Chapter 53: Neoplasms

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True neoplasms of the paranasal sinuses are uncommon in the general population. The most common are squamous cell carcinomas, which occur with a frequency of less than 1:200.000 per year (Batsakis, 1979). Because sinus malignancy initially mimics benign disease, the diagnosis usually becomes evident only after an advanced stage has been reached, thus explaining the relatively poor prognosis. Additionally, as indicated by Ketcham, "no one physician, no matter how intense his interest or ideal his patient referral patterns might be, can accumulate the experience to make him a paranasal sinus (cancer) specialist" (Ketcham, 1985).

This chapter first discusses material common to all sinus tumors: anatomy, symptoms, physical findings, diagnostic assessment, and surgical options. Although these lesions are rare, numerous histologic subtypes must be differentiated and are discussed individually, since management varies significantly among them.

Anatomy

Assessment and surgical management of this group of tumors demands an intense personal effort to understand the complex anatomy of the sinuses and skull base. Such knowledge is best obtained through anatomic dissection and repeated review of the anatomy of the human skull. It may be helpful to have a skull available in the operating room for inspection during complex resections, especially those that approach or traverse the skull base.

Pertinent to this discussion are the structures contiguous to each of the sinuses, since they define the extent of disease and determine the design of therapy.

Maxillary antrum

The most significant structures prognostically are above and behind the maxillary antrum. Superiorly is the orbit and the ethmoid sinus. The posterior border approaches the pterygoid plates, the pterygoid space, and the infratemporal fossa. The other boundary areas are more easily resected en bloc and thus are less worrisome when involved. Anteriorly is the canine fossa, inferiorly the alveolus, and medially the nasal cavity.

Ethmoid sinus

As with the antrum, the least worrisome areas of spread from the ethmoid sinus are inferiorly into the antrum or medially into the nasal cavity. Of much greater concern is spread superiorly, where the fovea ethmoidalis and the cribriform plate provide little barrier to intracranial spread. Evidence of intracranial spread requires an anterior craniofacial resection. The lateral wall consists of the thin bone of the lamina papyracea, which also provides little resistance to neoplastic invasion.

Sphenoid sinus

Located in the center of the head, the sphenoid sinus relates to very complex and vital structures. For this reason, complete resection of a malignant tumor involving the sphenoid sinus is usually not possible. The optic nerve and the pituitary gland are superior, and the internal carotid superior artery and the cavernous sinuses sit laterally. The bone of the lateral wall is usually less than 0.5 mm thick, and not uncommonly is totally dehiscent (Fuji et al, 1979). Anterior to the sinus are posterior ethmoidal cells, and inferiorly are the vidian nerve and the nasopharynx.

Frontal sinus

The least frequently involved by malignant tumor, this sinus is bounded anteriorly by the soft tissues of the scalp, inferiorly by the ethmoidal cells and the orbit, and posteriorly by the anterior cranial fossa.

It is important to realize that all of the sinuses have venous communications with intracranial veins, which provide yet another venue for lethal spread of neoplasm.

Lymphatics

Ohngren (1933) emphasized the importance of a precise understanding of the lymphatic drainage of the nose and paranasal sinuses as a prerequisite to adequate therapy. He contested the established dictum that these tumors do not often metastasize. He held that they do metastasize but that clinical evidence of early metastasis is absent, since the primary drainage is to the lateral and retropharyngeal nodes. He believed that the relative success of his therapy hinged on electrocoagulation and irradiation of the rich lymphatic channels passing from the nose and paranasal sinuses posteriorly to the retropharynx.

The following is a precise description by Ohngren (1933) of the lymphatics of this region:

Ever since first demonstrated by Mascagni the regional lymph nodes of the tumors under review here have been the subject of close study by, chiefly, Simon, Sappey, Kuttner, Key, Retzius, Cuneo, Marc Andre, Most, Turner, Broekaert, Grunwald and Moore. The view I have arrived at regarding the metastasis of these tumors is the outcome of a critical study of the works by the above authors as well as of observation on my own cases.

Close anatomical investigations by the above-mentioned authors have revealed that the lymph channels from the whole of the nasal cavity inside the vestibule from the lateral as well as the medial and lower walls pass in a backward direction towards the choana running together into a plexus in the lateral part of this. From this plexus a few large lymph channels are given off backwards above as well as below the eustachian cushion towards the posterior pharyngeal wall. Here they pick up finer branches from the nasopharynx before entering the retropharyngeal space. The lateral retropharyngeal lymph nodes receive most of these lymph vessels; only small branches pass directly upwards emptying into the medial jugular glands located just below the base of the skull. From the lateral retropharyngeal lymph glands the lymph is conducted in stems of larger calibre which empty into a node belonging to the deep

jugular chain and usually located at the level of the carotid bifurcation. It has generally been left to this node to determine the presence or not of metastases in the tumors we are concerned with. Obviously such methods must lead to erroneous conception of the frequency of metastases in the case of tumors of the lateral nasal wall, since the node is nothing but a secondary localization of metastases whereas the primary metastases located in the retropharyngeal nodes have escaped the attention of the observer.

Symptoms

The most common symptoms associated with malignant tumors of the paranasal sinuses are facial or dental pain, nasal obstruction, and epistaxis. These early signs represent the impact of a necrotic infected mass situated within the paranasal sinus and/or nose. As the disease progresses, it infiltrates adjacent structures, giving rise to additional symptoms as follows:

1. *Diplopia or vision loss* is most often a manifestation of tumor mass compressing or invaing the orbit and may also result from direct involvement of the optic or oculomotor nerves at the orbital apex or the cavernous sinus.

2. *Epiphora* is caused by obstruction or infiltration of the lacrimal duct situated in the anteromedial aspect of the maxilla.

3. *Facial swelling and malocclusion* result from bone destruction and advancement of the tumor into the soft tissues of the face or mouth.

4. *Trismus* signals far-advanced tumor invading the muscles of mastication, most commonly the pterygoid muscles.

5. *Neck mass,* palpable metastatic adenopathy in the jugular chain, is another sign of advanced disease, since the first-echelon nodes are located in the parapharyngeal region.

