-year survival.
2. Complete surgical excision with free margins offers the best chance of cure. However, this is not possible in a significant number of patients.
3. For patients with advanced disease, multi-modality treatment including endoscopic debulking and chemoradiation should be offered to patients.

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Sino-Nasal Intestinal-Type Adenocarcinoma

38

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38.1 Case Presentation

A 66-year-old woman presented with a 6 month history of left nasal obstruction and facial pain. She had experienced scanty purulent nasal discharge but no epistaxis. A nasal endoscopy revealed a left fleshy nasal mass. A CT scan of the sinuses revealed a left sino-nasal mass with minimal bony destruction (Fig. 38.1a, b). An endoscopic biopsy under general anaesthesia demonstrated an intestinal-type adenocarcinoma. MRI imaging of the face/neck and CT of the thorax and abdomen displayed that the malignancy was confined to the left paranasal sinuses.

The patient was offered a craniofacial resection but declined for cosmetic reasons. An endoscopic resection of the tumour was carried out with the resection margins free of disease. The patient continued to suffer from left facial pain and subsequent biopsies revealed early recurrence of the disease in the left anterior and posterior ethmoids. This was treated with further endoscopic resection. Further check biopsies revealed recurrence of the disease treated with 5-Fluorouracil (5 FU) topical chemotherapy. Topical chemotherapy was again used a year later to treat a recurrence of the tumour, located in the posterior ethmoid sinuses. Further surveillance imaging and biopsies confirmed the patient was free of disease (Figs. 38.2, 38.3 and 38.4) The patient remains free of disease 12 years after the initial treatment.

38.2 Background Knowledge

Adenocarcinoma of the paranasal sinuses is rare and generally follows an aggressive clinical course Craniofacial Resection has been the mainstay of treatment for many years and represents the gold standard of surgical resection. However, endoscopic resections now represent a viable alternative with fewer complications. As well as this, chemotherapy is an important treatment option and can be used to augment the effects of surgery. Adenocarcinomas are relatively radioresistant.

38.2.1 Craniofacial Resection

Craniofacial resection (CFR) was first described in 1963 and has since been considered the standard treatment for malignancies involving the

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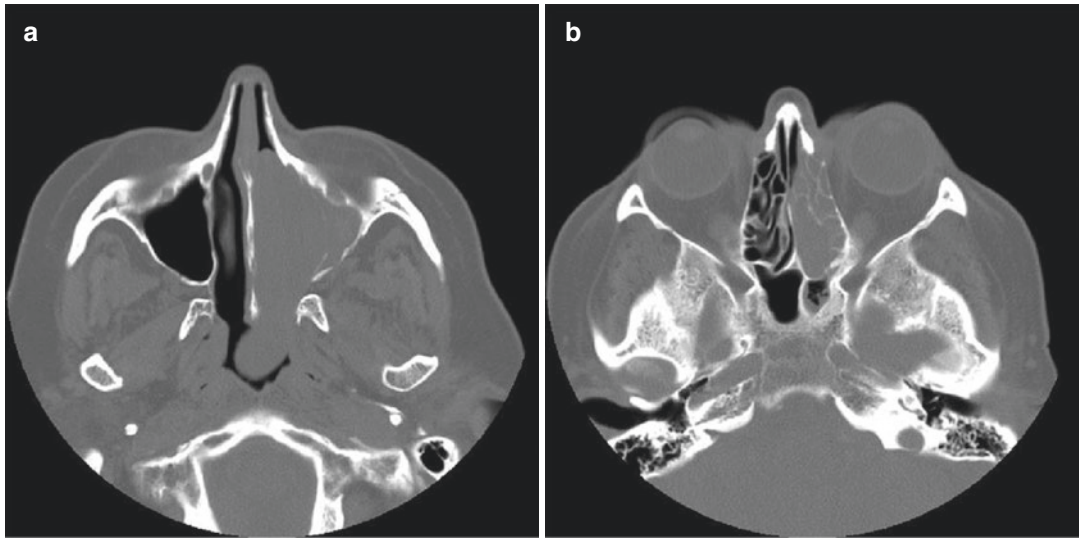

Fig. 38.1 Axial CT scans (**a**) CT scan showing a mass in the left nasal cavity, and (**b**) CT scan showing the mass in the left ethmoid sinuses, The mass was initially thought to be an inverted papilloma, but following histology it was shown to be an intestinal-type adenocarcinoma

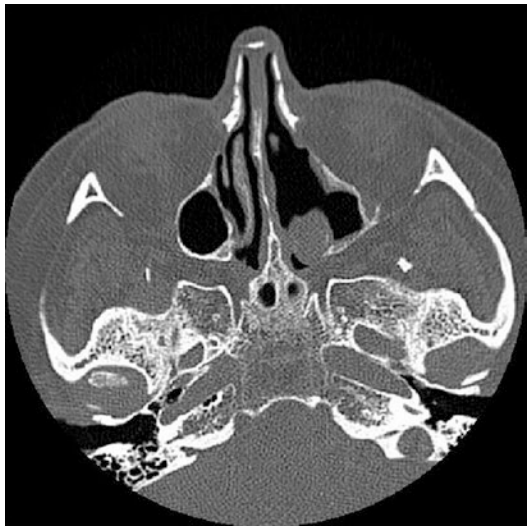

Fig. 38.2 Axial postoperative CT scan demonstrating a polyp in the left maxillary sinus

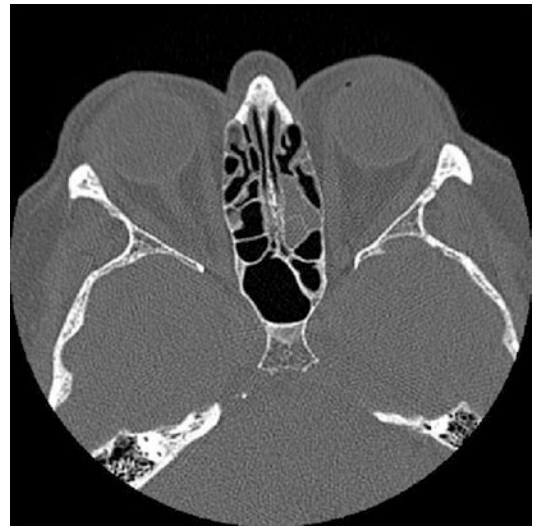

Fig. 38.3 Axial postoperative CT scan demonstrating mucosal thickening in the left ethmoid sinuses

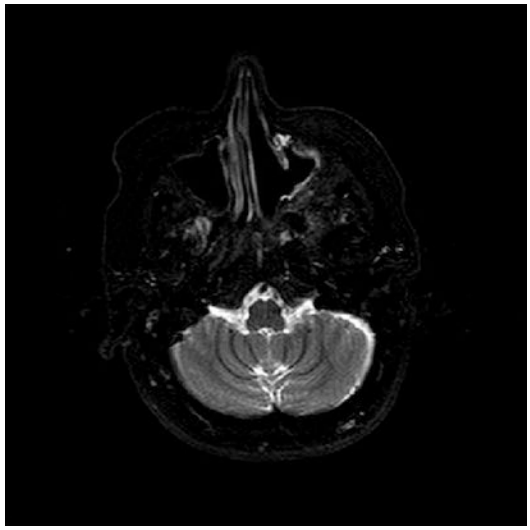

Fig. 38.4 Axial postoperative MRI scan showing an area of high signal in the patient's left maxillary antrum, indicating the need for a biopsy to be performed

anterior skull base. Whilst CFR has been shown to have low recurrence rates, it is also associated with a long recovery and complication rates as high as 40% in some studies.

38.2.2 Endoscopic Resection as a Viable Alternative

A number of studies compared the effects of both CFR and Trans-nasal Endoscopic Resection (TER). Eloy et al. reported that there were no significant differences in complication rates, postoperative survival, or metastasis between the two procedures. Moreover, they found that hospital stays were significantly shorter in the TER group of patients. TER were associated with better cosmetic outcomes. Podboj and Smid found that the operating time was on average an hour shorter, and blood loss during surgery was less than half of that with traditional external approaches. As well as this, postoperative radiotherapy, which may be delayed by wound healing with CFR, can be administered immediately following TER.

Appropriate patient selection is crucial for the success of TER. Patients with tumours invading the orbit, skin, or lateral recess of the frontal sinus are better managed with conventional CFR. Endoscopic resections are often 'piece meal' or 'segmental' resections rather than 'en bloc' ones. This necessitates a mapping of the resection and 'frozen section' biopsies of the remaining sino-nasal tissue in the patient following the resection.

Recurrence of the malignancy postoperatively for both endoscopic and traditional methods has been reported in several studies. Recurrence is the primary cause of cancer-related death in patients with ethmoidal malignancies and therefore is an important aspect of any treatment.

38.3 Clinical Approach

38.3.1 History

A history of unilateral nasal obstruction and nasal discharge raises suspicion of a sino-nasal malignancy particularly so if associated with epistaxis or blood-stained discharge. It is important to explore relevant history of risk factors such as smoking, exposure to hard wood dust, previous history of malignancy of the head and neck and previous radiotherapy.

A history of visual disturbance, constant headache, dental pain and loosening of teeth/ill-fitting dentures should also be sought.

38.3.2 Examination

This should include a complete ENT examination with a nasal endoscopy, assessment for orbital signs and neck palpation. A full ophthalmological assessment should be sought in the presence of any visual symptoms or imaging evidence of extension of the disease to the orbits.

38.3.3 Investigations

Thin slice CT scan and MRI of the sinuses are essential prior to biopsy. Once the histological diagnosis is confirmed, staging imaging (MRI neck and CT thorax and upper abdomen) is required.

38.3.4 Treatment

All patients should be discussed in the Multidisciplinary Team meeting. Patients should be closely involved in the clinical decision making process. This patient opted not to have a cranio-facial resection for aesthetic reasons. The pros and cons of external versus endoscopic approaches should be clearly highlighted to patients.

There is level III as well as level IV evidence to suggest that 5 FU is effective as a topical treatment in patients with sino-nasal adenocarcinoma. The treatment regimen is onerous and in this patient included weekly applications of 5 FU on ribbon gauze applied to the ethmoid sinuses. The treatment is associated with significant pain due to the intense inflammatory response to 5 FU. Adequate analgesia is required.

Summary and Author's Comments

Sino-nasal adenocarcinoma is usually treated with endoscopic or external cranio-facial resection depending on the experience of the surgical team and extent of disease. Adenocarcinoma may be relatively radioresitant and the role of radiotherapy is usually limited to palliative treatment of patients with unresectable tumours. There is a role for the use of topical 5 FU.

Important considerations for the surgical planning include orbital and dural involvement. There should be a detailed discussion with patients regarding the decision to exentrate the orbit if the involvement is beyond the orbital periosteum. Not all patients accept this and an alternative would be to offer radiotherapy for residual disease which in itself carries a risk to vision. Patients with significant dural involvement or intracranial extension should be considered for surgery by an endoscopic skull base surgical team.

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Sino-Nasal Undifferentiated Carcinoma

39

Hisham S Khalil

39.1 Case Presentation

A 61-year-old woman presented with a seven-month history of recurrent right epistaxis and increasing nasal obstruction. A nasal endoscopy revealed a fleshy right nasal poly. CT and MRI scans of the paranasal sinuses were requested. A diagnostic nasal endoscopy and biopsy under general anaesthesia confirmed the lesion to be a sino-nasal undifferentiated carcinoma.

39.2 Background Knowledge

SNUC are a very rare group of sino-nasal tumours of unknown aetiology. It has an estimated incidence of 0.02 per 100,000. Most patients with SNUC are diagnosed at a late stage in their disease, with a poor prognosis. Pathological examination of SNUC typically reveals large tumours with fungating and poorly defined margins that invade adjacent structures. The histologic appearance is characterized by sheets, trabecular, and ribbon-like arrangements of small to medium-size undifferentiated cells. These cells often have high nuclear to cytoplasmic ratio, high mitotic rate, and prominent tumour necrosis Lymphovascular and neural invasion are often also identified. It is immunohistochemically distinct from other sinonasal malignancies such as lymphoma, neuroendocrine carcinoma, mucosal melanoma, nasopharyngeal carcinoma, and olfactory neuroblastoma. Cytokeratin staining is positive while staining for leucocyte common antigen (LCA), S-100 protein, vimentin, in situ hybridization for Epstein-Barr encoded RNA (EBER), synaptophysin and calretinin are generally negative.

The rarity of these tumours means there is a lack of a strong evidence base. A 2017 retrospective review by Khan et al. noted five-year survival of 42.2% in s cohort of 460 patients included in the analysis. Most of the evidence-base for treatment comes from case series and case reports. Given the aggressive nature of these tumours, multi-modality treatment is the best approach including attempts to completely excise the tumour surgically, chemoradiation.

39.3 Clinical Approach

39.3.1 History

The patient gave a history of unilateral nasal obstruction and recurrent epistaxis. There was a slight reduction of her sense of smell and a mild facial pressure. She had no visual symptoms and was otherwise fit and well. Her symptoms did not improve with a steroid nasal spray prescribed by her GP. She was subsequently referred to an ENT Surgeon.

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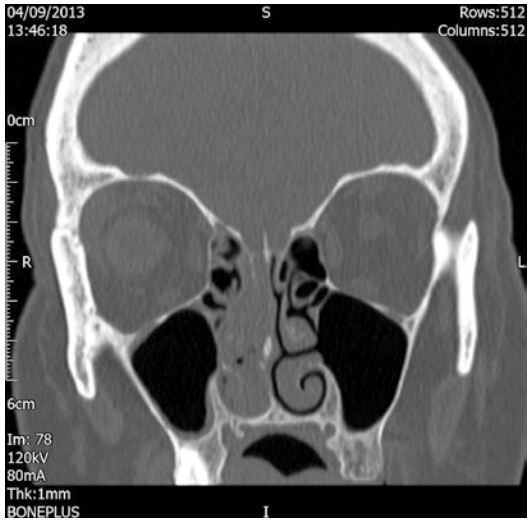

Fig. 39.1 Coronal CT scan of the paranasal sinuses with a soft tissue mass in the right nasal cavity and an intact cribriform plate. The right lamina papyracea and the bony nasal floor appear intact

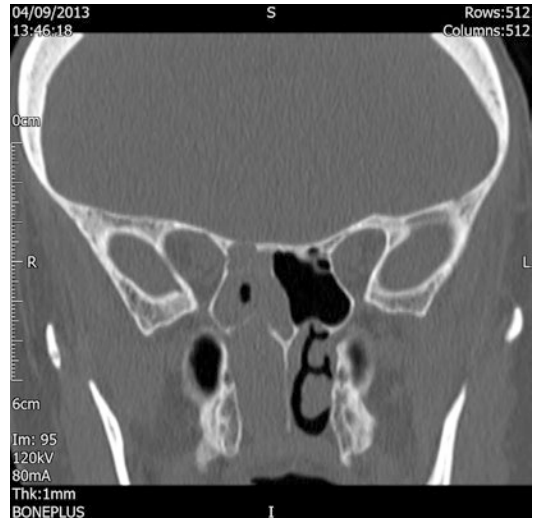

Fig. 39.3 A coronal CT scan of the paranasal sinuses with a soft tissue mass in the right sphenoid. There is apparent destruction of the bone of the planum sphenoidale

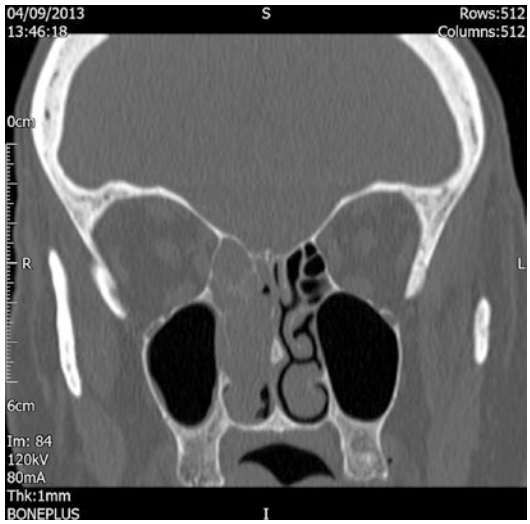

Fig. 39.2 A coronal CT scan of the paranasal sinuses demonstrating involvement of the right posterior ethmoid sinuses and an intact skull base

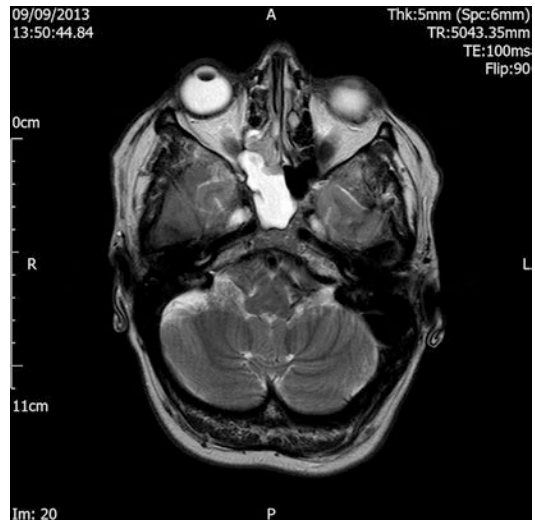

Fig. 39.4 T2-weighted axial MRI of the paranasal sinuses with a hyperintense lesion in the right sphenoid sinus extending to the right posterior ethmoids (mucocele) and abutting a heterogenous lesion in the right posterior ethmoid sinuses (suspected tumour)

39.3.2 Examination

A nasal endoscopy revealed a fleshy right nasal mass filling most of the right nasal cavity. The left nasal cavity, ears, throat and neck were normal. There were no orbital signs. A subsequent ophthalmological assessment was normal.

39.3.3 Investigations

An initial CT scan of the paranasal sinuses revealed a soft tissue mass filling the right nasal cavity, in contact with the right side of the nasal septum, involving the right medial wall of the

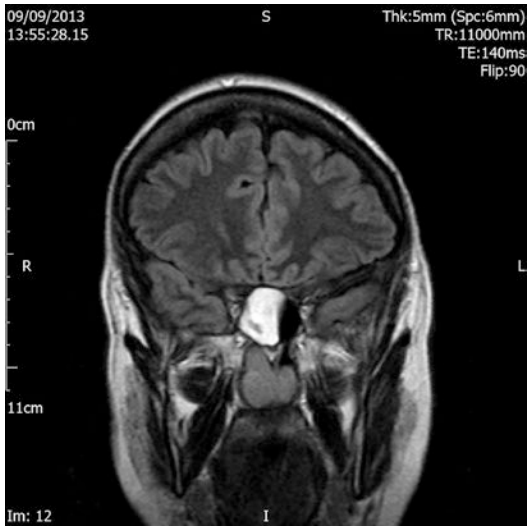

Fig. 39.5 Coronal T2-weighted MRI of the paranasal sinuses demonstrating a hyperintense lesion in the right sphenoid sinus, consistent with a mucocele

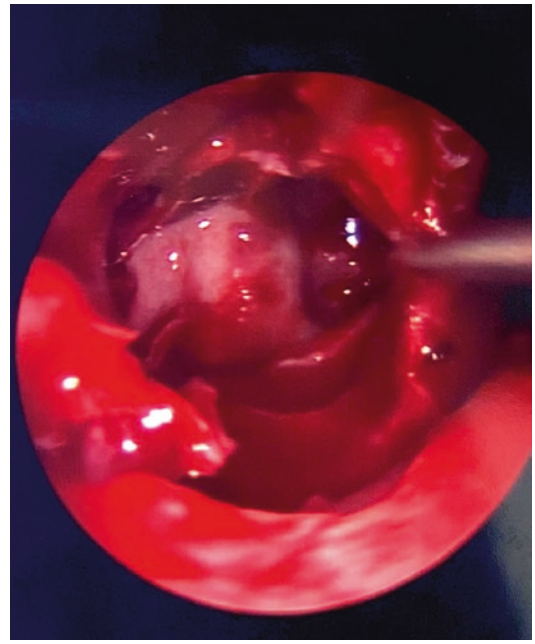

Fig. 39.6 Endoscopic view of the right posterior ethmoid complex during surgery demonstrating tumour tissue abutting the lamina papyracea

maxillary sinus, the posterior ethmoid sinuses and abutting the right medial orbital wall. The cribriform plate appeared intact. There was opacification of the right sphenoid sinus and apparent erosion of the bone of the planum sphenoidale (Figs. 39.1, 39.2 and 39.3).

A subsequent MRI scan of the paranasal sinuses revealed the soft tissue mass within the right sphenoid sinus to be a mucocele, responsible for the erosion of the bone of the right planum sphenoidale (Figs. 39.4 and 39.5).

The biopsy of the right sino-nasal lesion revealed a sino-nasal undifferentiated carcinoma. Staging MRI scan of the Neck and CT Thorax and upper abdominal cavity were normal. The definitive staging was T3N0M0 (Stage III).

39.3.4 Treatment

The patient was discussed in the Head and Neck and Skull Base MDTs with a recommendation to proceed with endoscopic resection of the sino-nasal tumour. The patient was counselled regard-

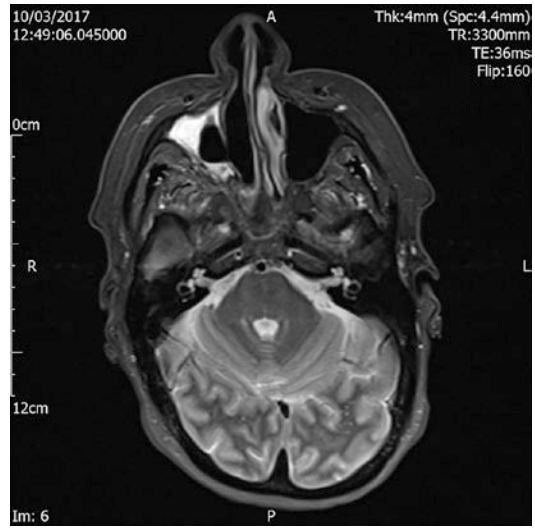

Fig. 39.7 Postoperative Axial MRI of the paranasal sinuses demonstrating excision of the tumour with some inflammatory changes in the mucosa of the right maxillary sinus

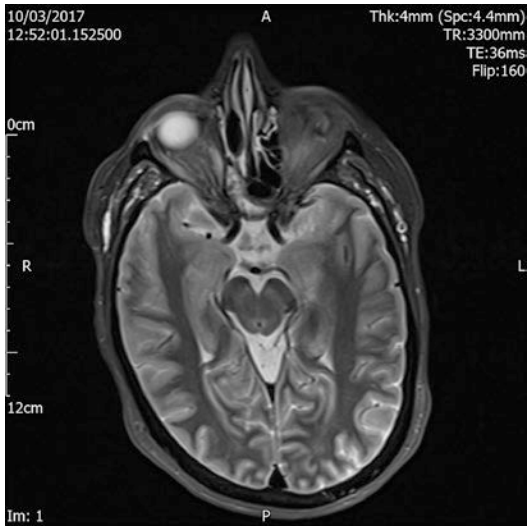

Fig. 39.8 Postoperative axial MRI scan of the paranasal sinuses demonstrating a clear right ethmoidal complex and right orbit. There is some mucosal thickening of the right spheno-ethmoidal recess but no evidence of recurrent disease

ing the possible need for an orbital exentration given the close proximity of the tumour to the medial orbital wall. She declined this option and agreed to undergo postoperative radiotherapy if there were positive margins.

Surgery was carried out with segmental endoscopic resection of the sino-nasal tumour using Computer-Assisted Image guidance. The tumour was shaved from the right medial orbital wall, anterior and posterior ethmoids (Fig. 39.6) and spheno-ethmoidal recess. An endoscopic medial maxillectomy was carried out with excision of the nasal septum in a small area contiguous with the tumour. A right sphenoidotomy revealed a mucocele with no evidence of tumour in the sphenoid sinus. The left nasal cavity and nasal floors were intact. 'check' biopsies were obtained after the resection. The patient had an uneventful postoperative recovery.

All margins were free of disease except for the region of the posterior ethmoids/medial orbital wall. The patient therefore underwent postoperative radiotherapy for 6 weeks. She had surveillance with MRI scans and diagnostic nasal endoscopies for the first 5 year after completion of treatment. She is currently undergoing annual review with surveillance MRI scans of the paranasal sinuses (Figs. 39.7 and 39.8). She remains free of disease 7 years after surgery.

Summary and Author's Comments

1. SNUC usually present late and have an aggressive course.
2. The surgical approach will depend on the presence or absence of involvement of the orbit, dura and brain tissue.
3. Shared decision making with the patient and the recommendation of the MDT is required.
4. There is no evidence to suggest that radical surgery confers better treatment outcomes compared to more conservative surgical approaches combined with radiotherapy and chemotherapy.

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Sino-Nasal Malignant Melanoma

40

Hisham S Khalil

40.1 Case Presentation

A 52-year old man presented with a few month history of recurrent left epistaxis and left nasal obstruction. An nasal endoscopy revealed a pigmented soft tissue mass in the nasal cavity. This was confirmed by CT (Figs. 40.1 and 40.2) and MRI imaging. A biopsy confirmed the lesion to be a malignant melanoma. This was excised through a total septectomy via a lateral rhinotomy approach (Fig. 40.3). A year later, he developed a satellite malignant melanoma lesions in the posterior ethmoid and sphenoid sinuses (Fig. 40.4). These were treated by endoscopic excision followed by postoperative radiotherapy. The patient remained free of disease but passed away of unrelated auses 9 years after his initial presentation (Fig. 40.5).

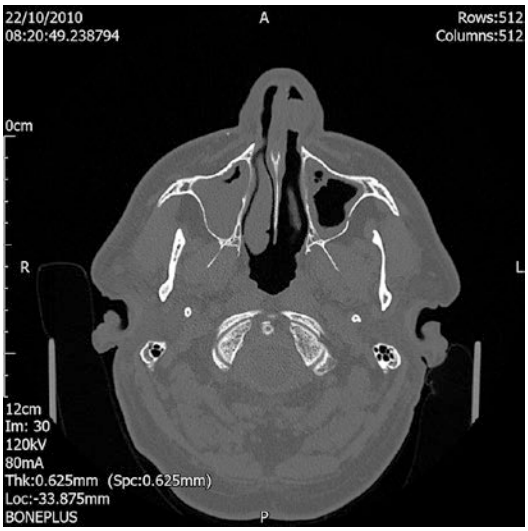

Fig. 40.1 Axial CT scan of the paranasal sinuses demonstrating a soft tissue lesion arising from the left side of the nasal septum

40.2 Background Knowledge

Sino-nasal malignant melanomas are rare tumours that accounts for between 0.5 and 2% malignant melanomas. They have a relatively poor prognosis due to late presentation. Risk factors and aetiology remain unclear. The incidence is slightly higher in white men over the age of sixty. Most patients present with unilateral epistaxis and nasal obstruction. The histology usually reveals an undifferentiated tumour with the diagnosis clinched with immunohistochemistry e.g. demonstrating positivity for vimentin, S100, and HMB45 (human melanoma black 45).

The best chance of cure is with complete surgical excision and free margintes. Radiotherapy and chemotherapy are reserved for local recurrence and distant metastasis. A meta-analysis of survival in primary sino-nasal malignant melanoma with various treatment modalities demonstrates that there is no survival advantage for combined radiother-

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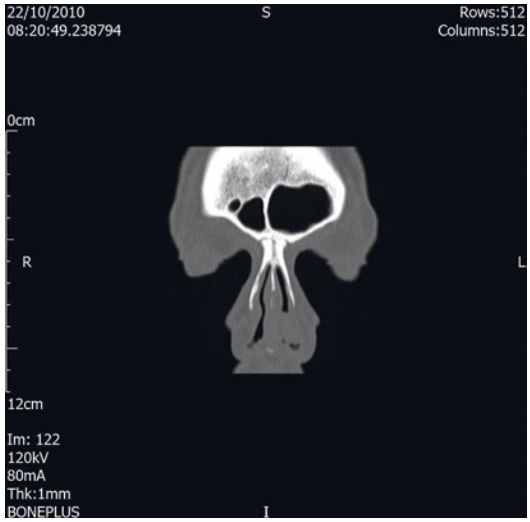

Fig. 40.2 Coronal CT scan of the paranasal sinuses demonstrating a soft tissue lesion arising from the left side of the nasal septum

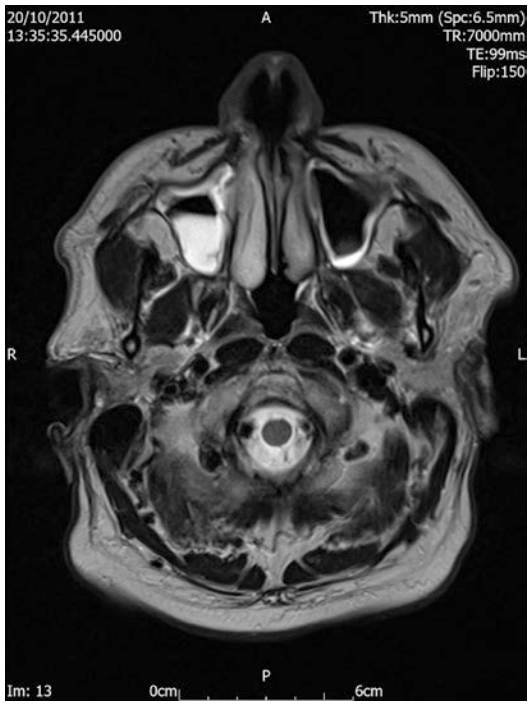

Fig. 40.3 Postoperative T-2 weighted axial MRI scan demonstrating the total septectomy. There is fluid in the right maxillary sinus

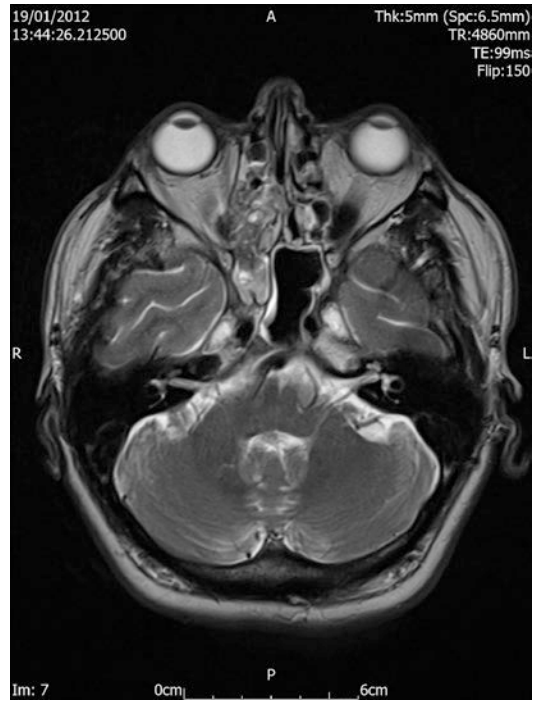

Fig. 40.4 Axial MRI scan demonstrating a recurrent lesion in the left posterior ethmoid and sphenoid sinuses

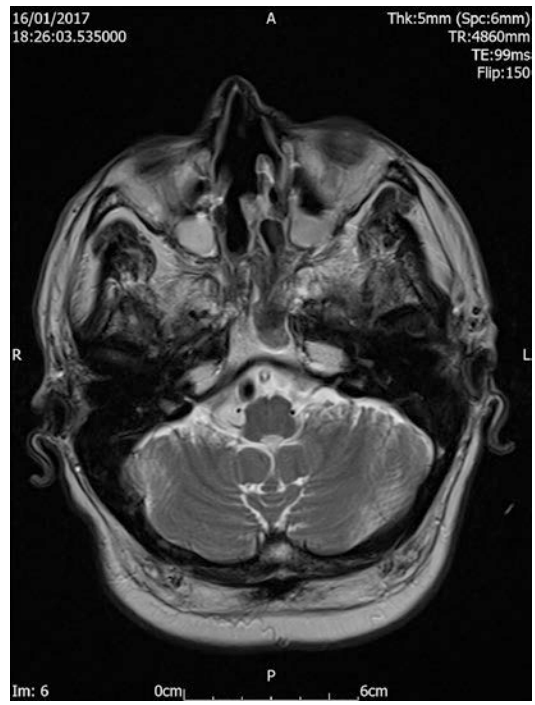

Fig. 40.5 Axial MRI scan with no evidence of disease following the treatment of the recurrent malignant melanoma with endoscopic excision and postoperative radiotherapy

apy + surgery or chemoradiotherapy + surgery versus surgery alone. There was a significant overall survival advantage for surgery + chemotherapy versus surgery alone and versus chemotherapy alone. The average overall survival was 27.41 months consistent with a poor prognosis. The risk of local recurrence (31–85%) and distant metastasis (25–50%) is high. Good loco-regional recurrence may still be associated with distant metastasis.

40.3 Clinical Approach

40.3.1 History

A history of recurrent unilateral epistaxis and nasal obstruction is the most common presentation. It is important to inquire about symptoms related to metastatic lesions.

40.3.2 Examination

A nasal endoscopy revealed a pigmented soft tissue lesion arising from the left side of the nasal septum and filling the anterior third of the left nasal cavity. The rest of the nasal cavities and postnasal space were normal. Examinations of the ears, throat and neck were unremarkable. The patient didn't have any orbital signs. Nasal malignant melanoma lesions may not always pigmented.

40.3.3 Investigations

A full blood count is essential to rule out anaemia as a result of recurrent epistaxis. Other laboratory investigations include renal profile and liver function tests. Like all sino-nasal tumours, investigations for sino-nasal malignant melanoma will include CT and MRI scans of the paranasal sinuses to delineate the lesion and assesses involvement of the orbit, skull base, dura and brain, MRI of the neck and CT of the thorax and upper abdominal cavity for staging purposes.

A baseline MRI scan of the sinuses is required 3 months following completion of treatment and then repeated every 6 months for the first 3 years of treatment. The practice thereafter varies, but an annual surveillance MRI scan of the sinuses is recommended.

40.3.4 Treatment

The mainstay of treatment of sino-nasal malignant melanoma is total excision with free margins. In this patient, the best chance to achieve this was through a lateral rhinotomy incision and total excision of the nasal septum with the aid of a Ballenger knife. The excision margins were free of disease. Close surveillance included 2-monthly follow up of the patient with nasal endoscopy and a surveillance MRI (Fig. 40.3).

A recurrence was detected almost a year following the initial treatment with the recurrence of the epistaxis and the detection of satellite deposits of a pigmented lesion in the left spheno-ethmoidal recess on nasal endoscopy. The recurrence was delineated on a MRI scan with involvement of the left sphenoid and posterior ethmoid sinuses (Fig. 40.4). A further biopsy and an endoscopic spheno-ethmoidectomy confirmed recurrent a mucosal malignant melanoma. Following a MDT review, the patient had postoperative radiotherapy. The patient remained free of disease 7 years later including on a MRI scan 5 years later (Fig. 40.5). The patient passed away from unrelated causes 9 years after his initial presentation.

Summary and Author's Comments

Sino-nasal malignant melanoma is a rare tumour that tends to present late with overall poor prognosis. The best chance of cure lies with complete surgical excision and free margins.

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Fibro-Osseous Lesions of the Craniofacial Skeleton

41

Hisham S Khalil and Abdulaziz Abushaala

41.1 Case Presentation

A 19 year old man presented with a few years history of increasing headaches. Over the preceding few months, his friends and family noticed a slight protrusion of his left eye. In the last two months, he started experiencing double vision. CT and MRI scans confirmed the presence of fibrous dysplasia of the left frontal sinus with a secondary mucocele formation extending into the left orbit. The mucocele was drained and the fibrous dysplasia reduced through a combined craniotomy and endoscopic sinus surgery. The patient symptoms resolved postoperatively, and he was discharged 5 years later.

41.2 Background Information

Benign fibro-osseous lesions of the craniofacial skeleton (BFOL) are a variant group of intraosseous disease processes that share similar microscopic features characterized by hypercellular fibroblastic stroma containing various combinations of bone or cementum-like tissue and other calcified structures. Ossifying fibroma (OF) and fibrous dysplasia (FD) are the most common fibro-osseous lesions.

41.2.1 Ossifying Fibroma

Ossifying fibroma affects patients in the third to fourth decade of life. It shows preference for the mandible and maxilla. It is classified into Ossifying fibroma of odontogenic origin (cemento-ossifying fibroma), and Juvenile ossifying fibroma. It may present as Facial swelling or disfigurements; it may cause signs and symptoms associated involvement of adjacent vital structures including the sinuses and orbit. Complete surgical excision is required to avoid the risk of recurrence.

41.2.2 Fibrous Dysplasia

Fibrous dysplasia is a disease of growing bones that presents mostly in children and adolescents. It has three varieties which in order of frequency are the monostotic form, a polystotic form and the rare and most severe form, the *McCune-Albright syndrome* which affects young girls with endocrine disturbances, small stature and pigmented skin lesions.

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The aetiology of fibrous dysplasia remains unknown. One theory is that the pattern and distribution of fibrous dysplasia depends on which tissues contain the mutated G_s-α (*GNAS1*) gene and is in turn affected by such factors as genomic imprinting.

The management of fibrous dysplasia depends on the symptoms. The disease may become quiescent after puberty. However, persistent growth requires surgical excision with or without reconstruction. Bisphosphonate therapy can also be prescribed. Malignant degeneration has been reported in a few cases of fibrous dysplasia, most of which are osteosarcomas and less frequently, fibrosarcoma, or chondrosarcoma. It is therefore important to keep patients with fibrous dysplasia under long-term follow-up.

41.3 Clinical Approach

41.3.1 History

A history of preceding trauma is not unusual in patients with fibrous dysplasia. The patient had sustained a head injury as a child. Depending on the site of the cranio-facial skeleton affected, there may be a history of asymmetry of the face or visual disturbance (proptosis and diplopia), atypical facial pain and headaches, nasal obstruction, anosmia and hearing loss. The gradual onset of symptoms may sometimes be missed by the patient.

41.3.2 Examination

There was evidence of left proptosis with mainly downward displacement of the left eye. There was no obvious asymmetry of the facial skeleton. A nasal endoscopy was unremarkable. An Ophthalmological assessment was normal apart from the mild left proptosis and some restriction of the left eye movement on lateral gaze with diplopia.

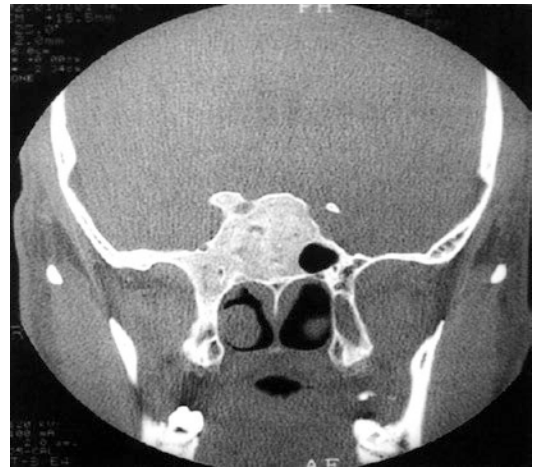

Fig. 41.1 Coronal CT scan of the paranasal sinuses demonstrating fibrous dysplasia of the right sphenoid sinus and most of the left sphenoid sinus

41.3.3 Investigations

The diagnosis of fibrous dysplasia as mainly made through imaging. A combination of CT and MRI imaging is required to clinch the diagnosis. The 'ground glass' appearance is the most common appearance. (Fig. 41.1) Other findings include thickening of the cranial cortices and the formation of cysts within the bone and mucocele formation.

A biopsy serves to confirm the diagnosis and exclude malignant transformation. This usually demonstrates. The histology is characterised by low to moderately cellular fibrous stroma surrounding irregular, curvilinear trabeculae of woven bone.

Summary and Author's Comments

BFOLs have overlapping clinical, radiological and histopathological features.

The accurate diagnosis often requires a multidisciplinary teams' collaboration to reach an accurate diagnosis and offer the appropriate treatment plan. Long-term follow up is required for patients with BFOL to detect any recurrence or malignant transformation.

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Osteomas of the Paranasal Sinuses

42

Abdulaziz Abushaala and Hisham S Khalil

42.1 Case Presentation

A 52-year-old Caucasian woman presented with an incidental finding of a bony lesion in the left frontal sinus on a CT scan of the paranasal sinuses. This was requested to investigate chronic rhinosinusitis that had failed to improve despite optimal medical therapy. A diagnosis of a left frontal sinus osteoma was made. The patient underwent functional endoscopic sinus surgery. The frontal sinus osteoma was not removed, and the patient was followed for a few years without progression of the osteoma.

42.2 Background Knowledge

Osteomas are slow-growing benign bony tumours. They are the most common benign tumours of the paranasal sinuses. They are found incidentally in about 3% of CT scans of the paranasal sinuses. They can appear as sporadic or part of Gardner syndrome. Osteomas are commonly found in the frontal sinus (80%), followed by the ethmoidal sinuses (15 %), maxillary sinuses (5%) and rarely in the sphenoid sinus.

There are three theories behind the development of paranasal sinus osteomas, none of which are universally accepted. Macroscopically, they can be pedunculated or have a sessile, broad base. Histologically, they are osteogenic tumours composed of mature bone which can be categorized into Ivory, mature or mixed histological types.

Osteomas are mostly asymptomatic; however, they may become symptomatic as they increase in size and causes obstruction of the sinus ostia or extent into the orbit, intracranially or cause facial asymmetry. The signs and symptoms of osteomas will depend on their location. They may present with symptoms of sinusitis or its complications, symptoms of orbital invasion like diplopia, proptosis, exophthalmos, epiphora, headache, facial pain or other neurological symptoms.

Asymptomatic osteomas require no active treatment. However, symptomatic osteomas require surgical resection by an endoscopic approach, external surgery or a combined approach, depending on the location and the size of the tumour. There have been no reports in the literature of malignant transformation of osteomas.

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42.3 Clinical Approach

42.3.1 History

The patient had symptoms of rhinosinusitis with bilateral nasal obstruction, intermittent purulent nasal discharge, reduced smell and occasional headaches. There was no history of a localised facial pain in relation to the osteoma site. The patient had a history of bronchiectasis and was on maintenance treatment with cyclical antibiotics, pulsed oral steroids, steroid inhalers and bronchodilators.

42.3.2 Examination

There was no evidence of facial swelling, asymmetry or tenderness. Standard signs of chronic rhinosinusitis were present with bilateral grade III inflammatory-looking polyps and mucopus in both nasal cavities.

42.3.3 Investigations

CT scans of the paranasal sinuses are the investigations of choice(Fig. 42.1). Histological confirmation is seldom require. Annual imaging is used initially to determine the rate of growth but is not required for the long term if the patient is asymptomatic and to avoid the risk of irradiation.

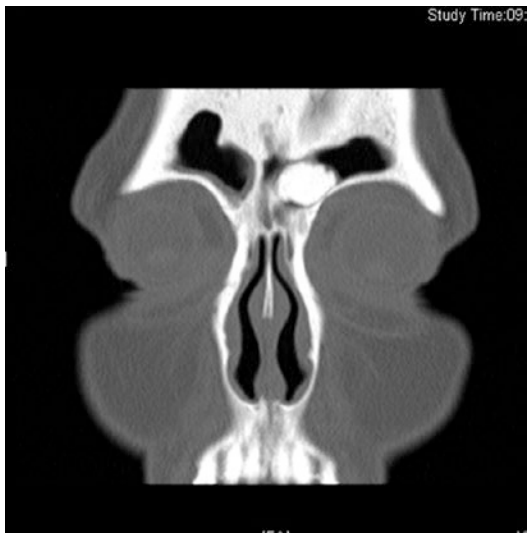

Fig. 42.1 Coronal CT of the paranasal sinuses revealing a left frontal sinus osteoma with good aeration of the frontal sinus

42.3.4 Treatment

A shared decision with the patient was taken not to excise the osteoma as it was deemed it was not contributing to her symptoms. This was to avoid potential risks of the surgery such as CSF rhinorrhea. Subsequent scanning confirmed this was an appropriate approach.

Excision is indicated in symptomatic patients for frontal sinus osteomas, this can be through an endoscopic extended frontal recess approach (Draf I-III), a mini-osteoplastic flap, an external frontal sinustomy or in larger osteomas a combined external and endoscopic approach. The osteomas are easier to excise endoscopically if they have a pedunculated base.

Summary and Author's Comments

Osteomas of the paranasal sinuses are often incidental findings on imaging. Surgical excision is only warranted in symptomatic patients and the approach will depend on the site and size of the osteoma as well the experience of the surgeon.

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Part VII

Paediatric Rhinology

Acute Rhinosinusitis

43

Elizabeth Kershaw and Aristotelis Poulios

43.1 Case Presentation

A 4 year old boy presents with a 12 day history of coryzal symptoms, cough, green rhinorrhea, nasal obstruction with mouth breathing and temperatures. He has recently had an upper respiratory tract infection, from which he seemed to be recovering. He is typically a mouth breather and a loud snorer with occasional apnoeic episodes, even when well.

He has no known comorbidities, and was born at term without complications. His GP advised Mum to try a nasal aspirator, which has helped remove some of the discharge from his nose but otherwise his symptoms have persisted.

On examination, he appears generally unwell and has a temperature of 38.3. Anterior rhinoscopy demonstrates oedema and pus in the floor of the nose (Fig. 43.1). Percussion over the maxillary sinuses is painful to the patient. The oropharynx is largely unremarkable apart from a trail of mucopurulent discharge from the post nasal space.

This patient was given a course of oral antibiotics and intranasal steroids and their acute symptoms resolved.

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43.2 Background Knowledge

43.2.1 Anatomy

The paranasal sinuses are air filled spaces surrounding the nasal cavity lined by respiratory mucosal epithelium. They begin to develop at 25–28 weeks gestation as sacculations of the mucosa, which then grow to invade the bones surrounding the nasal cavity to form sinuses. They continue to develop throughout childhood. The maxillary sinus is the first to develop and become aerated; it has two phases of growth, in the first three years of life and then again from 7–18 years of age. The ostium lies at or above the level of the floor of the nose, lower than in adults. The sphenoid sinus does not begin to develop until 3 years of age. The ethmoid sinus is present during childhood and pneumatization begins in the 4th year of life, with the ethmoid air cells developing during puberty. The frontal sinus develops last, pneumatization begins from age 2.

The paranasal sinuses are present to aid filtration, humidification and warming of inspired air. They lighten the skull, allow resonance for speech, absorb shock from trauma and contribute to facial growth.

