



Nose and Paranasal Sinuses

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1 Introduction

Nose and paranasal malignancies refer to a wide variety of cancers seen in the nose and adjacent paranasal regions. But less than 1% of the malignant tumours identified in the head and neck area are neoplasms. These tumours typically have mild first signs, making early detection challenging. Because of this, the time between the onset of symptoms and the final diagnosis is often 6 months. Furthermore, these tumours have a variety of histological presentations, making diagnosis difficult. Nasal and paranasal malignancies are grouped by the World Health Organization (WHO) into 44 unique histological categories, which are roughly classified into epithelial and non-epithelial groups [1], as given in Table 1.

Information on surgery and histological distribution of benign sinonasal tract tumours from 1298 patients who had treatment at the university hospitals in Brescia and Varese [2] is described in Table 2.

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Table 1 Paranasal sinus malignancy histologic classification by the World Health Organization [1]

Epithelial malignancies
Squamous cell carcinoma
• Verrucous carcinoma
• Basaloid squamous cell carcinoma
• Papillary squamous cell carcinoma
• Spindle cell carcinoma
• Adenosquamous carcinoma
• Acantholytic squamous cell carcinoma
Lymphoepithelial carcinoma
Sinonasal undifferentiated carcinoma
Salivary gland-type carcinomas
• Adenoid cystic carcinoma
• Acinic cell carcinoma
• Mucoepidermoid carcinoma
• Clear cell carcinoma not otherwise specified
• Epithelial-myoepithelial carcinoma
• Myoepithelial carcinoma
• Carcinoma ex pleomorphic adenoma
• Polymorphous low-grade adenocarcinoma
Adenocarcinoma
• Intestinal-type adenocarcinoma
• Non-intestinal-type adenocarcinoma
Neuroendocrine tumours
• Typical carcinoid
• Atypical carcinoid
Small cell carcinoma, neuroendocrine type
Soft tissue malignancies
Fibrosarcoma
Malignant fibrous histiocytoma
Rhabdomyosarcoma
Leiomyosarcoma
Angiosarcoma
Malignant peripheral nerve sheath tumour
Bone and cartilage malignancies

(continued)

Table 1 (continued)

Chordoma
Chondrosarcoma
Mesenchymal chondrosarcoma
Osteosarcoma
Hematolymphoid malignancies
Langerhans cell histiocytosis
Extranodal natural killer/T-cell lymphoma
Diffuse large B-cell lymphoma
Extramedullary plasmacytoma
Extramedullary myeloid sarcoma
Histiocytic sarcoma
Neuroectodermal malignancies
Ewing sarcoma
Primitive neuroectodermal tumour
Olfactory neuroblastoma
Melanotic neuroectodermal tumour of infancy
Mucosal malignant melanoma
Germ cell malignancies
Teratoma with malignant transformation
Sinonasal teratocarcinosarcoma

Accurately staging nose and sinus tumours is difficult despite the advancements in endoscopy and the widespread availability of modern imaging methods such as MRI and CT scans. Surgery and radiation are the two main treatments; however, these treatments can cause significant health issues including facial deformation, difficulty in chewing and in some cases, blindness. Sinonasal malignancies have a worse prognosis compared to other head and neck region malignancies, making quality of life considerations an important factor when assessing treatment options.

Table 2 Benign tumours of the sinonasal tract by histology and surgical approach [2]

Tumour histology	Surgical approach			Total
	Endoscopic	Combined	External	
Inverted papilloma	592	75	12	679
Osteoma	115	54	13	182
Juvenile angiofibroma	147	1	6	154
Lobular capillary hemangioma	68	0	0	68
Fibrous dysplasia	58	3	1	62
Schwannoma	16	3	0	19
Cavernous hemangioma	16	1	0	17
Ossifying fibroma	11	0	0	11
Hamartoma	46	0	0	46
Glioma	8	3	1	12
Pleomorphic adenoma	7	0	0	7
Miscellaneous	33	8	0	41
Total	1117	148	33	1298

2 Surgical Anatomy of the Nose and Paranasal Sinuses

The paranasal sinuses within the nose have extensive and complex architecture. It is essential to have an understanding of the intricate surgical anatomy due to the close proximity of these structures to crucial components such as the orbit and brain. Here is a brief overview.

2.1 Ethmoidal Sinus

- The ethmoid bone is divided into two parts in the midline that looks like a labyrinth. The basal lamina of the middle turbinate further divides this into anterior and posterior cell groups.
- The ethmoidal sinus is made up of a fragile skull base, and lamina papyracea forms its superior and lateral limits.
- The emissary vein, which is anterior to crista galli, and the olfactory fibres in the cribriform region form the path of spread to the anterior cerebral fossa.
- The distance in millimetres from the anterior ethmoidal foramen to the anterior lacrimal crest (24 mm), from the posterior ethmoidal foramen to the anterior ethmoidal foramen (12 mm) and from the optic nerve to the posterior ethmoidal foramen (6 mm) is an established formula for identifying the anterior ethmoidal artery, posterior ethmoidal artery and the optic nerve.
- The cribriform niche can range from 15.5 to 25.8 millimetres in length and from 0 to 15.5 millimetres in depth.
- During a craniofacial resection, the dura should be carefully removed since it is closely applied to the cribriform region.

2.2 Sphenoid Sinus

- The internal carotid artery and optic nerve are clearly visible on the lateral wall of the sphenoid sinus.

- In 20% of cases, the lateral wall has a varying depth, with an opticocarotid recess that is dehiscent.
- Relation:
 - Cavernous sinus, pterygoid canal and foramen rotundum that press into the sinus cavity laterally.
 - The pituitary gland is situated above.
 - The cells of the posterior ethmoidal sinus that extend lateral and superior to the sphenoid sinus are known as the poster superior onodi cell. It is more vulnerable to injury during instrumentation when it is present because the carotid artery and optic nerve are frequently accessible in its lateral wall.

2.3 Frontal Sinus

- Varying in size and form.
- The front ethmoidal cell influences the drainage to the middle meatus, resulting in an hourglass-shaped structure rather than a duct.

2.4 Maxillary Sinus

- The inferior turbinate, the lacrimal bone, the bulla of the ethmoidal sinus, the uncinat process, the perpendicular plate of the palatine bone and the nasal cavity itself form its medial wall.
- The maxillary sinus is related to the cheek anteriorly.
- Moreover, premolar and molar roots connect them inferiorly to the mouth cavity.
- Furthermore, the pterygoid plates, part of the sphenoid bone, form its posterior relation.
- Additionally, the pterygomaxillary fissure, which contains the terminal portion of the maxillary artery, is related posteriorly to the sinus.
- Inside the pterygopalatine fossa is the maxillary nerve, along with the pterygopalatine ganglion in its neural compartment, as well as the maxillary artery's terminal branch and its branches in its vascular compartment.

- Lastly, the infratemporal fossa is located between the pharynx's wall and the mandible's ascending ramus and contains the branches of the mandibular nerve, the lateral pterygoid muscles, the pterygoid venous plexus and the maxillary artery.

2.5 Orbit

- The medial wall comprises a thin bone called lamina papyracea, while the inferior is thin and linked to the maxillary sinus.
- An inferior orbital fissure links the pterygo-palatine fossa medially and the infratemporal fossa laterally.
- The superior orbital fissure connects to the cavernous sinus.

2.6 Nasal Septum

- Composed of the ethmoid's perpendicular plate, vomer and quadrilateral cartilage.

2.7 Lymphatic Drainage

- Moderately poor, it drains from the nose and paranasal sinuses to the jugulodigastric and retropharyngeal nodes.
- From the vestibule into the cervical group of lymph nodes through the anterior septum and columella.

3 Pattern of Tumour Spread

The distribution of the nose and paranasal sinus involvement by malignancy is as follows:

- The maxillary sinus is the most common site of nose and paranasal malignancy, accounting for 50–70% of the cases.
- The nasal cavity is the second most common site, accounting for 15–30%.
- The ethmoidal sinus is the third most common, accounting for 10–20%.

- Frontal sinus and sphenoidal sinus are the least common sites, accounting for less than 1%.

3.1 Local Spread

- As the size of the malignant lesion increases, it consumes the sinus in which it is located and erodes any surrounding bone.
- The dura, periosteum and perichondrium act as a barrier to limit the spread of the tumour, but further growth may involve these structures.
- If the dura, periosteum and perichondrium are breached, the tumour may spread to the intracranial and intra-orbital areas.

3.2 Regional Spread

- At presentation, approximately 10% of patients have metastasized lymph nodes.
- In cases where the lesion is centrally situated, both bilateral groups of lymph nodes may be impacted.

3.3 Distant Metastasis

- Adenocarcinoma has a higher chance of distant metastasis compared to squamous cell carcinoma, at 20% and 10%, respectively.
- Common sites for distant metastasis are the bone, brain, liver, lung and skin.

4 Clinical Evaluation

4.1 Endoscopy

It is essential to have a comprehensive endoscopic examination of the nasal cavity if someone is thought to have a malignant tumour in the nose or paranasal sinuses. These tumours may be visible due to their ulcerated form or may be difficult to identify in endoscopy. The tissue around the tumour may also become swollen, resulting

in a polypoid response. However, a biopsy in an outpatient clinic may not yield any information. Therefore, it is recommended to consult with a specialized medical professional and receive additional testing, if needed, to accurately determine the presence of a tumour.

4.2 Imaging

The imaging modality accurately identifies the characteristics of the disease, such as if there is bony erosion, involvement of the skull base, intracranial involvement, perineural invasion and orbital extension, in order to stage the extent of the disease.

4.3 Computed Tomography (CT)

- Bone erosion can be accurately determined by CT scan.
- CT is inadequate for accurately determining the disease's stage.
- It can be hard to tell apart from post-obstructive change, periorbital involvement and dural involvement using a CT scan.
- To gain a better understanding of the anatomy and extent of the disease in the nose and paranasal sinuses, axial and coronal images are necessary.

4.4 Magnetic Resonance Imaging (MRI)

- Precontrast T1-weighted MRI imaging can provide information on the extent of disease involvement in the soft tissue. It is important to assess any periorbital, infratemporal and cerebral space involvement.
- Postcontrast T1-weighted MRI with fat saturation may be useful to darken lipid-rich regions such as the periorbita and can show any perineural invasion of the geniculate ganglion and skull base foramina.
- T2-weighted MRI imaging can be used to re-evaluate the tumour size after CT scans and

identify secretions and oedema that may have been overlooked.

- Additional sequences such as FLAIR or diffusion-weighted imaging may be necessary depending on the tumour.

4.5 Positron Emission Tomography (PET)

- Positron emission tomography (PET)/CT enables accurate localization of malignant tumours by combining functional and structural imaging.
- The procedure relies on the uptake of 18-fluorodeoxyglucose (18-FDG) by cancer cells, which have higher levels of glucose transporters and hexokinase than normal cells.
- However, PET/CT is limited in its ability to interpret results due to inflammation in the sinonasal area and is used for pre-treatment staging and post-treatment monitoring for metastases.

4.6 Biopsy

A tissue diagnosis is essential when examining tumours. If a biopsy is performed in an outpatient clinic, it is important to have the necessary resources on hand to address any potential bleeding. Under general anaesthesia, biopsies increase the likelihood of a successful diagnosis and allow for the collection of samples directly from the sinus. If the Caldwell-Luc method has the potential to spread the tumour or complicate a future resection, it should not be used.

5 Surgical Approach

5.1 Endoscopic Approach

- The endoscopic approach was initially introduced for inflammatory conditions in the 1980s, and the technique gradually evolved for skull base and tumour surgery.

- Drills, debridors and bipolar diathermy are used during dissection in addition to the usual endoscopic sinus tools.
- The endoscopic method is of great use when the disease is limited to the nose and paranasal sinus.
- Endoscopic methods are contraindicated when the orbital contents, frontal sinus, dura lateral to the ethmoidal roof or nasal bones are involved; however, these techniques can occasionally be paired with transcranial approaches or orbital exenteration.

5.2 Open Approach

To facilitate the various bony resections, it is necessary to use the appropriate soft tissue access procedure, of which three are commonly described:

- Lateral rhinotomy.
- Weber-Fergusson.
- Midfacial degloving.