6. *Hearing loss* usually results from nasopharyngeal extension of the tumor causing serous otitis. This finding is important, since nasopharyngeal extension of disease is a contraindication to surgery.

7. *Facial numbness* is a manifestation of tumor invasion of portions of the trigeminal nerve.

Physical Findings

Until the tumor has infiltrated a cranial nerve or facial bones or grown sufficiently to obstruct a sinus ostium, it will silently advance in size and extent. Discovering an asymptomatic sinus tumor at an early stage is truly a rare and fortuitous event. The most common features includes the following:

1. *Nasal, facial, or intraoral mass.* The intranasal mass is often necrotic, but polypoid mucosa may obscure underlying tumor tissue in the nose. Facial swelling results when an

antral tumor erodes into the soft tissues of the cheek. The widening of the upper alveolar ridge or development of loose, nonvital teeth may be the earliest sign of inferior bony invasion. A palatal mass and ulceration are evidence of more advanced disease.

2. *Proptosis.* Mild protrusion of the eye may be consistent with tumor compression of the periorbita without frank invasion, but usually it reflects intraorbital tumor and implies advanced disease.

3. *Cranial nerve deficits.* The commonly involved cranial nerves are the second (CN II), third (CN III), fourth (CN IV), two branches of the fifth (CN V1 and CN V2), and the sixth (CN VI). Involvement of cranial nerves is a manifestation of advanced disease and indicates a poor prognosis (Ketcham et al, 1974; Weisberger and Dedo, 1977).

Diagnostic Assessment

The choice of diagnostic studies should be guided by the results of a thorough head and neck examination, including a careful assessment of cranial nerve function. Assuming a working diagnosis of neoplasm based on the history and physical examination, the subsequent studies are directed toward establishing the extent of the lesion and its histology.

Imaging studies

CT scan

When obvious evidence of neoplasm exists, the evaluation should move directly to computed tomography (CT) scanning. Baselines sinus radiographs are an unnecessary expense in this setting, whereas the CT scan is helpful to achieve a three-dimensional image of the lesion. Jing et al (1978) have stated that CT is equal to tomography for assessment of bony involvement but superior for assessment of the soft tissue extension. Standard coronal tomography, however, has a place. CT "has been found to have limitations in the delineation of soft tissue disease in areas of high contrast in tissue density (such as dental fillings) and in the evaluation of possible intracranial tumor extension in isodense, avascular lesions" (Jing et al, 1978). CT scanning may also be misleading in evaluation of the orbital floor, where an axial CT scan may give the impression of a mass because of "partial voluming" of the thin bone (Mancuso and Hanafee, 1982). CT scanning is particularly helpful for evaluation of tumor involvement of the retroorbital and orbital apex region. There should be a "central low density region of fat surrounding the optic nerve with a radial arrangement of the extraocular muscles" (Mancuso and Hanafee, 1982). Loss of this plane implies advanced isease, since the veins in this area have a direct contact with the cavernous sinus. CT scanning is also valuable in assessing infiltration of the nasopharynx, where normal anatomy consists of a very thin layer of mucousa over the medial pterygoid plate. Thickening in this region implies tumor infiltration (Mancuso and Hanafee, 1982). When evaluation of intracranial extension is necessary, contrast-enhanced CT scanning may improve the definition of tumor from adjacent brain (Seeger, 1984).

Angiography

Angiography may be appropriate, especially if the lesion demonstrates enhancement during initial CT study or if it approximates the carotid system. It will also be necessary in the evaluation of unusual tumors involving the sphenoid sinus and skull base. In the instance of vascular tumors involving the sinuses, angiography is essential for assessment of tumor extent and delineation of feeding vessels and as an approach for embolization when it is deemed necessary and safe (Overholt et al, 1978). Digital-enhancement angiography would seem to be preferable to conventional angiography, since it is more rapily performed with less need for selective catheterization and requires smaller amounts of contrast (Seeger, 1984).

Ultrasound

Ultrasound B-mode scanning is helpful in assessing orbital masses but is not as precise as CT scanning in defining the borders and extent of lesions.

Magnetic resonance imaging

The impact of MRI on the preoperative assessment of tumor extent has been dramatic. The combination of CT scan to evaluate changes in the bony architecture of the cranial and facial structures with MRI to define soft tissue extent of disease has introduced a new era in planning surgery for sinus neoplasms. The particular benefit of MRI is its ability to differentiate tissue density and predict with near-perfect accuracy the difference between tumor bulk and retained secretions in the sinuses. By using T1 and T2 weighting with gadolinium dye enhancement, the experienced neuroradiologist is able to predict with considerable accuracy the true extent of the tumor. It has been our experience that surgical planning based on these two modalities has been very satisfactory.

Biopsy

Often the lesion will present a surface that lends itself to biopsy at the time of initial clinical presentation. In this instance a biopsy specimen obtained using local anesthesia and a biting punch forceps will move the diagnostic evaluation along rapidly. Infiltration of a local anesthetic containing epinephrine may diminish bleeding. Tumors contained within the sinus cavities should be biopsied transnasally, since transcutaneous or transmucosal approaches may breach the margins of a later en bloc resection. One must be mindful of the possibility of a vascular tumor or an encephalocele when biopsying unilateral nasal masses. Usually palpation of the tumor with an instrument will demonstrate its solid nature. If the mass is soft or cystic, the patient should be asked to perform a Valsalva maneuver while the mass is being observed. Expansion of the mass implies an intracranial connection or a major venous connection. If doubts still exists, it is best to precede the biopsy with aspiration using a fine-gauge spinal needle. If the needle aspirate returns cerebrospinal fluid (CSF) or active bleeding, CT scanning and angiography should be considered before a biopsy is performed.

Aspiration cytology provides another avenue for diagnosis. Martin (1957) recommended it for deep tumors of the antrum. It is particularly useful for tumors that cause proptosis and present along the medial aspect of the orbit. Since the diagnosis in this setting includes benign lesions such as an orbital pseudotumor, histologic evaluation before surgical

invasion is most appropriate. When concern for damage of the orbital contents exists, the aspiration needle can be positioned using CT guidance.