Imaging of the paranasal sinuses during acute rhinosinusitis has demonstrated the predilection of sinus involvement; the maxillary and ethmoid sinuses (60%), sphenoid sinus (35%), and frontal

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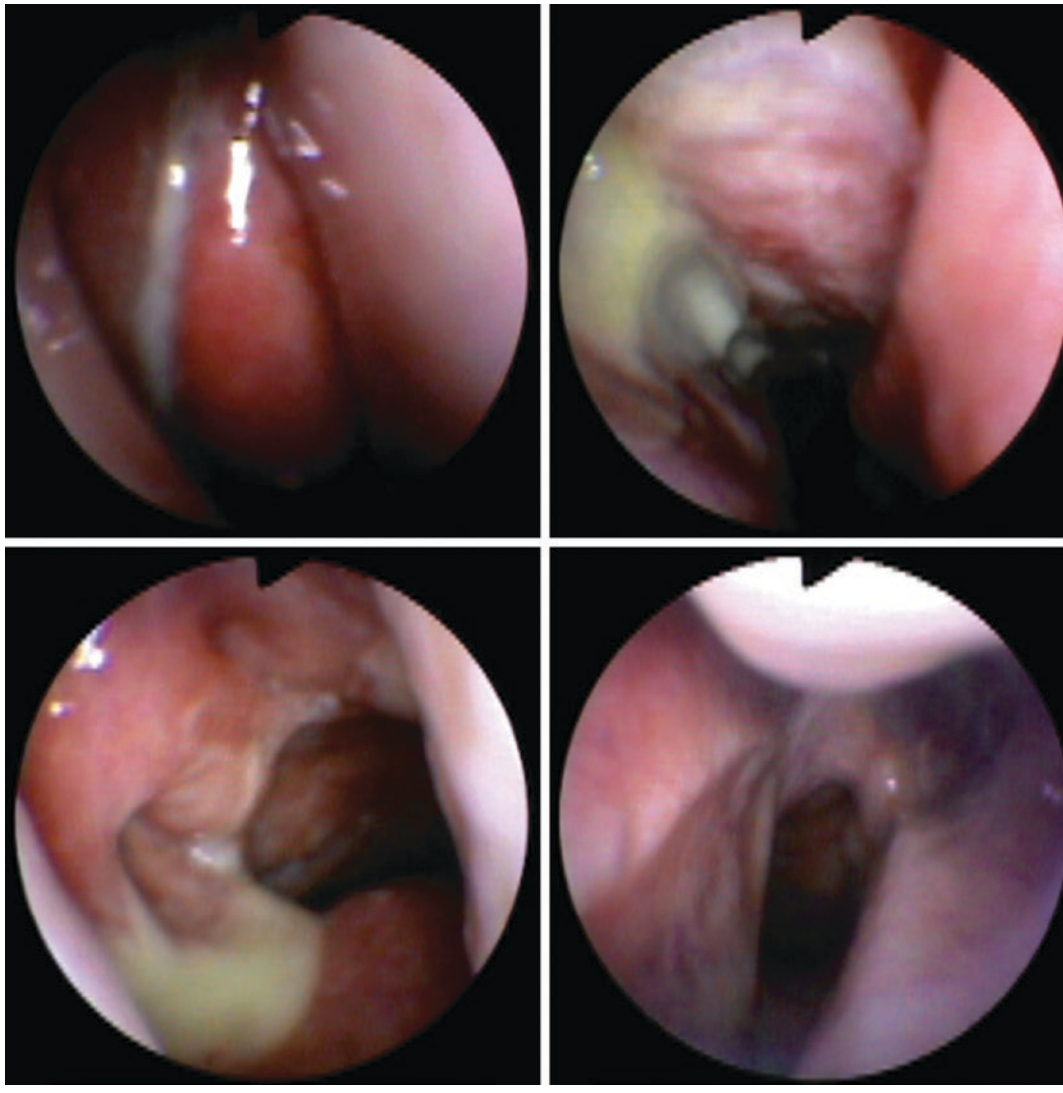

Fig. 43.1 Purulent discharge in right and left middle meatus

sinus (18%). This is in keeping with the chronological development of the sinuses during childhood.

43.2.2 Pathophysiology

Acute rhinosinusitis (ARS) is thought to occur on the background of an upper respiratory tract infection (URTI), the pathogens responsible are the same. The URTI will result in edema of the nasal mucosa and increased secretions, this in turn can occlude the sinus ostia, obstructing drainage of mucous and allowing stagnation. The build of up secretions within the sinuses, results in the symptoms of pain, discolored rhinorrhea and fever.

Gwaltney et al described the theory that ARS was a result of nose blowing in the presence of URTI, with pathogens being forced into the paranasal sinuses under pressure. Oedema and mucociliary dysfunction during URTI together impede the clearance of mucous and pathogens.

43.2.3 Microbiology

ARS is mostly common viral, caused by rhinovirus, influenza A and B, parainfluenza, respiratory syncytial virus, adenovirus, or enterovirus.

Bacterial ARS is typically a result of infection from Streptococcus pneumonia, Hemophilus influenza and Moraxella catarrhalis. Less frequently, Staphylococcus aureus, Streptococcus pyogenes and Streptococcus viridans may be isolated. In approximately a third of cases, the cause is polymicrobial, which is relevant when deciding on antimicrobial treatment. Penicillin resistance has been shown to be increasing.

Predisposing factors include allergy, impaired ciliary function, gastro-esophageal reflux disease, immunodeficiency including diabetes, and congenital disorders including cystic fibrosis and primary ciliary dyskinesia.

43.3 Clinical Approach

43.3.1 Diagnosis

Acute rhinosinusitis is diagnosed based primarily on the clinical history and examination of the patient.

In taking a history, the current symptoms should be listed including the onset, duration, preceding events, any fluctuation in severity, presence of nasal obstruction, rhinorrhea, facial pain, headache, cough, fever. Any previous similar episodes or history of upper respiratory tract infection, the use of antibiotics in the past. Other medical illnesses or conditions, allergy, social circumstances including home and education.

Examination of the patient includes general observations, evidence of fever. A thorough assessment of the ears, nose and throat. Anterior rhinoscopy will show evidence of mucosal oedema, mucopurulent discharge and inflammation; if tolerated a flexible endoscope can be used to assess the nasal cavity and may permit microbiology swabs being taken from the middle meatal antrum. Neurological assessment should be performed, as well as examination of the orbits, ocular mobility and vision to rule out complications of acute sinusitis.

Differential diagnoses will include foreign body, choanal atresia/stenosis, dental disease, adenoidal hypertrophy or inflammation, allergic rhinitis. The history and examination will guide the final diagnosis.

According the latest European Position Paper on Rhinosinusitis and Nasal Polyps (EPOS) published in 2020, acute rhinosinusitis is defined as:

Sudden onset of two or more of the symptoms:

- Nasal obstruction/blockage/congestion
- Or discolored nasal discharge
- Or cough (daytime and night-time)

For <12 weeks (with symptom free intervals if problem is recurrent); with validation by telephone or interview'

Acute bacterial rhinosinusitis (according to EPOS) is the presence of at least three of:

- Discolored nasal discharge (with unilateral predominance) and purulent secretion in the nasal cavity
- Severe local pain (with unilateral predominance)
- 'double sickening' – deterioration after an initial milder phase of illness
- Elevated ESR/CRP
- Fever >38C

Radiological and microbiological investigations are not indicated in uncomplicated ARS. Antral washout with swabs for microbiology is useful in severe or resistant cases. Alternatively, swabs can be taken from the maxillary antrum under endoscopic guidance.

43.3.2 Treatment

In the new EPOS 2020 it is emphasized that the treatment of almost all patients with ARS, adults and children, should be symptomatic. If needed this can be combined with local corticosteroids. Intranasal steroids have been demonstrated to show significant benefit in the treatment of ARS.

They reduce mucosal oedema which in turn improves drainage of the paranasal sinuses. Antibiotics should be used only when there are severe symptoms suggesting a bacterial ongoing acute rhinosinutitis. These can be high fever, relapsing after a short period of getting better, severe facial pain and raised ESR. If antibiotics are needed then Amoxillin-clavalunate (Co-amoxiclav) and cephalosporins are recommended, and where the patient has allergies, macrolides can be used. Ideally consider referring to the local antibiotic formulary. Falagas suggested that the use of antibiotics resulted in resolution of symptoms more rapidly, but that patients would ultimately recover spontaneously.

Surgical intervention is only necessary in the treatment of complications of acute rhinosinusitis.

Routine follow up in the ENT clinic is not required for uncomplicated acute rhinosinusitis.

Summary and Author's Comments

1. Acute rhinosinusitis is diagnosed based primarily on the clinical history and examination of the patient.
2. Differential diagnoses will include foreign body, choanal atresia/stenosis, dental disease, adenoidal hypertrophy or inflammation, allergic rhinitis.
3. Treatment is symptomatic in most cases, combined with local corticosteroids if needed.

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Chronic Rhinosinusitis

44

Elizabeth Kershaw and Aristotelis Poulios

44.1 Case Presentation

An 8 year old girl has been seen by her GP several times in the past 3 months with persistent coryzal symptoms, cough, green rhinorrhea, nasal obstruction with mouth breathing. She complains that her head feels full.

She has no known comorbidities and was born at term without complication. Her GP advised Mum to try a nasal aspirator, which has helped remove some of the discharge from her nose, and she has had several courses of oral amoxicillin with only short-lived benefit.

On examination, she appears well in herself. Anterior rhinoscopy demonstrates oedematous enlarged inferior turbinates. Percussion over the maxillary sinuses is painless to the patient. The oropharynx is largely unremarkable apart from a trail of mucopurulent discharge from the postnasal space.

This patient was given a prolonged course of doxycycline for 4 weeks and intranasal steroids to be used alongside a saline rinse. Her and her parents were advised that she would need to use the intranasal steroid spray long term to best control her symptoms.

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44.1.1 Background Knowledge

44.1.1.1 Anatomy

The paranasal sinus are air filled spaces surrounding the nasal cavity lined by respiratory mucosal epithelium. They begin to develop at 25–28 weeks gestation as sacculations of the mucosa, these then grow to invade the bones surrounding the nasal cavity to form sinuses. They continue to develop throughout childhood. The maxillary sinus is the first to develop and become aerated; it has two phases of growth, in the first 3 years of life and then again from 7 to 18 years of age. The ostium lies at or above the level of the floor of the nose, lower than in adults. The sphenoid sinus does not begin to develop until 3 years of age. The ethmoid sinus is present during childhood and pneumatization begins in the 4th year of life, with the ethmoid air cells developing during puberty. The frontal sinus develops last, pneumatisation begins from age 2.

The paranasal sinuses are present to aid filtration, humidification and warming of inspired air, they lighten the skull, allow resonance for speech, absorb shock from trauma and contribute to facial growth.

44.1.1.2 Pathophysiology

Kennedy et al. describe the cycle that results in chronic rhinosinusitis. The initial insult is of mucosal congestion or an anatomical variant resulting in closure of the osteomeatal complex

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and blockage of drainage from the sinus. This results in stagnation of mucous, thickening of secretions and a change in pH. Mucosal gas metabolism changes, which in turn damages cilia and epithelium. The combination of changes allows for bacterial growth to occur in a closed cavity with inflammation and bacterial infection. The mucosa thickens further, resulting in closure of the ostium and so the cycle continues.

Chronic rhinosinusitis is likely multifactorial. Predisposing factors include allergy which results in inflammation of the nasal mucosa, with obstruction of the sinus ostia. Adenoidal hypertrophy is believed to contribute by providing a reservoir of bacteria which can infect the sinuses, as well as obstructing drainage via the postnasal space. Impaired ciliary function including cystic fibrosis and primary ciliary dyskinesia prevents the movement of mucous from the sinuses allowing stasis. Gastro-oesophageal reflux disease leads of inflammation of the mucosa in the post nasal space, and immunodeficient conditions such as diabetes leave the patient less able to fight off infection.

44.1.2 Clinical Approach

44.1.2.1 Diagnosis

Ascertaining a history in keeping with chronic rhinosinusitis is often difficult in a child. It is challenging to differentiate persistent symptoms lasting more than 12 weeks, from recurring symptoms that completely resolve in between episodes. Symptoms of CRS are similar to those present during URTI, allergic rhinitis, adenoidal hypertrophy or adenoiditis.

Examination is not as simple as for adults. Performing nasal endoscopy can be difficult, and this is the optimal technique for assessing disease in the nasal cavity and around the middle meatus. First examine the ears to look for evidence of middle ear effusion or Eustachian tube dysfunction. The otoscope can then be used to perform anterior rhinoscopy, assessing the quality of the mucosa and looking for any purulent discharge or obvious polyps. If nasal endoscopy is possible, the presence or absence of concha bullosa should be confirmed, and the middle meatus can be viewed for evidence of polyposis or purulence. If polyps are seen then cystic fibrosis must be considered as a diagnosis.

According the latest European Position Paper on Rhinosinusitis and Nasal Polyps (EPOS) published in 2012, rhinosinusitis is defined as:

'Inflammation of the nose and paranasal sinuses characterised by two or more symptoms, one of which should be either nasal blockage/obstruction/congestion or nasal discharge (anterior/posterior nasal drip): +− facial pain/pressure +− cough (in adults, loss of smell)

And, either:

– endoscopic sign of nasal polyps
– and/or mucopurulent discharge primary from middle meatus
– and/or oedema/mucosal obstruction primarily in the middle meatus

and/or: CT changes with mucosal change within the osteomeatal complex and/or sinuses'.

Chronic rhinosinusitis is defined by EPOS 2012 as:

'Inflammation of the nose and paranasal sinuses characterized by two or more symptoms, one of which should be either nasal blockage/obstruction/congestion or nasal discharge (anterior/posterior nasal drip):

– +− facial pain/pressure
– +− cough
– Persisting for >12 weeks

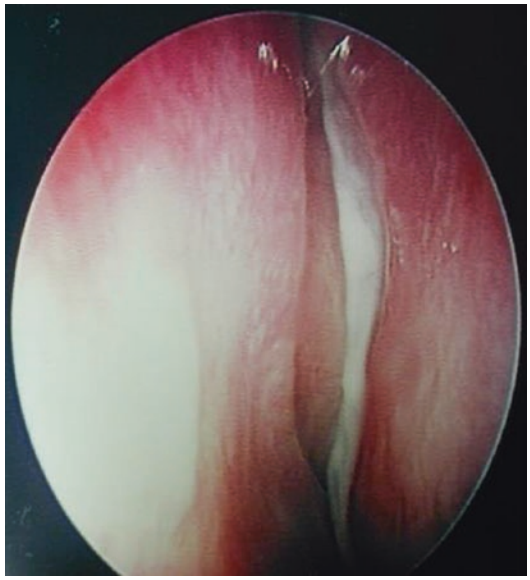
CT imaging of the paranasal sinuses can be used in patients who have symptoms of chronic rhinosinusitis but have not responded to therapy.

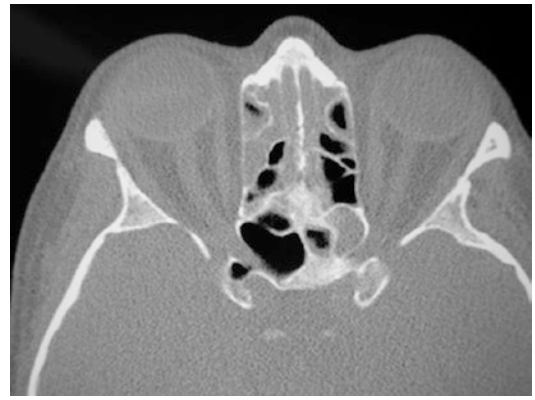
44.1.2.2 Medical Treatment

Saline irrigation is widely used. It can be prepared at home or bought in pre-prepared solutions. It reduces the amount of debris and mucous in the nose, which allows topical treatments to better access the nasal mucosa.

Topical corticosteroid sprays and drops can be used to reduce the amount of oedema and inflammation in the nose, which will help open the ostium to allow drainage from the sinuses.

Antibiotics can be used where patients have an acute exacerbation of chronic rhinosinusitis. Clarithromycin is used most frequently. The duration of treatment is controversial, with most





clinicians agreeing that a minimum of 4 weeks should be prescribed.

44.1.2.3 Indications for Surgery

Surgical intervention is much less common in paediatric patients with chronic rhinosinusitis than in adults. It is most frequently implemented in the management of severe complications of CRS, such as medial subperiosteal orbital abscess or complicated frontal or sphenoidal sinusitis. It is also recommended in cases where the patient has difficult to manage nasal polyposis, such as in cystic fibrosis, or if there is a diagnosis of antrochoanal polyp or inverted papilloma. When the patient's sinusitis is aggravating their coexisting pulmonary disease, for example in asthma or immunodeficiency, there is a weaker indication. Where the concern is impact on quality of life, or simple persistence of mucosal disease despite maximum medical therapy, surgery is less likely to be advised.

44.1.2.4 Surgical Intervention

As chronic rhinosinusitis is often multifactorial, surgery is considered once medical management has been optimised. The adenoid pad has been shown to contribute to paediatric chronic rhinosinusitis, particularly in children under the age of 12. Adenoidectomy should be considered as first line surgical therapy, with 70% of patients demonstrating improvement in their symptoms after surgery.

Ramadan et al. compared adenoidectomy alone, with adenoidectomy alongside maxillary sinus irrigation or balloon sinuplasty with improved outcomes shown at 1 year where adjunctive therapy was used alongside adenoidectomy (87.5% vs. 60.7% and 80% vs. 52.6% respectively).

Endoscopic sinus surgery can also be considered, although there is a trend toward avoiding this as there is associated risk to the nasolacrimal duct and orbit.

Summary and Author's Comments

1. Ascertaining a history in keeping with chronic rhinosinusitis is often difficult in a child.
2. The main therapy is medical with surgical therapy reserved only for those who do not respond to medical therapy.
3. Adenoidectomy should be considered in younger children who have not responded to medical treatment.

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Allergic Rhinitis

45

Elizabeth Kershaw and Aristotelis Poulios

45.1 Case Presentation

Five year old boy with nasal obstruction, persistent rhinorrhea, mouth breathing, sneezing. His symptoms seem to be worse during spring and summer months. He has undergone adenoidectomy in the past along with grommet insertion. However this did not improve his nasal symptoms and he has persistent middle ear effusions now that the grommets have extruded.

His parents have trialed saline nasal sprays which he has tolerated well. Prior to referral, he had not yet had any intranasal corticosteroid treatment as they are often not licensed for use in children under the age of six and the GP was not happy to prescribe them.

He has undergone skin prick testing which has confirmed allergy to grass. He was started on an antihistamine alongside intranasal corticosteroids. On review, despite good compliance with the medications prescribed, his symptoms persist and his school have commented that he is drowsy in class and that other children have started teasing him about his rhinorrhea.

He has been referred to the paediatric immunologist for consideration of immunotherapy for his grass allergy.

45.2 Background Knowledge

45.2.1 Anatomy

Nasal mucosa is the mucous membrane that lines the nasal cavity. It consists of respiratory and olfactory epithelium, depending on its function. The respiratory epithelium is classified as ciliated pseudostratified columnar epithelium and is made up of ciliated cells, globlet cells and basal cells. The ciliated cells are motile and work to sweep mucous and dirt out of the nose and sinuses; their optimal function is dependent on the quality of the mucous. Goblet cells produce mucous which forms a protective layer coating the nasal mucosa and traps particles that are then transported out of the nose by the cilated cells. In asthma, globlet cell hyperplasia results in overproduction of mucous; there are some schools of thought that believe the same occurs in allergic rhinitis although the evidence for this is limited.

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45.3 Pathophysiology

Allergic rhinitis is the type of allergy that affects the greatest number of people, thought to be 10–30% of the population in western civilisation. It occurs due to a type 1 hypersensitivity reaction. Patients with allergic rhinitis produce IgE in response to exposure to an allergen. This could be house dust mite, pollen, grass, dog, cat etc. The IgE antibodies then bind to mast cells within the nasal mucosa, and the mast cell is now 'sensitised'. Subsequent exposure to the allergen results in the allergen binding with the IgE on the mast cell, which activates the mast cell.

Mast cell activation leads to immediate and late phase reactions. In the immediate phase, histamine is released which causes vasodilation and increased permeability of the blood vessels in the nasal mucosa. In the hours after exposure, cytokines are released which attract leukocytes such as eosinophils, which move into the nasal mucosa and produce their own inflammatory products. This is turn results in further congestion, rhinorrhea and sneezing.

Allergic rhinitis can be classified as perennial (throughout the year, usually due to house dust mite, mould or animal) or seasonal (typically related to pollen exposure). It can be further classified:

- Intermittent <4 days per week or < 4 consecutive weeks
- Persistent >4 days per week or > 4 consecutive weeks
- Mild: normal sleep, no impairment of daily activities, work or school, symptoms not troublesome
- Severe: sleep disturbance, impairment of daily activities, work or school

45.4 Clinical Approach

45.4.1 Diagnosis

The patient will typically describe rhinorrhea, itching, sneezing, and nasal congestion. The onset, duration and severity of the symptoms will allow classification of the type and degree of allergic rhinitis. There is frequently a family history of allergic disease. It is important to ask about the presence of pets in the household, or at homes frequented by the patient.

Examination of the patient should include anterior rhinoscopy and nasal endoscopy where tolerated. Typically, they will have swollen nasal turbinates with excessive mucous in the nasal cavities. They may have middle ear effusions as a result of eustachian tube dysfunction related to mucosal thickening. Associated signs include swollen eyelids and conjunctival oedema. Children may characteristically wipe their nose in an upward sweep with the palm of their hand, known as the 'nasal or allergic salute', which can result in a crease across the nose which is described as the 'transverse nasal crease'.

Differential diagnosis can include adenoidal hypertrophy, deviated nasal septum, nasal polyps. These are all obstructive in nature and thorough examination of the patient should rule them out. Infectious rhinitis is usually seen in cases of undiagnosed sinusitis. Vasomotor rhinitis is more commonly seen in the adult population.

Diagnosis of allergic rhinitis can be confirmed with either skin prick testing or a RAST blood test. Skin prick testing involves applying allergen to the skin and then using a lancet to prick the skin (a new lancet should be used for each allergen). The area is then assessed and compared to a negative and positive control area, to demonstrate whether an urticarial reaction has occurred. The patient should not be taking antihistamine in the preceding days as this can affect the results. Skin prick testing is not tolerated by all children, particularly young children. Blood IgE levels can be tested to demonstrate allergy to specific allergens.

45.4.2 Medical Treatment

The first line of treatment is avoidance of the allergen, where known. House dust mite allergy can be improved by changing behaviours at home. The patient's family should be advised to reduce excess humidity where possible. Bedding

should be either hot washed frequently or placed in the freezer along with stuffed toys. Stuffed toys and unnecessary cushions should be kept away from the bed. Bookshelves accumulate dust and should be kept out of the bedroom. If it financially feasible, hard floors are preferable over carpet.

If the allergy is to a pet, then this is an emotional decision to be made by the family. They must weigh up the importance of the pet to the family, and the severity of the child's symptoms.

Avoiding pollen and grasses is incredibly difficult as they are airborne in the atmosphere, although keeping the windows closed during pollen season can be beneficial as well as using air conditioning if possible.

Antihistamines counteract the effects of histamine. They reduce the degree of oedema and itching in the nose. They can be taken orally or topically and can be bought over the counter or prescribed by a doctor. Older antihistamines frequently have a sedating effect and if taken in the morning can affect the child's ability to concentrate at school. Newer options are less sedating and often preferred by patients.

Intranasal corticosteroids can improve all symptoms, reducing nasal congestion as well as sneezing, itching and rhinorrhea. There are combination corticosteroid-antihistamine preparations available which can be used in moderate-severe persistent allergic rhinitis where antihistamine monotherapy has been unsuccessful.

Nasal saline irrigation can improve symptoms by reducing the allergen load within the nose.

Immunotherapy can be used for patients with refractory allergic rhinitis or those who suffer from side effects from taking medications, and the patients need to be referred to and assessed for suitability by an immunologist. This involves exposure to the allergen, allowing the patient's immune system to build resistance and reduce the intensity of symptoms on exposure. There are two options available, injections or sublingual tablets. The injections are given over 3 to 5 years and involve a gradually increasing dose of allergen until a maintenance dose is achieved, after which the time between injections is gradually increased. Sublingual tablets or drops are started prior to the allergy season starting and are continued for up to 3 years. Both have been shown to be effective in reducing patient symptoms and may prevent progression of rhinitis to asthma in children.

Topical decongestants are often popular amongst patients as they are available over the counter and often give instant relief from nasal congestion. They do not treat allergic rhinitis. They are contraindicated in patients with hypertension or cardiovascular disease, pregnant women and men with prostate enlargement. They are effective for several hours, but prolonged use typically results in rhinitis medicamentosa with worsening symptoms. Oral decongestants don't result in rhinitis medicamentosa but should also be avoided in those with cardiovascular disease.

Summary and Author's Comments

1. The first line of treatment is avoidance of the allergen, where known.
2. Antihistamines, intranasal corticosteroids, nasal saline irrigation and immunotherapy are other forms of treatment.

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Septal Deviation

46

Elizabeth Kershaw and Aristotelis Poulios

46.1 Case Presentation

Eight year old boy presents with right sided nasal obstruction. He denies any rhinorrhea or facial pain. He is otherwise well. He reports very occasional scanty epistaxis that resolves with anterior pressure. There is no history of childhood trauma.

When questioned, Mum admits that childbirth was complicated, requiring forceps delivery. She has always wondered if her sons face was injured as his nose has never looked like her or her husband's.

Past treatments have included antihistamines, steroid nasal sprays and salt water rinsing. None of these changed the sensation of a blocked nasal cavity.

The patient allows thorough examination. The septum is deviated into the left nostril. There is no evidence of polyp disease or any other obstructive cause.

46.2 Background Knowledge

46.2.1 Anatomy

The nasal septum is a vertical structure in the midline that separates the two nasal cavities. It is made from cartilage and bone. The bones contributing to the bony part, include the vomer, ethmoid, maxillary, palatine and nasal bones.

The anterior portion of the septum is cartilage. Inferior to this is the crest of maxilla. Posteriorly, the cartilage is attached to the vomer inferiorly and the perpendicular plate of the ethmoid bone superiorly. The vomer sits on the crest of the palatine bone. The crest of the nasal bones from the most superior part of the nasal septum.

46.3 Pathophysiology

Nasal septal deviation can be present at birth, or can occur later in children. When present at birth, this has either occurred during fetal development or during delivery.

Traumatic causes include injury due to contact sport, falls, fights or motor vehicle accidents amongst others. Occasionally deviation can be the result of medical intervention, such as inserting breathing apparatus via the nasal cavity.

With all traumatic causes, the result is dislocation of the bone or cartilage from its adjacent structures, with the formation of fracture lines

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and protrusion of the nasal septum into one or both nasal cavities narrowing the lumen and reducing airflow.

Airflow is dependent on resistance. Poiseuille's law tells us that the rate of airflow is proportional to the fourth power of the radius of the conduit, or nasal cavity in this case. A small decrease in the cross sectional area of the nasal cavity, will have a substantial effect on the airflow.

Inflammatory conditions, such as rhinitis, can make the patient aware of an already deviated nasal septum by further narrowing the nasal cavity.

46.4 Classification

Over time there have been many classifications of nasal septum deformity or deviation. Firstly, we can classify the degree of obstruction by looking at how far the septum is deflected from the midline to the lateral nasal wall. Secondly, the shape of the deformity is described. This can be as extensive as Mladina et al. listed seven types of deformity; Type 1: unilateral vertical septal ridge in the valve region that does not reach the valve itself, Type 2: unilateral vertical septal ridge in the valve region touching the nasal valve, Type 3: unilateral vertical ridge located more deeply in the nasal cavity, Type 4: S-shaped, Type 5: almost horizontal septal spur, Type 6: massive unilateral bone spur, and Type 7: variation. More recently Lee and Baker have given a more simplified classification; the caudal septum is straight but deviated from the midline and is usually displaced from the maxillary crest, C-shaped septal deformity in the vertical plane, C-shaped septal deformity in the horizontal plane, S-shaped septal deformity in the horizontal plane, and S-shaped septal deformity in the vertical plane.

46.5 Clinical Approach

46.5.1 Diagnosis

A child with nasal obstruction should have a thorough history and examination to rule out other causes such as allergic rhinitis, choanal atresia, adenoidal hypertrophy, pyriform aperture stenosis or nasal polyps. There are various conditions that can lead to nasal obstruction, and the treatment of each is different.

46.5.2 Medical Treatment

Saline nasal douching, steroid nasal sprays and antihistamines can benefit those patients who have an element of rhinitis or rhinosinusitis compounding the underlying structure issue of a deviated nasal septum.

46.5.3 Surgical Treatment

Once a diagnosis of deviated nasal septum is confirmed, and medical therapy has not been beneficial symptomatically for the patient, a discussion can be had regarding the surgical options. This is somewhat different than what is in adults in that any nasal surgical procedure in children may affect the relevant growth centres and result in a cosmetic deformity. The patient must be informed of alternative therapy options, such as continuing with medical management or not taking any treatment at all. They must also be informed of the risk of surgical intervention, which includes nosebleed, septal haematoma, infection and septal abscess, septal perforation, change in shape of the nose, tooth or nose numbness. Most ENT surgeons elect to perform the operation when the children reach the age of 16 years. If however, the operation is deemed absolutely necessary then a minimal septoplasty with very little removal of cartilage can be performed.

46.5.4 Follow up

Patients undergoing septoplasty are usually seen 6 weeks post operatively, to ensure that the procedure was successful. Not all patients will feel that their symptoms are fully resolved and they should have been counselled before the

surgery regarding this to manage their expectations.

Summary and Author's Comments

1. Nasal septal deviation can be present at birth which occurred during fetal development or during delivery or from a nasal trauma.
2. Surgery is an option but as minimal as possible due to the theoretical risk of damage to the relative growth centres.

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Antrochoanal Polyp

47

Heidi Jones and Aristotelis Poulios

47.1 Case Presentation

You see an 11 year old girl in clinic. She tells you she has had increasing unilateral nasal flow obstruction and rhinorrhea for the past few months. There is no history of pain or anosmia, and she feels generally well. Her parents tell you that she has recently become a loud snorer. She has no other significant medical history.

Examination demonstrates a right sided nasal mass which appears to arise from the middle meatus, and extends posteriorly to the nasopharynx. The pedunculated mass is just visible via the oral cavity. Examination of the left nasal cavity is unremarkable, as is the rest of your ENT examination.

CT scanning confirms the presence of an antrochoanal polyp, ACP. The ACP is excised en toto, via an endoscopic intranasal approach. There is no evidence of recurrence at subsequent follow up.

47.2 Background Knowledge

ACP is a benign mass which originates in the maxillary antrum, and extends to the nasal cavity or nasopharynx via the middle meatus or an accessory ostium. The classical description is of a unilateral mass with a cystic antral portion and solid polypoid nasal portion. The location and consistency described give rise to the dumbbell shape reported.

Whilst ACPs are also found in adults, they are comparatively more common in the paediatric population and represent up to 42% of all polyps.

The exact pathogenesis is unknown. One theory is that they develop from an intramural cyst which expands and then overspills into the nasal cavity via the least resistant path, the maxillary sinus ostium. Anatomical variations have been shown to be a risk factor in one paper. In children allergic ACPs are more common than inflammatory ACPs, suggesting allergy as a risk factor, and studies have demonstrated a positive allergy history in up to 50% of paediatric patients with ACP. In adults inflammatory ACPs are comparatively more common.

47.3 Clinical Approach

47.3.1 Presentation

Patients usually present with unilateral nasal flow obstruction and rhinorrhea. Often they report a mass seen in the nasal cavity or oropharynx. Larger ACPs can cause snoring and a sensation of foreign body in the throat.

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47.3.2 Diagnosis

The diagnosis is usually made clinically, and confirmed on imaging. CT shows a hypodense mass. There should be no associated bony destruction, but there may be a dilated ostium secondary to mass effect of the enlarging ACP.

47.3.3 Surgical Treatment

Management is surgical. Complete excision is advocated so as to reduce the risk of recurrence. This is usually performed via functional endoscopic sinus surgery, FESS. Reported recurrence rates are up to 15%, making it necessary to follow up patients for at least 2 years.

Summary and Author's Comments

1. ACPs are unilateral benign lesions
2. ACPs do not cause bony destruction
3. Treatment is by complete excision using FESS

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Cystic Fibrosis

48

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48.1 Case Presentation

A 3 year old girl is referred with nasal obstruction by the Paediatricians. She was born at term with rectal prolapse, and was found to carry the cystic fibrosis gene during her newborn screening. She is under the care of the Paediatricians as her pulmonary function tests are abnormal and she has recurrent chest infections.

The Paediatrician had noticed that she had fleshy masses in her nose, and was concerned regarding the possibility of nasal polyps. She has already been started on a course of topical nasal steroids, and has had courses of antibiotics to cover her chest infections. These have minimal effect on her nasal airflow.

She is started on a course of anti-pseudomonal antibiotics alongside the intranasal steroids and her progress monitored. A conversation is had with the parents regarding the options of surgically removing the polyps. The ENT surgeon then communicates this plan with the Paediatricians to ensure that surgery is only considered if it is felt that it will help the patient's nasal symptoms and respiratory function.

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48.2 Background Knowledge

48.2.1 Anatomy

Cystic fibrosis is an autosomal recessive hereditary disease. The responsible gene was discovered by Dr. Tsui in 1989 and was found on chromosome 7q. So far scientists have documented over 1900 mutations, and by creating a database they hope to find mutation-targeted therapies. The gene codes for the production of cystic fibrosis transmembrane regulator (CFTR) which is a membrane-associated protein thought to be responsible for regulating chlorine and sodium ion transport across epithelial cell membranes.

48.2.2 Pathophysiology

Cystic fibrosis affects nearly all exocrine glands resulting in the production of viscous eosinophilic secretions that blocks the duct lumen. In the nose, these thickened secretions block the ostia of the paranasal sinuses. Stagnation of mucous within the sinus is accompanied by bacterial colonization, typically with *Pseudomonas aeruginosa*, *Haemophilus influenza* and anaerobes. A hypoxic environment develops which damages the cilia and causes mucosal oedema and inflammation, resulting in long term damage to the mucosal lining of the sinuses.

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Nasal polyps are pale, fleshy masses which usually arise from the middle meatus bilaterally in the presence of inflammation.

48.3 Clinical Approach

48.3.1 Diagnosis

Newborn screening in the UK includes cystic fibrosis, looking for the presence of immunoreactive trypsinogen (IRT) in dried blood spots.

Pre-natal testing can be performed where there is thought to be a risk of cystic fibrosis. The foetus must inherit a mutated copy of the CFTR gene from both parents, so if one parent is negative then there is no need to further test the other parent or perform amniocentesis or chorionic villus sampling. However, it should be noted that negative test results do not always mean the foetus does not have cystic fibrosis as not all the gene mutations are known.

Most commonly, the sweat test is used to diagnose cystic fibrosis. This involves giving the patient pilocarpine to stimulate sweating, performed using iontophoresis, a process where electrodes are placed onto the substance and onto the skin, producing a voltage gradient on the skin to allow transdermal drug delivery. The sweat is collected and analysed for abnormal amounts of sodium and chloride.

Almost all children with cystic fibrosis will have radiological evidence of sinus mucosal abnormalities by the age of 1, and 20–40% of these children will develop nasal polyps.

Nasal polyps are not frequently seen in the paediatric population, so their presence should always trigger investigations to rule out cystic fibrosis as an underlying diagnosis if not already known. Polyps can also be a result of a foreign body or tumour.

Examination of the nose is challenging in the children. Anterior rhinoscopy will often demonstrate large polyps, however if possible flexible nasal endoscopy should be performed to confirm the extent of the disease.

48.3.2 Medical Treatment

Topical nasal steroids and saline douching are the first line therapy for nasal polyps and sinusitis. Antibiotic therapy should ideally be anti-pseudomonal, however, ciprofloxacin is not recommended for use in children due to the risk of tendon rupture and seizure, so IV treatment may be necessary.

48.3.3 Indications for Surgery

Surgery is recommended in cases where the disease is refractory to optimal medical management. In patients with underlying lung disease, sinus infections can aggravate this, and in those cases the patients have shown symptom improvement but no improvement in pulmonary function.

Surgical risks are higher due to the amount of mucosal inflammation increasing the risk of bleeding. Patients with cystic fibrosis can have vitamin K deficiency due to poor gut absorption, and a coagulation screen should be performed prior to consideration of surgical intervention. Vitamin K supplements can be provided if necessary.

48.3.4 Surgical Treatment

Functional endoscopic sinus surgery with nasal polypectomy is the procedure performed for these children. The polyps will typically return, however improving the drainage pathways from the sinuses should prevent episodes of recurrent sinusitis or chronic rhinosinusitis from developing.

48.3.5 Follow Up

Cystic fibrosis patients are under the long term care of the Paediatricians. The ENT surgeon will see the patient postoperatively to ensure there are no complications, or that further surgery is not required. The paediatricians can refer the patient back to the ENT department if the nasal symptoms deteriorate.

Summary and Author's Comments

1. Although not every child with nasal polyps will also have cystic fibrosis, this needs to be investigated in all children with nasal polyps.
2. A coagulation screen should be performed prior to consideration of surgical intervention.
3. Surgical risks are higher due to the amount of mucosal inflammation increasing the risk of bleeding.

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Choanal Atresia

49

Heidi Jones and Aristotelis Poulios

49.1 Case Presentation

You are called to see a neonate in respiratory distress. The obstetric history and birth were uncomplicated. You are told that the neonate became distressed and cyanotic soon after birth. The cyanosis appeared to settle on crying but resumed when crying stopped. Currently there is an oropharyngeal airway in situ, the neonate appears well and settled. Nasal mist test is negative, and bilateral choanal atresia is seen on flexible nasendoscopy.

A CT of the nasopharynx confirms mixed (bony and membranous) choanal atresia. Further investigation for features of CHARGE syndrome are negative.

The neonate goes on to have a repair via an endoscopic transnasal approach with a good functional outcome.

49.2 Background Knowledge

Choanal atresia is the anatomical obstruction of the usually patent posterior nasal aperature. This is thought to be secondary to failure of regression of the nasal buccal membrane during embryonic development. It is more common in females, 2: 1.

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The obstruction can be unilateral (70%), or bilateral (30%). The obstructing tissue is membranous, osseous, or most commonly a mixture of both.

Although choanal atresia can occur in isolation (50%), it is important to recognise the association with CHARGE syndrome (ocular coloboma, heart defects, choanal atresia, growth retardation, genital anomalies, and ear abnormalities), so as to identify associated diagnoses.

49.3 Clinical Approach

49.3.1 Diagnosis

The diagnosis is a clinical one, which is confirmed on imaging.

Presentation is varied, depending on whether the obstruction is bilateral or unilateral.

Unilateral atresia can cause mild or no symptoms. Consequently, it may be identified in adulthood, or incidentally, for example when it is not possible to pass a nasogastric tube on one side of the nose. Mild symptoms include nasal discharge and nasal airflow obstruction.

Bilateral atresia presents as respiratory distress and cyanosis in the neonate. This is because neonates are obligate nasal breathers. When the neonate cries, and therefore inhales via the mouth, the cyanosis settles. This pattern of

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Table 49.1 Screening for associated features of CHARGE syndrome

Anomaly	Screening
Ocular **C**oloboma	Opthalmology review
Heart defects	Cardiac echo
Choanal **A**tresia	Nasendoscopy and CT scan of paranasal sinuses
Growth **R**etardation	Paediatric review
Genital anomalies	Ultrasound of renal tract
Ear anomalies	Audiological assessment

cyclical cyanosis is classical of bilateral atresia. In the short term, the respiratory distress can be addressed by the placement of an oropharyngeal airway, until definitive treatment occurs.

CT scanning is the investigation of choice. It confirms the diagnosis, and delineates the extent and nature (osseous versus membranous) of the atresia.

As well as investigation of the atresia itself, it is essential that all patients additionally be screened for other features of CHARGE (Table 49.1).

49.3.2 Treatment

Asymptomatic, unilateral atresia can be managed conservatively.

For bilateral, or symptomatic atresia, the management is surgical. The literature suggests that earlier surgical management is associated with higher success rates. Whilst a number of surgical approaches are described in the literature, most commonly a transnasal endoscopic approach is used. The thickened posterior vomer is removed, and the membrane is perforated and dilated.

Some surgeons advocate the use of postoperative nasal stenting, so as to prevent reocclusion of the choanae. This remains controversial owing to the associated complications, including infection and nasal septal perforation. Where stenting is not used, early re-examination and resection of granulation tissue is warranted to reduce the risk of reocclusion.

Follow up with repeated endoscopy is recommended for a minimum of 1 year, to assess for reocclusion. Routine follow up imaging is not routinely recommended.

Summary and Author's Comments

1. The use of an oropharyngeal airway can be used in the emergency management of respiratory distress in the neonate with choanal atresia
2. Choanal atresia may be isolated, or part of CHARGE syndrome; all patients must be screened for the associated anomalies
3. CT scanning aids surgical planning
4. The preferred surgical approach is transnasal endoscopic.

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Pyriform Aperture Stenosis

50

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50.1 Case Presentation

You are called to see a neonate who has a history of respiratory distress and cyanosis associated with feeding. The paediatric team were unable to pass a nasogastric tube bilaterally, and wonder if the neonate might have choanal atresia. The obstetric history and birth were uncomplicated. Nasal mist test indicates partial obstruction, and you are unable to pass a paediatric flexible nasendoscope beyond the anterior nasal cavity.

CT scanning confirms pyriform aperture stenosis.

Although there is some response to nasal decongestants, the child fails to thrive and as such goes on to have surgical repair via an endo oral sublabial approach. On subsequent clinical follow up, the neonate has no further symptoms and is thriving.

50.2 Background Knowledge

Pyriform aperture stenosis is the congenital narrowing of the narrowest part of nasal cavity. The pyriform aperture is the most anterior and narrow part of the nasal cavity and is formed by the horizontal and nasal processes of the maxilla, the anterior nasal spine, and the nasal bones. Stenosis is thought to be secondary to the overgrowth of the maxilla during embryonic development, although the precise pathology is unknown.

The presentation varies according to the degree of stenosis. Pouseille's law dictates that even a small reduction in the normal diameter of aperture can result in a significant reduction in air flow. Milder cases may be symptomatic only during periods of feeding or distress. Because neonates are obligate nasal breathers, the most severe cases present with respiratory distress and may require endotracheal intubation. Pyriform aperture stenosis may be mistaken for choanal atresia, a more common cause of nasal obstruction. In pyriform aperture stenosis, the obstruction is more anterior, which can help differentiate the diagnosis clinically.

Pyriform aperture stenosis can occur in isolation or can be associated with other craniofacial anomalies, including the presence of a mega incisor (a large, single, central maxillary incisor) and holoprosencephaly.

50.3 Clinical Approach

50.3.1 Diagnosis

A diagnosis of nasal obstruction is made clinically. Whilst there may be clinical suspicion about pyriform aperture stenosis, diagnosis is made on CT scanning. Thin (1–1.5 mm) axial

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slices are taken in line with the hard palate. Classic features include overgrowth and medial displacement of the nasal processes of the maxilla. A pyriform opening of <11 mm in a full term neonate is generally considered diagnostic. Commonly there is an associated bony ridge on the caudal aspect of the palate, as well as abnormal dentition.

Where there is evidence of a mega incisor, holoprosencephaly should be suspected and investigated with MRI.

50.3.2 Medical Treatment

Mild cases can be managed with saline nasal drops and topical decongestants, which can provide temporary improvement until the nasal bones grow and the stenosis effectively widens.

Nasal stenting can be used to effectively widen the stenosis. However they can be difficult to manage and are associated with soft tissue complications, such as nasal septal perforation.

50.3.3 Surgical Treatment

Marked stenosis associated with respiratory distress or failure to thrive requires surgical management.

Most commonly a sublabial, endonasal approach is adopted. This approach allows good exposure of the pyriform aperture, following which the stenosis can be drilled. Drilling must be avoided at the floor of the nasal cavity, and anterior to inferior turbinate so as to avoid iatrogenic injury to the developing teeth and the nasolacrimal ducts, respectively. Stenting is often used for the first post-operative week to prevent adhesions. Once the surgical site has healed, no further follow up is required.

Balloon dilatation has been trialled as an alternative to surgical drilling for patients refractory to medical management. Balloon dilators are inflated within the nasal cavity, effectively out fracturing the inferior turbinates, so as to widen the pyriform aperture. Whilst the authors report good outcomes, the studies are small, and this method is yet to be widely adopted.

Summary and Author's Comments

1. Pyriform aperture stenosis should be suspected in cases where there is respiratory distress and it is not possible to pass a narrow tube beyond the nasal inlet
2. The diagnosis along with associated anomalies is made radiologically
3. Mild cases can be managed conservatively with topical decongestants
4. Severe cases require surgical correction of the stenosis.

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Meningoencephalocoele

51

Heidi Jones and Aristotelis Poulios

51.1 Case Presentation

You see a 2 year old child in the outpatient department. His parents report a longstanding history of unilateral clear rhinorrhea. On examination there is no external deformity but you note a pale mass within the right nasal cavity. There is no other significant past medical history of note, and the child is otherwise medically well.

CT and MRI scanning demonstrate a right sided meningoencephalocoele, which protrudes into the right nasal cavity from a large defect in the region of the foramen caecum.

The Patient undergoes surgical repair via an endonasal approach, with no post-operative complications. They are subsequently discharged from follow up at 1 year post op.

51.2 Background Knowledge

Meningoencephalocoeles are protrusions of both the meninges and the brain through a bony defect in the skull (Fig. 51.1). They can present as a rare congenital deformity, or as a secondary defect following trauma. Secondary meningoencphalocoeles are beyond the scope of this text.

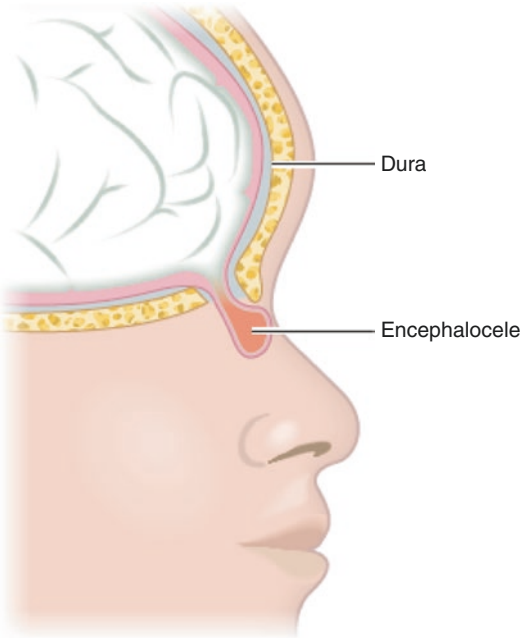

Fig. 51.1 Meningoencephalocoeles are protrusions of both the meninges and the brain through a bony defect in the skull. Encephaloceles are protrusions of brain parenchyma

The congenital variant occurs as a result of failed closure of the anterior neuropore during the 4th week of embryological development. They are classified according to their anatomical location as either frontoethmoidal or basal.

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51.3 Clinical Approach

51.3.1 Diagnosis

Presentation is variable, and often related to the size of the defect. Small meningoencephalocoeles may produce minimal or no symptoms. The main symptom is of nasal obstruction, which in infants can lead to feeding difficulties and failure to thrive. Less frequently there may be a visible deformity or nasal mass. The meningoencephalocoele can become larger, owing to pulsation of the brain. Consequently, symptoms can develop and progress with time. CSF leakage can cause clear rhinorrhea. Smaller defects may cause no symptoms but can lead to recurrent meningitis. Therefore, a meningoencephalocoele must be excluded in patients with recurrent meningitis.

When clinically apparent, meningoencephalocoeles are classically seen medial to the middle turbinate, and present as a pale mass.

Clinical suspicion is confirmed on imaging. CT provides a surgical road map, and is essential to facilitate safe excision.

51.3.2 Management

Management is surgical in all cases, and is performed in collaboration with neurosurgeons. Early operative repair is encouraged, before the meningoencephalocoele has had the opportunity to grow or impact on the developing craniofacial structure or lead to potentially fatal complications. This is balanced against the technical challenge of operating in the smaller field of a younger patient.

