Chapter 18: Non-healing granulomata and tumours of the nose and sinuses

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The non-healing granulomata and tumours of the nose and sinuses both present with nasal obstruction and epistaxis. A tumour-like mass is seen on nasal inspection and the diagnosis is usually made by biopsy.

Non-healing granulomata

Many chronic inflammations of the nose are characterized by the formation of granulation tissue infiltrated by chronic inflammatory cells, namely granulomata. Most of these granulomata are the result of a specific infectious organism and are termed 'specific granulomata'. In others, the aetiology is less clear and they are termed 'non-specific granulomata' or more familiarly to the otolaryngologist as the 'midline non-healing granulomata'.

Generally, the correct diagnosis can be established by either histological or microbiological examination. Table 18.1 lists some of the more common types of nasal granulomata. The specific granulomata are discussed in Chapter 8.

Table 18.1 Nasal granuloma

<table>
<thead>
<tr>
<th>Specific</th>
<th>Non-specific</th>
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<tr>
<td>Syphilis</td>
<td>Wegener's granulomatosis</td>
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<tr>
<td>Tuberculosis</td>
<td>Lethal midline granuloma (midfacial lymphoma)</td>
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<td>Lupus vulgaris</td>
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<td>Leprosy</td>
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<td>Sarcoidosis</td>
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<td>Rhinosporidosis</td>
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<td>Mucormycosis</td>
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<td>Aspergillosis</td>
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<td>Histoplasmosis</td>
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<td>Blastomycosis</td>
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<td>Sporotrichosis</td>
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<td>Leishmaniasis</td>
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Non-specific granulomata

These lesions, more frequently called the non-healing midline granulomata of the nose, have for many years, been the cause of considerable confusion to both clinicians and pathologists. There have been many apparently different clinical entities described with detailed but non-specific histological appearances. Fortunately, over the last decade both
clinical and pathological studies have clarified the situation and most clinicians recognize two main groups: Wegener's granulomatosis and the lethal midline granuloma (nasal lymphoma). Both groups have an appropriate therapeutic regimen and, provided the correct diagnosis is made early, the prognosis has been dramatically improved.

**Wegener's granulomatosis**

Wegener's granulomatosis is a systemic disease of unknown aetiology. It may present to the otolaryngologist at various sites in the head and neck, but primarily it involves the upper and lower respiratory tracts and the kidneys. Other parts of the body are less commonly involved. It is distinguished histologically from polyarteritis nodosa by the typical formation of granulomata.

Wegener's description of rhinogenic granulomatosis in 1939 was a classic paper in which he described both the clinical and pathological features of the condition. The essential histological features are: necrotizing granulomata of the upper and lower respiratory tracts; focal necrotizing glomerulonephritis of the kidneys; and systemic vasculitis.

**Clinical features**

The original concept of the condition was of a fulminating disease leading to early death from renal failure. The patient often presents with a persistent 'cold', complicated by a blood-stained nasal discharge. Nasal examination at this stage reveals thickening of the mucosa with some ulceration and crust formation. The appearances are similar to atrophic rhinitis, but the patient is obviously unwell. Biopsy of the nasal granulations may be reported on as non-specific chronic granulation by an inexperienced pathologist, but careful examination of multiple biopsies will generally demonstrate the diagnostic epithelioid necrotizing granulomata, fibrinoid necrosis and focal vasculitis. However, the rapid clinical deterioration with evidence of systemic involvement of both the lungs and kidneys confirms the diagnosis. The chest X-ray shows localized areas of infarction which may proceed to cavity formation. Urinalysis will often show red cells, casts and proteinuria, and tests of renal function will demonstrate a decreased creatinine clearance; the erythrocyte sedimentation rate is raised. Untreated, the patient rapidly progresses to renal failure and death within six months.

However, it is now recognized that there is a more benign natural history with one or other of the main sites being primarily involved with little obvious involvement of the other sites. Careful investigation will generally confirm the diagnosis by demonstrating decreased renal or pulmonary function. This modified presentation, when primarily involving the nose, is the usual source of confusion to the otolaryngologist. The patient complains of nasal obstruction, crusting and occasional epistaxis, and the usual initial diagnosis is atrophic rhinitis. Sinus X-rays will show a thickened lining membrane, and culture of the nasal secretions often grows *Staphylococcus aureus*. In such cases the true diagnosis will only be obvious on investigation and it is important to obtain an erythrocyte sedimentation rate, urinanalysis, creatinine clearance and chest X-ray. The nasal biopsy is important and should be referred to an experienced head and neck pathologist for an opinion. The need for a confirmatory renal biopsy in such cases is best discussed with the renal physician, for early diagnosis and treatment will prevent the development of the crucial renal failure.
Other sites in the head and neck, occasionally involved either by direct spread or as separate entities, are the middle ear, eyes and orbit, the palate and oral cavity. Systemically the skin, joints, heart and central nervous system may be involved.

