The Upper Jaw ("Midface") and Sinuses: Part II

19

John Hanratty and Michael Perry

19.1 Swellings and Lumps Around the Midface

The most common causes of swelling in this area are i) infections, ii) dental cysts and iii) tumours. Iatrogenic conditions that affect the bone, such as ORN and MRONJ may also present with swelling, although this is perhaps less obvious to the patient, who will also have significant discomfort and other symptoms. Disorders arising primarily within the bone, such as fibrous dysplasia and Paget's disease are uncommon and tend to present slowly and with few other symptoms. Of all these possible conditions, infections are by far the most common and are usually dental or skin in origin, although they can be related to the maxillary sinus. These are usually mixed infections, containing multiple species of aerobic/anaerobic bacteria. Infections arising in the skin are another common group and are discussed in the chapter on the skin. All these infections are very important anatomically. Spread of infection can quickly involve the orbit (orbital cellulitis) and (rarely) the cavernous sinus. Dacrocystitis may also occur here. This is an infection of the nasolacrimal duct. It usually presents as swelling higher up on the midface, along the side of the nose (Fig. 19.1).

19.1.1 Dental Infections

Odontogenic infections in the upper jaw arise either usually from dental caries or periodontal infections that have extended beyond the alveolar bone to involve the fascial spaces around the face and oral cavity. These infections tend to spread along planes of least resistance from the supporting structures of the affected tooth. In the

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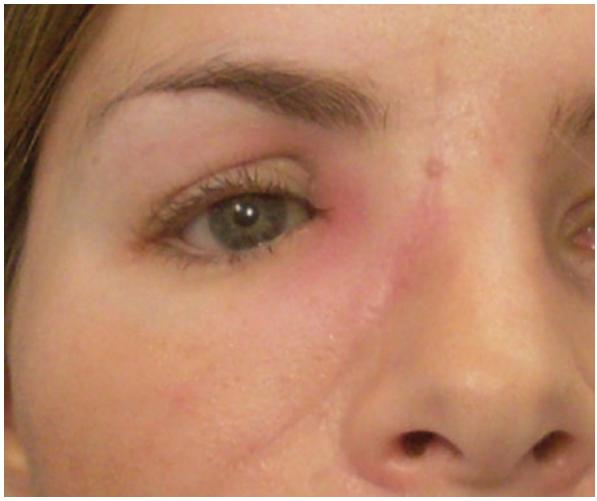

Fig. 19.1 Swelling alongside the nose can have several important causes. Note the displacement of the medial canthus, suggesting lacrimal pathology

maxilla, the alveolar bone is weakest on the buccal side throughout. Infections often present as worsening toothache. Initially the infection is contained within the supporting bone, resulting in exquisite pain on biting or tapping ("tenderness to percussion"). However if left untreated the infection may either discharge via the overlying bone, or spread into the surrounding fascial spaces. Initially the tissue planes serve to contain the spread of infection, but eventually these are breached and a rapidly spreading cellulitis occurs. In other patients a small abscess may spontaneously discharge into the mouth, or less commonly through the skin. Several facial spaces exist which can become distended with serous fluid, pus, blood or (rarely) tumour. Soft tissue fascial space infections specifically related to the maxilla and midface include the buccal space and canine fossa, although infection can rapidly spread to involve other nearby spaces.

19.1.1.1 Buccal Space

This is the most commonly infected tissue space. Patients often presents with a typical 'fat face'. Infections here arise primarily from mandibular or maxillary premolar or molar teeth, the apices of which lie outside of the buccinator muscle attachments. They are readily diagnosed because of marked cheek swelling but there is minimal trismus or systemic symptoms. The space lays anteriorly between the buccinator muscle and superficial fascia and skin. Posteriorly it passes between the masseter muscle and parotid fascia, and the overlying platysma. Inferior it is bounded by the insertion of the parotid fascia into the mandible, superiorly by the zygomatic arch. The buccal space contains the buccal fat pad. It communicates with the pterygoid space.

19.1.1.2 Canine Fossa

Infections here originate from the maxillary incisors and canines and manifest as dramatic swelling of the upper lip, canine fossa, and frequently the periorbital

tissues. Pain is usually moderate, and systemic signs are minimal. Occasionally, direct extension of infection into the adjoining antrum leads to purulent maxillary sinusitis. The space is bounded by the facial muscles around the orbicularis oris (levator labii superioris, levator anguli oris, zygomaticus minor and major) and the overlying skin. Infections usually originate from the upper canine or first premolar teeth. Depending on the length of their roots, infections can spread either between these muscles, or it can track and discharge into the upper buccal sulcus within the mouth. Non-dental causes of swelling include skin infections.

19.1.1.3 Masticator Space

These infections often originate from the third molar tooth. The masticator spaces consist of the masseteric, pterygoid, and temporal space components. These spaces intercommunicate with each other, as well as the buccal and deeper pharyngeal fascial spaces. The clinical hallmark of infection here is trismus with deep seated toothache. Swelling may not necessarily be prominent since the infection is deep to large muscle masses. When swelling is present, it tends to be brawny and indurated. This Masticator space itself is complex. It is bounded laterally by the temporalis fascia, zygomatic arch and masseter muscle and medially by the medial and lateral pterygoid muscles. The temporalis muscle and mandibular ramus further divide the space into superficial and deep compartments. The superficial compartment contains the submasseteric space below and the superficial temporal space above. The deep compartment contains the superficial pterygoid space (or pterygomandibular space) below and the deep temporal space above. The superficial pterygoid space communicates with the deep pterygoid space. The superficial and deep temporal spaces together are also known as the infratemporal fossa space.

Temporal space infections usually arise from posterior maxillary molar teeth. Swelling may be limited to the side of the face in the preauricular region and over the zygomatic arch and may be confused with a buccal space infection. As infection progresses, the cheek, eyelids, and whole side of the face may be involved. Infection here is potentially very serious and it can extend directly into the orbit via the inferior orbital fissure, resulting in orbital cellulitis. Infratemporal space infections also arise from upper wisdom teeth. Clinically there is marked trismus and pain are present, but there may be very little swelling. Extension of infection into the orbit through the inferior orbital fissure can also occur. It may also extend to the lateral pharyngeal wall, resulting in dysphagia.

19.1.1.4 Parotid Space

The parotid space is enclosed by the superficial layer of the deep cervical fascia. It is formed by the splitting of the deep cervical fascia, to enclose the parotid gland. The fascial covering is generally thin, but thickens to form the stylo-mandibular ligament. This is thus an incompletely enclosures space because the superomedial aspect of the gland is not covered. Infection can therefore track superiorly and into the parapharyngeal space. The parotid space contains the parotid gland, the parotid lymph nodes, the facial nerve, the external carotid artery and retromandibular vein.

Infections in the parotid space often occur in dehydrated, debilitated patients with poor oral hygiene who develop salivary duct obstruction. Pain, oedema, and erythema in the region of the parotid are typically observed with fever. Trismus is a later finding.

19.1.1.5 Upper Lip

Infections involving the upper lip can result in severe swelling, usually deep to orbicularis oris. These usually drain into the mouth. They are commonly due to periapical infections of the upper incisor teeth.

Whilst the majority of dental infections can be treated simply with antibiotics and appropriate dental treatment, once they have spread beyond the confines of the alveolar bone they are potentially serious infections. Infections can then rapidly spread throughout the fascial space and patients can quickly become unwell. Several well described pathways are known. These include

i) Infections related to the posterior teeth of both the maxilla and mandible commonly spread to the submasseteric or pterygomandibular spaces.
ii) Infections from these teeth can also track upwards into the infratemporal fossa between the attachments of the lateral pterygoid and temporalis muscle and into the temporal fossa resulting in to scalp abscesses.
iii) Infections can spread into the paranasal sinuses and further still, into the skull, meninges, cavernous sinus/other sinuses and brain via the pterygoid plexus.
iv) Infections have also been found to spread downwards via the neck into the chest wall, mediastinum and the pericardial, pleural and prevertebral spaces.

19.1.2 Spread of Infections

Spread beyond the confines of the local area and along the fascial spaces usually occurs from a spreading bacterial infection of dental origin. However, other possible causes include maxillary sinusitis and infections originating in the skin. Although initially mild, if untreated they can rapidly progress and in some cases become life threatening. The direction and extent invasion depends on the local anatomy, including which tooth the infection has originated from and in which jaw. Virulence of the organism and the patient's ability to fight infection ('host resistance') are also important factors. Severely immunocompromised patients (notably diabetic) are particularly at risk for rapidly spreading orofacial infections and bacteremia. The face has a very rich blood supply which typically helps in its resistance to infection. However, the rich valveless venous network communicates with the cavernous sinus, thus potentially allowing infections to drain retrogradely into the cranium. These venous communications are mostly around the orbit (angular and ophthalmic veins) and via the pterygoid venous plexus.

Clinical features of fascial space infections vary from the ambulant patient with mild cellulitis to the gravely ill, toxic and bed bound patient, requiring urgent admission and drainage. Worrying features include

i) Facial swelling and pain
ii) Fever and chills
iii) Halitosis
iv) Bleeding gums with minor trauma
v) Facial or neck swelling and tenderness
vi) Inability to open the jaw (trismus)
vii) Difficulty in swallowing (dysphagia)
viii) Dyspnea with inspiratory stridor

Patients can present with a localised, or more commonly diffuse swelling on the face. There may also be cervical lymphadenopathy and a history of antecedent toothache. With advanced infections the patient is often unwell and feverish. Urgent investigations include a full blood count (increased white cell count) and blood cultures. A very raised ESR indicates bacteraemia or septicaemia. If the patient is very ill and dehydrated, the haematocrit will be raised and the Urea and Creatinine will be elevated. It is also important not to forget to take a random serum glucose sample—facial space infections are often the first presentation in an undiagnosed diabetic.

The use of imaging depends on the site and severity of the infection. Intraoral (periapical) views can show the health of the affected tooth/teeth crowns and roots. These are commonly taken in otherwise healthy patients with caries (decay) and periodontitis (gum disease). Alternatively an OPT provides a very good first line survey image. Whilst plain soft tissue x-rays are also useful to see swelling and gas in the soft tissue spaces, these are more or less obsolete now due to the availability of far more precise methods. Occipitomental and true lateral views are useful if sinusitis is suspected, these will typically show opacity of the sinus and sometimes a fluid level. Today CT scanning is the first choice of imaging for clinically significant soft tissue infections, or infections involving bone. Abscesses and inflammation can be defined and used as a baseline for comparison later. Magnetic resonance imaging may be used in selected cases if the differential diagnosis includes a tumour. Although MRI demarcates soft tissue infection well, CT scan is cheaper, more readily available and has no adverse effects on patients with metallic implants. Ultrasound may be used to determine if a swelling is oedematous only, or contains pus. This is to some extent academic—if the swelling is significant, it still needs to be drained, even if it is just oedema.

All infections in the head and neck region should be assessed and managed with urgency. Neglected they can quickly result in major complications including sepsis, shock, orbital cellulitis, intracranial infections or airway related issues. With regards to midface infections, indicators of severity include

i) Trismus
ii) Difficulty swallowing
iii) Difficulty talking
iv) Swollen eyelids

v) Blurred vision
vi) Headaches
vii) Gross swelling
viii) Systemic upset
ix) An obvious fluctuant swelling.