The aims of surgery are twofold; to reduce the meningoencephalocoele, and to repair any resultant deformity. Surgery can be done in a single or in two stages.

Traditionally a transcranial approach was adopted to reduce the meningocoele. More recently an endoscopic transnasal approach is commonly used. It is less invasive and produces no visible external scarring. Those in favour also report fewer complications and a reduced in patient hospital stay. However, the narrow operating field causes significant technical difficulties.

The mass is separated from any nasal attachments and delivered back into the cranial vault, following which the defect is repaired with a synthetic or an autologous graft such as fat, cartilage, or fascia lata. The patient should be followed up to assess for complications, such as CSF leak, meningitis, and recurrence.

Summary and Author's Comments

1. Small meningoencephalocoeles may produce little or no symptoms
2. A meningoencephalocoele should be excluded in all patients with recurrent meningitis
3. Early surgical repair is encouraged
4. Although there are technical challenges, endoscopic repair is gaining favour.

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Nasal Glioma

52

Heidi Jones and Aristotelis Poulios

52.1 Case Presentation

You see a 2 month child in clinic. Her parents have noticed a fleshy swelling protruding from the right nostril. It doesn't seem to be growing and does not appear to be associated with any other symptoms. There is no change to size when the child is distressed. The obstetric history and birth were uncomplicated.

Clinically there is evidence of a blue/red fleshy mass within the nasal cavity. It is mobile, and seems to be attached to a stalk. Furstenberg's test is negative (i.e. the mass does not pulsate or increase in size with pressure on the ipsilateral jugular vein).

A CT scan demonstrates a lesion which appears to arise from the right septum. An MRI demonstrates no intracranial extension or associated abnormality. The child goes on to have the mass excised via an endoscopic, intranasal approach. Histology demonstrates a nasal glial heterotopia, NGH, which has been completely excised. The child is discharged at 1 year's follow up, with no evidence of recurrence.

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52.2 Background Knowledge

52.2.1 Anatomy-Pathophysiology-Histology

NGHs are rare congenital nasal masses, which are composed of heterotopic neural tissue. The pathogenesis is thought to be similar to that of a meningoencephalocoele; initially there is failed closure of the anterior neuropore during embryological development, which results in failed regression of the forebrain. In a meningoencephalocoele the protrusion of the meninges and brain persists, whilst in a nasal glial heterotopia the connection obliterates, leaving an isolated mass of neural tissue. Although this mass may sometimes be connected to the brain via a stalk, the stalk does not contain neural tissue or a CSF channel, distinguishing it from a meningoencephalocoele (Fig. 52.1).

NGHs can be extranasal, intranasal, or both. Presentation is dependent on the location of the mass. Extra-nasal heterotopias can sometimes be identified on prenatal ultrasound scanning. Clinically, NGHs present with a visible mass that is usually in the glabellar region but can be between the glabella and the nasal tip. It is classically blue/purple, firm, and non-pulsatile. Intranasal masses can be mistaken for intranasal polyps, and very small masses may not be clinically obvious. Unlike meningoencephalocoeles, there is no change to size, visible pulsa-

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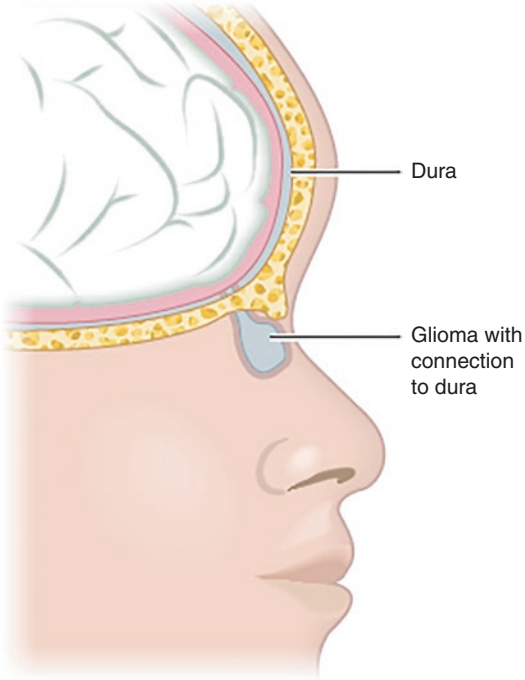

Fig. 52.1 Anatomical relationships of gliomas

tion with increased intracranial pressure (Furstenberg sign) as there is no intracranial component.

Although they do not grow, they can impact on the growth of the surrounding nasal skeleton, and therefore result in the development of deformities. There may be associated nasal obstruction, epiphora, epistaxis, and CSF rhinorrhea.

52.3 Clinical Approach

52.3.1 Diagnosis

Imaging is essential; whilst CT facilitates assessment of any bony defects, MRI allows better soft tissue assessment, as well as screening for any associated intracranial connection.

Diagnosis is confirmed on histological analysis. The mass is comprised of rests of heterotopic, non-neoplastic cells. The term glioma is misleading in that they are not neoplastic.

52.3.2 Surgical Treatment and Post-op Care

Management is surgical in all cases. Early operative repair is encouraged, before the nasal glial heterotopia has had the opportunity to impact on the developing craniofacial structure or lead to potentially fatal complications associated with intracranial connections. This is balanced against the technical challenge of operating in the smaller field of a younger patient.

The aims of surgery are twofold; remove the nasal glial heterotopia and to repair any resultant deformity. Intranasal masses can be removed via an endoscopic intranasal approach, whilst extra nasal masses are removed via an open approach. Several open approaches are described, including lateral rhinotomy, external rhinoplasty, midline nasal incision, and bicoronal incision. The exact approach adopted is dependent on the nature of the mass; in particular its size, location, and any associated craniofacial deformity.

Up to 20% of NGHs are associated with a stalk that connects with the brain. Regardless of the approach adopted, it is essential to remove the stalk of the mass, so as to reduce the risk of meningitis and recurrence.

There is a reported recurrence rate of up to 10%, making it essential to follow these patients up. Similarly it is important to identify complications which warrant further surgical management, such as meningitis, and nasal deformity.

Summary and Author's Comments

1. Glioma is a misnomer; histology shows non neoplastic heterotopic cells.
2. NGHs are distinct from meningoencephalocoeles in that there is no direct intracranial connection.
3. Surgical management aims to remove the mass, remove any associated stalk, and address any deformity.
4. There is recurrence in up to 10% of cases, making follow up mandatory.

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Nasal Dermoids

53

Heidi Jones and Aristotelis Poulios

53.1 Case Presentation

A 2 month old boy is brought to the ENT outpatient department. His parents describe a history of nasal dorsal mass which has been present since birth. There is a central punctum. More recently this punctum has been discharging intermittently. Clinically there is evidence of a dorsal mass with a central sinus from which a hair is protruding. The child has received no treatment thus far.

A CT scan demonstrates a nasal dermoid cyst, with no intracranial extension.

The patient undergoes an extracranial excision. At the time of surgery, there is no evidence of a sinus.

There is no evidence of recurrence of the cyst at outpatient follow up.

53.2 Background Knowledge

During normal embryological development, the nose is formed by three layers; ectoderm, mesoderm, and cartilaginous capsule. The frontal and nasal bones are separately formed by ossification of the mesoderm. Whilst there remains some debate as to the exact origin of a nasal dermoid, the most widely accepted theory is that of incomplete obliteration of the fonticulus frontalis. In early embryology, the dura extends to the skin by a small projection. During development, the nasal process of the frontal bone extends inferiorly, thereby separating this projection of dura from the skin. As the projection of dura recedes, the nasal ectoderm can follow, thus forming a cyst or sinus (Fig. 53.1).

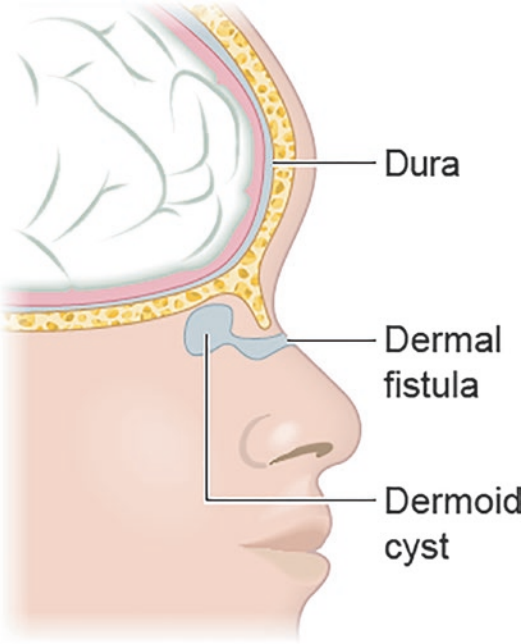

Fig. 53.1 Nasal dermoid due to incomplete obliteration of the fonticulus frontalis

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53.3 Clinical Approach

53.3.1 Pathology

Nasal dermoids typically present with a midline swelling between the columella and glabella. There may be an associated opening of a sinus tract with recurrent discharge and infections. The presence of protruding hair is pathognomic. Presentation is often at birth, and most commonly within the first few years of life.

The differentials of a midline nasal swelling include: Glioma, encephalocoele, epidermoid cyst, haemangioma, teratoma, neurofibroma, arteriovenous malformation, lipoma.

53.3.2 Diagnosis

Diagnosis is radiological, using MRI and CT to assess both the extent of the cyst as well as assessing the bony and soft tissue anatomy involved.

Hartley et al. proposed a classification system based on the cysts extent:

- Superficial
- Intraosseous
- Intracranial extradural
- Intracranial intradural

53.3.3 Surgical Treatment

Surgical excision is essential, so as to prevent infection and reduce bony deformity. Preoperative knowledge of the extent of the nasal dermoid is essential in surgical planning, to allow en bloc excision.

Table 53.1 Proposed surgical treatment based on the extent of the pathology

Superficial lesions	Local excision
Intraosseous tracts	Open rhinoplasty and drilling of the frontonasal bones for access
Intracranial extension	Bicoronal flap and frontal craniotomy *or* anterior small window craniotomy (less invasive)

The extent of surgery is guided by the extent of the cyst, as shown in Table 53.1.

Summary and Author's Comments

1. Nasal dermoids are rare embryological anomalies which present in infancy/childhood.
2. CT scanning is essential for diagnosis and surgical planning
3. Classification is based on anatomical extent
4. Treatment is by surgical excision

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Adenoidal Hypertrophy

54

Elizabeth Kershaw and Aristotelis Poulios

54.1 Case Presentation

4 year old boy was brought to the clinic by his parents who described that he was a loud snorer, keeping the family awake at night. On a recent holiday where the family shared a room, they had noticed that he would stop breathing for periods of up to 10 s at night, followed by a loud gasp before he continued snoring. He mostly breathes through his mouth, and eats noisily as he cannot close his mouth when eating.

The family deny any significant issues with rhinorrhea or other coryzal symptoms, there is no history of allergy type symptoms. He is otherwise well.

On examination, he is sat in the clinic breathing through an open mouth. A cold metal spatula is placed under his nose, with minimal misting. He has grade III tonsils (Fig. 54.1). His tympanic membranes are dull with evidence of a middle ear effusion. His hearing is satisfactory.

He is sent for polysomnography which confirms moderate obstructive sleep apnea. He undergoes adenotonsillectomy, and on review in the outpatient clinic his parents confirm that his apnoeic episodes have fully resolved. On examination, his middle ears are aerated.

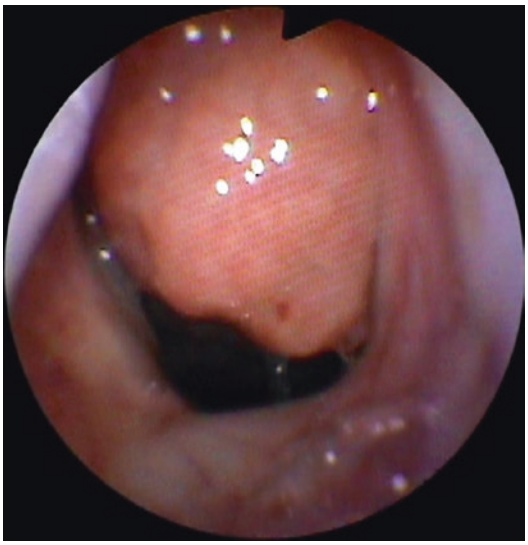

Fig. 54.1 Endoscopic photograph of the postnasal space, demonstrating grade III adenoids

54.2 Background Knowledge

54.2.1 Anatomy

The adenoids are also known as the pharyngeal tonsil, part of Waldeyer ring of lymphoid issue which also includes the palatine and lingual tonsils. Hypertrophy is defined as the increase in

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volume of tissue due to its cells enlarging in size, as opposed to hyperplasia which occurs when the number of cells increases. The mucosa covering the adenoids is typically a combination of pseudostratified columnar and specialised squamous epithelium.

At birth the adenoids are very small. They grow in size, reaching their maximum at about the age of 5, and then from 7 years of age they begin to atrophy. The degree of airflow through the nasopharynx is dependent on the size of the adenoid and the size of the nasopharynx.

54.2.2 Pathophysiology

The adenoids are lymphoid tissue, and when infected they increase in size. The normal response to resolution of infection, is that the adenoid tissue will again reduce in size. In case of recurrent infection, or if bacteria and debris become lodged within the tonsil, chronic infection can result in persistent enlargement of the adenoids.

Infection can lead to the loss of commensal bacteria normally found covering the adenoids, with pathogens being isolated. These can disrupt the balance of cell types, resulting in a relative loss of ciliated cells and mucociliary stasis.

Adenoidal hypertrophy can affect the paediatric patient in several ways. Firstly, it can obstruct the postnasal space either fully or partially, resulting in an inability to breathe through the mouth. The patient subsequently mouth breathes, resulting in altered speech, halitosis, snoring and on occasion apnoeic episodes. They can appear to be failing to thrive as their difficulty breathing can interfere with the ability to eat normally, resulting in reduced oral intake.

The enlarged adenoids can physically obstruct the medial end of the eustachian tube in the nasopharynx, with the patient suffering from eustachian tube dysfunction or hearing loss secondary to otitis media with effusion. If left undiagnosed, the tympanic membrane can become retracted and potentially cause erosion of the ossicles, perforate or develop cholesteatoma.

Obstructive sleep apnoea is a consequence of adenoidal hypertrophy, along with snoring which is seen more commonly. The normal neck musculature works to maintain airway patency, despite the presence of enlarged adenoids (with or without tonsillar hypertrophy). When the child falls asleep, the muscle tone decreases and the pharyngeal walls collapse inwards. In patient with adenoidal hypertrophy, this can result in complete closure of the airway. At this point, the child looks as though they are trying to breathe with movement of the chest but no movement of air. This is known as an apnoeic episode. When there is less than 50% of normal air movement, but not complete obstruction, this is hypoponea. Episodes lasting 10 s or more are those recognised as significant. The patient's carbon dioxide levels start to rise, which is detected by central chemoreceptors. The patient awakens to some degree, with the return of muscle tone and reintroduction of normal breathing.

If obstructive sleep apnoea is left untreated, the child is often restless throughout the night and difficult to rouse in the morning. They are excessively sleepy throughout the day with periods of hyperactivity and poor behavior. They often fall behind their peers at school because of their inability to concentrate. Long term complications of hypoxia include damage to the cardiovascular system with an increased risk of heart failure, stroke and ischaemic heart disease.

54.3 Clinical Approach

54.3.1 Diagnosis

The history obtained from the child and parents is the most useful tool. They will typically describe nasal obstruction, loud mouth breathing throughout the day, inability to chew food with the mouth closed, snoring and, less frequently, apnoeic episodes at night. It is important to ask about hearing due to risk of associated otitis media with effusion.

Examining the paediatric patient can be challenging. The physician should be able to rule out other causes of nasal obstruction, such as septal

deviation, chronic rhinosinusitis, allergic rhinitis, which will assist in making the diagnosis of adenoidal hypertrophy. If tolerated, flexible endoscopy can be performed to assess the size of the adenoids. Radiography is not routinely used in the UK, with correlation between X-ray and endoscopy findings being poor.

The gold standard investigation for obstructive sleep apnoea is polysomnography, which involves the use of an electroencephalogram, electrooculogram, electromyogram and electrocardiogram. This is not available in all centres, and the sleep study that is most frequently used measures only heart rate and oxygen saturations and is much more rudimentary.

Medical management is reserved for treatment of adenoiditis; however it will not resolve the problems associated with enlarged obstructing adenoid tissue.

Adenoidectomy is recommended for patients with a diagnosis of obstructive sleep apnoea. This is usually performed alongside tonsillectomy in the paediatric population, as unsuccessful adenoidectomy will lead to the patient requiring a further general anaesthetic and surgical procedure. It is also recommended for patients with conductive hearing loss secondary to otitis media with effusion, when ventilation tubes alone have been tried, and the effusion has returned.

Adenoidectomy can be performed using several techniques. Most commonly performed using an adenoid curette, which is a quick, inexpensive method. Technique is important, to avoid leaving residual tissue or damaging adjacent structures. Monopolar suction diathermy is performed under indirect vision with a mirror. This allows more accurate reduction of the adenoidal tissue with disruption of the biofilm. A microdebrider can be used alongside a rigid nasal endoscope to reduce the adenoid tissue under direct vision. Coblation adenoidectomy performed alongside nasal endoscopy is also described with good results.

Patients undergoing adenoidectomy are typically follow up in the outpatient clinic postoperatively to ensure resolution of their symptoms. Adenoidal regrowth can occur, more frequently in children under 5 years of age. These children may require revision adenoidectomy if still symptomatic.

Summary and Author's Comments

1. Adenoids grow in size, reaching their maximum at about the age of 5, and then from 7 years of age they begin to atrophy.
2. Obstructive sleep apnoea is a consequence of adenoidal hypertrophy, along with snoring which is seen more commonly.
3. Adenoidectomy is recommended for patients with a diagnosis of obstructive sleep apnoea.

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Facial-Nasal Trauma

55

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55.1 Case Presentation

You see a 7 year old boy, who presents acutely having fallen from his bicycle, and struck his face to the ground. The patient has already been assessed by the trauma team. They have concerns regarding facial trauma, but report no other significant injury elsewhere.

Clinically, you note significant facial bruising and swelling, and the nose appears deviated. There is no nasal septal haematoma. The mechanism of injury is in keeping with the facial injuries observed, and the child has a good rapport with his parents. There is no physical evidence of recent previous injury. He has no significant past medical history.

A CT scan of the facial bones confirms the suspected nasal fracture. There are no associated nasoethmoidal or facial fractures. The patient goes on to have a closed reduction under general anaesthetic at 7 days post injury, with a good cosmetic outcome.

55.2 Background Knowledge

Trauma is the leading cause of paediatric morbidity and mortality. In general the incidence of facial fractures increases with age. As the childs age and mobility increase and their level of supervision decreases, the velocity and therefore the severity of injury tends to increase.

Although the fracture patterns seen in the paediatric population tend to be similar to that seen in adults, the incidence of each is different owing to the anatomical differences of the paediatric facial skeleton. Whilst the sinciput is most prominent in infancy, as children develop, the midface and mandible become relatively more prominent (Table 55.1).

55.3 Clinical Approach

55.3.1 Diagnosis

As in any case of paediatric injury it is essential to be mindful of non-accidental injury, NAI. The clinician must consider the following signs; whether injury is in keeping with the mechanism of injury, whether the mechanism is in keeping with the child's development, whether there are

Table 55.1 Relative incidence of facial fractures in children, as reported by Alcalá-Galiano et al.

Fracture location	Incidence (%)
Nasal	59
Mandibular	22
Orbital	10
Frontal	5
Midfacial	4
Complex (le fort/ naso orbito ethmoid	2

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any signs of previous / healing injuries, whether there has been a delay in presentation, and whether the child has an appropriate rapport with their accompanying guardian.

An underlying fracture should be considered where there is significant swelling, asymmetry, or mobile segments. Clinical suspicion is best confirmed with CT. As with adults, a nasal septal haematoma should be clinically excluded in all children with facial trauma.

Paediatric facial fractures usually occur in the setting of severe trauma and as such are unlikely to occur in isolation, with associated neurocranial injuries most common. As is true for any trauma case, it is essential that the clinician does not miss any associated injuries through focusing on distracting facial injuries. All patients should have a trauma survey completed in line with ATLS principles.

55.3.2 Medical/Surgical Treatment

A large proportion of facial trauma, including mandibular, orbital, frontal, midfacial, and complex fractures, is usually managed by the maxillofacial team. Although specific detail is beyond the scope of this book, the approach is usually more conservative than that for adults, so as to prevent growth disturbance.

As with adults, isolated traumatic nasal deformities are manipulated at an interval of 1–2 weeks, allowing any associated swelling to subside for thorough assessment of the appearance of the nose. Septal haematomas should be drained urgently under general anaesthetic to reduce the risk of necrosis, infection, and growth disturbance.

Summary and Author's Comments

1. Be mindful of non-accidental injury in children presenting with facial injuries
2. Ensure associated injuries are not missed in the presence of a facial injury
3. Management aims to reduce fractures by the most conservative method, so as to allow normal development of the growing skeleton.

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Part VIII

Olfactory Disorders

Anosmia: Conductive

56

Sarantis Blioskas

56.1 Case Presentation

A 35-year-old female was referred to the ENT OPD with a long history of anosmia (>12 months). The patient reported significant longstanding nasal obstruction and occasional nasal discharge that has troubled her for years and was treated with decongestant and nasal steroid sprays. Reduction of smell was gradually evident, especially during acute exacerbation of symptoms in the course of viral infections. In the last 12 months, there was almost constant complete anosmia resulting in a significant decline in quality of life. The patient reported that she could no longer tell when she needed to change diapers to her baby, and she had food beyond the expiration date because she was unable to detect spoiled food. On examination, anterior rhinoscopy revealed grade 4 nasal polyposis that almost fully occluded both nasal cavities, extending to olfactory clefts bilaterally (Fig. 56.1). Flexible nasendoscopy was impossible to perform due to complete occlusion by polyps. A CT imaging of sinuses revealed extensive chronic rhinosinusitis with polyps with bilateral osteomeatal complex obstruction and polyps extended upwards to olfactory clefts (Fig. 56.2). The patient was prescribed maximal medical therapy, including oral and nasal steroids and oral antibiotics, and was listed for functional endoscopic sinus surgery. Conservative treatment was unsuccessful in treating her anosmia; however, following surgery patient reported a significant improvement in symptoms.

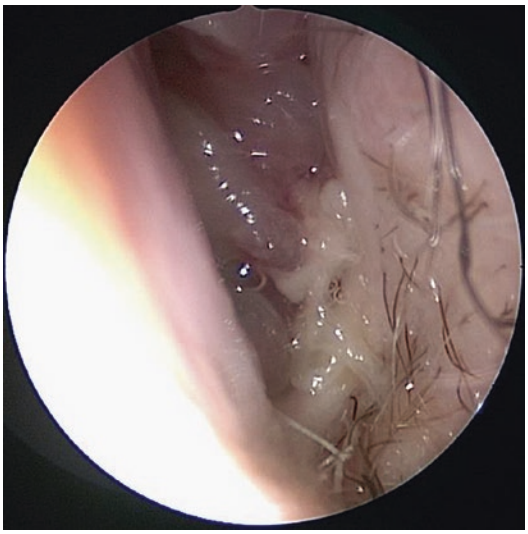

Fig. 56.1 Grade IV nasal polyposis

56.2 Background Knowledge

In all living organisms, including humans, olfaction can play a significant role in terms of feeding, territorial adjustment, as well as detection of

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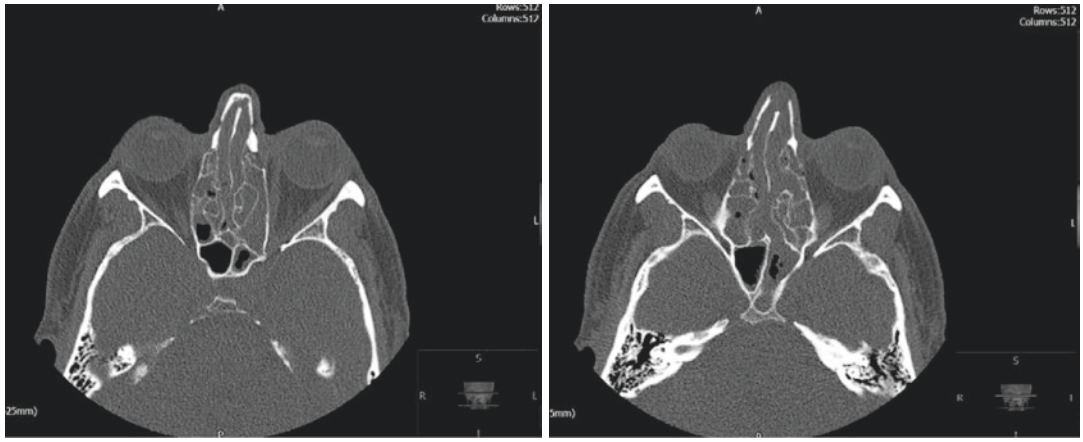

Fig. 56.2 Diffuse CRS with nasal polyps resulting in conductive anosmia

harmful conditions such as fire, gas, and spoiled food. Thus olfactory impairment should be considered as a significant deficit.

In terms of physiology, odorants are mostly hydrophobic compounds that vary widely in structure, yet their common characteristic is that they can stimulate olfactory receptors which are located in the olfactory epithelium at the roof of the nasal cavity. Olfactory receptors of the nasal cavity detect and interact with these chemical ligands and their interactions induce neuronal signals that are transmitted to the olfactory bulb and ultimately to the corresponding areas of the brain

Odorants reach olfactory receptors during the process of nasal inhalation and exhalation. Consequently, any form of nasal obstruction can limit the access of odorants to their corresponding areas of neural interaction by limiting the nasal airflow towards these areas. Since physiologically, there is a direct relationship between the olfactory response and the number of odorant molecules delivered to the olfactory receptors, reduced nasal airflow leads directly to the reduced or diminished olfactory ability.

However, such changes in nasal airflow impact on olfactory functions, not merely in terms of access. Sinonasal disease and nasal mucosal changes can affect olfactory processing at many levels, leading to the overall decline in olfactory ability noticed by the patient.

56.3 Clinical Approach

In terms of classification, smell disorders are broadly classified as conductive and sensorineural and such differentiation is particularly important in clinical consultations to establish the initial work-up. The potential causes of nasal airway blockage that can potentially lead to conductive anosmia can be roughly classified as in Table 56.1.

For cases of conductive anosmia, the diagnostic algorithm can generally be as follows.

56.3.1 History

The importance of a detailed history cannot be overemphasized. The time course (sudden onset or gradual declination) of symptomatology is of great importance. In the same concept, the patient can also recall a temporal connection between the sudden or gradual onset of anosmia and infection such as rhinitis or rhinosinusitis. The presence of additional nasal symptoms like nasal discharge, postnasal drip, nasal obstruction, facial pain, sneezing and itching, as well as comorbidities like allergies, inflammatory diseases (ANCA vasculitis, sarcoidosis, etc.) is of paramount importance to be sought for. The same applies to potential medication intake (e.g., chronic use of decongestants/rhinitis medica-

Table 56.1 Potential causes of conductive anosmia

Rhinitis	Inflammatory	Infectious	Acute viral rhinitis Acute bacterial rhinitis Primary atrophic rhinitis Secondary atrophic rhinitis
		Non-infectious	Allergic rhinitis Occupational rhinitis Drug-induced rhinitis Idiopathic rhinitis NARES
	Non-inflammatory		Idiopathic rhinitis (sympathetic dysregulation) Drug-Induced Cocaine sniffing Hormonal rhinitis Food-induced rhinitis
Rhinosinusitis	Acute		
	Chronic	Without polyps	
		With polyps	
Tumours	Benign	Inverting papilloma Neurilemoma Ossifying fibroma Osteoma Angiofibroma Adenomatoid hamartoma	
	Malignant	Squamous cell carcinoma Adenocarcinoma Lymphoma Melanoma	
Anatomical factors	Septal deviation Postoperative synechiae Turbinate hyperplasia Stenosis / scarring Choanal atresia		
Granulomas	ANCA vasculitis Churg-Strauss syndrome Sarcoidosis Infectious Midline T cell destructive granuloma		
Congenital disorders	Congenital ciliary defects Cystic fibrosis		

mentosa) or drug abuse (e.g. cocaine sniffing). Additionally, it is significant not to forget to investigate whether the nature of the disorder is continuous or fluctuating, as usually fluctuating symptoms indicate conductive issues.

56.3.2 Clinical Examination

A thorough head and neck clinical examination alone can sometimes accurately pinpoint the cause of conductive anosmia. Anterior rhinoscopy is essential, yet it should always be accompanied by flexible or rigid nasal endoscopy. Endoscopy should always include the area of the olfactory cleft. The presence of polyps, high septal deviation, rhinitis or rhinosinusitis and/or tumors of the nose or anterior skull base should be accurately established through endoscopy. At the same time, the recently proposed Olfactory Cleft Endoscopy (OCES) Scale should be used in the evaluation of the olfactory cleft. The presence of nasal discharge (clear or colored) should also be evaluated. In cases where clinical examination

has indicated an acute cause of conductive anosmia, repeat of the clinical examination after adequate treatment of the acute cause is always recommended.

56.3.3 Rhinomanometry

Rhinomanometry can provide an objective method for evaluating nasal airflow.

56.3.4 Olfactory Function Evaluation

It is always useful to add olfactory function tests in the diagnostic test battery, although their value in conductive anosmia can be limited. Several tests are based upon odor naming and are relatively short and easy to perform. Specific tests like the "Cross-Cultural Smell Identification Test" (CCSIT), the UPSIT family ("University of Pennsylvania Smell Identification Tests"), the "Sniffin' Sticks", the "European Test of Olfactory Capabilities", or the Connecticut Chemosensory Clinical Research Center Test (CCCRC-Test), etc., can all be used. However, as many of these tools include verbal components, one has to consider the influence of cognition and language. Also, the age of the patient has to be kept in mind when interpreting the test results.

56.3.5 Imaging

Both CT and MRI scans can be ordered if indicated by history and clinical examination. However, especially in conductive anosmia, CT scans of paranasal sinuses are superior in identifying a group of patients with the sinonasal disease who may benefit from medical and / or surgical treatment. CT scans can also be superior in detecting slight opacification of the olfactory cleft, which sometimes cannot be adequately visualized endoscopically. Nevertheless, if the non-conductive nature of the anosmia cannot be excluded, then MRI of the brain can be invaluable in differentiating from other causes.

Treatment of conductive anosmia depends on the specific cause, since eliminating the causative factors usually leads to direct improvement of olfactory function as well. Thus, in cases of temporary anosmia due to viral rhinitis, often, no specific therapy is required. In cases of allergic rhinitis, the use of antigen avoidance, nasal steroids, and antihistamine medication is well established. Similarly, treatment of rhinosinusitis with or without polyps includes the use of nasal steroids, oral steroids, antibiotics. The role of functional endoscopic sinus surgery and hyposensitisation are also evidence-based. Surgical options are also the mainstay of treatment in cases of conductive loss of smell due to nasal lesions or anatomical factors (e.g., septoplasty, turbinate surgery, the release of nasal adhesions).

Summary and Author's Comments

1. Anosmia is broadly classified as conductive and sensorineural and such differentiation is particularly important in clinical consultations to establish the initial work-up.
2. The importance of a detailed history cannot be overemphasized.
3. Nasal endoscopy alone (especially of the olfactory cleft) can sometimes accurately pinpoint the cause of conductive anosmia.
4. In terms of treatment, eliminating the causative factors usually leads to direct improvement of the olfactory function.

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Anosmia: Sensorineural

57

Sarantis Blioskas

57.1 Case Presentation

A 43-year-old female was referred by her GP with a three-month history of gradual declination of smell that led to complete anosmia. The patient did not report nasal blockage, nasal discharge, or facial headaches. She was generally fit and well and reported no medication intake, allergies or alcohol intake. She used to be an occasional smoker, yet she discontinued smoking more than 10 years ago. The patient denied any history of head trauma, toxin exposure or family history of anosmia or neurological conditions. Apart from anosmia and consequent taste disturbance, the patient also reported chronic fatigue and occasional vision abnormalities that she described as "blurred like sensation" as well as occasional loss of depth perception. She also vaguely reported a recent difficulty in remembering everyday events, yet no loss of long term memory. All head and neck clinical examination including cranial nerves examination was unremarkable. Anterior rhinoscopy and flexible nasendoscopy revealed a normal nasal cavity and postnasal space, clear olfactory clefts, no signs of rhinitis and only a minimal/clinically unimportant deviation of the nasal septum. In view of her medical history we decided to undertake imaging (MRI) of her brain and blood work to detect levels of Zinc, vitamin B12, renal and liver function. Laboratory tests indicated normal findings; however Brain MRI revealed evidence of neural demyelination, indicative of a potential neurodegenerative disease (multiple sclerosis). The patient was referred to the neurology department for further evaluation and treatment.

57.2 Background Knowledge

Attempts have been made to classify different forms of anosmia according to the anatomic topography of presumed pathology, thus differentiating between conductive anosmia (resulting from blockage of odourant transmission to the olfactory neuroepithelium), sensorineural anosmia (resulting from damage/loss of the olfactory neuroepithelium or nerve) and central anosmia (resulting from damage/loss of the olfactory processing pathways of the central nervous system).

Although, due to limitations in such a classification, modern classifications tend to rely more on specific underlying aetiology, yet this traditional approach remains valuable.

In terms of sensorineural anosmia, physiology suggests that odor transduction involves a cascade of events that occur at the olfactory epithelium and olfactory bulb, engaging receptors located in the cilia of olfactory neurons. Injury,

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aging, disease, or infection, can interfere with these neuronal processes and result in impairment of olfactory performance.

57.3 Clinical Approach

In terms of aetiology sensorineural anosmia can be significantly variable. Table 57.1 roughly summarizes different kinds of causes that can lead to sensorineural anosmia.

For cases of sensorineural anosmia, the diagnostic algorithm can generally be as follows.

- History

As in conductive anosmia, thorough history taking is essential. Yet particularly in cases where sensorineural anosmia is suspected, the presence of upper respiratory infections, particularly during the period before the onset of anosmia, should be directly inquired. History should also emphasize general neurological symptoms (e.g., tremor, motor symptoms, loss of memory, visual symptoms) as sensorineural anosmia could be the first symptom of neurological disease. Head trauma and drug intake (including chemotherapy) or toxin exposure (including questions on alcohol dependence or smoking) should also be sought. Family history may prove useful in cases where congenital anosmia is suspected, and in those cases, questions concerning other syndromic attributes should also be considered. In terms of onset, unlike anosmia attributed to sinonasal disease, post-infectious or posttraumatic anosmia is mostly characterised by sudden onset loss. However, in posttraumatic cases, there could be a gap of days or weeks between the trauma and recognition of the symptoms.

- Clinical examination

A thorough head and neck clinical examination including anterior rhinoscopy and flexible or rigid nasal endoscopy of the olfactory cleft is again a prerequisite. Nevertheless, in sensorineural anosmia, full cranial nerve and peripheral nervous system examination, including tests of memory and cognition, is of great importance.

Table 57.1 Causes of sensorineural anosmia

• Post-infectious	Upper respiratory tract infections Viruses (common cold, influenza) Bacteria Fungi Microfilaria HIV	
• Post-traumatic	Fracture cribriform plate	
• Associated with neurological disease	Epilepsy Myasthenia gravis Stroke Parkinson's disease Alzheimer's disease MS	
• Associated with exposure to drugs/toxins	Anaesthetics (local) Antimicrobials Antithyroid medications Chemotherapy Alpha-receptor antagonists	Acids Benzene Cadmium Chlorine Ethyl acetate Formaldehyde Hydrazine Hydrogen sulphide Lead Mercury Nitrous gases Paint solvents Silicon dioxide Trichloroethylene Zinc gluconate
• Congenital	Kallmann syndrome Turner's syndrome Bardet Biedl syndrome	
• Associated with aging		
• Other possible causes	Iatrogenic damage Tumours Multiple systemic co-morbidities Diabetes mellitus Hypertension Vitamin B12 deficiency Psychiatric conditions Migraine Radiotherapy Alcohol dependence Smoking	
• Idiopathic		

- Olfactory function evaluation

In general olfactory function, evaluation could be undertaken either subjectively, through patient-

reported assessment or objectively through psychophysical tests and/or electrophysiological studies and magnetic resonance imaging.

Subjective assessment can make use of visual analogue scales and Likert questionnaires, nevertheless it should be noted that self-assessment tends to be generally unreliable.

Psychophysical assessment uses several tests to determine threshold, discrimination and odor identification (see also Conductive anosmia chapter).

Electrophysiological studies include electroencephalography (EEG) and electro-olfactography (EOG), with the latter usually used exclusively in the research setting. Functional imaging is useful in identifying brain activity in response to odorous stimuli, either through positron emission tomography (PET) or functional magnetic resonance imaging (fMRI).

- Imaging

Unlike conductive anosmia where computed tomography scans of nose and paranasal sinuses are superior, in cases of sensorineural anosmia MRI scans of the brain are advantageous in investigating structures like the olfactory neuroepithelium, the olfactory bulb and higher pathways and detecting intracranial neoplasms, traumatic brain injury and signs of neurological diseases (e.g. degenerative conditions).

MRI scanning can also be used to calculate the olfactory bulb volume and olfactory sulcus depth, which can affected by multiple causes like exposure to toxins, congenital malformations and neurodegenerative diseases.

- Laboratory tests

Laboratory investigations to exclude hepatic, renal and endocrine disorders may be obtained. Deficits in vitamins (B12) or minerals may also be investigated.

- Olfactory epithelium biopsy

Biopsy of olfactory epithelium biopsy is generally not undertaken only in a research setting.

In terms of treatment, specific treatment is achievable only in a minority of cases. That applies particularly in cases of anosmia following viral infections and post-traumatic anosmia where no treatment has proved effective. In a clinical setting though treatment with nasal steroids irrespective of aetiology is common practice. Oral steroids also seem to be useful, although evidence is not definitive. There is also some contradicting evidence on the efficiency of mineral (Zinc) and vitamin (A, B12) supplements alone or in combination with steroids. Insufficient evidence also exists for phosphodiesterase inhibitors and intranasal calcium buffers.

Apart from medication, olfactory training therapy (repeated daily exposure of a subject to a range of odourants) has proven successful in different aetiologies of anosmia, although the specific mechanism remains unknown.

Finally, patient reassurance and education remain essential. In the same concept, warn for dealing with potential risks (smoke and natural gas detectors or date checking for spoiled food) should be emphasized.

Summary and Author's Comments

1. Attempts have been made to classify different forms of anosmia according to the anatomic topography of presumed pathology. Sensorineural anosmia results from damage/loss of the olfactory neuroepithelium or nerve.
2. In terms of aetiology sensorineural anosmia can be significantly variable, including post-infectious, post-traumatic and congenital causes, as well as aging, neurological disease and exposure to drugs/toxins.
3. Regarding treatment, specific treatment is achievable only in a minority of cases. Medication and olfactory training therapy can be useful; however, regardless of the efficiency of management, patient reassurance and education remain essential.

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Cacosmia

58

Sarantis Blioskas

58.1 Case Presentation

A 72-year-old male was referred to the ENT department complaining of a four-month history of constant cacosmia. The patient reported that after an episode of upper respiratory tract infection, which was treated with a short course of antibiotics, he started sensing an overwhelming smell of "rotten vegetables." His GP tried decongestants, nasal steroid sprays and oral antibiotics with no particular improvement. Apart from the annoying smell, the patient also reported occasional frontal headaches and a sensation that "phlegm is always stuck at the back of his nose" and he could not blow it out. He experienced a significant decline in his quality of life due to cacosmia since it has affected his appetite to the point that he started experiencing weight loss. On examination, anterior rhinoscopy and flexible nasendoscopy revealed significant purulent discharge on bilateral middle meatus and mucopurulent post nasal drip. The rest of the ENT examination was unremarkable. A CT of the patient's sinuses depicted evidence of chronic rhinosinusitis with bilateral blockage of osteomeatal complexes. Maximal medical treatment with oral antibiotics (Doxycycline 100 mg OD), tapering oral steroids and nasal steroids were administered for a period of 4 weeks, which significantly improved nasal blockage and diminished headaches, yet it failed to control the "persisting smell of rotten vegetables." On these grounds, we suggested Functional Endoscopic Sinus Surgery, with repeated irrigations of the diseased sinuses (Fig. 58.1). Following surgery, the patient was treated with long term nasal steroids and saline nasal douching. On his follow up 6 weeks following surgery the patient described that the "stinging odor was now gone."

58.2 Background Knowledge

Unlike anosmia, which is a quantitative dysfunction of the olfactory sense, cacosmia ('dysosmia') is usually a form of "parosmia," thus a qualitative dysfunction in the presence of an odour or better described as a distorted perception of an odour stimulus.

However, unlike the definition of the more general term "parosmia," "cacosmia" is generally referred as a negatively perceived olfactory distortion that usually occurs with a stimulus present (aka a form of parosmia) or absent (aka a form of "phantosmia").

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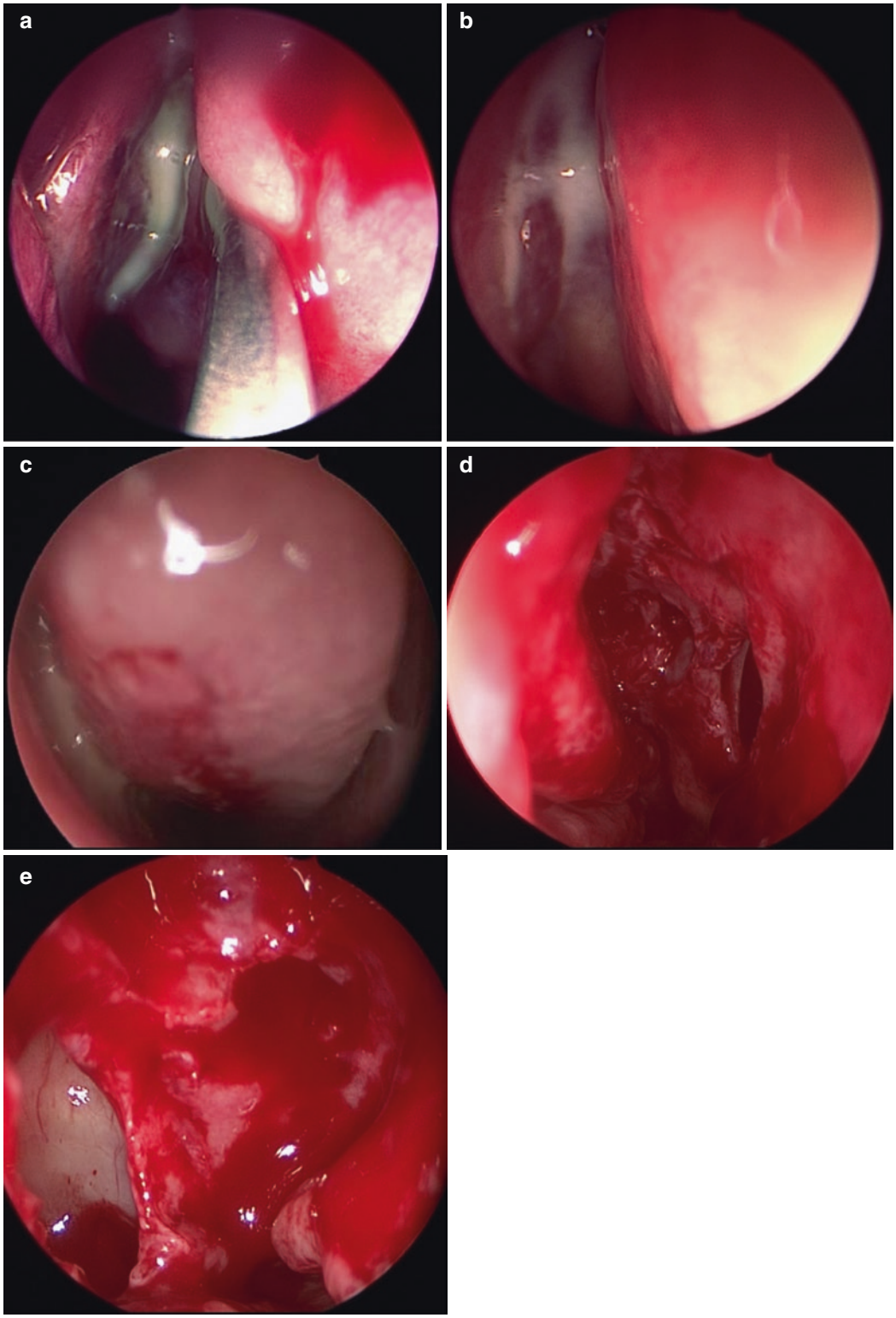

Fig. 58.1 (**a**–**e**) Intraoperative photographs of the case. Purulent discharge is notable in both middle meatus. Adequate clearance was achieved

58.3 Clinical Approach

In terms of aetiology, most of the causes already described for sensorineural anosmia, can also cause qualitative disorders like cacosmia. In particular, cacosmia has been described as a common occurrence after upper respiratory tract infections. In a more recent survey cacosmia was observed in 12.9% of patients with flu-like symptoms whose biopsies showed moderately impaired olfactory mucosa, yet not in those where olfactory mucosa was completely destroyed, suggesting that in these cases cacosmia can be attributed to impaired receptor cells that are still connected to higher olfactory centers.

In addition, cacosmia can also be described during the exacerbation of symptoms of sinonasal disease (chronic rhinosinusitis) because of purulent nasal discharge, especially in cases of fungal rhinosinusitis. The same applies to cases of atrophic rhinitis and rhinitis medicamentosa.

Neurodegenerative diseases (MS, Parkinsons) as well as malignancies of both the nose/paranasal sinuses and the central nervous system have been linked to symptoms of cacosmia. Finally, head trauma remains a common cause.

As far as the diagnostic approach is concerned, the diagnostic algorithm of quantitative olfactory dysfunction (including history, nasal endoscopy, CN examination, imaging, olfactory function tests) remains applicable and useful.

However, it should be stressed that qualitative dysfunction like cacosmia are usually solely based upon the patient's report and is very difficult to quantify by means of psyschophysical testing. Nevertheless, a comprehensive test, such as the "Sniffin' Sticks", could achieve an overall assessment of such a dysfunction. Apart from that, the impact of cacosmia can be graded by measuring (a) the frequency of occurrence: (daily occurrence giving 1 point, otherwise 0 points) (b) the intensity (very strong giving 1 point, otherwise 0 points) (c) the social effects (e.g., weight loss. Significant change of habits, yes giving 1 point and no 0 points). The sum score represents the degree of the disorder. It should be noted that cacosmia can be very debilitating and lead to decreased appetite, weight loss, and depression.

Imaging-wise, findings to support the diagnosis of qualitative olfactory disorders are small OB volumes compared with patients with unimpaired olfaction. There is no particular laboratory testing suggested for patients with cacosmia, although there are metabolic diseases that can occur through sensation of unpleasant odors such as trimethylaminuria or fish-odor syndrome.