If none of the preceding methods achieves satisfactory material for histologic examination, the tumor must be approached directly. A surgical procedure appropriate to the lesion must be selected (such as frontal trephination, transseptal sphenoidectomy, intranasal antrostomy, or external ethmoidectomy). The operation should be designed to avoid or minimize disruption of later en bloc resection. Possibly, biopsy will be the only surgical procedure carried out during the patient's treatment. In that case wide drainage of the sinus should be carried out to allow for the discharge of necrotic debris that results from subsequent radiation therapy.

Surgical Options

Extirpation of paranasal sinus tumors is a challenging exercise in surgical planning, which may be considered in three phases. First, one must assess the bony and soft tissue structures to be included for en bloc resection. Second, the approach must be designed to provide adequate exposure while preserving functional tissue and cosmetic integrity whenever possible. Third, the repair should be planned to use prosthetics and/or soft tissue techniques to best advantage. See Fig. 53-1 for anatomic reference.

External ethmoidectomy (Fig. 53-2)

Indications

The most limited operation to be discussed, external ethmoidectomy is appropriate for removal of benign tumors of the ethmoidal region and as an approach to biopsy and drainage for tumors of the sphenoethmoidal region and the medial orbit.

Bony excision

The limits of bony resection include the medial orbital wall and the ethmoidal labyrinth.

Surgical approach

The approach is through an incision on the lateral wall of the nose.

Benefits

It allows excellent cosmesis and preservation of functional tissue.

Limitations

It will not provide en bloc excision for any but the most limited tumors (middle turbinate). Typical of any lateral nasal incision, there is a tendency to form a fistula to the nasal cavity when this area is irradiated.

Inferior medial maxillectomy (Fig. 53-3)

Indications

Inferior medial maxillectomy is designed for resection of the medial wall of the antrum and the inferior turbinate. It is most often used for management of an inverted papilloma.

Bony excision

The margins extend laterally to a vertical line dropped from the infraorbital foramen, inferiorly to the floor of the nose, superiorly to the lacrimal fossa and the middle meatus, and posteriorly to the dorsal end of the inferior turbinate.

Surgical approach

A lateral rhinotomy incision is frequently used. The same bony excision may be achieved through a sublabial approach, although an en bloc resection may be more difficult with this approach (Conley and Price, 1979).

Benefits

The procedure allows adequate exposure and resection for limited tumors while preserving functional tissue and providing a very acceptable cosmetic result.

Limitations

It provides en bloc removal of a limited area. When an inverted papilloma is being removed, careful monitoring using a frozen section is often necessary, especially at the posterior margin.

Medial maxillectomy (Fig. 53-4)

Indications

Medial maxillectomy may be used for larger benign or intermediate tumors involving the entire lateral nasal wall but without extension to the orbit, anterior cranial fossa, lateral maxilla, or alveolus.

Bony excision

The block removed contains the lateral nasal wall, including all turbinate tissue, and the contents of the ethmoid and maxillary sinuses.

Surgical approach

An extended lateral rhinotomy incision provides excellent exposure. In some instances the same procedure may be accomplished by combining an ethmoidectomy incision with a

sublabial incision.

Benefits

An en bloc resection may be accomplished with little cosmetic deformity resulting.

Limitations

The posterior and superior margins of resection should be monitored using a frozen section where indicated. Removal of all turbinate tissue results in an abnormal nasal cavity, often requiring chronic management of crusting.

Radical maxillectomy (Fig. 53-5)

Indications

Radical maxillectomy is the standard operation for advanced carcinoma of the maxilla. Considerable debate exists regarding the proper application of this procedure, since the amount of tissue to be removed depends on a careful assessment of the extent of tumor involvement (Sakai et al, 1983; Terz et al, 1980). Another uncertainty relates to the establishment of proper indications for removal of the orbital contents in combination with radical maxillectomy (p. 946).

Bony excision

Complete radical maxillectomy includes removal of the maxilla along with the nasal bone, the ethmoid sinus, and in some instances, the pterygoid plates.

Surgical approaches

The Weber-Fergusson incision is used with extensions around the eyelids to preserve those soft tissues. A skin or dermis graft is used to line the defect (Bryce, 1984). A preformed obturator can be of great assistance in the immediate postoperative period by acting as a support for packing.

Benefits

This operation is adequate treatment for malignant tumors confined to the maxilla and those with extension to the facial soft tissues, palate, or anterior orbit but without invasion of the ethmoidal roof, posterior orbit, or pterygoid region. When the procedure is supplemented by irradiation, one may expect a cure rate approximating 30% (Jackson et al, 1977; Sisson, 1970; Wang, 1983). There is, however, continuing debate regarding the indications for resecting the orbital contents in patients with advanced paranasal sinus carcinoma. As indicated by Ketcham and Van Buren (1985) the decision to excise the orbital contents has been made on the basis of achieving an "en bloc" resection of the tumor. However, Ketcham implied that as imaging became more precise more selective resection might be possible. Perry et al (1988) emphasized the impact of CT scanning on surgical planning and documented that selective preservation of the orbital contents is appropriate and

achieves adequate local control in properly selected cases. This finding is consistent with the experience of Weymuller et al (1980) who also could not demonstrate an improved local control rate or survival advantage when the orbital contents were included in maxillectomy procedures.

Limitations

Even when orbital exenteration is included, a maxillectomy fails to provide an adequate resection when the tumor has escaped superiorly (ethmoidal roof) or posteriorly (orbital apex, pterygoid region). When these areas are demonstrably involved, one must decide whether to undertake craniofacial resection or use a regimen as described by Sakai et al (1983), which relies on chemotherapy, curettage, and irradiation.

Craniofacial frontoethmoidectomy (Fig. 53-6)

Indications

Craniofacial frontoethmoidectomy is specifically designed to provide en bloc resection for tumors of the ethmoidal bone and frontal regions. By including exposure of the anterior cranial fossa, the procedure makes possible complete resection of the ethmoid and frontal sinuses, and dural resection may be included when necessary.

Bony excision

Resection may include the anterior cranium (including the frontal sinus), the floor of the anterior cranial fossa, the ethmoid labyrinth (and the eye when necessary), and the nasal septum. If the tumor crosses the midline, the procedure may be performed bilaterally.