Not all fascial space infections require urgent admission. If minor they can be prescribed antibiotics and referred to their own dentist for urgent assessment of the teeth. Odontogenic orofacial space infections are usually polymicrobial involving both strict anaerobes and facultative bacteria found within dental plaque and gingival crevice. The most prevalent anaerobic bacteria include gram-positive cocci, such as *Peptostreptococcus* spp., and gram-negative rods, such as *Bacteroides* spp., *Fusobacterium* spp., *Prevotella* spp. and *Porphyromonas* spp. The most prevalent aerobes are facultative gram-positive cocci such as *Streptococcus mutans,* and viridans streptococci. Facultative gram-negative bacilli and *S. aureus* (including MRSA) are uncommon in immunocompetent hosts but may be more important in immunocompromised patients. Empiric antimicrobial therapy should therefore be initiated promptly in such patients. The choice of specific antimicrobial regimens for odontogenic orofacial infections is empirical based on anticipated causative pathogens and immune status of the host. Currently penicillin plus metronidazole or a β-lactam-β-lactamase inhibitor combination (e.g., ampicillin-sulbactam) is one suitable regime. Clindamycin, doxycycline, or moxifloxacin is an alternative for penicillin-allergic patients. In immunocompromised hosts, additional broad-spectrum coverage for facultative gram-negative bacilli should be considered. Methicillin-resistant *Staphylococcus aureus* (MRSA) infection should be treated initially with vancomycin, or ideally after discussion with your local microbiologist.

Further treatment depends on the extent of the infection, its location, the patient's general health and the response to previous treatments. Drainage of buccal, canine fossa and submasseteric abscesses may be possible intraorally, or via the neck (for buccal and submasseteric abscesses). Ideally the incision is placed in a suitable skin crease, although this is not the main consideration. A retromandibular approach enables drainage without going through masseter muscle, but gravity-aided drainage may then not be possible. It is also important to avoid injury to the retromandibular vein, external carotid artery and facial nerve, all of which are at risk. Submandibular access avoids these structures and drainage under the influence of gravity is better. Incisions are classically taught to be 'two finger-breadths' below the lower border of the mandible to prevent injury to the mandibular branch of the facial nerve. For other midface abscesses intraoral drainage may be possible. An incision in the upper labial or buccal sulcus directly over the abscess will quickly enter the cavity. With more extensive abscesses an incision along the anterior border of the ramus of the mandible can provide access to the submasseteric and pterygomandibular spaces. At the same time the causal tooth/teeth should be removed. Surgical bone removal should be avoided if at all possible as his increases the risk of osteomyelitis.

19.1.3 Maxillary Sinusitis

Sinusitis is an inflammatory process involving the mucosal lining of the paranasal sinuses. Because infection causes inflammation of both the sinuses and the nasal cavity, the term "rhinosinusitis" is commonly used—see also the chapter on the nose. A major underlying factor in the aetiology of acute sinusitis is blockage of the sinus ostium. This commonly affects the maxillary sinus opening, just under the middle turbinate (Figs. 19.2, 19.3 and 19.4).

As a result mucus is retained, stagnates and can then become secondarily infected. Viral infections of the upper respiratory tract are implicated in most cases of rhinosinusitis, but other conditions such as allergic rhinitis, asthma, aspirin sensitivity, immunosuppression, genetic predisposition, cigarette smoking and chronic dental infection may be responsible. These all contribute to inflammation around the osmium and impaired sinus drainage. It has also been suggested that oxygen depletion within the sinus (from normal gas resorption) can result in a negative pressure, which promotes the ingress of bacteria from the nasopharynx. Organisms include S. pneumoniae, H. influenzae and M. catarrhalis. Nose blowing also forces organisms into the sinus. Patients can present with nasal obstruction/congestion, nasal discharge (anterior or posterior nasal drip), toothache and facial pain or pressure, particularly over the cheeks. These commonly develop following an upper

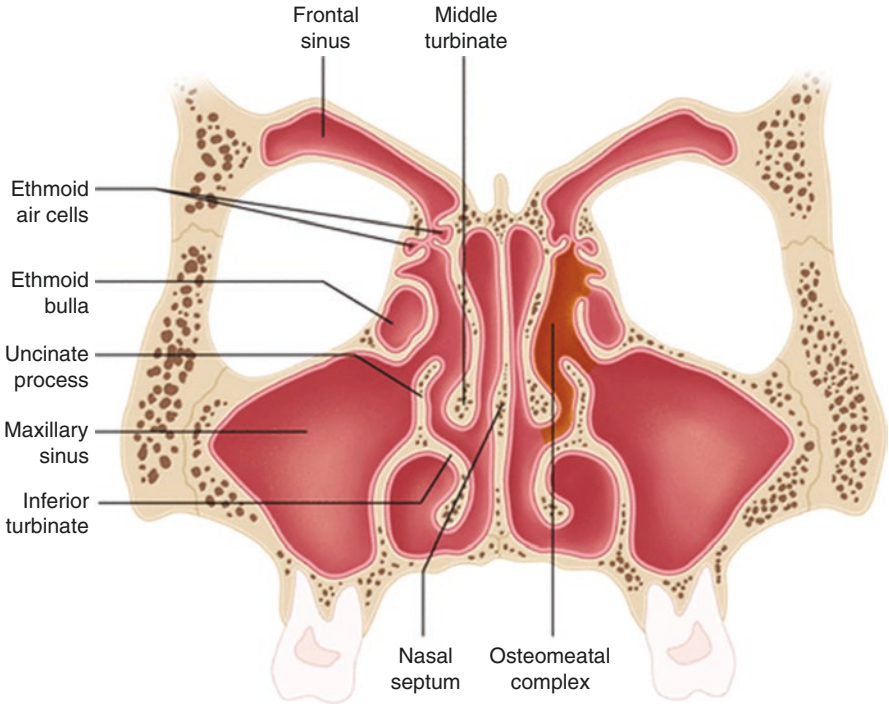

Fig. 19.2 Anatomy of the paranasal sinuses and orbits

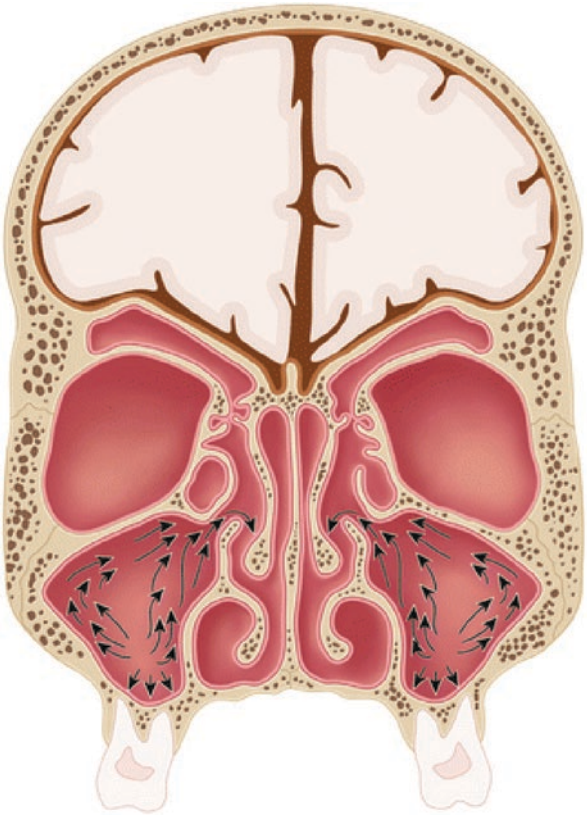

Fig. 19.3 Schematic of mucociliary clearance. A coronal section through the skull, including the paranasal sinuses. Mucociliary clearance from the maxillary sinus is represented by the arrows showing directional sweeping to move mucus out of the sinus by the ciliated sinonasal epithelium

respiratory tract illness. In a small number of patients periapical infections arising from one or more upper molar teeth, or oro-antral communications following extraction of a molar tooth, can also result in bacterial sinusitis. Patients with 'odontogenic' maxillary sinusitis comprises about 10% of all bacterial sinusitis. Patients with AIDS are at increased risk of invasive fungal sinusitis. Aspergillus fumigatus has been most frequently reported.

Sinusitis can be acute or chronic. In acute sinusitis symptoms persist for a few weeks, but always resolve in under 12 weeks. Patients usually present with a unilateral discoloured or purulent nasal discharge, post nasal drip, localised pain and a fever >38 °C. These infections are mostly viral in origin, although a small number will develop a secondary bacterial infection. Septal deviation, polyps, foreign bodies within the nasal cavity and rarely a sinonasal tumour are predisposing factors. Chronic sinusitis is diagnosed when symptoms last more than 12 weeks. These commonly include nasal blockage, nasal discharge, facial pressure and hyposmia/anosmia. Pain is often less severe but loss of smell is more common. On examination there is nasal inflammation, mucosal oedema and a mucopurulent discharge. Patients describe pain over the maxillary and sometimes frontal sinuses. This is characteristically worse on bending forward and on percussion over the sinus. Swelling is rare (Fig. 19.5).

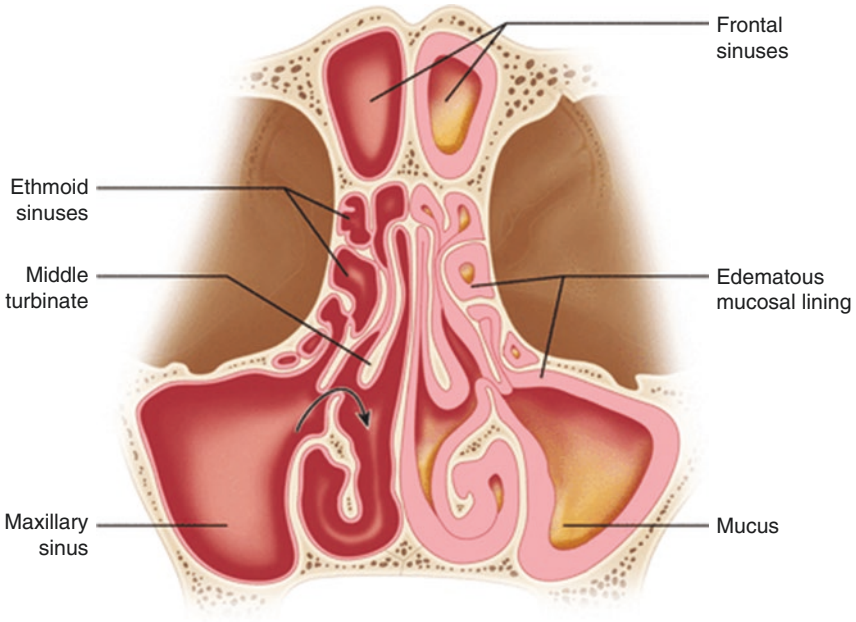

Fig. 19.4 Schematic showing obstruction of normal mucociliary clearance by sinonasal mucosal edema on a coronal section through the paranasal sinuses. On the left, there is normal mucociliary clearance of mucus through the natural opening of the maxillary sinus. On the right, edema of the sinonasal mucosa obstructs the natural opening of the maxillary sinus leading to resultant mucus stasis

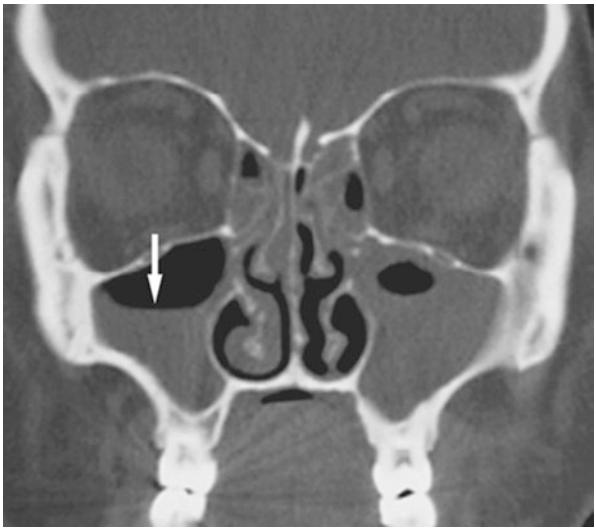

Fig. 19.5 Acute rhinosinusitis secondary to common cold. Coronal CT shows fluid-level in right maxillary sinus (*arrow*), probably also in left sinus, and mucosal thickening of ethmoid cells bilaterally. Normal bone structures

Investigations are not always necessary unless complications are suspected. Plain X-rays are largely obsolete today, but if taken they may show a fluid level in the frontal or maxillary sinuses. CT scans will show fluid and thickened sinus mucosa. Occasionally a maxillary sinus tap may be performed to confirm a bacterial aetiology, but this is not routine practiced and is usually reserved for patients who do not respond to treatment. Nasopharyngeal swabs may be taken. Patients with recurrent or chronic sinusitis should be assessed for underlying allergies. Approximately 60% of patients with chronic sinusitis have allergies to house dust mites, pet fur and fungi. Complications of maxillary sinusitis includes spread to the adjacent paranasal sinuses (pansinusitis) and from there to the anterior cranial fossa and orbit. Cavernous sinus thrombosis, subdural empyema, intracranial abscess, meningitis or focal neurological signs have all been documented. Most cases of orbital cellulitis are secondary to sino-nasal disease, particularly ethmoiditis. Frontal sinusitis may lead to osteomyelitis of the frontal bone (Pott's puffy tumour) (Figs. 19.6, 19.7 and 19.8).