Treatment of cacosmia and qualitative disorders, in general, can be challenging. Treatment options generally aim at reducing the overall functionality of the olfactory apparatus in order to reduce the unpleasant sensation. Hypertonic saline nasal drops, and nasal steroids can be useful. Furthermore, treatments like a nose clip, topical cocaine hydrochloride to induce anesthesia of olfactory epithelium, or even surgery to section olfactory fibers along the cribriform plate are options for extreme cases. The same applies to the use of gabapentin that acts centrally to suppress olfaction.

Summary and Author's Comments

1. "Cacosmia" is generally referred to as a negatively perceived olfactory distortion that usually occurs with a stimulus present (aka a form of parosmia) or absent (aka a form of "phantosmia").
2. Cacosmia has been described as a common occurrence after upper respiratory tract infections.
3. Qualitative dysfunction like cacosmia are usually solely based upon the patient's report and is very difficult to quantify by means of psyschophysical testing.
4. Treatment of cacosmia and qualitative disorders in general can be challenging.

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Phantosmia

59

Sarantis Blioskas

59.1 Case Presentation

A 54-year-old male was referred to the ENT outpatient clinic by his GP with a 4-month history of a reported "odor of roses" and a degree of loss of smell. In particular, the patient said that he experienced "loss of smell and taste," with occasional ability to smell. Yet, even then, "everything appeared to be smelling like roses," although there were no flowers there. The olfactory hallucination appeared on both nostrils equally and usually lasted for no more than 5 min each time. The patient reported no other nasal symptoms and only occasional headaches, which nevertheless when occurring were particularly severe and persisting, and were treated unsuccessfully with simple pain medication (Paracetamol and Ibuprofen). The patient was a light smoker and occasional alcohol drinker and he was under medication for high blood pressure (Ramipril). Thorough clinical history did not reveal evidence of nasal disease, previous head trauma, family history of olfactory disorders or neurological symptomatology. However, careful questioning indicated a significant time link between phantosmia and headaches, as hallucinations seemed always to precede attacks of pain in the form of an "aura." Additionally, when asked, the patient could remember sensitivity to light and/or sound when experiencing these "throbbing headaches." Clinical head and neck examination was unremarkable and cranial nerve examination did not reveal any deficits. Flexible nasendoscopy depicted evidence of mildly rhinitic mucosa, consistent with smoking, with no further findings. As history and clinical examination suggested that the patient suffered from what appeared to resemble migraine attacks, it was decided to be referred to a neurology clinic for advice. The patient was put on medication for migraines (triptan) and on his follow up 3months after the initial consultation, he reported never to have experienced any "smell of roses" since being started on medication.

59.2 Background Knowledge

Phantosmia is a qualitative dysfunction in the absence of an odour (i.e. an odour is perceived without concurrent stimulus, an 'olfactory hallucination'). The actual prevalence of phantosmia is unknown since objective assessment is challenging, yet phantosmia occurs in almost 25% of patients reporting subjective olfactory dysfunction and seeking medical attention in specialized olfactory centers.

Phantosmia is usually part of more general complex olfactory issues, combining both quantitative and qualitative dysfunctions, thus it is more

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often seen in patients who also suffer a degree of loss of olfactory ability. Despite that, phantosmia can also occur in people with an otherwise normal sense of smell.

59.3 Clinical Approach

The cause of phantosmia is not fully clarified, yet it may be attributed to a number of etiologies. Such etiologies include chronic rhinosinusitis, neurologic diseases such as epilepsy or neurodegenerative disorders, spontaneous or post-traumatic intracranial hemorrhage and psychiatric conditions.

A clinically useful differentiation regarding phantosmia aetiology is presented in the following table (Table 59.1).

As far as the diagnostic approach is concerned, obtaining a clinical history remains essential and should among all include a history of seizure disorder, migraine symptoms, memory loss, psychiatric/neurologic disorders, cognitive dysfunction or recent acute or chronic rhinosinusitis. As mentioned, alleviation of symptoms with unilateral occlusion usually points towards peripheral phantosmia and should be investigated through history taking. Much like cacosmia, quantification of symptoms in cases of phantosmia can be challenging. It should rely on evaluating the frequency of occurrence, the intensity and the social effects (e.g., weight loss, significant change of habits).

Table 59.1 Types of phantosmia

Types of phantosmia	Pathophysiology	Characteristics
Peripheral phantosmia	Dysfunction at the level of the olfactory receptors and neurons, leading to a distorted representation of stimulus	• Intermittent, usually caused by odorant stimuli, and often localized on one side • Relieved by nasal obstruction or topical cocaine (causing anosmia)
Central phantosmia	Dysfunction of the cortical olfactory pathways	• Constant, unremitting and bilateral • Remains unchanged

Through physical examination patient's general demeanor and psychiatric health should be assessed. A standard examination of the olfactory cleft through nasal endoscopy, and detailed cranial nerve examination as well as evidence of motor and sensory impairment is also important. A dental examination should be included for the oral diseases that may produce a foul odor.

Diagnostic work-up should include olfactory testing to determine the extent of olfactory dysfunction. At the same time, magnetic resonance imaging should be the modality of choice to rule out neoplasm, cerebrovascular or sinus disease.

Management of phantosmia can be divided into medical treatment and surgical modalities, and the choice is usually dictated by the underlying cause, as established by the diagnostic workup. Thus, in cases of peripheral phantosmia, medical therapy may prove ineffective, and surgical intervention should be suggested to patients that have failed multiple trials of medication.

Medical therapy usually targets the specific etiology of phantosmia. Thus, phantosmia associated with epilepsy may be treated with antiepileptic medication, whereas migraine-associated phantosmia can be treated with topiramate. Steroid therapy in patients with rhinosinusitis, lamotrigine and treatment with gabapentin or aripiprazole (antidepressant therapy) for patients with underlying psychiatric illness are also included in potential medical therapeutic options. The use of topical cocaine HCl for anesthetizing olfactory neurons has been proposed in the past; however, undesired effects can occur. Finally, the role of olfactory training in cases of phantosmia remains controversial.

Surgical modalities include resection of the olfactory bulb through neurosurgical approaches using a bifrontal craniotomy as well as endoscopic intranasal approaches of excision of the epithelium of the olfactory cleft. Such procedures include either the risks and morbidity associated with a craniotomy or the potential complications of endoscopic anterior skull base surgery, including CSF leak and meningitis. Given the risks associated, no consensus exists regarding optimal management.

Summary and Author's Comments

1. Phantosmia is a qualitative dysfunction in the absence of an odour (i.e. an odour is perceived without concurrent stimulus, an 'olfactory hallucination')
2. Phantosmia etiology includes chronic rhinosinusitis, neurologic diseases such as epilepsy or neurodegenerative disorders, spontaneous or post-traumatic intracranial hemorrhage and psychiatric conditions
3. A clinically useful differentiation regarding phantosmia aetiology divides peripheral and central causes of phantosmia.
4. Management of phantosmia can be divided into medical treatment and surgical modalities and choice is usually dictated by the underlying cause

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Taste Disorders

60

Sarantis Blioskas

60.1 Case Presentation

A 42-year-old female was seen at the ENT outpatient clinic on her first follow up after a combination of chemotherapy and radiotherapy treatment for a T3N0M0 SCC of the palate. She had her last radiotherapy treatment completed about 1 month ago, and following that, she reported a constant "metallic taste" and a general disordered sense of taste. Quoting the patient's description, "everything tasted flavorless, indifferent, flat, and slightly metallic and only when excessive salt was added, there was a very light pinch of flavor." Although taste impairment reduced her appetite, the patient reported only minimal weight loss. History taking included detailed questions on potential olfactory loss, yet the patient denied any symptoms from her perspective. Clinical examination revealed evidence of dried oral mucosa (xerostomia) with no presence of ulcers or mucositis; however, a slightly erythematous appearance of the tongue surface was noted. Bimanual examination of salivary glands revealed moderately decrease saliva flow. Dental hygiene was satisfactory, and the rest of the head and neck examination did not show remarkable findings. The patient was reassured as she was communicated that usually 6 months to 1 year after radiotherapy, taste sensation recovers to its previous level. Self-coping strategies were suggested including increasing water intake, frequent teeth brushing, mouth rinse before meals with salt-water or ginger ale, avoid smoking, and selecting foods with strong flavors including lemon, spices or pickled foods. As taste changes can impact the enjoyment of eating or drinking and potentially lead to malnutrition, small, frequent meals and snacks, attractive presentation of foods and preferring enjoyable high calories strong-flavored puddings, sauces or gravies were also encouraged. The patient was also referred to a cancer center dietitian for help with food selections. On her following follow-up appointments, the patient reported satisfactory gradual improvement.

60.2 Background Knowledge

Taste is arguably the most significant external sensory system. People with a reduced or diminished sense of taste, like head and neck cancer patients suffering from a radiation-induced loss of taste, rapidly become nutritionally compromised and usually require a chronic nasogastric tube for feeding.

In general, qualities regarding the sense of taste can be divided into sweet, sour, salty, bitter and savory. Taste is then combined with olfaction and the intrinsic odors of different foods create their unique identification termed "flavor." Thus,

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contrary to popular belief, taste provides little information regarding the identity of a food. Instead, the "taste" of specific foods is accomplished via the olfactory and visual modalities and maintained by retronasal olfaction.

In terms of basic anatomy, taste receptors are located in several fields within the oral cavity, including various areas of the tongue, the soft palate, and regions of the pharynx and even laryngeal areas. These receptors predominantly reside within multicellular rosebud'-shaped structured clusters labeled 'taste buds,' which at the tongue occur in papillae. Thus, there are three types of taste papillae: fungiform papillae scattered in the anterior 2/3 of the tongue, vallate papillae at the middle of the posterior 1/3 and foliate papillae at the sides of the posterior 1/3 of the tongue.

The receptor cells in taste buds come in direct contact with oral solutions via microvilli at their apical ends. The afferent taste neural information is transmitted to fibers inside the taste buds and through them to ganglia of cranial nerves (CN) VII, IX, and X. Subsequently, neural stimulus projects onto the nucleus of the solitary tract located at the dorsal medulla at the brainstem and finally via the thalamus to the primary opercular and insular taste cortex, the orbitofrontal cortex, the cingulate gyrus and the amygdala.

60.3 Clinical Approach

It needs to be stated that true deficits in the sense of taste are only a minority out of the patients that seek medical attention because of sensory problems, with olfactory problems being the majority. When true taste issues occur they most frequently involve taste losses, although taste phantoms do also occur.

Taste disorders can be roughly classified into

1. **Quantitative taste disorders**: which include ageusia (complete loss of taste) or hypo−/hypergeusia (partial loss of taste/increased taste capacity),
2. **Qualitative taste disorders**: which include parageusia and phantogeusia (triggered and permanent taste distortion accordingly) and
3. **Other kinds of taste disorders**: that usually refer to taste agnosia (difficulty identifying taste stimulus despite effectively perceived).

Nevertheless, it should be noted that the kind of taste disorder does not necessarily correspond to the underlying cause as well. Thus, taste disorders can be attributed to a different group of causes, which most of them can result in the same disorder (e.g., hypogeusia). Potential causes of taste disorders are the following (Table 60.1):

In the general population, isolated taste disturbances are rare, thus taste testing in clinical practice is mainly restricted to specialized departments. Diagnosis can be facilitated with the following tools:

(i) Patient History. Although the patient's history can be the key to understanding the disorder, it should be stressed that in case of taste disorders, it can be unreliable as patients can rate their taste function inaccurately, or even not notice taste deficiency at all. Thus, history taking should emphasize specific questions about the production of saliva, swallowing issues, history of head and neck surgery (including the middle ear), trauma, and, most importantly, drug intake and potential comorbidities (e.g., diabetes, neurological conditions). Questions about olfactory loss should also be included since combined olfactory and gustatory impairment is common.

(ii) Taste testing. Testing of the sense of taste includes the use of chemicals applied orally. Different tests exist and they can be generally divided into whole mouth tests (assessing overall taste) and regional tests (assessing specific areas independently). The former may include liquids (the three-drop method) or alternatively tasting tablets or wafers, whereas the latter usually uses taste strips, disc filter paper or liquid tastants.

(iii) Electrogustometry (EGM). First introduced in 1958, EGM involves the application of weak anodal electric currents to specific regions of the mouth and measure electric taste thresholds. Advantages include avoiding the use of chemical solutions, measuring

Table 60.1 Causes of taste disorders

Cause	Characteristics
Age	Taste generally decreases with age, although the loss of taste is usually much less pronounced than the corresponding loss of olfaction. Aging leads to raising of taste detection threshold for standard stimuli, however much smaller decline exists for suprathreshold stimuli.
Postoperative/ posttraumatic disorders	Mainly qualitative (taste distortion) changes and they occur after either head injury or ENT surgery. Quantitative taste deficits (ageusia, hypogeusia) often go unnoticed thus, their incidence is largely unknown. Operative procedures that can be potentially complicated with postoperative taste disorders include middle ear surgery (via injury to chorda tympani), tonsillectomy and oropharyngeal procedures in general (probably due to disturbance to the lingual branch of the IX nerve), microlaryngoscopy, tracheal intubation and generally procedures that include lingual compression, and finally dental procedures. In the same concept lay the cases of head and neck cancer patients undergoing oncologic surgery with or without radiation therapy. Regarding trauma, disorders can be due to head injury and less frequently caustic ingestion.
Neurological	*Peripheral* causes include syndromes affecting the facial and more rarely the glossopharyngeal nerve, like Bell's palsy, neuroborreliosis, herpes zoster, polyneuropathy, and neoplastic processes affecting the submandibular region or the skull base. *Central* causes include cerebrovascular disorders, CNS tumors, epilepsy, Alzheimer's, multiple sclerosis and neurodegenerative diseases in general, however it is extremely rare in these cases to have isolated taste disorders alone. There are neurological causes with unclear localization, like familiar dysautonomia, hereditary ataxia, Machado-Joseph disease, genetically determined global thermoanalgesia, Guillain-Barre syndrome and Creutzfeldt-Jakob disease.
Drugs and toxic substances	Antimicrobial drugs (antifungal like amphotericin B, antibiotics like penicillin, metronidazole and tetracycline), anti-inflammatory (diclofenac), antihypertensive (ACE inhibitors like captopril, Ca^{++} channel blockers like amlodipine, nifedipine, diuretics like amiloride), antihyperlipidemics (statins), neurologic medication (carbamazepine, levodopa, amitriptyline), antineoplastic drugs (methotrexate) and many more. Other toxic substances, including industrial compounds, tobacco, and alcohol, may also adversely affect the taste.
Chronic conditions	Liver failure from various causes, (cirrhosis, hepatitis, sclerosing cholangitis), nutritional status (zinc, vitamins), uremia due to chronic renal failure, endocrine disorders including diabetes and thyroid disease, and autoimmune diseases (Sjogren's syndrome, amyloidosis)
Postviral/ idiopathic taste disorders	Idiopathic taste disorders are the most commonly diagnosed causes of dysgeusia and they either follow an upper respiratory tract infection or are of entirely unknown etiology, reflecting the poor clinical knowledge of such disorders even today.

side differences in patients with taste disorders (for example assessing chorda tympani injury) and assessing both the gustatory and the trigeminal system. Electric taste thresholds also show high test-retest reliability. Disadvantages include the fact that electrogustometry typically evokes sour or salty taste, whereas many taste disorders particularly affect the bitter taste; thus, electrogustometry may fail to identify clinically relevant damage. Furthermore, electric taste thresholds correlate well with regional but not whole-mouth chemical taste thresholds thus, electrogustometry may be unable to reflect real-world taste experience accurately.

(iv) Magnetic Resonance Imaging. The neural outcomes of taste perception may be visualized functional magnetic resonance imaging (fMRI). Yet, such modalities are not routinely used in clinical practice.

Treatment of taste disorders focuses mainly on the treatment of the underlying disease. This includes both the management of local causes and/or neurological disease, and the thorough revision of any medication received. Self-coping strategies like oral hygiene, increasing water intake, avoiding strongly smelling or tasting meals and eating small meals more frequently can also prove useful. Specific therapeutic options are limited and mainly consists of zinc

and a combination of systemic steroids and vitamin A. However, existing evidence is neither convincing nor equivocal. Nevertheless, zinc replacement therapy remains the treatment of choice, particularly in cases of idiopathic taste disorders (usually 140 mg/day for 4 months). Finally, it should be noted that in many cases a spontaneous improvement is possible.

Summary and Author's Comments

1. Taste is arguably the most significant external sensory system, yet true deficits in the sense of taste are only a minority out of the patients that seek medical attention
2. Taste disorders can be roughly classified into quantitative disorders, qualitative taste disorders and miscellaneous
3. Electrogustometry (EGM) was first introduced in 1958 and involves the application of weak anodal electric currents to specific regions of the mouth and the measurement of electric taste thresholds
4. Treatment of taste disorders focuses mainly on the treatment of the underlying disease

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Part IX

Systematic Diseases Affecting the Nose and the Paranasal Sinuses

Granulomatosis with Polyangitis (GPA)

61

Mohamed Morsy and Marios Stavrakas

61.1 Case Presentation

A 43-year-old lady presenting with longstanding bilateral nasal obstruction, anosmia, bloody crusts and blood-stained nasal discharge. She also had bilateral hearing loss. She tried some topical steroid nasal sprays and saline nasal douches before without much improvement. The patient also gave a history of cough, haemoptysis and dyspnoea, particularly on exertion, as well as haematuria. External nose examination has shown a saddle nose deformity. On examination by anterior rhinoscopy, the nasal mucosa shows multiple areas of ulceration and granulations. On endoscopic examination, the nasal cavity was filled with multiple crusts, and some adhesions between the septum and lateral nasal wall could be seen. Both tympanic membranes were markedly retracted, showing a clinical picture of otitis media with effusion. Endoscopic examination of the larynx has shown subglottic stenosis. C- ANCA was positive. Kidney function tests were elevated. CT scan of nose and sinuses has shown non-specific mucosal thickening and evidence of bone destruction. Renal biopsy has shown granulomatous inflammation with vasculitis and necrosis.

61.2 Background Knowledge

Granulomatosis with polyangiitis (GPA), previously known as Wegener's granulomatosis, is a systemic autoimmune disease of unknown aetiology. There is evidence that GPA is an autoimmune disease in which Anti Neutrophil Cytoplasmic Antibody (ANCA) plays a role, causing tissue damage by stimulating degranulation of toxic oxygen radicals from leucocytes. Other theories have supported that ANCA may activate neutrophils, leading to inflammation or that circulating complexes of ANCA and neutrophil degranulation products provoke a Type 3 hypersensitivity reaction. Finally, evidence is growing for a genetic association of GPA, especially familial association, mostly in relation to major histocompatibility complex (MHC). Genetic associations correlate best with ANCA specificity. The Genome-Wide Association Study in GPA demonstrated an association of PR3-ANCA with MHC, class II, DP (*HLA-DP*), serpin peptidase inhibitor class A (*SERPINA1*), and proteinase 3 (*PRTN3*), whereas MPO-ANCA was associated with MHC, class II, DQ (*HLA-DQ*). Histologically, GPA is characterized by granulomatous inflammation of the upper and

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lower respiratory tract with necrotizing vasculitis of small and medium-sized blood vessels and focal or proliferative glomerulonephritis. Classically, it affects the nose, lungs and kidneys. It can occur in a localized form affecting only the nose.

61.3 Clinical Approach

61.3.1 History

Most patients present with nasal symptoms. Rhinologic presentations occur in up to 90% of patients, including nasal obstruction, crusting, bloody nasal discharge, epistaxis, hyposmia or anosmia, pain, epiphora, or change in the shape of the nose. Otological symptoms are not uncommon, primarily conductive (< Otitis Media with Effusion) or even sensorineural hearing loss. Subglottic stenosis can occur in one-fifth of these patients. Lower respiratory tract symptoms such as cough, haemoptysis and dyspnoea, and renal symptoms such as haematuria can also occur. It is interesting to mention that nasal symptoms vary from 55% to 90%, 40–50% of patients have chronic sinusitis, and acute bacterial or fungal sinusitis is not uncommon. With regards to symptoms from various systems at presentation, the figures are the following: Head & Neck (H&N) 73%, lower respiratory tract 48%, and kidneys 20%. In the H&N, up to 80% have sinonasal and around 50% have tracheal involvement. The diagnosis is based on clinical criteria, and other cANCA vasculitides such as Churg Strauss disease should be excluded.

61.3.2 Examination

Examination reveals friable nasal mucosa with ulceration and granulations, often with bloody crusts and adhesions. There may be a septal perforation, and in advanced cases there may be loss of normal internal nasal architecture with a single large cavity. The saddle nose deformity can also occur due to loss of dorsal support. It is essential for the ENT surgeon to distinguish disease activity from damage, as this plays an important role in the planning of medical treatment and adjusting the doses of immunosuppressants. Birmingham Vasculitis Activity Score (BVAS) is a tool that scores five items: bloody nasal discharge/crusts/ulcers/granulomata, paranasal sinus involvement, subglottic stenosis, conductive hearing loss and sensorineural hearing loss. There are other modified tools for the same purpose, such as the ENT/GPA Disease Activity Score (ENT/GPA DAS) and also scoring tools for the damage (Vasculitis Damage Index (VDI).

61.3.3 Investigations

Cytoplasmic ANCA (c-ANCA) specific for proteinase3 (PR3) is 90% sensitive and 98% specific for generalized GPA; the sensitivity falls to 50% in localized disease.10% of patients with GPA will have a positive test for (p-ANCA) against myeloperoxidase (MPO), and up to 30% may have a negative ANCA initially. This result may change over time and could come back positive if repeated after a few months; therefore, cANCA may be used to monitor disease activity. The biopsy of the nasal mucosa is not always diagnostic. Lung and renal biopsies have a higher yield. Histological diagnosis of GPA shows the presence of granulomatous inflammation, vasculitis and necrosis. CT scan of the sinuses may show nonspecific findings and may be normal in the first stages of the disease. Pathological characteristics on the scan include septal erosion, mucosal thickening, and bony changes (Fig. 61.1).

61.3.4 Medical Treatment

Treatment should be multidisciplinary, with involvement of ENT, rheumatology, renal and respiratory teams. Involvement of the rheuma-

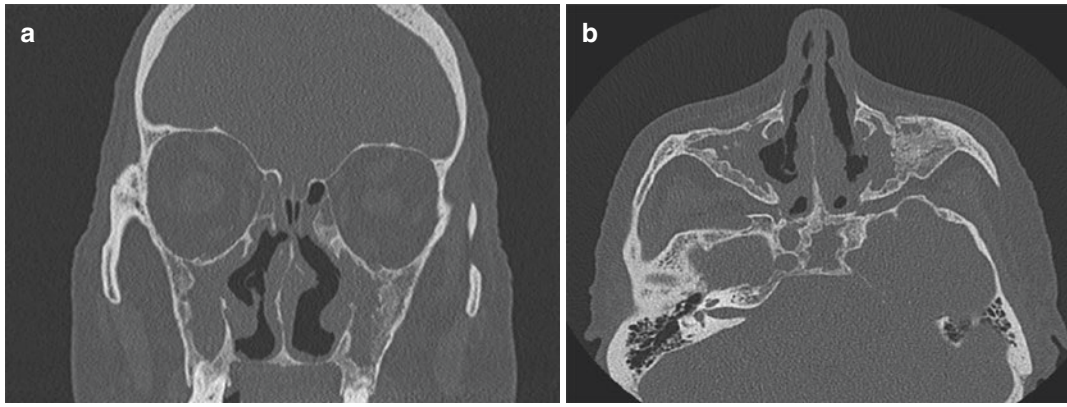

Fig. 61.1 (**a**, **b**) A patient with GPA, who underwent bilateral middle meatal antrostomies in the past. The maxillary sinuses remain entirely filled with inflammatory soft tissue and there is marked sclerosis of the sinus walls. The middle turbinates have been eroded

tology team is essential in the medical treatment and long-term follow up of the disease activity of patients with GPA. Being an autoimmune disease, it is treated by corticosteroids and immunosuppressive medications. The introduction of cyclophosphamide has further improved remission rates to 90%, and steroid-sparing agents such as azathioprine, methotrexate and mycophenolate mofetil are then used to maintain remission. Monoclonal antibodies such as rituximab and infliximab are also being used effectively.

Optimising the nasal condition by saline nasal irrigation and topical steroid sprays provides symptomatic relief for patients with nasal symptoms.

Hearing aids are the treatment of choice for hearing loss in these patients, as grommet insertion for otitis media with effusion in GPA would result in chronic otorrhea.

61.3.5 Indications for Surgery

- For reconstruction of nasal dorsum in saddle nose deformity. Reconstruction in patients with GPA appears to be safe and effective if the disease is in remission before reconstruction is started (in our practice-at least 1 year in remission). L-shaped rib cartilage graft is a good option. Because revision rates for patients with GPA undergoing external nasal reconstruction seem to be higher than with normal septorhinoplasty, preoperative counseling about this fact is warranted.
- For subglottic stenosis, upper airway endoscopy and regular balloon dilatation, intralesional injection of triamcinolone and/or endolaryngeal laser treatment.
- Endoscopic Sinus surgery should be better avoided as adhesions are likely to make the outcome worse; similarly, surgical repair of septal perforation is unlikely to be successful. Therefore, surgery is generally not advised, except for carefully selected cases.

61.3.6 Follow Up

Regular follow up is required to monitor the disease activity and change the medications or modify the dosage accordingly. Nowadays, joint ENT-Rheumatology clinics are gaining popularity as they achieve fewer hospital visits, early recognition of relapse and disease activity and consequently achieve better disease control.

Summary and Author's Comments

1. GPA is an autoimmune disease characterized by granulomatous inflammation of upper and lower respiratory tracts, necrotizing vasculitis of small and medium blood vessels and glomerulonephritis.
2. Nasal manifestations are the most frequent presentations, including crusts, adhesions, granulations, septal perforation and saddle nose deformity.
3. C-ANCA is a specific test, but it could be negative in the early stages of the disease.
4. Steroids and steroid-sparing drugs are the mainstays of treatment. Multidisciplinary team approach and involvement of rheumatology are important.
5. Surgery should be generally avoided, with rare exceptions-carefully selected cases.

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62 Eosinophilic Granulomatosis with Polyangiitis (EGPA or Churg-Strauss Syndrome)

Mohamed Morsy and Marios Stavrakas

62.1 Case Presentation

A 52-year-old female patient presenting with worsening nasal symptoms including bilateral nasal obstruction, blood-stained nasal discharge & anosmia. She was diagnosed with allergic rhinitis. She gave a history of late-onset asthma, which she developed about 15 years ago. More recently, she started presenting cardiomyopathy and peripheral neuropathy. On examination, nasal polyps could be seen.

62.2 Background Knowledge

Eosinophilic granulomatosis with polyangitis is a systemic autoimmune condition. It is characterized by a triad of asthma, systemic vasculitis affecting small-to-medium-sized blood vessels, and eosinophilia (peripherally and in mucosal lesions). The asthma is characteristically late-onset and, together with allergic rhinitis and nasal polyps, may precede the vasculitis by several years. Common nasal presentations include nasal obstruction, anosmia and rhinorrhea, which are the most common sinonasal symptoms, occurring in up to 95% of patients. Less common nasal symptoms include sneezing, crusting & epistaxis. Nasal polyps are a common finding on examination (nearly 60% of patients with EGPA also have a degree of nasal polyposis at presentation). Destructive lesions may be seen on examination, but these are not as common as in GPA.

The disease has 3 phases which may overlap:

- prodromal phase (allergic stage): in which nasal & respiratory symptoms last for several years. Almost all patients experience asthma and/or allergic rhinitis.
- eosinophilic stage: peripheral eosinophilia >10%, both in the blood and tissues
- systemic vasculitis: affecting the heart, peripheral nervous system, the gastrointestinal tract and the kidney.

62.3 Clinical Approach

Almost all patients present initially with asthma and/or allergic rhinitis. Nasal obstruction, anosmia and rhinorrhea are the commonest nasal symptoms, but it can also present with crusts and epistaxis.

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Lanham's criteria can be used to aid diagnosis (all should be present)

1. Asthma
2. Peak eosinophilia >1.5 x 10^9 cells/L
3. Systemic vasculitis, two or > extrapulmonary sites

The American College of Rheumatology 1990 criteria for the diagnosis of EGPA (Churg-Strauss) require at least **four** of the following:

1. asthma;
2. eosinophilia of greater than 10% differential;
3. mono- or polyneuropathy due to vasculitis;
4. nonfixed pulmonary infiltrates;
5. abnormalities of the paranasal sinuses;
6. and extravascular eosinophils on biopsy.

This gives a sensitivity of 85% and a specificity of 99.7%.

Examination
Diagnostic nasal endoscopy commonly shows nasal polyps.

Investigations
Anti-neutrophil cytoplasmic antibodies (ANCAs) are strongly associated with three vasculitides: GPA, eosinophilic granulomatosis with polyangiitis (Churg-Strauss syndrome) and microscopic polyangiitis (MPA).

EGPA is another ANCA-associated vasculitis, but pANCA (against MPO) is only positive in 31–50% of cases. Histological diagnosis requires the presence of extravascular eosinophilic granulomas and necrotizing vasculitis, but a nasal biopsy is generally of low diagnostic certainty. Skin, nerve, muscle or lung tissue has a higher positive yield. There is a pan-opacification of the paranasal sinuses on CT scan, due to chronic rhinosinusitis (CRS) with nasal polyps (Fig. 62.1).

62.3.1 Medical Treatment

Again, this should be multidisciplinary, involving ENT, rheumatology, renal, cardiology & respiratory physicians. Immunosuppression is the mainstay treatment, with corticosteroids and steroid-sparing agents (such as azathioprine and cyclophosphamide), but rituximab and interferon alpha are also being used. Topical steroid nasal sprays and saline nasal douches are used to control nasal symptoms.

62.3.2 Surgical Treatment

Endoscopic sinus surgery could be done to remove nasal polyps and also in the case of

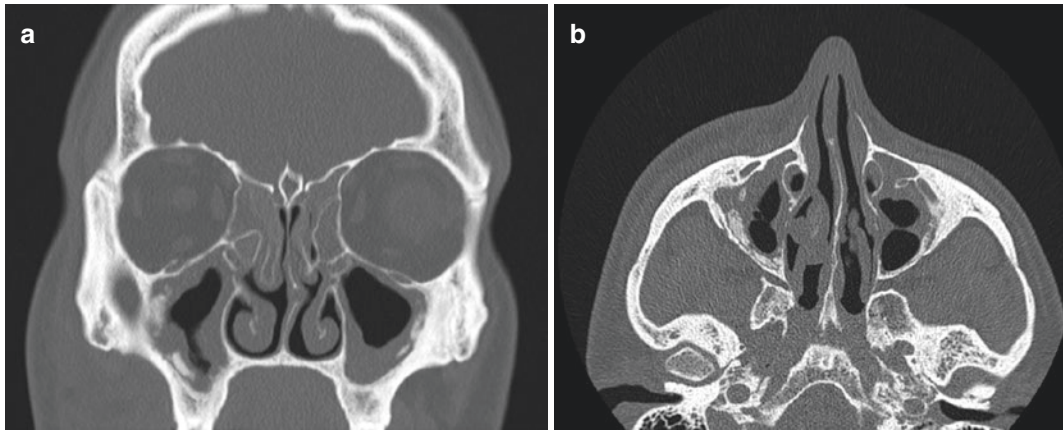

Fig. 62.1 (**a**, **b**) EGPA. Extensive polyposis throughout the ethmoid air cells with secondary obstruction of the frontal sphenoid and maxillary antra. Within the maxillary antra the surrounding bone is thickened with areas of calcification within the surrounding soft tissue material, which indicates chronicity of the changes

mucocele formation. Careful selection of the patients and assessment of indications for surgery is important.

Summary and Author's Comments

1. Eosinophilic granulomatosis with polyangitis is one of the ANCA-associated vasculitides, and should be suspected if a patient is presenting with asthma and nasal polyps and manifestations of systemic vasculitis.
2. Cardiac involvement is the most frequent cause of death in EGPA.
3. Treatment should be multidisciplinary, with steroids and steroid-sparing drugs being the mainstay of treatment.
4. Topical steroid sprays and saline nasal douches are used to control nasal symptoms, with endoscopic sinus surgery being reserved for polyp removal and marsupialization of mucoceles.

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Relapsing Polychondritis

63

Konstantinos Geronatsios

63.1 Case Presentation

A 57-year-old male patient was referred to our clinic with a three-month history of worsening nasal congestion, nasal discharge, midfacial pain mainly involving the nasion, minor sporadic nose bleeding, low-grade fever, weight loss and fatigue. He was treated at first with a 12-day course of Amoxicillin—Clavulanic acid combined with steroid nasal spray with no improvement, and after a month with a 14-day course of Cefuroxime, steroid nasal spray and a 12-day course of oral steroids (Methylprednisolone), with a slight improvement of his nasal symptoms. A CT sinus scan was also performed, which showed mucosal thickening of the sinuses and the nasal cavity. We performed a thorough examination, following a detailed history taking, which was otherwise unremarkable. He only mentioned an episode of right knee swelling 6 months ago, which settled completely. Nasal endoscopy using a 0° rigid nasal endoscope revealed swollen and friable nasal mucosa, mild crusting and nasal discharge (Figs. 63.1 and 63.2). We also noticed an acute progression of saddle nose deformity. We ordered blood tests that demonstrated elevated WBC, CRP, ESR, and Immunoglobulin G, mild anemia, and positive ANA, ASMA. The patient was admitted to the hospital for further investigation. During his stay in the hospital, we performed a nasal mucosa biopsy which was not diagnostic. On the fourth day of his hospital stay, the patient started feeling severe pain and swelling on his right ear, which evolved to perichondritis. Two days after, he experienced the signs and symptoms of perichondritis on the left ear. We performed an audiogram, which revealed a bilateral middle-grade sensorineural hearing loss (Fig. 63.3). The patient fulfilled three of six Mc Adam criteria for relapsing polychondritis. He was referred to a rheumatology department for further evaluation and treatment.

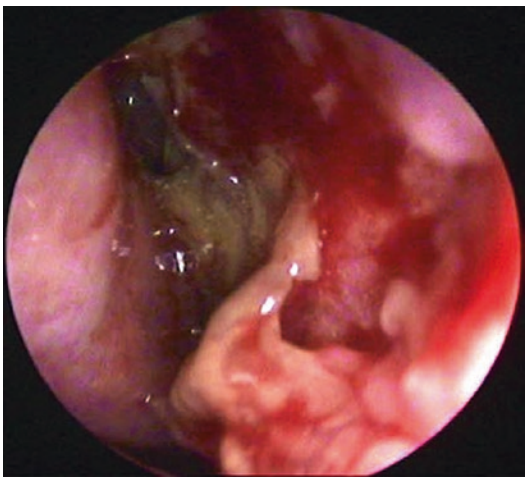

Fig. 63.1 Right nasal cavity

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63.2 Background Knowledge

Relapsing polychondritis (RP) is a severe, rare, progressive, multisystemic autoimmune disease. RP involves cartilaginous structures and mainly the ears (90%), nose (20–60%) and laryngo-tracheobronchial tree (50%). It can also affect the middle and inner ear (cochlear and vestibular damage), eyes (20–60%), peripheral joints (33–70%), cardiovascular system (10–25%), central nervous system (3%) skin—oral mucosa (36%) and renal system (22%).

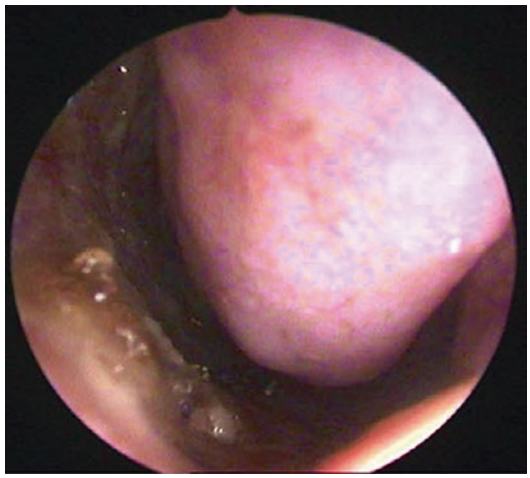

Fig. 63.2 Left nasal cavity

RP's etiology is still unknown. HLA-DR4 is considered to be related to the disease. Also, cartilage-specific circulating autoantibodies, such as antibodies against collagen II, IX, and XI, may be detected. Antibodies to protein Matrilin-1 can also be found in patients with respiratory symptoms due to RP.

Biopsy and pathologic features of an active inflammatory cartilage lesion, depend on the stage of the disease. Mixed inflammatory infiltration with lymphocytes, neutrophils, plasma cells, eosinophils extending into cartilage can be noticed. Also, as the disease progresses, granulation tissue and fibrosis are possible pathologic features, sometimes with granular deposition of IgG and C3.

63.3 Clinical Approach

63.3.1 Diagnosis

RP can affect several organs, and it's heterogeneity and episodic symptoms can cause a significant delay in diagnosis. If left untreated, it can be life-threatening. It is not uncommon that the diagnosis may be delayed even for years from the onset of symptoms. The median age of onset is between the fourth and the fifth decade of life.

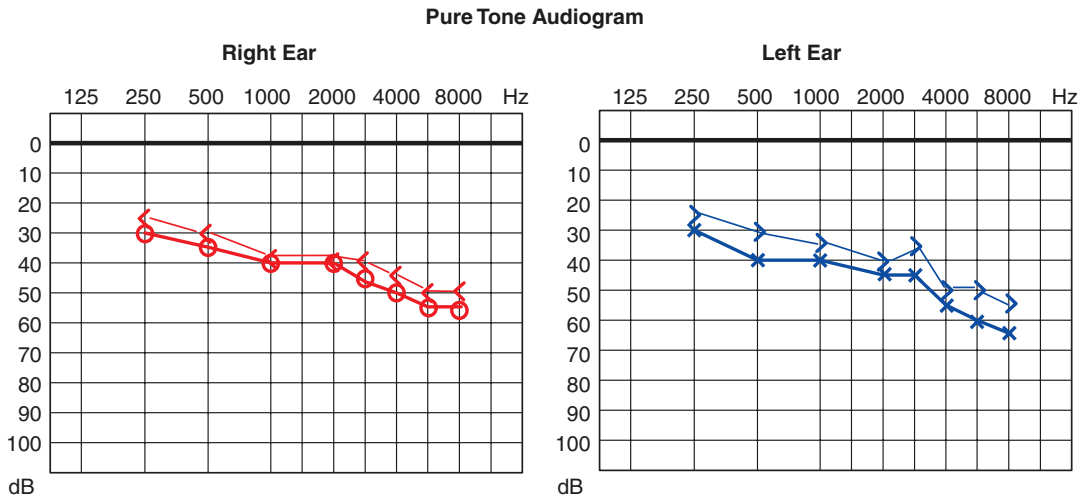

Fig. 63.3 Pure tone audiogram of the patient

A thorough history and physical examination are essential. The gradual onset or the episodic nature of symptoms is crucial for the clinician. There is no specific test for RP. ESR and CRP may be elevated, while anemia and leukocytosis are possible laboratory findings. ANA, ANCA, RF may also be positive. RP can be associated with other autoimmune diseases, such as Rheumatoid Arthritis, Systemic Lupus Erythematosus, Ankylosing Spondylitis, Granulomatosis with polyangiitis, Churg–Strauss syndrome, and Myelodysplasia. The main symptoms depend on the affected systems, and are the following:

- General symptoms: fever, fatigue, weight loss, oral lesions, skin rash.
- Auricular symptoms: pain, perichondritis, swelling and tenderness usually of both ears, external auditory canal obstruction.
- Respiratory: dyspnea, respiratory distress, wheezing, respiratory infections, stridor, choking, tracheobronchomalacia.
- Nasal symptoms: nasal blockage, difficulty breathing, epistaxis, swollen nose, saddle nose deformity, nasal chondritis, pain at the base of the nose, rhinorrhea, septal perforation.
- Ocular: scleritis, episcleritis, conjunctivitis, iritis, retinopathy, optic neuritis.
- Cardiovascular: aortic valvular disease, pericarditis, vasculitis, aneurysms, abnormal heart rhythm, myocardial infarction, descending thoracic aortic aneurysm.
- Musculoskeletal system: arthritis, arthralgia, myalgia.
- Audiovestibular system: sensorineural hearing loss, vertigo, otitis media, stapes fixation—conductive hearing loss.
- Central nervous system: headache, psychosis, V and VII cranial nerve palsy, ataxia, hemiplegia, seizures.
- Renal system: microhematuria, proteinuria, tubulointerstitial nephritis, IgA nephropathy, glomerulonephritis, membranous nephropathy.

Clinicians should be aware of the diagnostic criteria of RP according to McAdam et al., which are the following:

1. Recurrent chondritis of both auricles
2. Non-erosive inflammatory polyarthritis
3. Nasal chondritis
4. Ocular inflammation
5. Respiratory tract chondritis
6. Audiovestibular damage

- Histological confirmation is not required. Three or more clinical features are necessary for the diagnosis of RP.
  - Modified criteria were proposed by Damiani and Levine. They suggested the following:

    At least one of the six clinical features suggested by McAdam, with histological confirmation

    or

    Two of six criteria indicated by McAdam and positive response to corticosteroids or dapsone
  - Another modification of McAdam criteria was proposed by Michet et al., who suggested:

    Confirmed inflammation in two of three cartilages (auricular, nasal, laryngotracheal)

    or

    Confirmed inflammation in one of the above three cartilages, combined with two minor criteria among hearing loss, vestibular dysfunction, seronegative polyarthritis, ocular inflammation

63.3.2 Treatment

There are still controversies in the treatment of relapsing polychondritis, because of the rarity and the unknown etiology. The medical treatment varies and depends on the activity of the disease and the number of systems involved.

Medical treatment modalities include NSAIDs and aspirin for the control of pain and inflammation, dapsone, colchicines, and systemic steroids. As second-line treatment in severe disease, other immunosuppressants can be used, such as Cyclophosphamide, Cyclosporine, Azathioprine, Methotrexate (combined or not with systemic corticosteroids). The efficacy of biological agents such as Infliximab, Rituximab, Abatacept, Anakinra, Etanercept, and Certolizumab is also under investigation.

Surgical treatment is indicated in patients with complications related to RP and may include tracheostomy, tracheoplasty, balloon dilatation, tracheal stent, saddle nose deformity repair, and aneurysm—cardiac valve repair.

63.3.3 Follow-Up

RP is a progressive disease, usually with fluctuating symptoms. Its severity and morbidity vary and are related to age, sex (male > female), presence of cardiac and airway complications, multisystemic disorder, systemic vasculitis, hematologic malignancy, and renal involvement.

According to Trentham and Le, the survival rate is 94% at 8 years. In earlier studies, the 5-year survival rate was 70%, decreased in cases of systemic vasculitis.

Summary and Author's Comments

1. Relapsing polychondritis is a severe, rare, progressive, multisystemic autoimmune disease, and a cause of death in delayed diagnosis or if left untreated.
2. A thorough history and physical examination are critical to the diagnostic procedure. Clinicians should be aware of this disease and McAdam—modified McAdam criteria.
3. Nasal manifestations of the disease are not specific and nasal septum biopsy may be helpful in some cases.
4. Medical treatment is still a challenge and the surgical treatment is indicated in cases of complications.

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Sarcoidosis

64

Mohamed Morsy and Marios Stavrakas

64.1 Case Presentation

A 42-year-old female patient presenting with bilateral nasal obstruction, blood-stained nasal discharge, facial pain & crusts. She has also noticed redness in the skin on the tip of her nose. She has also noticed a change in the quality of her voice and progressive dyspnea. The nasal examination has shown bloody crusts, erythematous inflamed nasal mucosa and septal perforation with granulations in the nasal cavity. A chest x-ray has shown bilateral hilar lymphadenopathy and routine blood tests have picked up hypercalcemia.

64.2 Background Knowledge

Sarcoidosis is a chronic granulomatous disease of unknown aetiology. It is characterized by non-caseating granulomas involving the lungs, the upper respiratory tract, the skin, and lymph nodes. Ninety percent of patients with sarcoidosis will have thoracic involvement either within the lung itself or affecting hilar lymph nodes. The Krespi staging classifies sarcoidosis into mild (stage I), moderate (stage II) or severe (stage III).

64.3 Clinical Approach

Nasal manifestations precede the onset of skin lesions by several years. These include bilateral nasal obstruction, blood-stained nasal discharge, facial pain, anosmia & crusts. Up to 9% of patients experience sinonasal disease. Cutaneous lesions are also common with a typical purple-red discoloration of the tip of the nose, known as lupus pernio, as well as subcutaneous nodules. Patients with sinonasal involvement tend to have a more severe systemic disease, a longer history of disease and require systemic treatment in higher doses than controls with sarcoidosis but without sinonasal involvement.

64.3.1 Examination

The nasal examination can show septal perforation and erythematous nasal mucosa with granulations. The nasal cavity could be filled with crusts. The nasal tip can show lupus pernio. The laryngeal examination can show supraglottic lesions, mainly affecting the epiglottis.

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64.3.2 Investigations

Sarcoidosis was previously diagnosed using the Kveim test, but this has been withdrawn in the United Kingdom due to health and safety reasons. Increased angiotensin-converting enzyme (ACE) levels are characteristic. Serum calcium may be elevated in systemic disease, and chest x-ray shows bilateral hilar lymphadenopathy, or other abnormalities, depending on the stage of the disease. Histology shows non-caseating granuloma and finally, CT of the paranasal sinuses may show non-specific changes consistent with CRS.

64.3.3 Medical Treatment

Management should be multidisciplinary with a respiratory physician being involved. The mainstay treatment is systemic steroids and steroid-sparing agents. Topical treatment for the nose includes nasal douching, topical steroids and lubricants. Intralesional steroids may be used in cutaneous or laryngeal lesions.

64.3.4 Surgical Treatment

Surgery (e.g., rhinoplasty) should be avoided if possible but may be considered if the patient has been in remission for several years or for complications (e.g., CO_2 or Nd:YAG laser to adhesions, nasal stenosis or lupus pernio). The same applies to endoscopic sinus surgery, and patient selection should be cautious, and prognosis must be guarded.

Summary and Author's Comments

1. Sarcoidosis is a systemic granulomatous disease, which can present with nasal manifestations similar to other granulomatous diseases affecting the nose.
2. Lung involvement is common in up to 90% of patients.
3. Treatment should be multidisciplinary, with steroids and steroid-sparing drugs being the mainstay of treatment.
4. Topical steroid sprays and saline nasal douches are used to control nasal symptoms, with endoscopic sinus surgery being reserved for selected cases.

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Cocaine Abuse (Cocaine-Induced Midline Destructive Lesions—CIMDL)

65

Mohamed Morsy and Marios Stavrakas

65.1 Case Presentation

A 23-year-old male patient presenting with worsening nasal symptoms, including a feeling of bilateral nasal obstruction, blood-stained nasal discharge and whistling noise during breathing. He gave a history of cocaine sniffing. Nasal endoscopy revealed a large septal perforation and remodeling of the other structures of the nasal cavity. He was advised to discontinue cocaine use, started on nasal douching and nasal ointments, and was listed for a septal button insertion.

65.2 Background Knowledge

Long-term cocaine abuse can induce granulomatous inflammation and destruction of the nose, sinuses and palate that may be clinically indistinguishable from GPA or other granulomatous conditions. This process is caused by the marked vasoconstrictive effect of cocaine, particularly if it is mixed with levamisole. A history of intranasal substance abuse should, therefore, be sought in all patients presenting with such symptoms.