Surgical approach

The approach described by Johns et al (1984) allows excellent exposure with preservation of the supraorbital rim to diminish the cosmetic impact of the surgery. Although removal of the eye is unpleasant to consider, Ketcham et al (1974) have documented an improved survival when orbital resection is combined with craniofacial resected (57% versus 26% without exenteration).

Benefits

Craniofacial exposure provides direct visualization of the cribriform plate and the fovea ethmoidalis and the potential for en bloc removal. It also provides wide exposure to allow effective repair of dural tears, thus diminishing the change of a postoperative CSF leak and intracranial infection. In addition, the exposure afforded would allow intraoperative irradiation or placement of a radioactive implant.

Limitations

If the tumor extends to the sphenoid sinus, the cavernous sinus, or transdurally, en bloc resection cannot be achieved.

Extended craniofacial resection (Fig. 53-7)

Indications

In actuality there is no one definitive extended craniofacial resection to be described. Extensive tumors involving the anterior skull base, including certain tumors with involvement of the pterygoid plates, can be surgically approached, with the resection to include any and all of the structures outlined below. Each operation is individually tailored according to the extent and nature of the tumor to be excised (Smith, 1990; Janecka, 1990).

Bony margins

The posterior line of resection is defined by the oval foramen, the round foramen, and the internal carotid artery. The excision may extend through the sphenoid sinus and up to the contralateral optic nerve. The remaining margins are those of the craniofacial frontoethmoidectomy and the radical maxillectomy.

Surgical approach

A team including a neurosurgeon and a head and neck surgeon should perform this operation. A combination of bicoronal and anterior or lateral facial incisions is used for exposure. The closure is improved by including a split-galea flap to cover the dura (Schramm, 1979).

Benefits

Thorough exposure and complete excision of otherwise unresectable tumors are the main benefits of this procedure.

Limitations

In discussing the limitations of the craniofacial approach, Ketcham et al (1974) have cited clear-cut pterygoid plate erosion and cranial nerve invasion as indicators of inoperability. Even though this operation has extended the limits of surgical resection and in so doing has resulted in numerous cures, it still must be accepted that it does not provide adequate en bloc resection for tumor at the orbital apex, in the nasopharynx, or deeply infiltrating the pterygoid space.

Benign Neoplasms

Osteomas and chondromas

Osteomas are most commonly found in the frontal sinus; the ethmoid sinus and maxilla follow, in that order. When located away from the sinus ostium, an osteoma is silent and only discovered incidentally during radiographic examination. In this instance it should be followed with an interval radiograph in 1 to 2 years to assess growth. Osteomas are formed of mature lamellar bone and cause symptoms only when they interfere with sinus drainage or possibly when they impinge on the dura. When the osteoma causes a mucocele, obliterative

surgery or wide drainage is recommended (osteoplastic frontal sinus obliteration or external ethmoidectomy).

Chondromas may develop anywhere in the sinonasal tract. Batsakis (1979) has emphasized the tendency to underestimate the aggressive nature of these neoplasms. According to him, the histologic differentiation between benign and malignant tumors is "incompletely defined". Because of this histologic uncertainty, resection should be more aggressive than with an osteoma and consideration should be given to "fairly radical treatment".

Schwannomas and neurofibromas

Schwannomas and neurofibromas are indolent tumors arising from peripheral nerve components. A schwannoma is an isolated encapsulated lesion, whereas a neurofibroma is woven into the nerve and is often one of multiple lesions. The tumors cause symptoms by slow progressive growth that may distort tissues by pressure or become symptomatic by obstruction of a sinus ostium. During evaluation a contrast-enhanced CT scan may be performed. Neuromas and neurilemmomas demonstrate a characteristic irregular patchy appearance (Mancuso and Hanafee, 1982). These lesions are managed by conservative local resection. One should recall that malignant transformation occurs in about one of eight patients with multiple neurofibromatosis (von Recklinghausen's disease) (Hyams, 1984).

Ossifying fibromas and cementomas

Although somewhat difficult to differentiate histologically from fibrous dysplasia, ossifying fibromas may be diagnosed by including clinical and radiographic criteria. They occur in an older age group, primarily young adults, and are typified radiographically by a sclerotic bony margin that is evident at surgical resection, when the lesion often "shells out" (Hyams, 1984).

Cementomas are described as a variant of ossifying fibromas with a "cementum-like osseous element". These, too, are appropriately managed by local excision, but if any tumor is left, a recurrence is likely.

Odontogenic tumors

This rare group, accounting for only 1% of all jaw tumors, includes ameloblastoma and the calcifying epithelial tumor of Pindborg (McClatchey, 1979). They are locally aggressive, requiring resection with a small margin of normal tissue to prevent recurrence (Hyams, 1984). The vast majority of odontomas are better characterized as hamartomas, emphasizing their benign nature and their evolution from a developmental abnormality in odontogenic material (McClatchey, 1979).

Intermediate Neoplasms

Inverted papillomas

Inverted papillomas usually present as polypoid unilateral nasal masses. On occasion they may be found in association with allergic nasal polyps, which accounts for the need to always submit labeled, separate specimens from each side of the nose when performing routine polypectomy. The histology consists of infolded epithelium that may be squamous, transitional, or respiratory. The incidence of frank malignant change approximates 10% (Hyams, 1970, 1971).

Even when not malignant, these lesions must be treated with respect, since inadequate excision is likely to result in recurrence (Cummings and Goodman, 1970). Most often, a lateral rhinotomy incision combined with medial maxillectomy will allow adequate margins (Myers et al, 1981).

Meningiomas

Extracranial meningiomas, which arise from ectopic arachnoid tissue, are very rare tumors. They have a variable histology and may require electron microscopy for identification (Leipzig and English, 1984). Occasionally an intracranial meningioma will invade the sinuses or orbit. In this setting plain skull films may reveal hyperostosis of the ethmoidal region. Further evaluation should consider CT scanning and angiography or MRI (Seeger, 1984). Surgical excision is the only form of definitive management although radiation therapy may be effectively used to "stabilize" inoperable lesions.