Management of acute sinusitis is mostly medical. Paracteomol or ibrupofen may be prescribed to reduce pain and fever. Intranasal decongestants (maximum 10 days for adults only) and irrigation of the nose with saline may relieve congestion and nasal discharge. It is important to remember that acute sinusitis is caused by a virus in more than 98% of cases and takes several weeks to resolve. Antibiotics are therefore only indicated if there are features of bacterial infection, or if the patient has a co-morbidity that puts them at risk of complications. Commonly prescribed

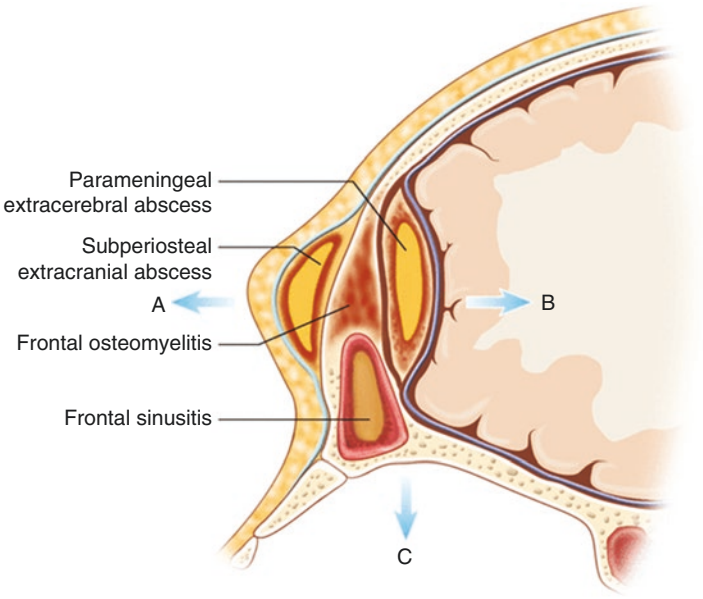

Fig. 19.6 Main topographic complications of Pott's puffy tumor. (**a**) Extracranial extension, (**b**) intracranial extension, and (**c**) orbital extension

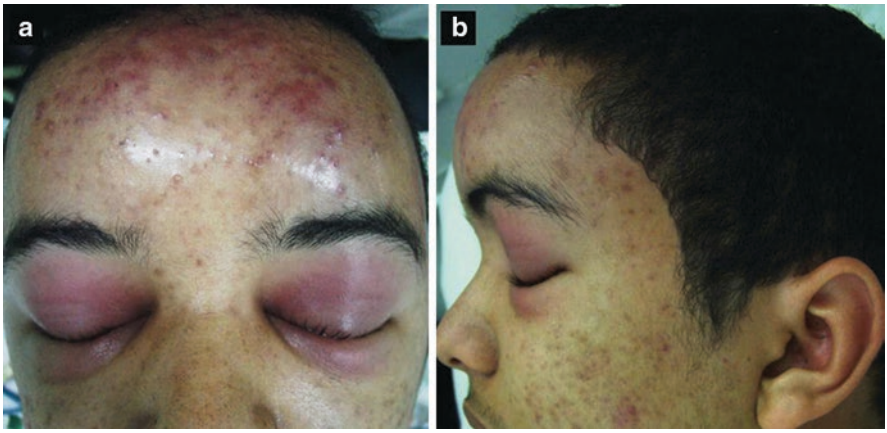

Fig. 19.7 Case 3.1. Bifrontal tender, fluctuant swelling, facial erythema, and periorbital cellulitis in a 15-year-old boy previously treated for paranasal sinusitis. Frontal view (**a**) and lateral view (**b**)

antibiotics are Amoxicillin or phenoxymethylpenicillin for 7 days. Doxycycline erythromycin or clarithromycin may be used if there is a penicillin allergy. Surgery is rarely indicated but sinus lavage (endoscopically or via trephination of the canine fossa) may be required to decompresses the sinus. Treatment of chronic rhinosinusitis usually involves topical nasal steroids, along with treatment of any underlying causes or allergies. Antibiotics may be indicated in patients who fail to respond, or in those who have severe symptoms with evidence of persistent bacterial infection. Endoscopic sinus surgery may be required to improve sinus drainage.

19.1.4 Osteomyelitis of the Upper Jaw

Osteomyelitis can be defined as "an inflammatory condition of the bone that usually begins as an infection of the medullary cavity, rapidly involves the haversian systems and quickly extends to the periosteum of the area". Before antibiotics, osteomyelitis of the jaws was a common and often a fatal infection in the maxillofacial region. However the discovery of antibiotics and other antimicrobial agents and development of better surgical options, plus hyperbaric oxygen therapy (although still somewhat controversial) has greatly improved the incidence and prognosis of this disease. Osteomyelitis of the midface is uncommon and is mostly associated with odontogenic infections. However it can occur following extractions, trauma, or irradiation, and is also seen in patients taking bisphosphonates (MRONJ—see the chapter on the mandible). Osteopetrosis is a rare metabolic bone disease characterised by a generalised increase in skeletal mass. This also increases susceptibility to osteomyelitis of the jaws. However in all these cases, infection is more common in the lower jaw. This is because the upper jaw has a relatively better blood supply. Before antibiotics became available both were frequently fatal. Acute osteomyelitis is less common than chronic osteomyelitis and today patient rarely present with

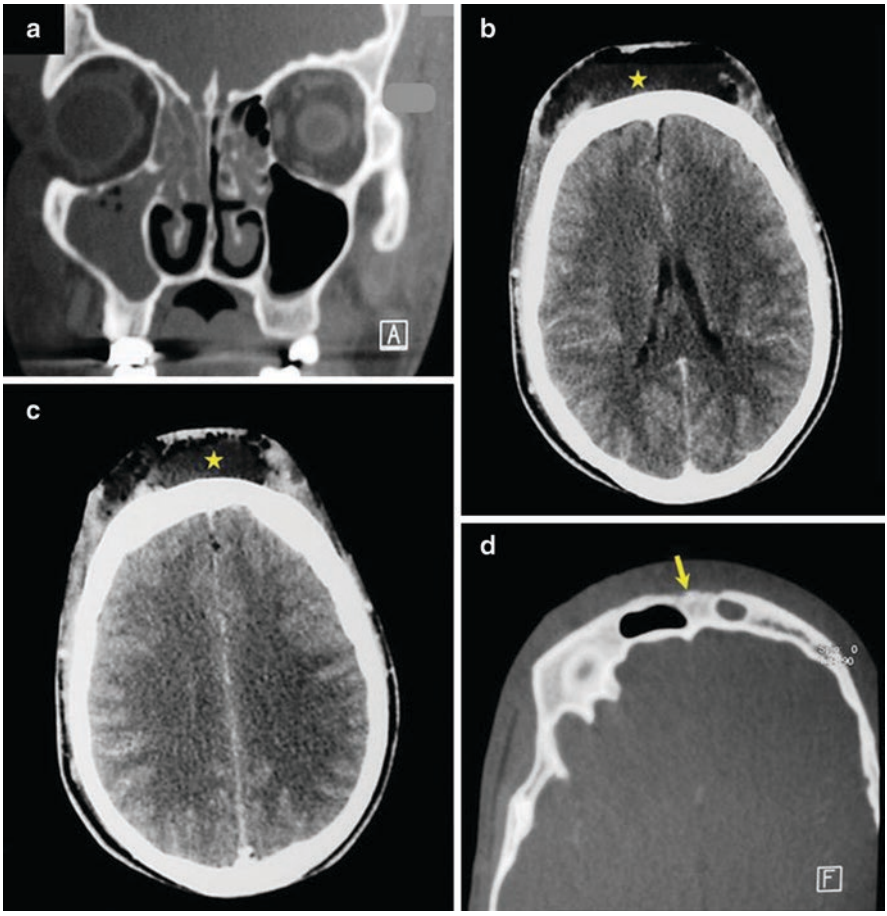

Fig. 19.8 Case 3.1. (**a**) In the same patient, a coronal maxillofacial CT scan shows right paranasal pansinusitis. (**b**, **c**) Axial cranial CT scan (*star*) with bubbles of gas showing the presence of gas-forming micro-organisms. (**d**) Note the erosion of the outer frontal bone table (*arrow*)

obvious suppuration. A small amount of pus exuding from around a tooth is more likely to be a periodontal abscess, however if multiple adjacent teeth are involved, mobile and the overlying soft tissue are inflamed, there is probably some acute osteomyelitis present.

Clinical features of osteomyelitis depend on the site, type and extent of infection and may include pain, swelling, erythema and halitosis. Over recent years there appears to have been a change in clinical presentation. This is mainly due to the increasing incidence and complicating aspects of immunosuppressive illnesses or treatments such as diabetes mellitus, HIV infections and immune suppressing drugs. This condition is therefore commonly seen in smokers, diabetics, alcoholics and other known high risk groups. Rare cases secondary to sickle cell have been reported, presumably secondary to infarction of the bone. In severe infection pain can be unremitting, throbbing and deep seated.

Chronic osteomyelitis can arise from previous acute osteomyelitis following inadequate treatment or systemic factors. It can present with a simple fistula or with purulent discharge and a segment of poorly vascularised, atrophic mucosa found adhered to the bone. Chronic infection has less intense pain, but it is still deep-seated and throbbing. The surrounding tissues are usually soft, swollen, erythematous and tender, secondary to inflammation and oedema, but this may progress to a firm subperiosteal abscess. Intraoral examination may reveal friable, bleeding granulation tissue, mobile teeth and exposed necrotic bone with sequestrum formation in the upper alveolus. In such cases it is important to ensure that these features are not those of a malignancy. Usually there is an obvious source such as a decayed tooth, which may be tender and mobile. Most patients are either malnourished or immune deficient.

Investigation usually include CT and MRI to define the extent of infection and exclude malignancy. Radiography may reveal osteolysis, periosteal reaction or sequestra. The differential diagnosis is wide and includes benign ossifying and non-ossifying fibroma, infection of the minor salivary glands and non-specific chronic lymphadenitis and Ewing's sarcoma, osteosarcoma, chondrosarcoma, non-Hodgkin's lymphoma and metastatic disease of the jaws. Therefore often an initial biopsy is required. Fungal osteomyelitis although rare, also needs to be considered as there is no difference in clinical presentation between this and bacterial infection. Radioisotope 99mTc bone scanning was once popular, but is perhaps less used for diagnosis now due to its poor resolution and better alternatives. Nevertheless it may have a role in follow up and determining resolution of infection. More recently, Positron emission tomography has shown promise in mapping out margins of metabolic activity to differentiate between normal bone and affected bone. This is particularly helpful if resection and reconstruction are indicated for residual non-vital bone.