65.3 Clinical Approach

65.3.1 Clinical Presentation

The patient is usually an adolescent or young adult with a history of intranasal substance abuse. It often presents with a feeling of bilateral nasal obstruction and bloody crusts or blood-stained nasal discharge. Presentation with septal perforation and saddle nose deformity is also common. Midline nasal and facial destruction can also occur.

65.3.2 Examination

The nasal mucosa looks markedly congested and inflamed with bloody crusts. Destruction of the nasal septum with septal perforation is frequent. Midline nasal and facial destruction can happen. Other possible differential diagnoses of the same presentation include invasive fungal sinusitis, midline lethal granuloma (T-cell lymphoma), squamous cell carcinoma, and GPA.

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65.3.3 Investigations

ANCA is often positive in this condition, with PR3 reactivity in more than 50%, which compounds the similarity with GPA. Still, there are some subtle differences in caspase 3 and 9 expression and ANCA reactivity with neutrophil elastase that can distinguish the two conditions. A high apoptotic cell index on biopsy specimens from patients with CIMDL may also help to differentiate CIMDL from GPA.

If cocaine abuse is highly suspected, but there is no history of intranasal substance abuse given, it can be checked in urine or hair sample, after consenting the patient accordingly.

65.3.4 Medical Treatment

Cessation of cocaine abuse should be the first action take. Topical nasal treatments to provide symptomatic relief and prevent built-up of crusts such as regular saline irrigation and emollients such as vaseline. Surgical reconstruction is often challenging, with poor outcomes. Septal buttons can help in the symptomatic relief.

Summary and Author's Comments

1. Cocaine sniffing can result in granulomatous inflammation of the nose, similar to that of GPA.
2. Long term cocaine abuse can lead to midfacial destruction.
3. Cessation of cocaine sniffing and saline nasal irrigation is the mainstay of treatment.
4. Surgical repair of septal perforation could be attempted after stopping cocaine sniffing, but the outcomes are variable.

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Hereditary Haemorrhagic Telangiectasia (HHT)

66

Marios Stavrakas, Manuela Cresswell, and Hisham S Khalil

66.1 Case Presentation

A 32-year-old woman with Hereditary Haemorrhagic Telangiectasia (HHT) and recurrent severe epistaxis had partial and temporary improvement of nosebleeds with topical ointments and laser ablation. She had frequent hospital admissions for repeated laser ablation every 3–4 months with the need for multiple general anaesthetics.

She developed anterior septal perforation secondary to previous laser ablation to the nasal septum, resulting in crusting and whistling. A decision was made to use a septal button to close the perforation as opposed to surgical closure. The patient has since noticed a significant improvement with reduction of the frequency and severity of epistaxis. Over the last 5 years she has had the septal button changed twice.

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66.2 Background Knowledge

Hereditary haemorrhagic telangiectasia (HHT) is an autosomal dominant disorder with variable penetrance. The estimated prevalence is 1 in 5000, but there is a broad geographic variability. Histologically, HHT is characterised by dilated venules and a lack of contractile elastic fibres. These changes also may lead to arteriovenous malformations (AVMs) and telangiectasia formation, and all the elements mentioned above predispose to bleeding.

66.3 Clinical Approach

66.3.1 Diagnosis

The most frequent clinical manifestation among HHT patients is epistaxis, which affects up to 98% of the HHT population. Other signs include telangiectasias of the lips, tongue, oral cavity and gastric mucosa. Clinical diagnosis is based on the Curacao criteria:

1. Epistaxis that occurs spontaneously on more than one occasion
2. Telangiectasias at characteristic sites including the nose, fingers, oral cavity
3. Visceral lesions such as pulmonary, hepatic or cerebral AVMs

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Table 66.1 Investigations if HHT is suspected or confirmed according to clinical criteria

Central nervous system involvement	• MRI head	Necessary to screen children as the AVMs are high flow and have a more aggressive natural history A child with a negative MRI should be rescanned in adulthood
Pulmonary involvement	• Transthoracic contrast ECHO with agitated physiological saline • CT thorax • Angiography	Transthoracic contrast ECHO is recommended as the initial screening tool for pulmonary involvement
Gastrointestinal involvement	• Upper endoscopy (OGD)	Endoscopy is not recommended unless a patient is symptomatic or has anaemia that is disproportionate to the degree of epistaxis he or she is experiencing
Hepatic involvement	• Doppler U/S • CT abdomen	Known HHT patients with elevated liver function tests or clinical symptoms should be investigated

4. Family history of HHT (first-degree relative diagnosed with HHT via the same criteria)
   (a) Definite 3–4
   (b) Possible 2
   (c) Unlikely 0–1

Screening is essential as cerebral and pulmonary AVMs may have catastrophic and life-threatening consequences (Table 66.1). In these cases, treatment should be considered even in asymptomatic patients. When patients are symptomatic with gastrointestinal, liver, or oral cavity bleeding, treatment should also be considered.

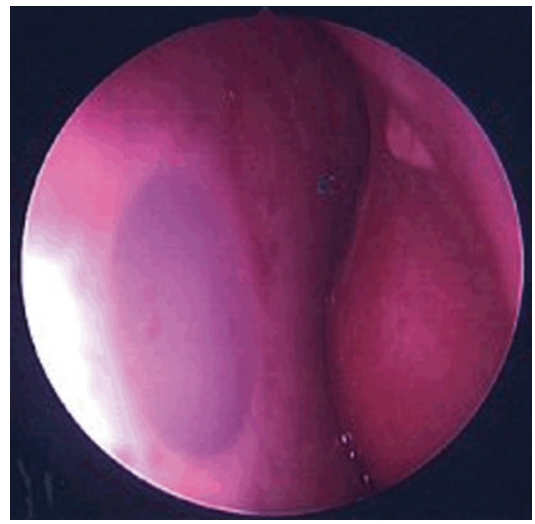

Fig. 66.1 A nasal endoscopic view of the left nasal cavity with the septal button in situ and telangectatic vessels visible in the area covered by the septal button

66.3.2 Treatment

Surgical closure with mobilisation of septal flaps would have been challenging given the bleeding risk in HHT. The patient underwent nasal septal button placement (Fig. 66.1), following which she described not only improvement of nasal crusting but a marked reduction in the frequency and severity of epistaxis. The septal button had to be changed twice over the last 2 years due to nasal discomfort. It is proposed that nasal septal button placement can reduce turbulent nasal airflow and improve surface protection of nasal mucosa from dryness, preventing need for repeated surgical procedures.

Most surgeons adopt a stepwise approach to HHT treatment, paying attention to the symptoms, the frequency of epistaxis and possible Hb drop.

- Conservative measures:
- Avoidance of triggers, prevention of nasal dryness and crusting, use of saline douches, saline-based nasal gels, mupirocin ointment.
- Medical treatment:
  – Topical oestrogen, aiming to induce septal mucosal metaplasia.

  - Topical/PO/IV tranexamic acid, aiming to stabilise clots and influence endothelial cells directly.
  - Bevacizumab, which is a VEGF inhibitor, aiming to affect angiogenesis. It has been injected submucosally and showed promising results. Although it may need to be administered repeatedly or indefinitely, it allows significant symptom-free periods.
  - Tamoxifen, an anti-oestrogen drug commonly used for breast Ca.
- Surgical
  - Coagulation/coblation.
  - Laser photocoagulation.
  - Septodermoplasty: replacing the diseased mucosa with a full or split-thickness skin graft, laying on top of the remaining perichondrium. This has been shown to reduce the need for multiple laser coagulations by up to 57%.
  - Young's procedure: closure of the nasal vestibule. It is effective, but the surgeon should take into consideration the changes that will take place, including anosmia and taste disorders.

Our preferred method of coagulation is photocoagulation with KTP laser, taking advantage of its flexible fibre to reach the various abnormal sites and the effective ablation of the blood vessels due to its wavelength. If necessary, the procedure can be staged to avoid septal perforation. Also, we try to avoid nasal packing but if required, we prefer dissolvable packing materials.

66.3.3 Follow Up

HHT patients remain under regular follow up and coagulation of the vessels is repeated as necessary. Sometimes we prefer to stage surgical interventions and alternate sides of coagulation aiming to prevent complications such as septal perforation.

Summary and Author's Comments

1. Key histopathological feature is the lack of contractile elastic fibres.
2. Minimal instrumentation and dissolvable packing material are important in the acute setting.
3. Treatment is based on the frequency and severity of nosebleeds, available equipment and surgical expertise.

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Facial Pain

67

Marios Stavrakas and Hisham S Khalil

67.1 Case Presentation

A 47-year old female patient was referred to the Nasal Disorders clinic to investigate her for persistent facial pain. A provisional diagnosis of chronic rhinosinusitis was made and the patient had a course of intranasal corticosteroids without any encouraging results. She described the pain mainly as a feeling of pressure and discomfort over the nasal bridge, nasion and periorbital regions. There was no history of migraines or headaches, the patient described intermittent nasal obstruction but had no other concerns. Flexible nasendoscopy and CT scan of the sinuses were normal, without any signs of chronic inflammation or any other sinonasal pathology. After excluding other causes of headaches and facial pain, the diagnosis of midfacial segment pain was made and the patient was started on Amitriptyline 10 mg once daily at night. This improved her symptoms and no further actions were taken apart from monitoring the effect of medical treatment and the need for dose adjustment.

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67.2 Background Knowledge

67.2.1 History

History taking is targeted at excluding sinonasal pathology and malignancy (primary or metastatic). Many of the patients that are referred with facial pain do not suffer from chronic sinusitis but from some other type of headache, so we need to focus on identifying characteristics of alternative causes of facial pain or headache.

67.2.2 Examination

Head and neck examination including flexible nasendoscopy is necessary and aims to identify any sinonasal pathology, cervical lymphadenopathy or cranial nerve deficits. Inspecting the face for any lesions, swelling or colour changes of the skin is prudent. Identification of trigger points, signs of temporomandibular joint disfunction, oral and dental pathology may assist in selecting the appropriate diagnostic pathway.

67.2.3 Investigations

CT scan of the sinuses is helpful and we believe it should be performed when investigating persistent facial pain with normal clinical examination.

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67.2.4 Treatment

Amitriptyline or Nortriptyline can be used for midfacial segment pain. We start with a low dose, e.g. 10 mg once a day (nocte) and titrate the dose according to the patient's response, usually at 2–3 week intervals. It is advised to check for any other current medications that may interact with them and warn the patients about potential side effects. If treatment is ineffective and cessation is decided, gradual reduction of the dose is recommended and the patient needs to seek medical advice before doing so. More complex cases may require involvement of a Neurologist or the pain team.

ENT surgeons see a significant number of patients with facial pain, referred as sinonasal pathologies.

67.3 Clinical Approach

A significant proportion of the patients who are referred to ENT for investigation of their facial pain do not have any sinonasal pathology. West and Jones (2001) found that approximately 25% of their referrals had normal nasal endoscopy and CT of the paranasal sinuses. Among these patients, the most prevalent diagnosis was midfacial segment pain. Moreover, the additional value of CT scan is doubtful, as anatomical variations can be seen in asymptomatic patients too.

Intranasal contact points have been regarded as a potential cause for facial pain for several years but this does not seem to be accurate according to more recent studies. The removal of these contact points rarely results in total elimination of the pain and the postoperative improvement of some symptoms can be attributed to neuroplasticity or cognitive dissonance. This agrees with other studies which support that surgery does not help significantly patients with midfacial segment pain and may even worsen the pain in 25% of this group.

Table 67.1 summarises the differential diagnosis of facial pain of non-sinugenic origin.

Table 67.1 Common types of headache and facial pain

Diagnosis	Characteristics
Migraine	Common type of headache, characterised by a combination of neurological, gastrointestinal and autonomic symptoms. Affects 8.1 in 1000, M:F = 1:6. About 70% of patients have a positive family history. It can present with or without aura and symptoms may include photophobia, phonophobia, visual disturbances, nausea, vomiting, vertigo and fatigue. Headaches can be unilateral (60%) or bilateral (40%) and can last for 4–72 h.
Tension-type headache	Can be divided in episodic and chronic. These headaches are characterised by generalised pressure or tightness in the head. The discomfort is not affected by activities and nausea, phonophobia or photophobia are not prominent symptoms. A significant number of migraine sufferers (94%) have co-existing tension-type headache.
Cluster headaches	They comprise headaches with signs of cranial autonomic hyperactivity. M:F = 9:1, prevalence 0.1–0.4% and age of onset is usually 20–40 yo. Attacks occur for several weeks or months, then remit, leaving the patients pain free for several months or years, then the pain recurs. 20% present Horner's syndrome. Nausea and vomiting are uncommon. Trigger factors: alcohol, vasodilator drugs, apnoea-induced hypoxaemia.
SUNCT	Short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT), is characterised by the extremely short duration of attacks (20–120 s) and high frequency (30–100 times/day). They occur during daytime and the pain is confined to the distribution of the V1. It may be mistaken for trigeminal neuralgia, but the characteristic symptoms are conjunctival injection and tearing.
TMJ dysfunction	Refers to the group of muscle and joint disorders around the TMJ. It is a common problem and affects 30–46% of population, with pain occurring in 5–15%. Most TMJ disorders present with pain in front of the ear and carry an element of myofascial pain.

Table 67.1 (continued)

Diagnosis	Characteristics
Trigeminal neuralgia	Incidence is 3–5/100.000, with a peak onset between 50 and 60 yo. The most characteristic element is agonizing, paroxysmal sharp pain confined strictly to one or more branches of the trigeminal nerve. Mild flushing may occur, and the pain is almost always unilateral. About 80% of cases are associated with vascular compression and trigeminal neuralgia may present in up to 5% of patients with multiple sclerosis.
Post-herpetic trigeminal neuralgia	Associated with HSV infection. Neuralgia consists of pain persisting in the affected dermatome (typically affects the first division of the trigeminal nerve). May be associated with scarring, loss of pigmentation and hair, burning pain or dysaesthesia.
Midfacial segment pain	This is a form of tension-type headache, which has all the same qualities of the pain but it affects the face and may involve the nasion, under the bridge of the nose, either side of the nose, the periorbital region, retro-orbitally or across the cheeks. It is described as a dull ache, feeling of pressure or tightness. It can be chronic or episodic and the skin and soft tissues of the area may be sensitive to touch.

Other types that may cause orofacial pain are glossopharyngeal neuralgia (similar to trigeminal neuralgia, located in the distribution of the glossopharyngeal nerve), burning mouth syndrome (chronic sensation of burning in the oral cavity and tongue, without any dental or medical causes), medication-overuse headache (chronic pain due to chronic overuse of analgesics, especially ergotamine, triptans, simple analgesics, opioids and combination analgesics) and atypical facial pain (persistent idiopathic pain that does not fit into any other diagnosis).

The treatment is basically medical and depends on the diagnosis. Refractory cases may benefit from an opinion from an OMFS colleague or a neurologist. In some units, there are joint facial pain clinics between ENT and OMFS surgeons, Neurologists and Anaesthetists specialised in pain.

Summary and Author's Comments

1. Facial pain is associated with a very heterogenous group of diagnoses.
2. History and detailed clinical examination can guide the clinician to the correct management pathway.
3. It is essential to have in mind the basic characteristics of each pathology and involve other specialties if deemed necessary.
4. Sinus surgery does not necessarily improve facial pain unless there is confirmed sinonasal pathology.

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Part X

Orbital Pathology

Epiphora

68

Georgios Vakros, Marios Stavrakas,
and Hisham S Khalil

68.1 Case Presentation

An 83 year old woman presented with a history of bilateral epiphora and conjunctival discharge. She was awaiting bilateral cataract extractions which had to be postponed till the epiphora and discharge were treated. She underwent a combined ENT and Ophthalmological assessment prior to listing for bilateral endoscopic dacryocystorhinostomy (DCR) under general anaesthesia. Her symptoms resolved and she was able to proceed with her cataract surgery.

68.2 Background Knowledge

Epiphora or watery eye is the excessive tearing which results from the imbalance between the production and the drainage of tears. There are several etiologies that will influence that balance ranging from tears overproduction to drainage failure as well as defective tear removal mechanism. The identification of the specific cause requires a complete evaluation of the lacrimal system, the ocular surface and eyelids. Only by following a systematic approach the treating clinician will be able to provide the correct treatment for the patient, as epiphora tends to be a very troubling symptom, which often is managed poorly.

68.2.1 Anatomy and Physiology of The Lacrimal System

68.2.1.1 Secretory System

The main secretory gland of the lacrimal system is the lacrimal gland (LG). It is located within the lacrimal gland fossa of the frontal bone, at the superolateral aspect of the orbit. The LG is divided into 2 lobes, the orbital and the palpebral. These two lobes are separated by the levator palpabrae aponeurosis. It is an exocrine gland and the excretory lacrimal ducts empty 5 mm above the lateral tarsal border in the conjunctival cul-de-sac, after passing through the levator aponeurosis and Muller's muscle. The LG receives sensory innervation (afferent) from the ophthalmic branch of the trigeminal nerve and its efferent consists of both sympathetic and parasympathetic nerves. Afferent nerves from

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the ocular surface activate the efferent parasympathetic nerves thus stimulating the production of tears.

68.2.2 Drainage System

The tears drainage system originates at the level of the puncta located medially on the margin of the upper and lower eyelid. The normal position is to be opposed on the globe, within the tear lake in order to ensure good drainage of tears. The puncta are surrounded by the ampulla giving its fleshy elevation at the eyelid margin. The ampulla leads to a vertical segment of the canaliculus which extends for 2 mm prior turning 90°, continuing 8–10 mm medially until they join to form the common canaliculus which finally joins the lacrimal sac. The common canaliculus separates from the lacrimal sac by the valve of Rosenmuller and its function is hypothesized to be a preventative mechanism of tears reflux. The lacrimal sac lies within the bony fossa formed by the anterior and posterior lacrimal crests. It is surrounded by the medial cantus, which wraps around the anterior and posterior aspects of the sac. Part of the sac is extending few millimeters above the medial canthal tendon. Inferiorly, the lacrimal sac transitions into the nasolacrimal duct (NLD), which in adults it measures 12–18 mm. It travels in a posterolateral fashion through the bone and opens into the nose through an ostium under the inferior turbinate. The inferior meatus is partially covered by a mucosal fold (valve of Hasner) and this is located 30-35 mm from the external nares.

There are several valves within the lacrimal system in which not all have a functional purpose. The most clinically significant are the valves of Rosenmuller (preventing air reflux when blowing the nose) and Hasner (prevents regurgitation of fluids from the nose up). The following figure (Fig. 68.1) demonstrates the known valves of the lacrimal system.

Tears spread across the ocular surface with the blinking mechanism by the action of the orbicularis oculi. The muscle directs the tears medially into the punctum and the fibers surrounding it help it into pumping the tears down the lacrimal system.

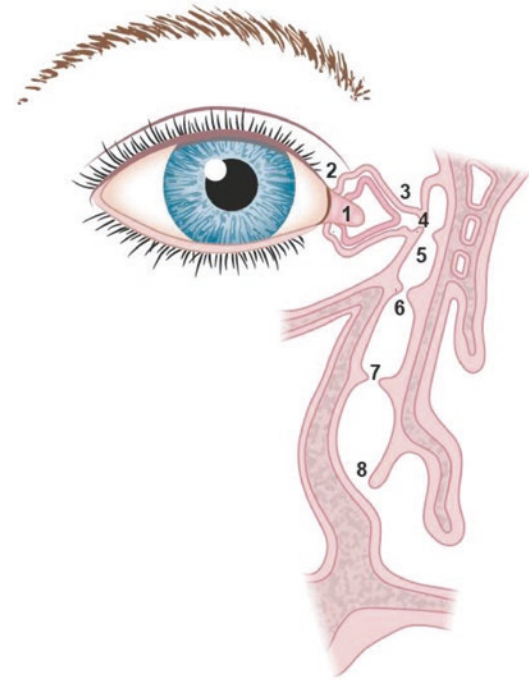

Fig. 68.1 Anatomy of the valves of the lacrimal system: (1) Bochdalek, (2) Foltz, (3) Huschke, (4) Rosenmuller, (5) Medial palpebral ligament, (6) Bernard/Krause, (7) Tailefer, (8) Hasner (adapted from Snell and Lemp)

68.2.3 Diagnosis

The tearing patient requires careful and systematic evaluation in order to reach the accurate diagnosis and deliver targeted treatment. A focused history is of vital importance as it can guide towards the underlying etiology while the clinical examination and testing confirms the diagnosis.

The commonest cause of epiphora is due to unstable tear film, which manifests as dry eye syndrome. This originates either from dysfunction of the meibomian glands, oil secreting glands located posteriorly to the eyelashes leading to evaporative dry eye (EDE) or from aqueous/mucous deficiency related to lacrimal gland/goblet cell dysfunction leading to aqueous deficient dry eye (keratoconjunctivitis sicca; KCS) or mixed mechanism. The symptomatology of dry eye is variable though most patients usually present with ocular irritation e.g., foreign body sensation or burning/itchy sensation as well as intermittent blurriness of vision and redness.

Their symptoms tend to worsen in the presence of wind, air conditioned or dry environment and when the patient is outside the house than inside. These types of patients require evaluation by an ophthalmic team since slit lamp examination is needed for evaluation with the use of fluorescein of the conjunctival and corneal surface.

In addition, abnormalities in lid position also can cause symptoms of epiphora. The patient can describe either persistent or intermittent epiphora. Lid laxity influences the function of the normal tear dispensing mechanism resulting in pooling of tears and overflow. This can be in the form of medial ectropion where the disorientation of punctum in relation to the ocular surface—normal punctum is in contact with the ocular surface—leads to failure of the drainage of tears down the punctum and thus overflow. Furthermore, the presence of entropion leads to corneal irritation from the eyelashes resulting in reflex lacrimation. To assess overflow and normal flow of fluorescein, the *dye disappearance test* (DDT) is utilized as an essential part of the clinical examination.

DDT is useful for assessing the presence or absence of sufficient outflow of the lacrimal system. It is particularly useful in children since any other tests require sedation or a very co-operating child. The DDT is performed by applying a drop of fluorescein 2% or a moistened fluorescein strip in the conjunctival fornix. The examiner observes the tear film distribution; preferably with the use of cobalt blue light filter fitted on the slit lamp, direct ophthalmoscope or pen torch. The observation time is for 5 min and the examiner assessing the clearance of dye from the tear meniscus. If dye persists following the observation interval, this indicates lacrimal drainage obstruction. DDT can be normal in the presence of intranasal obstruction.

In order to assess the integrity of the lacrimal system, additional tests are those related to irrigation of the drainage system. A drop of topical anesthetic (proxymetacaine 0.5% or oxybuprocaine 0.5% or tetracaine 1%) is instilled in the inferior fornix. The lower punctum is dilated with a punctum dilator, if narrow, and a 26 gauge irrigating cannula is placed in the canalicular system. It is advanced 2 mm vertically and then about 2–3 mm horizontally while lateral traction of the lower eyelid is applied to avoid canalicular kinking. Clear saline is then injected and the results noted regarding reflux and whether mucous or fluorescein is present (Fig. 68.2). Warn the patient regarding the feeling of fluid at the oropharynx.

Other tests that can be done to assess the patency of the lacrimal system include Jones I and II.

A. *Jones I* is investigating the lacrimal system outflow under physiologic condition. It is performed in a similar manner to the DDT with the only difference being that the examiner passes a cotton tip applicator in the inferior nasal meatus at the level of the ostium of the NLD at 2 and 5 min. Presence of fluorescein between 2 to 5 min indicates a patent duct while absence indicates obstruction though it does not highlighting at which level.
B. *Jones II* is following Jones I test, if the former is negative (no fluorescein picked up). After flushing the residual fluorescein from the conjunctival fornix, a cotton tip applicator is inserted in the nose at the lower nasal meatus (or use the pre-existing one if Jones I test is negative). A 26-gauge cannula is then inserted in the canalicular system and the system is irrigated with normal saline. If the solution retrieved from the nose is clear, this is indicates that the fluorescein (or in reality the patient's tears) cannot enter the lacrimal outflow system suggesting of obstruction. If fluorescein is retrieved, this indicates partial NLDO.

In normal circumstances, there should be no or very minimal reflux from the upper canaliculus. If difficulty found while advancing the irrigating cannula and inability to irrigate fluid this is highly suggestive of total canalicular obstruction (Fig. 68.2a). This will also be associated with pain. If saline can be irrigated successfully but it refluxes from the upper canaliculus without lacrimal sac distention palpable, then it is suggestive of complete common canalicular blockage (Fig. 68.2b). If mucoid material refluxes through

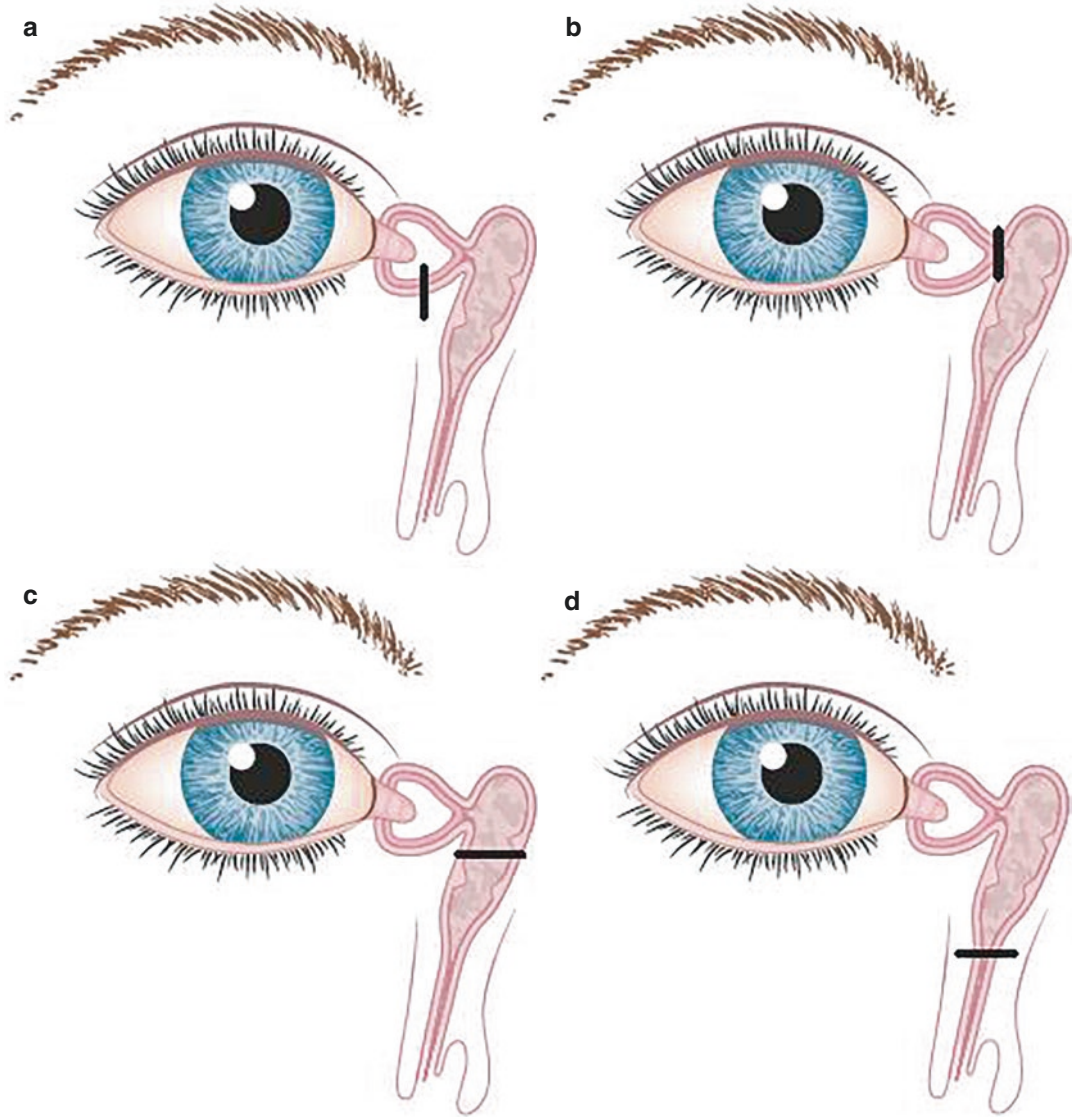

Fig. 68.2 Possible areas of lacrimal system obstruction (black line). (**a**) At the level of canaliculus, (**b**) at the level of common canaliculus, (**c**) at the level of NLD, (**d**) partial obstruction

the opposite punctum with lacrimal sac distention, this is suggestive of complete NLDO (Fig. 68.2c). Finally, if some reflux occurs through the opposite punctum with some of the saline irrigating the system, then the diagnosis is partial NLDO or NLD stenosis (Fig. 68.2d). A patent canalicular system can be present in functional NLDO.

Other additional tests, which are particularly useful in assessing anatomical and functional abnormalities, are contrast dacryocystography (DCG) and dacryoscintigraphy (DSG) respectively. In the former, a radiolucent dye is injected through both canaliculi followed by a computerized digital subtraction imaging. This provides excellent anatomical information regarding the nasolacrimal system and is particularly useful in identifying the problem in cases of failed DCR for example. In case of DSG, radionucleotide eye drops are given to the patient to use and a

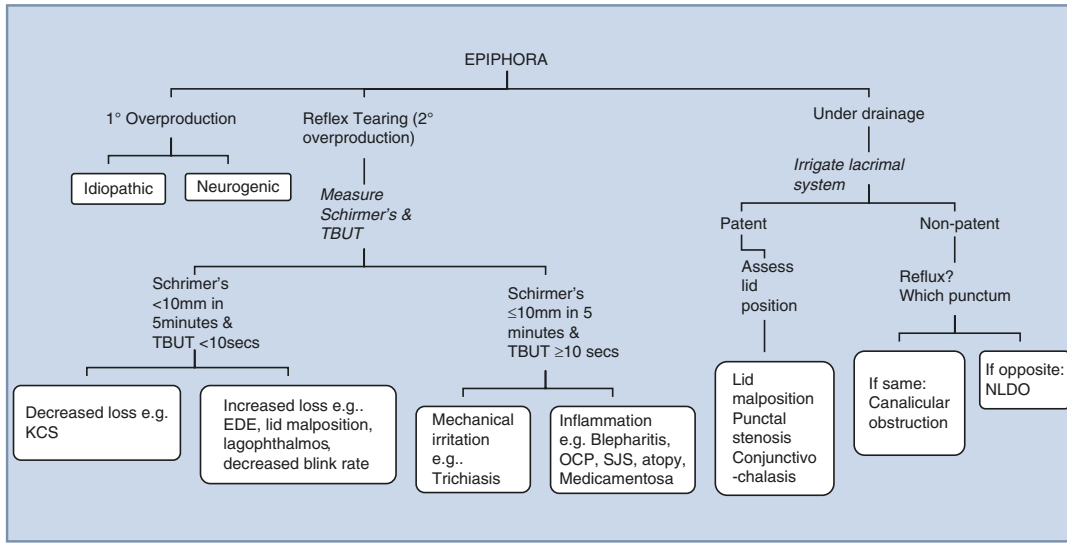

Fig. 68.3 Diagnostic algorithm for epiphora (Adapted from Price and Richard). *TBUT* tear breakup time, *KCS* keratoconjunctivitis sicca, *EDE* evaporative dry eye, *OCP* ocular cicatricial pemphigoid, *SJS* Stevens-Johnson syndrome, *NLDO*: nasolacrimal duct obstruction, *Medicamentosa* drug related adverse/side effects

scintigram is taken in order to assess the tear flow. In normal circumstances, the canaliculi an the lacrimal sac should be visible within 10–12 s and within 10–30 min in the nasal cavity. This is useful in cases of functional NLDO though it does not provide anatomical detail. Finally, computerized tomography or magnetic resonance imaging is used in cases of craniofacial injury or congenital deformities or in case of suspected malignancy and they can also provide information regarding coexisting sinus or nasal disease.

In addition to the clinical tests, physical examination signs can also provide with valuable information regarding the cause of the epiphora and can aid to the underlying diagnosis. Measuring the blinking rate (normal blinking occurs every 10 s) and whether the eyelids close fully can guide towards an ocular surface driven epiphora. Part of the lacrimal system test is to palpate the lacrimal sac for distention. It is important to document whether there were any signs of tenderness as this can be a sign of acute inflammation. The following diagram (Fig. 68.3) can be used as a memoir for the diagnostic approach.

68.2.4 Management

After a careful evaluation and once diagnosis is reached, the appropriate treatment plan needs to be decided. Epiphora related to nasolacrimal duct obstruction requires different approach between children and adults.

In children, the NLDO is congenital and the initial management step is to perform Criggler massage as this can help the opening of the valves (usually related to either persistent valve of Rosenmuller or Hasner). Recurrent use of topical antibiotics is not indicated, as this is ineffective and also conjunctivitis is not related to an active infection. If the initial approach fails (if it hasn't resolved by the age of 1–2), then probing with lacrimal syringing under general anesthetic is required, which is successful in the majority of cases. If is unsuccessful, this can be either due to anatomical abnormality or rarely, creation of false passage during the probing. In such cases, nasolacrimal intubation with silicone tubes in situ for 2–5 months can have up to 90% success rate. An alternative to intubation is that of balloon catheter dilatation that can be an alternative

approach and studies have suggested 77–90% resolution in comparison to intubation. Lastly, if all of the above have failed and the child suffers from dacryocystitis or dacryocystocele then dacryocystorhinostomy (DCR). DCR has both an endonasal and external approach with the former being highly successful and less complicated than external approach. In failed DCR, a repeat DCR can take place though the decision depends on what are the findings on the DCG, as the patient might need conjunctivodacryocystorhinostomy (CDCR) (rare).

In adults the most common cause of epiphora is due to reflex lacrimation (2° overproduction) or eyelid related abnormality and therefore, referral to the oculoplastic surgeon or comprehensive ophthalmologist is indicated for evaluation and treatment. Involutional entropion or ectropion need to be corrected; the former requires retractor re-insertion with lateral canthopexy or tarsal strip while the latter horizontal lid tightening with lateral canthopexy or tarsal strip. Additionally, in cases of punctual ectropion, medial spindle with/without lid tightening can be very successful.

In some cases where the patient has punctual stenosis especially those with previous infection or chronic inflammation, then a 3- snip punctoplasty can help with the symptoms if the NLD is patent.

In cases of where the level of partial obstruction is at the level of canaliculus or common canaliculus, the initial step can be the use of silicone tubes intubation that stays in place for 2–3 months and that can be effective. If however, there is a complete obstruction then the next step is CDCR with the insertion of Pyrex tube (Jones tube) or other types. In cases of canalicular block, balloon canaliculoplasty or localized canalicular excision with re-anastomosis or canalicular stents have been utilized as well as DCR with lacrimal intubation (retro tubes).

Finally, in the presence of a non-patent lacrimal system (NLDO), the surgical options are influenced by the location of the occlusion. In case of NLDO then the treatment of choice is endoscopic or external DCR. Both techniques have a very high success rate however; each of them has its own complications and challenges. Table 68.1 indicates the pros and cons of both approaches.

Table 68.1 Comparison of endonasal with external DCR (adapted from Anijeet et al.)

Endonasal DCR	External DCR
Advantages	
No external scarring	Easier access to the lacrimal sac beyond medical canthus
Direct access to rhinostomy site with excellent visualization of the intranasal anatomy allowing correction of any intranasal pathology	High predictability and excellent visualization of lacrimal sac and duct and any internal pathology e.g. tumor
Avoids damage to the medial canthal tendon and orbicularis thus retaining lacrimal pump function	Accurate anastomosis of lacrimal sac and nasal mucosa
Shorter operating time with faster recovery	Better if mucocele present
Disadvantages	
Slightly lower success rates especially with re-operation	External facial scar
Poorer access to the superior aspect of lacrimal sac	Interferes with lacrimal pump function
Higher cost of equipment	Post-operative infection/ skin abscess
More technically challenging	

The endonasal surgical approach is as follows:

- 0° scope
- Infiltration with local anaesthetic and decongestion of middle meatus with 1:10,000 adrenaline on neuro patties
- Elevation of posteriorly based mucosal flap to expose the lacrimal bone
- Removal of bone (lacrimal and frontal process of the maxilla) in order to expose the lacrimal sac
- Probing of canaliculi through puncta
- Incision of lacrimal sac
- O'Donoghue silicone tubes and Watski sleeve for stenting
- Flap trimmed accordingly and put back to cover exposed bone
- Light packing if required

In the postoperative period it is important to warn the patient over the possibility of haemorrhage and clear instruction should be given regarding this. Furthermore, nasal washes (without nose blowing) with solutions such as Steri-Mar® spray will aid in removal of hard clots and soothes the nasal airway. The role of oral antibiotics is controversial. Antibiotic/steroid containing eye drops are commonly used in clinical practice along with nasal steroid spray.

The success rates for external DCR has been reported from 89.8% to 100% in comparison to the endonasal, which has been reported from 74.1% to 97% however, a variation in technique and definition as success is variable between these studies. The factors that appear to be influencing success are surgical experience, adequate rhinostomy, use of silicone stents and postoperative follow-up.

68.3 Clinical Approach

68.3.1 History

The patient gave a history of waking most mornings with 'sticky eyelids'. She also suffered from bilateral watery eyes which interfered with her ability to read. This was worst when exposed to the wind outdoors. The Ophthalmology team referred her for consideration of bilateral Dacryocystorhinostomies after syringing suggested bilateral nasolacrimal duct obstructions (NLDO). The patient had a history of atrial fibrillation and was on low dose Aspirin. She did not have any other relevant past medical history.

68.3.2 Examination

The patient was assessed in the lacrimal clinic providing a joint assessment by an ENT and Ophthalmic Surgeon. There was evidence of an increased tear film bilaterally and regurgitation of mucous on pressure on both lacrimal sacs. There was some laxity of both lower eye lids but no ectropion. There was increased staining of both conjunctival sacs following the fluorescein dye test which failed to clear. There was filling of the lacrimal sac and delayed regurgitation of saline from the upper lid puncti when syringing through the lower puncti and vice versa with some mucus. A diagnosis of a bilateral postsaccal NLDO was made. A flexible nasal endoscopy revealed a modest deviation of the nasal septum but was otherwise normal. The patient was offered bilateral endoscopic or external DCRs. She chose the endoscopic approach as it would have had a shorter duration for bilateral procedures.

68.3.3 Treatment

Endoscopic DCR is carried out as a joint procedure with an Ophthalmic surgeon. The authors do not stop patients taking Aspirin prior to the surgery but request they stop any other antiplatelets such as 'Clopidogrel', 7 days prior to the date of surgery.

The nasal cavities are prepared with a cophenylcaine spray (Lidocaine and Phenylephrine) in the anaesthetic room followed by the application of neurosurgical patties immersed in 1 in 100,000 epinephrine to both middle meati. This is followed by infiltration of the junction of the middle turbinate with the lateral nasal wall with 2% lidocaine/1:80,000 epinephrine. The diagnosis is confirmed on the table by lacrimal probing (Fig. 68.4). A rectangular mucosal flap is then

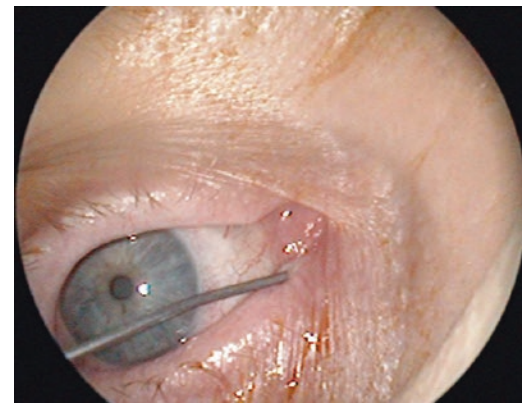

Fig. 68.4 Probing through the lower punctum at time of surgery

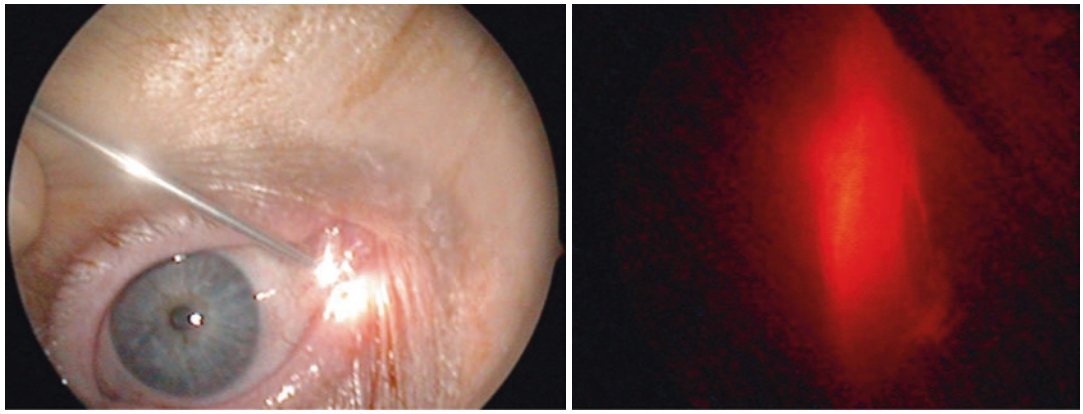

Fig. 68.5 Illumination of the right lacrimal sac using a vitreo-retinal probe

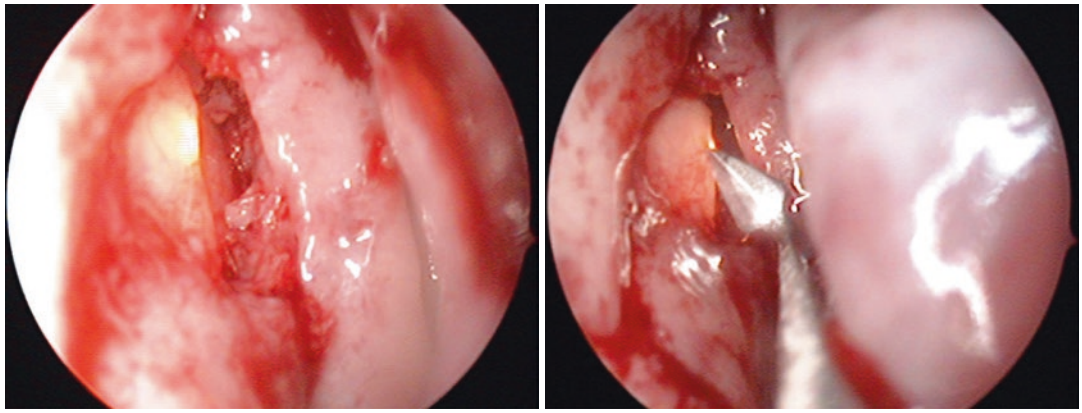

Fig. 68.6 Illuminated right lacrimal sac after removal of the bone over it and use of a keratotomy to incise the medial wall of the lacrimal sac

raised to expose the ascending process of maxilla and the uncinate process.

The bone over the lacrimal sac is then removed using a Kerrison forceps, aided by the illumination of the lacrimal sac using a vitreo-retinal probe (Fig. 68.5). The medial wall of the lacrimal sac is incised using a keratotomy knife (Fig. 68.6). The mucosa of the medial wall is then completely removed as opposed to raising flaps. O'Donoghue silicone tubes are then inserted through the upper and lower canaliculi and then approximated together using a staple, a Watski sleeve or tying them together. The silicone tube is then pulled gently through the loop in the inner angle of the eye to ensure it is neither too tight or lax (Fig. 68.7). An absorbable nasopore pack is used if there is a risk of bleeding. The patient is warned against rubbing the eyes and given a saline nasal douche for 2 weeks and antibiotic eye drops for a week. The stents are removed in 8 weeks in the ENT outpatients by cutting the loop of the silicone tube in the inner angle of the eye and then removing the stent from the nasal cavity under endoscopic control.

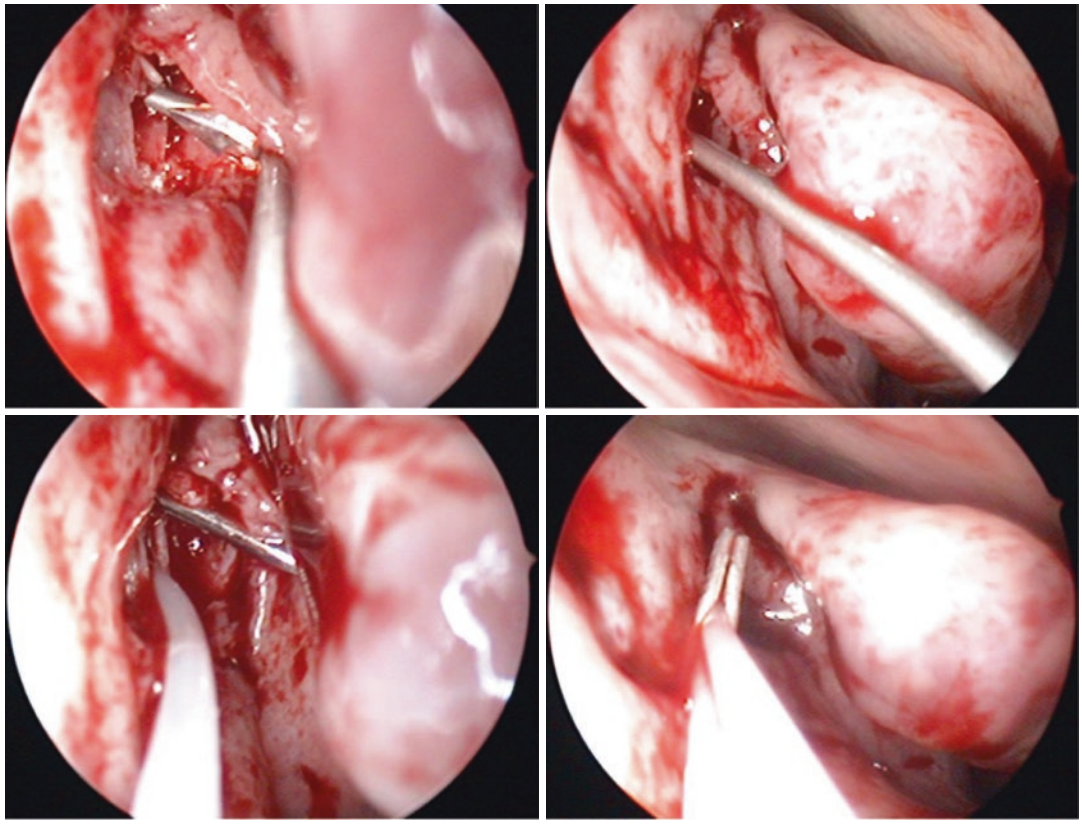

Fig. 68.7 Insertion of O'Donoghue silicone tubes and stapling of tube ends to avoid dislodgment

Summary and Authors' Comments

1. The management of epiphora is best done by an Ophthalmic surgeon or in a joint lacrimal clinic with an ENT Surgeon depending on available resources.
2. The assessment should exclude the numerous causes of epiphora other than nasolacrimal obstruction including conjunctival, corneal and lid causes. An assessment of the site of nasolacrimal obstruction is then made using probing, syringing and the fluorescein dye test.
3. Endoscopic DCR is best reserved for cases of nasolacrimal obstruction that involve the lacrimal sac or beyond in the nasolacrimal duct. It is not suitable for common canalicular (pre-saccal) obstructions.
4. An ENT assessment is essential to exclude nasal pathology that may contribute to epiphora e.g. a nasal mass and to assess the patency of the nasal cavity and the possible need for septal surgery for access if an endoscopic approach is to be carried out.