5.3 Combined Approach

It involves both the endoscopic and the open approach to facilitate the excision of the locally advanced extensive lesion of the nose and paranasal sinuses.

most common in adolescent male patients [4]. The lateral nasopharynx is where the lesions develop, and they are hormonally sensitive [4]. The internal maxillary artery (IMA) is the source of their blood supply. Despite being a benign tumour, juvenile nasopharyngeal angiofibroma can seriously harm nearby tissues by local invasion.

An examination of 120 cases from the Mayo Clinic characterized the clinical characteristics of juvenile nasopharyngeal angiofibroma [5]. The typical patient was 15 years old (range 7–29 years). The most typical symptoms included nasal obstruction, epistaxis and nasal discharge; OME and reduced hearing can be present [4–6]. Although the tumours may protrude into the nasal cavity, they are frequently only detectable through a nasal cavity and nasopharyngeal examination by an endoscope [5].

CT or MRI with contrast is typically used to confirm the diagnosis and reveals an enhancing mass with involvement and enlargement of the pterygomaxillary fissure [5, 7].

Depending on the extent of the condition, many categories are used to stage it, some of which are shown in Tables 3 and 4.

Different modalities of imaging are shown in Figs. 1, 2, 3, 4 and 5.

Intranasal biopsy of these lesions should be avoided because of the risk of life-threatening bleeding [8].

6 Angiofibroma

Angiofibroma is a highly vascular, histologically benign lesion that can induce severe epistaxis in juvenile nasopharyngeal angiofibroma [3]. It is

6.1 Case 1

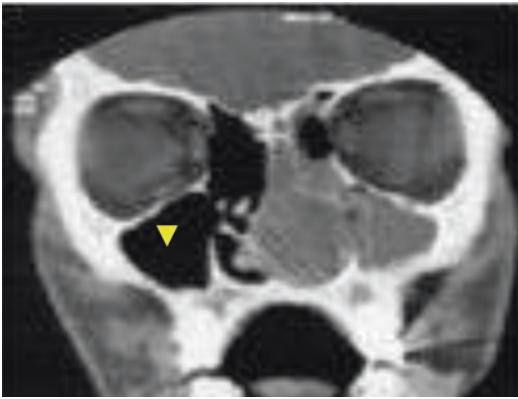
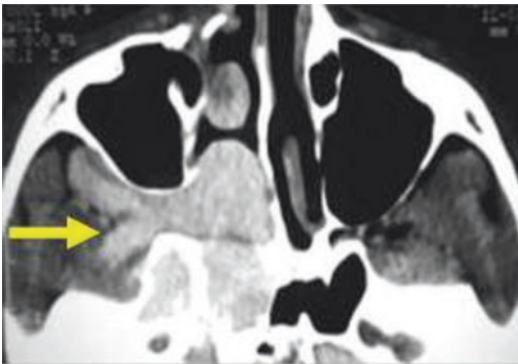
A 19-year-old male patient presented with a 4-month history of self-limiting nasal bleeding. Contrast-enhanced CT and MRI scan shows an enhancing lesion in the right nasal cavity with

Table 3 Andrew's classification (modified Fisch) of JNAs

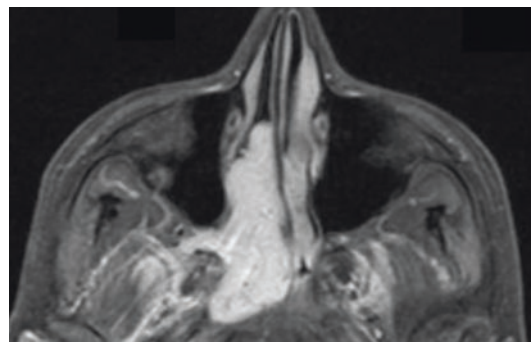
Stage	Description
I	Tumour limited to the nasal cavity and nasopharynx
II	Tumour extension into the pterygopalatine fossa, maxillary, sphenoid or ethmoid sinuses
IIIa	Extension into the orbit or infratemporal fossa without intracranial extension
IIIb	Stage IIIa with small extradural intracranial (parasellar) involvement
IVa	Large extradural intracranial or intradural extension
IVb	Extension into the cavernous sinus, pituitary or optic chiasm

Table 4 Radkowski's classification (modified session's classification) of JNAs

Stage	Description
Ia	Limited to the nose and nasopharynx area
Ib	Extension into one to more sinuses
IIa	Minimal extension into the pterygopalatine fossa
IIb	Occupation of the pterygopalatine fossa with or without orbital erosion
IIc	Infratemporal fossa extension with or without cheek or pterygoid plate involvement
IIIa	Erosion of the skull base (middle cranial fossa or pterygoids)
IIIb	Erosion of the skull base with intracranial extension with or without cavernous sinus involvement

**Fig. 1** CT scan in the coronal plane shows the extension of the lesion into the nasal cavity and the maxillary sinus**Fig. 2** Lesion originating from the sphenopalatine foramen and involving the nasopharynx, PPF and ITF/stage IIc

limited extension to the pterygopalatine fossa. Patient was planned for excision by endoscopic approach.

**Fig. 3** MRI shows the lesion involving the nose and extending into the pterygopalatine fossa/stage II

6.1.1 Operative Technique

- Cottonoids soaked in lignocaine and mixed with an adrenaline solution should be used to sufficiently clear the nasal cavity due to congestion.
- The inferior turbinate, middle turbinate and axilla of the middle turbinate should all be identified before the zero-degree Hopkins endoscope is inserted into the nasal cavity.
- Good exposure without injuring the surface of the tumour.
- Middle turbinectomy, ethmoidectomy, broad anrostomy or sphenoidotomy are procedures that can be used to accomplish exposure.
- Establishing the proper dissecting plane between the angiofibroma and the surrounding tissue.
- Removing the posterior maxillary wall to reach the infratemporal and pterygopalatine fossae.

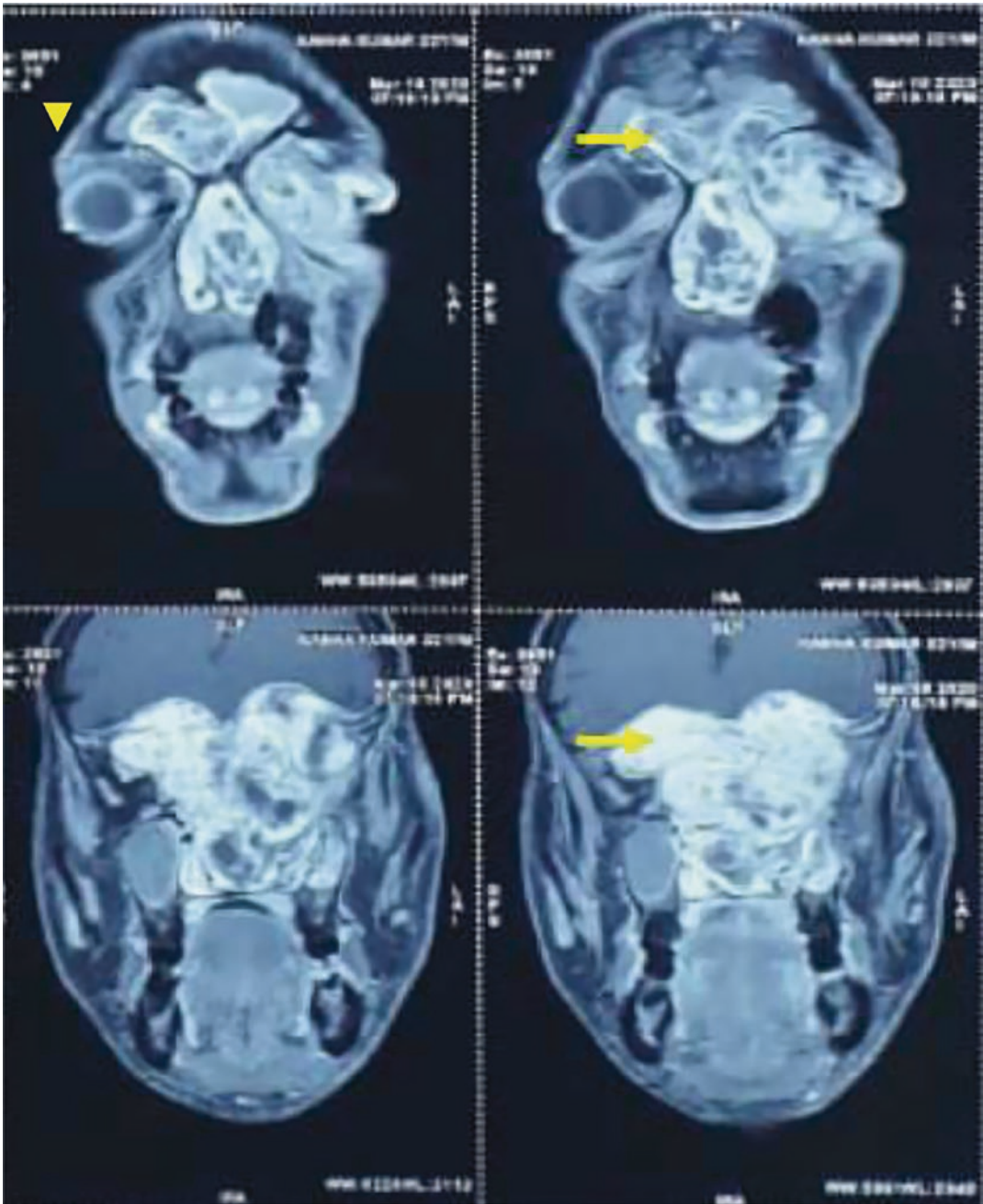


Fig. 4 MRI scan: a coronal plane shows the lesion with intracranial extension

- If a posterior septectomy is necessary to provide for four-hand method exposure to the nasopharynx.
- Identifying and managing the maxillary artery.
- Haemostasis can be achieved by:
 - Irrigation by warm saline.
 - Packing with cottonoids.
 - Using haemostatic material like fibrillar and Surgicel.
- Drilling of the pterygoid root—To prevent recurrence.

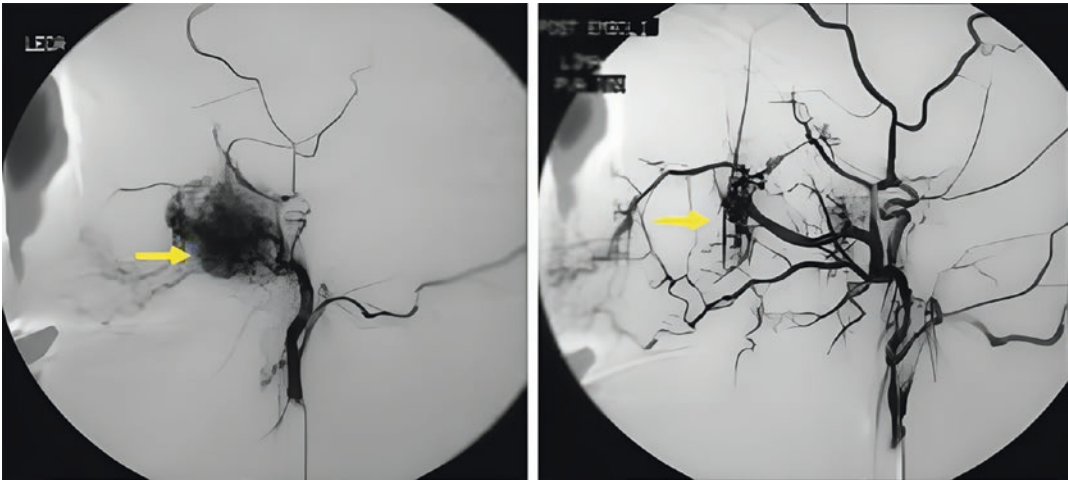


Fig. 5 DSA showing tumour blush, feeder branch from IMA (a), and tumour disappears following embolization (b)

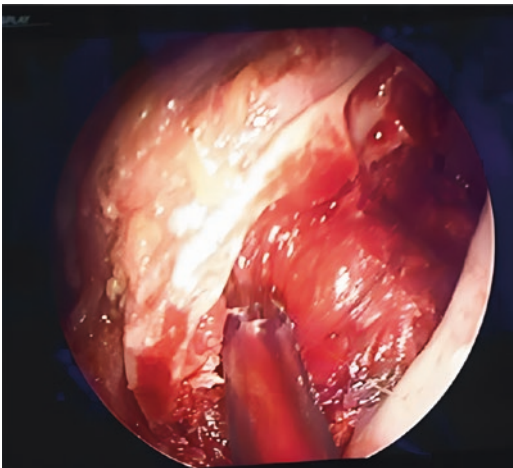


Fig. 6 Creating a plane between the lesion and the surrounding normal mucosa of the nasal cavity by blunt dissection



Fig. 8 Excised specimen of angiofibroma

- Few steps of endoscopic surgery are depicted in Figs. 6, 7 and 8.