Infections are usually mixed, comprising of facultative anaerobes, such as the non-haemolytic streptococci viridans group and haemolytic Streptococci. These pass into the bones from the infected tooth or periodontal socket. Haematogenous spread is rare. Specific infections (tuberculosis, syphilis and actinomycosis) are uncommon, although a number of recent reports have suggested that methicillin resistant staphylococci may be more common than previously thought. This should be considered in osteomyelitis which does not respond to treatment. Actinomycosis is an unusual but specific infection which can result in recurrent and chronic jaw abscesses. These can discharge large amounts of pus, containing bright yellow granules ("sulphur granules"). This should be considered in any patient with chronic bone abscesses with discharging sinuses. Fungal osteomyelitis is very rarely seen. Mucormycosis is an opportunistic fulminant fungal infection, which mainly infects immunocompromised patients. This fungus invades arteries leading to thrombosis and necrosis of hard and soft tissues. It is commonly seen associated with diabetic patients. Rhizopus arrhizus, produces the enzyme ketoreductase, which enable the fungus to utilise the patient's ketones. This should be distinguished from non-invasive fungal infections. Aspergillosis is the second most common fungal infection after candida. This can also be invasive in nature when involving the maxillary sinus although it not cause as much bone destruction compared to mucormycosis (Figs. 19.9, 19.10, 19.11, 19.12 and 19.13).

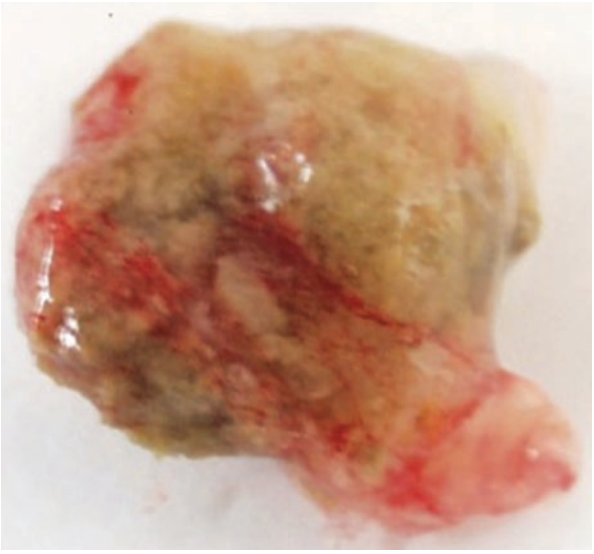

Fig. 19.9 Allergic fungal rhinosinusitis (AFRS). This surgical specimen of sinus contents demonstrates the typical eosinophilic mucin. The gross appearance can be similar to a fungus ball

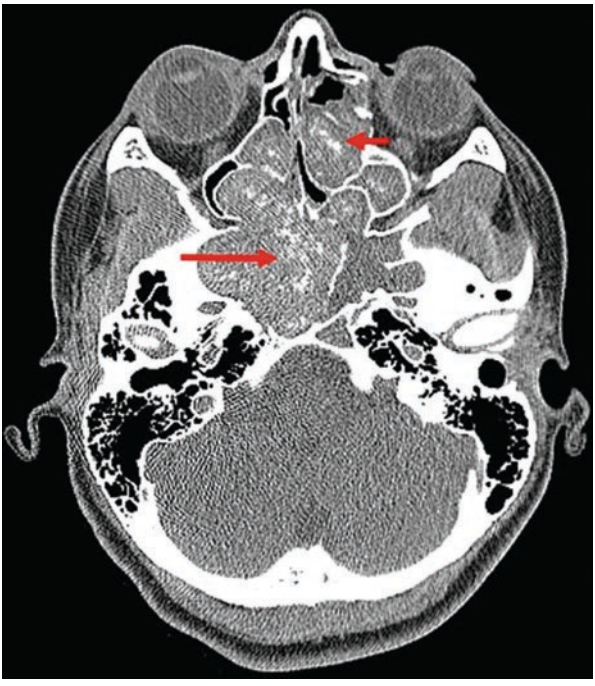

Fig. 19.10 Allergic fungal rhinosinusitis (AFRS), computed tomography (CT). Axial CT shows mucocele formation with hyperdense sinus contents (arrows)

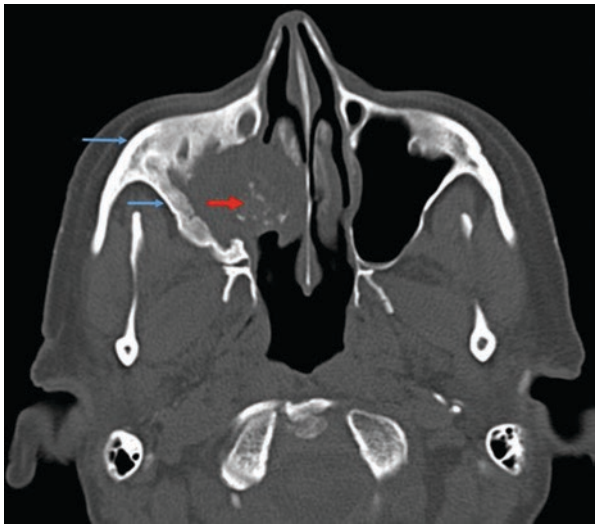

Fig. 19.11 Fungus ball, computed tomography (CT). Axial CT shows an opacified right maxillary sinus with hyperdense material medially (red arrow). Note the pronounced sinus wall hyperostosis (blue arrows) resulting from chronic inflammation

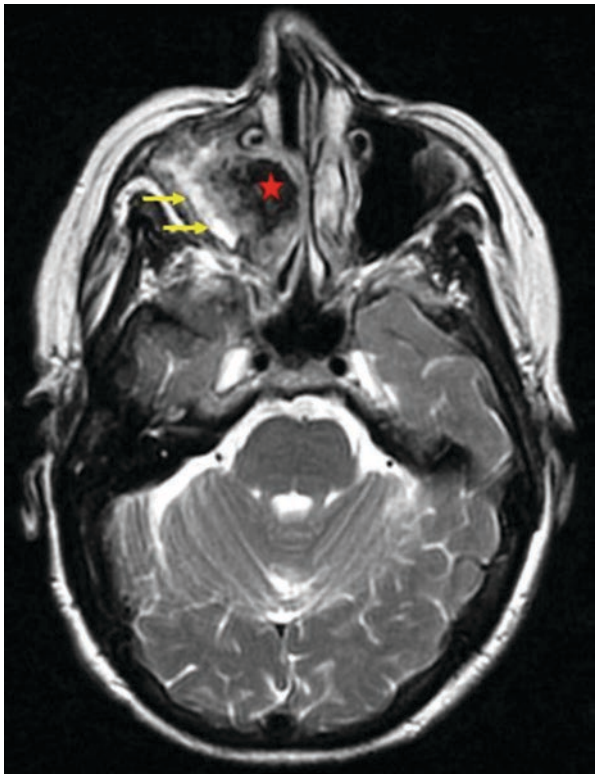

Fig. 19.12 Fungus ball, magnetic resonance imaging (MRI). Axial T2 MRI from the same patient as in Fig. 19.11, showing low signal (red star) corresponding to the region of the medial sinus and nasal cavity containing the fungus ball, and bright signal (yellow arrows) from inflamed mucosa on the lateral sinus walls

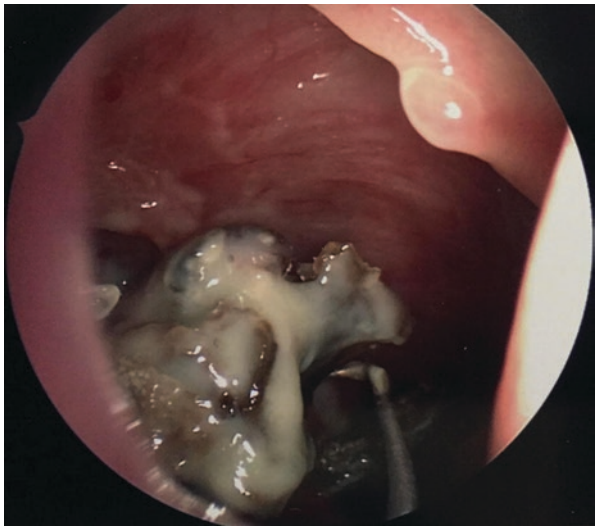

Fig. 19.13 Fungus ball in the maxillary sinus, endoscopic view. Note the variegated debris with variable consistency and surrounding purulent exudate

Acute osteomyelitis needs urgent referral for admission, intravenous antibiotics and drainage of pus. Empirical antibiotics are started while waiting for the results from culture and sensitivity. The correct choice of antibiotic is important as this must be able to penetrate bone effectively. If no local protocols exist discussion with a microbiologist is advised. However, chronic osteomyelitis can often be managed as an outpatient with appropriate long term antibiotics (4–6 weeks) and followup. The decision to admit for intravenous antibiotics depends on a number of factors including the i) severity of symptoms, ii) signs of systemic involvement (rare), iii) the extent of the infection (patients usually require CT) and iv) patient compliance. Any associated contributing factors should also be identified and treated. Surgical debridement is usually required. This aims to provide drainage to the area of infection, remove any sequestra, teeth and foreign bodies and promote tissue perfusion. Treatment ranges from simple sequestrectomy, to segmental resection and reconstruction in recalcitrant cases. Hyperbaric oxygen has also been reported to help in some cases, but the results are variable and access to hyperbaric chambers may not be easy. Itraconazole and fluconazole (along with surgical debridement) are often prescribed for fungal infections.

19.1.4.1 Actinomycotic Osteomyelitis

This is a relatively rare but specific chronic granulomatous infection, which is more common in the mandible. Although completely curable, if not detected early it can result in chronic extensive tissue destruction. Actinomycosis is a saprophytic infection that is characterised by granulomatous and suppurative lesions producing multiple sinus tracts. The organism, Actinomycetaceae are slow growing Gram-positive, non-acid fast, anaerobic or microaerophilic bacteria. Infection can present in an acute, subacute, or chronic form. It may involve only soft tissue or bone, or the two together. Infection spreads contiguously, frequently ignoring tissue planes and

surrounding tissues and organs. Diagnosis is often delayed and is usually based on histopathology following biopsy. Management of actinomycotic osteomyelitis is surgical debridement of necrotic tissue with prolonged antibiotics for 3–6 months.

19.1.4.2 Infantile Osteomyelitis

This is a rare but life-threatening infection that can be easily misdiagnosed. Early diagnosis and treatment is essential. In addition to an infective aetiology, osteomyelitis is predisposed to by genetic, toxic and environmental factors. Symptoms commonly include pain, high fever, rapid pulse, in a clearly unwell child. Untreated severe complications include airway involvement, intracranial infection, dacryocystitis, blindness and death.

Most cases occur secondary to hematogenous spread of bacteria, possibly related to traumatic injuries and open oral wounds. Suppurative mastitis in the infant's mother and osteopetrosis have also been implicated. A common pathogen is Staph aureus. Drainage of pus and intravenous antibiotics are usually required. First choice antibiotics include penicillin and cephalosporin for several weeks. Regular CRP levels are required to monitor the course of the disease.