**Treatment**

The essential aim is to control the renal involvement as the usual cause of death is renal failure. High doses of steroids (40-60 mg/day) often result in a rapid clinical improvement, but long-term control or cure depends on the use of the cytotoxic drugs, azathioprine or cyclophosphamide. Cyclophosphamide, a widely used alkylating agent, is the drug of choice in North America (used in doses of 2 mg/kg per day). Its main side-effects are well documented, but the occurrence of haemorrhagic cystitis may be confusing in Wegener's granulomatosis. With the long-term use of drugs necessary in Wegener's granulomatosis, sterility, particularly in males, is a problem. In the UK, the drug of choice has been the antimetabolite, azathioprine (3 mg/kg per day). This drug is used widely as an immunosuppressant in transplant surgery. Its main side-effects on the liver and bone marrow are close related and, with the control of the condition, the dosage can usually be decreased. The variation of the dosage requires considerable experience, and is best monitored by the clinical improvement and by the fall in the erythrocyte sedimentation rate. In particularly severe cases, both drugs can be used concurrently with benefit. Long-term use of the cytotoxics beyond the period of active disease is probably necessary to prevent relapse. In dealing with relapses, it is important to exclude concurrent sinus infection in an immunosuppressed patient with disordered nasal anatomy and physiology. Opportunistic fungal infections are not uncommon. After a prolonged period of inactive disease some patients are able to stop all medication, but continued close supervision is necessary. Management of the nasal cavities during the active phase requires regular irrigations and the use of glucose-in-glycerin nose drops to reduce crusting. Sinus drainage surgery is necessary if there is an associated sinus infection. Correction of the common saddle deformity is best left until control of the disease is well established.

**Non-healing midline granuloma**

The title of this condition is satisfactory for current clinical usage, but with a better understanding of the underlying pathology a more suitable term will arise in due course. Clinically, the condition can readily be differentiated from Wegener's granulomatosis - the slow progressive destruction of the nose and midfacial region by an apparent chronic inflammatory response is much greater than seen in the latter. There is remarkably little systemic disturbance and no evidence of pulmonary or renal involvement, death eventually following intercurrent infection or cachexia. Originally described by McBride in 1897, the condition has been of interest and confusion to both clinicians and pathologists over the years with a variety of aetiologies being suggested. More recently the consensus of opinion has been that the condition is probably a malignant lymphoma (Kassell, Echevarria and Guzzo, 1969), the variable clinical picture being the result of different degrees of immunological control in individual patients. Harrison (1974), on clinical grounds, used radiotherapy with success and, in 1977, Michaels and Gregory found common histological features suggestive of lymphoma in a group of patients with this disorder. The features of widespread necrosis with atypical cells were termed 'necrosis with atypical cellular exudate' by them and were considered to be consistent with a histiocytic lymphoma. Furthermore, four of their cases had
similar lymph node metastases confirming their view that this was a malignant lymphoma. More recently Ishii et al (1982), using immunofluorescent techniques, demonstrated that the cell-surface markers of the malignant cells in lethal midline granuloma had the same cell-surface phenotype as is usually found in human peripheral T cells. Consequently, they concluded that lethal midline granuloma was a nasal T-cell lymphoma. T-cell lymphomata tend to develop in such extralymphatic sites as the skin and nasal mucosa. They also found the histological appearance to be consistent with T-cell lymphoma.

**Treatment**

The condition responds well to local radiotherapy, consistent with the histological diagnosis of lymphoma. Initially low non-curative doses appeared satisfactory for control, but with experience, relapses became common and now full lymphoma curative doses are used to the midfacial region and regional lymph nodes. The use of steroids and cytotoxics has been completely without success, supporting the different aetiology to Wegener's granulomatosis. Following curative radiotherapy, surgical debridement and reconstruction can be used to minimize the deformity, or a nasal prosthesis can be worn.

In conclusion, most workers now consider that non-healing granuloma should be subdivided into two categories: the well-accepted Wegener's granulomatosis, and the malignant nasal lymphoma. This latter group contains the polymorphic reticuloses. Whether there is a third small group of 'idiopathic' midline granulomata of non-lymphoma origin is becoming increasingly uncertain and, with full experienced pathological investigation, this possibility will probably disappear completely.

**Nasal tumours**

A wide variety of tumours of different histological type are found in the nasal cavities and paranasal sinuses. The more common types are listed in Table 18.2. Benign tumours are not uncommon, but malignant tumours are rare constituting less than 1% of all malignancies (3% of head and neck tumours). The presenting symptomatology of all tumours is similar and in some cases histological examination is necessary to decide whether any particular tumour is malignant.

**Benign tumours**

**Papilloma**

Squamous papillomata of the skin of the nasal vestibule and anterior septum are quite common. Usually they are treated in the outpatient department by cautery or cryosurgery. However, if they recur they should be excised for histological examination, as the early case of squamous carcinoma of this area is readily curable whereas the missed case is often incurable.