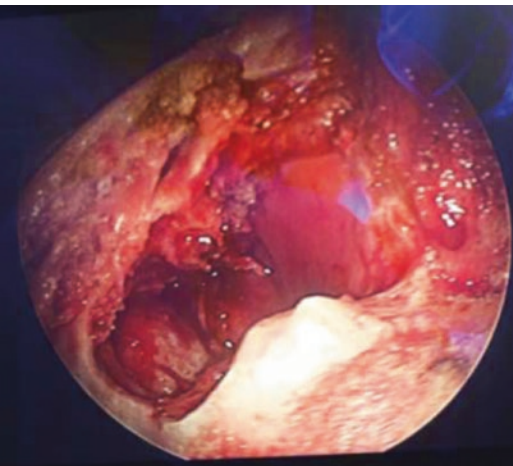


Fig. 7 Endoscopic view of the nasal cavity after the excision of the lesion

6.2 Case 2

A 17-year-old male presented with intermittent nasal bleeding for 3 months and a mass protruding from the right nostril for 1 month. Contrast-enhanced CT and MRI scan shows an enhancing lesion in the right nasal cavity with extension to the pterygopalatine fossa and no intracranial or intra-orbital extension. The patient was planned for excision by open approach by Weber-Ferguson incision.

6.2.1 Operative Technique: Caveats

- The maxilla is better exposed by the Weber-Ferguson incision.
- To ensure cosmesis, the transverse limb should be positioned near the lid edge, often in the first crease.
- To reduce postoperative oedema, any lateral extension into a crow's foot should travel in an inferolateral orientation.
- It is beneficial to bend the incision forward across the nasal bones in the medial canthal area, where there is the highest risk of skin loss as a result of radiation, for further support.
- The mucosal incision runs anteriorly across the alveolus and turns laterally at the intersection of the hard and soft palates, passing behind the maxillary tuberosity.
- The incisions in the soft tissue are all gradually dissected free when the face skin flap is elevated on a submuscular plane.
- Haemostasis is achieved.
- The nasal and maxillary cavity is packed with ribbon gauze.

Pre-operative imaging, surgical steps and follow-up of the same patient are depicted in Figs. 9, 10, 11, 12, 13, 14, 15, 16 and 17.

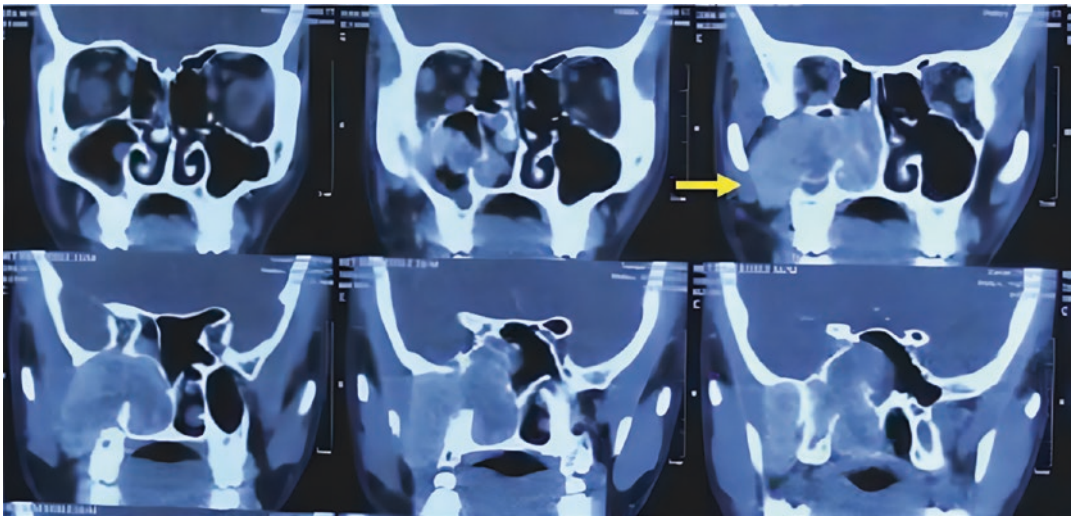


Fig. 9 Hondousa sign widening of the gap between the ramus of the mandible and body of the maxilla because of the lesion

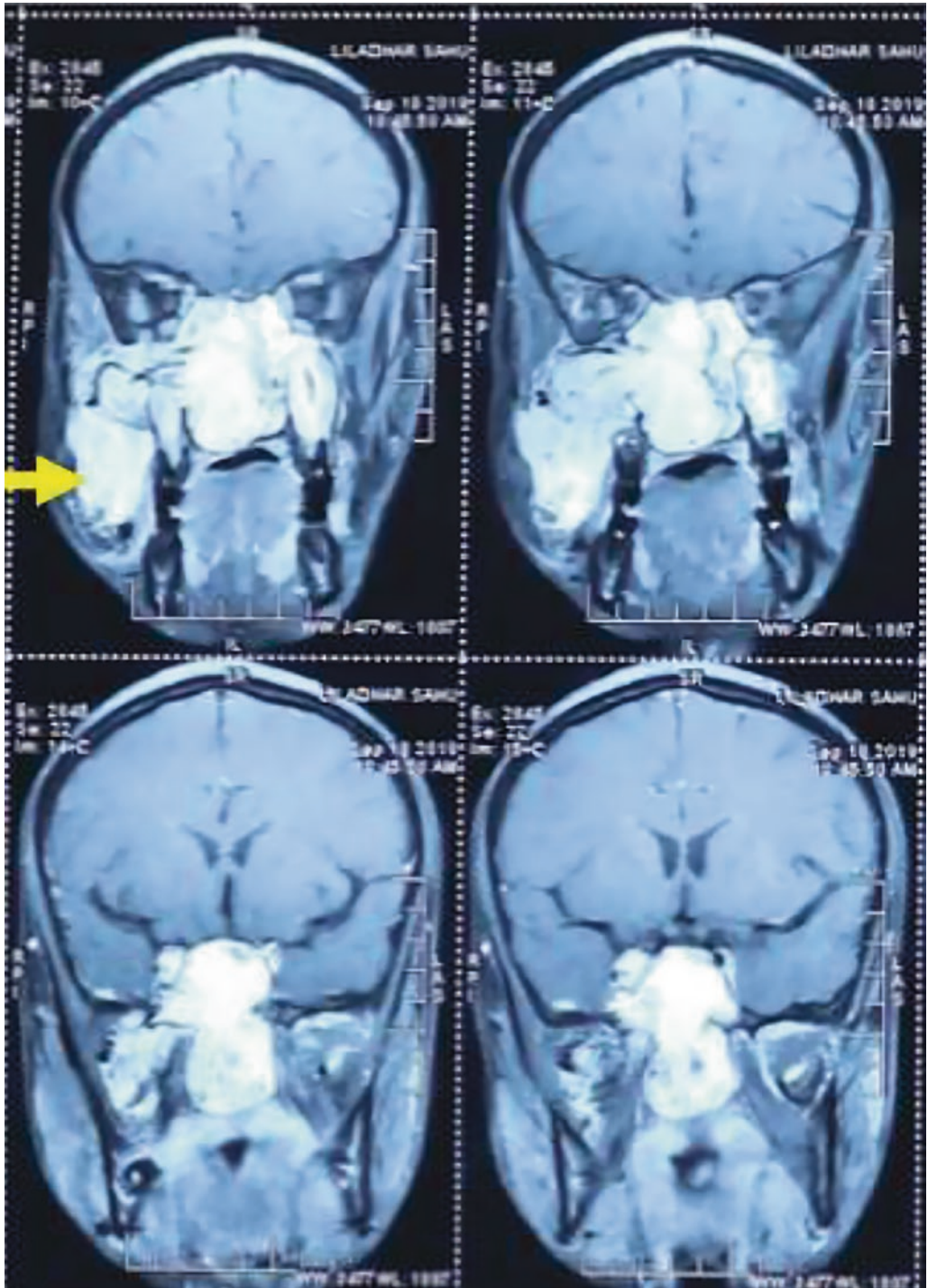


Fig. 10 Hondousa sign as seen in MRI

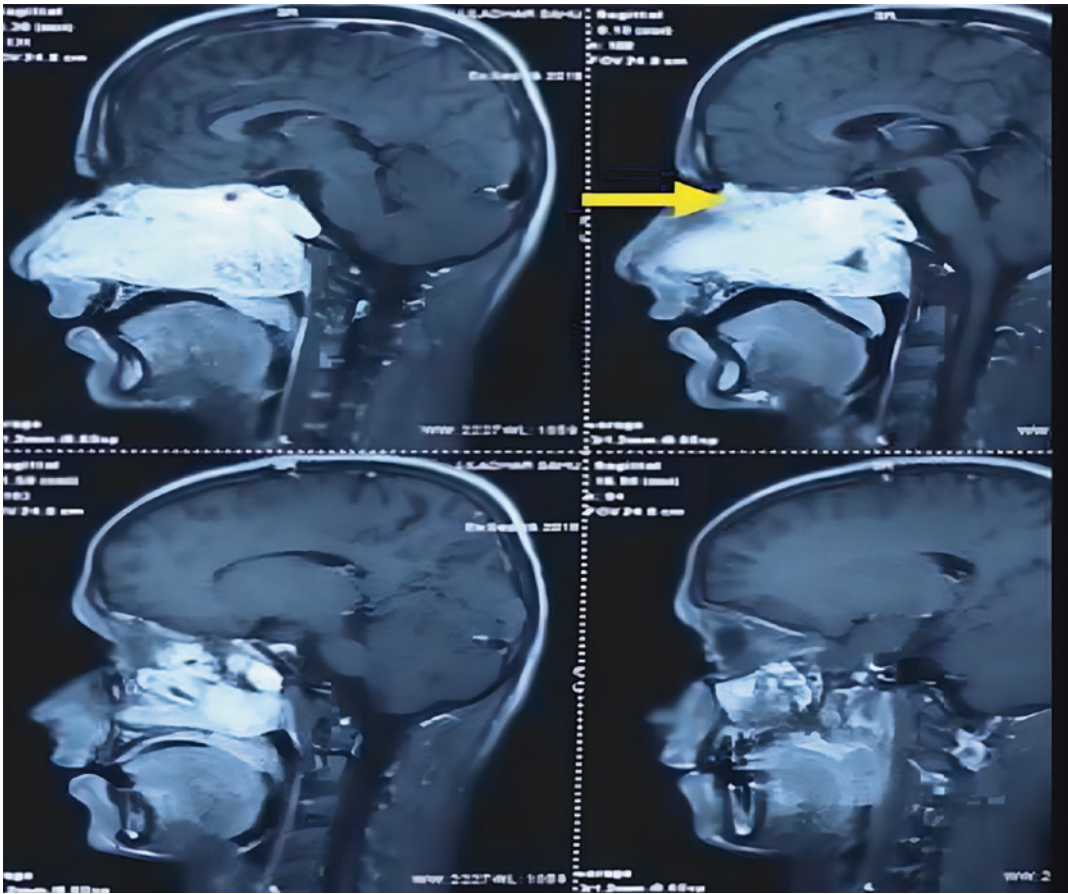


Fig. 11 Para-sagittal view in MRI showing the lesion reaching up to the cribriform plate



Fig. 12 Angiofibroma protruding through the right anterior naris: the lesion can be seen extending through the right anterior naris



Fig. 14 Exposure of the tumour after removal of the anterior and medial wall of the maxilla



Fig. 13 Elevation of the right cheek flap after the modified Weber-Fergusson incision in the subperiosteal plane and exposure of the anterior wall of the maxilla



Fig. 15 Excised angiofibroma specimen



Fig. 16 Follow-up after 6 months: an endoscopic view showing local control of the disease and well-healed scar over the right nasolabial region



Fig. 17 Follow-up after 1 year: the scar is barely visible

7 Inverted Papilloma

Inverted papillomas develop when the epithelium invades the underlying connective tissue stroma, which is more common in adults and rare in children. In the nasal vestibule, middle turbinate, septum or lateral nasal wall, they appear as unilateral red, polypoid lumps. A malignant change may occur in 2% of people [9].