19.1.5 Odontogenic Cysts and Other Tumours of the Midface

This is a very complex topic. Cysts arising in the upper (and lower) jaw are usually defined as a pathologic cavity partially or totally covered by epithelial tissue. The term "odontogenic" refers to structures that arising from the same tissues that make up the teeth. Odontogenic lesions are an important aspect of head and neck pathology. These lesions arise from odontogenic-forming tissues and consist of two main types i) odontogenic cysts and ii) odontogenic tumours. As a group, these two pathologies form the commonest cause of non-infective swellings in both the upper and lower jaws. They show different degrees of aggressiveness and biological behaviour although the vast majority are benign in nature. However some may develop aggressive behaviour, resulting in local infiltration, bone destruction and a high growth and recurrence rates. The pathogenesis of odontogenic lesions is not fully known but it appears that several different cell types play important and varied roles. Interactions between epithelial and stromal cells are critical in the control of growth and clinical behaviour of these lesions. A wide range of epithelium-associated processes and products have been implicated in the biological behaviour of odontogenic lesions, such as increased expressions of various proliferative markers, impaired expression of tumour suppressor genes and abnormal cell-cycle pathways. Non-epithelial myofibroblasts in the stroma play an important role in the production of extracellular matrix components such as collagen and angiogenic factors. These have been shown to secrete metalloproteinases and TGF-β1 which can remodel the extracellular matrix and affect tumour growth and progression. Myofibroblasts are found in higher concentrations in odontogenic keratocysts and some tumours, suggesting an important role.

Other than a few embryonic cysts which result from the inclusion of epithelium along embryonic lines of fusion (called non-odontogenic cysts), most jaw cysts are lined by epithelium which is odontogenic in origin. These are referred to as odontogenic cysts. Odontogenic cysts account for around 10% of all lesions diagnosed in the oral cavity. Many of these cysts occur in the maxilla. Odontogenic cysts are divided into two main types according to their origin-developmental (such as dentigerous cysts) and inflammatory (such as radicular cysts). Developmental cysts are of unknown origin, but do not seem to result from inflammation. They are usually asymptomatic, but do have the potential to become very large, resulting in bony expansion and erosion. Like odontogenic tumours, some odontogenic cysts can be aggressive.

Today, Oral pathology is a subspecialty of pathology, with pathologists specialising solely in this area of diagnosis and pathologies of the orofacial environment. Consequently the classification of odontogenic cysts and tumours has become very comprehensive and complex. One classification includes

1. Benign odontogenic tumours
   - Ameloblastoma
   - Squamous odontogenic tumour
   - Calcifying epithelial odontogenic tumour (Pindborg tumour)
   - Ameloblastic fibroma
   - Calcifying odontogenic cyst
   - Odontoma
   - Odontogenic fibroma
   - Myxoma (odontogenic myxoma, myxofibroma)
   - Cementoblastoma
2. Malignant odontogenic tumours
   - Malignant ameloblastoma
   - Primary intraosseous carcinoma
   - Malignant variants of other odontogenic epithelial tumours
   - Malignant changes in odontogenic cysts
   - Odontogenic sarcomas
   - Odontogenic carcinosarcomas
3. Non-neoplastic bone lesions
   - Fibrous dysplasia of the jaws
   - Cemento-osseous dysplasia
   - Periapical cemental dysplasia (periapical fibrous dysplasia)
   - Cherubism (familial multilocular cystic disease of the jaws)
   - Central giant cell granuloma
   - Aneurysmal bone cyst (ABC)
   - Solitary bone cyst
   - Traumatic bone cyst of jaw
   - Simple bone cyst of jaw
   - Haemorrhagic bone cyst

This is not an exhaustive list.

Odontogenic cysts are defined as cysts that arise from odontogenic epithelium in the tooth-bearing regions of the jaws. Proliferation and/or degeneration of this epithelium leads to cyst development. They form a diverse group and their histological classification is also very complicated. Cystic jaw lesions may be considered as epithelial or non-epithelial, odontogenic or non-odontogenic, developmental, or inflammatory in origin. The vast majority of patients present with a well defined, corticated, radiolucency in the bone, often discovered by chance following a routine dental assessment. However, growth of some cystic lesions can cause damage to adjacent structures and expand into the maxillary sinus and orbit. This can result in facial asymmetry, loose or displaced teeth and occasionally pathological fractures.

Many cysts can be diagnosed with reasonable certainty from an X-ray, although some require biopsy. Many share similar clinical and imaging characteristics. Often they are related to an unerupted tooth. This relationship is very helpful in diagnosis. Although various radiographic features have been described, most of them first appear as unilocular radiolucencies and later evolve into multilocular lesions. Thus, there is considerable overlap in both the clinical and radiographic characteristics of many these lesions, which makes the diagnosis difficult. In particular, cystic ameloblastomas can be confused with locally aggressive keratocysts. CT and MRI are therefore occasionally required to assess large or deeply sited lesions. Radiographically, some cysts may have characteristic features—some lesions contain radiopaque foci within the radiolucency. Examples include calcifying odontogenic cysts, adenomatoid odontogenic tumours, calcifying epithelial odontogenic tumours and odontomas. The presence and special patterns of calcification may help differentiate these lesions. However all should be referred to a specialist for confirmation. The more common cysts encountered include

- Dentigerous cyst
- Odontogenic keratocyst (may be considered as an intraoral BCC)
- Periapical cyst
- Residual cyst of the jaw
- Traumatic bone cyst of jaw
- Stafne cyst

Other causes of a 'cyst-like' lesion in the jaws include

- Botryoid Cyst
- Ameloblastoma
- Metastases, including lymphoma
- Squamous cell carcinoma invading the bone
- Multiple myeloma
- Periapical abscess
- Giant cell granuloma
- Aneurysmal bone cyst
- Glandular odontogenic cysts

This list is not exhaustive but also illustrates the difficulty in triaging and diagnosis cysts and growths of the jaws. These lesions require histological confirmation as many require definitive treatment. Although odontogenic cysts are benign lesions, carcinomatous degeneration can occur in up to 3% of cases. Long periods of chronic inflammation have been suggested as a predisposing factor to malignant transformation of cystic epithelium. These tumours are sometimes referred to as primary intraosseous carcinoma, or jaw malignancies derived from odontogenic epithelial remnants. Patients present with pain and increasing swelling, although in some cases the tumour may be asymptomatic and is discovered during routine dental radiography.

Several treatments have been described in the management of maxillary cysts, although none has been accepted globally. Decompression, marsupialisation, enucleation and resection of lesions are all undertaken, depending on the size, site and aggressiveness of the lesion. Since many of these also occur in the lower jaw, these are discussed in that chapter.

Like odontogenic cysts, odontogenic tumours are a varied group of lesions with diverse clinical behaviour and histopathologic types. These range from hamartomatous lesions to malignancy. Because these tumours arise from odontogenic tissues, they are also unique to the jaws. Those occurring within the bones are referred to as central, those in the overlying mucosa are called peripheral. Classification of odontogenic tumours is based on their various interactions between odontogenic mesenchyme and epithelium. Thus tumours are classified as epithelial, mesenchymal or mixed, depending on which histological component gives rise to the neoplasm. The majority of benign tumours arise de novo, whereas malignant tumours usually arise from their benign precursor. Treatment is usually excision. Specimens are tested for specific diagnostic and prognostic tumour markers.

19.1.6 Maxillary Tumours

The majority of tumours arising in the maxillary and paranasal sinuses present with advanced disease. Consequently the cure rates are generally poor. Squamous cell carcinoma (SCC) is the most frequent type of malignant tumour, seen in about 70–80% of cases. Carcinoma arising in the bone is an extremely rare condition. Primary intraosseous squamous cell carcinoma (PIOSCC) is defined as "a squamous cell carcinoma (SCC) arising within the jaw bones, which has no initial connection with the oral mucous membrane. These may arise from the lining of an odontogenic cyst or de novo from presumed odontogenic cells. Papillomas are also distinct entities that may undergo malignant degeneration. These cancers initially grow within the bony confines of the sinuses and remain asymptomatic until they erode and invade adjacent structures. Metastasis to the neck is infrequent. Cancer of the maxillary sinus is the most common of the paranasal sinus cancers. Tumours of the ethmoid sinuses, nasal vestibule, and nasal cavity are less common, and tumours of the sphenoid and frontal sinuses are very rare. Industrial exposure to hard wood dust, benzene and nickel may be related to cancer of the paranasal sinus and nasal

cavity. A subgroup of paranasal sinus and nasal cavity SCC are associated with human papilloma virus (HPV) infection. HPV-positive patients may have a better prognosis than those who are HPV-negative.

Patients can present with epistaxis, rhinorrhoea, nasal obstruction, altered sensation infra-orbital nerve, skin erythema and skin fixation. Thought they may occasionally present with oral symptoms of pain, palatal swelling, tooth mobility.

Although there is no cartilage present in the adult upper jaw, chondrosarcoma, a rare malignant cartilaginous tumour with elements of hyaline cartilage differentiation, can also occur. This has been suggested to originate from remnants of the embryonic cartilage involved in nasal and septal development, and from Meckel's cartilage in the mandible. Chondrosarcoma is a slow-growing tumour which in the head and neck can arise in various sites, including the anterior maxilla and the base of the skull. It accounts for about 1% of all chondrosarcomas within the body. On examination, the cut surface of the tumour is said to have a translucent, blue-grey or white sheen, which resembles hyaline cartilage. Clinically, they form a heterogeneous group of lesion with a diverse morphologic features and behaviour. Radiographically, tumours may appear to have poorly defined borders with cortical expansion and destruction. These radiolucencies often contain scattered and foci of calcification. Tumours may be graded on the basis of rate of mitotic activity, cellularity and other histological features, which are important in determining the risks of recurrence and metastasis. 5-year survival is around 90% for patients with Grade 1 tumours and patients with Grade 2 and 3 have a 5-year survival of about 55%. Treatment is multimodal and includes surgical resection and chemoradiotherapy.

Multiple myeloma has also been reported to occur in the maxilla. This condition is one of a group of plasma cell neoplasias, in which neoplastic proliferation of lymphoid B cells occurs. Also included are, solitary bone plasmacytoma and extramedullary plasmacytoma. Multiple myeloma consists of clonal proliferation of abnormal plasma cells throughout the bone marrow, with various degrees of differentiation. The tumour cells usually produce large amounts of monoclonal light or heavy chain immunoglobulins that can be detected in serum or urine. Its cause is unknown, but some occupations, exposure to certain chemicals, overdose irradiation, viruses and genetic factors have been suggested as possible factors.

The disease is more frequently in elderly men and in African Americans.

Symptoms usually result from lytic bone disease, anaemia, renal failure, bone pain, fatigue and recurrent infections. Myelomatous involvement of the maxilla is very rare, but may present as an expansile jaw lesion. Oral manifestations of MM are the first sign of the disease in about 10% of the patients. These include swelling, pain, numbness, bleeding, mobile teeth, xerostomia, root resorption and jaw radiolucencies, with occasional fracture. Diagnosis is confirmed based hematologic and biochemical findings (monoclonal immunoglobulin, Bence-Jones proteins), urinanalysis, the presence of multiple punched-out lesions on skeletal radiography and following biopsy. Treatment involves mainly irradiation and chemotherapy and more lately, autologous transplantation for young patients. Although the prognosis is generally poor, this may be affected by patient's age, tumour site, renal involvement, and histological features.

19.1.6.1 Osteomas of the Upper Jaw

These are rare. An osteoma is a slow growing benign osteogenic lesion that is characterised by proliferation of either the cancellous or cortical bone. Depending on its location, it may be classified as central (arising from the endosteum), peripheral (arising from the periosteum) or extra-skeletal (a soft tissue osteoma that usually develops within the muscle). In the upper jaw and elsewhere in thread and neck, these should be considered in the differential diagnosis of any slow-growing, non-tender, bony hard, lump. The term "ivory osteoma" is sometimes used to refer to osteomas with a very thick cortical component. These can occur within the sinuses, most common in the frontal sinus, followed by the ethmoidal and maxillary sinuses. Most cases are very slow growing and otherwise asymptomatic, presenting as a lump, swelling or asymmetry. The pathogenesis of osteoma is unknown, but has been suggested to be either a true neoplasm, a developmental anomaly or a reactive mechanism to trauma or infection. Multiple osteomas may be a feature of Gardner's syndrome, which includes sebaceous cysts, multiple supernumerary teeth and colorectal polyposis. Lesions appear as a sessile or pedunculated mushroom-like mass. Treatment if symptomatic, is surgical removal.