Transitional cell papilloma or inverted papilloma was originally described by Ringertz (1938), and its potential for malignancy was discussed by Osborn (1970) and Hyams (1971. The diagnosis is made histologically, the deep invaginations of the epithelium into the stroma being the typical features. They are usually found unilaterally, and there is a male
predominance of 5:1. They may be present at any age, but are found most commonly in the fifth decade. Treatment is by surgical removal (radiotherapy is not indicated even for recurrences). Local intranasal removal tends to be followed by recurrence, probably due to the difficulty in seeing the extent of the involved mucosa on the complex anatomy of the lateral nasal wall. In those cases where repeated or rapid recurrence occurs, removal by a lateral rhinotomy approach is more effective in preventing recurrence. The important feature of these tumours is their tendency to undergo malignant change, in about 2-5% of cases. It must also be remembered that the papilloma may be present simultaneously with carcinoma in about 5-10% of cases; consequently careful follow-up and examination of all material removed surgically is important.

Table 18.2 Tumours of nose and paranasal sinuses

<table>
<thead>
<tr>
<th>Benign</th>
<th>Malignant</th>
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<tr>
<td>Epithelial</td>
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<tr>
<td>Adenoma</td>
<td>Squamous cell carcinoma</td>
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<tr>
<td>Papilloma</td>
<td>Adenocarcinoma</td>
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<tr>
<td></td>
<td>Anaplastic carcinoma</td>
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<td></td>
<td>Transitional cell carcinoma</td>
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<td></td>
<td>Malignant melanoma</td>
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<td></td>
<td>Salivary gland tumours</td>
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<td></td>
<td>Adenoid cystic carcinoma</td>
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<td></td>
<td>Malignant pleomorphic</td>
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<td></td>
<td>Aesthesioneuroblastoma</td>
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<tr>
<td>Non-epithelial</td>
<td></td>
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<tr>
<td>Fibroma</td>
<td>Fibrosarcoma</td>
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<tr>
<td>Haemangioma</td>
<td>Angiosarcoma</td>
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<tr>
<td>Nasal glioma</td>
<td>Haemangiopericytoma</td>
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<tr>
<td>Neurilemmoma</td>
<td>Meningioma</td>
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<tr>
<td>Chondroma</td>
<td>Chondrosarcoma</td>
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<tr>
<td>Osteoma</td>
<td>Osteogenic sarcoma</td>
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<td></td>
<td>Lymphosarcoma</td>
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<td></td>
<td>Rhabdomyosarcoma</td>
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<td>Plasmacytoma</td>
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<td>Odontogenic tumours</td>
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<td>Fibro-osseous tumours</td>
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Haemangioma

Haemangiомata may be found anywhere in the nasal cavities, but commonly are found on the anterior part of the septum, where they are called the 'bleeding polyp of the septum'. They are probably not true tumours but vascular malformations (Osborn, 1959). Recurrence is common unless the base of the polyp is excised from the septum.
Fibroma

Simple fibromata are occasionally seen as single firm polyps in the nose, but do not grow to any size.

Neurofibroma

Neurofibromata involving any of the nerves inside the nose may grow to quite a large size and a lateral rhinotomy is often necessary for complete excision.

Osteoma and other osseous tumours

Harrison (1984) reviewed this group of tumours and emphasized the need to correlate the clinical features with both radiological and histopathological findings if one is to understand the natural history of each tumour and plan its management. The benign osteomata are the commonest tumours in this group and are often found as an incidental finding in the frontal sinus on X-ray. The majority are asymptomatic and do not progress rapidly. However, they tend to occur at an earlier age in Arabs and often grow to quite large sizes in this ethnic group. Surgical excision is indicated where they cause symptoms and demonstrate an increase in size. Complete removal is essential and the base attachment must be included. As the base is often in contact with the underlying dura, a craniofacial type of approach is sometimes indicated (Cousins, Lund and Cheesman, 1987).

Fibrous dysplasia, originally described by von Recklinghausen in 1891, is now divided into two types - the multiple polyostotic lesions of Albright’s syndrome and the monostotic lesions more familiar to the otolaryngologist involving the bones of the skull. The management of the monostotic foci is discussed fully in Chapter 14, and consists of limited surgery to reduce the cosmetic defect. Radiotherapy may cause malignant change and is contraindicated.

Tumours and cysts of dental origin

Ameloblastoma and dental cysts are mentioned in Chapter 14.

Malignant tumours

Cancer of the nasal cavities or paranasal sinuses is a highly lethal condition and particularly unpleasant by its obvious nature both to the patient and the family. The results in the past have been unsatisfactory with a 30% overall 5-year survival. Frazell and Lewis (1963) commented that the unsatisfactory results could be attributed to a number of factors: the disease was invariably advanced on presentation; the complex anatomy of the region and close relationship to the orbit and skull base; and the reluctance of surgeon and radiotherapist to treat aggressively for fear of increasing the natural mutilation of the disease. Hopefully, over the last two decades there has been an improvement in this situation.

