Paparella: Volume III: Head and Neck

Section 2: Disorders of the Head and Neck

Part 1: Nose and Paranasal Sinuses

Chapter 10: Tumors of the Nose and Paranasal Sinuses

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Tumors of the nose and paranasal sinuses are uncommon, and most head and neck surgeons will diagnose only a handful of such cases throughout their career. Malignant tumors of the sinonasal tract account for 0.2 to 0.8 per cent of all human malignancies and only 3 per cent of malignant tumors of the upper aerodigestive tract (Grant and Silverberg, 1970); for example, there is one new case of maxillary sinus carcinoma diagnosed per 200.000 persons annually (Batsakis, 1979). Most patients with tumors of the nose and paranasal sinuses are diagnosed relatively late in the course of their illness, as the early symptoms of paranasal sinus tumors mimic those of chronic sinusitis and it is difficult to adequately examine the paranasal sinuses in the office. Even the newer radiographic imaging techniques offer little help in differentiating between neoplastic and inflammatory processes in this region. As a result, the head and neck surgeon should be aggressive in biopsying nasal and sinus lesions that are in any way suspicious and that do not respond to the usual antibiotic and decongestant regimens.

Tumors of the nose and paranasal sinuses were described by Hippocrates and Galen, who felt that treatment only spread the tumor and shortened the patient's life. The French surgeon Gensoul, in 1827, performed the first total maxillectomy in a patient with osteogenic sarcoma. Rogers, in 1824, was the first American to partially remove the upper jaw and maxilla. At the turn of the century, the use of cautery to destroy tumors in the paranasal sinuses was described. Radiation therapy was introduced after 1910 and was used in combination with electrocautery in the treatment of maxillary sinus tumors (Watson, 1942).

During the first half of the 20th century, antrostomy with curettage and cauterization or radiation was the treatment of choice for maxillary sinus malignancies. Martin remarked on the poor salvage rate for carcinoma of the paranasal sinuses treated with curettage and recommended initial radical surgical removal of the tumor (Martin, 1948).

In 1954, Smith and colleagues reported the first series of cases employing craniofacial resection for carcinoma of the paranasal sinuses (Smith et al, 1954). Of his three patients, one had unresectable disease at the time of operation, one was considered to have achieved palliation, one the other was free of disease at 1 year of follow-up. Since that time, radical surgical excision has become the preferred treatment modality for malignant tumors and some benign tumors of the nose and paranasal sinuses. Radiation as a sole modality is reserved for palliation and recurrent disease in those patients who are too debilitated to undergo complete excision. Radiation therapy is also used in conjunction with radical surgical excision to decrease the risk of recurrence.

Anatomy

An understanding of the anatomic relationships of the midface and anterior cranial fossa is critical to the proper management of tumors of the nose and paranasal sinuses. The paranasal sinuses are thin-walled structures that communicate with the nasal cavity through a series of natural ostia. These anatomic weaknesses provide for easy dissemination of tumor from its primary site to adjacent structures. Frequently, it is the symptoms resulting from extension of the tumor to contiguous structures that finally bring the patient to a physician's attention.

For an in-depth discussion of the anatomic relationships of the nose and paranasal sinuses, the reader is referred to Volume I of this book. Pertinent anatomic highlights will be presented in this chapter.

The *nasal cavity* is divided into two sections by the cartilaginous and bony nasal septum. The septum is relatively resistant to tumor invasion. The nasal septum is frequently seen displaced into the contralateral nasal cavity by the tumor, especially in benign neoplastic processes. The lateral walls of the nasal cavities are perforated by the numerous ostia of the paranasal sinuses and the nasolacrimal duct. The turbinates project from the lateral nasal walls and are more frequently the primary site of tumor development than is the septum. The cribriform plate composes the roof of the nasal cavity. Tumors involving the ethmoidal sinuses usually extend into the anterior cranial fossa through this relatively thin structure.

The *maxillary sinus*, or antrum of Highmore, is the largest paranasal sinus. It is pyramidal, with its apex directed laterally and its base adjacent to the lateral nasal wall. The alveolar process of the maxilla forms the floor of the antrum, and the roof of the sinus is contiguous with the orbital floor. The posterior wall of the maxillary sinus is anterior to the pterygomaxillary fossa. The ostium of the maxillary sinus drains into the middle meatus with an accessory ostia frequently being present. Anterior ethmoidal air cells are often seen bulging into the superomedial corner of the sinus. The infraorbital nerve exits the maxilla through the anterior superior aspect of the sinus.

The ethmoidal sinuses consist of a honeycomb of thin-walled air cells located between the lateral nasal wall and the medial wall of the orbit (lamina papyracea). The roof of the ethmoidal complex is formed by the floor of the frontal sinus and the anterior cranial fossa. Inferolaterally, the ethmoidal sinuses are adjacent to the maxillary sinus. The posterior ethmoidal cells share bony walls with the sphenoidal sinus. The drainage of the ethmoidal sinuses is somewhat variable, with ostia of the anterior air cells present at multiple sites in the middle meatus. The ostia of the posterior ethmoidal sinuses are located in the superior meatus.

The *frontal sinus* is located in the frontal bone and is usually a paired structure; however, lesser bony septa may compartmentalize this sinus. The posterior wall of the frontal sinus composes the anterior wall of the cranial fossa. Inferiorly, the orbits and ethmoidal sinuses border the frontal sinus. Drainage is through the nasofrontal duct, which may empty directly into the nose or first into the ethmoidal infundibulum.

The *sphenoidal sinus* is a midline structure that is divided into two unequal cavities by a septum. Its anatomic relationships make the sphenoidal sinus a critical structure when attempting to assess the resectability of paranasal sinus or nasopharyngeal tumor. Superiorly, the sinus borders the pituitary gland, frontal lobes, and the cavernous sinus. The pons and the basilar artery are located behind the posterior wall, and the nasopharynx is inferior. The carotid arteries lie lateral to the sphenoidal sinus and may be dehiscent within the sinus, protected only by dura and sinus mucoperiosteum. Superomedial to the carotid arteries and anterior to the pituitary gland is the optic chiasm. Drainage into the nasal cavity is through the sphenoidal ostium on the face of the sphenoid bone.

The *nasopharynx* is a complex structure with important anatomic relationships. Anteriorly, the nasopharynx communicates with the nose through the posterior choanae. Inferiorly, it communicates with the oropharynx through the pharyngeal isthmus. The two lateral walls are penetrated by the eustachian tube orifices. Only the superior and posterior walls contain no ostia. Superiorly, the nasopharynx is lined with pseudostratified ciliated columnar epithelium. However, this changes to stratified squamous epithelium at its lower extent. The mucosa of the nasopharynx contains goblet cells, minor salivary glands, and lymphoid tissue. The adenoids usually involute before adolescence. The nasopharynx has abundant bilateral lymphatic drainage. The primary drainage sites are the nodes of Rouviere, which are located in the lateral pharyngeal space at the skull base. Secondary drainage occurs to the jugulodigastric node group.

The nasopharynx is suspended from the skull base by the pharyngeal aponeurosis. Because of its location, there is early involvement of adjacent structures by nasopharyngeal tumors. The carotid artery, cavernous sinus, and cranial nerves III, IV, V1, V2, and VI are frequently involved; the abducens nerve is involved earliest. Extension of tumor into the middle fossa occurs early through the foramen lacerum.

Epidemiology

There is very good evidence for environmental factors contributing to the incidence of both sinonasal and nasopharyngeal carcinomas. In the US and the UK, nasopharyngeal carcinoma is responsible for 2 per cent of head and neck malignancies and 0.3 per cent of all malignancies. In China, cancer of the nasopharynx accounts for 18 per cent of all malignancies and 56 per cent of head and neck malignancies (Digby et al, 1941). This high rate is felt to be secondary to both environmental and genetic factors.

Several occupational groups have been demonstrated to be at increased risk for the development of sinonasal carcinomas. They include nickel refiners, woodworkers, leather workers, textile workers, petroleum refiners, isopropyl manufacturers, and chrome pigment manufacturers (Roush, 1979). Squamous cell carcinoma primarily develops in nickel workers, and these tumors are usually located in the nasal cavity.

Adenocarcinoma is the most frequent tumor found in woodworkers, with the preferred location being in the ethmoidal sinuses or in the superior nasal vault. Forty per cent of the tumors in boot and shoe manufacturers are adenocarcinomas, with the most frequent site being the ethmoidal sinuses. Although there has been no specific carcinogen isolated in either of these two industries, aldehydes, aflatoxin, and chromium are suspected agents (Roush, 1979).

In addition, the Epstein-Barr virus (EBV) has been implicated as a causative organism in nasopharyngeal carcinoma.

Table 1. Tumors of the Nose, and Paranasal Sinuses

Malignant Tumors

Squamous cell carcinoma
Lymphoreticular tumors
    Extranodal lymphoma
    Extramedullary plasmacytoma
Esthesioneuroblastoma
Salivary gland tumors
    Adenoid cystic carcinoma
    Adenocarcinoma
    Mucoepidermoid carcinoma
    Other tumors
Melanoma
Sarcomas
    Chondrosarcoma
    Rhabdomyosarcoma
    Fibrosarcoma
    Angiosarcoma
    Hemangiopericytoma
Metastatic tumors

Kaposi's Sarcoma

Benign Tumors

Papilloma
Inverting papilloma
Meningioma
Neuroma
Chordoma
Juvenile nasopharyngeal angiofibroma
Osseous tumors
    Osteoma
    Ameloblastoma
    Fibrous dysplasia
    Cherubism.