Hemangiomas

On rare occasions a primary hemangioma will occur in the maxilla. Hyams (1984) has emphasize the possibility of an erroneous diagnosis when a pyogenic granuloma is mistaken for a true hemangioma. More frequently, the maxilla will become involved with a soft tissue hemangioma of the face or a vascular malformation of the skull. Hemangiomas are typified by rarefaction on radiographs, although they become sclerotic as they mature. The lesions are typically asymptomatic, and extirpation is indicate only for bleeding or significant discomfort. When resection is necessary, it should be preceded by angiography, and selective embolization should be seriously considered as an adjunct to surgical resection (Biller et al, 1982).

Hemangiopericytomas

Hemangiopericytomas typically occur in the nose but may involve the sinuses. According to Batsakis (1979), "If there is one common denominator for hemangiopericytomas, it is the lack of uniformity in appearance, growth and biological behavior". Clinical behavior may vary considerably from a slowly enlarging rubbery mass to an infiltrating aggressive neoplasm. It is Batsakis' opinion that these tumors are locally aggressive and must be respected as tumors that are likely to recur and that are associated with delayed recurrence, making 5-year survival an inadequate measure of cure.

Malignant Neoplasms

Epidemiology

Numerous environmental agents are considered to have a causal relationship with carcinomas of the paranasal sinuses. The following are all associated with squamous cell carcinomas: aflatoxin found in certain foods and dust; chromium, nickel, mustard gas, polycyclic hydrocarbons, and other organic chemicals, usually from manufacturing processes; and mesothorium (Thorotrast), a radiopaque dye used as a contrast medium within the antrum (Keane et al, 1981). Wood dust has a particular association with adenocarcinoma of the ethmoid sinus (Klintenberg et al, 1984).

Most series document squamous cell carcinomas to be the most common histologic type, with an incidence of roughly 80%. Adenoid cystic carcinomas and adenocarcinomas are next in frequency (approximately 10%). Numerous other tumors complete the list in small numbers.

Epithelial malignancy

Staging

Staging is appropriately discussed only in the context of carcinomatous tumors, since the other lesions are so uncommon that they do not warrant such attention (or such confusion). As Harrison (1978) has suggested, the staging system should allow differentiation between distinct stages of disease progression. Unfortunately, most of these tumors present with such advanced disease that the subtle differences between cases may not be relevant to clinical management or prognosis. Therefore "one must conclude that attempts at classifying this tumor finitely are both impractical and unrealistic" (Harrison, 1978).

Ohngren (1933) emphasized the ominous significance of tumor spread in a posterior or superior direction. The imaginary line extending from the medial canthus to the angle of the jaw gives a rough estimate of the dividing line between tumors that may be resected with a good or poor prognosis. He developed a staging system based on personal experience that included topography (tumor location and extent), histology (three grades), and cervical metastases (presence or absence). That the system was effective is borne out by his statistics, which clearly show a difference in 3-year "freedom from symptoms" between groups I (82%), II (63%), and III (25%).

Other systems have been proposed by experienced surgeons. Each represents a personal variation of the generally accepted schemes, but they differ from one another sufficiently to make direct comparison of results impossible, thus negating one of the primary goals of any staging system. The interested reader is referred to Harrison's (1978) article for further discussion and references.

In the USA we are probably best served by consistently using the format outlined by the American Joint Committee on Cancer (AJCC), although an adjustment based on Harrison's (1978) criticism should be seriously considered. In particular, orbital apex involvement and pterygoid muscle involvement should be placed in the T4 category. It also appears that

Ohngren's (1933) inclusion of tumor differentiation may be relevant to the staging of these tumors. That this may be a reasonable contention is borne out by Wang's (1983) statistics regarding 3-year survival according to the cell type: squamous cell, 34% (15 of 44 patients); undifferentiated carcinoma, 29% (4 of 14 patients); transitional cell carcinoma, 40% (2 of 5 patients); and carcinoma with inverted papilloma, 75% (3 of 4 patients). However, Shidnia et al (1984) could not confirm a relationship between histology and survival.

The advent of combined CT and MRI scanning for neoplasms of the paranasal sinuses will allow for more accurate anatomic assessment of individual cases. This improved definition of the tumor boundaries should also allow more precise staging. It is my contention that MRI and CT scanning should be required for staging of these neoplasms.

Staging criteria: primary tumor (T)

Tx Minimum requirements to asses primary tumor cannot be met
T0 No evidence of primary tumor
Tis Carcinoma in situ
T1 Tumor confined to antral mucosa of infrastructure with no bone erosion or destruction
T2 Tumor confined to suprastructure mucosa without bone destruction, or to infrastructure with destruction of medial or inferior bony walls only
T3 More extensive tumor invading skin of cheek, orbit, anterior ethmoid sinus, or pterygoid muscle
T4 Massive tumor with invasion of cribriform plate, posterior ethmoid sinus, sphenoid sinus, nasopharynx, pterygoid plate, or base of skull.

Management

Definitive treatment has included surgery, irradiation, and electrocoagulation in varying combinations. It does not seem that substantial improvements have been achieved in the past 50 years if Ohngren's (1933) 3-year statistics are used as a standard of comparison (group I, 82%; group II, 63%; and group III, 25%). More recently, adjunctive chemotherapy has been introduced, but its impact on outcome is still uncertain.

Maxillary sinus. Early (T1 and T2) lesions may be treated by surgical resection (maxillectomy) or irradiation. Although Wang (1983) has not recommended definitive irradiation, he has reported curing 4 of 4 patients with T2 maxillary sinus tumors with that modality alone. Bryce (1984) has also stated that irradiation alone (5500 rad) is an effective way to manage early tumors but that 25% to 30% of patients will die of local recurrence.

The majority of patients have advanced (T3 or T4) disease when first seen, which most authors agree requires combined therapy for optimal control (Bryce, 1984; Sakai et al, 1983; Shidnia et al, 1984; Wang, 1983). Considerable variation exists within each reporting institution regarding the application of surgery and irradiation. All series are retrospective and present a mixture of patient populations, usually receiving the following types of management: surgery for cure, surgery with preoperative or postoperative irradiation, and irradiation with surgical salvage. This hodgepodge of retrospective data leaves one in a muddle, although some trends are becoming apparent.