The rarity of these tumours, which constitute less than 1% of all malignancies (3% of head and neck tumours), means that many primary physicians will not see a single case in the whole of their professional careers. Their consequent relative unawareness of the condition
and the similarity of the symptoms with the more common inflammations of the upper respiratory tract results in a failure to think of the true diagnosis before the tumour extends beyond the bony margins of the sinus. The average delay between first noticeable symptom and diagnosis is 6 months. Unfortunately, this situation is unlikely to improve in the UK due to the scarcity of primary physicians with extra training in otolaryngology. Hopefully, in other countries with a higher proportion of otolaryngologists, earlier presentation will become more common. The greater availability of computerized tomographic (CT) scanning is also likely to result in the earlier detection of these tumours in those patients whose symptoms take them to other specialties such as neurology and ophthalmology.

There is an increasing tendency to refer these cases to major centres, and this often results in better management. The surgeon and radiotherapist being more experienced are more able to provide the very individual treatment regimen that is so often necessary with these tumours. The use of CT scanning enables the precise delineation of the tumour extent, and careful planning of both the radiotherapy and subsequent surgical resection.

Aetiology

The upper jaw is one of the few sites in the head and neck where a definite aetiology has been established for some tumour types.

Adenocarcinoma of the nasal cavity and sinuses is known to be common among woodworkers (Acheson et al, 1968). Esme Hadfield in her Hunterian Lecture (1970) showed the incidence of adenocarcinoma to be 10 times greater in High Wycombe compared with the rest of Buckinghamshire, and Acheson et al (1982) showed the skilled furniture maker, particularly the machinists, had a cumulative risk of at least 1 in 120 during their industrial lifetime of acquiring the disease (similar to carcinoma of the bronchus). The occupational risk was recognized by the Government and, in 1969, in the UK, adenocarcinoma of the nasal sinus in woodworkers in the furniture industry became a prescribed disease under the 1959 National Insurance (Prescribed Diseases) Regulations and it is the duty of the doctor to acquaint patients of their rights. The particular type of wood machined also appears to be significant, the African mahogany being the most dangerous. It is interesting to note that this wood is often used in fires by the Bantu tribesmen of South Africa who have the highest incidence of upper jaw cancer in the world, although in those cases, squamous cell carcinoma is the more common type.

Barton (1977) discussed the role of nickel as a carcinogen in squamous cell carcinoma in nickel workers. In Norway, the modification of the industrial process and a screening programme among the workers has resulted in a decline in incidence.

Pathology

A large variety of different tumour types has been described in the upper jaw and Table 18.2 lists the more common types. The most common histological type is squamous cell carcinoma, present in 80% of cases.

The primary site is not always easy to determine with several different sinuses commonly involved by the time the patient presents. The majority (60%) of tumours appear
to be of antral origin, 30% arise in the nasal cavities, and the remaining 10% arise from the ethmoids. Primary frontal and sphenoid tumours are very rare.

Palpable cervical lymphadenopathy is present in about 15% of cases on presentation. This small figure is because the lymphatic drainage of the paranasal sinuses is to the retropharyngeal nodes and thence to the lower deep cervical chain. Consequently, the early involved nodes are not easily palpated in any area of the neck.

**Presentation**

The presentation of each particular case depends on the primary site, the direction and extent of spread. Nasal cavity tumours present with the nasal symptoms of obstruction and epistaxis. Ethmoidal tumours also present with nasal symptoms, but also may have early orbital symptoms such as proptosis and epiphora, with diplopia being a late symptom. Frontal sinus tumours tend to present solely with orbital symptoms. Sphenoid sinus tumours generally present late to neurologists with neurological symptoms.

It is instructive to look at the potential presentation of antral tumours. Tumours within the antral cavity are unlikely to present early unless fortuitously they involve the infraorbital nerve giving a change in facial sensation, or alternatively bleed giving rise to epistaxis. Any epistaxis in an elderly patient who is not hypertensive requires radiological investigation, but the sinus X-rays are best deferred for 7-14 days to allow resolution of any inflammation associated with nasal packing. When the tumour breaches the antral walls, definite signs and symptoms become more obvious, their exact nature depending on the particular wall eroded.

Invasion of the nasal cavity leads to nasal obstruction and epistaxis and the tumour is often clearly seen. Less commonly, the tumour causes ethmoidal polyposis and apparently normal nasal polyps are seen; hence it is essential to examine histologically all material removed from the nose. Inferior spread to involve the palate and alveolus may result in presentation to the dentist with either an ill-fitting denture or loose tooth. Frank ulceration of the palate is a late symptom. Anterolateral spread into the soft tissues of the face may result in epiphora by involving the lacrimal sac. Facial swelling and disordered sensation are more common. Anterior spread is more likely to result in palpable cervical lymphadenopathy. Posterior spread into the infratemporal fossa and skull base may cause less obvious symptomatology, loss of trigeminal function, and trismus occurring from involvement of the pterygoid muscles. Spread into the nasopharynx may result in deafness as a result of eustachian tube dysfunction. Superior spread into the orbit causes early proptosis by increasing the volume of the orbital contents, direct involvement of the nerves and muscles occurring late.