Diagnosis

Symptoms and Signs

The most common symptoms in patients with tumors of the nose and paranasal sinuses are unilateral nasal obstruction (48 per cent), facial or palatal swelling (41 per cent), facial

pain (41 per cent), nasal discharge (37 per cent), and epistaxis (35 per cent) (Tabb and Barranco, 1979). The average delay from the onset of symptoms to diagnosis is 8 months (Gullane and Conley, 1983). This delay is greater for adenocarcinoma than for squamous cell carcinoma. The correct diagnosis is frequently overlooked because other disease processes are thought to be present. Many patients later diagnosed as having carcinoma relate vague past histories of "sinusitis". *Orbital* complaints are less common, occurring in about 25 per cent of patients, and include diplopia, decreased visual acuity, periorbital edema, and proptosis (Schechter and Ogura, 1972).

Oral symptoms include palatal ulcers and swelling. These patients may experience an inability to wear previously well-fitting dentures. Dentulous patients may develop pain referred to the teeth, and many patients undergo dental extractions as the initial therapy for this discomfort. Trismus and trigeminal paresthesias indicate tumor invasion into the pterygomaxillary fossa. Persistent oroantral fistula after tooth extraction should raise the suspicion of a malignant process in the antrum.

Nasal symptoms include the obstructive symptoms of rhinitis and sinusitis, which are identical to those of early carcinoma. Unilateral nasal obstruction, or rhinorrhea, particularly if blood-stained, is suspicious for malignancy.

Any patient presenting with symptoms suggestive of sinonasal neoplasia requires a complete head and neck examination. This includes examination of the nasopharynx, either with a nasopharyngeal mirror or a fiberoptic nasopharyngoscope. Although cervical metastasis is uncommon in sinonasal malignancy, it is often the presenting symptom in nasopharyngeal carcinoma. Therefore, the neck, including the posterior triangle area, should be carefully examined. The oral cavity should be inspected without dentures in place to appreciate any change in palatal architecture or tumor extension.

In extensive disease, there will be gross involvement of the nasal cavity. In less advanced cases, the symptoms are subtle. There may be bulging of the medial wall of the maxillary sinus into the nasal cavity or a purulent exudate from one of the sinus ostia. Any asymmetry of the nasopharyngeal walls deserves biopsy, especially in the presence of cervical adenopathy.

Any patient who fails to respond to conventional treatment for nasal obstruction or discharge deserves a biopsy to rule out malignancy. The biopsy specimen can be obtained transnasally if the tumor is visible or transantrally through a Caldwell-Luc approach. The majority of tumors originate in the maxillary sinus, and 75 to 80 per cent of them are squamous cell or anaplastic carcinomas. Salivary gland tumors account for 6 to 17 per cent of malignancies (Gullane and Conley, 1983).

Radiography

Plain sinus radiographs are usually the first study obtained in the evaluation of paranasal sinus disease. Certain neoplastic processes - for example, osteomas and fibrous dysplasia - produce characteristic images on plain radiographs. Other benign tumors cannot be differentiated from inflammatory or malignant disease, requiring other imaging modalities to aid in the diagnosis preoperatively.

In advanced malignancies, a soft tissue mass and bony destruction will be apparent on plain films. Early malignant tumors in the paranasal sinuses are impossible to differentiate from inflammatory disease unless there is evidence of bone erosion. The same problem exists for primary nasal tumors. Tumors of the nasopharynx can be diagnosed on plain radiographs only if there is erosion of the skull base.

Polytomography is an improvement over conventional radiographic techniques. Fine detail of bone destruction in a narrow plane can be discerned with this modality. However, a large dose of radiation is given to the patient to perform this study, and polytomes are inadequate to demonstrate orbital, intracranial, or pterygomaxillary space invasion by tumor.

The addition of computed tomography (CT) has greatly improved the preoperative diagnosis of sinonasal pathologic conditions because of its ability to demonstrate both soft tissue and bone densities on the same image. The selective uptake of intravenous contrast material by tumor helps to differentiate it from other soft tissue structures. Extension of tumor into the orbit, intracranial cavity, soft tissues of the face, and pterygomaxillary fossa is easily identified.

One of the major drawbacks of CT scanning is its inability to distinguish between tumor mass and mucosal congestion secondary to obstruction by tumor Kondo et al, 1982). This differentiation is critical when extension into the posterior ethmoidal or sphenoidal sinuses is suspected. Unless there is concomitant bony destruction, it may be necessary to biopsy the suspicious areas to rule out involvement by tumor.

The other problem with CT scanning is the relative difficulty in obtaining coronal images. Coronal scans derived from axial sections do not contain the fine detail required to assess skull base erosion. True coronal scans obviate this problem, but many patients cannot tolerate the positioning necessary to obtain the scan. Despite their drawbacks, CT images of malignant tumors of the sinonasal tract almost always correlate with the surgical findings (Virapongose et al, 1986).

The introduction of magnetic resonance imaging (MRI) has helped solve some of these problems. MRI can visualize images in any plane desired (axial, coronal, sagittal, and so on) without repositioning the patient and without any loss in detail. There is no radiation exposure involved in MRI and no loss of detail from dental amalgams.

Compared with CT scanning, MRI shows better soft tissue contrast without resorting to contrast infusion but is not as effective in demonstrating bone detail. Visualization of bony structures is dependent on surrounding soft tissue producing a signal; the bony structures are seen in silhouette. MRI has been especially useful in determining tumor extension in nasopharyngeal tumors. Figure compares CT with MRI in a patient with undifferentiated nasopharyngeal carcinoma. Inflammatory disease in the paranasal sinuses generates an intense signal on MRI (Moore et al, 1986). This feature may be beneficial in differentiating between inflammatory and neoplastic processes.

At this time, there is no published study indicating whether MRI has overall superiority over CT in the diagnosis of tumors in the nose and paranasal sinuses.

Pathologic Features

Malignant Tumors

Staging

The American Joint Committee on Cancer has formulated standard tumor, nodes, and metastases (TNM) staging for malignant tumors of the maxillary sinus and nasopharynx (American Joint Committee on Cancer, 1980). Tumors arising primarily from the nose or the other paranasal sinuses are much less common, accounting for only 12 per cent of paranasal sinus tumors in one series, precluding the accumulation of meaningful statistics (Badib et al, 1969).

Ohngren's line is an imaginary line drawn between the medial canthus of the eye and the angle of the mandible. This line divides the maxillary antrum into the infrastructure (anteroinferior) and the suprastructure (posterosuperior). This line is significant in determining the stage of antral tumors. T1 tumors are confined to the antral mucosa of the infrastructure with no evidence of bony erosion. In T2 lesions, the tumor is confined to the antral mucosa of the suprastructure, with no evidence of bony destruction, or is located in the infrastructure with destruction of the inferior or medial osseous walls. T3 tumors may invade the orbit, anterior ethmoidal sinuses, pterygoid muscle, or the skin of the cheek. T4 tumors are massive lesions with invasion of the cribriform plate, the posterior ethmoidal cells, the sphenoidal sinus, the nasopharynx, the pterygoid plates, or the skull base.

The nodal (N) and metastatic (M) portions of the staging system are identical to those for other head and neck tumors. In an N0 neck, there is no clinically positive adenopathy palpable. An N1 neck contains a single clinically positive homolateral node not greater than 3 cm in diameter. The N2 neck is divided into two subgroups: N2a contains a single clinically positive homolateral node greater than 3 cm but less than 6 cm in diameter, and N2b involves multiple clinically positive nodes, none greater than 6 cm in diameter. The N3 neck is divided into three groups, with N3a containing one or more homolateral nodes greater than 6 cm in diameter, N3b containing bilateral positive nodes, and N3c containing only clinically positive contralateral nodes. Metastatic disease is assessed as M0 for no known distant metastasis and M1 for the presence of distant metastasis.

Malignancy of the nose and paranasal sinuses is primarily a local disease. Ashley and colleagues (1977) demonstrated at autopsy that only 25 per cent of maxillary sinus carcinomas metastasize. However, tumor is restricted to the maxillary sinus in only about 25 per cent of patients, with bony destruction apparent in up to 80 per cent of cases (Harrison, 1971).

Squamous Cell Carcinoma

Epithelial cell carcinoma accounts for more than 70 per cent of malignant neoplasms in the nose and paranasal sinuses. The maxillary sinus is involved in 70 per cent of cases, followed by the nasal cavity (20 per cent) and other sinuses (10 per cent) (Robin et al, 1979). Unfortunately, the disease is limited to the maxillary sinus at the time of diagnosis in only 25 per cent of cases. The numerous natural ostia and thin bony walls of the paranasal sinuses make spread from one compartment to another quite easy.

The symptoms for squamous cell carcinoma of the sinonasal tract are very similar to those for all malignant lesions of the nose and paranasal sinuses. Most patients have a long-standing history of "sinusitis" prior to the diagnosis. It is uncertain whether the chronic inflammation is a contributing factor in the development of carcinoma. The basic rule to follow is if a patient presenting with complaints of sinus disease does not respond to a routine course of medical management, a further workup is necessary to rule out neoplastic disease.

Maintaining a high index of suspicion is the only way that the diagnosis of malignancy in the nose or paranasal sinuses will be made early in the course of the disease. The average delay from onset of symptoms to diagnosis is 8 months (Gallagher and Boles, 1970). In order to confirm the diagnosis, biopsy material is needed. This is usually obtained through a transnasal approach, if there is visible tumor present, or via a Caldwell-Luc approach. In patients who have no visible tumor in the nose and are too debilitated to tolerate a Caldwell-Luc procedure, biopsy material can sometimes be obtained through a transnasal antrostomy.

In the nasal cavity, the lateral walls, especially the turbinates, are frequently involved. Cervical metastases occur in 17 per cent of patients. Carcinomas of the nasal septum are much less common and tend to occur at the mucocutaneous junction. Because it is a midline lesion, cervical spread can occur to either side and is found in approximately 10 per cent of cases. For small lesions, both surgical excision and radiation therapy provide equally good results. For larger lesions, combination therapy is necessary to control the disease. In general, carcinomas of the nasal cavity are diagnosed at earlier stages than are their counterparts in the paranasal sinuses. As a result, 5-year survival statistics for nasal cavity carcinomas are approximately double those for tumors located in the paranasal sinuses.