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Dacryocystitis

69

Georgios Vakros, Marios Stavrakas,
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69.1 Case Presentation

A 75-year-old lady was referred with an 8-month history of a swelling on the lateral wall of her nose, close to her medial canthus. The swelling was fluctuating in size, with mild tenderness and discomfort in the previous 6 months. The patient reported a history of previous infection in the area, which was treated successfully with a course of oral antibiotics. The diagnosis of a nasolacrimal cyst, which caused the previous episode of dacryocystitis, was confirmed by CT and MRI scans. The current episode was treated with antibiotics and the case revisited a few weeks later for definitive treatment, in the form of endoscopic marsupialisation. The patient is still under follow up, without any recurrence.

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69.2 Background Knowledge

Dacryocystitis consists of inflammation that occurs in the lacrimal sac and is usually due to infective aetiology and it can be sequelae of nasolacrimal duct obstruction (NLDO). It can be divided in acute, chronic and congenital with risk factors being structural predisposing conditions of the face and head (midface abnormalities) as well as obstructive conditions of lacrimal system (nasal fractures with deviated septum, lacrimal sac tumours/cysts, outflow obstruction due to e.g. narrow meatus, nasal polyps, chronic rhinitis etc.). The aetiology lies usually with bacterial colonization, most commonly *Staphylococcus epidemidis* or a*ureus*, Streptococci sp. as well as anaerobic bacteria and gram-negative bacteria. In the presence of blockage, the lack of tear flow and sequestration within the lacrimal system encourages bacterial overgrowth manifesting as dacryocystitis.

69.3 Clinical Approach

69.3.1 Anatomy

See epiphora Chap. 68.

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69.3.2 Diagnosis

Diagnosis of acute dacryocystitits is mostly clinical and relative straight forward. If acute,the patient presents with an acutely painful, red swelling over the medial canthal region with associated epiphora or purulent discharge. On palpation of the area, there is a firm, tender nodule inferior to the medial canthus (nodule superior to medial canthus is highly indicative of tumour of the lacrimal sac and usually associated with blood stained discharge on palpation). Purulent discharge can be expressed from the puncta, if tolerated by the patient and in some instances, a dacryocutaneous fistula can occur leading to spontaneous drainage. Congenital acute dacryocystitis in newborns or infants is a potentially life-threatening condition as it can manifest at times as a preseptal or even orbital cellulitis leading to brain abscess, meningitis and ultimately death by systemic sepsis.

In the majority of cases, no imaging investigations are required as the diagnosis is done on clinical examination. However, if presentation is atypical, inflammatory etiology should be suspected (sarcoidosis, granulomatosis with polyangiitis) and appropriate tests should be performed (ANA, ANCA, ACE). CT scan can be useful in assessing any occult malignancy or bony abnormality especially in the context of previous trauma. DCG or DSG are adjunctives in such cases as it will assist with the assessment of the lacrimal anatomy. Irrigation of the nasolacrimal system (*see epiphora chapter*) is not indicated in the acute setting as it will cause significant discomfort to the patient and the clincial benefit it provides is minimal.

Finally, nasal endoscopy can be useful in assessing the nasal passages in case of tumors, hypertrophy of inferior turbinate and septal deviation and allows direct observation of the internal meatus.

69.3.3 Management

The initial intervention for acute dacryocystitis is targetted towards resolving the infection. Broad spectrum antibiotics such as flucloxacillin or amoxicillin with clavulanic acid and performing warm compresses to encourage drainage. The role of incision and drainage remains debatable since such intervention encourages the formation of dacryocutaneous fistula. The definitive treatment once the acute phase has resolved in DCR (external or endonasal) as it will help with reducing the risk of recurrent infection.

In chronic dacryocystitis, the symptomatology and clinical presentation is rather different. The patient presents with chronic epiphora, which maybe associated with recurrent or chronic unilateral conjunctivitis. This indicates the presence of mucocele, which is clinically evident by either the presence of a painless swelling at the inner canthus or by the expression of mucopurulent canalicular discharge on lacrimal sac pressure. Treatment of chronic dacryocystitis is again with DCR and chronic use of antibiotics is unnecessary.

Summary and Author's Comments

1. Dacryocystitis is a condition, which requires prompt treatment as it can complicate to orbital cellulitis.
2. Any mass above the medial canthal tendon should alert the physician of a tumor of a lacrimal gland especially in the presence of blood stained discharge.
3. Chronic conjunctivitis of unknown etiology can be a presenting feature of chronic dacryocystitis and therefore,

requires diagnostic probing/irrigation of the nasolacrimal system.
4. Following the first episode, if DCR is not performed then the patient is at risk of future attacks.
5. Endonasal or external DCR have similar success rates.

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Globe Proptosis (Exophthalmos)

70

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70.1 Case Presentation

A 30 years old man presented with 4 weeks of a slowly progressive globe proptosis of his left side. He described the sentation of pressure behind his eye associated with some discomfort especially on extreme lateral gaze. He also reported polydipsia. He was otherwise well without any other symptoms. He has no other past medical history or any previous surgical history. He was not on any regular medications. Examination revealed 20/20 vision with full color vision and visual fields. He had a 3 mm non-axial, non pulsatile proptosis (normal right eye 20 mm), with restriction on the left lateral gaze. Otherwise, normal ophthalmological examination. A CT scan revealed a left ethmoidal mucocele. He underwent endonasal drainage surgery and the eye proptosis resolved (20 mm both eyes) along with full range of ocular motility.

70.2 Background Knowledge

Globe proptosis is the abnormal protrusion of the globe due to orbital pathology that decreases the available orbital volume resulting in the anterior bulging of the globe. It can be divided into axial proptosis and eccentric proptosis (dystopia) and it can be unilateral or bilateral. In general, clinicians nowadays for ease or habit use the term exophthalmos to describe the protrusion of the globe however, we should be aware that exophthalmos tends to mostly be used in the context of thyroid eye disease (TED).

70.2.1 Anatomy

The orbit is an open cone-shaped cavity where the apex is the optic canal and the base is the orbital rim. It is 40 mm wide, 35 mm high and 45 mm deep and thus, its volume is 30 cc. The orbit is formed from the following bones:

- Roof—lesser wing of sphenoid and orbital part of frontal bone. This is adjacent to the anterior cranial fossa and frontal sinus. Any defect in the roof, will give a pulsatile proptosis due to the cerebrospinal fluid pulsation.

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- Medial wall—maxillary, lacrimal, ethmoid and sphenoid bone. The lamina papyracea separates the ethmoidal cells from the orbit and several nerves and blood vessels perforate through it and therefore, orbital cellulitis is frequently associated with ethmoidal sinusitis.
- Floor—zygomatic, maxillary and palatine bone. The posteromedial part of the maxillary bone is much thinner than the anterior part and therefore, is most commonly involved in "blowout" fractures. Inferiorly to the orbital floor is the maxillary sinus and thus, any carcinoma of the sinus will cause displacement of the orbit superiorly, rather proptosis.
- Lateral wall—greater wing of sphenoid and zygomatic bone. This is adjacent to the temporalis fossa and middle cranial fossa. Due to its configuration, the anterior half of the globe is protruding beyond the lateral orbital margin and thus vulnerable to lateral trauma.
- Orbital apex—Optic canal and superior orbital fissure. The optic canal transmits the optic nerve with its surrounding meninges and the ophthalmic artery with its sympathetic plexus. The orbital fissure is divided into superior and inferior. The superior orbital fissure is an opening of the lesser and the greater wing of sphenoid and it transmits lateral to medial the lacrimal, frontal and trochlear nerve (CN-IV) along with the upper and lower divisions of oculomotor nerve (CN-III), nasociliary nerve and abducent nerve (CN-VI). The superior ophthalmic vein passes through the lateral part of the fissure and connects to the cavernous sinus.

The walls of the orbit are surrounded by a loose periosteum called periorbita. It is continuous through the foramina of the skull and to the dura at the optic canal, superior orbital fissure and ethmoidal canal. Its firm points of attachment are at the cranial sutures and at the various foramina. At the orbital apex, the periorbita is thicker and forms the common tendinous ring, a fibrous ring that serves the common origin of the four recti muscles. The periorbita at the anterior part of the orbit gives rise to sheets that enter the eyelids and form the orbital septum. The septum is the foremost most significant anatomical landmarks as this separates the orbital contents from eyelid and works as a barrier to the spread of infections. Part of the periorbita also gives rise to the trochlea for the superior oblique tendon, encloses the lacrimal sac and also forms the periosteum of the nasolacrimal canal. The sensory innervation of the periorbita is through branches of the trigeminal nerve.

Retro bulbar masses tend to push the globe anteriorly unless they originate at the same plane as the globe (intraconal). In addition, in cases of penetrating trauma, the presence of fat at the wound indicates perforation of the orbital septum and is indicative of the severity of the injury as well as higher chance of complications associated with it, for example orbital hemorrhage or retained foreign body in the orbit or globe involvement.

70.3 Clinical Approach

70.3.1 Diagnosis

In cases like this it is important to assess the patient both from the ophthalmic side as well as systemically, since the proptosis might be a manifestation of an underlying disease.

As with any disease, a meticulous clinical history needs to be taken, as this will be an important aid in forming an accurate differential diagnosis. From the ophthalmic side of things, the rate and duration of proptosis is vital. Infective etiologies or post-traumatic will progress more rapidly than inflammatory. The presence of pain, changes in vision, diplopia or transient episodes of loss of vision should alarm the attending clinician as it warrants rapid intervention. Likewise, a systems review will highlight a systemic underlying pathology in cases where the orbital involvement is a secondary feature.

Clinical examination is the only way to confirm the presence of true proptosis versus the possibility of pseudoproptosis. High myopia, contralateral enophthalmos, shallow orbits, contralateral ptosis, upper lid retraction and buphthalmos (enlargement of the globe usually due to uncontrolled intraocular pressure from

birth onwards, causing stretching of the coats of the eye resulting in abnormal axial length of the globe and widening of the cornea) are few causes that can cause confusion to the treating physician. A good way to identify that is to review old photographs, as this will help identifying facial asymmetry or a change in appearance.

In cases of true proptosis, the patient can be examined by looking down with the physician looking down from behind in order to assess whether the corneal apex protrudes beyond the superior orbital rim. Objectively, it is measured with a Hertel exophthalmometer and the interlateral canthal distance together with the actual measurement of how much in millimeters the globe protrudes from the orbital rim are documented for monitoring purposes. A normal range would be between 22 and 24 mm but suspicion should be raised if there is a big asymmetry between the two eyes (>2 mm). Additionally, the measurements vary between male and females as well as races and also if the patient's eyes are not centrally aligned. Even though is the only quantitative method of measuring he proptosis, is highly variable between clinicians so attention to the examination technique is important.

Other aspects of the examination include: neck examination for lymph nodes or neck masses, optic nerve assessment [visual acuity, color vision, pupillary defects (including relative afferent pupillary defect) and confrontation visual fields], ocular motility and lid examination; as this can give clues of thyroid eye disease or inflammatory orbital disease, and cranial nerve examination as facial numbness can indicate a neuropathic spread of squamous cell carcinoma. In addition, an ophthalmic input is paramount and requires assessing the function of the eyelids and appearance of conjunctival vessels as "corkscrew" appearance combined with conjunctival hyperemia can indicate a carotid-cavernous fistula (CCF). Also, the ophthalmic specialist can assess the corneal integrity and sensation as well as the retina and optic nerve structure-function. Finally, the function of the extra ocular muscles needs to be performed (forced duction test) as this will provide important sings for restrictive versus neurogenic disease.

There are in addition, special maneuvers that need to perform as part of the examination. Retropulsion of the globe as well as auscultation can help when there is a suspicion of an orbital space occupying lesion for example encephalocele or a CCF or in the context of thyrotoxicosis. Furthermore, asking the patient to hold his breath on brace position and then sitting upwards or blowing through the end of a 2-milliliter syringe (Valsalva maneuver) will allow the examiner to elicit vascular lesions such as orbital varices. Any symptoms indicative of sinus disease warrant a detailed endoscopic examination by the ENT surgeon.

The causes of proptosis are a lot but a useful diagram (Fig. 70.1) aids to a differential diagnosis:

In addition to the clinical examination, specific laboratory studies can assist in forming a diagnosis. The number one diagnosis of unilateral or bilateral proptosis in an adult is thyroid eye disease, thus, a thyroid function panel including T3, anti-thyroid peroxidase antibodies and anti-thyroid stimulating hormone receptor antibodies are compulsory. Additional tests include complete blood count in case of infective causes or if the patient has signs of sepsis, then blood cultures must be obtained prior to starting broad spectrum antibiotics.

In any patient with proptosis where the underlying diagnosis is unclear, imaging is highly encouraged and needed for the diagnosis but also because it will assist in forming an appropriate management plan. Computerized tomography (CT) of the brain and orbits with thin "slices" of 1–3 mm will allow the evaluation of an orbital mass and it's association with bony structures (in case of bone invasion for example) and the surrounding tissues. When ordering CTs, it is highly advisable to use intravenous contrast (if permitted based on the renal function) as this will assist in highlighting inflammatory processes or tumors with high vascularity or vessel engorgement. In addition, CT will highlight calcified material in tumors much easily than Magnetic Resonance Imaging (MRI). Finally, CT venogram is the investigation of choice when CCF is suspected or any other pathology that potentially involves the cavernous sinus.

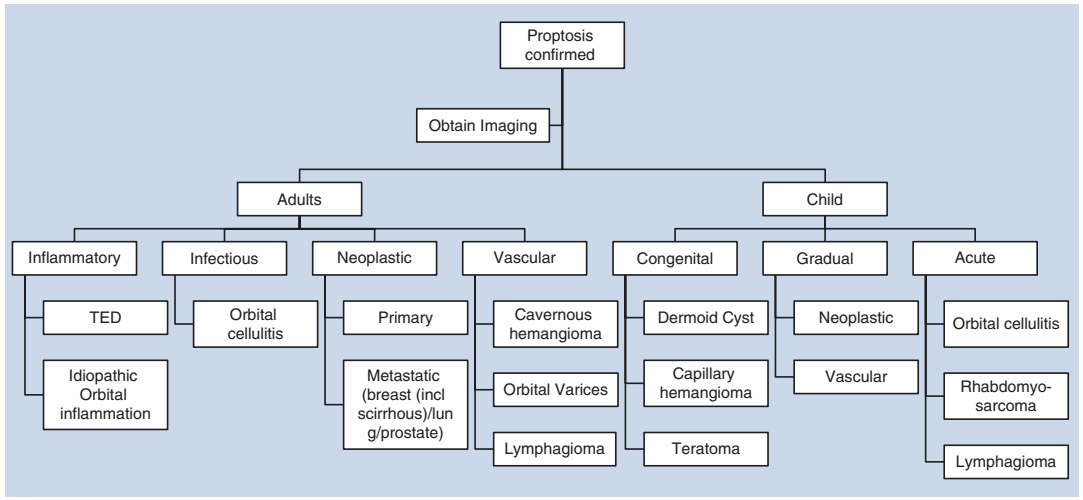

Fig. 70.1 Diagnostic approach to proptosis (adapted from Maguire et al.)

MRI is a better investigation for soft tissues and its resolution gives better anatomical information regarding the morphology of the extra-ocular muscles in case of thyroid eye disease, vascular lesions and the brain. It can also be used safely in children, as it has no radiation. Again it is used with the aid of intravenous contrast along with other sequences such as magnetic resonance angiography as it can help to understand vascular lesions or inflammatory lesions better.

Amongst the radiological investigations, orbital ultrasound offers an advantage is diagnosing vascular lesions such as in case of a dilated superior ophthalmic vein in cavernous sinus thrombosis or varices or lesions in the mid anterior half of the orbital cavity. Combined with Doppler it can provide information regarding flow of the suspicious lesion. Table 70.1 lists how common conditions appear on CT and MRI scan.

Table 70.1 Common orbital lesions and radiological correlation (Yanoff and Duker)

Most common orbital lesions which are well-circumscribed on CT/MRI	Most common orbital lesions which are ill-circumscribed on CT/MRI
Children	
Dermoid cyst	Capillary hemangioma
Lymphangioma	Orbital pseudotumor
Optic nerve glioma	Plexiform neurofibroma
Rhabdomyosarcoma	Leukemic infiltrate
Adults	
Cavernous hemangioma	Orbital pseudotumor
Neurofibroma	Leukemic infiltrate
Fibrous histiocytoma	Metastasis
Lymphoproliferative disorders	Lymphoproliferative disorders
Neurilemmoma	Primary malignant tumor

70.3.2 Management

The aim of the treatment approach is to either cure the underlying disease or establish a plan that ensures the patient remains symptom free and/or reduce co-morbidity.

Recurrent measurements of exophthalmos, visual and color acuities, pupillary responses and extra-ocular muscle motility along with visual fields (in case of thyroid eye disease) are paramount for the management of such patients.

Pharmacotherapy is the initial approach for most conditions and recent advances in the treatment especially of thyroid eye disease will be a game-changer in the near future.

Artificial tears act as humectants in the eye and in the recent decades there has been a large increase in the preparations available with various properties. Regardless the choice, the broad categories used are those in eye drops format (carboxymethylcellulose or hyaluronic acid or hypromellose based) or ointments (paraffin

based). The purpose of such medications is to decrease the discomfort that is caused by the dysfunction of the blinking reflex and tears distribution on the eye (see epiphora chapter). Lubricants are harmless and can be used in any case of exophthalmos.

In case of TED, both the ENT and ophthalmic physicians must be in co-ordination with the endocrinologist regarding controlling the thyroid abnormalities, as uncontrolled disease will inevitably make the eye condition worse and can even lead to loss of vision. TED can manifest in hyper-, hypo- and euthyroid states. The disease activity needs to be assessed and there are several grading systems for assessing TED activity (EUGOGO, CAS). In general, worsening score highlights the need for emergency treatment. In moderate or severe disease, intravenous pulsed steroids remain the mainstream treatment as their function has immediate effect. Orbital irradiation is an alternative treatment modality, though its effect takes a while to occur and therefore, it is not the treatment of choice for sight threatening disease. In sight threatening disease the role of emergency surgical decompression is important. In January 2020, the Food and Drug Administration (FDA) has recently approved the use of biologics for treating thyroid eye disease for Teprotumumab (Tepezza). These autoantibodies bind to the insulin-like growth factor 1 receptor (IGF-1R) that is overexpressed in orbital fibroblasts, B and T cells and thyrotropin receptor at the adipose cells and fibroblasts; which are increased in active thyroid eye disease. As a result, they attenuate the actions of IGF and thyroid stimulating hormone by blocking the production of pro-inflammatory cytokines and hence decreasing edema, adipogenesis and expansion of the orbital contents. Once the condition is stable, then surgery can be recommended on the basis of cosmesis or debilitating diplopia or corneal exposure. The order in which surgery is offered is: Orbital decompression (2,3 or 4 wall decompression) followed by strabismus surgery followed by eyelid surgery. The rationale for such approach is that decompression surgery will alter the strabismus and affect the eyelid position. Thus, not all patients require all the three types of surgery.

Orbital cellulitis is a potentially life threatening condition which requires early recognition and escalation along the systemic sepsis treatment protocol. After confirming the diagnosis (see above), the patient is admitted for broad-spectrum antibiotics (common organisms are Staphyloccoccus aureus and Streptococcus pneumoniae) and joint care of the ENT surgeon and Ophthalmologist. ENT's involvement is in the management of subperiosteal abscess or drainage of sinus infection (ethmoidal sinus is the commonest) while the ophthalmologist ensures the good ocular function, initially with intense monitoring of the visual and optic nerve function and depending on progression, it becomes less frequent. Any sings of concern or deterioration, prompts for emergency surgery.

Likewise, orbital (retro bulbar) haemorrhage consists of another ocular/orbital emergency. Usually the presentation is part of a poly-trauma or extensive facial trauma and thus any emergency physician should be taught to manage it at the initial stages. The triad of painful proptosis—tense orbit associated with loss of ocular function (vision, pupillary reflexes and restricted motility) in the context of trauma should alert the treating clinician and lateral canthotomy with inferior cantholysis and maybe superior cantholysis should be performed. This is performed before the patient undergoes CT, as delay in treatment can be catastrophic. If however, vision is not threatened then hyperosmotic agents such as intravenous mannitol 20% 1 g/kg over 45 min or acetazolamide 500 mg and methylprednisolone 100 mg for neuroprotection can be given.

The steps of lateral canthotomy with cantholysis are highlighted below:

1. Infiltrate anesthesia into the lateral canthus and lateral portion involving the eyelid.
2. An artery clip is used to crush the lateral canthal angle as this minimizes the bleeding.
3. Use Stevens scissors (or any big scissors) to cut the lateral canthus for approximately 5–7 mm (canthotomy).
4. Grasp the lower eyelid near the lateral canthal angle with toothed forceps and pull slightly medially to place traction. A tight band is felt

between the orbicularis and the conjunctiva, inferior to the lower edge of the incision.
5. Strum the canthal tendon with the Stevens scissors and cut inferiorly through the tendon avoiding the conjunctiva if possible (inferior cantholysis).
6. Full release of the inferior cru of the tendon will release the lower eyelid, which will be now freed from its attachment.
7. A gush of blood will be released from the corner and the orbit will become less tense, with progressive resolution of symptoms and recovery of the pupillary reflexes.
8. Repeat this process for the superior cru of the tendon, if the orbital tension remains and hasn't relieved haemorrhage.

Systemic vasculitis (granulomatosis with polyangiitis) or idiopathic orbital inflammation (IOI) can manifest with either unilateral but most commonly bilateral proptosis. Initially oral non-steroidal anti-inflammatory medications can be used however; their role tends to be adjuvant to high dose steroids, which are tapered over months until resolution of symptoms. If rebound symptoms occur, either they require longer tapering or systemic immune suppression. It is important to acquire tissue biopsy prior initiating immune suppressants if possible, and if it is not going to cause significant injury to the surrounding healthy tissues. In steroid-resistant cases or those patients that steroids are not tolerated, orbital radiotherapy can be used as an alternate approach.

In proptosis related to vascular abnormalities (varices, cavernous hemangioma, lymphangioma), a surgical intervention is only required if there is compromise of visual function (optic nerve decompression; Table 70.2) or unacceptable cosmetic changes. In the majority of cases, conservative treatment with annual or biannual visual function assessment is good practice. Hemangiopericytomas are generally undergoing a wide surgical excision with adjuvant radiotherapy, if incompletely excised since it can undergo malignant transformation, while arteriovenous malformations are either undergoing embolization therapy (interventional radiology) or can be surgically excised.

Table 70.2 Indications and surgical steps of optic nerve decompression (Custer)

Optic nerve decompression[a]
• Indications – Traumatic neuropathy – Thyroid eye disease associated with optic neuropathy – Vision loss due to idiopathic ICH – Fibro-osseous lesions – Neoplasms – Mucoceles – Pseudotumour cerebri – Ischaemic optic neuropathy – Acute optic neuropathy associated with acute retinal necrosis syndrome – Optic nerve meningioma – Osteopetrosis
• Determination of potential benefit is strongly associated with the cause. Causes of complete disruption of the optic nerve will not recover regardless surgical decompression, while cases secondary to oedema, haematoma or moderate bony compression may respond favourably to decompression.
• Technique – Ethmoidectomy and wide sphenoid antrostomy – Occasionally a four-hand technique is required and, in that case, a posterior septectomy is necessary – Identification of the lamina – Removal of the lamina papyracea 0.5–1 cm anterior to the optic nerve tubercle – If the bone is thick, eggshell with diamond burr – Decompress medially 180° – Beware of infero-medial course of ophthalmic artery (15%) – Incise sheath and annulus of Zinn if necessary → supero-medially. We avoid incising the sheath in cases of fungal sphenoiditis to avoid any intracranial complications (Fig. 70.2). – Optic nerve decompression and orbital decompression can be done simultaneously on indicated cases

[a]Technique can vary between surgeons

The choice between medical and surgical treatment of primary orbital tumors or metastatic tumors depends on the age of presentation along with the type of tumor. The treatment decision is usually referred to a cancer multidisciplinary team meeting (MDT) as often multiple specialties are involved. In children, orbital rhabdomyosarcoma is the commonest malignant orbital tumor and early resection with adjuvant chemo/radiotherapy is indicated, according to the Intergroup

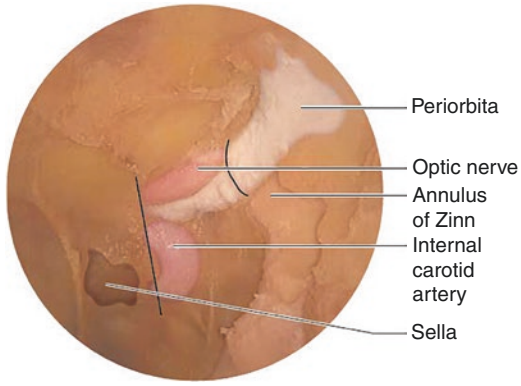

Fig. 70.2 Cadaveric dissection of the optic nerve in the sphenoid sinus

Rhabdomyosarcoma Study Group. In adults, optic nerve or sphenoid wing meningiomas tend to be observed or can be surgically excised if there are concerns regarding optic nerve involvement. Optic nerve gliomas, which are commonly associated with Neurofibromatosis type I require excision. In metastatic disease, as the tumor tends to be invasive, usually chemotherapy combined with external beam radiotherapy and in rare occasions wide excision or exenteration of the orbital contents consists of the management approach. Lastly, the treatment approach for lacrimal gland tumors (pleomorphic adenoma or adenocarcinomas, adenoid cystic carcinomas) depends on the radiological appearance (pleomorphic adenoma is the only one that appears well-circumscribed on CT) and a complete en bloc removal of the gland (pleomorphic adenoma) or radical orbitectomy with any other bone involved or with exenteration followed by adjuvant chemo/radio-therapy is the treatment of choice (pleomorphic adenocarcinoma or adenoid cystic carcinoma or metastasis).

Summary and Author's Comments

1. Globe proptosis is a clinical sign that is the presenting sign of a long list of causes
2. The number one cause of unilateral of bilateral proptosis in adults is TED and in children is orbital cellulitis
3. Onset, duration and speed of evolution can direct the clinician to the underlying cause
4. Painful proptosis is more associated with inflammatory conditions while painless is characteristic of neoplastic conditions
5. In most cases, a multidisciplinary approach is required for the interpretation of the diagnostic tests as well as for treatment planning.

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Part XI

General Rhinology

Septoplasty: Standard Approach

71

Sarantis Blioskas

A 45-year-old Caucasian male was referred to the ENT Department by his GP complaining about bilateral nasal blockage, significantly worse on the left side that dated back more than 10 years before referral. The patient did not report significant nasal discharge, headaches or facial pain, and no decline in his sense of smell. He did recall having sustained nasal trauma during late childhood, which was never investigated or treated, yet he did not report any link between that and his nasal blockage. On examination, there was no significant alar collapse and the Cottle sign was positive. Anterior rhinoscopy revealed a "C" shaped deviation of the nasal septum towards the left side, with compensatory hypertrophy of the right inferior turbinate (Figs. 71.1 and 71.2). Flexible nasendoscopy confirmed the above-mentioned findings. The patient was given the option of surgery and after consent was obtained, he underwent a septoplasty under general anesthesia. The cartilaginous nasal septum was realigned to the midline by a combination of submucosal cartilage resection and repositioning, as well as scoring of cartilage on the convex side. Quilting sutures and bilateral absorbable packing were utilized to avoid septal hematoma.

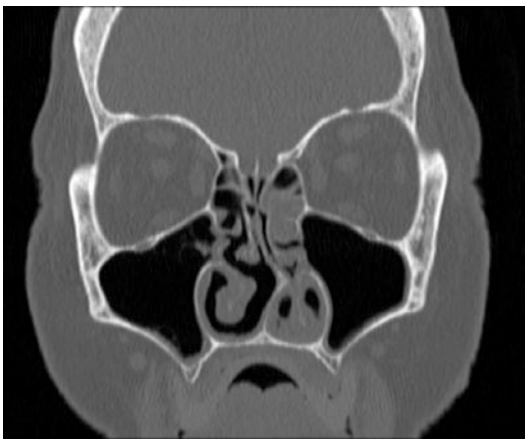

Fig. 71.1 CT of the paranasal sinuses (coronal) demonstrating septal deviation to the left resulting in nasal obstruction and hypertrophy of the right inferior turbinate

The patient was followed up 2 months after the procedure and he reported a significant improvement of his nasal airway.

71.1 Background Knowledge

To comprehend the rationale of septoplasty, one should obtain a sound knowledge of relevant clinical anatomy. The nasal septum consists of the cartilaginous septum, the perpendicular plate of the ethmoid bone, the vomer and the structures that constitute the septal framework.

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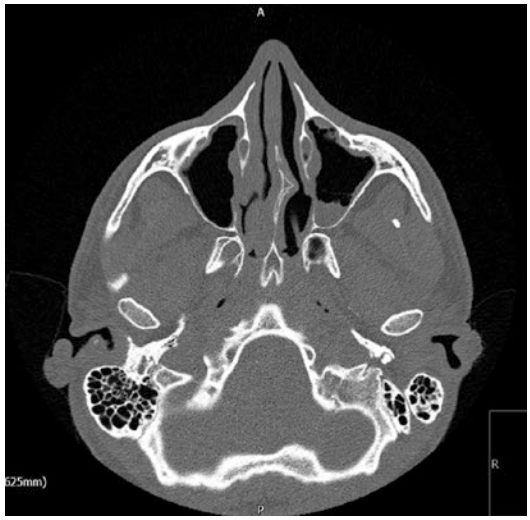

Fig. 71.2 CT of the paranasal sinuses (axial) of the same patient

The cartilaginous septum consists of the quadrangular cartilage. From a surgical point, critical elements to be identified are its caudal end and the processus lateralis ventralis, which is a broadened area where the cartilage is connected to the premaxilla, as well as the sphenoidal process which is an extension of the cartilaginous septum posteriorly between the perpendicular plate and the vomer.

The perpendicular plate of the ethmoid bone is a bony plate superiorly attached anteriorly to posteriorly to the nasal spine of the frontal bone, the inferior surface of the cribriform plate and posteriorly to sphenoidal crest and vomer.

The vomer is a quadrangular bone posteriorly attached to the sphenoid bone. Its posterior margin forms the medial wall of the choanae and its inferior margin is connected to the maxillary and palatine crests.

The "surrounding structures" of the nasal septum constitute the septal framework. They are the anterior nasal spine, the maxillary crest and palatinal crests and the premaxilla with the premaxillary table and wings. These structures are joined with the nasal septum and the understanding of these connections is of the utmost importance in septal surgery.

The nasal septum is covered with mucoperichondrium and mucoperiosteum (for cartilaginous and bony parts, respectively). The mucoperichondrium consists of the epithelium, the lamina propria and the perichondrium, with the latter consisting of two layers, an outer layer of loose fibers and an inner layer of densely packed fibres adjacent to the cartilage. Elevating the mucoperichondrial (and mucoperiosteal) flap in the proper plane—that is, under the inner perichondrial layer—during septoplasty is a critical step of the procedure, since it ensures significantly less intraoperative bleeding and minimizes the risk of tears created at the elevated flaps.

71.2 Clinical Approach

The deviation of the nasal septum is far from being rare. Ideally, nasal septum would be a completely straight midline structure, yet this is hardly the case. Most individuals suffer from some degree of deviation or curvature of the nasal septum, but that should always be differentiated from patients suffering from symptoms due to their deviation of the nasal septum.

The deviation of the nasal septum has been recognized as a source of a variety of problems. Nasal obstruction is the predominant feature for which patients with deviated septum usually seek medical help. Nasal septal irregularities can also result in an aesthetic compromise of the nasal shape. Overall, septal deviation becomes clinically significant when it results in functional or aesthetic morbidity.

The etiology of septal deviation is often congenital, although it could also be attributed to nasal trauma or iatrogenic causes. Diagnosis is usually achieved through psimple anterior rhinoscopy. Nasendoscopy can improve diagnostic accuracy and depict posterior spurs or deviations that can be overlooked with anterior rhinoscopy. Imaging (in the form of a CT scan of the paranasal sinuses) is rarely necessary and may be considered if sinonasal pathology is investigated or if there are concerns about the integrity of the keystone area. Finally, rhinomanometry and acoustic rhinometry can provide a detailed outline of the points of maximal nasal obstruction and effectively differentiate patients most likely to be improved by a septoplasty.

Septoplasty, with or without turbinate reduction, is the most commonly performed surgical

procedure to address persistent nasal obstruction. Although septoplasty is not a standard procedure since it involves a variety of possible techniques, and it is every time tailored to the individual patient's needs, it generally revolves around a few core principles:

- Submucosal resection of bony or cartilaginous deviations with care to identify a precise subperichondrial pocket and maintain this plane of dissection is of utmost importance. It remains a gold standard approach since advocated by Killian and Freer more than a century ago.
- Hemitransfixion incision and the approach described by Cottle and Loring has displaced Killian's incision as for the gold standard (Fig. 71.3).

- The significance of addressing all deviated areas of both the bony and the cartilaginous septum to achieve maximal improvement in airway patency is clear.
- Even though all deviated portions should ideally be addressed, the nasal valve area bounded by the nasal septum, the upper lateral cartilage, and the anterior surface of the inferior turbinate, is of paramount importance in improving nasal obstruction and special attention should be drawn in maximizing patency.
- Septoplasty is not a procedure of "resecting" but a procedure of "reshaping." This principle suggests that ideally, all excised cartilage and bone should be trimmed or crashed, reshaped and replaced between the raised mucoperichondrial flaps.
- Any surgical resection of the bony and cartilaginous parts of the septum should not compromise nasal stability. Adhering to the "L" strut principle (sparing 1–1.5 cm struts at the caudal and dorsal septal areas) avoids complications like tip collapse and saddle nose deformity. It ensures that there will be no significant unwanted aesthetic alterations of the external nose. The "keystone area," which is the junction between the dorsal part of the cartilaginous septum and the nasal bone, bony septum, and upper lateral cartilages, is crucial at providing support and destabilization should be avoided.
- No single method of correction can be effective in treating all cases. There is a large variety of different techniques in correcting the deviated septal parts, including cartilage scoring, cartilage disarticulation ("swinging door"), spur resection, suture placement, etc. There are also specific techniques to address particular problems like caudal deviations ("tongue in grove," columella pocket, columellar graft, etc.), dorsal deviations (spreader grafts, freeing the dorsal septum from upper lateral cartilages extracorporeal techniques, etc.). A detailed description of these techniques is beyond the scope of this chapter. It should be emphasized, though, that a surgeon should be familiar with the majority of these techniques to be able each time to tailor his approach to treat the specific problems appropriately.
- Most methods can be approached through an endonasal approach. Yet complex deviations may sometimes require an external approach (open rhinoplasty approach).
- We can avoid septal hematoma formation by using nasal packing (dissolvable or not) or "quilting sutures." Dissolvable nasal packing or sutures are well tolerated by the patients and this is our recommended practice.

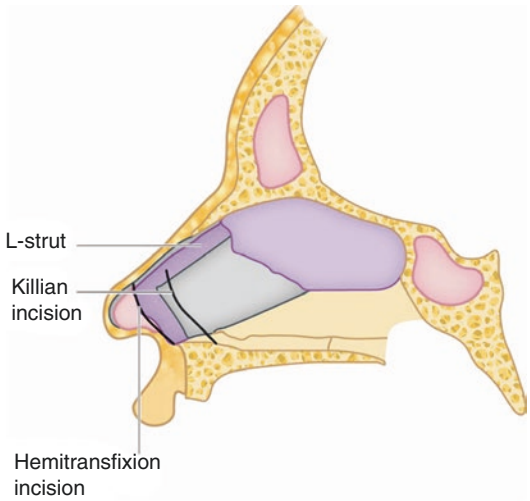

Fig. 71.3 Hemitransfixion and Killian's incisions. The cartilaginous L-strut should be preserved to maintain adequate support

Despite septoplasty being one of the most common ENT operations in adults, its effectiveness remains a matter of debate in terms of evidence-based medicine. The level and quality of data are far from being convincing. The need for further evidence regarding the benefit of the intervention has been widely declared; nevertheless, the scarcity of evidence has not hampered the widespread practice of septoplasty as a routine procedure to treat nasal blockage.

In light of the lack of objective evidence, one should always be reminded of potential risks and complications of the procedure. Apart from the failure to achieve the intended results, a patient undergoing septoplasty may encounter postoperative bleeding (being the most common complication), as well as septal hematoma, septal perforation, hyposmia, infection/septal abscess, postoperative pain and/or numbness, adhesions and even temporary reduced visual acuity. Equally important, nasal deformity (saddle nose or collapsed tip) and consequent psychological implications, can result in a severe decline in patient's quality of life.

Summary and Author's Comments

1. To comprehend the rationale of septoplasty, one should obtain a sound knowledge of the relevant clinical anatomy.
2. Most individuals suffer from some degree of deviation or curvature of the nasal septum, yet that should always be differentiated from patients suffering from symptoms due to their deviation of the nasal septum.
3. Diagnosis is usually achieved through plain anterior rhinoscopy, yet nasal endoscopy can improve diagnostic accuracy. Rhinomanometry and acoustic rhinometry can provide a detailed outline of the points of maximal nasal obstruction.
4. Septoplasty involves a variety of possible techniques, yet it generally revolves around a few core principles.
5. The current body of evidence does not support firm conclusions on the overall effectiveness of septoplasty.

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Septoplasty: Endoscopic Approach

72

Sarantis Blioskas

A 32-year-old female was referred to the ENT Department by her GP complaining about bilateral nasal blockage, occasional clear nasal discharge, and postnasal drip. The patient had tried courses of intranasal steroids and oral antibiotics prescribed by her GP, which led to partial and temporary relief of symptomatology, and she was thus referred for further care. On examination, anterior rhinoscopy and flexible nasendoscopy revealed bilateral grade III polyps and a septal spur narrowing the right middle meatus. A CT scan of the sinuses confirmed the presence of chronic rhinosinusitis with polyps, as well as a septal deviation towards the right side. Patient was given maximal medical therapy, and she was followed up 1 month after completion. She reported a recurrence of symptoms after treatment, and in the light of that, she was given the option of functional endoscopic sinus surgery and endoscopic septoplasty. All the risks and complications were discussed and the patient consented for surgery. The option of an endoscopic septoplasty was preferred compared to a traditional septoplasty, in the context of simultaneous endoscopic sinus surgery. The patient was followed up 6 weeks after the procedure and she reported a significant improvement regarding her symptoms of nasal blockage, postnasal space drip and loss of smell. She was postoperatively prescribed with long term intranasal steroids and nasal douching.

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72.1 Background Knowledge

Deviated nasal septum is one of the most common causes of nasal obstruction, and its surgical correction through septoplasty represents a widespread routine practice. Despite that traditional approaches being the gold standard, the advent of endoscopic techniques has revolutionized nasal surgery in general. Thus, despite their main indication still lies in paranasal surgery, endoscopic approaches for correcting septal deviations alone or as an adjunct to functional endoscopic sinus surgery are gaining in popularity.

Endoscopic septoplasty was first introduced in 1991 by the pioneering work of Lanza and Stammberger and since then multiple endoscopic techniques, each with its advantages and limitations, have been proposed.

Traditional septoplasty is usually termed "headlight" septoplasty and it is performed under direct visualization using a nasal speculum. Thus there are inherent limitations regarding visualization, particularly evident when addressing irregularities of the posterior septum due to narrow field of view and limited illumination. Furthermore, the comparative relationship between the lateral nasal wall structures and the

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deviated nasal septum is not always easily determined because of the distortion of normal nasal anatomy by the insertion of the nasal speculum. Some of these drawbacks are effectively bypassed with the use of endoscopes.

72.2 Clinical Approach

Endoscopic septoplasty is gaining popularity because of certain distinct advantages. The main benefit, as in all endoscopic procedures, is the better visualization of septal deformities. This improvement in visualization is accomplished through the magnification of structures, improved illumination, and better access to narrow areas. Due to these unique characteristics, the endoscopic approach allows more conservative and less invasive surgery, thus minimizing potential adverse outcomes. In this concept, dissection of mucoperichondrial and mucoperiosteal flaps is done under constant and close visualization and mucosal disruptions are recognized immediately, and their size is limited by meticulous dissection.

A distinct advantage of endoscopic septoplasty is its inherent ability to transmit the intraoperative image to a monitor that makes it readily available to larger audiences, including trainees and/or medical students, as well as enables supervisors to inspect the procedure when undertaken by residents closely. Thus, it represents an excellent teaching tool. This is particularly important because the learning curve of the procedure was traditionally long due to limited visual access to trainees.

Finally, endoscopic septoplasty utilizes the same standard instrumentation used for functional endoscopic sinus surgery. As the two procedures usually combine, as access to the middle meatus can be limited by septal deviations, endoscopic septoplasty performed in conjunction and before endoscopic sinus surgery improves surgical outcome with minimal time consumption.

Regarding surgical techniques, most of the same core principles established for traditional septoplasty still apply for the endoscopic approach as well. Submucosal dissection in a sub-mucoperichondrial plane remains the way to go. However, the hemitransfixion incision is rarely utilised and usually replaced by a Killian incision directly anterior of the deviation to be corrected. The reason for that is that the inherent disadvantage of poor access is balanced by the increased visualization provided by the endoscope. Once the flaps are raised, deviated parts of the cartilaginous and bony septum can be resected (and ideally reinserted) using a Jansen-Middleton punch in a twisting motion. The use of powered instrumentation, which is routinely available during functional endoscopic sinus surgery, could also be considered. Adequacy of the surgical correction can be assessed by returning the mucosal flaps to the midline and inspecting the nasal airway bilaterally while palpating areas of residual deviation (Figs. 72.1, 72.2, 72.3, 72.4, 72.5, and 72.6).

A special technique made possible through the endoscopic approach, in particular, is the notion of "directed septoplasty". This modification is particularly useful for addressing isolated septal deformities (e.g., spurs) in the absence of greater deviations. It involves a horizontal incision under endoscopic visualization, directly on the apex of the spur to be addressed and elevation of a superior and an inferior mucosal flap. The

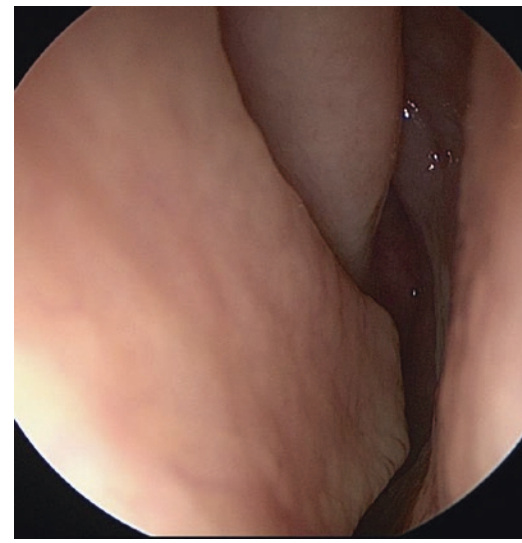

Fig. 72.1 Endoscopic view of the septal deviation

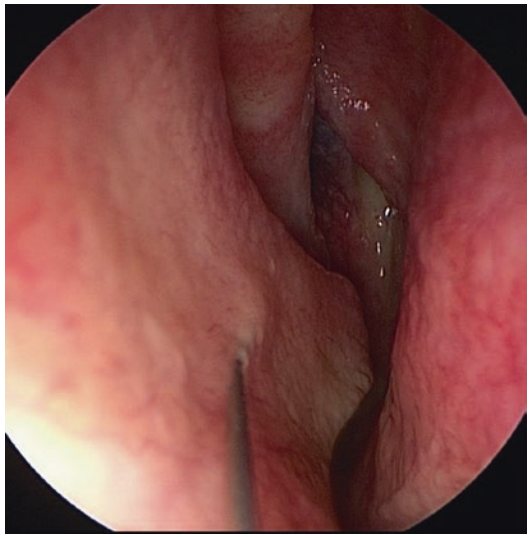

Fig. 72.2 Infiltration with local anaesthetic

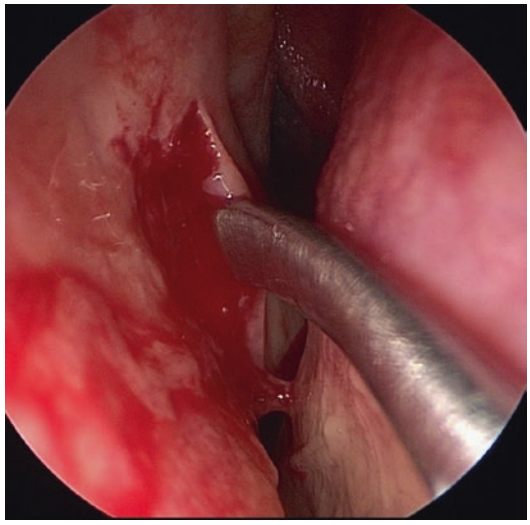

Fig. 72.3 Killian's incision and elevation of the mucoperichondrial flap

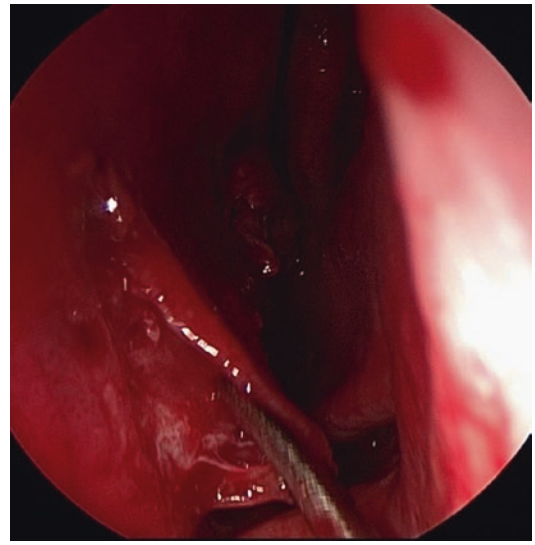

Fig. 72.4 Elevation of the flap, chondrotomies and dissection on the contralateral side

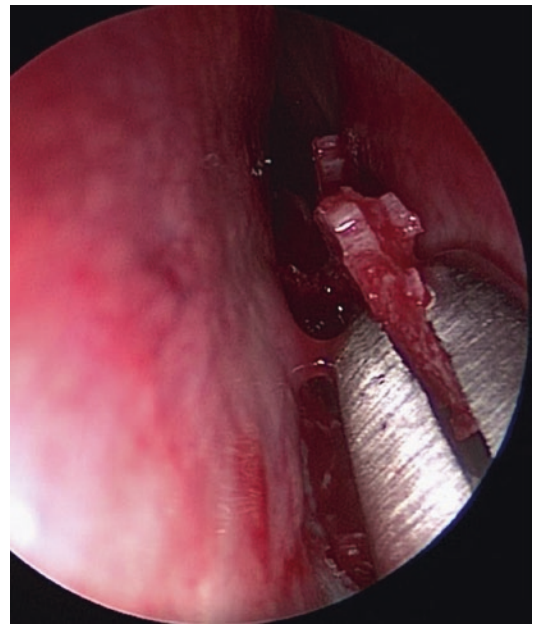

Fig. 72.5 Removal of the deviated part of the septum

deformity is excised through powered instrumentation or through-cutting instruments and the flaps are redraped. Directed septoplasty results in very limited dissection, which ensures quicker postoperative healing, fewer complications, and less operative time.