19.1.6.2 Reparative Giant Cell Granuloma of the Maxilla

Giant cell reparative granuloma accounts for around 10% of all benign lesions of the jaws. Histologically, these are similar to other giant cell lesions such as cherubism and aneurysmal bone cyst (discussed in the chapter on the lower jaw). Granuloma often occur in the maxilla, but they may also be seen in the mandible. The lesion is usually slow-growing and affects children and young adults. More aggressive lesions are rare and despite their benign histological features, may clinically appear malignant. These are not true neoplasms but rather a reactive process, possibly triggered by trauma or inflammation. Granulomas may be classified according to their location, as central (bone) and peripheral (gingival tissues). Patients may present with pain and facial swelling. Although these are generally considered benign, there have been reports of metastasis and malignant transformations to osteosarcoma or fibrosarcoma. Radiological appearances are nonspecific, necessitating biopsy for definitive diagnosis. Treatment is generally surgical. Radiotherapy has been reported to induce malignant transformation. Nonsurgical treatments for small, slow growing lesions may include calcitonin and intralesional corticosteroids.

19.1.7 Extramedullary Haematopoiesis (EH)

During the course of several haematological pathologies (lymphomas, leukemias, thalassemia, myelofibrosis, chronic haemolytic anaemia, etc.), when the haematopoietic function of the bone marrow is insufficient, extramedullary haematopoiesis (EH) can occur. This is defined as haematopoiesis occurring in organs outside of the bone marrow. It occurs in diverse conditions, including foetal development, normal immune responses and pathological circumstances. EH can also occur during an active immune responses to pathogens. Most frequently, this occurs in the spleen

and liver to produce antigen-presenting cells and phagocytes. EH also occurs when the marrow becomes inhabitable for stem and progenitor cells in certain diseases notably myelofibrosis (in which the marrow cells are replaced with connective tissue). EH can also occur in response to increased production of erythropoietin—seen in chronically anaemic patients (e.g. chronic haemolytic anaemia). In the skull, marrow expansion causes widening with thinning of the outer table and thickening of the inner table, with remodelling of the skull. Only rarely does this process involves the facial bones but it has been reported to involve the mandible, maxilla, nose and orbit. Consequently it may be misdiagnosed as sinusitis. In one report a rare case of diffuse craniofacial compensatory marrow expansion was seen in a patient with β-thalassemia. This presented as near-complete obliteration of both maxillary sinuses and partial nasal obstruction.

19.1.8 Osteoradionecrosis (ORN)

ORN of the upper jaw is less common than the lower, due to its relatively better blood supply. Radiation reduces the vascularity of the irradiated tissues by stimulating a pathological process known as endarteritis obliterans. This results in a progressive, hypovascular, hypoxic and hypocellular environment, with diminished cellular activity and poor wound healing. This makes the bone and surrounding soft tissues vulnerable to trauma (including tooth extraction) and infection. If secondary infection develops, it can spread through the non-vital bone. The clinical spectrum of presentation of ORN is wide. Patients usually present with a non-resolving painful mucosal ulcer with evidence of exposed bone or sequestrum. There may also be limitation of mouth opening, usually about 3–6 months following radiotherapy. Other symptoms and signs include orocutaneous fistula, increasingly mobile teeth or pathological fracture. Radiological appearances include a moth eaten appearance to the bone, which is best seen on CT. These findings can easily be confused with malignancy unless a careful history is taken. Management is based on treating any superadded infection, meticulous oral hygiene, analgesia and nutritional support. Surgical debridement may be required, but this is kept to a minimum. In severe or unremitting cases resection of the bone and reconstruction using free tissue transfer may be undertaken.

19.1.9 Medication Related Osteonecrosis of the Jaw (MRONJ)

Medication related osteonecrosis of the jaw has been defined as necrosis of the jaw bone, related or unrelated to dental procedures, persisting for more than 6–8 weeks, which is refractory to conservative treatment, in patients having no history of prior radiotherapy and who have been treated with bisphosphonates for a general disease causing bone resorption. Bisphosphonates are synthetic analogues of inorganic pyrophosphates which were discovered in the late 1960s. They are a class of drugs which influence bone metabolism, specifically by inhibiting osteoclastic action.

Because this reduces bone loss, they are often prescribed in patients with multiple myeloma, bony metastasis, Paget's disease of bone, fibrous dysplasia and osteoporosis. However osteonecrosis can occur as a side effect in both jaws. This is secondary to avascular osteonecrosis of the jaw. Trauma and infection increase the need for bone turnover and therefore these accelerate localised bone necrosis. Until recently, bisphosphonates were administered predominantly as an intravenous infusion. However they are now available in tablet form and can be taken orally. Drugs include alendronic acid (Fosamax), Clodronate, pamidronate and zoledronate. Patients often present between several months to several years after commencement of the drug with pain and swelling affecting the mucosa of the jaw. Fistulae may develop. This may initially be confused with chronic osteomyelitis, ORN or even malignancy. CT usually show regions of mottled bone and sequestrum formation. Other adverse effects include renal failure, arthralgia, fever, muscle pain and hypocalcaemia. Treatment involves cessation of any smoking, meticulous oral hygiene using chlorhexidine and minimal debridement (smoothening of sharp bone edges and removal of loose necrotic bone). Swabs should be taken to look for any coexisting pathogens. Necrotic bone itself is not painful and therefore any symptoms such as pain, cellulitis and cutaneous fistulas are suggestive of secondary infection and may indicate long-term antibiotics (and sometimes antifungal agents). The first choice antibiotic is usually an oral penicillin, tetracycline or ciprofloxacin. Cessation of the drug, if not contraindicated usually helps. This can often allow the recovered bone to demarcated non vital bone which can then be removed. Ozone therapy (Ozonytron for 20 days) has been reported to have a positive effect on bone defects by virtue of oxidation and stimulation of endogenous antioxidant systems. However this is controversial. If long term use of bisphosphonates is required, "drug holidays" (monthly breaks in administration) have been reported to reduce the risks of developing BRONJ. In patients who are to be commenced on bisphosphonates, a thorough examination of the dentition and oral cavity and treatment of any pre existing dental (periodontal) disease should be undertaken at least 1 month before commencement of the drug. Bisphosphonate-associated osteonecrosis of the auditory canal in a patient treated with Zoledronic acid has also been reported.

19.1.10 Paget's Disease (Osteitis Deformans)

Paget's disease is a chronic condition of bone characterised by an abnormality in the normal bone remodelling process. Normal bone is in balance with cells that act to lay down new bone and others to take up old bone. This ability to 'turnover' bone is important for maintaining the normal calcium levels in our blood. Paget's disease is a disorder of bone remodelling that typically begins with excessive bone resorption followed by an increase in bone formation. It is more common in England, Scotland, central Europe and Greece, as well as countries settled by European immigrants. It is uncommon in Scandinavia and Asia. Two types are described. When a single site is affected by Paget's disease, it is referred to as the monostotic type. When multiple sites of bone are affected it is referred to as the polyostotic. Usually the bones

become fragile and misshapen. Over time they become mechanically weaker, expanded, more vascular and more susceptible to fracture than normal adult bone. There is also an extremely rare form of Paget's disease in children, referred to as juvenile Paget's disease. Most patients have no symptoms, but when they occur, the most common complaint is bone pain. Although rare for the maxilla to be involved, patients usually present with bone pain associated with marked deformity. Clinical examination may reveal excessive warmth, from hypervascularity and paraesthesia of the infraorbital nerve as a result of nerve compression. These symptoms may be confused with sinusitis or a tumour. Complications of Paget's disease include

i) Fractures and deformities. Affected bones break more easily. Extra blood vessels in these deformed bones cause them to bleed more during repair
ii) Osteoarthritis. Misshapen bones can increase the amount of stress on nearby joints, which can cause osteoarthritis.
iii) Heart failure. Extensive Paget's disease may require increased cardiac output and subsequent pump failure.
iv) Bone sarcoma. This occurs in less than 1% of people with Paget's disease.

Diagnosis of Paget's disease is made following imaging and lab tests. The first indication of Paget's disease is often abnormalities on plain films. These may show areas of bone reabsorption, enlargement of the bone and characteristic deformities of the disease of bone. Bone scan will reveal "hot spots". Alkaline phosphatase will be elevated. Treatment is not always required if asymptomatic. However, if the disease is active (indicated by an elevated alkaline phosphatase level) and is affecting high-risk sites, such as the skull or spine, bisphosphonates may be prescribed. Calcitonin (Miacalcin), a naturally occurring hormone involved in calcium regulation and bone metabolism may also be required. Side effects include nausea, facial flushing and irritation at the injection site. The outlook is generally good, particularly if treatment is given before major changes have occurred.

19.1.11 Fibrous Dysplasia (FD)

This is a disorder of bone growth of unknown origin, in which normal bone is replaced by immature fibrous bone. It can occur in any part of the skeleton but the skull and face are commonly involved. Patients present with a smooth hard swelling or deformity usually in childhood or early adulthood. During the growth spurt this may become painful. Two types of FD are described.

- Monostotic (Involving a single bone, or adjacent bones, such as the upper and lower jaw
- Polyostotic (Involving many bones).

Leontiasis ossea is a rare form of FD in which there is extensive involvement of the maxilla, with encroachment into the orbital cavities, mouth and paranasal

sinuses. This results in a wide facial appearance likened to a lion's face. The most severe form of polyostotic fibrous dysplasia is known as McCune-Albright syndrome, which includes endocrine diseases (precocious puberty) and skin pigmentation. Fibrous dysplasia may also be associated with Neurofibromatosis. The condition is said to burn itself out during puberty but exceptions are well known. Management include bisphosphonates and surgical contouring of and cosmetic deformity (Fig. 19.14).

19.2 Facial Pain and Numbness

These are rather imprecise, but commonly used terms that are used to describe a wide range of symptoms involving different anatomical regions. "Facial pain" can be perceived in the cheeks, orbits, teeth, side of the face and the head. It can take many forms, from a dull aching quality, to a sharp and incapacitating nature. In many cases a careful history can often help identify the cause of the pain. Due to its wide ranging effects, this topic is therefore also discussed in other chapters in this book. Many patients will have more than one pain diagnosis and may also have an underlying psychiatric or personality disorder which predisposes to chronic pain. This can alter the presentation, perception and management of their pain. Increasingly it is being recognised that some patients have an increased risk of developing chronic facial pain. It is now known that there are neural markers for fear and anxiety and other personality traits which can exacerbate chronic pain.