Primary carcinomas of the frontal or sphenoidal sinuses are rare, accounting for less than 1 per cent of all paranasal sinus tumors. These sinuses are usually involved secondary to direct extension from the other sinuses, primarily the ethmoidal sinuses.

Carcinoma of the maxillary sinus is twice as common in men as in women, and more than 95 per cent of patients are older than 45 years of age (Batsakis, 1979). In addition to the frequent history of chronic sinus disease, 26 per cent of patients have oral symptoms ranging from pain in the maxillary teeth or loosening of the teeth to a change in the contour of the hard palate or gingivo-buccal sulcus. Following the extraction of maxillary teeth, there may be a persistent oroantral fistula. Trismus may occur if the tumor has extended into the pterygoid fossa. A frequent complaint is the inability to wear previously well-fitting dentures.

A unilateral nasal discharge of epistaxis is present in 35 per cent of patients, and nasal obstruction is present in half (Tabb and Barranco, 1971). When large tumors erode the medial wall of the antrum, the tumor may be visualized within the nasal cavity. Ocular symptoms are present in 25 per cent of patients and are the initial complaints in 5 per cent of patients (Larsson and Martenson, 1954). Ocular symptoms are the result of extension of the tumor through the floor of the orbit and may include diplopia, decreased visual acuity, periorbital edema, or proptosis.

Growth of the tumor through the anterior wall of the maxilla will result in symptoms referable to the soft tissue of the face. Asymmetry of the face may occur and is often associated with facial numbness secondary to invasion of the infraorbital nerve. In advanced

cases, the tumor may erode through the skin.

During the course of the disease, cervical node metastases develop in approximately 35 per cent of patients and distant metastatic disease in 10 per cent. The incidence of nodal metastasis without invasion of the oral cavity is half that seen when the oral cavity has become involved with the disease.

Except for the rare early carcinoma of the maxillary sinus, 5-year survival statistics are poor. Gullane and Conley (1983) demonstrated a 1-year survival rate in patients with cervical nodal disease of only 37 per cent, and all these patients were dead of disease within 3 years.

As illustrated in Table 2, most patients with squamous cell carcinoma of the maxillary sinus present with T3 and T4 lesions (150 of 216 patients). These advanced tumors, with extension into contiguous structures, are difficult to cure. However, because of the infrequency of late recurrences, 5-year survival statistics generally represent cure rates. Combined treatment with both surgery and radiation therapy, given either pre- or postoperatively, provides the best chance for survival. Because of the relative rarity of carcinoma of the maxillary sinus and the variations in treatment protocols among different institutions, strict comparisons of various treatment protocols are difficult. Surgical excision attempts to remove all evidence of tumor, preferably with an en bloc resection. The type of procedure employed is dependent on the amount of tumor extension to adjacent structures. For small lesions, a radical maxillectomy with preservation of the orbital contents is performed. For tumors located in the suprastructure, the palate can sometimes be preserved. Unless there is palpable nodal disease, a neck dissection is not routinely performed.

Table 2. Five-Year Survival of Patients with Squamous Cell Carcinoma of the Maxillary Sinus

Author	T1	T2	T3	T4
Bush and Bagshaw	0/0	0/1	5/14	1/12
Gallagher and Boles	1/1	5/12	9/27	0/9
Lee and Ogura	3/5	5/11	5/18	5/13
St.-Pierre and Baker	0/0	6/8	6/21	7/36
Totals	4/6	16/41	25/80	13/70
Percentage	67%	39%	31%	18%.

In larger tumors, all diseased structures must be removed. This usually involves performing an orbital exenteration and resecting the hemipalate and facial skin when indicated. Massive tumors involving the cribriform plate, with or without extension into the cranial cavity, are best approached through a craniofacial resection. These massive resections may offer an improved chance for survival. In the series of Ketchum and colleagues (1973), 54 patients who underwent craniofacial resection were followed for a minimum of 3 years. Of these patients, 26 individuals were free of disease, and 6 individuals died from unrelated causes but were free of disease at the time of death.

Primary ethmoidal carcinomas are relatively rare; ethmoidal sinus involvement is generally the result of spread from the maxillary antrum. Unlike maxillary sinus carcinoma, there is only a slight male preponderance. Nasal obstruction with epistaxis is more common than in antral carcinomas. Extension through the cribriform plate occurs early and is usually accompanied by anosmia. A craniofacial resection is required to allow removal of the tumor with good surgical margins. Survival statistics parallel those of large maxillary sinus carcinomas. Death usually occurs secondary to the inability to control local disease.

Lymphoreticular Tumors

Lymphoma. Extranodal head and neck sites are present in 10 per cent of patients with non-Hodgkin's lymphoma. Of these, 66 per cent occur in Waldeyer's ring (including the nasopharynx and tonsils), and 16 per cent occur in the nasal cavity and paranasal sinuses (Jacobs et al, 1986). The majority of lymphomas in the nose and paranasal sinuses are of the histiocytic and diffuse lymphocytic types (Batsakis and Sciubba, 1985). The symptoms and age and sex distribution are similar to those of squamous cell carcinoma, requiring a biopsy to differentiate between the two. Systemic symptoms are unusual.

The antrum and ethmoidal sinuses are most frequently involved, but there is usually local extension to the soft tissue of the face at the time of diagnosis. Current treatment for early lesions with favorable histologic features consists of radiation therapy to the primary site and the neck. For lesions with a poorer prognosis, radiation is combined with chemotherapy. Five-year survival rates depend on the histologic grade of the lymphoma. Most authors quote a 5-year survival of 50 to 70 per cent after radiation therapy (Batsakis, 1979).

Extramedullary Plasmacytoma. Sixty per cent of all extramedullary plasmacytomas occur in the nose, nasopharynx, and paranasal sinuses. There is a male to female ratio of 4:1, and most patients present between the ages of 40 and 70 years (Batsakis, 1979). These tumors may be either polypoid or sessile, are red, and rarely ulcerate. The more aggressive lesions tend to be soft and friable. After the diagnosis of plasmacytoma has been returned, the patient must undergo a thorough evaluation to determine that this is not a local manifestation of multiple myeloma.

The clinical course with this tumor is variable, and the histologic appearance is not a useful criterion in determining aggressiveness. The disease may remain localized or evolve into systemic multiple myeloma after a latent period. Because of this, there is no one treatment of choice. Most surgeons would favor excising the tumor widely. Radiation can be given as part of a planned combination therapy or may be held in reserve for recurrences.

Esthesioneuroblastoma

Berger and co-workers first described "L'esthesioneuroepitheliome olfactif" in 1924. To date, barely 200 cases have been recorded in the world literature. The relative rarity of this tumors, however, may be the result of misdiagnosis, as it is easily confused with other tumors arising in this area. Esthesioneuroblastoma is a neurogenic tumor of olfactory epithelium. In humans, this epithelium is limited to the upper surface of the superior turbinate, the upper nasal septum, and the cribriform plate.

The tumor can occur at any age, but almost two thirds of these patients are between the ages of 10 and 34 years (Bailey and Barton, 1975). There is a slight male predominance. As with all intranasal tumors, symptoms are nonspecific and include epistaxis, anosmia, and nasal obstruction. Because of this lack of symptoms, most patients are diagnosed late in the course of the disease. Esthesioneuroblastoma usually appears as a red or fleshy mass in the nasal vault. Symptoms of local invasion, such as proptosis, are usually evident at diagnosis.

The diagnosis is made by biopsy. Adequate tissue is needed to prevent confusion with other undifferentiated round cell tumors of the nasal cavity, especially when secondary infection is evident. Although some histologists have subdivided esthesioneuroblastomas by histologic appearance, there is no difference in the clinical course among them. All esthesioneuroblastomas contain intercellular neurofibrillary matrices and vascular septa dividing clusters of small round cells (Batsakis, 1979). A high mitotic index, prominent nucleoli, and cellular pleomorphism mitigate against the diagnosis.

Although slow growing, all esthesioneuroblastomas are locally invasive and carry a 20 per cent rate of metastasis. The cervical lymph nodes and lungs are most frequently involved. If inadequately treated, esthesioneuroblastoma will invariably recur.

No treatment combination has been overwhelmingly successful in the management of esthesioneuroblastomas. These tumors are variably radiosensitive. It appears that combined surgery and radiation therapy provides the highest survival statistics. Baily and Barton (1975) demonstrated a 45 per cent 5-year survival for patients treated with either surgery or radiation. When the two modalities were combined, the 5-year survival rate increased to 67 per cent. Other authors prefer surgical excision primarily and reserve radiation for persistence or recurrence (Skolnik et al, 1966). Because of the slow growth of these tumors, 5-year statistics may not be accurate reflections of the long-term clinical course.

Because of the location of esthesioneuroblastomas, surgical excision is usually accomplished through a craniofacial approach.

Salivary Gland Tumors

Malignant salivary gland tumors account for only 6 to 17 per cent of malignant tumors in the nose and paranasal sinuses (Gullane and Conley, 1983). The relative incidence of these tumors in the sinonasal passages differs greatly from that in the major salivary glands. For example, pleomorphic adenomas, the most common parotid tumor, are unusual in the nose and paranasal sinuses. The most common salivary gland tumors in the nose and paranasal sinuses are, in descending order of frequency, adenoid cystic carcinoma, adenocarcinoma, pleomorphic adenoma, mucoepidermoid carcinoma, and undifferentiated carcinoma. Acinic cell carcinoma, carcinoma ex-pleomorphic adenoma, and oncocytomas are rare (Batsakis, 1979). In general, all malignant salivary gland tumors in the sinonasal tract are aggressive lesions that are usually recognized only after there is massive extension of the disease.