The 2017 WHO classification of sinonasal papillomas are divided into:

- Inverted
- Oncocytic
- Exophytic

7.1 Case 1

A 52-year-old male smoker presented with recurrent inverted papilloma. He was operated thrice outside.

7.1.1 Operative Technique of Medial Maxillectomy: Caveats

- For well-differentiated or low-grade malignant tumours, inverted papillomas and other tumours with a small extent on the lateral wall of the nasal cavity or the medial wall of the

maxillary antrum, a medial maxillectomy is suggested.

- Depending on the size and location of the tumour, the surgical procedure involves either a modified Weber-Ferguson incision or a lateral rhinotomy.
- Technically, it is frequently challenging to remove the operative specimen from a medial maxillectomy in a Monobloc method. Mobilization of an ethmoid tumour needs to be done slowly and with great caution due to the sensitivity of ethmoid air cells.
- The anterior bony wall of the maxilla is reached by deepening the skin incision into the soft tissues and muscle of the upper lip and cheek.
- The incision is prolonged superiorly through the soft tissues all the way to the orbit's bony edge.
- The whole medial wall of the maxillary antrum, the side's inferior turbinate, ethmoid air cells and lamina papyracea are all marked for excision.
- The infraorbital nerve close to the orbital rim is carefully protected while the cheek flap is lifted.
- An antrotomy of the anterior wall allows access to the maxillary antrum.
- To provide digital access to the maxillary antrum, a sizable piece of the anterior wall of the maxillary antrum is removed.
- The inside of the maxillary antrum is carefully examined to ascertain the size of the tumour.
- If a medial maxillectomy is feasible, a periosteal elevator is used to lift the medial wall of the orbit's periosteum along the medial orbital rim and then remove it from the lamina papyracea.
- The continuity and shape of the bony rim of the orbit's inferior and inferomedial edges are preserved with extreme care.
- The medial ocular periosteum is dissected as far posteriorly as feasible. The anterior and posterior ethmoidal arteries are transected, ligated or electrocoagulated as they leave the lamina papyracea during this procedure.
- Once the orbital contents have been sufficiently mobilized in an extraperiosteal plane, a dry piece of gauze is placed for haemostasis between the orbital contents and the lamina papyracea.

- The dissection of the nasal ala and its medial retraction, which permits access to the nasal cavity, are the current areas of emphasis.
 - The medial wall of the maxillary antrum is divided in a horizontal plane at the base of the nasal cavity using a curved osteotome.
 - As observed via the anterior wall antrotomy, this technique is carried out with delicate strokes with a mallet over the osteotome until the posterior edge of the maxillary antrum is reached.
 - Similar to this, the medial wall's bony incision is continued cephalad, all the way to the medial wall of the orbit.
 - The maxillary orbital surface, the nasal bone and the orbital surface of the frontal bone can all be broken with an osteotome to release the lamina papyracea.
 - Finally, the complete surgical specimen encompassing the inferior turbinate and the middle turbinate with the lower ethmoid air cells is removed by transecting the posterior attachment of the surgical specimen close to the posterior choana using angled scissors.
 - By electrocoagulating the bleeding spots across the sliced bone surfaces, haemostasis is stabilized.
 - With a fine burr, the surgical flaps' sharp bone spicules are smoothed off.
 - The nasal cavity and the maxillary antrum are packed using a ribbon gauze roll. The anterior naris is used to remove the packing.
 - Using Prolene suture material for the skin and absorbable interrupted sutures for the soft tissues, the remaining incision is stitched close in two layers.
- Pre-operative imaging and surgical steps of the same patient are depicted in Figs. 18, 19, 20, 21, 22, 23, 24 and 25.

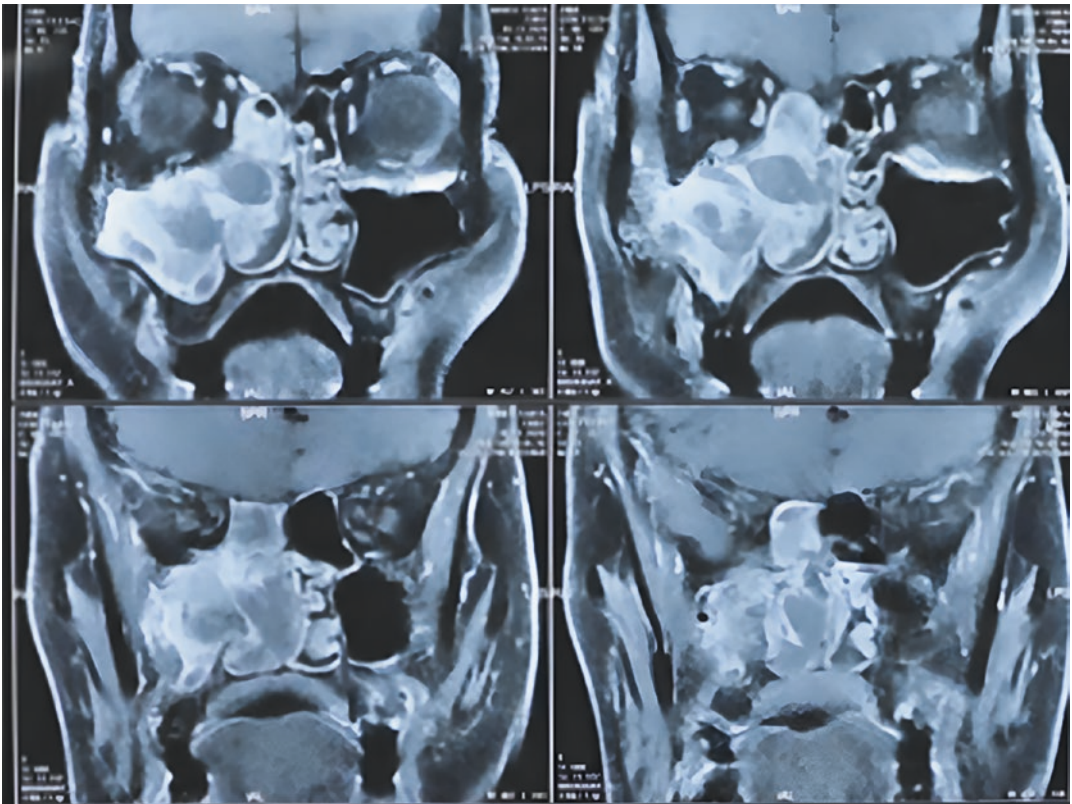


Fig. 18 MRI scan: a coronal plane showing a lesion involving the right medial wall of the maxilla and the lesion extending into the right maxillary, right anterior ethmoid and right nasal cavity

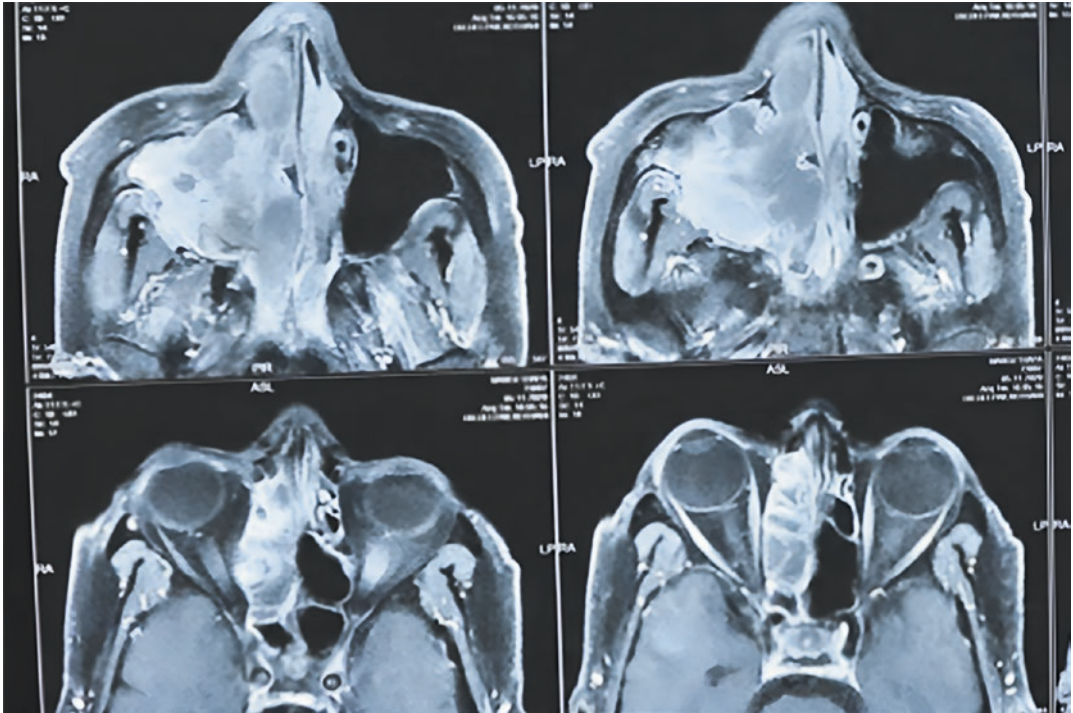


Fig. 19 MRI scan: an axial view showing an extensive lesion involving the right medial wall of the maxilla and the lesion extending into the right maxillary, right anterior ethmoid and right nasal cavity



Fig. 20 Marking of the skin incision



Fig. 21 Elevation of the cheek flap and exposure of the anterior wall of the maxilla

7.1.2 Postoperative Care

- The nasal packing was removed after 72 h.
- However, until the gap has been completely epithelialized, aggressive nasal irrigation and sufficient humidity are essential to eliminate clots and crusts.

- The patient was instructed on nasal irrigation with a catheter, since the anterior nares are the sole route to the maxillary antrum.
- Practically little cosmetic distortion and little functional impairment are the outcomes of the surgery.

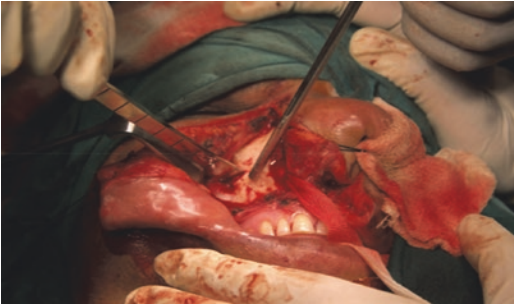


Fig. 22 Opening the anterior wall of the maxilla



Fig. 23 Maxillary cavity after opening the anterior wall of the maxilla and excision of the tumour from the maxillary sinus and the medial wall of the maxilla



Fig. 24 Specimen of inverted papilloma following medial maxillectomy



Fig. 25 Closure of the skin incision

- The patient binocular vision is normal, and the globe is properly aligned with the opposing side.

8 Giant Cell Tumour

Deep and superficial soft tissue can include soft tissue giant cell tumours, which are uncommon tumours with little malignant potential. These tumours have mononuclear components made up of round, oval or spindle cells as well as equally distributed multinucleated large cells. There are no atypia or obvious signs of mitotic activity. Desmin, cytokeratins, smooth muscle actin and TRAP immunohistochemical stains are quite useful for diagnosis. Since this type of tumour is extremely uncommon to occur in the nasal cavity, soft tissue tumours must be considered in the differential diagnosis of nasal obstruction.