Furthermore, ease of dissection is facilitated by avoidance of the anterior decussating fibers. In the same concept "directed septoplasty" can include a posteriorly placed incision, directly anteriorly to the focal deviation to be addressed, achieving the same advantages. Finally, the same overall notion of a minimally invasive approach can be implemented in treating septal perforations.

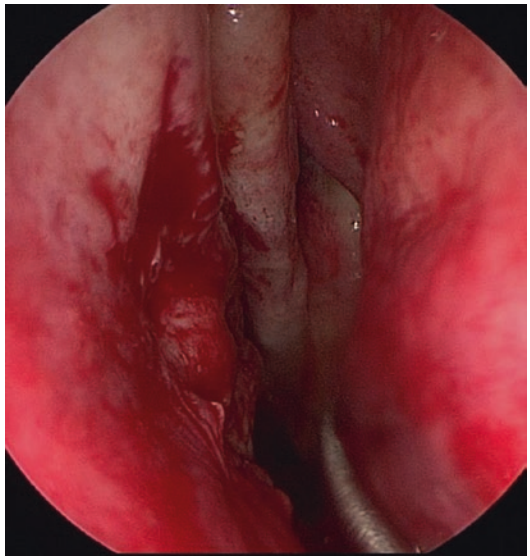

Fig. 72.6 The mucosal flap is put back in place and the incision can be closed with absorbable sutures

The main issue to be evaluated in terms of evidence-based medicine is the comparison between endoscopic and traditional septoplasty. In terms of clinical outcomes, although there is generally a scarcity of data, yet literature seems to suggest that endoscopic septoplasty is compared favorably to the traditional approach. Chung et al. in one of the largest series sizes published in the literature reported complete resolution of nasal obstruction in 70% of 112 endoscopic septoplasty patients with an additional 20% reporting partial improvement over a 13-month mean follow-up. Other authors also reported favorable outcomes. Apart from evaluating the efficiency of the endoscopic septoplasty alone, studies comparing the outcomes of the two approaches are of utmost importance. Such reports have shown similar patient outcomes, surgical results, and comparable operative times.

In terms of postoperative complications, these are the same for endoscopic and "headlight" septoplasty and include epistaxis, scarring and crusting, septal hematoma, septal perforation, synechiae formation, dental numbness, cerebrospinal fluid leak, and persistent nasal obstruction due to residual septal deviation. In one of the largest series published, 4.3% of patients reported transient dental pain or hypesthesia, while the incidence of epistaxis or septal hematoma was less than 1%, the frequency of septal perforation was 3.4% and synechiae formation reached 2.6%. Similar results were reported by other authors and these rates are comparable with those reported in the literature for traditional headlight septoplasty.

The wealth of studies that describe individual experiences with each technique, as well as the numerous studies that compare clinical outcomes, attest to the lack of management consensus today. If we were to undertake an effort to highlight indications and contraindications for endoscopic septoplasty, relative indications would include more posterior deviations of the septum without the involvement of the caudal septum, isolated focal spurs, revision cases, septal perforations and procedures undertaken in conjunction with endoscopic sinus surgery when improved access to the sinuses is necessary. On the other hand, contraindications to endoscopic septoplasty would include conditions necessitating an open septorhinoplasty approach (e.g., dorsal deviations and crooked nose), and severe caudal deflections. The rationale behind these contraindications is that the comparative advantage of increased visualization via the use of endoscopes is diminished if an open approach is used, as the latter provides adequate access and visualization through a "headlight" approach as well. Moreover, challenges in controlling the endoscope when operating in the caudal septal area (that is too anteriorly) can pose a significant disadvantage.

Ultimately, surgeons should be trained to both techniques in order to take advantage of the unique benefits offered by each approach and decide on the approach based on the location and severity of the septal deformity and the individual patient's needs. Additionally, a "combined approach" using both endoscopes and direct visualization should always remain in their surgical armamentarium.

Summary and Author's Comments

1. Despite that traditional approaches being the gold standard, the advent of endoscopic techniques has revolutionized nasal surgery.
2. The main advantage, as in all endoscopic procedures, is the better visualization of septal deformities.
3. Most of the same core principles established for traditional septoplasty still apply for the endoscopic approach.
4. Relative indications for endoscopic septoplasty include posterior deviations, isolated focal spurs, revision cases, septal perforations and endoscopic sinus surgery when improved access to the sinuses is necessary. Contraindications include conditions necessitating an open septorhinoplasty approach and severe caudal deflections.

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Turbinate Hypertrophy: Turbinate Surgery

73

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A 26-year-old female was referred to the ENT Department by her GP complaining about bilateral nasal blockage, notably worse in the course of occasional viral infections, constant clear nasal discharge and occasional sneezing. The patient had tried courses of intranasal steroids and oral antihistamines prescribed by her GP, which improved her nasal discharge and sneezing; however, her nasal blockage was not improved significantly and the patient was experiencing troubled sleep and declination in her overall wellbeing and quality of life. On examination, anterior rhinoscopy and flexible nasendoscopy demonstrated evidence of allergic rhinitis and significant hypertrophy of inferior turbinates bilaterally. The septum was not deviated and there was no evidence of alar collapse. Skin prick testing was undertaken and depicted allergy to house dust mites. Given that the patient has already tried several courses of antiallergic medication, with minimal improvement regarding her nasal blockage, she was offered the option of a turbinate reduction under general anesthesia. Superficial linear diathermy of the inferior turbinates bilaterally was performed, utilizing a monopolar electrocautery needle (20 W). No nasal packing was necessary and a chlorhexidine dihydrochloride and neomycin sulfate nasal ointment and a salty water spray were prescribed for postoperative treatment. The patient was reviewed 6 weeks following treatment and she reported a significant improvement regarding her nasal blockage and sleep quality.

73.1 Background Knowledge

The nasal turbinates (or conchae) are three bony shelves (inferior, middle, and superior turbinate) that project from the lateral wall of the nasal cavity. Each of these curved bones histologically comprises three layers: mucosa, lamina propria and bone. Pseudostratified ciliated columnar respiratory epithelium covers mucosal surfaces, yet the anterior edge of the inferior turbinate, in particular, is covered by stratified squamous epithelium. The ciliated epithelium is continually beating providing this way constant propelling of the mucous blanket from the front of the nasal cavity toward the nasopharynx, ensuring cleaning and filtering of the upper respiratory tract. Furthermore, the turbinates maximize the effective intranasal surface area for humidification and warming of inspired air.

The lamina propria contains loose connective tissue and a complex array of arteries, veins, and venous sinusoids, which overall resemble erectile tissue. Blood circulation in this rich submucosal cavernous plexus and multiple arteriovenous

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anastomoses creates a cyclic alternating constriction and dilation of the turbinate vasculature and consequently congestion and decongestion of the turbinate erectile tissue every 2 to 7 hours, known as the nasal cycle. This cycle is dictated by autonomic fibers from nerves originating from the pterygopalatine ganglion. Thus, generally decreased sympathetic activity or increased parasympathetic activity, resulting in nasal congestion and obstruction.

Hypertrophy of the turbinates is one of the most common causes of nasal obstruction, to the point that up to 20% of the population can suffer from chronic nasal obstruction secondary to turbinate hypertrophy (Fig. 73.1). The inferior turbinate is thought to play a significant role in the overall development of nasal obstruction since its anterior portion lies in the nasal valve area, which provides approximately 50% of total airway resistance; thus, even minor engorgement in inferior turbinate's erectile tissue can result in significant resistance.

On the other hand, the role of turbinates in nasal obstruction apart from mechanical can also be functional. Thus, low contact of nasally inhaled air with the turbinate mucous membrane can result in the subjective perception of obstruction, even in the presence of a normally patent airway. As a result, preservation of normal mucosal function when attempting to address turbinate hypertrophy surgically is of paramount importance.

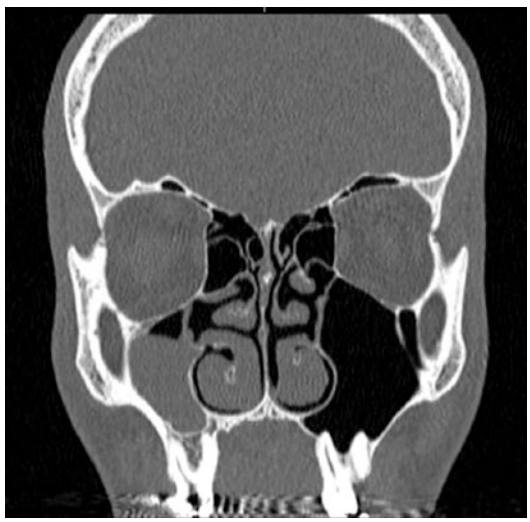

Fig. 73.1 Bilateral inferior turbinate hypertrophy

73.2 Clinical Approach

There are various causes of turbinate hypertrophy that can include conditions that cause permanent turbinate hypertrophy producing chronic nasal obstruction, such as allergic rhinitis, vasomotor rhinitis, or chronic hypertrophic rhinitis. Compensatory hypertrophy can also be seen in the setting of nasal septal deviation, particularly on the opposite side of the convex deviation. Upper respiratory infections and drugs or hormones may also induce turbinate dysfunction, usually intermittent or temporary. Similarly, turbinate engorgement in the recumbent position tends to manifest as nocturnal congestion that alternates sides, especially in the older population. Finally, anatomic bony turbinate enlargement due to progressive ossification throughout adulthood has been mentioned as a rarer form of turbinate hypertrophy.

In terms of diagnosis, turbinate hypertrophy is usually easy to diagnose through anterior rhinoscopy and nasal endoscopy. Acoustic rhinomanometry provides an objective means of assessing nasal obstruction and particularly nasal valve function. It should, however, be born in mind that turbinate hypertrophy is rarely an isolated condition, so diagnosis most commonly involves diagnosing the underlying disease, causing turbinate hypertrophy, such as allergic and non-allergic rhinitis, vasomotor rhinitis, chronic hypertrophic rhinitis, etc.

First-line treatment for turbinate hypertrophy involves conservative medical management, which should be attempted first, if not contraindicated, before surgical intervention. Nasal blood vessels are extremely sensitive to sympathomimetic medications; thus the topical implementation of alpha receptor agonists such as oxymetazoline and phenylephrine can treat short term obstruction, however, rebound vasodilation in the form of rhinitis medicamentosa after repeated application should be taken into account. Histamine is a potent vasodilator in the nose, so

antihistamines acting on the H1 receptors in the nose prevent the binding of histamine and, therefore dilation of the venous sinuses. Leukotriene receptor antagonists and intranasal steroids are also considered cornerstones of medical management. However, if medical therapy fails to alleviate obstructive symptoms, surgical intervention should attempt to reduce turbinates size while maintaining normal nasal physiology.

Although there are many options for turbinate reduction, there is no consensus on the best and most effective technique. Alternatives include office procedures for decreasing the bulk and thickness of the submucosal tissue of the inferior turbinate, as well as turbinate surgery that is usually undertaken under general anesthesia, and in some instances, it can include bony removal as well, to treat potential bone hypertrophy.

- Electrocautery (Diathermy). Electrocautery can be applied either directly on the mucosa or in a submucosal plane. Linear diathermy (usually referred as DITS (Diathermy of Inferior Turbinates)) involves a monopolar diathermy needle used in a linear fashion on the turbinate mucosal surface, whereas submucosal turbinate cauterization could mean either a monopolar or bipolar probe electrode, inserted submucosally via a stab incision on the anterior turbinate edge and advanced all through the posterior edge. Submucosal diathermy aims at inducing a reduction in volume through fibrosis and wound contracture while preserving turbinate mucosa. Nevertheless, as electrocautery can produce very high temperatures approaching 800 °C, submucosal thermal damage can potentially compromise turbinate vasculature and place turbinate at risk of severe ischemia. Linear diathermy, on the contrary, though advantageous in that perspective, does, however, carry a greater risk regarding damage to surrounding mucosa and decreased mucociliary clearance (Figs. 73.2, 73.3, and 73.4).
- Radiofrequency Ablation. The method lies in inducing tissue reduction via ion agitation and increase of tissue temperature, as well as tissue vaporization, through local submucosal delivery of low-frequency energy. A specially adapted electrode is inserted into the turbinate at one or more points and energy is delivered in the deep mucosa causing thermal lesions by rising tissue temperature at a controlled fashion up to 60 °C to 90 °C to prevent surrounding tissue damage. As in electrocautery, fibrosis and wound contraction leads to tissue volume reduction, yet in a more precise and targeted manner. In the same concept, coblation is a form of radiofrequency ablation that uses a plasma field created by the radiofrequency current generated between bipolar

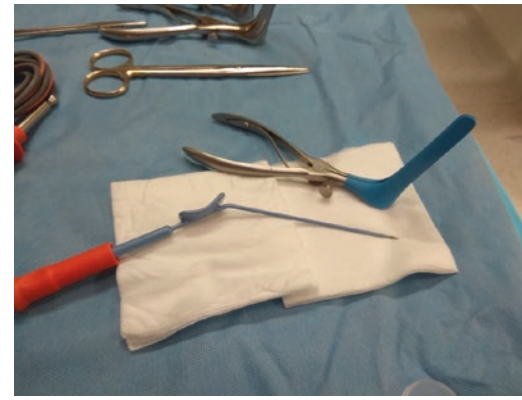

Fig. 73.2 Abbey diathermy needle and insulated Killian;s speculum

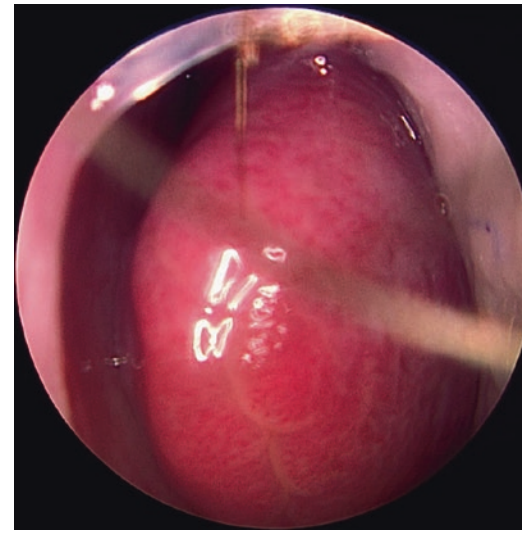

Fig. 73.3 Endoscopic view of hypertrophic inferior turbinate

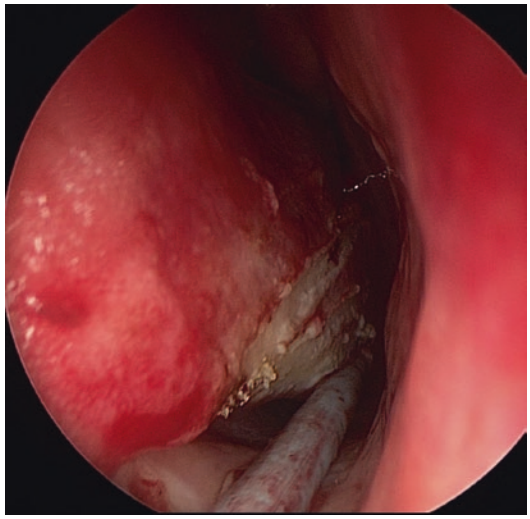

Fig. 73.4 Linear cautery for turbinate reduction

electrodes to ablate soft tissues with minimal thermal damage to surrounding healthy tissue.

- Cryotherapy. Cryotherapy involves the application of a cryoprobe to the surface of the inferior turbinate. Cryotherapy induces the formation of intracellular ice crystals and extended destruction of goblet cells due to the large amounts of water inside their membranes. Tissue reduction is achieved through freezing around −85 °C for 60 to 75 seconds. Cryotherapy is effective in nonallergic rhinitis but fails to achieve notable results in cases of allergic rhinitis. Results are most of the time temporary and mucosal destruction and scarring can potentially hamper successful treatment of nasal obstruction.
- Laser Surgery. Various types of laser have been used to reduce the volume of turbinate hypertrophy, including CO2, argon, KTP, Nd: YAG, Ho: YAG, and diode lasers. In principle, a laser is an excellent tool for a controlled coagulopathy of soft tissue, as heat from the laser leads to coagulation of the surrounding tissue and scarring of the turbinates leading to a reduction in size. The effect of laser application on mucociliary clearance is widely debated. While some advocate that irreversible damage to epithelium can decrease clearance, other studies have shown no long-term effect.
- Turbinate surgery. There are multiple surgical techniques in treating turbinate hypertrophy, including turbinectomy, lateralization, submucosal resection of turbinate bone, turbinoplasty with or without the use of powered instrumentation, and in the greater sense, even Vidian neurectomy can be listed in the same category.
  - Turbinectomy (resection of the inferior turbinate) became one of the most frequently practiced rhinological surgical procedures in the first quarter of the twentieth century. Nevertheless, disastrous postoperative sequelae, including postoperative atrophic rhinitis, "secondary" ozena and "empty nose syndrome," has raised significant concerns and method was practically abandoned in favor of more conservative techniques.
  - Lateralization by outfracturing the turbinate was introduced as early as 1904 by Killian. It is widely used as an adjunct to septoplasty procedures, as it is a fast, conservative, function-preserving method with almost zero side effects, although results are reasonably limited.
  - Submucosal resection of the turbinate bone was presented around 1910, as an alternative to the more aggressive techniques and was later refined by House in the 1950s and again by Mabry in the 1980s when the term "turbinoplasty" was introduced. The main rationale of the procedure is that preserving the outer mucosa intact and removing submucosal tissue and turbinate bone. The technique can involve either manual or powered instrumentation (microdebrider). A vertical incision is made in the anterior head of the inferior turbinate and soft tissue is separated from the turbinate bone by blunt dissection. Turbinate bone is removed with or without debulking of the submuco-

sal soft tissue using standard forceps or powered turbinate microdebrider type blade [microdebrider-assisted inferior turbinoplasty (MAIT)]. Care is taken not to perforate the mucosal flap, which is then repositioned.

It cannot be overemphasized that despite the variety of techniques, the cornerstone in turbinate surgery remains the attempt to widen the nasal airway while respecting nasal physiology. It is wise to mention the presence of an iatrogenic disorder named "empty nose syndrome" (ENS). In this condition not rarely encountered after drastic reduction of inferior turbinate mucosa (mainly following turbinectomies) paucity of the mucosal surface area leads to the presence of paradoxical nasal obstruction despite an objectively wide, patent nasal fossa. Thus, preserving as much as possible of the normally functional turbinate mucosa is a prerequisite in successfully addressing turbinate hypertrophy.

Summary and Author's Comments

1. Hypertrophy of the turbinates is one of the most common causes of nasal obstruction, to the point that up to 20% of the population can suffer from chronic nasal obstruction secondary to turbinate hypertrophy.
2. There are various causes of turbinate hypertrophy that can include conditions that cause permanent turbinate hypertrophy producing chronic nasal obstruction, such as allergic rhinitis, vasomotor rhinitis, or chronic hypertrophic rhinitis.
3. First-line treatment for turbinate hypertrophy involves conservative medical management, which should be attempted first, if not contraindicated, before surgical intervention.
4. Although there are many options for turbinate reduction, there is no consensus on the best and most effective technique.
5. It cannot be overemphasized that despite the variety of techniques, the cornerstone in turbinate surgery remains the attempt to widen the nasal airway, while respecting nasal physiology.

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Septal Lesions

74

Sarantis Blioskas

74.1 Case Presentation

A 52-year-old male was referred from his GP for an over six months history of soreness at the area of the nasal bridge, along with nasal blockage and occasional discharge. The patient described a long history of nasal deformity due to nasal skin thickening, flushing, and persistent redness, as well as papules and pustules. The nasal bridge also appeared flattened and expanded (Fig. 74.1). On flexible nasendoscopy, there was a gigantic mass arising from the dorsal part of the nasal septum, projecting to the nasal cavity bilaterally (Fig. 74.2). An MRI scan depicted an infiltrative mass centred within the nasal septum extending anteriorly into the nasal soft tissues and premaxillary fat. It measured approximately 5.9 cm AP by 3 cm transverse diameters in the axial plane (Figs. 74.3 and 74.4). There were aggressive features with bony destruction of the nasal septum. An examination under anesthetic and biopsy of the lesion was arranged, and histology revealed high-grade dysplasia and squamous cell carcinoma. After discussion of the case to a Head and Neck MDT meeting, the patient underwent induction chemotherapy followed by endoscopic excision of the lesion (endoscopic septectomy).

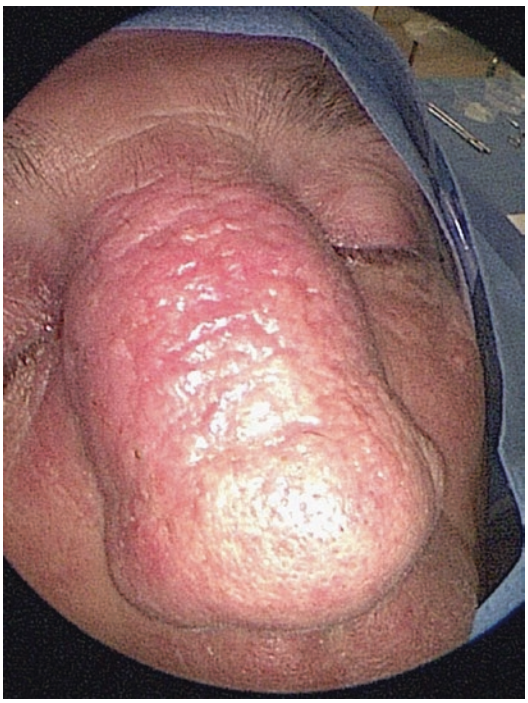

Fig. 74.1 Skin changes and widening of the nasal dorsum due to the underlying pathology

74.2 Background Knowledge

The nasal septum, an integral part of nasal anatomy, is the primary provider of support to the overall nasal structure. The septum, unlike most of the nasal structures, is singular, and it can be

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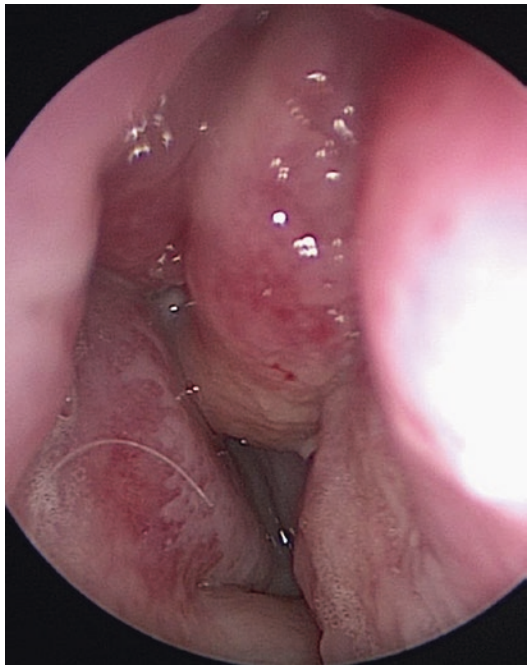

Fig. 74.2 Endoscopic photograph showing the lesion attached to the nasal septum

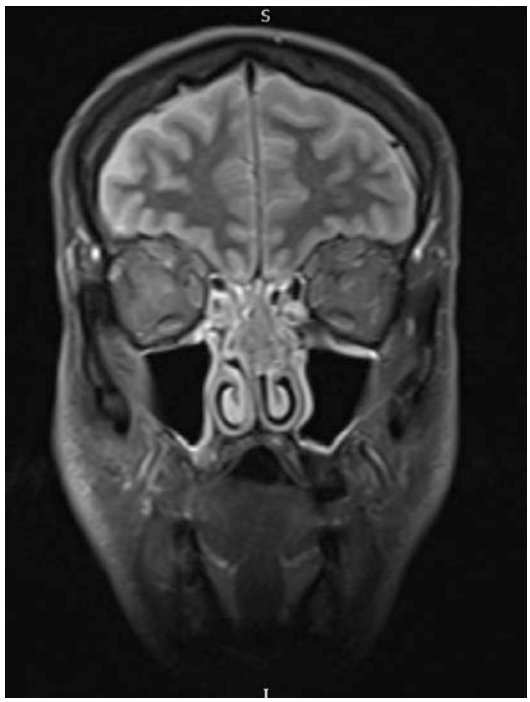

Fig. 74.3 Gadolinium-enhanced MRI scan showing the mass involving the nasal septum

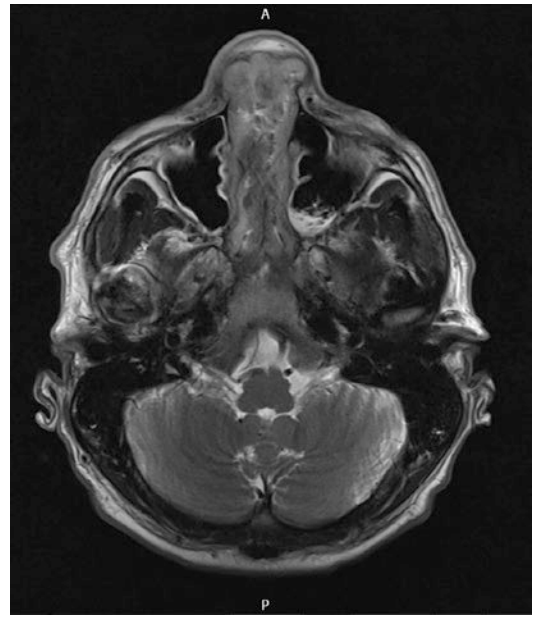

Fig. 74.4 Axial MRI demonstrating the extent of the pathology

divided into a bony and a cartilaginous portion. The former comprises the perpendicular plate of the ethmoid postero-superiorly and the vomer posteroinferiorly and the later to the septal (quadrangular) cartilage anteriorly. Both the bony and the cartilaginous are covered by mucoperiosteum and mucoperichondrium.

In general, tumors and lesions of the nasal septum are rare. Both benign and malignant nasal septal lesions arise either from the epithelia of the nasal mucosal lining, the nervous and blood supply of the septum, or directly from the cartilaginous and the bony parts themselves. Although the vast majority of minor mucous and serous glands are located in the lateral nasal wall, tumors deriving from such structures like pleomorphic adenomas could originate from the nasal septum.

In terms of anatomical landmarks, one should always keep in mind the presence of the nasoseptal swell body. It is located in the anterior septum and is comprised of submucosal erectile tissue. This discrete area could appear as a suspicious lesion, and consideration should be given regarding differential diagnosis from small septal swellings.

74.3 Clinical Approach

More than 90% of neoplasms of the nasal septum are malignant. (Table 74.1),

Table 74.1 Nasal septal lesions

Malignant lesions	
Squamous cell carcinoma	Although primary squamous cell carcinomas of the nasal septum constitute only 9% of all malignant tumors of the nasal cavity, they represent the most prevalent malignant tumor of the septum accounted for more than half the cases in a large series. There are no disease-specific findings; thus a high index of suspicion is necessary. The best method of treatment is wide surgical excision and immediate repair of the defect. Cosmetic results in larger tumors have been achieved with adhesive or bone-anchored prosthetics
Malignant melanoma	Primary malignant melanoma of the nasal cavity accounts for less than 1% of all melanomas. The commonest site of origin within the nasal cavity is the nasal septum, followed by the inferior and middle turbinates. It is a highly aggressive neoplasm carrying a poor prognosis. Early diagnosis with a high index of suspicion is critical because this type of tumor is extremely aggressive, yet symptoms are initially nonspecific. The presence of a pigmented lesion in the nasal cavity should raise the suspicion of malignant melanoma. An incisional biopsy is used to establish a definite diagnosis. The mainstay of treatment as in all melanoma cases involves complete resection with wide negative margins; nevertheless, this is often not achievable due to delayed diagnosis and adjacent critical structures like the skull base and the orbit. Radiation therapy as a primary or as an adjunctive treatment, has also been used with not very encouraging results.
Adenoid cystic carcinoma	Adenoid cystic carcinomas typically arise from seromucinous salivary tissue in the major and minor salivary glands and it is rare in the nasal cavity. When it appears nasally, it can carry a worse prognosis than in any other head and neck areas. It is generally a slow-growing tumor with nonspecific presenting symptoms; thus it can reach large dimensions before becoming symptomatic. Adenoid cystic carcinoma spreads hematogenously with lungs being the commonest metastatic site, as well as perineurally, with the latter being a pathognomonic factor and the cause for high rates of local recurrence. Lymphogenous metastases are extremely rare.
Metastatic renal cell carcinoma	Renal cell carcinoma is the third most common infraclavicular neoplasm to metastasize to the head and neck, yet the pattern of metastasis to the nasal cavity is unclear. Although metastases of renal cell carcinoma tend to occur multifocally, nasal metastases are usually single. Epistaxis is the most common presenting symptom due to the vascularity of the tumor. This can also become an issue during diagnostic biopsies, as intraoperative bleeding can be severe. The mainstay of treatment includes the excision of solitary metastatic lesions after nephrectomy. Systemic immunotherapy with interleukin two or interferon α is also utilized, whereas radiotherapy is of little use, as renal cell carcinomas are considered generally radioresistant tumors.
Mucoepidermoid carcinoma	Mucoepidermoid carcinoma is the most common malignant salivary gland tumor. Histologically, mucoepidermoid carcinoma can be classified into a low, intermediate or high malignancy grade. Histological classification affects the treatment algorithm. Surgical resection alone has been advocated for low-grade mucoepidermoid carcinoma. However, in case of high-grade tumors, postoperative radiation is utilised. Lymphogenous metastasis should be treated with appropriate neck dissection +/− postoperative radiotherapy.
Other malignant neoplasms also include transitional cell carcinoma, reticulum cell sarcoma, adenocarcinoma, Chondrosarcoma, osteosarcoma, anaplastic carcinoma, Histiocytic lymphoma, Verrucous carcinoma, neuroendocrine carcinoma, carcinoma ex-pleomorphic adenoma.	

(continued)

Table 74.1 (continued)

Benign lesions	
Angiofibroma	Angiofibromas typically base on the posterolateral wall of the nasal cavity near the superior margin of the sphenopalatine foramen. Yet, they can rarely occur outside the nasopharynx, and in these instances, they are termed extranasopharyngeal angiofibroma (ENAs). Their most prevalent location is the maxillary sinus, while the nasal septum is an infrequent location of origin. Extranasopharyngeal angiofibromas are reported in the literature as case reports as they are quite rare and their different location, biological history and clinical behaviour (symptoms, age and sex predilection) compared to that of nasopharyngeal angiofibromas has led to doubt as to whether they should be considered as separate entities. Nasoseptal angiofibromas can occur at the anterior one-third of the nasal septum, the bony cartilaginous junction, or the ethmoidal perpendicular plate. Initial symptoms usually are epistaxis alone or with nasal obstruction as well as a well-circumscribed lobule nodular mass covered by intact mucosa into the nasal cavity having a slow-growing but locally invasive pattern. Thus, despite being uncommon, nasal septal angiofibromas should be considered during the evaluation of epistaxis, although they can present a diagnostic challenge and generally, a high index of suspicion is essential. Differential diagnosis may include lobular capillary hemangioma and sinonasal-type hemangiopericytoma. Regarding diagnosis, CT scan and magnetic resonance imaging (MRI) are used to determine the tumour site and its extension while hypervascularity, will dictate arteriography prior to surgical procedures in order to arrange potential embolization. Surgical excision of the lesion is advocated as the treatment of choice, mainly through an endoscopic and endonasal approach with or without preoperative embolization. Recurrence is rare. Office biopsies should be avoided, given the risk of brisk hemorrhage.
Schwannoma	Schwannoma is a benign slow-growing neurogenic tumour arise from the sheath of any myelinated nerve fiber. Between 25% and 45% of such tumours occur in the head and neck region, the eighth cranial nerve (vestibulo-cochlear nerve) is the most typical site, while potential sites include the scalp, face, parotid gland, tongue, soft palate, pharynx, parapharyngeal space, larynx, trachea, middle ear, internal and external auditory meatus, and neck. Only 4% of the lesions involve nasal and paranasal cavities and schwannoma of the nasal septum is still the rarest. The lesion is almost always solitary and is predominantly observed between 25 and 55 years of age, without any prevalence of race or sex. In descending order the posterior nasal septum, followed by midportion of the nasal septum, and anterior nasal septum give rise to such tumors, which originate from either the sympathetic nerve to the septal blood vessels, the parasympathetic nerve to the septal mucous glands, or the sensory nerve to the septum (nasopalatine nerve and the anterior and posterior ethmoidal nerves). Symptoms and signs depend on the site of origin and the extent of the lesion and could include nasal obstruction, anosmia, deformity of the nasal pyramid, headache, and epistaxis. Septal schwannoma commonly displays a polypoid appearance without any distinctive features and CT scan findings are not specific. Magnetic resonance imaging (MR) is superior to CT in differentiating a tumour including findings like the target sign and fascicular sign on the T2-weighted sequence. Still, nasal septal schwannoma diagnosis primarily depends on biopsy or complete excision of the mass. The treatment of choice for nasal septum schwannoma is surgical resection of the mass, since schwannomas have the potential of malignant transformation. Excision may be achieved by external approaches (lateral rhinotomy with external ethmoidectomy, the Caldwell-Luc approach, midface degloving) or endoscopic endonasal surgery with the latter replacing the former in the majority of the cases. The condition is typically curative with rare postoperative recurrence, yet more evidence is needed to elucidate the association between safety margin and tumor recurrence.

Table 74.1 (continued)

Benign lesions	
Pleomorphic adenoma	Pleomorphic adenomas arise mainly in major salivary glands, yet a small minority have been reported in the nasal cavity, paranasal sinuses, nasopharynx, oropharynx, hypopharynx, and larynx, with the most favoured site of origin in the upper respiratory tract being the nasal cavity and in particular the nasal septum although the vast majority of minor mucous and serous glands are located in the lateral nasal wall and turbinates. Intranasal pleomorphic adenomas are quite rare, slow-growing tumours presenting between the age of 30 and 60 years and being slightly more common in women. Despite their benign nature, they may develop malignant behavior (ex-pleomorphic carcinoma). Typical presenting features include unilateral nasal obstruction, epistaxis, nasal deformity, epiphora, and mucopurulent rhinorrhea. Nasal pleomorphic adenoma appears as a pink-grey unilateral mass with a polypoid aspect, smooth surface and a firm or soft consistency. Surgical excision with clear histological margins is generally accepted as the treatment of choice. Surgical approaches include endonasal endoscopic resection, external rhinoplasty, midfacial degloving, and lateral rhinotomy. The choice of surgical technique is founded on the size and location of the lesion. Intranasal pleomorphic adenoma has a low rate of recurrence compared with recurrence rates of the major salivary gland tumors. Clinical follow up is still necessary to exclude the malignancy and reduce the loco-regional recurrence rate.
Chondroma	Chondromas are benign lesions with well-defined boundaries, composed of hyaline cartilage tissue. Such lesions are mostly seen in the hands and feet of adults, only 10% of all cartilaginous tumors arise from the head and neck region (most commonly observed in the larynx and the ethmoido-sphenoidal area). Lesions of the nasal septum are exceedingly rare. Although chondromas are benign, they are locally invasive and also tend to recur after removal. They can also have a tendency towards sarcomatous change. Their symptoms in the nasal cavity depend on the site, size, and rate of growth of the tumor. Nasal chondromas are characterized by slow growth and do not cause pain; thus they are usually diagnosed incidentally. On palpation, chondromas are generally non-tender, fixed, and of firm consistency. On CT images, they are well-defined homogenous masses and in MR images, chondromas have a higher signal intensity in T2 than in T1. Biopsy is the only specific means of diagnosis. Differential diagnosis should include well-differentiated chondrosarcoma and the pathologist might have difficulty in separating these two lesions. Local resection is the accepted treatment and as chondromas tend sarcomatous change, long-term follow-up is strongly recommended.
Papillomas (Exophytic / inverted)	Histologically, the sinonasal papillomas are divided into three different subtypes: Inverted papillomas, columnar cell papillomas, and exophytic papillomas. Exophytic papillomas are predominantly localised on the nasal septum, while the others almost always occur on the lateral nasal wall or in the adjacent sinuses. Nasal septal papillomas are also known as fungiform papillomas are also known as septal papillomas, everted papillomas, papillomatosis and Ringertz tumours. The consensus in most of the literature is that malignant change in fungiform papillomas is rare Controversy exists about whether rare septal inverted papillomas are true inverted papillomas or whether they should be considered differently from those of the lateral nasal wall and paranasal sinuses. Nevertheless, the clinical course of these patients suggests that inverted papillomas of the nasal septum behave like inverted papillomas elsewhere and require wide surgical excision and careful follow-up. Fig. 74.5
Other benign lesions include myoepithelioma, fibrous histiocytoma, adenomatoid hamartoma	

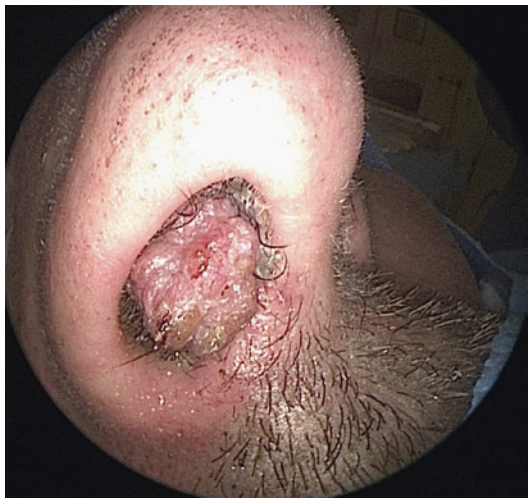

Fig. 74.5 Exophytic nasal papilloma, attached to the anterior nasal septum

Summary and Author's Comments

1. Both benign and malignant nasal septal lesions arise either from the epithelia of the nasal mucosal lining, the nervous and blood supply of the septum, or directly from the cartilaginous and the bony parts themselves
2. More than 90% of neoplasms of the nasal septum are malignant.
3. Benign lesions include angiofibromas, schwannomas, pleomorphic adenomas, chondromas and papillomas (Exophytic / inverted). Malignant lesions include SCC, melanomas, adenoid cystic carcinomas, metastatic renal cell carcinomas, and mucoepidermoid carcinomas

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Septal Perforation: Surgical Repair

75

Sarantis Blioskas

75.1 Case Presentation

A 37-year-old female patient was referred to the ENT department due to nasal congestion, recurrent epistaxis and nasal crusting. The patient underwent a septoplasty and diathermy of inferior turbinates, about one year prior to referral and a right cartilaginous septal spur was resected. The patient reported good postoperative results, but two months after surgery, the nasal blockage recurred and crusting appeared to be exacerbated. The patient reported no underlying conditions, no previous substance misuse or intranasal medications and she was not a smoker. On examination, flexible nasendoscopy revealed two <0.3 cm septal perforations at the middle part of the quadrangular cartilage, in close proximity. There was reasonable crusting on the periphery and the rest of the nasal mucosa appeared healthy and well healed. The rest of the head and neck examination was unremarkable and blood tests did not reveal evidence of underlying vasculitis or granulomatous disease (cANCA/pANCA/PR3 negative). The patient was given the option of surgical correction with a favourable prognosis due to the size and location of the perforations or the fitting of a septal button. The patient opted for the former, and under general anesthesia, an endonasal approach was utilized to elevate mucoperichondrial flaps. The margins were reapproximated with basting sutures. Silastic splints were placed and preserved for three weeks. On her follow up two months later, the patient was pleased with the result.

75.2 Background Knowledge

Nasal septal perforations despite general belief can be quite common, with some reports raising their prevalence up to 0.9% of the general population. Yet, it should be noted that patients with septal perforations constitute a highly diversified group, ranging from patients unaware of their perforation (incidental finding during routine clinical examination) to patients suffering from severe recurrent nosebleeds and nasal symptoms with a significant decrease of their quality of life. Thus the initial challenge is to differentiate those cases that require additional workup and surgical treatment.

Theoretically septal perforations can occur on any part of the nasal septum; nevertheless their vast majority occurs at the anterior part. The anterior septum is formed of the quadrangular cartilage, which is covered by mucoperichondrium. Depending on the specific cause, the usual pathophysiologic process involves ischemic damage of the mucosal layer and underlying perichondrium. As the cartilage bears no

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individual blood supply and depends on its overlying perichondrium to sustain vitalization, destruction of the perichondrium leads to gradual cartilage necrosis. This process inevitably leads to the formation of a septal perforation. Once such a perforation is formed, mucosal edges quickly epithelialize over the edges of the destructed cartilage, and this process prevents closure of the defect during normal healing.

75.3 Clinical Approach

Septal perforations can arise from different underlying causes. Potential factors that could lead to perforation of the nasal septum are summarized in Table 75.1.

In terms of symptomatology, a septal perforation can range from being asymptomatic to result in severe nasal destabilization and deformity. More commonly clear perforations with well-healed margins are not causing any issues and they could be incidentally discovered. Nevertheless, sometimes a perforation can lead to an alteration of the normal intranasal laminar airflow that can cause an annoying whistling sound. Furthermore, extensive crusting at the periphery of the perforation could result in significant nasal obstruction, while chronic picking of it, can lead to progressive enlargement of the defect. In the same context, removal of crusted, dried secretions that collect at the edges of the perforation can result in recurrent epistaxis. Poor nasal hygiene and chronic infective secretions

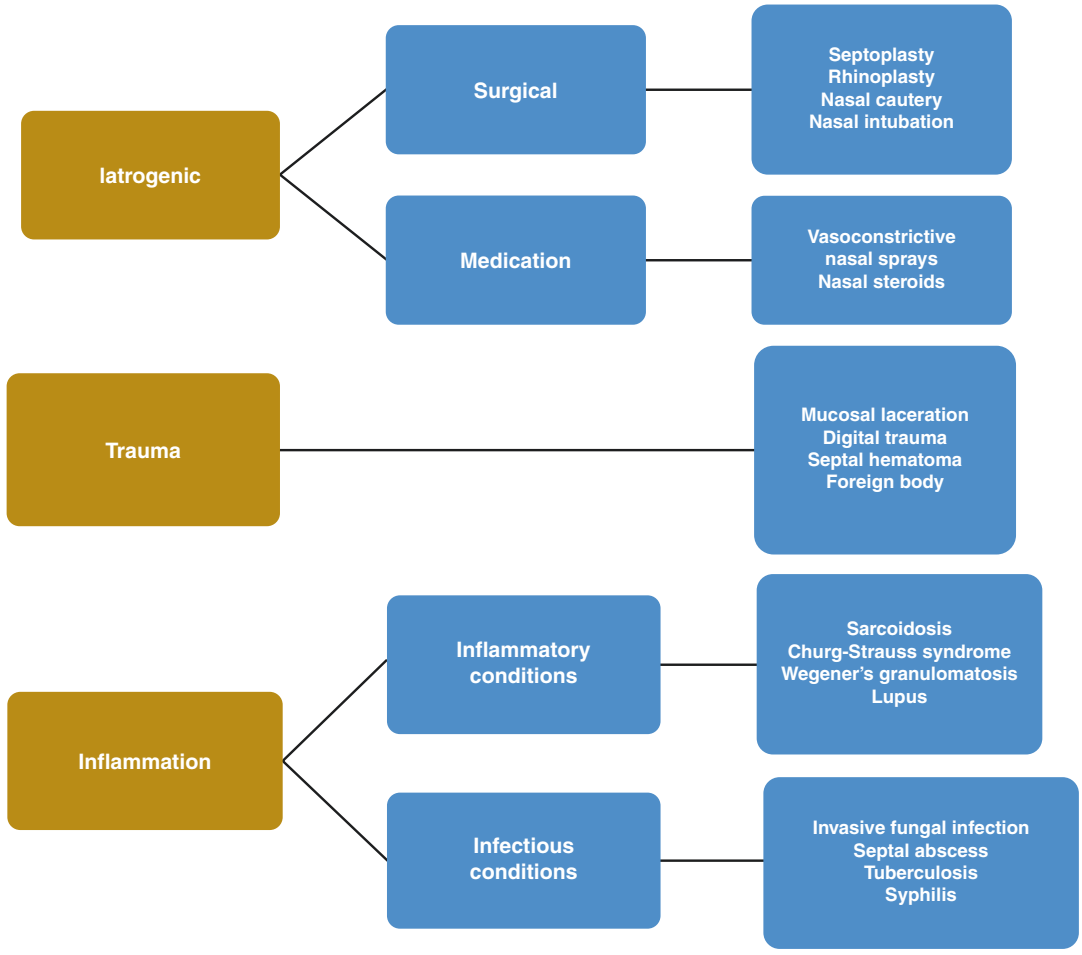

Table 75.1 Causes of septal perforation

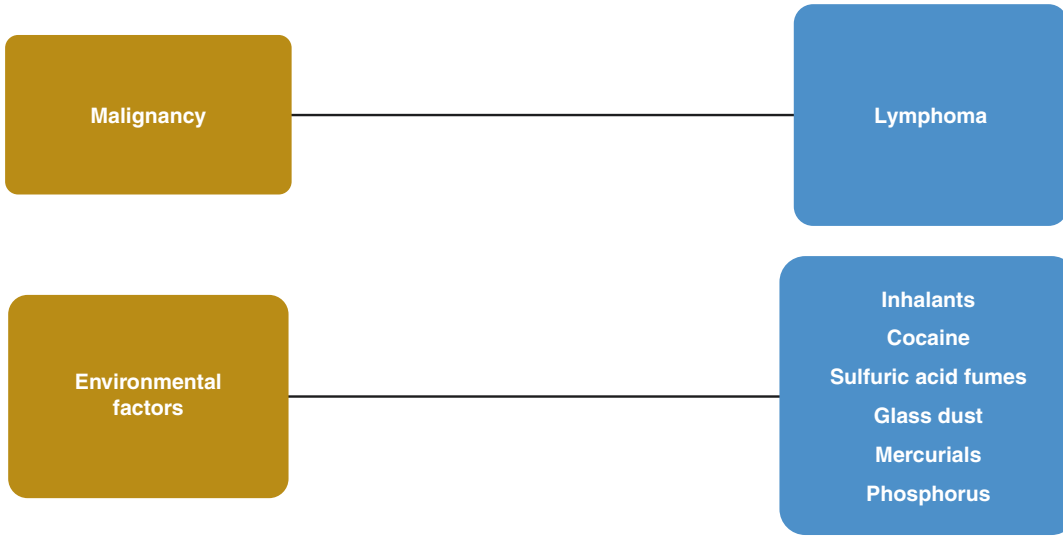

Table 75.1 (continued)

could result in persistent low-grade perichondritis, and consequently, infection leading to progressive enlargement of the perforation. Such an enlargement could compromise the supporting role of the septum and lead to internal nasal valve collapse, nasal tip collapse and/or saddle-nose deformity.