Adenoid Cystic Carcinoma. One third of all salivary gland tumors in the nose and sinuses and half of the salivary gland tumors in the nasopharynx are adenoid cystic carcinomas. They are also known as cylindromas because of their histologic appearance. Adenoid cystic carcinomas of the nose and paranasal sinuses have the poorest prognosis of

all cylindromas, which is due to both their late diagnosis and their tendency toward early perineural invasion.

Most patients present with epistaxis and nasal obstruction. Cheek numbness may occur secondary to invasion of the infraorbital nerve. Tumors located in the nasopharynx are commonly associated with serous otitis media and diplopia (Batsakis, 1979). These tumors may be histologically divided into low grade and high grade. Although perineural invasion, perivascular invasion, and bone invasion are equally common in both grades, vascular invasion is more frequent in high-grade tumors (Batsakis and Sciubba, 1985). Perineural invasion usually occurs along the maxillary and mandibular divisions of the trigeminal nerve.

There is a male to female ratio of approximately 12:7, and the median age at diagnosis is 50 years for low-grade tumors and 56 years for high-grade tumors.

Treatment involves radical excision of the primary tumor followed by radiation therapy. Presence of tumor at the surgical margin is associated with a high recurrence rate, even with the addition of radiation therapy. Chemotherapy is not felt to be of benefit at this time. Like their counterparts in the major salivary glands, cylindromas of the sinonasal tract have a low rate of regional node spread (14 per cent); however, 40 per cent of patients will demonstrate distant metastases (Spiro et al, 1974). The most favored sites of spread are the lungs and bones.

Because of the propensity for late distant metastases, 5-year and even 10-year survival statistics may be inadequate for determining cure rates. Spiro and co-workers (1974) described a cure rate of only 7 per cent for lesions of the nose, paranasal sinuses, and larynx, as compared with 10-year cure rates of 29 per cent for adenoid cystic carcinomas of the parotid gland. Ten per cent of deaths are secondary to distant metastases; most occur as a result of uncontrolled local disease.

Adenocarcinoma. The relationship between wood-workers and leather workers and adenocarcinoma of the nose and paranasal sinuses has already been discussed. There are three basic histologic forms: papillary, sessile, and alveolar-mucoid. Papillary adenocarcinoma is the type commonly associated with woodworking. In general, these tumors can be divided into low-grade and high-grade histologic types.

Low-grade adenocarcinomas of the sinonasal tract tend to occur high in the nasal cavity and the ethmoidal sinuses, as compared with adenoid cystic carcinomas, and tend to extend into the adjacent sphenoidal sinus and orbits. High-grade tumors are more often located in the lower portion of the antrum and are more diffuse and extend into the middle cranial fossa and pterygoid plates. Intraneural and perineural spread can occur with high-grade tumors (Batsakis, 1979).

The male to female ratio is 3:1 for low-grade tumors and 1:1 for high-grade adenocarcinomas. The median age at diagnosis is 62 years. The clinical behavior of both grades of adenocarcinoma is similar to that of adenoid cystic carcinoma. Regional node metastasis is less than 30 per cent, but distant metastases are far less common than in cylindroma (16 per cent) (Spiro et al, 1973). Metastases are usually found in the high-grade type.

Treatment involves radical surgical excision followed by radiation therapy. Because of the high location of these lesions in the nose and sinuses, excision often requires craniofacial resection. Neck dissection is performed if there is concurrent nodal disease. Nodal disease at the time of primary resection is associated with only a 10 per cent salvage rate (Spiro et al, 1973). As with adenoid cystic carcinoma, death is usually secondary to uncontrolled local disease.

Other Salivary Gland Tumors. Mucoepidermoid carcinoma is the third most frequent salivary gland tumor found in the nose and paranasal sinuses. The nasal cavity and maxillary sinus are most frequently involved. Their clinical course is similar to that of adenocarcinoma.

Benign mixed tumors (pleomorphic adenoma) are unusual in the sinonasal cavity and usually arise from the nasal septum. Wide local excision is curative. Malignant mixed tumors (carcinoma ex-pleomorphic adenoma) are very uncommon and arise most frequently in the maxillary sinus. They are extremely aggressive tumors.

Melanoma

The incidence of new cases of malignant melanoma is two per 100,000 population annually. Of these 15 to 20 per cent occur in the head and neck region. Less than 2 per cent of all melanomas originate in the nose or paranasal sinuses. Metastasis to the nose or paranasal sinuses is quite rare (Batsakis and Sciubba, 1985).

These lesions are predominantly found in the nasal cavity. The anterior part of the nasal septum is the most common location, followed by the middle and inferior turbinates. The maxillary antrum is the most frequently involved sinus. Even though the olfactory area has pigmented cells, there have been no cases reported of melanomas arising from this area.

Mucosal melanomas are thought to arise from melanocytes in the mucosa of the nose and sinuses and not from a precursor nevus. Although melanocytes are present in the nasal cavities of both white and black persons, this is primarily a disease of white persons. Mucosal melanomas in general are rare before puberty and tend to occur in patients 15 years older than those with cutaneous melanomas of the head and neck region. There is no sexual predilection.

Symptoms are similar to those in other sinonasal neoplasms; however, 80 per cent of these patients complain of epistaxis. The tumor appears as a fleshy mass that may be multicentric. The tumor may or may not be heavily pigmented. There is no difference in the clinical course between melanotic and amelanotic tumors (Blatchford et al, 1986).

Mucosal melanomas have a poorer prognosis than their cutaneous counterparts. The clinical course is one of aggressive local recurrences, regional node metastasis, and distant metastases. The recommended treatment is surgical and involves wide local excision of the involved structures. If nodal involvement is evident at the time of surgery, a neck dissection is included in the procedure. Radiation therapy is not beneficial, and there is currently no efficacious chemotherapeutic regimen. Nasal melanomas appear to have a slightly better prognosis than do other mucosal melanomas.

Next to anorectal melanomas, upper respiratory tract melanomas have the poorest prognosis. A mean survival from the time of biopsy of 2.3 years has been cited by Gallagher (1970). The 5-year survival rate was 11 per cent, and only 0.5 per cent of affected individuals lived 10 years.

Sarcomas

Rhabdomyosarcoma. The most common soft tissue malignant tumor found in patients less than 15 years of age is rhabdomyosarcoma. Occurrence in the head and neck accounts for more than 40 per cent of cases. Rhabdomyosarcoma of the nasopharynx and paranasal sinuses accounts for 25 to 20 per cent of all extraorbital cases (Batsakis, 1979). Rhabdomyosarcoma is much more common in whites than in blacks, with an annual incidence of 4.4/million in the white population (Batsakis et al, 1980).

In the head and neck, the most prominent type of tumor is the embryonal form, followed distantly by the alveolar form. Because of the age group involved, early nasopharyngeal rhabdomyosarcoma is usually misdiagnosed as adenoidal hypertrophy. The correct diagnosis is usually considered only after regional spread has already occurred. The prognosis for patients with rhabdomyosarcoma of the head and neck is poorer than for those with peripheral tumors.

The prognosis for patients with rhabdomyosarcomas in the parameningeal areas (nasopharynx, paranasal sinuses, and middle ear) is especially poor. Unlike rhabdomyosarcomas in other sites, these tumors demonstrate direct extension into the central nervous system (CNS). Meningeal extension is associated with a median survival of 9 months. Because of the rapid clinical course in these patients, there is frequently no evidence of distant metastases. In the series reported by Newman and Rice (1984), 91 per cent of patients with parameningeal tumor sites developed CNS extension, and 64 per cent presented with evidence of CNS disease.

The Intergroup Rhabdomyosarcoma Study advocates the use of combination therapy (radical surgery, radiation therapy, and chemotherapy) for the treatment of all rhabdomyosarcomas. They have demonstrated an increase in the 2-year survival rate from less than 30 per cent to 70 per cent for all head and neck tumors (Donaldson et al, 1973). In the University of California at Los Angeles (UCLA) series, the only two survivors who had extension of rhabdomyosarcoma to the CNS underwent combination therapy that included intrathecal chemotherapy and radiation to the brain and spinal cord (Newman and Rice, 1984).

Fibrosarcoma. Fibrosarcoma was once felt to be relatively common, but recent improvements in the diagnostic capabilities of pathologists has markedly reduced the apparent indices. It is now thought that it composes only 5.5 per cent of all soft tissue sarcomas. Any sinus can be the primary site, although the antrum is most commonly involved. Extension beyond the primary site usually occurs before the diagnosis has been made (Batsakis et al, 1982).

The lesions generally appear as a polypoid mass that is not encapsulated. Unlike fibrosarcoma elsewhere in the head and neck, the histologic grade does not correlate well with the clinical outcome (Batsakis, 1979). Metastases are unusual and are usually related to

uncontrolled local disease. The appearance of metastases may be delayed up to 10 years from the time of primary treatment (Batsakis et al, 1982). The treatment is surgical, but postoperative radiation therapy may be of some benefit. If the surgical margins are free of tumor, the prognosis is relatively good in comparison with other sarcomas of the nose and paranasal sinuses.

Angiosarcoma. Angiosarcoma of the nose and paranasal sinuses is extremely rare. Less than 25 cases have been cited in the literature. Angiosarcomas are rapidly growing tumors that occur during middle age. The location in the nose and paranasal sinus is associated with a better overall prognosis because of the tumor's size at diagnosis and the higher level of differentiation found in angiosarcomas of the upper airway (Panje et al, 1986).

Treatment is surgical and involves a wide local excision with neck dissection if there is evidence of nodal spread. Radiation is useful for palliation. There are not enough cases to determine precise prognostic statistics, but angiosarcomas elsewhere in the head and neck are associated with a 5-year survival rate of 50 per cent, half with persistent or recurrent disease.