8.1 Case 1

8.1.1 Operative Technique of Infrastructure Maxillectomy: Caveats

- The radiological imaging of giant cell tumour of the right maxilla is seen in Figs. 26 and 27, which includes the upper alveolar ridge and spreads from the area of the second premolar tooth socket to the area of the second molar socket.
- Before beginning an infrastructure maxillectomy, a proper characterization of the extent of the tumour requires appropriate radiographic examination with CT scans in the axial and coronal planes.
- Under general anaesthesia, nasotracheal intubation into the opposing nasal cavity is used to accomplish the surgery.
- Around the visible and palpable tumour, an incision is made in the mucosa of the gingivo-buccal sulcus and that of the hard palate, with appropriate mucosal margins.
- If teeth are available, a suitable tooth is pulled far from the tumour's edge through the socket of the bone being removed, protecting the integrity of the neighbouring remaining tooth.

- The mucosal incision is prolonged intraorally into the soft tissues and up to the bone anterior wall of the maxilla.
- Before dividing the bones, this incision is expanded circumferentially through the soft tissue to separate all the soft tissue attachments of the hard palate and the lower part of the maxilla.
- An osteotome is utilized to split the remaining bone attachments and remove the specimen in a Monobloc using bone cuts through the previously described mucosal incision.
- The bottom part of the maxillary antrum, which was taken in one piece together with the surrounding hard palate and the maxilla's alveolar process, is seen in the anterosuperior view of the surgical specimen (Fig. 31).
- It is not necessary to curette out the remaining antrum if the mucosa does not exhibit any chronic inflammatory alterations.
- To keep the packing in place, a dental obturator that had previously been made is now linked to the remaining teeth.
- Nasal packing has to be done.
- The packing may be taken out in 2–3 days, and a temporary dental obturator is made while waiting for the surgical defect to com-

Fig. 26 An expansile radiolucent lesion involving the anterolateral wall of the right maxilla

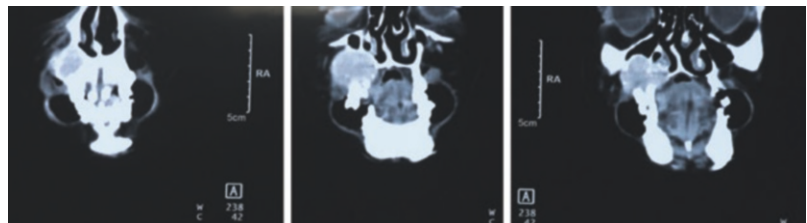
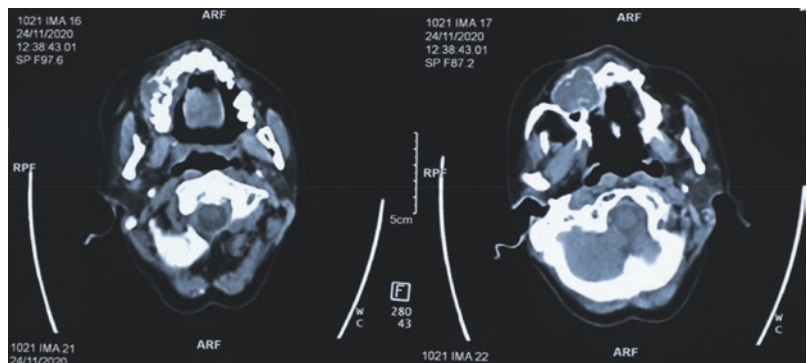


Fig. 27 CT scan: an axial view showing an expansile radiolucent lesion present between the right upper second premolar and first molar tooth



pletely epithelialize. At that moment, maxillofacial prosthodontists create a permanent removable dental prosthesis.

Pre-operative imaging and surgical steps of the same patient are depicted in Figs. 28, 29, 30, 31 and 32.



Fig. 28 Marking of the skin incision



Fig. 29 Elevation of cheek flap exposing the anterior wall of the right maxilla



Fig. 30 Surgical site after infrastructure maxillectomy, maxillary antrum and the cut end of the upper alveolus visualized

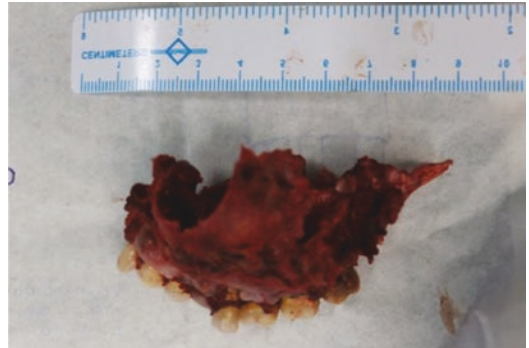


Fig. 31 Specimen showing en-bloc resection



Fig. 32 Final closure

9 Squamous Cell Carcinoma of the Maxilla

The most frequent sinonasal cancer is still squamous cell carcinoma. However, it can be challenging to pinpoint the specific location of the tumour because these areas are frequently afflicted, as well as the nasal cavity and anterior ethmoid. Most often, it starts in the maxillary sinus. Differentiation levels vary and may worsen with time [10], while often, a combination of surgical and chemoradiation therapy is performed. However, chemoradiation alone may be curative in sinonasal carcinomas with poor or undifferentiated carcinoma. Rarely is the columella or nasal septum the main location. Due in part to the potential of bilateral cervical node metastasis, these tumours have a dismal prognosis.

9.1 Case 1

A 51-year-old man presented with a 3-month history of growth on the left side of the maxillary sinus involving the hard palate. A biopsy was suggestive of moderately differentiated squamous cell carcinoma.

9.1.1 Operative Technique: Caveats

- A subtotal maxillectomy effectively eliminates the infrastructure and superstructure of the maxilla together with the whole maxilla, with the exception of the orbital floor.
- It is referred to as a “complete maxillectomy” if the orbital floor is also removed during the procedure. To avoid ptosis of the globe in the situation, the orbital floor rebuilding should be taken into account.
- An orotracheal tube is used to maintain the patient’s general anaesthesia when they are put on the surgical table.
- Using the Weber-Ferguson incision, the maxilla was exposed.
- For the elevation of the cheek flap, a skin incision is created with a scalpel, and electrocautery is then performed, which offers good haemostasis.
- The gingival sulcus and the whole thickness of the upper lip are split.
- The superior labial artery has to be tied up if it is bleeding.
- An incision is made in the mucosa of the upper gingivobuccal sulcus, staying close to the gingiva, to raise the upper cheek flap. It is elevated full thickness, staying directly above the maxillary periosteum until its posterolateral side is revealed.
- The infraorbital nerve and its entrance into the soft tissues of the face are exposed by continuing to elevate the cheek flap in this area.
- The infraorbital nerve should be carefully maintained if only the lower part of the maxilla is to be removed in order to maintain the cheek’s cutaneous sensations.
- In order to reach the pterygomaxillary fissure, it is crucial to raise the cheek flap all the way back to the posterolateral surface of the maxilla. This will expose the zygoma’s underside.
- By slicing the soft tissues along the ala of the nose and through the mucosa of the lateral wall of the nasal cavity, access is gained to the nasal cavity.
- On the other side, a mouth gag is now used, and the oral cavity is opened as widely as possible to reveal the hard palate and alveolar process.
- Just below the infraorbital foramen, the anterior wall of the maxilla is divided using a high-speed power saw with a very tiny blade.
- To produce the desired line of resection through the anterior wall of the maxillary antrum and to enable the excision of the lower half of the maxilla as the surgical specimen, the bone cut is prolonged anteriorly and posteriorly.
- The infraorbital nerve and its entrance into the soft tissues of the face are exposed by continuing to elevate the cheek flap in this area.
- To expose the alveolar process, the hard palate, the zygoma and the area around the posterolateral aspect of the maxilla, the oral cavity is opened as far as feasible.
- The line of fracture is carried between two teeth if there is space between them. It is advised to extract one tooth along the antici-

- pated line of transection of the alveolar process since, if the teeth are intact, it is extremely possible that the final tooth on the remaining alveolar process will become loose and maybe fall out.
- The bone that was previously cut between the previously formed transverse line of transection and the alveolar process is connected using the power saw once again.
 - The hard palate mucosa is now the centre of attention. A tongue depressor is used to ensure sufficient exposure with the mouth open wide.
 - In order to maintain good mucosal margins in all directions, an incision is created in the hard palate mucosa surrounding the primary tumour using a needle-tip electrocautery.
 - The tumour is circumvented by anteriorly starting an incision at the maxillary tubercle and curls anteriorly.
 - Prior to the alveolar process holding the left-side incisors and canines, the incision is extended behind it.
 - To finish the specimen's circumferential mobilization, the mucosal incision comes to a point where it meets the removed first molar tooth's socket.
 - The length of the hard palate's mucosal incision is deepened into the mucoperiosteum and up to the bone.
 - The incision in the hard palate mucosa and a part of the soft palate is visible in a close-up image of the surgical field.
 - Along the line of the mucosal incision, the hard palate is divided using a power saw.
 - Due to bleeding from the palatine arteries, internal maxillary artery branches and the posterior wall of the maxilla as well as the pterygoid fossa, there is significant bleeding at this stage of the surgery.
 - It is imperative to speed up this procedure, since attempts to stop the bleeding are fruitless until the surgical specimen is removed.
 - An osteotome is used to join the fracture lines after all the bone incisions have been performed with the power saw, allowing the specimen to be rocked over the soft tissue attachments.
 - The posterior soft tissue attachments of the specimen (the pterygoid muscles) are split with electrocautery or Mayo scissors, and the surgical specimen of the bottom half of the maxilla is removed.
 - The internal maxillary artery, sphenopalatine artery and smaller blood vessels of the soft palate are often the sources of bleeding at this stage.
 - The internal maxillary artery can be ligated to stop bleeding, or a chromic catgut suture ligature can be inserted through the pterygoid muscle stumps.
 - However, bleeding from the sphenopalatine artery is seldom controlled by ligation; as the vessel's stump is typically in a bony cleft, electrocoagulation is the best method for achieving haemostasis.
 - Now a burr has been used to smooth out all the jagged spicules on the bony margins.
 - Interrupted absorbable sutures are used to approximate the cut margins of the mucosa on the soft palate's anterior and posterior borders.
 - Bacitracin solution is used to irrigate the wound, and blood clots are removed.
 - Starting at the roof of the antrum, ribbon gauze is applied digitally to the defect and held in place by light digital pressure so that it may fit into the antrum's nooks and corners.
 - The dental obturator that was previously made is now applied and fastened with wires. To restore the hard palate section that was removed, the obturator is connected to the remaining teeth. If the patient is missing teeth, drill holes are used to connect the obturator to the remaining alveolus.
 - The dental obturator's placement restores the missing hard palate tissue, enabling the patient to swallow liquids and soft meals without any trouble right away after surgery.
 - Utilizing non-absorbable sutures for the skin and absorbable interrupted sutures for the subcutaneous tissues, the skin incision is stitched shut in two layers.
 - To produce a higher aesthetic effect, careful consideration is given to an accurate approximation of the skin margins.

- In that sense, precise realignment of the skin borders along the ala and the nasal cavity's floor is crucial.
- On the first day following surgery, there is a mild amount of oedema of the lower eyelid and cheek, but this swelling is temporary and often goes down on its own in 2 to 3 days.
- To lessen postoperative oedema, place ice packs over the cheek.
- The majority of patients can handle a soft diet a day following surgery.
- Pre-operative imaging, surgical steps and follow-up of the same patient are depicted in Figs. 33, 34, 35, 36, 37, 38 and 39.