**Investigation**

The objectives of investigation are to obtain a histological diagnosis and to determine the extent of the tumour. A biopsy may be readily obtained from tumour presenting in the nasal or oral cavities. If the tumour is within the antrum, a biopsy is best obtained by an intranasal antrostomy which will also provide drainage during radiotherapy. Biopsy by a Caldwell-Luc approach was previously recommended because it allowed better visualization of the tumour within the sinus and also palpation of the antral walls. This is more
appropriately done by CT scanning, and there is always a potential danger of removing the bony barrier between tumour and the facial soft tissues, even if subsequent radiotherapy 'sterilized' the area. In making the biopsy good representative samples must be taken, and any overlying necrotic tissue must be previously removed.

The extent of the disease is best determined radiologically and the modern CT scanner has revolutionized the assessment, both by demonstrating the full posterior-superior spread of the tumour, and also by showing where a more conservative approach is possible, such as around the orbit. Unfortunately, the full potential of CT scanning is not always realized particularly in departments where these tumours are seen uncommonly. The section of the most appropriate tomographic cuts and the variation in window settings are important to demonstrate the extent of both bone and soft tissue involvement. In the absence of an experienced radiologist, these are best determined at the time of the scan by discussion between the clinician and radiographer. Scans performed by a radiographer alone, and subsequently sent for later reporting by a radiologist, rarely give the necessary information. Axial cuts are readily obtained on all machines, but the present coronal and sagittal reconstructions offered by some machines are unsatisfactory for the detail required, and true coronal cuts obtained by putting the patient through the scanner in the submentovertical position are much better. The use of contrast is of particular value in assessing soft tissue involvement.

Magnetic resonance (MR) imaging is becoming more widely available; this allows scanning in three planes and, with increasing experience, promises to be able to distinguish more accurately between tumour spread and a mucocoele in adjacent sinuses.

Angiography apart from vascular tumours has little benefit to offer.

Nasoendoscopy may be valuable in early tumours, but with the tumours usually encountered the associated bleeding and mucocoele formation makes the procedure difficult, and the information obtained is generally inferior to radiology.

**Classification**

The classification of tumours is considered an essential feature of cancer management. It enables the individual clinician to plan an appropriate method of treatment based on experience, and also allows comparison between different treatment regimens. Unfortunately, the complex anatomy and late presentation with extensive disease involving more than one site has made classification difficult, different systems have been suggested and there has been a failure to agree on a standard UICC (International Union Against Cancer) classification. The first practical classification was proposed by Sebileau (1906) and he divided the upper jaw into three regions: suprastructure, mesostructure and infrastructure. Lederman (1970) adapted Sebileau's classification to the TNM system, but unfortunately it has not found wide acceptance. Harrison (1978) has discussed in detail the problems with the various classifications. The main objection to all systems is the doubt that each case can be accurately recorded as to site and extension. With the advent of CT scanning, this objection has been largely overcome and it is to be hoped that a universally acceptable classification will be found. At the present time the only accepted classification is the American Joint Committee on Cancer (AJCC) system for the maxillary antrum.
Treatment

There is no widespread agreement on treatment regimens for upper jaw cancer. This is partly because of the relative rarity of the tumours, which means few clinicians develop a wide experience, but is more because of the lack of a standard system of classification which prevents meaningful comparisons between different centres.

The wide variation in tumour extent on presentation, and the differing response depending on histological type, means that each case must be considered separately, and the management planned on general principles.

The condition is naturally mutilating and death tends to be delayed and unpleasant; consequently many patients require some form of active treatment, if only for palliation. No single modality of treatment has been shown to be successful (Robin and Powell, 1981) and in most centres a combination of radiotherapy and surgery is used. Failure to achieve cure is generally due to local disease and, despite many different drug combinations, the use of systemic chemotherapy has not shown any significant benefit. Local chemotherapy with the intra-arterial infusion of cytotoxics has been disappointing apart from control of pain, and this can usually be more easily achieved by other means. More recently the use of topical 5-fluorouracil cream in conjunction with low dose irradiation and subradical surgery has shown good results (Kneght et al, 1985). The present author, however, has had little success with this modality in palliation following radical surgery, and more evidence is needed before widespread use can be recommended.

Early tumours localized to the primary site can probably be cured by either radiotherapy or surgery, but for the vast majority of cases full dose radiotherapy followed at an interval of 4-6 weeks by radical surgery is the recommended approach. Preoperative radiotherapy probably reduces the viability of the tumour, both at the primary site and within the lymphatic drainage area not encompassed by any subsequent surgery. With the modern sources available, the irradiation is concentrated in the tumour area sparing the skin, ensuring little subsequent surgical complication. Some clinicians advocate primary surgery and postoperative radiotherapy in the belief that primary debulking helps the radiotherapy; it also allows the residual areas of tumour to be identified histologically. This latter approach is of value in slow growing tumours, such as the adenoid cystic carcinoma, adenocarcinoma, and chondrosarcoma.