Surgery alone. Surgery alone is generally accepted as adequate management for tumors isolated to the antral mucosa (Jackson et al, 1977). As stated above, most authors agree that T3 and T4 lesions require combined therapy. Terz et al (1980) have taken a more aggressive surgical posture and reported dramatic improvement in survival for patients with T3 and T4 lesions when craniofacial resection was performed. According to these authors, craniofacial resection achieved an astonishing 3-year disease-free survival of 72% (15 of 22 patients) among patients who had T3 or T4 tumors. The numbers are even more surprising when one realizes that irradiation was not used. However, reporting separately on patients from the same institution and the same time period as that of Terz et al, Amendola et al (1981) compared the outcome of 19 patients who underwent "radical craniofacial surgery" with that of 20 patients treated with megavoltage irradiation and a Caldwell-Luc procedure. Their reported 3-year survival of the group undergoing radical craniofacial surgery was 37%, whereas the irradiated group had a 3-year survival of 50%. Were the surgical patients reported by Terz et al identical to those reported on by Amendola et al? If so, how do we resolve the dichotomy of reporting and in what light should we hold any retrospective report concerning an uncommon disorder that is typically managed on an individual basis?

Surgical management should include consideration of immediate rehabilitation. This is usually best achieved with prosthetics that allow immediate restoration of function and yet are removable to allow inspection of the maxillary cavity. Preformed palatal plates may be used to support transoral packing in the postoperative period and may later be modified to occlude the oronasal defect. If one is blessed with a talented prosthetic department, the aftermath of a craniofacial-orbital resection can be reasonably well camouflaged.

Combined irradiation and surgery. With T3 tumors little difference exists between preoperative and postoperative irradiation. Postoperative irradiation will often control microscopically positive resection margins (Shidnia et al, 1984). With the more advanced lesions, especially those where resectability is questionable, preoperative irradiation appears to be the procedure of choice.

A planned preoperative dose of 5500 rad is used in Toronto, followed by radical resection (Bryce, 1984). If no tumor is found, a 5-year survival of 77% occurs; if tumor is found, the survival drops to 23%; overall survival is 50%. Shidnia et al (1984) have also noted the favorable effects of a tumor-free specimen. Their group of patients receiving irradiation followe by surgery had an absolute 30-month survival of 28% (9 of 30 patients), whereas their group of patients receiving surgery followed by irradiation had a survival of 45% (11 of 24 patients). Shidnia et al have suggested that the latter group was selected for more favorable lesions, since the study was not designed in a prospective manner. For operable lesions (meaning those not extending to the nasopharynx, skull base, or pterygoid fossa), Wang (1983) favors radical surgery to remove tumor bulk and establish drainage followed by 5000 to 5500 rad in 6 weeks.

The most emotionally charged decision regarding the management of advanced maxillary sinus tumors is the need for orbital exenteration. The experience of Ketcham et al, which indicates a doubling of survival with removal of the orbital contents for the management of ethmoid neoplasms (57% versus 26%), provides strong encouragement to perform orbital exenteration for advanced operable disease. However, analysis of the benefits of orbital exenteration in two other studies leaves this issue open to question (Perry, 1988;

Weymuller, 1980).

Radiation therapy. Wang (1983) takes special care to shield the eye and the frontal lobe. He has reported a 24% NED (no evidence of disease) for his 34 patients treated with irradiation alone. In contrast, Ellingwood and Million (1979) have intentionally included the orbit to include the probable routes of tumor extension within the irradiation field. Their cure rate of 71% suggests a successful regimen. However, all patients treated with this regimen lost vision in the treated eye. Amendola et al (1981) have also documented a 3-year absolute survival of 55% using an average of 6600 rad for lesions that were primarily T4. Then fields were angled posteriorly to avoid the contralateral eye. No complications of irradiation are mentioned in this article, and the authors stress the improved quality of life when irradiation for cure is contrasted with craniofacial resection.

Perhaps the most significant variation from traditional management has been reported by Sakai et al (1983). They have accumulated more than 780 patients since 1957 at Kobe and Osaka Universities in Japan. Their cumulative experience demonstrates impressive support for their current regimen, which consists of 5000 rad, continuous intraarterial 5-fluorouracil (5-FU) infusion, tumor reduction (via the Caldwell-Luc procedure), and immunotherapy. In the reported time period they have seen a rise in 5-year survival from 20% (282 patients treated with 7000 rad and maxillectomy, 1957-1966) to 54% (134 patients treated with the current regimen, 1976-1979). They have simultaneously seen a substantial decrease in the functional disability encountered by their patients. An analogous regimen has been reported from Rotterdam, Holland, with similarly encouraging results in a series of 60 patients (2-year survival, 76% 5-year survival, 65%) (Kneght et al, 1984).

Inoperable neoplasms. Irradiation may be used to determine operability in tumors that initially appear inoperable. In this situation Wang (1983) delivered 6000 rad in 6 weeks, with surgery to follow. He encouraged aggressive surgery if the response to irradiation appeared to make the lesion operable. If margins were subsequently positive, a further boost of local irradiation was considered. Interestingly, Wang had better results with this more advanced group (survival of 58% or 11 of 19 patients) than he did with smaller lesions, in which he favored surgery to be followed by irradiation (survival of 36%, or 5 of 14 patients). The rationale for treating patients with "curative" doses of irradiation when their lesions are initially inoperable is borne out by the number of long-term survivors seen in this group of patients (Shidnia et al, 1984; Wang, 1983). This result is probably a manifestation of the heterogeneous nature of the inoperable patients; some are inoperable by virtue of tumor extent, whereas others are medically unsuitable or have refused surgery outright.