Hemangiopericytoma. Approximately one quarter of all hemangiopericytomas occur in the head and neck. They are unpredictable neoplasms, both histologically and in their clinical course. These tumors are formed by pericytes and represent only 1 per cent of all vasoformative tumors. There is no sexual predilection and there is a slight peak incidence in middle age.

Hemangiopericytomas are rapidly growing, painless lesions. In the nose and sinuses, they are generally removed early secondary to obstructive symptoms. They are usually gray and rubbery in appearance, despite their vascular nature. In the sinonasal area, the nasal cavity is most frequently involved (60 per cent) followed by the sphenoethmoidal complex (30 per cent), the antrum, and the nasopharynx (less then 5 per cent) (Batsakis and Rice, 1981).

The treatment is surgical, with the benefits of radiation therapy unproved. Hemangiopericytoma appears to be a less aggressive tumor when it is located in the head and neck than when it is located in the rest of the body. There is an overall recurrence rate of 57 per cent, with more than half occurring more than 5 years after diagnosis (Batsakis, 1979). Patients with hemangiopericytoma must be followed for the rest of their lives to assess tumor recurrence. Distant metastasis is uncommon.

Chondrosarcoma. Chondrosarcoma of the nose or paranasal sinuses is rare. In the sphenoethmoidal area, the differentiation among chondrosarcoma, chondroma, and chordoma may be very difficult. Because of the high local recurrence rate, there is a poor long-term prognosis, although the clinical course may be protracted. The local recurrence rate for chondrosarcoma in the head and neck is 85 per cent, versus 15 per cent for tumors elsewhere in the body (Harwood et al, 1980).

The disease is treated with radical surgical excision, reserving radiation for palliation (Vener et al, 1984). Chondrosarcoma of the nasal septum is extremely rare. As of 1979, there were only four cases of chondrosarcoma of the nasal septum in the literature (Batsakis, 1979).

Metastatic Disease

There are fewer than 100 reported cases of metastatic carcinoma to the nose and paranasal sinuses from sites caudal to the clavicle. The most commonly encountered primary tumor is renal cell carcinoma, followed by lung and breast carcinomas. As with primary tumors of the nose and paranasal sinuses, the symptoms are nonspecific (Batsakis and Sciubba, 1985). Renal cell carcinoma metastatic to the nose often presents as epistaxis. Evidence of metastatic disease may antedate the discovery of the primary tumor, especially with renal cell carcinoma.

Unless the metastasis to the nose or paranasal sinuses is the only evidence of metastatic disease, treatment is in the form of radiation and chemotherapy.

Kaposi's Sarcoma

Although Kaposi's sarcoma is a malignant tumor, because of its relationship with the acquired immune deficiency syndrome (AIDS), it will be discussed separately. Until recently, Kaposi's sarcoma was an unusual neoplastic disease of the vascular system, generally found in sub-Saharan Africa and the Mediterranean (Batsakis and Rice, 1981). The disease, generally found in men in their fifth to seventh decades of life, presented with cutaneous symptoms. Visceral and mucosal lesions occur as part of the generalized disease, and lymphadenopathy may antedate the cutaneous lesions. Lesions in the nasal cavity are half as common as in the oropharynx and usually coexist with cutaneous lesions on the extremities.

AIDS has been defined by the Centers for Disease Control (CDC) as "the appearance of Kaposi's sarcoma in individuals younger than 60, and/or life-threatening opportunistic infections in individuals with no known predisposing cause for those infections" (Update AIDS - US, 1982). At this writing, AIDS is predominantly found in the homosexual and bisexual communities, in intravenous drug abusers, and in patients requiring frequent transfusions of blood products (ie, hemophiliacs). The HTLV III virus has been implicated as the causative organism and is transmitted through contaminated body fluids, such as semen and blood.

Because of the protean manifestations of AIDS, head and neck complaints are common. In a series of 72 patients with AIDS, 29 individuals had oral and facial lesions consistent with Kaposi's sarcoma. Epistaxis occurred in one patient, and nasal erythema was seen in two patients (Rosenberg et al, 1984). The diagnosis of mucosal Kaposi's sarcoma is usually made clinically; however, if there are no other symptoms suggestive of AIDS, a biopsy may be necessary.

Currently there is no curative treatment for AIDS. Therapy is directed toward control of opportunistic infections. Chemotherapy is beneficial only for non-AIDS-related Kaposi's sarcoma.

Benign Tumors

Papilloma

Papillomas are commonly found in the nose and paranasal sinuses and may be multiple. Most papillomas are nonkeratinizing squamous cell lesions, and although they are histologically similar to the lesions found in recurrent respiratory papillomatosis, there is no pathogenic relationship. Simple excision is curative, but new growths may occur.

Inverting Papilloma

There is much controversy in the literature surrounding the inverting papilloma, much of which stems from the numerous synonyms for this neoplastic disease, including schneiderian papilloma and soft papilloma. Inverting papilloma is a true epithelial neoplasm characterized by hyperplastic endothelium inverting into the underlying stroma. A single layer of respiratory columnar epithelium lining the surface is usually present (Batsakis, 1979).

Inverting papilloma most often arises from the lateral nasal wall in the area of the middle meatus. It only rarely arises from the nasal septum or the paranasal sinuses. At diagnosis, it is often found completely filling the nasal cavity and may involve adjacent structures. The maxillary antrum and the ethmoidal sinuses are most frequently involved, but the orbit and sphenoidal and frontal sinuses may also contain tumor. When malignancy is not present, there is no true invasion of surrounding structures but rather displacement and destruction by pressure necrosis.

Although present in all age groups, inverting papilloma has a peak incidence in the fifth and sixth decades of life. There is a male to female ratio of 3:1. The most common symptom is unilateral nasal obstruction, often in association with rhinorrhea (Lawson et al, 1983). Epistaxis is common, but there is little pain unless secondary infection or malignancy is present. There is no known relationship to allergy.

Although there is no pathognomonic radiologic finding, a large nasal mass with destruction of the lateral nasal wall is suggestive of inverting papilloma. Bony sclerosis secondary to synchronous chronic sinusitis may be present. The diagnosis is confirmed by biopsy.

The treatment of inverting papilloma is surgical. Complete excision is necessary to prevent recurrence. The high rates of recurrence quoted in the literature represent incomplete excision of the tumor. Extension of the tumor into the nasofrontal duct, supraorbital ethmoid cells, lacrimal fossa, or infraorbital recess of the maxillary sinus are associated with a high recurrence rate (Vrabec, 1975). Most recurrences occur within 2 years. The type of resection performed is dependent on the size and extent of the tumor. In most instances, a medial maxillectomy is required to remove all the tumor. For very small lesions, a more conservative excision may be attempted. Unless there is an associated malignancy, mutilating surgery should be avoided. Radiotherapy is ineffective in controlling these tumors and may promote malignant transformation.

There is a known association between inverting papilloma and malignancy. Unfortunately, because of referral patterns and the histologic criteria employed, the quoted incidence ranges from 2 to more than 50 per cent (Lawson et al, 1983). Hyams, in his large series from the Armed Forces Institute of Pathology (1971), found a rate of 13 per cent. It is not known whether this association represents malignant degeneration of an inverting papilloma or the simultaneous presentation of carcinoma and inverting papilloma.

Meningioma

Extracranial meningiomas, excluding orbital meningiomas, are very rare. Although there are different histologic types, all meningiomas arise from meningothelial arachnoid cells. Extracranial meningiomas are histologically identical to their intracranial counterparts, and most are the result of extracranial arachnoid cells clusters (Granich et al, 1983). These clusters tend to occur along the tracts of the cranial nerves and in association with the skull foramina.

The treatment for these tumors is surgical, usually involving a medial maxillectomy. Complete removal is curative. There is no role for radiation therapy in the primary treatment of nasal meningiomas.

Neuroma (Schwannoma)

Approximately 25 per cent of schwannomas occur in the head and neck; few occur in the nose and paranasal sinuses. Solitary neurofibromas without associated von Recklinghausen's disease are rare (Batsakis and Sciubba, 1986). Symptoms depend on the location of the tumor within the nose and paranasal sinuses. The treatment is complete surgical excision.

Hemangioma

Hemangiomas may occur either intraosseously in the nasal bones or, more frequently, in the mucosa of the nose and paranasal sinuses. The male to female ratio is 1:4, and most patients are middle-aged. Pain is present in most cases, and a history of local trauma is often elicited. On x-ray film, a translucent area with spicules of bone radiating out from a central core is seen in intraosseous nasal hemangiomas. Complete excision is curative.

Hemangioma of the mucosa may be difficult to differentiate from inflammatory polyps (Batsakis, 1979). These tumors are most commonly seen in the anterior nasal septal area and are often associated with Osler-Weber-Rendu disease or von Hippel-Lindau disease. Polypoid lesions may be treated with local excision, including the septal perichondrium. The diffuse hemangiomas often seen in Osler-Weber-Rendu disease are difficult to control. These patients often present with a history of frequent copious epistaxis. In these patients, a septodermoplasty, excision of the involved nasal septal mucosa, and reconstruction by placing a dermal graft over the bare septal cartilage, can be attempted. After this procedure, increased nasal crusting occurs because of the lack of mucus-secreting cells on the septum. It is often impossible to remove all the involved mucosa; as a result, these patients often have recurrence of their hemangioma at the excision margin.

Chordoma

Chordomas probably arise in detached remnants of the notochord, and one third occur at the skull base. They account for 0.2 per cent of nasopharyngeal tumors (Mills, 1984). Diplopia is the most common presenting symptom, followed by fronto-occipital headache, visual field defects, and nasal obstruction. The peak incidence is in the third and fourth decades of life, and there is a slight male predominance.

