Chapter 187: Differential Diagnosis of Neoplasms of the Anterior and Middle Cranial Fossae

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Tumors of the anterior (ACF) and middle cranial fossae (MCF) have often been considered inoperable because of the anatomic complexity of adjacent structures. The development of sophisticated surgical approaches to the skull base by otolaryngologists - head and neck surgeons has made tumors affecting this region resectable. Many lesions cannot be adequately removed by strictly intracranial or extracranial approaches alone. A combined approach involving multiple surgical teams is often the only way to achieve a beneficial result.

Evaluation of the extent and histology of lesions in the ACF and MCF can be difficult. These regions are affected by a wide range of pathologic processes arising intracranially, or within the intervening bone. Pathology in these areas may not be externally visible until it is far advanced. The naturally occurring foramina and fissures allow both benign and malignant tumors to pass freely within the bone without necessarily causing erosion. This problem has often resulted in late diagnosis and a lack of appreciation for the tumor's true extent. Biopsy is not always safe or even possible.

Surgical Anatomy

The adult chondrocranium extends from the root of the nose to the superior nuchal line and consists of both an intracranial and extracranial surface. The floor of the ACF on each side of the midline is formed mainly by the orbital plate of the frontal bone. The floor descends anteromedially to a thin plateau that is the roof of the ethmoid sinuses; then more medially its posterior aspect becomes the cribriform plate. This 1 mm thick, perforated based continues as the planum sphenoidale, which extends laterally as the lesser wing of the sphenoid.

Tumors of the orbit are related to much of the inferior surface of the fossa, but intracranial extension in this area more often arises from the ethmoid labyrinth and the sphenoid sinus. The posterior walls of the frontal sinuses also form part of the anterior aspect of the ACF floor.

The butterfly-shaped MCF has a more complex anatomy, containing many foramina and fissures. Within its anterior wall, between greater and lesser sphenoid wings, the superior orbtial fissure connects the orbit with the MCF. The anterior wall also provides potential communication between the MCF, the pterygopalatine fossa, and the inferior orbital fissure. Viewed from below, the lateral aspect of the MCF floor forms the roof of the infratemporal fossa, which not only contains the muscles of mastication, internal maxillary artery, and the mandibular division of the fifth cranial (trigeminal) nerve (CN V), but also communicates with the pterygomaxillary fissure, nasopharynx, and parapharyngeal space.

Radiologic Assessment

In patients with suspected ACF and MCF lesions, the use of high resolution contrast-enhanced computed tomography (CT) and gadolinium-enhanced magnetic resonance imaging (MRI) is invaluable in defining the extent of a lesion, determining if it is resectable, suggesting its type, guiding a biopsy if warranted, and, in the case of resectable tumors, allowing the surgeon to plan a resection that will maximize benefit to the patient while minimizing morbidity.

Despite their unquestionable utility, CT and MRI do have specific limitations. Some patients are unable to maintain the required position long enough for these studies to be conducted. Sedation may be useful if the study is deemed necessary. Neck stiffness can prevent the patient from extending the neck sufficiently for coronal CT. These views can be reconstructed from axial scans but with a significant loss of resolution. MRI is superior to CT because high-resolution views in multiple planes can be constructed without requiring repositioning and rescanning.

CT is most useful to detect bone erosion, intracranial extension, and tumor vascularity. MRI is inferior to CT to detect bone erosion but more useful to detect soft tissue invasion, great vessel distortion, and dural involvement.

MRI and CT three-dimensional reconstruction images make spatial relationships easier to comprehend. The images can be constructed to view the anatomy in question at a variety of angles and from a variety of perspectives. The complex anatomy of the ACF and MCF makes three-dimensional reconstruction a highly useful technique for evaluating the relationship between lesions and normal structures in these areas. Unfortunately, this technology is still experimental and only available at a few centres.