Diagnostic workup related to septal perforations aims to determine the underlying aetiology before recommending or not a surgical repair. Diagnosis should include:

- History taking: Although perforation may pass unnoticed, history should look for nasal obstruction, an audible whistle during nasal breathing, nasal crusting, epistaxis, and copious nasal discharge. Questions should also inquire about nasal surgical procedures including cautery, prior nasal trauma or foreign body septal injury, cocaine use, excessive use of vasoconstrictors or nasal steroids and excessive digital manipulation of nasal crusting.
- Clinical evaluation: Flexible or rigid nasal endoscopy is invaluable in evaluating septal perforations. Endoscopy can provide information about the size (even measured through a paper ruler) and the site (anterior or posterior, high or low) of the perforation. Both facts may affect the choice of surgical repair. Also, the presence of well-healed or crust and ulcerous periphery could dictate further workup (biopsy) and selection of treatment.
- Laboratory testing: Laboratory findings are essential in investigating certain inflammatory conditions. Thus, elevated antineutrophil cytoplasmic autoantibody (c-ANCA) can point towards Wegener's granulomatosis (currently named c-ANCA vasculitis), especially when combined with a high erythrocyte sedimentation rate (ESR) and rheumatoid factor (RF). Elevated serum angiotensin-converting enzyme (ACE) and serum calcium levels can be indicative of sarcoidosis, whereas elevated perinuclear-staining antineutrophil cytoplasmic antibodies (p-ANCA) and peripheral blood eosinophilia could suggest Churg-Strauss syndrome
- Biopsy: Biopsy from the periphery of the perforation is an option in ulcerous, crusting and inflamed perforations to rule out T-cell lymphoma. Literature suggests that only clinically malignant perforations are worthwhile biopsying. Biopsies of the superior margin of the perforation should be avoided because they contribute to the vertical diameter of the defect and increase the difficulty of the eventual closure.
- Imaging: Imaging is not an essential part of the diagnostic battery. A sinuses CT scan can

be helpful to look for co-existing sinus disease, whereas chest radiograph may exhibit mediastinal adenopathy in cases of sarcoidosis.

In terms of surgical repair, it should always be stressed that septal perforation repair is elective, carries the risk of failure and conservative alternatives can minimize symptomatology. Nevertheless, small perforations have higher rates of successful repair than larger ones, so there is a theoretical advantage in deciding to undertake early surgical repair.

The rates of success can be influenced by various factors. First of all the causative factor should be treated prior to undertaking an attempt to repair. This is particularly important for cocaine use induced perforations, where it should be made certain that patients have abandoned cocaine use. In the same context, patients should abandon chronic use of intranasal vasoconstrictors, and for patients suffering from c-ANCA vasculitis, surgery should be delayed until systematic treatment achieves disease remission. Given the above are guaranteed, successful operation depends on the size and location of the perforation, the remaining cartilage and the presence of scar and granulation tissue.

Regarding surgical technique, either the endonasal or the open rhinoplasty approach can be utilized. Generally, defects measuring less than 5 mm can be closed by the endonasal approach, although moderate-sized perforations up to 2 cm have also been successfully treated with this approach. When the endonasal approach is used, mucoperichondrial flaps are elevated and reapproximated with basting sutures (Figs. 75.1 and 75.2). Usually an interpositional graft can be inserted between the mucoperichondrial flaps to reinforce the repair like bone, cartilage, periosteum and acellular dermal allograft. Silastic splints are routinely placed and preserved for at least 2 to 3 weeks.

The external or open rhinoplasty approach provides superior access and is usually reserved for larger defects. It involves a standard inverted-v mid-columellar incision. Mucoperichondrial flaps are elevated up to the region of the perforation and posterior and inferior to it. The maxillary crest and nasal floor are included in the flap and dissection extends laterally to the insertion of the inferior turbinate bone to the lateral nasal wall. A lateral releasing incision just below the insertion of the inferior turbinate creates a bipedicled mucoperichondrial flap, which is medialy advanced to close the perforation. The bipedicled flap can be converted to a rotational flap through a transverse mucosal incision extending from the nasal spine, across the anterior nasal sill, directed toward the lateral wall, in cases where additional

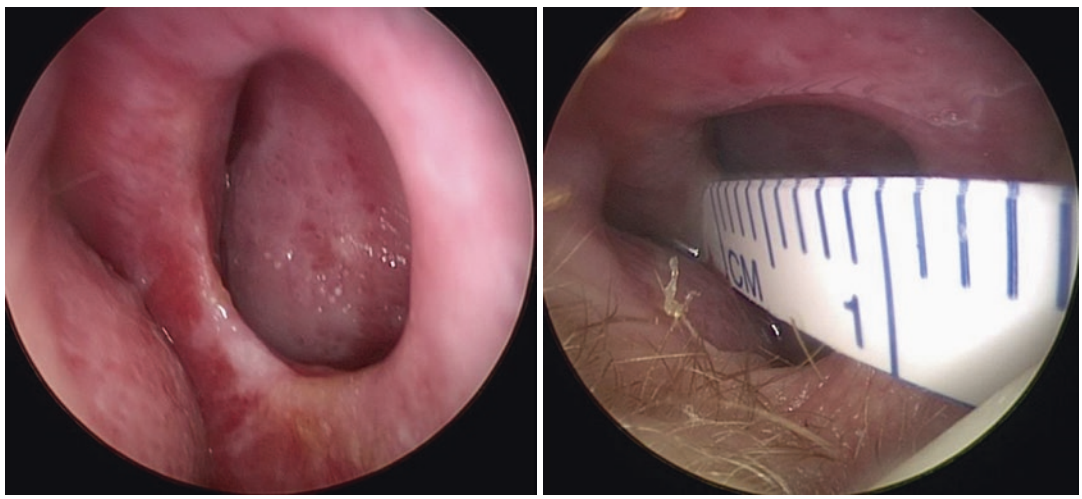

Fig. 75.1 Septal perforation measured intraoperatively

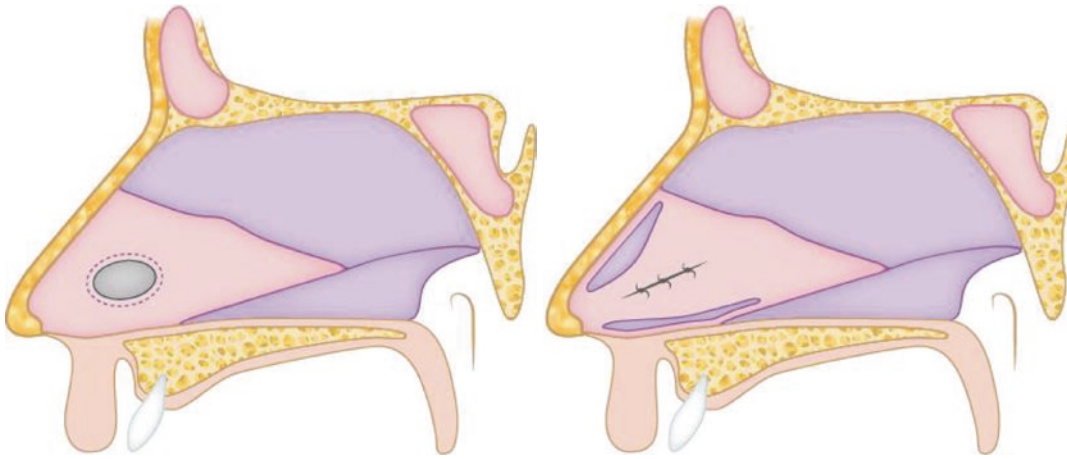

Fig. 75.2 Technique employed by the authors for relatively smaller septal perforations

mucosal mobility is necessary. In cases of even larger defects, separation of the mucoperichondrium from the undersurface of the lower lateral cartilages, and recruitment of the mucoperichondrium from areas above the perforation could also be utilized. Again, an interpositional graft can be inserted between the mucoperichondrial flaps. The areas of the exposed nasal floor are left to re-mucosalize and as above silastic splints are routinely placed against the septum.

Perforations larger than 2 cm are not likely to be efficiently treated with intranasal flaps. In these cases the following options exist:

(i) Inferior turbinate pedicled flap. It can be performed through an endonasal approach and is particularly useful in caudal perforations. An incision is made at the posterior–inferior aspect of the inferior turbinate, and a flap, including mucosa and submucosa (along with bone if necessary), is raised. The distal portion of the flap is pulled forward, split open and sutured into the septal perforation. This flap procedure requires a second stage 3 weeks later to divide the pedicle. The division can be performed in an outpatient setting under local anesthetic.

(ii) Anterior ethmoidal artery septal flap (Castelnuovo flap). Endoscopic repair of anterior septal perforations can be achieved via a unilateral septal flap pedicled by the anterior ethmoidal artery. The posterior border of the flap is fashioned vertically along the septum, 0.5 to 1.0 cm posterior to the septal projection of the axilla of the middle turbinate and continued along the nasal floor, reaching the lateral wall of the posterior portion of the inferior meatus. Then, turns parallel to the septum until it reaches the anterior portion of the inferior meatus. At this point, it becomes perpendicular to the septum, reaching the inferior border of the perforation. This maneuver creates a larger superiorly based rotation advancement flap, supplied by the anterior ethmoidal artery. The perforation on the contralateral side is not covered. (Fig. 75.3)

(iii) Tunneled sublabial mucosal lap. This procedure uses oral mucosa by incising the ipsilateral buccal mucosa, leaving the mucosa adjacent to the frenulum intact. A medially-based pedicled flap is raised and a midline sublabial-nasal fistula is created so the flap can be brought up into the nose and tunneled between the previously elevated septal mucoperichondrial flaps.

(iv) Facial artery musculomucosal flap. This flap can be utilized to treat perforations up to 4 cm. The donor area is the buccal mucosa immediately subjacent to the facial

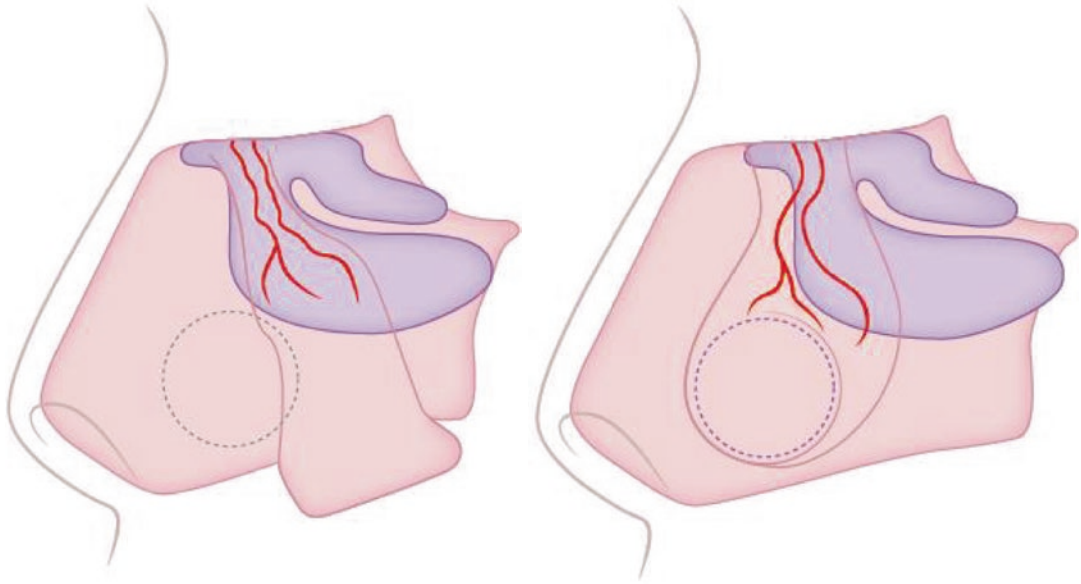

Fig. 75.3 Anterior ethmoidal artery septal flap

artery and extends to the inferior gingivobuccal sulcus. The mucosa is harvested along with the underlying vessel and then tunneled through a subperiosteal dissection into the piriform aperture where it is sewn into the perforation. The contralateral side can be skin grafted or let to granulate.

(v) Radial forearm free flap. This flap is based on the radial artery, which is anastomosed to the facial artery and sewn into the septal perforation.

(vi) Staged tissue expansion. A staged tissue expansion followed by reconstruction of the defect two weeks later can also be an option.

(vii) Posterior septectomy. Posterior septectomy has been reported as a simple alternative to treat large nasal septal perforations, by addressing their symptoms. The removal of high stress points along the posterior margin may explain why posterior septectomy can be an effective treatment option.

Summary and Author's Comments

1. Nasal septal perforations can be quite common and their symptomatology range from being asymptomatic to resulting to severe nasal destabilization and deformity.
2. Septal perforations can arise from different underlying causes.
3. Diagnostic work-up includes detailed history, nasal endoscopy, laboratory tests and potential biopsy
4. In terms of surgical repair, it should always be stressed that septal perforation repair is elective, carries the risk of failure and conservative alternatives can minimize symptomatology. Regarding surgical technique either the endonasal or the open rhinoplasty approach can be utilized.

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Septal Perforation: Septal Button

76

Sarantis Blioskas

76.1 Case Presentation

A 30-year-old police officer was referred to the ENT department due to an annoying whistling sound during nasal inhalation, that was apparent to both him and his partner. The patient did not report any other nasal issues, such as nasal obstruction, discharge, recurrent epistaxis or other ENT related symptoms. He denied any underlying conditions, previous substance misuse, nasal medication or surgery and he was not a smoker. The patient did report that he was aware of an anterior septal "hole" that he was seeing on mirror self-examination, though he could not recollect the first time he was made aware of it. However, the patient used to be an active martial arts athlete and had sustained repeated and severe nasal injuries that were not medically investigated or treated at the time, he could not report a direct link between the two. On examination, both anterior rhinoscopy and flexible nasendoscopy revealed an >1 cm anterior septal perforation (Fig. 76.1). There were no nasal crusting or bleeding on the periphery and the nasal mucosa appeared healthy and well healed, despite lack of any prior conservative treatment. The rest of the head and neck examination was unremarkable. Since the whistling sound was the symptom reported, the patient was given the option of either surgical correction (with a guarded prognosis due to size and location of the perforation) or the fitting of a septal button. The patient opted for the latter and a septal button was trimmed to appropriate dimensions and fitted under general anesthesia. On his follow up two months after surgery, the patient was pleased with the result.

76.2 Background Knowledge

The pathophysiology, symptomatology and diagnostic workup of a septal perforation are analyzed in the previous chapter and the reader is kindly requested to review them prior to embarking on studying further.

Irrespective of the etiology of septal perforation, whistling sounds, crusting and recurrent nosebleeds remain the primary concern of patients. Inserting a septal button can be an ideal alternative to surgical management, as it is a fast, low cost, minimally invasive procedure that bears excellent results and low failure rates. Furthermore, compared to surgical correction, insertion of septal button carries the advantage that in failure to meet the patient's expectations, it can be easily removed with minimal discomfort for the patient.

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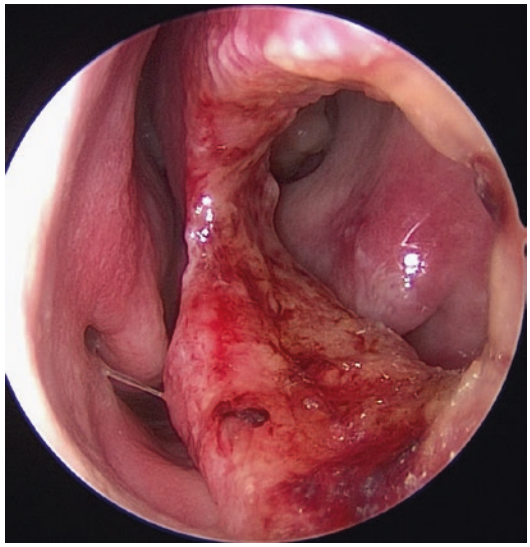

Fig. 76.1 Large anterior septal perforation

76.3 Clinical Approach

Regarding conservative treatment, initial management of septal perforation begins with improving nasal hygiene. All factors that contribute to the enlargement of septal perforation like substance misuse, vasoconstrictive agents, recurrent nasal trauma or even smoking should be eliminated and the patient should be counseled to avoid digital cleaning.

Routine nasal irrigation with saline solution or regular humidification through steam inhalation can improve the condition of nasal mucosa and help reduce the build-up of crusts. Nasal ointments, usually vaseline-based, applied regularly prevent the drying and hardening of crusted material, further alleviating symptoms of nasal blockage and discomfort. In the setting of visible mucosal inflammation, an antibiotic-based ointment might be preferable. It is not rear that conservative treatment alone successfully manages complaints of pain, dryness and crusting at the perforation site.

In cases where long term conservative treatment has ultimately failed to resolve symptomatic complaints of the patient and as the risk of progressive enlargement of the perforation is always present, the patient could be given the option of a septal button insertion. Prefabricated buttons are typically made of soft silicone and are comprised of a flexible hub connecting two pliable discs, which allow them to adapt to the curvatures and irregularities of the septum. Septal buttons come in various sizes and can be inserted into position under general or local anesthesia.

Different maneuvers can be used to facilitate proper placement of the button. An endoscopic view of the nasal cavity can significantly aid placement and minimize insertion trauma or displacement. Most of the times one side of the disk is folded and passed or pulled through the perforation with alligator, cup or bayonet forceps. The flanges should be seated in the region of the internal nasal valve superiorly and contact the nasal floor inferiorly. Other placement techniques involve the "sutured rosette" technique or facilitating a vertical incision in one of the disks and rotating it through the perforation in place, much like a corkscrew. (Fig. 76.2).

Despite that commercially available buttons come in various sizes, nasal septal buttons can be difficult to accurately size due to the often irregular shape of septal perforations. Obturators that are too small may become dislodged, while those which overlap the perforation too much may lead to nasal obstruction and crusting. It is almost always necessary to measure the size of the perforation and trim the button to the appropriate size. A simple ruler under endoscopic vision can be very helpful, while other sizing techniques can also be useful. In cases of extremely large perforations, where commercially available buttons prove to be of suboptimal fit and at high risk of dislodgement and ongoing symptoms, a custom made button can be manufactured. Imaging can be helpful in customizing septal buttons, while the use of modern 3D printing can provide optimal prosthetic closure. When such equipment is unavailable, combining two buttons can manifest solutions to difficult problems.

After a septal button is inserted, it can remain in place for long periods, yet this duration is very dependent on the patient's diligence with good nasal hygiene and proper care of the prosthesis. Treatment with saline irrigation and vaseline-

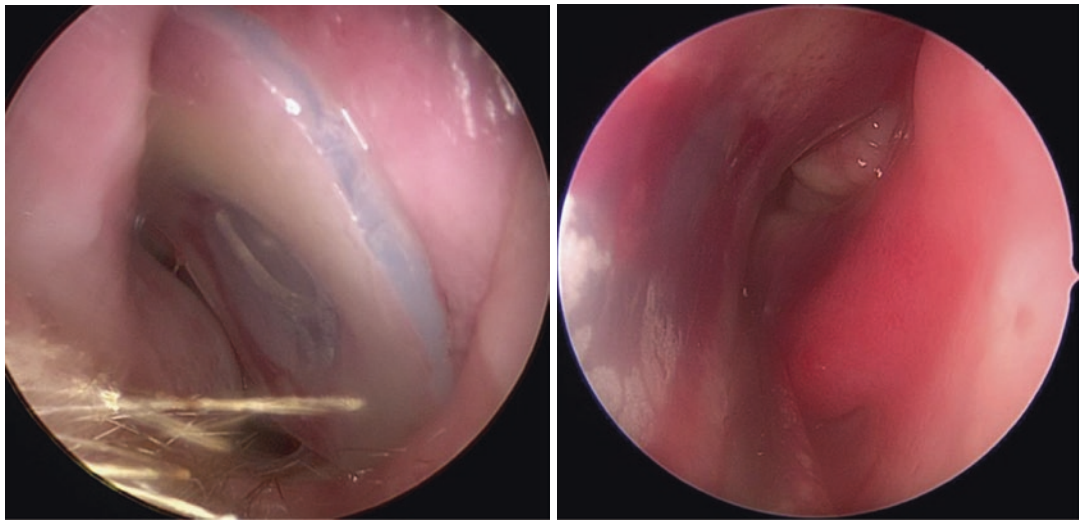

Fig. 76.2 Endoscopic photograph after the insertion of a silicone septal button

based ointments is still necessary to ensure the results of placing the prosthesis are not compromised by crusting and recurrent infections.

Unfortunately, septal buttons have been associated with several complications. Although they are usually placed to treat recurrent nosebleeds, there are instances where they increase the frequency of epistaxis. As already mentioned, with meticulous care, crusted material can collect around the flanges. Improper sizing can cause intranasal pain, granulation tissue on nasal floor, chronic discomfort and dislodgement of the button, especially after sneezing. Finally, movement of the axis may contribute to a steady erosion of the perforation edges and eventual enlargement of the defect. Generally, contraindications are the presence of acute infection with osteitis, chronic septal disease (Wegener), neoplasia and extremely large perforations. Indications for device removal include: chronic pain or discomfort, excessive crusting and epistaxis, biofilm colonization causing infection, need for resizing or need for ongoing cleaning and maintenance.

Regarding long term results, a landmark study depicted a removal rate in 45 patients of 67%, with large perforations and those that are due to septal resection being associated with a poor prognosis. Symptom score improvement for all symptoms except snoring was 55%, but for the main nasal symptoms, it was 70%. A meta-analysis on prosthetics for nasoseptal perforations (10), demonstrated an overall success rate of 65%, and few reports of complications—only one fungal infection and 9 unspecified infections—in 706 cases.

Summary and Author's Comments

1. Inserting a septal button can be an ideal alternative to surgical management, as it is a fast, low cost, minimally invasive procedure that bears excellent results and low failure rates.
2. Regarding conservative treatment, initial management of septal perforation begins with improving nasal hygiene. After a septal button is inserted, it can remain in place for long periods. Yet, this duration is very dependent on the patient's diligence with good nasal hygiene and proper care of the prosthesis.
3. Literature suggests an overall success rate of 65% and a few reports of complications.

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Nasal Vestibule Infection

77

Sarantis Blioskas

77.1 Case Presentation

A 26-year-old female attended the emergency department of the hospital complaining of a five-day course of nasal soreness, located mainly on the left vestibule and radiating to the nasal dorsum and left maxillary area. The patient reported nasal congestion and crusting on the left nostril as well as progressively worsening redness and erythema covering the left vestibule area and extending to the left cheek. Occasional left nasal purulent discharge was also evident. Clinical examination was challenging due to tenderness, yet anterior rhinoscopy and flexible nasendoscopy revealed evidence of vestibule hair folliculitis and boils in the nasal dorsum, covered by crusting and mucopurulent secretions. A nasal swab was obtained and microbiology depicted positive cultures of Methiciline Sensitive Staphylococcus Aureus (MSSA). The rest of the head and neck examination and examination of the cranial nerves were unremarkable and there was no evidence of bacteremia or neurological deficits. Blood work revealed an anticipated raised of WBC and CRP. The patient was treated with a combination of topical mupirocin ointment and salty water nasal irrigation, as well as intravenous Flucloxacillin (1 g QDS). The initial spread of nasal cellulitis and facial extension was marked, and after 24 hours, a significant clinical improvement of both tenderness and spread of erythema was evident. The patient was switched to oral antibiotics for 2 weeks and the clinical course was uneventful.

77.2 Background Knowledge

The nasal vestibule is the most anterior part of the nasal cavity constituting a specialized organ with very characteristic structures and specific functions. The vestibule is surrounded almost entirely by its half of the nasal lobule, bounded laterally by the ala, or wing, medially by the mobile septum and columella, superiorly by the cul-de-sac and limen vestibuli, and posteriorly by the skin lying on the alveolar process of the superior maxilla. Its inferior limitation is the nostril (external naris). Thus, it is enclosed by the cartilages of nose and lined by stratified squamous keratinized epithelium. Inside the vestibule are small hairs called vibrissae, which filter dust and other matter that are breathed in. Within the vestibule, the epithelium loses its keratinised nature and undergoes a transition into typical respiratory epithelium before entering the nasal fossa, so the nasal vestibule is the area where the respiratory tract makes its first contact with the outside world of air.

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Nasal vestibulitis is a localized infection of the hair-bearing nasal vestibule, caused predominantly by Staphylococcus aureus (S.aureus). The area is an ecological niche of Staphylococcus aureus, with estimated carriage rates of 20% in the general population. The vestibule is limited above and behind by a ridge, the limen nasi, over which the skin becomes continuous with the nasal mucous membrane. Apparently, the staphylococcal cells flourish here in the relative absence of human defenses and/or are capable of withstanding the local antibacterial defenses. Although no studies to date have investigated the incidence or prevalence of nasal vestibulitis, it appears to be a common condition, especially in the elderly.

Although generally, nasal vestibulitis is a minor infection, its significance lies on potential complications that may arise, which can be dramatic and even life-threatening. Thus, nasal vestibulitis may be accompanied by mid-face cellulitis and abscess formation, which can require immediate drainage. In case of such a scenario, the anatomical characteristics of the vascular supply of the mid-face becomes of paramount clinical importance. Surgical anatomy indicates the significance of the valveless venous system connections between the facial and angular veins and the superior ophthalmic vein and cavernous sinus. The facial vein is also interconnected with the cavernous sinus via the inferior ophthalmic vein and pterygoid plexus. In this concept, infection from the vestibule area may potentially spread to venous walls and transmitted intracranially, causing cavernous sinus thrombosis and associated intracranial complications. In these paths, the nasal vestibule and generally the mid-facial skin are usually referred to as the "danger area."

77.3 Clinical Approach

Nasal vestibulitis is a common infection affecting mostly the adult population. Evidence depict no sex predilection, while interestingly, there appears to be a side predilection, with vestibulitis and abscess formation favoring the right side. This could be related to right hand predominance and possibly to nasal picking. Childhood representation is generally rare, although one could expect the prevalence of nasal vestibulitis to be higher in children, as nose picking and nose blowing are more frequent in children.

Regarding aetiology, nasal vestibulitis can happen spontaneously, but there are factors that appear to contribute, including minor topical trauma such as nose-picking, hair plucking, excessive nose blowing, and topical nasal steroid therapy. It is hypothesized that the central pathophysiological event that gives rise to the inflammatory cataract is an epithelium breach that serves as the port of entry for infection. A separate patient category involves cancer patients. Systemic, antineoplastic therapy, such as taxane- and VEGF-related therapies, predispose to nasal vestibule infections. It seems than in these patients, the same scheme applies, with disruption of the nasal epithelium, allowing for overgrowth of colonizing organisms. Yet, in addition cancer-directed therapies, which can affect dividing epithelial cells and can cause immunosuppression, and targeted therapies, which have been associated with increased rates of skin and nail infections, increase the risk for nasal vestibulitis. Other predisposing factors range from environmental (e.g., low humidity, altered pH), and individual variables (e.g., smoking, immunosuppression), to medication use (e.g., diuretics, isotretinoin), surgery, infections (e.g., herpes simplex/zoster), and systemic conditions such as systemic lupus erythematosus.

In terms of symptomatology, nasal vestibulitis typically presents with severe pain, redness and swelling of the nasal vestibule and tip. Thick yellow crusting overlying nares and anterior nasal septum and severe tenderness during nasal tip manipulation or anterior rhinoscopy are also typical findings. Signs may also include ulceration, minor bleeding areas and small abscesses. A unique diagnostic sign is the presence of erythema and edema on the skin of nasal tip, which is usually referred to as the Rudolph Sign (as in Rudolph, The Red-Nosed Reindeer). Systemic symptoms and signs are not typical and may include fever and general decline.

Diagnostic workup begins with a history and basic head and neck clinical examination. An index of suspicion is necessary, yet clinical signs are so evident that clinical diagnosis is usually quickly established. Anterior rhinoscopy and flexible nasal endoscopy are required to gain perspective on the extent of the infection. Nasal swabs are routinely taken for culture before commencing treatment. The majority of positive results depict MSSA, as S. aureus nasal colonization has been estimated to be 30% in healthy individuals. However, with the worldwide spread of community-acquired MRSA (CA-MRSA) results may differ, since the skin and soft tissue infection (SSTI) is the most common clinical manifestation of CA-MRSA. Results of cultures are clinically significant since CA-MRSA nasal vestibulitis is associated with a more complicated clinical course and thus warrants more aggressive treatment. Laboratory findings usually indicate a rise in inflammatory markers (WBC, CRP). Imaging is rarely helpful in cases of nasal vestibulitis unless intracranial complications are suspected. In case of cavernous sinus thrombosis, findings of fever, headache, chemosis, proptosis, and cranial nerve III, IV, V, and VI palsies will raise concerns and an MRI and CT venogram should be undertaken.

Treatment of nasal vestibulitis is generally topical, at least for mild cases with a localized skin infection. Mupirocin is the most effective choice for topical treatment. The pain and erythema start to improve within 12 hours after topical therapy. Topical treatment may also include cleaning all the crust from the nasal vestibule, local application of cool compresses, emollients and/or other topical agents (e.g., retapamulin, polysporin, chlorhexidine, saline). In case of signs of a more diffuse infection, systemic antibiotic treatment should be initiated. These usually include antistaphylococcal agents such as ascloxacillin or flucloxacillin, either orally or intravenously. Such a more aggressive approach should also be considered in cases of facial cellulitis or in immunocompromised or diabetic patients, as well as in patients with other risk factors (e.g., hypercoagulability disorders). In case of a local abscess formation, surgical drainage may be necessary.

Most cases of nasal vestibulitis can be treated as outpatients. Nevertheless, admission is indicated in more complicated situations like cellulitis or abscess and cases not responding to treatment. The same applies of course in cases where there is suspicion of intracranial complications like cavernous sinus thrombosis. The assumed risk of intracranial complications associated with midfacial infection is the main reason for close surveillance or admission. Yet, evidence suggests that either these complications can be avoided with appropriate treatment or they are merely rare since they only appear in modern literature as case reports. In the management of intracranial venous thrombosis, anticoagulation with heparin is generally considered, to prevent further thrombosis and reduce the incidence of septic emboli.

Summary and Author's Comments

1. Nasal vestibulitis is a localized infection of the hair-bearing nasal vestibule, caused predominantly by Staphylococcus aureus.
2. Although generally, nasal vestibulitis is a minor infection, its significance lies in potential complications that may arise, which can be dramatic and even life-threatening. In these paths, the nasal vestibule and generally the mid-facial skin is usually referred to as the "danger area."
3. Nasal vestibulitis typically presents with severe pain, redness and swelling of the nasal vestibule and tip (Rudolph sign).
4. Treatment of nasal vestibulitis is generally topical, at least for mild cases. In case of signs of a more diffuse infection, systemic antibiotic treatment should be initiated.

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Part XII

Post-operative Complications

Orbital Injury

78

Marios Stavrakas and Hisham S Khalil

78.1 Case Presentation

A 36-year-old male patient with primary CRS with nasal polyposis was listed for FESS-polypectomy. Prior to surgery, the patient had a CT scan of his sinuses to aid planning. FESS was carried out by a trainee under senior supervision. While performing a right middle meatal antrostomy and anterior ethmoidectomy, fat was seen prolapsing in the nasal cavity after injuring and penetrating the lamina papyracea. Immediately, the senior surgeon took over and continued with the operation. The eye was checked and instrumentation with powered instruments was avoided. Further surgery in this area was abandoned. Postoperatively, the situation was explained to the patient and he had an uneventful recovery.

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78.2 Background Knowledge

In case of injury to the lamina papyracea as described above, it is crucial to identify the problem immediately. In our practice, we believe it is important to avoid any further use of powered instruments in the area of fat prolapse. Uncontrolled dissection there may result in further injury of the orbital contents, bleeding or damage to the medial rectus or the superior oblique muscles. It is also essential to assess the pupil and the orbit and make sure there is no dilation or orbital haematoma. We prefer light packing of the nasal cavity with a dissolvable pack. Postoperatively, it is essential to explain the situation to the patient, instruct them to avoid blowing the nose and warn them about potential periorbital ecchymosis. We monitor the eye observations and if there are any concerns regarding visual acuity or ophthalmoplegia/diplopia, early Ophthalmological assessment needs to be requested. If the patient undergoes revision surgery, it is necessary to repeat the CT scan of the sinuses preoperatively.

78.3 Clinical Approach

The paranasal sinuses are in very close proximity to the orbits, especially the ethmoid sinuses, and this anatomical relationship exposes the orbital contents to the risk of trauma during endoscopic sinus surgery. Orbital injury during endoscopic

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sinus surgery can result in orbital haematoma or trauma to medial rectus or superior oblique muscles. It is usually secondary to violation of the lamina papyracea and, at his time, orbital fat prolapses into the nasal cavity. This can be confirmed by applying pressure to the eye globe, a manoeuvre which transmits movement at the area of the suspected orbital penetration. The problem is that muscular trauma results in diplopia, despite any efforts to repair the injury. In case the lamina is accidentally penetrated, we should avoid using powered instruments, to prevent any further damage to the contents of the orbit. A minority of those patients develop postoperative periorbital ecchymosis, which settles with time.

Optic nerve damage is a rare complication and may result in loss of vision, directly postoperatively, or can have a delayed presentation. It can be caused by direct injury to the nerve (for example, in an Onodi cell) or indirectly. We should inspect the pupil intra- and postoperatively and suspect an optic nerve injury if it dilates rapidly during (either from globe ischaemia or from damage to pupillomotor nerves). Moreover, postoperative severe visual loss with a poorly reactive pupil and a relative afferent pupil defect are indicators of optic nerve damage. In such scenarios, we must involve an Ophthalmologist as soon as possible.

Avoiding any accidental injuries to the lamina is the best way of prevention. The surgeon must spend enough time studying the imaging preoperatively, identify the lamina intraoperatively, and if a small crack is suspected, the bulb-press test is a method to confirm this type of injury. Of course, in some cases where the anatomy is complex, image guidance can also help to prevent such problems.

Summary and Author's Comments

1. Orbital trauma can result in serious sequelae; therefore, prevention is the best way to approach the problem.
2. A careful study of the anatomy (Onodi cells, dehiscent lamina papyracea) is extremely important.
3. Meticulous dissection around the lamina papyracea, especially by unexperienced endoscopic surgeons, will also help to avoid any disastrous complications.
4. Early involvement of the Opthalmology team and continuous assessment of the vision for any deterioration is the first step in an unfortunate case of optic nerve injury.

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79 Post Adenoidectomy Haemorrhage

Marios Stavrakas and Hisham S Khalil

79.1 Case Presentation

A 7-year-old girl was brought back to the emergency department after undergoing adenoidectomy earlier on the day. Her parents were concerned when they noticed a relatively minor nosebleed. The patient was initially assessed in ED according to the ALS protocol. All observations were normal and there was no significant drop in Hb. It was decided to take the patient back to theatre for Examination Under Anaesthesia (EUA) and arrest of post adenoidectomy bleed. Postoperative recovery was uneventful, and the patient was discharged on the following day.

79.2 Background Knowledge

When approaching such a patient, the clinician should adopt a systematic assessment practice, according to APLS/ATLS principles. It is important to keep the child calm and take a brief history focusing on the time of onset, duration of bleeding, date of the surgery, surgical technique (if known) and indication for the primary surgical intervention. It is also essential to enquire about clotting disorders and other medical problems that may affect our efforts to achieve haemostasis. The clinician should attempt to stabilize the patient, fluid resuscitate them and monitor the observations closely. The next decision to make is about the necessity and timing of surgical intervention to arrest the bleeding. In our unit, we prefer the same technique as in primary adenoidectomy, by using monopolar suction diathermy. The bleeding usually comes from aberrant vessels or remnants of adenoidal lymphoid tissue. In extreme situations, when bleeding cannot be stopped, an option is to place posterior packs, keep the child intubated and try to address other contributing factors, for example, clotting disorders. A useful adjunct is tranexamic acid, which can be used in the absence of any contraindications.

79.3 Clinical Approach

The upper two-thirds of the pharynx are supplied by the ascending pharyngeal artery, which is usually the responsible vessel for post adenoidectomy haemorrhage. On some occasions, an aberrant vessel is responsible for the bleeding. It is a rare com-

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plication (about 0.5% of adenoidectomies) and predominantly occurs on the day of the surgery. Children with disordered clotting may have a higher risk of delayed bleeding and require close monitoring in the postoperative period.

Again, prevention and careful pre-operative assessment is the best way to avoid complications. We should pay attention to thorough history taking, aiming to identify clotting disorders and appropriate pre-operative workup, if indicated. When the patient is anaesthetised, it is worth examining the postnasal space to exclude any tumours (e.g., angiofibroma) but also check for any aberrant vessels.

Our approach in case of this complication should be systematic and thorough. Initial assessment, according to ATLS/APLS guidelines, should be followed by the decision about operating. Under general anaesthesia, we remove any clots and look for any actively bleeding vessels or adenoidal tissue remnants. Our preferred practice is to use a nasopharyngeal mirror and suction diathermy to arrest the haemorrhage. If this is not successful, packing the postnasal space is an option. This will most probably require ITU stay, as postnasal space packing is not tolerated very well by children. With regards to the timing of removal, this varies from 4 h to 12 h (overnight), followed by an assessment of haemostasis. The clinician must seek an expert opinion from a Haematologist if clotting problems are suspected. Finally, if all the above do not achieve adequate haemostasis, we should consider the role of interventional radiology.

Summary and Author's Comments

1. The surgeon needs to assess the patient carefully according to ATLS/APLS principles and make arrangements to stabilise them before going to theatre.
2. We follow a stepwise approach, with our first aim being to arrest the bleeding in theatre. Other steps may be followed if the bleeding is persistent.
3. Haematology or Paediatric input must be considered in cases where clotting/systematic disorders are suspected.

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Orbital Haematoma

80

Marios Stavrakas and Hisham S Khalil

80.1 Case Presentation

A 42-year-old man was referred to the ENT department by Ophthalmology for a biopsy of a right intra-orbital, extraconal lesion. (Figs. 80.1 and 80.2). The patient had existing visual symptoms, and it was anticipated they would develop intra-orbital bleeding/haematoma as a result of the biopsy. The patient was consented for a planned lateral canthotomy and cantholysis. This was not required as the patient did not develop any significant bleeding and the medial orbital compartment was decompressed by removing the lamina papyracea. The histology was confirmed as a low-grade lymphoma.

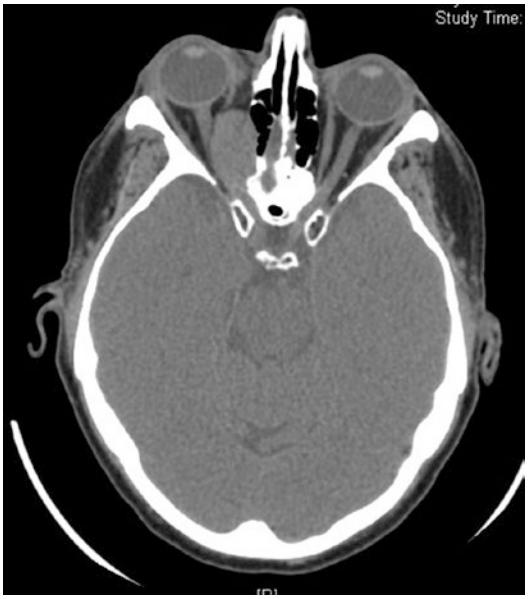

Fig. 80.1 Axial CT scan of the paranasal sinuses demonstrating an intra-orbital, extraconal tumour

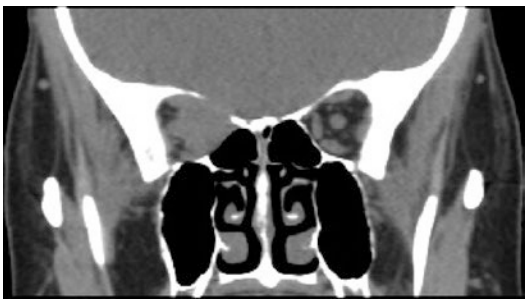

Fig. 80.2 Coronal CT scan of the paranasal sinuses demonstrating the same lesion

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80.2 Background Knowledge

Orbital hematoma is a serious complication of endoscopic sinus surgery, which may lead to blindness if prompt action is not taken. It can be caused either by venous or arterial bleeding, depending on the area of the injury. Thus, when the lamina papyracea and periorbita are breached, we can get a venous haematoma and arterial, when the anterior (rarely the posterior) ethmoid artery is transected and retracted into the orbit.

A small amount of blood in orbit (as little as 5 mL) can acutely increase intraorbital pressure to dangerous levels, with dramatic consequences to the visual acuity. A slow venous hematoma may never accumulate enough blood to become a problem (60 to 90-minute window). In contrast, an arterial hematoma accumulates faster and can increase the intracranial pressure to dangerous levels in only 15 to 30 minutes.

For this reason, the eyes are kept exposed during surgery and monitored for any dilation of the pupil or movement of the globe, especially when using powered instruments. Some signs that indicate potential orbital hematoma are ecchymosis, lid oedema with eye closure, proptosis, chemosis, profound hardness or firmness of the globe on palpation, and a dilated pupil.

If a surgeon has concerns during an endoscopic sino-nasal procedure, they should stop operating and remove any packing material if necessary. If the haematoma is expanding, there is a need to ask an Oculoplastics surgeon for help urgently. In an emergency, the sinus surgeon should proceed to lateral canthotomy and inferior cantholysis immediately. If there is adequate expertise, the surgeon should also decompress the obit endoscopically. Regular eye observations are required in the postoperative period, as instructed by the Ophthalmologist.

80.3 Clinical Approach

80.3.1 History

Orbital hematomas are more likely to occur during endoscopic tumour resections that involve the orbit. The risk is also higher in patients where the lamina papyracea has been breached by other pathologies such as mucoceles, diffuse sinonasal polyposis or previous endoscopic sinus surgery.

80.3.2 Examination

The prevention of orbital haematoma is best achieved by regular observation of the eye globe during endoscopic sinus surgery and planning for elective canthotomy and inferior cantholysis if the surgeon anticipates a high risk of orbital hematoma, as was the case in this patient. Bruising around the eye, sub-conjuctival haemorrhage, proptosis and increased intra-orbital pressure assessed by balloting are all signs of an orbital haematoma and orbital injury. Once the patient is stabilised through a lateral canthotomy/endoscopic decompression of the orbit the patient needs to have a comprehensive Ophthalmological assessment once they have recovered from a general anaesthetic.

80.3.3 Investigations

Careful assessment of pre-operative imaging may help predict a high risk of orbital haematoma as was the case in this patient with an intra-orbital tumour. (Figs. 80.1 and 80.2) If an orbital haematoma occurs and following emergency treatment and ophthalmological assessment, a postoperative CT scan is important to assess the extent of orbital injury.

80.3.4 Treatment

The anaesthetist should be alerted to the occurrence of an orbital haematoma to reduce the systemic blood pressure. Medical treatment to decrease intra-orbital pressure includes intravenous mannitol and acetazolamide, a carbonic anhydrase inhibitor. The head of the surgical table is also raised to reduce venous bleeding. Further treatment includes lateral canthotomy/inferior cantholysis and decompression of the orbit, depending on the surgeon's expertise. Early involvement of the Ophthalmologist is adviced.

Summary and Author's Comments

1. As with most complications, prevention and careful pre-operative preparation is the key to avoid them.
2. Again, a stepwise approach is recommended: identification→ call for help (Ophthalmologist, senior Rhinologist) → Lateral canthotomy& inferior cantholysis +/− medical treatment→ Removal of packing if that is necessary→ Orbital decompression (endoscopic vs. external approach) → Postoperative care

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Septorhinoplasty Complications

81

Marios Stavrakas and Hisham S Khalil

81.1 Case Presentation

A 38-year-old female patient was referred urgently by her general practitioner. Over the last week, she developed erythema, tenderness around the tip of her nose. A swab showed heavy growth of Pseudomonas. The patient had undergone septorhinoplasty elsewhere 10 years ago, with the use of alloplastic grafts. Our first priority was to remove the infected graft and explained to the patient that this may compromise the tip support, affect the shape of her nose and eventually may need revision surgery. The implant was removed successfully via a closed approach, the patient had an excellent recovery and further revision was not deemed necessary.

81.2 Background Knowledge

Septorhinoplasty is a complicated procedure that carries several potential complications, with post-operative deformities causing revision surgery in 5% to 15% of the cases (Table 81.1). Beyond the structural, functional, or aesthetic problems, it is of paramount importance to consider the psychological impact that such a complication may have on the patient. The following table gives a brief overview of the complications associated with this operation.

We believe that many of the complications, as mentioned above, can be avoided with careful patient selection, detailed pre-operative consultation, employing teach-back techniques and meticulous pre-operative planning. It is essential to assess any rhinoplasty candidates for body dysmorphic disorder and request an opinion from a clinical psychologist if needed. Important factors to consider are also the graft material selection (we prefer autologous cartilage graft or irradiated rib grafts) and post-operative care.

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Table 81.1 Basic characteristics of the most common postoperative complications of septorhinoplasty

Type of complication	Characteristics
Infection	Septal abscess, toxic-shock syndrome, dacryocystitis, intracranial infection following trauma to the skull base, infected grafts, reaction to suture material
Complications related to grafts	Infection, extrusion, foreign body reaction
Functional	Failure to improve the nasal airway, residual septal deviations, nasal valve stenosis, alar collapse
Psychological	
Structural - aesthetic	Polly beak-deformity = deep radix, over-resection of the bony dorsum, a prominent cartilaginous dorsum, as well as a drooping tip→ curved profile from dorsum to tip Over resection of the nasal dorsum - saddle nose deformity irregularities following humpectomy/ open roof deformity resulting in a wide nasal bridge Asymmetry of the nasal base Unfavourable rotation of the tip, hanging columella, alar retraction Loss of tip support due to distortion of the tip support mechanisms and failure to repair them Alar collapse
Wound healing problems	

81.3 Clinical Approach

81.3.1 History

There is a number of potential complications that has to do with the complexity of septorhinoplasty as an operation. History should focus on the previous surgery (primary/revision), the type of complication (infection/functional/aesthetic) and the time from surgery (primary/secondary). We should not forget to explore the psychological impact, as sometimes this is the patient's main problem.

81.3.2 Examination

Inspection, palpation and anterior rhinoscopy are usually enough to identify the problem. If the cardinal symptom is nasal obstruction, nasendoscopy may give us more information. We need to assess the skin envelope (sound breakdown/infection/foreign body reaction), the nasal valve area (collapse), and the septum (haematoma/abscess). Also, we should evaluate the tip support and always examine the nose for any significant deformities (polly-beak, saddle nose, wide dorsum, asymmetries).

81.3.3 Investigations

Swabs are taken in case of infection. PNIF or rhinomanometry are adjuncts in the evaluation of the nasal airway patency. We believe it is essential to take photographs, especially if revision surgery is planned.

81.3.4 Treatment

Treatment depends on the type of complication, with revision rhinoplasty remaining one of the main surgical options. Technical details are beyond the scope of this book.

Summary and Author's Comments

1. Careful patient selection and preoperative planning may save the clinician from unfavourable outcomes.
2. We advocate a cooling-off period between consultations, screening for body dysmorphic disorder and setting of realistic expectations, which should be confirmed with teach-back techniques and followed by clear documentation.
3. The surgeon should have adequate training before taking on rhinoplasties, not only to avoid any complications but also to be in a position to correct any abnormalities.

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