19.2.1 Facial Pain Syndromes

Trigeminal neuralgia and atypical facial pain are two common conditions which can be very difficult to treat. It is not always possible to distinguish these from each other and other diagnoses, but this is important because treatments and prognoses differ. All types of idiopathic facial pain syndromes should be regarded as a "diagnosis of exclusion", that is, all other causes of facial pain should be considered and if necessary investigated before making this diagnosis. Every now and then patients with unusual symptoms have significant underlying disease. Tumours and demyelinating conditions especially should be considered in all cases of complex facial pain. Idiopathic facial pain makes up a significant proportion of outpatient attendances. Four main symptom complexes are commonly described.

i) Facial arthromyalgia (FAM or Temporomandibular joint dysfunction syndrome)—See the chapter on Lower Jaw
ii) Atypical facial pain
iii) Atypical odontalgia
iv) Oral dysaesthesia (Burning mouth)—see the chapter on Mouth

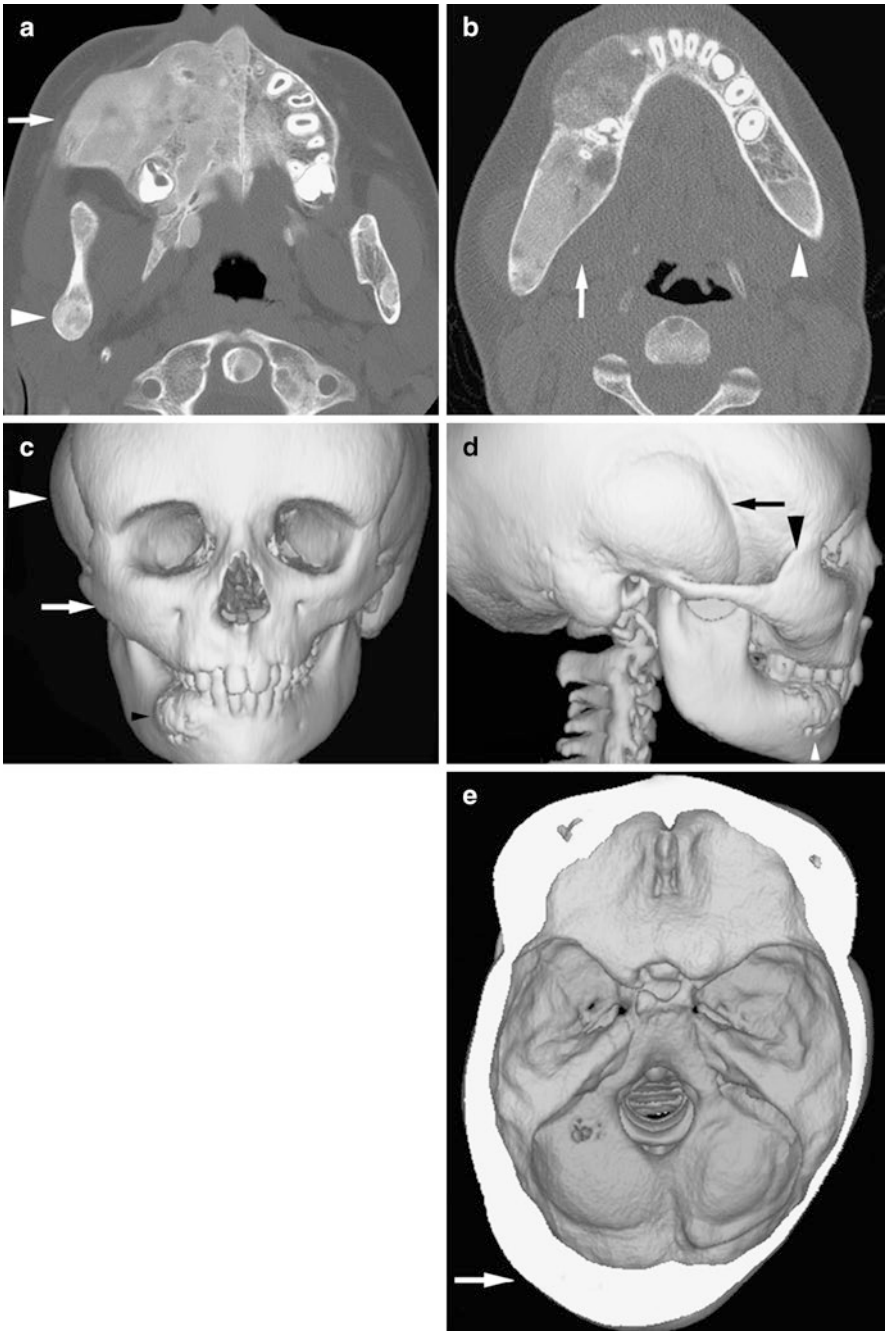

Fig. 19.14 Polstotic fibrous dysplasia right maxilla mandible and temporal bones

It has been suggested that these symptoms may form part of a whole-body pain syndrome, involving the neck, back, abdomen and skin. Stressful events and impaired coping ability are well known associations, but the precise aetiology of idiopathic facial pain remains unknown. Some reports have suggested that stress-induced neuropeptide inflammation within the tissues (e.g. TMJ) causes pain and local production of free radicals. Eicosinoids (a family of oxygenated derivatives of 20-carbon polyunsaturated fatty acids that includes prostaglandins, thromboxanes, leukotrienes, endocannabinoids, and isoecosanoids) have been isolated from affected tissues and are believed by some specialists to be responsible for unexplained pain in non-joint areas including the teeth.

19.2.1.1 Atypical Facial Pain (AFP)

Atypical facial pain (AFP) is classified as a "somatiform pain disorder". This has a psychological component to it and therefore diagnosis may require the services of a pain psychologist. The condition has many distinguishing features that make it a clinical entity in its own right and should not be used as a 'catch all' diagnosis, for seemingly unexplained facial pain. Nevertheless, it is essentially a diagnosis of exclusion that should only be made after all other possible organic causes have been excluded. Afflicted patients therefore often undergo extensive investigation.

Patients with AFP have no identifiable cause for the pain. It is typically described as being a deep, dull ache, sometimes fluctuating, sometimes continuous, with intermittent severe episodes that the patient can find no cause for. Often the pain has been present for several years and analgesics rarely affect its nature. It is most commonly bilateral, ill defined and its distribution cannot be explained on any anatomical basis. Patients may say they are kept from sleeping by the pain but usually look well-rested. When they do sleep, the pain does not wake them. A proportion of these patients may show traits of depressive illness or anxiety states, and they may also experience pain in other sites, such as the back or neck. Irritable bowel syndrome may also be associated. The patient's mood often does not correlate to the description of their symptoms and they may show exaggerated responses to examination and report a stressful life style.

In the first instance it is important to correctly diagnose AFP since treatment for this is entirely medical. Surgical procedures are not indicated. Often the ill-defined nature of the patient's pain results in unnecessary dental work being carried out, sometimes followed by litigation because it didn't work. In light of the association of atypical facial pain with some neuroses (particularly depression), and the suggestion that this has a psychogenic basis, first line treatments often use antidepressant agents. Dothiepin, a tricyclic anti-depressant, has been shown to be effective in reducing symptoms. Selective serotonin re-uptake inhibitors (SSRIs) are also a popular choice. However a clinical appropriate level of depression can also be associated AFP, understandably so, as a result of the negative effect the pain has on the patient's life. Depression and chronic pain may therefore exacerbate each other—the pain predisposing the patient to depression and the depression disrupting sleep

and heightening symptoms. The management of any depression should therefore be equally important and often requires specialist help.

19.2.1.2 Atypical Odontalgia

Atypical odontalgia, also known phantom tooth pain, or neuropathic orofacial pain, is characterised by chronic pain in a tooth or teeth, or in a site where teeth have been extracted or following root treatment, without an identifiable cause. The pain is typically clearly localised by the patient to the dentoalveolar tissues either where teeth are still present or have been lost. Over time, this may move from one site to another or spread to involve wider areas of the face or jaws. It is often described as a severe, continual throbbing pain, which can vary from mild to intense, especially with hot or cold stimuli. The pain may be widespread or well localised and may or may not be relieved by the injection of local anaesthetic. Frequently it is precipitated by a dental procedure and may move from tooth to tooth. It may last a few minutes to several hours. Atypical odontalgia may be a neuropathic pain or it may signify psychological issues. Some studies have found an association between atypical odontalgia and depression and anxiety. However, the significance of this is unclear. The pathologic mechanism seems to be dysfunction or "short-circuiting" of the nerves that carry pain sensations from the teeth and jaws. Areas of the brain that process pain signals have been suggested to undergo biochemical changes that result in a persistent sensation of pain despite a cause. Counselling, avoidance of unnecessary dental treatments and antidepressants form the basis of treatment. Some antidepressants activate serotonin and noradrenaline in the nervous system and thus affect the descending pain inhibitory system of the neurotransmission pathway. However, not all patients will respond adequately to these.

19.2.1.3 Complex Regional Pain Syndrome (CRPS)

This is a form of chronic pain that usually affects a limb, but which can occur following high energy injuries to the face. In some patients the pain can out of proportion to the clinical state. It is an uncommon problem and not well understood. Current reports suggest it is caused by injury or an abnormality to the peripheral and central nervous systems. Symptoms include

i) Continuous burning or throbbing pain
ii) Sensitivity to touch or cold
iii) Sensation of swelling and changes in skin temperature
iv) Muscle spasms, tremors and weakness
v) Decreased function

Symptoms may vary and occasionally spread to elsewhere. They may persist for months to years. The cause of complex regional pain syndrome isn't completely understood. It's thought to be caused by an injury to or an abnormality of the peripheral and central nervous systems. CRPS typically occurs as a result of a trauma or an injury.

19.2.2 Identifiable Causes of Facial Pain

19.2.2.1 Herpes Zoster (Shingles) (See Also the Chapters on the Eye and Lower Jaw)

This is an acute herpetic infection which can affect any sensory dermatome, but most commonly that of the trigeminal nerve—CN V. It is a secondary complication of a previous Varicella Zoster (VZ) virus infection. It usually affects older age groups, often involving the side of the face or forehead and presenting with burning or a tingling pain in the skin along with skin eruptions. These are confined to the distribution of the involved nerve. If eruptions occur near the orbit, or if shingles involves the ophthalmic branch of CN V, involvement of the eye is possible. This it is very painful and debilitating, and is often associated with severe complications. Urgent referral to ophthalmology is required. The condition has a very high incidence in the immunosuppressed. Post-herpetic neuralgia is chronic pain with skin changes following acute herpes zoster. There may be burning or a tearing sensation, or itching and crawling dysaesthesias in skin. In the acute phase, stellate ganglion blocks using local anaesthetic such as bupivacaine, may help for severe pain. Transcutaneous Nerve Stimulation (TENS), capsaicin cream, and tricyclic antidepressants are also useful. Recent reports have demonstrated that Botulinum toxin is effective for neuropathic pain, such as postherpetic neuralgia, trigeminal neuralgia, and peripheral neuralgia.

19.2.2.2 Trigeminal Neuralgia ('tic douloureux')

Trigeminal neuralgia (TN) is a disorder most commonly seen in middle aged and elderly patients. It is defined by The International Association for the Study of Pain (IASP), as "sudden usually unilateral severe, brief, stabbing, recurrent episodes of pain in the distribution of one or more branches of the trigeminal nerve". The International Headache Society (IHS) divides Trigeminal Neuralgia into two distinct categories i) Classical and ii) Symptomatic (or secondary). Classical TN includes patients in which no identifiable cause can be found for their TN, other than microvascular compression of the trigeminal nerve. Symptomatic TN describes those patients in which an identifiable cause can be found, such as a tumour, arteriovenous malformation or multiple sclerosis (MS). Not every patient with TN fulfils the IHS diagnostic criteria and therefore the diagnoses of "atypical" or "type II" Trigeminal Neuralgia has been suggested.

Most commonly the pain involves the lower two branches of the trigeminal nerve. With the exception of TN caused by multiple sclerosis, pain affects only one side of the face. The disorder is more common in women, with a peak incidence between 50 and 60 years of age. In young patients it may be an early feature of multiple sclerosis, HIV disease, or a lesion somewhere along the distribution of the trigeminal nerve (centrally or peripherally). Therefore do not make this diagnosis in young patients until they have been fully investigated. The cause of trigeminal neuralgia is probably multi-factorial with current evidence suggesting local nerve compression within the skull base, altered neuronal processing and possibly a demyelinating process. Neurovascular compression syndrome (NVCS) is defined as

the direct contact and mechanical irritation of the Trigeminal nerve (TN) by a nearby blood vessel. Other similar 'compression syndromes' have been described and include hemifacial spasm (CN VII), vestibulocochlear neuralgia (CN VIII), and glossopharyngeal neuralgia (CN IX). The transition zone between central myelin and peripheral myelin has been suggested to be an anatomical area susceptible to mechanical irritation.