Carotid angiography carries an estimated morbidity of 2.25% and mortality of 0.1% (Shugar and Mafee, 1982). Therefore, this imaging technique should be reserved for specific cases such as vascular lesions of unclear extent and lesions in which determination of carotid involvement is needed to plan treatment. When the carotid is involved, temporary balloon occlusion is a useful test. During the occlusion the stump pressure is measured and the patient is monitored clinically. In addition, xenon computerized or positron emission tomography may be used. Unfortunately, none of these methods is foolproof in predicting whether the patient can tolerate an en-block resection including the carotid. Superselective angiography with embolization of tumor feeding vessels is another technique that can facilitate the removal of lesions (such as angiofibromas and paragangliomas) by reducing intraoperative bleeding. Digital-subtraction angiography and other computerized enhancement techniques can produce images of higher resolution using lesser dye loads.

Neoplasms Involving Anterior Cranial Fossa

Even those tumors that actually arise from the ACF floor, such as olfactory neuroblastoma, rarely present with evidence of intracranial extension or cerebrospinal rhinorrhea. Involvement of surrounding areas, such as the orbit (proptosis) or the nasal cavity (obstruction or epistaxis), more often draws attention to the primary condition. Although biopsy of accessible tissue may confirm the histology, clinical examination combined with

radiologic assessment determines the macroscopic extent. The accuracy of these findings varies with each lesion, necessitating consideration of more common conditions occurring in the ACF.

Although neoplastic destruction of the ACF bone produces little pain, involvement of dura causes headache, either by distortion or inflammation. Infiltration of the related frontal lobe results in psychologic changes rather than variations in motor activity, as only the posterior precentral gyrus is concerned with voluntary movement (in the opposite side of the body).

Destruction of the olfactory nerves or distortion of olfactory bulbs eventually results in anosmia, although this may be preceded by parosmia, best defined as "distortion of the sense of smell" rather than diminution.

Benign neoplasms

A wide variety of benign tumors are found in the ACF that, although not metastasizing, may cause considerable disability by uncontrolled local growth. Because rate of growth is both variable and unpredictable, early diagnosis is important if permanent disability is to be avoided.

Osseous and fibroosseous conditions

Surgeons have recently realized that differentiation between the various fibroosseous lesions affecting the skull bones is only possible by correlating clinical features with radiologic and histologic findings. Before 1946, these lesions were classified somewhat arbitrarily on the relative amounts and character of bone and stroma present in the biopsy material. Consequently, osteoma, ossifying fibroma, and osteoblastoma might be grouped together despite markedly differing natural histories (Harrison, 1984).

Osteoma. Osteomas are benign bone lesions, characterized by bony excrescences, and usually arising in membranous bones. They are the most common fibroosseous lesions affecting the skull bones. Childrey (1939) noted an incidence of 0.42% in patients undergoing sinus radiography. The frontal sinus is probably the most common site of osteomas. Many are asymptomatic, although they may produce various clinical symptoms depending on site, size, and rate of growth. Osteomas may arise from the perpendicular plate of the ethmoid bone or involve the ACF floor by frontoethmoidal extension (Fig. 187-1) or via the posterior wall of the frontal sinus (Fig. 187-2). Hudolin et al (1961) reported a large frontal sinus osteoma that extended into the ACF and caused mental status changes, headache, incontinence, and sinusitis. Lautembach reported a female/male ratio of 3:1 with a median age in the 60s. On gross examination osteomas are well-circumscribed lesions with smooth or lobulated surfaces. Resorption of normal bone at the periphery of an osteoma may be seen, although the osteomas themselves are expansile, not infiltrative. Microscopically, osteomas are composed of varying amounts of cancellous bone, usually located in a central core and containing fatty marrow and osteoblastic activity, surrounded by a peripheral layer of compact bone. Radiologic appearance is highly variable and rarely shows the site of origin. Treatment of osteomas involves excision if the lesion is symptomatic. Complete removal is curative, although some patients, including those with Gardner's syndrome, may have synchronous or

metachronous lesions.