9.1.2 Postoperative Care

- Following a partial maxillectomy, the postoperative care of the patient focuses on maintaining excellent oral hygiene and caring for the face incision until sutures are removed.
- Because they act as a nidus for infection and can occasionally induce wound separation,

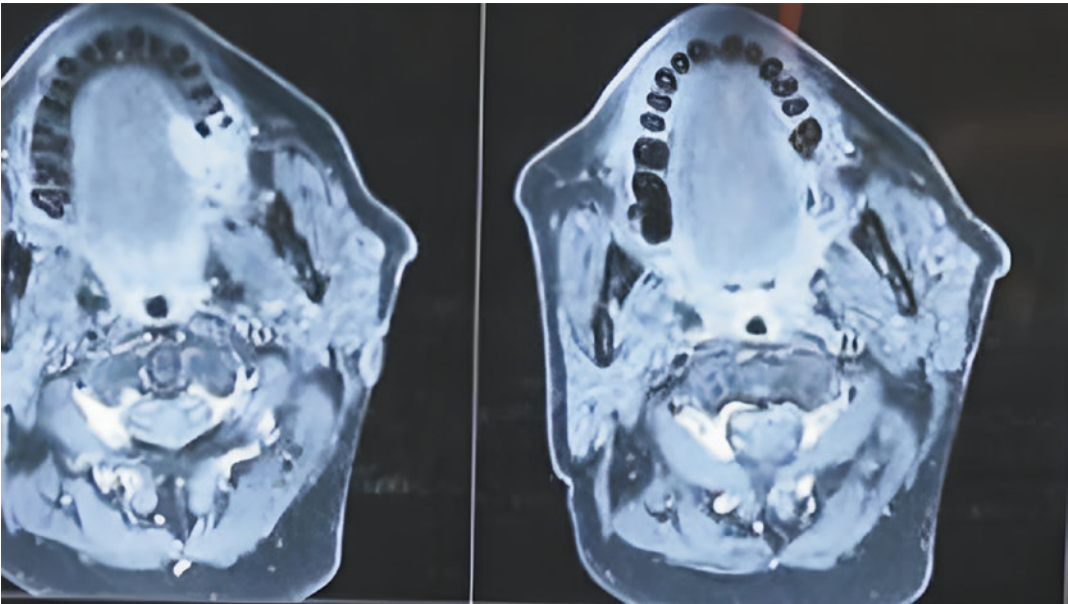


Fig. 33 MRI scan showing lesion



Fig. 34 Marking of the skin incision



Fig. 35 Elevation of the skin flap and exposure of the anterior wall of the left maxilla



Fig. 36 Surgical site after orbital plate preserving subtotal maxillectomy



Fig. 37 Surgical specimen showing the ulceroproliferative growth with adequate margin



Fig. 38 Closure of skin incision with obturator in situ

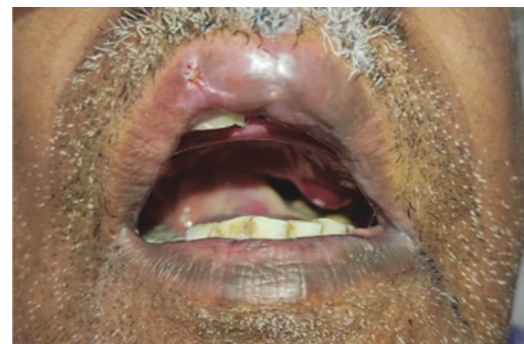


Fig. 39 Two weeks post-surgery with obturator in situ

suture line clots and crusts are carefully removed from above the suture line.

- Sometimes it's important to apply ice pack compresses to the cheek if there is chronic swelling or an inflammatory reaction.
- On the second postoperative day, the patient is instructed to regularly rinse and irrigate their mouths with a warm water and baking soda solution to maintain their mouths free of debris and secretions.
- On the third postoperative day, the packing is delicately removed.
- The dental obturator is removed by cutting the wires 1 week following surgery.
- The prosthodontist now creates an interim obturator, which is secured to the remaining teeth using clasps.
- In edentulous individuals, early retention of a prosthesis can be challenging and frequently disappointing.
- Up until the surgical defect's skin graft has completely healed, oral and nasal irrigations are maintained.

- Approximately 6–8 weeks later, a permanent dental obturator is made.
- The patient's ability to talk normally and consume all sorts of food is restored thanks to this permanent obturator.

10 Odontogenic Tumours

According to the WHO, ameloblastic fibroma (AF) and associated diseases are neoplasms made up of proliferating odontogenic epithelium embedded in cellular ectomesenchymal tissue that resembles dental papilla and with variable degrees of inductive transformation and creation of dental hard tissue [11].

Ameloblastic fibroma is an uncommon mixed odontogenic tumour that makes up about 2.5% of all odontogenic tumours [12, 13]. It typically affects those under 30 (70% of patients are younger than 20 years at presentation). There are no racial or sexual preferences [14–16]. Most often (approximately 80% of the time), AF devel-

ops in the mandible, with the remaining occurrences developing in the maxilla [14, 15, 17, 18]. AF patients typically experience discomfort, tooth eruption failure and/or jaw oedema [12, 14]. It is difficult to identify AF from a straight-forward ameloblastoma radiographically because it resembles a radiolucent multilocular or unicellular cyst [12, 14, 17].

10.1 Case 1

A 4-year-old female child presented with a growth in the right upper alveolus for 4 months, with a biopsy suggestive of ameloblastic fibroma.

Pre-operative imaging, surgical steps and follow-up of the same patient are depicted in Figs. 40, 41, 42, 43, 44, 45, 46, 47 and 48.

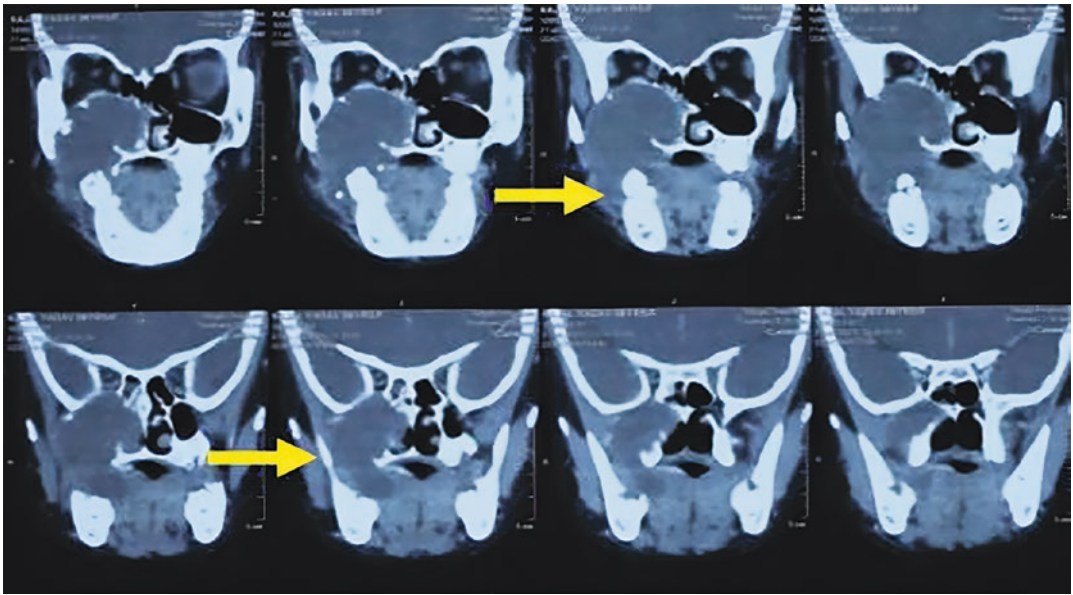


Fig. 40 CT scan: coronal cuts showing the lesion in the right maxilla with the erosion of the anterolateral wall of the maxilla



Fig. 41 Skin incision and elevation of cheek flap



Fig. 42 Elevation of the cheek flap, which shows the lesion eroding the anterolateral wall of the maxilla and extending to the oral cavity at the level of the upper gingivobuccal sulcus, smooth surface of the lesion in the oral cavity can be appreciated

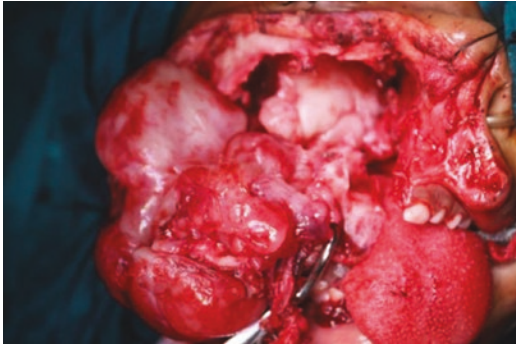


Fig. 43 Tumour exposed in the right maxillary sinus after removal of the anterior wall of the maxilla



Fig. 46 Surgical defect after orbital plate-preserving subtotal maxillectomy with an obturator in situ

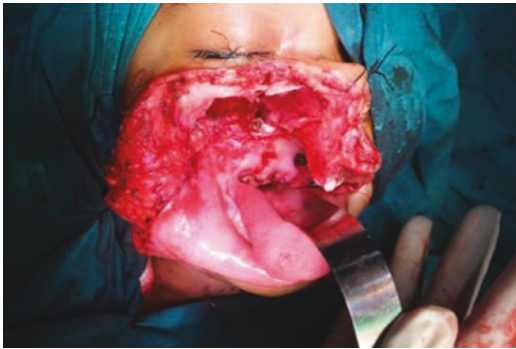


Fig. 44 Surgical defect after orbital plate-preserving subtotal maxillectomy



Fig. 45 Excised en-bloc specimen



Fig. 47 Postoperative follow-up after 6 months



Fig. 48 Follow-up after 1-year, locoregionally controlled disease

11 Osteosarcoma

The majority of instances of osteosarcoma (OS), a malignant mesenchymal tumour that seldom affects the maxilla, show as painful swelling of the area around the maxilla as their initial clinical manifestation. The most significant determining variables of prognosis are early diagnosis and extensive surgical excision of the tumour. To hasten the diagnosing process, careful consideration should be given to OS's atypical clinical manifestations.

11.1 Case 1

A 25-year-old man presented with a recurring tumour in the right nasal cavity had right partial maxillectomy with complete palatotomy and temporalis muscle rotation flap restoration after the first undergoing surgery for a giant cell lesion.

Pre-operative imaging, surgical steps and follow-up of the same patient are depicted in Figs. 49, 50, 51, 52, 53, 54, 55, 56, 57, 58, 59 and 60.

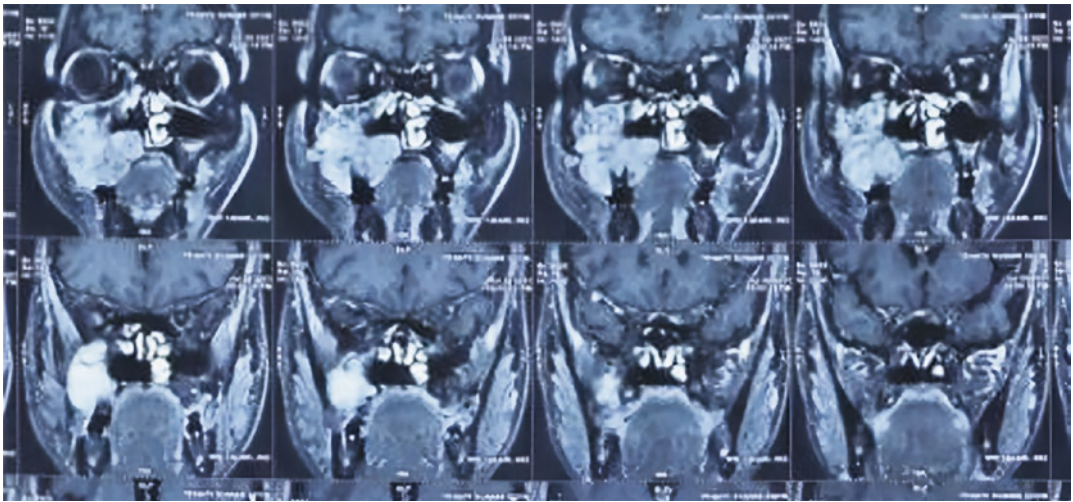


Fig. 49 MRI scan showing the lesion in the right maxillary sinus and involving the palate: lesion over the palate is seen crossing the midline anteriorly



Fig. 50 Marking the skin incision



Fig. 51 Elevation of the cheek flap over the anterior wall of the right maxilla in the submuscular plane

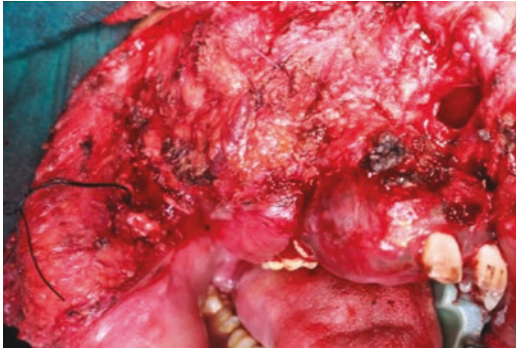


Fig. 52 Complete exposure of the tumour



Fig. 55 Skin marking for temporalis muscle rotation flap



Fig. 53 Surgical defect following extirpation of the tumour in the right maxilla by subtotal maxillectomy with total palatotomy