Lee et al (1974) have documented the potential value of intraarterial infusion chemotherapy (5-FU) and simultaneous irradiation for initially inoperable tumors. They achieved an absolute 2-year survival of 50% in this group with a very poor prognosis. Survival dropped to 28% at 5 years. Unfortunately, optic nerve damage was fairly prevalent and caused bilateral blindness on two occasions. Subsequent to this study Goepfert et al proceeded to develop a technique for "combined super-selective intrarterial and systemic chemotherapy". They have reported a 43% complete response rate and a combined complete and partial response rate of 91%. A number of these patients went on to receive radiation therapy and "extensive cranial facial surgery was avoided in seven of nine patients with CR and one patient with PR (near CR); these eight patients underwent radiotherapy directly after

completion of induction chemotherapy" (Lee et al, 1989). The utilization of selective chemotherapy and irradiation for advanced tumors of this area has tremendous appeal when contrasted to the horrendous defects created by surgical ablation of lesions in the axial region of the skull. It is hoped that follow-up information on these patients will reveal a sustained result leading to a change in our management.

Ethmoid/sphenoid and frontal sinuses. Ethmoid/sphenoid and frontal sinus tumors are especially likely to present with evidence of advanced disease. Tumor extension to the structures of the skull base can be expected at initial presentation in 50% of patients (Ellingwood and Million, 1979; Kitt and Panje, 1984).

The argument for craniofacial resection in patients with malignancy extending to or primarily arising from the ethmoid sinus has been presented by Ketcham et al (1974). En bloc tumor resection, improved assessment of disease extent, and better cosmesis provide the rationale for applying this technique. The researchers obtained a 5-year survival of 41% in a particularly difficult group of patients, many of whom were referred with advanced disease after failure of previous treatment. Ketcham et al noted a decrease in survival with preservation of the orbital contents (26%) as compared with orbital resection (57%). Further documentation of the techniques and applicability of cranial facial resection and its significant impact on improved local control and survival is provided by Shah (1987), Biller et al (1989), and Panje et al (1989).

Ellingwood and Million (1979) have stressed the importance of properly designed fields to include the whole orbit (when necessary) and skull base. Using primary irradiation, they achieved a 5-year actuarial survival of 59% and a 5-year NED of 50%. The incidence of major complications in this series was substantial and included blindness in every eye treated with irradiation of the whole orbit.

In Wang's (1983) series, 4 of 12 patients (33%) managed with irradiation alone survived 3 years, whereas 8 of 17 patients (47%) managed with surgery and postoperative irradiation survived 3 years NED.

Primary carcinoma of the frontal sinus is a very uncommon entity. According to a review article by Brownson and Ogura (1971), the survival is 1 of 33 patients. One would anticipate improvement of these statistics as craniofacial resection and appropriately designed irradiation fields are used.

Cervical lymph nodes. Involvement of cervical nodes will occur in about 25% to 35% of cases (Batsakis, 1979). Most authors agree that palpable cervical lymph nodes indicate a very poor prognosis (Weymuller et al, 1980). In contrast, the experience of Shidnia et al (1984) suggests that if neck disease is discovered at an early stage, it can be successfully managed with neck dissection. These authors advise inclusion of the ipsilateral neck and preauricular area in the initial field of irradiation for T3 and T4 tumors.

Adenocarcinomas. Adenocarcinomas have an epidemiologic association with woodworking, furniture making, and leatherwork (Klintenberg et al, 1984). They are more commonly found in the upper nasal cavity and ethmoid sinus and are usually lethal by virtue of local progression. In light of its propensity for aggressive local progression,

adenocarcinoma should be treated in a surgically aggressive fashion with en bloc resection when possible. It is hoped that statistics for this particular disorder will improve as application of craniofacial techniques becomes more frequent.

In contrast to adenoid cystic carcinomas, adenocarcinomas have a relatively low incidence of distant metastasis, but the long-range survival is equally poor (Batsakis, 1979). There is a range of histologic differentiation, with patients with the poorly differentiated form having a 5-year survival of no better than 20% (Hyams, 1984), whereas patients with well-differentiated papillary tumors have a better survival (Batsakis, 1979).

Adenoid cystic carcinomas. Adenoid cystic carcinomas occur with about the same frequency as adenocarcinomas but most commonly involve the antrum. They have a variable histologic pattern, and substantial biopsy tissue may be necessary before an accurate diagnosis can be established. Also part of the histologic picture is their propensity for invasion along nerve sheath structures. These tumors are more likely to express distant metastases than regional metastasis, although the most likely cause of death is relentless local neoplasm at the skull base (Batsakis, 1979). Initially high cure rates (5-year survival of 75%) shrink as time goes on, with the AFIP registry reporting a 15-year survival of 25% (Hyams, 1984).

Management relies on surgical resection with irradiation added if the margins are in question (Wang, 1983). Bryce (1984) has recommended using only one major modality (surgery or irradiation) at a time, since the 5-year survival is good and late recurrence is virtually a certainty. This posture is based on a desire to hold a potentially effective treatment modality in reserve. One wonders whether this is treating the physician or the patient, since we do not know if combined treatment diminishes the recurrence rate or increases the disease-free interval. A contrasting opinion has been offered by Horree (1974), who has recommended "three-modality therapy", including regional infusion with 5-FU, maxillary resection, and then irradiation. Neoplasms representing the entire spectrum of salivary gland malignancy have been reported to occur in the paranasal sinuses but are not discussed here.

Analysis of experience gained in the few centers that provide neutron irradiation has demonstrated a significant benefit of that form of radiation in the management of salivary gland neoplasms. When confronted with an adenoid cystic carcinoma, and particularly one that is otherwise untreatable, the use of neutron beam irradiation should be seriously considered (Koh, 1989).

Complications. The incidence of complications increases significantly when surgery breaches the intracranial space and also when high doses of irradiation are delivered to the orbit or intracranial contents.

Major complications of surgery are diminished by the use of perioperative antibiotics and by careful attention to dural closure including the use of pericranial flaps. The most significant complications include meningitis, brain abscess, CSF leak, and postoperative wound hemorrhage.

Cataracts are the most common complication of sinus irradiation (Bryce, 1984). Other radiation-induced lesions include osteoradionecrosis, keratitis, optic neuritis, and hypopituitarism. Injury to the optic nerve is made more likely with increased fraction size

(250 rad/fraction) and total dose (Wang, 1983). After high-dose irradiation, which includes the whole orbit, one can expect loss of vision in 100% of cases (Ellingwood and Million, 1979).