A mass in the nasopharynx covered with normal mucosa may be visualized. Extensive bone destruction and the presence of a soft tissue mass in the nasopharynx may be evidence on x-ray film. The diagnosis is confirmed by biopsy.

Because of its location, it is almost impossible to completely excise a skull base chordoma. Surgical excision is used to debulk the tumor, and some groups advocate the use of postoperative radiation therapy. There are several surgical approaches to the clivus for attempted resection of chordomas. The transcervical-transmandibular approach to the skull base provides the best exposure of the nasopharynx with proximal control of vascular structures (Krespi et al, 1986). Postoperative morbidity is secondary to the tumor resection required and not to the surgical approach and usually includes deficits of several cranial nerves.

Because of the inability to totally excise these tumors, the prognosis for patients with chordomas is poor, with a mean survival of 7 years (Higinbotham et al, 1967). Death is secondary to local extension of the disease.

Juvenile Nasopharyngeal Angiofibroma

Juvenile nasopharyngeal angiofibroma (JNA) is an uncommon benign, but locally destructive, tumor. It accounts for 0.05 per cent of head and neck tumors and is found almost exclusively in adolescent boys (Batsakis, 1979). JNA can grow to a large size before overt symptoms are present, which include unilateral nasal obstruction, epistaxis, rhinorrhea, and facial swelling. The mean duration of symptoms is 6 months (Bremer et al, 1986).

Angiofibromas originate at the posterolateral wall of the roof of the nose at the point at which the sphenoidal process of the palatine bone joins the vomer and the pterygoid process of the sphenoid bone (Neel, 1985). Grossly, tumor is usually seen to fill one or both sides of the nasopharynx and extends into the nasal cavity. Ulceration of the overlying mucosa is uncommon unless the tumor has been previously biopsied. The tumor is pink to red, lobulated, and rubbery. The main blood supply in JNA is the ipsilateral internal maxillary artery. However, in recurrent tumors and in tumors for which embolization has been attempted, bilateral arterial supplies are common.

As the tumor enlarges, it pushes the nasal septum to the contralateral side and causes anterior bowing of the posterior wall of the maxillary antrum. Further extension leads to involvement of the pterygomaxillary fossa, infratemporal fossa, and inferior orbital fissure. This causes cheek fullness and orbital symptoms. Intracranial extension is seen in less than 10 per cent of patients and can occur either through the sella medial to the carotid artery and lateral to the pituitary gland or through the middle fossa anterior to the foramen lacerum and

lateral to the cavernous sinus and carotid artery (Neel, 1985). Intracranial extension through the lateral route is more common and is amenable to resection; involvement of the cavernous sinus is difficult to manage surgically.

The diagnosis can usually be arrived at preoperatively based on the physical and roentgenographic findings. Opacification of one or more sinuses is generally seen on x-ray studies, most commonly of the maxillary sinus. The presence of anterior bowing of the posterior wall of the maxillary sinus is considered pathognomonic of JNA. Other signs include erosion of the greater wing of the sphenoid bone with characteristic widening along the lower lateral margin of the superior orbital fissure, erosion of the medial wall of the antrum or hard palate, and displacement of the nasal septum (Batsakis, 1979). Because the blood supply in unoperated tumors is constant, some groups no longer advocate the use of preoperative angiography.

The treatment of angiofibromas has changed over the years; currently surgery is the preferred treatment. There is no role at present for hormonal therapy in JNA. Except for very small tumors, the wide exposure offered by the lateral rhinotomy approach is required. Bone is removed as necessary to completely expose the tumor prior to manipulation of the tumor per se. In patients with intracranial extension, the surgical procedure must be customized to the amount of disease present. Tumor in the cavernous sinus is generally considered unresectable because of uncontrollable hemorrhage.

In the series of Bremer and associates (1986), there was an overall recurrence rate of 17 per cent. Only 5 per cent of patients with extracranial disease had recurrent disease, but 50 per cent of patients with intracranial extension demonstrated recurrent disease. Residual intracranial disease can be treated with radiation.

Osseous Tumors

Osteomas. Osteomas are slow-growing, benign tumors composed of mature bone and are found almost exclusively in the head and neck. The mandible is the most frequent location for osteomas. In order of frequency, the frontal, ethmoidal, and maxillary sinuses can be involved. Sphenoidal sinus osteomas are very rare (Batsakis, 1979). Symptoms are referable to the site of the tumor and are usually present for more than a year before the patient seeks help.

These tumors are occasionally diagnosed incidentally during the workup for an unrelated complaint. The treatment for osteomas is surgical but is not warranted unless the patient is symptomatic. At surgery, the tumor is attached to underlying bone by a pedicle. Often a rim of normal bone must be removed to allow extraction of the tumor.

Osteomas in the frontal sinus are best removed through an osteoplastic flap approach. External ethmoidectomy is performed to provide access to osteomas in the ethmoidal sinuses. For maxillary sinus tumors, a Caldwell-Luc approach affords adequate exposure for removal of the osteoma. Complete excision is curative.

Ameloblastomas. These constitute only 1 per cent of all tumors and cysts in the jaws. Eighty per cent of ameloblastomas occur in the mandible and 20 per cent occur in the maxilla

(Batsakis, 1979). Of the maxillary tumors, half are found in the molar area, a third are found adjacent to the antrum, and the remainder occur at other sites (Goodsell et al, 1977). The tumor commonly extends into the pterygomaxillary fossa, ethmoidal sinuses, and orbit. This extension is facilitated by the lack of thick, dense bone in the maxilla, as compared with the mandible. Patients present with complaints of an expanding mass in the cheek and local discomfort.

Histologically, the tumor tends to mimic the enamel organ. There are generally two microscopic patterns: (1) follicular, with epithelial islands in a fibrous stroma and (2) plexiform, with fibrous stromal islands intermixed with strands of epithelium. Although both patterns may be present in the same tumor, one predominates.

Because of certain recurrence with simple curettage, ameloblastoma of the maxilla must be excised en bloc with good surgical margins. This often involves performing a radical maxillectomy with preservation of the orbital contents.

Fibrous Dysplasia. This is one of a group of fibro-osseous lesions characterized by the replacement of normal bone by tissue containing collagen, fibroblasts, and varying amounts of osteoid tissue (Batsakis, 1979). Fibrous dysplasia can be either monostotic or polyostotic. The maxilla is the most frequently involved bone in monostotic fibrous dysplasia. Painless enlargement of the maxilla is the presenting symptom, and the disease usually begins in the first two decades of life. Involvement of several facial bones and even of the calvarium can occur and makes management more difficult.

The diagnosis of fibrous dysplasia can be made radiographically. There is a characteristic mottled or ground glass appearance to the affected bone and there is no discrete border around the lesion.

Fibrous dysplasia is slow growing and tends to stabilize during adulthood. For this reason, excision is not indicated unless there are functional or cosmetic problems resulting from the lesion. At surgery, it is usually impossible to remove all of the dysplastic bone. Instead, an attempt is made to correct functional and cosmetic complaints without performing mutilating surgery.

Radiation therapy in fibrous dysplasia is without merit and has been associated with an increased risk of malignant transformation to osteogenic sarcomas (Pecaro, 1986).

Cherubism is an autosomal dominant disease with 100 per cent penetrance in males and 50 to 70 per cent penetrance in females (Batsakis, 1979). Although the patient is normal at birth, fullness in the cheeks develops during the second or third year of life. The maxilla is involved in two thirds of the patients and the result is an arched palate, premature loss of the deciduous teeth, and failure of permanent teeth to erupt. The lesion progresses for the next 5 years, and cessation of growth occurs after age 10 years. The lesion may regress after puberty.

There is seldom any pain or functional disability associated with the disease. Surgery is restricted to improving cosmetic appearance when necessary. There is no role for radiation therapy in cherubism.

Treatment

Chemotherapy and Radiation Therapy

Most authors agree that surgery provides the only chance for cure in patients with malignant tumors of the nose and paranasal sinuses. Benign tumors are handled by surgical excision when indicated. However, for those patients who refuse surgery, are too ill to tolerate a surgical procedure, or whose tumors are unresectable, chemotherapy or radiation therapy, or both, can be attempted to control the disease process. There are few good studies comparing the effectiveness of nonsurgical protocols, making treatment difficult at best.

Intra-arterial infusion of chemotherapeutic agents in combination with radiation therapy has been attempted. Shibuya and colleagues (1982) employed simultaneous intra-arterial 5-fluorouracil (5-FU) and radiation therapy in 51 patients with T2 and T3 tumors of the maxillary sinus. Of the 34 patients who completed the protocol, 14 of them demonstrated recurrences between 4 and 42 months. The recurrences were usually noted in areas supplied by the ethmoidal and transverse facial arteries, the ethmoidal sinuses, and the lateral portion of the maxillary sinus. These areas were postulated to have received a lower concentration of cytotoxic agents during infusion.

Bush and Bagshaw (1982) analyzed the effectiveness of radiation therapy versus combined radiation and surgical therapy. Radiation therapy was used in 19 patients (9 of whom had T4 lesions), with a 5-year survival rate of only 15 per cent. Combined therapy was used in 13 patients (only 3 of whom had T4 lesions). Survival times were two to three times higher, depending on the surgical procedure involved.

Orbital complications from radiation therapy occur frequently in patients treated for paranasal sinus and nasopharyngeal malignancy. The exact dose required to produce ocular damage is unknown, but doses less than 5000 rad are felt to be relatively safe. Complications have occurred after as little as 1150 rad (Nakissa et al, 1983). The most common complications are lid edema and erythema and conjunctivitis. Visual loss can be acute, with progressive ocular wasting, or delayed, resulting from maculoretinal degeneration. When there has been no demonstration of orbital involvement by tumor, it is tempting to shield the ipsilateral eye from radiation. However, this may lead to inadequate irradiation and an increased risk of recurrence.