Combined therapy is also indicated for palliation because modern rehabilitation methods ensure little disability even following maxillectomy. Involvement of the facial skin is not a contraindication to treatment, and in practice many such cases do well. The involved area is best excised and repaired with a rotation flap of skin from either the forehead or cervical region, and occasionally microvascular free flaps have a part to play. The use of an on-lay prosthesis does much to restore the patient's confidence and may allow the return to a normal life.

Ideally, an operation should be planned to encompass the tumour in each individual patient, and over the years many different operations have been described. In practice there are two major procedures - lateral rhinotomy and total maxillectomy - and both can be extended by craniofacial resections for extensive tumours.
The basic technique of each operation is well described in the standard textbooks of operative surgery, but many points require special emphasis to ensure good tumour clearance and rapid rehabilitation. The selection of the operation depends on the preoperative assessment, but generally if the palate or zygoma is involved a total maxillectomy is indicated; in most other tumours a lateral rhinotomy will give good tumour clearance and requires little rehabilitation.

Skilled anaesthesia is essential; topical anaesthesia of the nasal mucosa with Moffet's solution and hypotensive general anaesthesia are of considerable benefit to the surgeon.

**Maxillectomy**

**Soft tissue approach**

The maxilla is best exposed by the Weber-Fergusson incision. The transverse limb should be placed close to the lid margin to prevent postoperative oedema of the lower lid and, in the medial canthal region where the potential for skin loss as a result of radiotherapy is greatest, it is helpful to curve the incision forward over the nasal bones for additional support postoperatively. A midline upper lip incision, if closed in three layers, is cosmetically more acceptable than a stepped incision. The mucosal incision along the midline of the hard palate swings laterally at the junction with the soft palate passing behind the maxillary tuberosity and then round the alveolus anteriorly. The facial skin flap is raised and all the soft tissue incisions are gently dissected free of the bone to allow the subsequent osteotomies.

**Osteotomies**

The maxilla is freed from the skull by osteotomies through the frontal process of the maxilla, through the body of the zygoma, through the midline of the palate, and the pterygoid plates need to be free posteriorly. The palatal osteotomy is placed in the floor of the nasal cavity and made either with a fissure burr or Gigli saw. The pterygoid plates are best separated from the maxilla with a curved osteotome, and subsequently dissected free from the muscles. The final two osteotomies are made with a fissure burr: medially through the ethmoid cells and frontal process of the maxilla after dividing the lacrimal sac; laterally the osteotomy is made through the body of the zygoma, except for those laterally placed tumours where the zygoma needs to be included in the resection, then the osteotomy is made in the lateral orbital wall below Whitnall's tubercle and through the zygomatic arch.

The remaining bony attachments are the posterior ethmoid cells and posterior antral roof, and these break readily on mobilizing the maxilla. The remaining soft tissue attachments are freed with Mayo scissors, and the maxilla removed. Bleeding from the internal maxillary artery is controlled initially by packing and then by application of a Ligaclip.

**Completion of resection**

Following removal of the maxilla, further tissue must be removed to ensure complete tumour clearance and promote drainage from the remaining sinuses. The ethmoid cells should be exenterated, and both the sphenoid and frontal sinuses opened widely. If there is obvious involvement of the orbital peristeum, orbital exenteration is generally indicated. The support
of the globe is complex and virtually all the medial and inferior orbital walls can be removed without the eye sinking. However, laterally, removal of Whitnall's tubercle gives considerable drooping of the eye which is best corrected by transposing the temporalis muscle medially. Orbital exenteration is achieved by an extraperiosteal dissection and transection of the muscle cone at the apex with Mayo scissors. Bleeding from the ophthalmic artery stops with local pressure. Following orbital exenteration, the eyelids are preserved but the lid margins and tarsal plates are excised to give a smooth skin lined cavity which accepts an on-lay prosthesis satisfactorily.

Postoperative spread into the pterygoid muscles is best managed by an alternative craniofacial procedure as further dissection after maxillectomy is complicated by venous bleeding from the pterygoid plexus.

**Rehabilitation**

Careful rehabilitation ensures minimal cosmetic and functional defect following maxillectomy. Healing of the bony cavity is fairly rapid, but it is advantageous to apply a split-skin graft to the back of the face flap. After resuturing the facial incision, the cavity should be immediately fitted with an obturator. An initial cover plate should have been constructed preoperatively to fit the palate. This is then built up with gutta percha to fill the cavity and to restore the normal facial contours. The main problem with this type of prosthesis is its weight, and help in retention is necessary. Retention is aided medially by creating a ledge on the floor of the nasal cavity by resecting the inferior part of the septum, and laterally a mucosal ledge can be made by suturing the anterior margin of the soft palate to the lateral labial mucosa over a short distance. If the prosthesis still tends to fall into the mouth, it can be secured by a circumzygomatic wire.