Therapeutic decision making. Taking into consideration the data reviewed above, some conclusions regarding therapy for advanced disease seem justifiable:

1. Most tumors are at an advance stage when first recognized and require aggressive multimodality treatment.

2. Survival and local disease control are better when the orbit is included in the therapeutic plan (that is, exenteration or irradiation).

3. A significant chance of metastasis to the primary (pharyngeal) and secondary (cervical) nodes exists. They should be included in the primary treatment plan.

4. Higher success rates have been reported with primary irradiation, and the associated functional and cosmetic impairment seems to compare favorably with that of surgery.

5. The best regimen may prove to be a combination of chemotherapy, irradiation, and limited surgical debridement. This, of course, requires the availability of sophisticated equipment and a cooperative effort by a multidisciplinary team.

Sarcomas

Osteogenic sarcomas and chondrosarcomas of the facial skeleton are encountered more commonly in the mandible than in the maxilla. They are usually lethal by virtue of relentless local progression. In that light, the most successful therapy is based on wide en bloc resection, with 5-year survival being in the 10% to 20% range. Irradiation seems to be an effective adjuvant for both tumors. De Fries and Kornblut (1979) have reported a survival of 30% with induction chemotherapy, preoperative irradiation, and wide-field ablative surgery. The multi-institutional analysis of outcome from neutron beam irradiation has identified facial sarcoma as an area in which significant improvement was identified. Neutron beam irradiation, when available, should be considered especially in the management of surgically unapproachable or unresectable lesions (Laramore, 1989).

Hyams (1984) has characterized a well-differentiated and a poorly differentiated form of fibrosarcoma. One must consider the diagnosis of aggressive fibromatosis when evaluating a fibrous tumor in young adults (Wilkins et al, 1975). All tumors in this group require en bloc excision for cure (Fu and Perzin, 1976).

Hematopoietic and lymphoid tumors

The paranasal sinuses are an uncommon primary site for lymphomatous tumors, but any of the multiple subtypes may occur. Since subtyping will determine both the therapy and the prognosis, the most important role of the otolaryngologist - head and neck surgeon is to provide adequate tissue in good condition for histologic processing. If a lymphoma is suspected, the pathology department should be notified before the biopsy is done so that fresh

specimens may be properly managed for subtyping.

Treatment is usually chemotherapy, although irradiation alone is adequate for stage I and stage II Hodgkin's disease (Wang, 1983).

Malignant melanomas

Melanomas of the paranasal sinuses are usually advanced at the time of discovery. In addition to symptoms typical of sinus tumors, the patient may note a black nasal discharge and have a pigmented mass within the nose. Frequently regional or distant metastases exist at the time of initial examination. In the AFIP series (Hyams, 1984), the 5-year survival was 11% and the 20-year survival was 0.5%, with a mean survival of 2.3 years. Although radical surgical removal is the standard of care, it is obviously not very effective. Irradiation has been considered inadequate, but adjustments in fractionation may provide better results, as suggested by Berthelsen (1984), who found local control in three of six patients with nasal melanomas treated with primary irradiation.

Paranasal Sinus Neoplasms in Children

Sinus tumors in children are usually benign. The common nasal sinus tumors of children are polyps arising on an allergic basis or in association with cystic fibrosis. The most common true neoplasms are fibroosseous lesions and tumors of dental origin. Mucoceles and hemangiomas are less frequently encountered. The malignant tumors are predominantly embryonal rhabdomyosarcomas, although epithelial malignancies may occur.

Tumors should be assessed radiographically as discussed earlier, and the diagnosis should be established by a transnasal biopsy with a generous tissue sample submitted for pathologic study, since special stains and electron microscopy may be necessary.

Tumors are usually managed surgically because (1) most lesions are radiosensitive, (2) irradiation in excess of 3000 rad will have a substantial effect on growth of the facial skeleton, and (3) there is a significant risk of radiation-induced malignant transformation. Surgery should be designed to respect the developing tooth buds and facial growth centers in the palate, nose, and zygomatic process of the maxilla. The tendency is toward conservative local resection, especially with tumors of dental origin, but some lesions will require en bloc resection for a curative effort.

Monostotic fibrous dysplasia is a lesion considered to be a developmental defect of bone (Batsakis, 1979; Hyams, 1984). As such, it is most common in the pediatric age group and is more common in the maxilla than in the mandible. According to Schramm (1979), in contrast to most bony and odontogenic tumors, fibrous dysplasia sometimes requires wide local removal, including partial maxillectomy, to achieve clear margins. Resection causing a major functional or cosmetic deformity (orbital exenteration, mandibulectomy) is not often warranted (Ramsey et al, 1968). Ossifying fibromas and cementomas, in contrast, are well delineated and may be managed by local excision with a minimal margin.

Rhabdomyosarcomas are the most common malignant neoplasm of the upper respiratory tract in the pediatric age group (age 15 or younger) in the AFIP records (Hyams,

1984). The tumor is aggressive, demonstrating rapid progression and dissemination. According to Hyams (1984), the histologic subcategories of juvenile rhabdomyosarcoma (alveolar, botryoid, and embryonal) have no relevance to the neoplasm's clinical behavior or the prognosis, whereas the Intergroup Rhabdomyosarcoma Study (IRS) had suggested an unfavorable prognosis for cytologic anaplasia or monomorphous round cell patterns (Maurer and Ragab, 1984). The best survival is obtained with clear surgical margins supplemented by irradiation an chemotherapy (Feldman, 1982). The IRS has evaluated results from 157 nonorbital and 54 orbital rhabdomyosarcoma case. They have found patients with parameningeal involvement (as indicated by CT evidence of skull base erosion or spinal fluid tests demonstrating pleocytosis as well as elevated protein and decreased glucose levels) to have a poor prognosis. This group of patients has been benefited by a treatment regimen combining chemotherapy and craniospinal irradiation.

Patients with a circumscribed local tumor and no evidence of distant metastases have an excellent prognosis when treated with multidrug chemotherapy and adjuvant irradiation. In most instances radical ablative surgery is not considered necessary to achieve a successful poutcome (Maurer and Ragab, 1984).