Fig. 56 Elevation of the scalp skin to expose the temporo-parietal fascia



Fig. 54 Surgical specimen



Fig. 57 Rotation of the temporalis muscle onto the right maxillary defect



Fig. 58 In setting the temporalis muscle to completely cover the maxillary defect

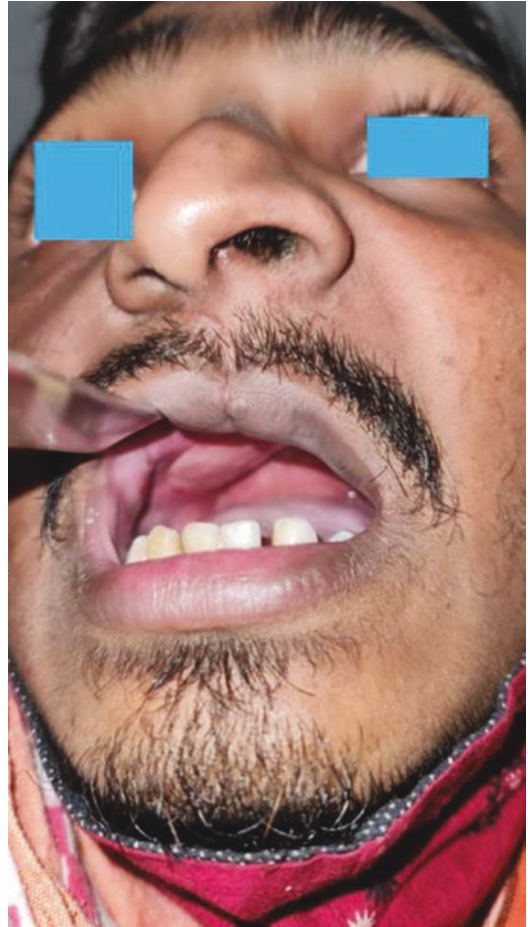


Fig. 60 Follow-up after 1 year showing locally well-controlled lesion



Fig. 59 Follow-up after 1 year after the completion of adjuvant radiotherapy with well-healed surgical scar

12 Rhabdomyosarcoma

Rhabdomyosarcomas are the most prevalent paranasal sinus malignancy in paediatric patients, with the orbit being the most frequent subsite overall. Rhabdomyosarcomas are among the tumours with tiny, rounded blue cells that may be seen on histology and are generated from the primitive mesenchymal tissue with myogenic differentiation. There are four categories in the current histologic categorization of

rhabdomyosarcoma [19]. The most prevalent kind, known as the embryonal type, often affects newborns and younger children (incidence of 55–65%). Botryoid and spindle cell subtypes are included in the embryonal categorization and are typically thought to have the best prognosis. The alveolar form (20–30% incidence) more frequently affects teenagers and has a worse prognosis potential, necessitating typically more intense multimodality therapy. The undifferentiated type is poorly characterized with no clear histologic myogenesis or differentiation, while the anaplastic type—previously known as the pleomorphic type—primarily affects adults. Due to their quick development and high rate of distant dissemination, both of these varieties have dismal prognosis. Given the proximity of important tissues to the head and neck, only a biopsy is frequently done there, since recurrence might be significant despite rigorous resection. Neoadjuvant chemoradiation

is encouraged by certain regimens to make tumours more amenable to excision. Overall, 5-year overall survival is good, notably for orbital rhabdomyosarcoma, which has a 95% survival rate, compared to a 74% survival rate for parameningeal locations [20].

12.1 Case 1

A left nasal mass has recurred in a 3-year-old kid. He underwent the first surgery in April 2021 using an endoscopic technique since his HPR suggested an AC polyp. A second biopsy of the recurring tumour again revealed evidence of an AC polyp, and the final HPE was reported as rhabdomyosarcoma.

Pre-operative imaging, surgical steps and follow-up of the same patient are depicted in Figs. 61, 62 and 63.

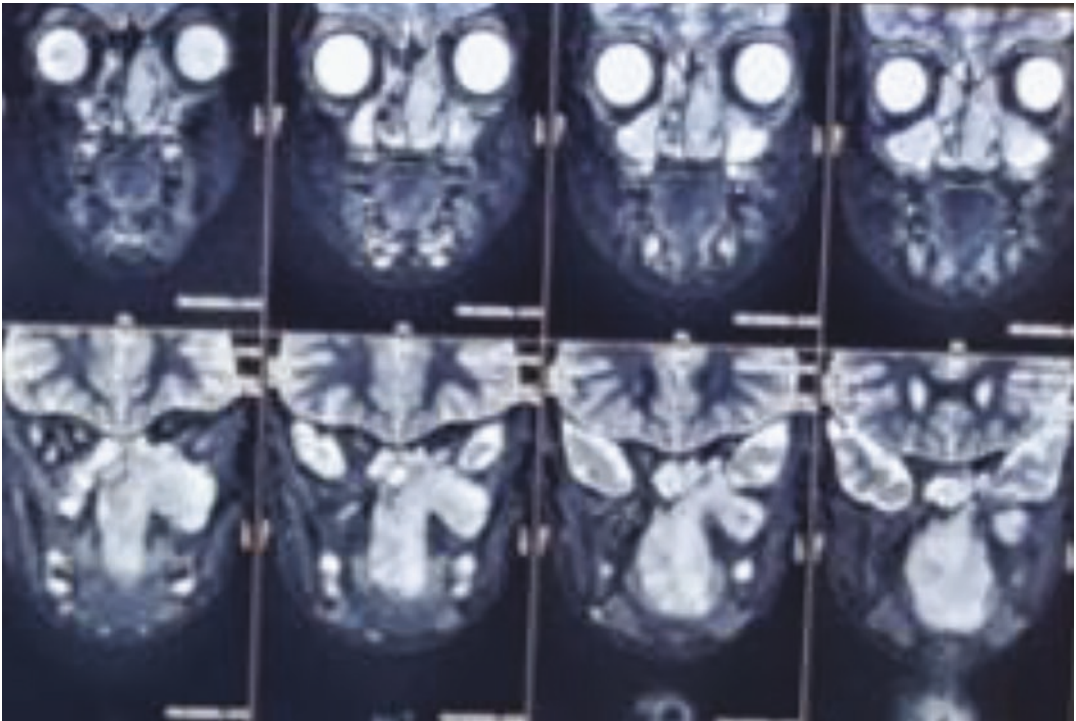


Fig. 61 MRI scan shows a hyperintense lesion present in the left maxillary sinus



Fig. 62 Closure of the skin incision



Fig. 63 Follow-up after 6 months showing a well-healed surgical scar

13 Olfactory Neuroblastoma (OAN)

The basal cells that make up the olfactory neuroepithelium are where OAN comes from. OAN makes up less than 5% of sinonasal cancers [21]. The incidence of this tumour has a bimodal distribution, with peaks at 20 and 50 years of age. Women are more likely to have it. A neuroendocrine tumour called OAN has the ability to create peptides that can lead to paraneoplastic illnesses. In the literature, cases of patients with Cushing's syndrome, OAN-producing vasoactive peptide-induced hypertension or improper antidiuretic hormone production have been reported.

OAN must be distinguished from small cell carcinoma, rhabdomyosarcoma, lymphoma, neuroendocrine tumour and sinonasal undifferentiated carcinoma (SNUC), all members of the category known as "small round blue cell tumours." Therefore, a histopathological assessment by an experienced pathologist is advised. OAN is normally devoid of keratins and exhibits the neuroendocrine markers neurone-specific enolase, synaptophysin and chromogranin. S-100 may only be positive at the tumour's perimeter, which can help distinguish OAN from sinonasal melanoma. Vimentin, actin and desmin negativity rule out rhabdomyosarcoma [22].

Hyams et al. [23], Showed that the degree of differentiation, the tumour architecture, the mitotic index, the nuclear polymorphism, the fibrillary nature of the matrix and tumour necrosis were used to construct a four-point histological grading system for OAN. This tumour may either be indolent (grade 2) or exceedingly aggressive (grade 4). On the use of cytogenetics in the OAN diagnosis, there is minimal information. While some people are lucky enough to live for more than 20 years, others are less fortunate and pass away within a few months from extremely aggressive illnesses with extensive metastases [24]. OAN can penetrate the dura and

anterior cerebral fossa up to 25% of the time. At the time of presentation, cervical node metastases are visible in 5% of patients, and distant metastases are found in around 7% of patients [24]. For OAN, disease-specific staging methods have been developed. The most useful and commonly used systems are those attributed to Kadish et al. [24], Morita et al. [25] and Dulguerov and Calcaterra [26].

13.1 Case 1

A 32-year-old lady presented with a mass in the nasal cavity involving the skin, pre-operative biopsy from the mass showed Esthesioneuroblastoma.

Pre-operative imaging, surgical steps and follow-up of the same patient are depicted in Figs. 64, 65, 66 and 67.

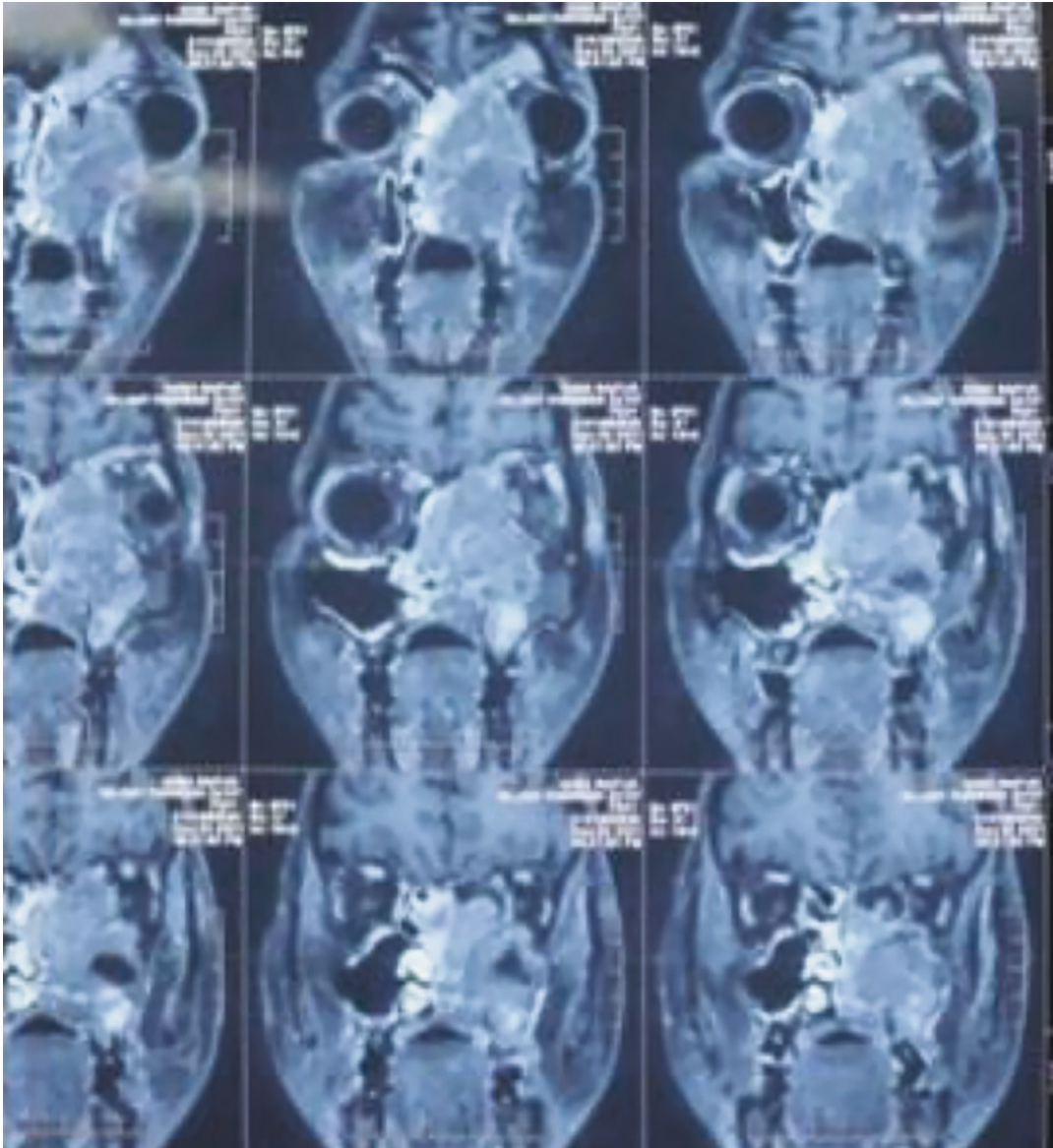


Fig. 64 MRI scan coronal section showing lesion involving the left nasal cavity, lesion seen involving the cribriform plate



Fig. 65 Marking of skin incision



Fig. 66 Elevation of the cheek flap



Fig. 67 Follow-up after 3 months with a well-healed scar over the left nasolabial fold region

crine, intestinal and undifferentiated. The least aggressive variety is papillary adenocarcinomas. The kind most frequently linked to tumours brought on by wood workers is intestinal. The prognosis for sessile and mucoid adenocarcinomas is the poorest [27].