The primary prosthesis is generally changed after 10 days, and progressively more sophisticated prostheses can be made over the next 4-8 weeks. The final prosthesis should be no more problem than a bulky upper denture.

**Lateral rhinotomy**

This approach gives good access to the nasal cavities, the ethmoids, the nasopharynx and sphenoid, and also to the pterygopalatine fossa. For more extensive tumours, an *en bloc* resection can be achieved by combining this operation with an anterior craniofacial approach.

The incision is cosmetically very acceptable as it passes along the lateral border of the nose and around the alar margin. The upper end should start just above the level of the medial canthus. The upper lateral cartilage is freed from the nasal bones at the pyriform opening and the soft tissue flap is elevated from the frontal wall of the maxilla and nasal bones. The orbital periosteum is elevated as for an external ethmoidectomy, and the lower part of the lacrimal sac is exposed by nibbling away the anterior lacrimal crest. The orbital contents can then be completely freed medially by dividing the sac low down, and also by freeing the insertion of the inferior oblique tendon and trochlea by sharp dissection from the orbital rim. Access to the anterior nasal cavities can be increased by removing the nasal bones with little cosmetic defect. However, it is more usual to include the lateral nasal wall and ethmoid complex in the resection. The bone is freed by osteotomies cut with a fissure burr - first
through the anterior wall of the maxilla just lateral to the medial antral wall. This osteotomy starts at the orbital rim lateral to the lacrimal fossa and extends inferiorly to the level of the nasal floor. Further osteotomies are made:

1. along the lower border of the lateral nasal wall in the inferior meatus, and
2. through the lamina papyracea just below the anterior ethmoid artery and also across the medial orbital floor.

Finally the upper osteotomy is continued forward through the frontal process of the maxilla and nasal bone. This frees the whole block of the lateral nasal wall and ethmoid complex, apart from their posterior attachments just in front of the optic and sphenopalatine foramen. In this region the bone is very thin and easily fractured by elevating the block medially. Virtually all the mucosa of the nose can be included as a cuff with the main specimen, the posterior and antral mucosal attachments being freed by scissors. The view obtained following the removal of this main block of tissue is excellent and the excision can be extended into the sphenoid and frontal sinuses or alternatively into the pterygopalatine fossa. At the completion of the procedure, the operative cavity is packed with a Whitehead's varnish pack for 7-10 days.

**Other surgical procedures**

*Palatal fenestration*

This operation was originally designed for the implantation of radium into the maxillary antrum, and was claimed to allow good postoperative visualization of the cavity. However, with the alveolus left intact the view is very limited and with modern prostheses the operation has no advantage over the classical maxillectomy.

*Anterior craniofacial resection*

Involvement of the cribriform plate region has long been known as one of the major reasons for failure to control ethmoidal neoplasms; some surgeons even saw it as a contraindication to surgery. Smith, Klopp and Williams (1954) described a surgical approach to this region which was subsequently developed by Ketcham et al (1973) and Clifford (1977). The present author's team has been using a modified craniofacial technique for 10 years (Cheesman, Lund and Howard, 1986) and feels that it must be considered the ideal procedure for most ethmoidal neoplasms. The operation uses a lateral rhinotomy approach for anterior access. This is supplemented by a small midline 'window' craniotomy giving access to the floor of the anterior cranial fossa. After shrinkage of the brain with controlled hyperventilation to reduce end-tidal PCO₂ to 22 mmHg, the dura is elevated from the roof of the ethmoids and cribriform plate and the area is encompassed with a cranial osteotomy. This osteotomy, in conjunction with those of the lateral rhinotomy, allows the en bloc resection of both ethmoid complexes. Involved dura can be excised and repaired with fascia lata. Involved brain can be excised, but cure is unlikely to result at this late stage, although palliation is excellent. The window craniotomy is wired back in place and the soft tissue is closed with remarkably little cosmetic defect. The combined approach not only gives excellent visualization of the ethmoid region, but readily allows extensions of the resection into the sphenoid, the orbit, the pterygopalatine fossa and the skull base centrally. The initial results
of this type of surgery in all series have seen a doubling of the cure rate for ethmoidal tumours to about a 60% 5-year cure rate.

**Lateral craniofacial resections**

The routine use of CT scanning shows that many antral tumours extend posteriorly to involve the infratemporal fossa. In the past, attempts to clear the infratemporal fossa anteriorly have not been very successful mainly due to venous bleeding from the pterygoid plexus. A middle fossa extradural approach to the foramen rotundum allows the roof of the infratemporal fossa to be freed and an *en bloc* resection of the medial infratemporal fossa is possible as part of a classic maxillectomy. The combination of this approach with an anterior fossa resection allows the *en bloc* removal of the orbit for extensive tumours involving the orbit.