Ossifying fibroma (fibrous dysplasia). Some investigators have argued that ossifying fibroma and fibrous dysplasia are distinct entities. The nomenclature is confusing, with seemingly endless synonyms applying to these lesions. This discussion groups these lesions together and refers to them as *ossifying fibromas.*

Ossifying fibromas are benign bone lesions characterized by their clear margins, slow expansion, and tendency to occur in the mandible and maxilla, from which they can extend to the ACF floor (Fig. 187-3). These lesions are often asymptomatic, but may produce gross cosmetic deformity because of their huge size. Pain and paresthesias are among the most common symptoms. Because of jumbled nomenclature, reports of the epidemiology of this lesion are often contradictory. In general, women are affected more often than men, and the lesions tend to occur mostly in the third and fourth decades. Microscopic examination of these lesions reveals persistence of woven bone in the central core of the lesion, although mature, lamellar bone often covers the lesion. For this reason biopsy can be hazardous, especially in the frontoethmoid or cribriform plate areas. When diseases leads to obstruction of the nasofrontal duct, secondary frontal sinus mucocele may develop.

Radiographic features are variable. Sometimes patchy calcification dominates (Fig. 187-4). More frequently central calcification is seen in these lesions. In their early stages they have an osteolytic radiolucent appearance. Later a well-circumscribed radiopaque calcified center can be seen surrounded by a radiolucent periphery. In many cases clinical and radiographic findings are pathognomonic, although the appearance of the lesions may at times mimic osteogenic sarcoma.

Treatment, when indicated for cosmesis or symptomatic disease, includes careful radiologic assessment to determine the extent of the lesion followed by complete extirpation, if possible, after which recurrence rarely occurs. If complete extirpation is not possible, surgery is undertaken to minimize cosmetic and functional deformities. Malignant degeneration is rare but is occasionally seen after irradiation. Therefore, treatment by irradiation is not recommended.

Osteoblastoma. Osteoblastoma is a rare lesion composing only about 1% of all primary bone tumors. Only 15% of osteoblastomas are found in the head and neck, usually within the maxilla, although they may involve the ACF floor. The most common complaint in patients with these tumors is dull, aching pain of insidious onset. The avergae age at the time of diagnosis is 17 years, and the male/female ratio is 2:1. On gross examination the tumors tend to be hemorrhagic and friable. Microscopically they are characterized by abundant osteoid material and absence of cartilage. Radiographic feagtures are often not distinctive. Commonly lesions are well circuscribed and radiolucent. Removal of the lesion by curettage is the treatment of choice, although many patients gain many years of symptomatic relief from incomplete resection. Highly vascular lesions may be better treated with radiation than with surgery. Malignant degeneration has been noted both with and without radiation treatment although in both cases this development is rare.

Osseous hemangioma. Osseous hemangioma, also known as hemangioma of bone, is a slowly growing benign lesion composed of capillary, cavernous, or venous blood vessels. Although rare, these lesions are said to represent more than 70% of all skull hemangiomas (Glasscock et al, 1984). In the skull, the mandible and the maxilla are the two most common sites of origin. The vast majority of osseous hemangiomas are asymptomatic, although vague insidious pain may be reported. Their incidence increases with age, but symptomatic causes seem to present most often in young adults. In the skull they have a male/female ratio of about 2:1. On gross examination osseous hemangiomas are nonencapsulated, brownish-red lesions. They arise in the diploë and extend by erostion through the inner and outer tables. Bulging of the tables, especially the outer surface, may be seen. Microscopically, thin-walled vascular channels are seen interspersed among bony trabeculae. CT may be of limited use, as contrast enhancement can be variable. Surprisingly, digital subtraction studies show only minimal "blushing". Although they may erode the base of the ACF, the dura is not involved and radical removal is often possible.