Patients usually complain of a sharp, intense, lancinating pain. It is abrupt in onset and typically lasts only a few seconds (2 min at maximum). Patients may report the pain as arising spontaneously, but these paroxysms can always be triggered by innocuous mechanical stimuli or movements. Patients usually do not experience pain between paroxysms. If they do report additional continuous pain, in the same distribution and in the same periods as the paroxysmal pain, they are considered to have Atypical trigeminal neuralgia (ATN), described below. Pain quickly spreads across the distribution of a branch of the trigeminal nerve. This is almost always unilateral, with over 30–40% of patients complaining of pain in both the maxillary and mandibular divisions. In about 20% of patients, the pain is confined to the mandibular division. Episodes may last up to several hours. It is not unusual for attacks to become more frequent and increasing intensity. Atypical trigeminal neuralgia (ATN), or type 2 trigeminal neuralgia, is a rare subtype with a varied presentation that is more difficult to diagnose. In contrast to trigeminal neuralgia, this can occur at any age. Symptoms often overlap with several other disorders, are a less intense burning or dull pain and can be associated with migraine, temporomandibular joint disorders or other musculoskeletal-type pain. They can vary widely and fluctuate from a mild aching to a crushing or burning sensation. Some patients may experience intense pain in one or in all three branches of the trigeminal nerve. ATN can be bilateral and can be "triggered" by light touch, talking, smiling, chewing, or a cool breeze. Some neurosurgeons have started to report that outcomes are different in these groups.

Currently there are no medical tests that can conclusively diagnose trigeminal neuralgia. However an MRI is usually indicated to look for other causes, notably demyelinating conditions, skull base tumours and neurovascular compression syndrome. Any positive response to medication is usually considered as supporting evidence for the diagnosis, which is otherwise made on the basis of a detailed history and examination. The mainstay of treatment remains medical, typically using anticonvulsant agents. Trigeminal neuralgia usually responds well to carbamazepine and/or amitriptyline, and a muscle relaxant such as baclofen. Carbamazepine remains the drug of choice with an initial regime of 100 mg three times daily being gradually increased to a maximum of 1200 mg daily, titrated against its effect. However some patients may develop side-effects such as tremor, dizziness, double vision and vomiting. Patients should also undergo regular monitoring of liver function and full blood count. Long-term treatment has been reported to cause folic acid deficiency (with megaloblastic anaemia) and hyponatraemia in the elderly. Alternative 'second line' drugs include phenytoin, sodium valproate, lamotrigine, baclofen. If affective, treatment can then be slowly withdrawn after 3–6 months. Surgical management is occasionally required. This

includes cryotherapy to the nerve and alcohol/glycerol injections. Neurosurgical decompression may be required in severe cases following image confirmation of nerve compression. This aims to relieve direct pressure on the trigeminal nerve by separating and padding the nearby blood vessels as they emerge from the brain stem. Alternatively, for patients not suitable for surgery, ablative procedures include radiofrequency thermocoagulation glycerol rhizotomy, balloon compression or Gamma knife. High resolution imaging is used to precisely define the anatomy which then enables a focused beam of ionising radiation to irradiate the proximal trigeminal nerve at its entry point near the pons. Results so far have been very promising with 50% pain relief for 4 years. However there is a risk of sensory changes which can impact on daily life.

19.2.2.3 Anaesthesia Dolorosa/Post Traumatic Trigeminal Neuropathy

This is a term used to define pain following surgical damage to the trigeminal nerve, most commonly at the Gasserian Ganglion after ablative treatment of TN. In other patients there may be a history of nerve injury as a result of facial trauma, dental extraction, root canal treatment, local anaesthetics or placement of implants. Pain usually develops within 3–6 months of treatment. Post traumatic trigeminal neuropathy (peripheral painful traumatic trigeminal neuropathy—PPTTN), can involve all three divisions of the trigeminal nerve to some degree. Symptoms vary from continuous pain, to daily bouts of pain lasting several hours. If there is definite nerve injury there may also be also numbness, dysaethesia, "pins and needles" (often described as "insects crawling over the face"), hyperalgesia and allodynia. Treatment is difficult, and often there is a poor response to medication. Over time, some sensory improvement may occur. Not all traumatic injuries result in pain. Some may result in sensory disturbances only.

19.2.2.4 Facial Migraine

There are several reports describing migraine-like features involving the face usually in the maxillary or mandibular divisions of the trigeminal nerve. Pain is episodic or chronic, severe and associated with nausea, photophobia and autonomic features. These respond to anti-migraine therapy such as triptans (see the chapter on the head).

19.2.2.5 Granulomatosis with Polyangiitis

Granulomatosis with polyangiitis (GPA) most commonly occurs in older adults, although these diseases have been reported at all ages. It is more common among white individuals. Symptoms are often non specific and include fatigue, fever, weight loss, arthralgias, rhinosinusitis, cough and dyspnea, urinary abnormalities, purpura, and neurological disability. They can progress slowly over months or very quickly over a few days. Patients may relapse with symptoms that are different from the initial presentation. It is therefore a very difficult disease to diagnose, often relying of the results of special investigations.

19.2.2.6 Osteopetrosis

Osteopetrosis is noted here, although it can involve other bones in the head and neck. The disease is caused by a failure of the osteoclasts to resorb bone, resulting in impaired remodelling and an excessive accumulation of bone. This defective turnover causes paradoxical increase is fragility in the affected bone. It may also affect tooth maturation and cause nerve entrapment and abnormal growth. Osteopetrosis can occur in infancy, childhood, or adulthood. It is most significant in younger age groups as the condition can also reduce marrow cavity volume and haematopoietic activity. In the head and neck, osteopetrosis can affect in the facial bones, sinonasal region, skull base, and temporal bone. Nasal stuffiness may occur from sinonasal malformation. Mastoid and middle ear malformations may also result in middle ear disease. Entrapment neuropathies can arise from involvement of the skull base foramina, and result in deafness, proptosis as well as hydrocephalus. The mandible can also become susceptible to osteomyelitis due to its compromised blood supply. All affected bones are fragile and can easily fracture.

19.2.3 Facial Numbness and Trigeminal Neuropathy

Facial numbness can be a difficult symptom to diagnose. In most cases either the cause is benign or it cannot be found. Common differential diagnoses for acute or subacute peri-oral and facial numbness include stroke, demyelinating diseases such as multiple sclerosis, tumours, infections, or psychiatric problems. Pathology can involve any or all of the divisions of the trigeminal nerve resulting in varied and sometimes widespread symptoms. Many patients who present with numbness resolve spontaneously, without a definitive diagnosed ever being made. In these cases a presumed viral aetiology is suspected (Trigeminal viral neuropathy). Nevertheless numbness, like pain should be regarded with suspicion, especially if it is unilateral and corresponds to the dermatome of a sensory nerve. This can occasionally be the presenting symptom of significant pathology. Important causes of numbness include

i) Following dental treatment (nerve injury). Numbness is most often seen secondary to trauma. Around 40% of cases report an injury of some type. The most common cause is removal of lower wisdom teeth, where numbness occurs in relation to the lower jaw (ID nerve) or mouth (lingual nerve). Injuries to the midface (notably zygomatic fractures) can injury the infraorbital nerve, resulting in numbers of the cheek, side of the nose and upper lip. Recovery is varied and depends on the severity of injury, treatment provided and age of the patient. In some cases nerve decompression either immediately or within the first 3 months may reduce pressure on the nerve and improve outcomes.
ii) Following trigeminal neuralgia
iii) Viral neuropathy. Temporary dysfunction of the nerve can occur following viral infection.

iv) Demyelinating diseases (notably MS)
v) Tumours (sinus, intracranial, skull base, nerve sheath). Rarely, pain and numbness are an initial sign of perineural infiltration from malignant tumours. Sensory loss within the distribution of the trigeminal nerve, should raise suspicion for the possibility of carcinoma of either the nasopharynx, the cerebellarpontine angle, or the base of the skull. In around half of these cases, numbness precedes other symptoms by several months.
vi) Sinus pathology (including large odontogenic cysts) affecting the infraorbital nerve
vii) Arteriovenous malformation
viii) Peripheral neuropathy (eg vitamin deficiency, diabetes mellitus, alcohol, lead poisoning, exposure to vinyl chloride.)
ix) Ischaemic or haemorrhagic lesions of the brainstem can also cause several isolated cranial nerve palsies. Small lesions in the brainstem can affect the trigeminal nerve only, although isolated trigeminal nerve injury is uncommon. Involvement of the spinal trigeminal tract and its nucleus can present as painful trigeminal sensory neuropathy. Infarcts of the midpons can also present as isolated trigeminal nerve neuropathy involving all three divisions.
x) Trigeminal neuropathy can sometimes herald the onset of systemic sclerosis, mixed connective tissue disease or Sjögren's syndrome. Bilateral trigeminal neuropathy is often related to mixed connective tissue disease or sarcoidosis.
xi) Congenital trigeminal anaesthesia (CTA) is a rare condition characterised by a congenital defect of all or part of the sensory component of the trigeminal nerve in children. It is a heterogeneous condition that can present as an isolated entity, or in association with other congenital abnormalities of the mesoderm, ectoderm and/or brainstem. It usually presents with painless keratopathy in a young child which may be mistaken for herpes simplex keratitis. Other features include a history of numbness or painless lesions on the face and cutaneous ulceration within the trigeminal dermatome. CTA should therefore be considered in all children with a painless keratopathy, especially if there are other features involving the head and neck (facial anomalies, cranial nerve palsies, brainstem-elated symptoms or developmental delay). Most children present in infancy or early childhood.

Trigeminal neuropathy is a relatively frequent problem that can be difficult to diagnose. Although most patients have a unilateral neuropathy secondary to focal lesions, a few present with a bilateral and symmetric numbness. Bilateral trigeminal neuropathy, is often related to connective tissue disease. This may extend to involve the motor component of the nerve. Autopsy studies have suggested that the disease involves the cell bodies of the sensory ganglia and motor nuclei. This has been named "facial onset sensory and motor neuronopathy" (FOSMN). FOSMN may be a primary neurodegenerative disorder or an immune-mediated neuropathy. Bilateral trigeminal sensory neuropathy may subsequently follow one of two clinical courses—in some patients the disease will remain isolated as a sensory neuropathy, in others it will progress to FOSMN. This latter condition is more serious and

potentially life-threatening, with progression of severe motor involvement. It is not known whether these are distinct diseases or similar pathophysiological processes. No diagnostic criteria can currently differentiate between the two and so follow up is important.

The onset of facial numbness thus requires careful investigation and sometimes long-term follow-up. Investigations are tailored towards the suspected cause and may include the following i) Full blood count and ESR/CRP, ii) Electrolytes (notably calcium, potassium and sodium), iii) Liver function tests, iv) Thyroid function tests, v) Vitamin levels and vi) Heavy metal and toxicology screening. Imaging may include MRI or CT. MRI is best for assessing for demyelinating, skull base and other intracranial diseases. CT is indicated in the assessment of the sinuses. However the most appropriate choice may require discussion with a radiologist. Trigeminal motor evoked potentials, nerve conduction studies (NCS), electromyographic (EMG) and nerve biopsy are seldom required. One condition that closely resembles FOSMN is Kennedy's disease, an X-linked hereditary disease that progressively affects primary sensory neurons and motoneurons. This can occasionally present with trigeminal symptoms Therefore all men should undergo genetic testing for Kennedy's disease. Management of trigeminal neuropathy is directed to any underlying cause identified. If none is found reassurance and review is all that is required.