Surgery

The following section is a brief discussion of the surgical procedures referred to earlier in this chapter. It is not intended to be a comprehensive presentation, and the reader is referred to a surgical atlas for a more detailed discussion.

Medial Maxillectomy

The medial maxillectomy was originally described by Sessions and Larson in 1977. In 1983, Sessions and Humphreys modified the procedure, eliminating the lip-splitting incision for certain cases and improving the management of the medial palpebral ligament. This procedure provides access to the entire nasal cavity and the maxillary, ethmoidal, and

sphenoidal sinuses, permitting en bloc removal of small tumors of the ethmoidal labyrinth and antrum. Inverting papillomas are usually removed using this approach.

Technique. The medial maxillectomy is begun with a lateral rhinotomy incision extending from the medial aspect of the eyebrow, down the lateral aspect of the nose halfway between the nasal dorsum and the medial canthus, around the ala, and ending at the nostril. The incision may be carried to the lip for greater exposure. The periosteum along the frontal process of the maxilla is incised and elevated along the face of the maxilla and the medial orbital wall. The frontoethmoidal suture line is identified, and the anterior ethmoidal artery is ligated. This is the superior margin of the resection, representing the level of the cribriform plate. The posterior limit of resection is at the level of the posterior ethmoidal artery. The lacrimal sac is divided to permit exposure of the inferior orbital wall.

A lateral osteotomy is performed; the bone is outfractured with the soft tissues of the nose and retracted with a suture through the ala. Dissection is continued on the face of the maxilla, taking care not to injure the infraorbital nerve. At this point, resectability is determined by opening the maxillary antrum and inspecting for extension of disease to the posterior, inferior, or lateral walls. Tumor in these areas requires a complete maxillectomy for resection. Bone cuts are now made across the pyriform rim at the level of the floor of the antrum and from the opening in the maxillary sinus to the orbital rim, just medial to the infraorbital nerve. The superior bone cut is made at the frontoethmoidal suture line and is connected with the osteotomy at the inferior orbital rim.

The intranasal cuts are made next. The first cut is made with an osteotome beneath the inferior turbinate. Soft tissues below the inferior turbinate are cut with a heavy scissors. Using a right-angle scissors, the soft tissue cut is extended superiorly in front of the pterygoid plates. At this point, the specimen is grasped and rocked free, dividing any remaining soft tissue attachments. After hemostasis is achieved, the nasal bone is repositioned, a suture is placed through the medial canthal ligament, securing it at the proper level, and the soft tissues of the face are reapproximated. The cavity is packed with antibiotic-impregnated petrolatum gauze. The resulting cavity is small and is relined with mucus without difficulty.

The specimen contains the lateral wall of the nose, the medial and superior walls of the maxilla, and most of the ethmoidal complex. The lacrimal sac is allowed to drain freely into the new nasal cavity. The most troublesome postoperative problem is nasal crusting, and the cosmetic and functional results are excellent.

Degloving Approach

The degloving approach was described by Sachs and associates in 1984. It provides access to both nasal cavities and maxillary sinuses without facial incisions and with minimal postoperative morbidity. It is especially useful in the complete eradication of inverting papilloma when used in association with a medial maxillectomy.

Technique. Bilateral gingivobuccal incisions are performed from the midline to the maxillary tuberosity, and the soft tissues of the face are elevated off the face of the maxilla bilaterally to the level of infraorbital nerve. Bilateral intercartilaginous incisions are made between the upper and lower lateral cartilages of the nose, and the soft tissues of the nose are

elevated off the nasal dorsum to the root of the nose. A complete transfixion incision is made, and the remaining soft tissue attachments between the nose and the midface are sharply dissected. The entire midface can now be retracted, exposing both nasal cavities and both maxillary sinuses.

After the exposure has been secured, the required excision can be performed. The facial structures are then returned to their original location, and the transfixion incision is reapproximated with absorbable sutures. The gingivobuccal incision is closed, and the nasal cavities are packed for hemostasis.

Total Maxillectomy

Total maxillectomy for the treatment of paranasal sinus malignancies was described in the early part of the 19th century. The technique of total maxillectomy has not changed significantly since that time, but the operative mortality then was 300 per cent and there was little hope of rehabilitation (Baredes et al, 1985). It is still a considerable surgical undertaking with the possibility of a significant blood loss. The term *maxillectomy* actually refers to a group of operations involving resection of the maxilla and possibly the orbit, palate, or soft tissues of the face, depending on the extent of tumor invasion.

Technique. If possible, preoperative impressions of the palate should be obtained to allow fabrication of a palatal prosthesis to be used intraoperatively. The skin incisions for total maxillectomy depend on whether an orbital exenteration is planned. The classic Weber-Fergusson incision begins midway between the dorsum of the nose and the medial canthus and extends inferiorly to the ala. The lip is split, and the incision is connected to the ala. If orbital exenteration is planned, the superior end of the incision is extended laterally along the margins of both the upper and lower eyelids and is connected past the lateral canthus. The lower lid incision is avoided if preservation of the orbital contents is planned to prevent prolonged postoperative edema. However, if additional exposure is required, this incision is completed. An incision is then made in the gingivobuccal sulcus to the maxillary tuberosity.

The skin flaps is then elevated off the face of the maxilla, leaving the periosteum attached to the maxilla. If there is involvement of the facial soft tissues by tumor, adequate margins must be left. If preservation of the orbital contents is planned, the orbit is explored to confirm the lack of tumor. This is accomplished by incising the periosteum at the level of the infraorbital rim and dissecting the periosteum off the orbital floor. Once the final decision to preserve the orbital contents has been made, the bone cuts can be performed. If orbital exenteration is planned, based on clinical and radiographic findings, the orbital contents are not manipulated.

The following description is for a maxillectomy with preservation of the orbital contents. Orbital exenteration will be discussed in the next section. The first bone cut is made with an osteotome, separating the nasal bone from the frontal process of the maxilla. The second bone cut is made at the level of the frontoethmoidal suture line and is extended down to the inferior orbital fissure posteriorly. The zygomatic arch is sectioned with a Gigli saw, as is the frontal process of the malar bone.

An incision is made intraorally, separating the hard and soft palates. The soft tissues of the hard palate are incised, and an ipsilateral central incisor is removed. The palate is transected with the Gigli saw or osteotome. Depending on the extent of invasion, the pterygoid plates may be removed by placing a curved osteotome behind them. At this point, the specimen is held in place only by soft tissue attachments, which are sharply dissected. When the specimen is removed, brisk bleeding occurs from torn branches of the internal maxillary artery.

After hemostasis is achieved, the orbital floor can be reconstructed with fascia or Marlex mesh. The cheek flap is covered with a split-thickness skin graft, and the flap is sutured back in place. If a palatal prosthesis has been previously fabricated, it is secured into position and the sinus cavity is packed with antibiotic-impregnated petrolatum gauze. The cosmetic and functional results are surprisingly good considering the size of the resection.

When exenteration of the orbital contents is planned, the previously described osteotomies are performed. The orbital contents are then dissected from the superior orbital rim back to the orbital apex. Downward traction on the mobile maxilla provides sufficient room to transect the optic nerve in the orbital apex. The specimen is removed and hemostasis is achieved as previously mentioned. As in maxillectomy with preservation of the orbital contents, the cavity is lined with a split-thickness skin graft, and the palatal prosthesis is inserted. The cheek flap is reapproximated and the cavity is packed.

Because of the loss of the orbit, and often of facial skin, the cosmetic appearance following this procedure is often not acceptable to the patient. Prostheses incorporating facial skin, eyelids, and globe improve the appearance significantly. Cheek fullness can be provided by an attachment to the palatal prosthesis that projects into the cavity. One of the advantages of this procedure is that recurrences are appreciated early by directly inspecting the cavity through the orbital defect.

Craniofacial Resection

Partial and total maxillectomies are sufficient when the tumor does not extend to the cribriform plate and the anterior cranial fossa. The craniofacial resection was designed to provide en bloc removal of tumors involving these structures while protecting the frontal lobes from injury. The first craniofacial resection for carcinoma of the paranasal sinuses was described by Smith and colleagues in 1954.

In 1963, the first large series of patients with paranasal sinus neoplasms approached via the craniofacial route was published (Ketcham et al, 1963). Ketcham and coworkers operated on 19 patients, 13 of them having failed more conservative therapy. There was one operative mortality. Six of 11 carcinoma patients and 5 of 7 sarcoma patients were free of disease for a median of 39 months. Six of the seven patients who failed therapy had surgical margins that contained tumor cells. Although in his later reports Ketcham describes using an enlarged midline bur hole through the frontal bone (Ketcham et al, 1973), we prefer the added exposure provided by a formal bifrontal craniotomy (Krespi et al, 1987).

Over the last 20 years, there have been only minor technical modifications to this procedure. Most of these have been directed toward improved decompression of the dura to

facilitate retraction of the frontal lobes. A lumbar subarachnoid drain is placed preoperatively and cerebrospinal fluid (CSF) is withdrawn during the operation. Mannitol may be infused in a dose of 1.0 to 1.5 gm/kg body weight to dehydrate the brain. The patient is hyperventilated to produce a respiratory alkalosis, resulting in cerebral vasoconstriction and a decrease in intracranial pressure. Renal compensation to the alkalosis does not occur for several days (Rafferty, 1984).

Technique. After general endotracheal anesthesia is induced, the lumbar subarachnoid drain is placed. The scalp is shaved, and the face and scalp are isolated from each with adhesive drapes. Temporary tarsorrhaphies are placed bilaterally, and the thigh is prepped for fascia lata or skin grafts. The neurosurgical team begins the operation. A bicoronal incision is made behind the hairline, and the scalp is mobilised subperiosteally to the supraorbital rims. The pericranium is left attached to the scalp flap to allow its use as a pedicled flap for repairing the defect in the anterior cranial fossa.