Juvenile angiofibroma

Juvenila angiofibroma (JA) is a benign, highly vascular tumor that arises almost exclusively in adolescent males. A rare tumor, it accounts for only 0.5% of all tumors of the head and neck. The tumor almost always arises in the sphenopalatine foramen and follows a typical pattern of growth. After filling the vacant spaces in the nasopharynx, sphenoid sinus, and nasal passage, the tumor grows laterally into the pterygomaxillary and infratemporal fossae from which it can extend into the cheek or, via the inferior orbital fissure, to the orbit. Through bone erosion, some tumors reach the anterior or middle cranial fossa and extend into the cavernous sinus via the sphenoidal sinus (Fig. 187-5).

The most common presenting symptoms are nasal obstruction and recurrent epistaxis, which is sometimes severe. In younger boys the tumors tend to be more aggressive and the symptoms more severe. On gross examination the tumors are reddish, nodular, and covered by a tan mucous membrane. Ulcertation of the mucous membrane over the site of the tumor is common. On microscopic examination the tumor consists of a heterogenous mixture of thin-walled endothelium-lined vessels and moderately cellular collagenous stroma. Plane films demonstrate a soft tissue mass filling the nasal cavity. Pterygoid plate displacement (Fig. 187-6) or anterior bowing of the posterior wall of the maxillary sinus are highly suggestive of JA, although these findings also have been reported with other nasopharyngeal tumors. CT scan is invaluable in revealing bony erosion and the extent of the tumor. Angiography produces a pathognomonic image of multiple tortuous vessels in the arterial phase, followed by a dense blush in the capillary phase. Angiographic diagnosis can obviate the need for biopsy, which can be dangerous because it can cause severe hemorrhage. Most JAs take their initial blood supply from the ipsilateral internal maxillary artery, but they can acquire vasculature from other arteries as they grow. Sphenoidal and ophthalmic branches of the internal carotid and the ascending pharyngeal arteries are frequently involved. Bilateral carotid angiography can reveal these additional sources and preoperative embolization can decrease intraoperative blood loss. A number of staging systems for JA have been proposed (Chandler et al, 1984; Fisch, 1983; Sessions et al, 1981) but they are beyond the scope of this discussion.

Although involution occurs over time, the mainstay of treatment for JA is surgery aimed at thorough resection of the lesion. Radiation has been used successfully as a primary therapy and for recurrences and inoperable tumors. Disadvantages of radiation therapy include frequent treatment failures in some series, the potential for inducing malignancy, the theoretical risk of stunting facial growth, and cataract formation if the eyes are irradiated.

Meningioma

Meningiomas are slow-growing tumors that differentiate along the lines of meningial constituents, especially arachnoid villi. Symptomatic meningiomas constitute 14% of all intracranial tumors. The most common location of these tumors is intracranially in the parasagittal region, although they are found in numerous intracranial and spinal locations. Primary meningiomas involving the skull base from below are rare; in such cases the surgeon must search diligently for an intracranial lesion. This search should include a CT scan repeated every 1 or 2 years postoperatively, as a primary intracranial lesion may not be evident when the extracranial disease is diagnosed. True extracranial meningiomas have been reported in ectopic extracranial arachnoid granulations trapped outside skull fusion lines (Granich and Goodman, 1983), in the middle ear, and around cranial nerves. Although they are usually well circumscribed and seldom metastasize, they spread directly along the plane of least resistance and extend into bone along haversian canals rather than destroying bone by erosion. Intracranial meningiomas can enter the orbit via the supraorbital fissure, the nasal cavity via the cribriform plate, or the paranasal sinuses via the MCF floor.

Pain, cranial nerve involvement, and other signs are related to individual sites. Involvement of the ethmoid sinuses can cause nasal obstruction and a mass may be visible in the nose. Proptosis can result from orbital involvement. Cranial nerve symptoms such as trigeminal neuralgia may be present. Meningiomas are most frequently diagnosed in middle-aged women. In some reports the female/male ratio is as high as 9:1.