14 Adenoid Cystic Carcinoma

Nine percent of sinonasal cancers are adenocarcinomas. Similar to SCC, it favours men and typically affects people in their sixth and seventh decades of life. The upper nasal cavity and ethmoid sinuses are the typical locations for adenocarcinomas. They seldom develop metastases and grow slowly. Sinonasal adenocarcinoma has a number of known histological subgroups, including papillary, sessile, mucoid, neuroendo-

14.1 Case 1

A 43-year-old lady presented with intermittent nasal bleed for 3 months and a mass in the left nasal cavity for 2 months; biopsy was suggestive of adenoid cystic carcinoma.

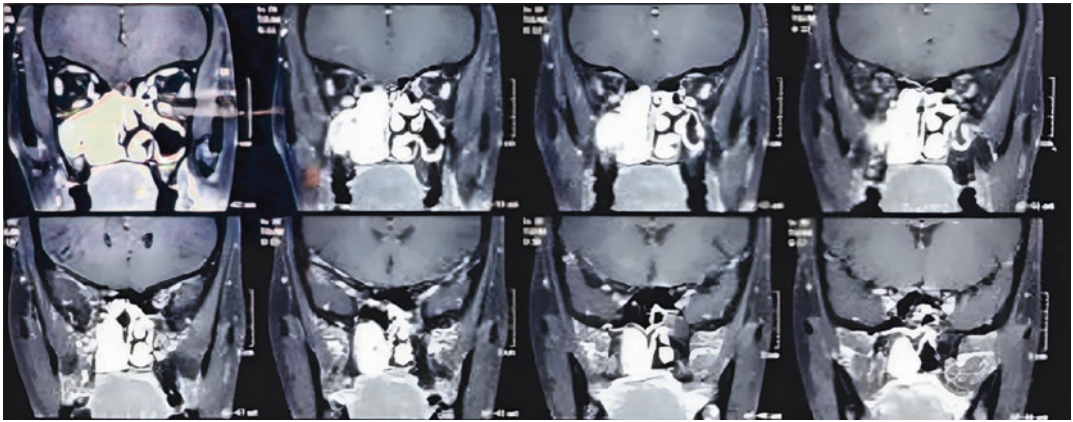


Fig. 68 MRI scan shows a lesion involving the right maxilla and right-sided nasal cavity



Fig. 69 Palate and orbital plate-preserving maxillectomy defect following surgical extirpation of the tumour



Fig. 70 Follow-up after 6 months

Pre-operative imaging, surgical steps and follow-up of the same patient are depicted in Figs. 68, 69 and 70.

15 Postoperative Management

- The conventional/Merocel pack is to be removed after 48 h, preferably in OT, to look for any bleeding at the surgical site.
- Saline irrigation is to be started after pack removal.
- Endoscopic suctioning and cleaning of the nose every week until crusting subsides.
- Regular follow-up.

16 Adjuvant Treatment

- Generally reserved for locally advanced primary tumours of the nose and paranasal sinuses.
- *Radiotherapy*:
 - Positive or close margins
 - Nodal metastasis
 - Unresectable tumours
 - Recurrence (in specific settings)
- *Chemotherapy* is used when previous surgery and radiation have failed.
- *Hormone therapy* has been proposed due to the androgen receptors associated with JNAs in an attempt to decrease tumour size and vascularity.
- *Oestrogen* has been shown to decrease the size and vascularity of the tumour but has feminizing side effects.

17 Mucormycosis

The coronavirus disease 2019 (COVID-19) pandemic has been one of the most significant global health crises in recent history. The World Health Organization (WHO) classified COVID-19, caused by severe acute respiratory syndrome coronavirus 2 (SARS-CoV2), a global pandemic in March 2020. As the world grapples with the devastating impact of this virus, several countries have witnessed a sharp rise in cases of a rare fun-

gal infection known as mucormycosis. In India, this epidemic of mucormycosis has emerged as a significant concern for healthcare providers. The sudden surge of this deadly fungal infection has put an additional burden on an already overwhelmed healthcare system in India. The fungal invasion of blood vessels results in mycotic thrombosis, ischemic infarction and necrosis of affected host tissues, making mucormycosis a potentially lethal infection.



Fig. 71 Mucormycosis of hard palate causing oro-nasal fistula



Fig. 72 Mucormycosis of the alveolar process of the hard palate



Fig. 73 Orbital extension of mucormycosis causing the frozen globe



Fig. 75 Gadolinium-enhanced T1-weighted coronal MRI showing heterogeneous lesion involving bilateral maxillary sinus right more than left with right orbital involvement

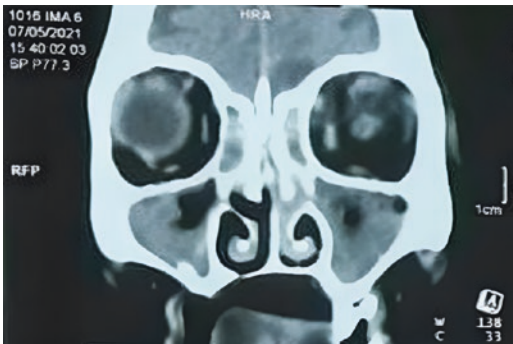


Fig. 74 Contrast-enhanced CT images—coronal section—heterogeneous lesion involving bilateral maxillary sinus, bilateral ethmoidal sinus and extension into the nasal cavity



Fig. 76 Excision specimen showing the infrastructure maxilla, black necrotic inferior turbinate and middle turbinate

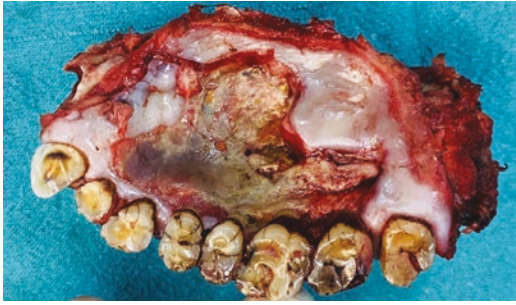


Fig. 77 En-bloc resection of the infrastructure of the maxilla, showing disease involving the hard palate



Fig. 78 Surgical site following left medial maxillectomy and left orbital exenteration

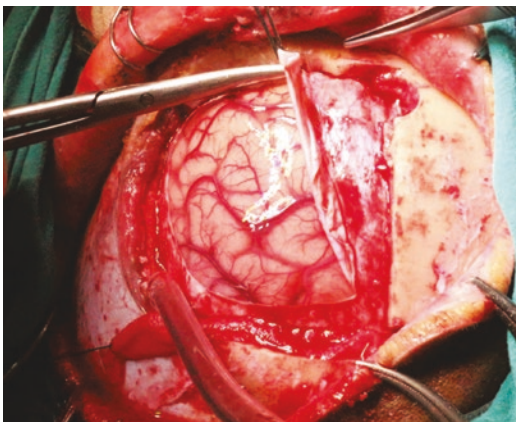


Fig. 79 Intracranial extension of mucormycosis for which craniotomy was performed



Fig. 80 Third month of follow-up of the patient, well-healed orbital cavity following exenteration with a well-healed scar which is barely visible

Various presentations of mucormycosis, intra-operative findings and the rehabilitation prosthesis are depicted in Figs. [71](#), [72](#), [73](#), [74](#), [75](#), [76](#), [77](#), [78](#), [79](#), [80](#), [81](#), [82](#), [83](#), [84](#), [85](#) and [86](#).



Fig. 81 Dental obturator lateral view



Fig. 82 View of dental obturator from below

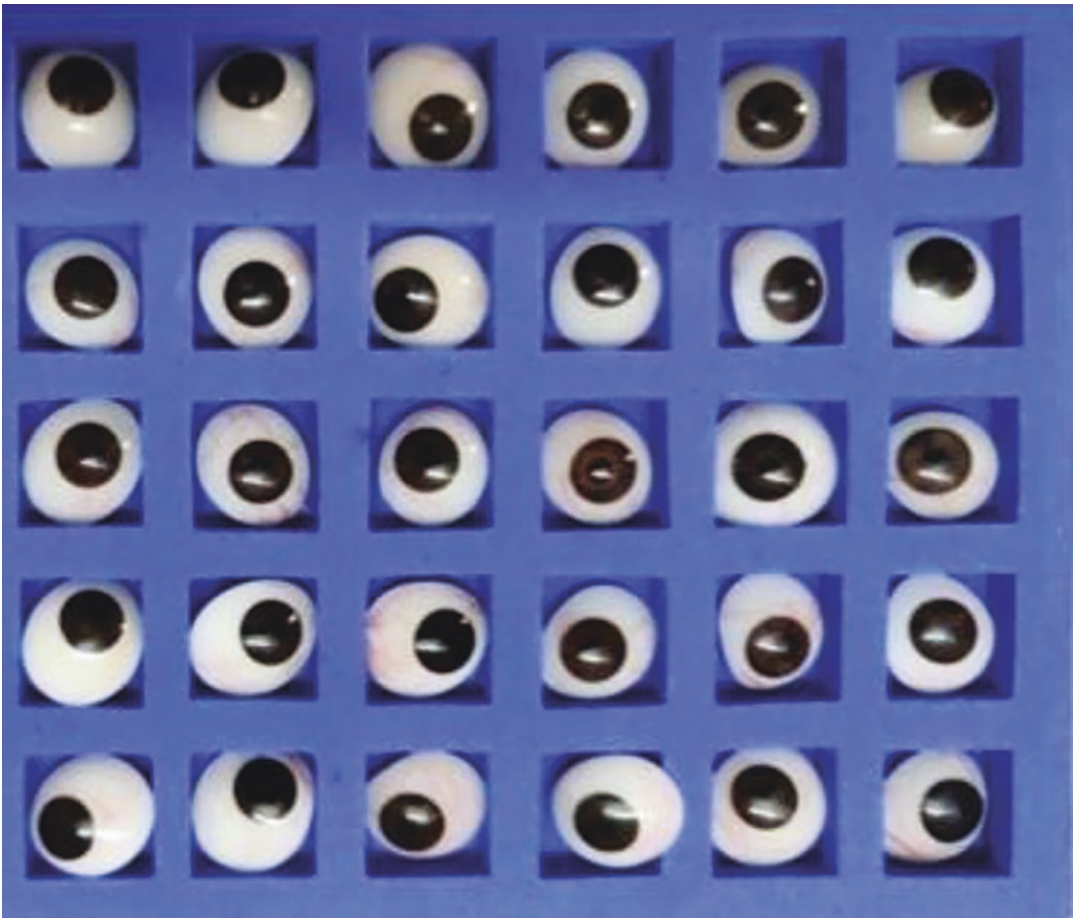


Fig. 83 Ocular prosthesis



Fig. 84 Follow-up case of right infrastructure maxillectomy before placement of the obturator



Fig. 85 Follow-up case of right infrastructure maxillectomy after placement of the obturator

Fig. 86 Before and after use of ocular prosthesis



18 Rehabilitation

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