**Orbital exenteration**

Attempts to preserve the orbital contents and reduce mutilation have, in the past, often resulted in orbital recurrence, requiring a second operation and decreased cure rate. Ketcham et al (1973) clearly showed that orbital exenteration for involvement of the orbital periosteum doubled the cure rate from 32% to 62% even in more advanced tumours. Consequently, it was the author's initial practice to remove the orbital contents if the orbital periosteum was involved with tumour. Histological examination however showed that the tumour rarely penetrated the orbital periosteum to involve the orbital fat. More recently, the author's team has practised resection of the orbital periosteum and, in the absence of involvement of the underlying orbital fat on frozen section, has preserved the orbital contents. This has been achieved with little disturbance of ocular function, and of 20 cases there has only been orbital recurrence in two cases; both had successful subsequent orbital exenteration. Medial defects in the orbital periosteum are repaired with split-skin grafts, but inferior resections must be repaired with fascia to prevent prolapse of the globe.

**Special problems**

**Malignant melanoma**

Malignant melanoma of the nasal mucosa is very rare, about 1% of all malignant melanomata. Usually they have a pigmented appearance, but amelanotic tumours are not uncommon, and are often diagnosed as anaplastic carcinomata unless the intracytoplasmic pigment is sought. They respond poorly to radiotherapy; chemotherapy is equally unsuccessful, and may aggravate the situation by altering the patient's immune status. Consequently, wide surgical excision is used, but the success of the operation probably depends more on the patient's immune competence. Virtually the whole of the nasal mucosa can be removed by a lateral rhinotomy approach and experience indicates that there is nothing to be gained by a full craniofacial resection. Lymph node metastases in the neck are usually isolated and can be removed individually, there being no evidence to support the use of a radical neck dissection. Local recurrences can often be controlled for many years with no active treatment and symptomatic debulking can be achieved by use of the laser or cryoprobe. Death from melanomatosis often follows some mild immunological challenge such as influenza.
Aesthesioneuroblastoma (olfactory neuroblastoma)

This malignant tumour is derived from the neuroectoderm. It may present to the rhinologist as a nasal tumour or alternatively to the neurosurgeon as an anterior fossa mass. Many of these tumours produce vasoactive hormones, and urinary assays of the metabolites, dopamine and 3-methoxy-4-hydroxymandelic acid have been used to monitor recurrence. The realization that this tumour is distributed on both sides of the cribiform plate and requires a craniofacial resection has resulted in an improved prognosis. For an early tumour, craniofacial resection offers the chance of complete cure, although at this time adjunctive curative radiotherapy is still recommended.

Adenoid cystic carcinoma

These tumours have a propensity to spread along the perineural spaces, and recurrences often occur at distant sites along the course of nerves supplying the primary area, many years after apparently successful local cure. They are not radiocurable, but do respond to radiotherapy. A combination of surgery and radiotherapy is indicated for these cases, surgery being used first to remove the bulk of the tumour and to indicate the likely areas of perineural spread. In the young patient, long-term cure is sought by attempting to include the involved nerves as far centrally as possible. In terms of an antral lesion, this entails a lateral craniofacial approach dividing the maxillary nerve at the foramen rotundum before it enters the wall of the cavernous sinus. If tumour is found within the nerve at this level, it is best controlled with radioactive implants, and iodine-125 seeds, with their long half-life, are a promising source for this type of tumour.

Adenocarcinoma

The adenocarcinomata or wood-workers' cancer generally involves the ethmoid sinuses. It is a relatively slow growing tumour and rarely metastasizes. It is said to be less radiosensitive than the squamous cell carcinoma and long-term cure rates are poor (less than 20% over 5 years). The initial results of craniofacial resections for these tumours in the ethmoid are encouraging and the current policy is to initially debulk the tumour intranasally. This is followed by a full course of radiotherapy and then 6 weeks later by a planned craniofacial resection of the ethmoids.

Tumours of the nasal cavity

These tumours are more common in males and, although 5% are bilateral, there is a predominant involvement of the right side of the nose, possibly related to the trauma of nose picking. The lateral wall is most commonly involved, 50% on the turbinates and, with decreasing frequency, the septum, vestibule, posterior choana and floor. Squamous cell carcinoma is the commonest histological type. As a group the do better than the paranasal sinuses with a 50% 5-year survival. For most sites either radiotherapy or surgery is used, but tumours of the vestibule and septum are best treated surgically. Tumours in these sites look innocuous, but there is a real danger of spread along the tissue planes of the cartilaginous nose and also into the upper lip. Local radical excision with careful histological control of the specimen is essential and, if the tumour is found near excision margins, postoperative radiotherapy should be given. If the tumour is restricted to one side of a cartilage plane local
resection is possible, but if it breaches the cartilage boundary, cartilaginous rhinectomy is necessary. Prosthesis rehabilitation should be used initially until tumour recurrence can be excluded. Delayed nasal reconstruction is possible, but many patients prefer a prosthesis which can be cosmetically very acceptable.