A bifrontal craniotomy is performed, with the inferior limit of the bone flap placed just above the supraorbital rims. No attempt is made to prevent entry into the frontal sinuses. If this occurs, the mucosa of the frontal sinus should be removed and the nasofrontal ducts packed with muscle or fat. After CSF is withdrawn through the lumbar catheter, the frontal lobes are elevated from the floor of the anterior cranial fossa back to the level of the planum sphenoidale. The dura is sharply dissected from the crista galli. The olfactory nerves are identified and transected as they emerge from the cribriform plate. If there is any tumor extending through the cribriform plate and involving the overlying dura, the dura is left attached to the specimen. This defect can later be grafted with fascia lata.

At this point, resectability is determined. If there is gross invasion of the frontal lobes or extension of tumor posteriorly to involve both optic nerves, the tumor is considered unresectable. All dural tears are meticulously closed to prevent a CSF leak, and if a fascia lata graft is necessary to repair a dural defect, it is sutured at this point. If the lesion is deemed unresectable, the bone flap is replaced and the procedure is terminated. If the tumor is resectable, the frontal flap is temporarily replaced, and the head and neck team begins the transfacial approach.

Facial exposure is provided through a Weber-Fergusson incision. The lip is split if necessary for additional exposure. The transfacial resection is determined by the extent of tumor involvement. For tumors arising from or extending to the maxillary sinus, a radical maxillectomy with orbital exenteration is usually necessary. For ethmoidal sinus tumors without orbital or maxillary sinus involvement, a medial maxillectomy is generally sufficient.

Exposure of the nasal cavity is obtained by a lateral osteotomy. The septum is divided anterosuperiorly, and the nose is mobilized to the contralateral side, exposing both nasal cavities. The septum is removed if involved with tumor, or if required for exposure, but every attempt is made to leave sufficient dorsal support to prevent postoperative deformity. The osteotomies required for the maxillectomy are performed with an osteotome and Gigli saw. If there is no involvement of the antrum, the palate is preserved.

At this point, the neurosurgeons return to the operative field and, using either drills or osteotomes, free the cribriform plate and any involved orbital floor from above. Anteriorly,

the cut should extend through the posterior wall of the frontal sinus. Posteriorly, the anterior wall of the sphenoidal sinus is resected. The lateral margins are determined by the tumor location. For lesions without orbital involvement, the lateral osteotomy on the side of the tumor includes the medial orbital wall. On the contralateral side, the osteotomy extends through the ethmoidal air cells. When there is orbital involvement, a large area of the orbital roof is included in the specimen.

After all the bony cuts have been made, the soft tissue attachments are sharply dissected, and the specimen is removed transfacially. Hemostasis is achieved, and the dura is carefully re-examined, searching for additional dural lacerations, which are most easily repaired through the transfacial route.

The pericranium is dissected off the frontal scalp flap and is rotated into the anterior fossa to serve as support for the frontal lobes. This flap will also decrease the risks associated with using fascia lata grafts for dural defects (Johns et al, 1981). A split-thickness skin or dermal graft is harvested and used to line the entire facial defect. Antibiotic-impregnated petrolatum gauze is used as packing to support the graft.

If a total maxillectomy has been performed, the palate prosthesis is inserted to facilitate early oral nutrition and maintain facial contour during healing. The frontal bone flap is replaced, and the skin of the scalp and face is closed. As with total maxillectomy, unless orbital exenteration has been performed, there is minimal change in the patient's appearance postoperatively.

In patients who have not previously undergone radiation, administration of 6000 rads of postoperative radiation is planned. Frequently, patients requiring craniofacial resection have failed more conservative approaches, usually including radiation therapy.

Complications. Following craniofacial resection, complications are relatively frequent, although usually minor. The largest series of patients has been reported by Ketcham and colleagues (1973). His group operated on 54 patients, 35 of whom experienced complications. There was no relationship noted between previous treatments and the subsequent development of complications. The most common complications were serous otitis (19 instances), orbitofacial edema (22 cases), and CSF leaks, 15 instances of which were considered minor. Most of the complications described, including all major CSF leaks, occurred in the first 31 patients. There were two deaths reported, both secondary to meningitis.

Schramm's series of 12 patients had a complication rate of 33 per cent (Schramm et al, 1979). Two patients had postoperative diplopia, and one patient had low back pain secondary to the intraoperative removal of CSF. The final complication was a CSF leak and subdural abscess with osteomyelitis of the frontal bone. This patient eventually recovered.

Of Sisson and co-workers' eight patients, complications developed in half (1976). Osteomyelitis of the frontal bone developed in two patients who had received preoperative radiation therapy, and was treated surgically. Two other patients bled from the internal maxillary artery after the packing had been removed. There were no cases of CSF leakage and no deaths.

A complete "take" of the skin graft lining the facial defect occurs rarely. However, this is not a problem, and epithelialization proceeds rapidly as long as the cavity is kept clean with saline irrigations. Some disorientation can occur during the first few postoperative days secondary to retraction on the frontal lobes. This resolves spontaneously.

Results. Long-term survival depends on the histologic type of the tumor in addition to the size of the lesion. Terz and associates (1980) reported a 3-year survival rate of 72 per cent for 22 patients with squamous cell carcinoma and 0 per cent for 2 patients with salivary gland carcinoma treated with craniofacial resection. Recurrent tumor developed locally in seven patients with squamous cell carcinoma, and cervical metastases developed in three patients.

In Schramm's series, 9 of 12 patients had been followed for 1 to 4 years without evidence of recurrence (Schramm et al, 1979). Four of these patients had squamous cell carcinoma and three had salivary gland tumors. One patient with melanoma died after 4 years, and the other two patients had been studied for less than a year. Two patients with esthesioneuroblastoma were free of disease 1 year postoperatively.

The series by Ketcham and co-workers (1973) is the most comprehensive. Their 54 patients were followed for a minimum of 3 years. Fifty per cent were free of disease. Six patients were free of disease and died of unrelated causes 19 months to 11 years after surgery. The median survival time for all patients was 8 years. Fifty-five per cent of 36 patients with carcinoma and 42 per cent of 14 patients with sarcoma were free of disease. There was no survivors among three patients with esthesioneuroblastoma.

Transcervical-Transmandibular Approach

The transcervical-transmandibular approach to the skull base, as described in 1984 by Krespi and Sisson, offers wide exposure of the middle cranial fossa with the advantage of proximal vascular control. It is useful for approaching tumors of the clivus and nasopharynx.

Technique. After a tracheostomy has been performed, a transverse neck incision is made from the mastoid tip to the midline and is carried up to the lip. The digastric muscle is released from the hyoid bone, and the sternocleidomastoid muscle is retracted laterally to allow exposure of the carotid vessels. The external carotid artery is ligated distal to the lingual artery. Cranial nerves X, XI, and XII are identified and preserved. The mandible is split in the midline, and the floor of the mouth is incised to the area of the retromolar trigone. The lingual nerve is identified and preserved if possible.

The incision is extended onto the soft palate, following the medial border of the upper dentition. The soft tissues of the palate are elevated to expose the nasopharynx and the bony palate. Exposure can be improved by removing the hard palate with Kerrison rongeurs. At this point, the entire nasopharynx is exposed, and resection of nasopharyngeal tumors can be accomplished.

Further dissection is required to remove tumors arising from the clivus, such as chordomas. With blunt dissection, a space is created between the superior constrictor muscles and the prevertebral fascia superior to the hypoglossal nerve. The styloglossus and

stylopharyngeus muscles and the glossopharyngeal nerve are divided. The oropharynx can now be retracted to the contralateral side. The eustachian tube is transected under direct vision by placing scissors medial to the internal carotid artery. This detaches the mucosa of the nasopharynx from the skull base. After the nasopharynx has been retracted to the contralateral side, the prevertebral fascia is incised. Retraction of the prevertebral musculature exposes the clivus and upper cervical spine. Removal of chordomas can be accomplished under direct visualization with conventional instruments or laser.

The closure begins by resuspending the nasopharynx from the skull base and replacing the hemipalate flap. A cricopharyngeal myotomy is performed, and the soft tissue closure is performed routinely. In dentulous patients, lingual splints may be required to prevent nonunion of the mandibulotomy. Although the approach per se does not involve the sacrifice of any cranial nerves, removal of the tumor is often associated with the sacrifice of one or more of these nerves. As a result, these patients may have swallowing difficulties and aspiration postoperatively.

Combined Therapy

Most patients with advanced tumors of the nose and paranasal sinuses will receive radiation therapy in addition to surgical excision, especially if there is a question of residual disease. There is controversy over whether radiation therapy should be employed pre- or postoperatively. Proponents of preoperative radiation contend that radiation therapy reduces the frequency of orbital exenteration. Preoperative radiation therapy is associated with a higher incidence of wound complications and interferes with the accurate assessment of surgical margins. Radiotherapists, however, feel that the reduced oxygen content of tissues after surgery interferes with the efficacy of radiation therapy.

Even with state of the art imaging techniques, accurate staging cannot always be achieved. Withholding radiation therapy until after surgery allows more accurate planning of treatment ports. In one study, 31 per cent of paranasal sinus cancers were underestimated, and only 3 per cent were overestimated preoperatively. The most frequent errors were located in the pterygopalatine and infratemporal fossae (Robin and Powell, 1981).

The benefits of postoperative radiation are demonstrated in the study of Lee and Ogura (1981). They treated 45 patients with T_4 lesions of the maxillary sinus with either radiation therapy or planned preoperative radiation followed by surgical excision. Twenty-eight of these patients had pterygoid plate involvement, 21 had sphenoidal sinus involvement, and 10 had invasion of the ethmoidal sinus-cribriform plate area. The 2-year disease-free rate was 26 per cent for the combined-therapy group and only 8 per cent for the radiation-only group.