On gross examination these tumors are tough, white-gray, well-circumscribed masses. Microsopic appearance shows lobulate groups of cells reminiscent of normal arachnoid granulations. Syncytial and fibroblastic cells are prominent and psammoma bodies are often present. Histologic type has little clinical relevance except for the angioblastic, anaplastic, and papillary variants, which have poor cell differentiation and which exhibit varying degrees of malignant behavior. CT scan typically shows hyperdense, well-circumscribed soft tissue with uniformly dense contrast enhancement and areas of calcification. Meningiomas arising from intracranial sites are frequently associated with hyperostosis (Fig. 187-7). Bone destruction may be seen with extracranial extension. Venous digital subtraction angiography shows a vascular stain with blood supply from the meningeal vessels.

Treatment is by complete surgical removal with an excellent prognosis. Overall recurrence at 5 years after surgical excision is 15%. The less differentiated variants discussed previously are exceptions and outlook is less optimistic.

Other benign neoplasms

Other benign neoplasms, such as salivary adenomas, chondromas, and neurofibromas, involve the ACF by pressure erosion or attachment to the cribriform plate. Coronal CT is adequate to show the resulting bone loss; direct intracranial extension has not been seen in such cases.

Malignant neoplasms

Paranasal sinus

The intimate relationship between lateral ethmoidal block and cribriform plate (Fig. 187-8) ensures that ethmoidal malignancy soon involves the ACF floor. In addition, variations in both size and number of the ethmoidal cells may result in extension into the orbital roof and sphenoid bone.

At least 80% of all malignant tumors arising within the paranasal sinuses and nasal passages are squamous carcinomas, with the maxillary sinus the most common primary site. The absence of a real barrier between antrum and ethmoid, however, ensures that most antral tumors eventually involve the ethmoidal labyrinth and therefore the ACF. All show a soft tissue mass on routine radiographic examination, and at least 70% show some evidence of bone destruction. This applies to all malignancies in this area, irrespective of histologic type.

The cribriform plate is rarely more than 1 mm in thickness, with an average length of 21 mm. Its breadth increases evenly (anteriorly to posteriorly), varying from 4 to 5.4 mm, and it is perforated to allow the central processes of olfactory cells to pass into the nose. Normal channels, therefore, exist to allow a tumor to pass intracranially from the nose without the need for bone erosion (Fig. 187-9). Extension into the frontal sinus may lead to destruction of sinus walls, although this may be difficult to distinguish from a secondary sinus mucocele.

Bone

Malignant bone tumors are uncommon, affecting primarily men around the third decade of life. Pain is prominent and at diagnosis bone destruction is widespread. The radiologic appearances are variable, depending on the amount of bone destroyed. The "sun ray" appearance depends on preservation of cortex, with new bone growing perpendicularly to the surface, and is found in at most 25% of patients. Although most often starting in the maxilla, both chondrosarcoma and osteogenic sarcoma rapidly spread to involve adjoining sinuses, orbit, and skull base. The diagnosis is suggested by the patient's age and the extent of disease and is confirmed by biopsy. Radiology merely confirms the widespread extent.

The 5-year survival rate is 50% for chondrosarcoma and 25% for osteogenic sarcoma. Complete excision is the only chance for cure.

Olfactory neuroblastoma

Olfactory neuroblastoma is a neuroendocrine tumor arising in the nasal cavity. Neuroblastomas represent at most 2% of all malignant nasal neoplasms. The tumor is thought to arise from olfactory epithelium of neural crest origin in the upper portion of the nasal cavity. This location leads to early involvement of the cribriform plate. Preformed holes in this portion of the ethmoid complex allow rapid penetration into the ACF, even without radiographic evidence of bone erosion. Harrison (1984) reported universal involvement of the dura in his series, although evidence of gross bony erosion (Fig. 187-10) occurred in only half the cases. Enhancement on CT can help define intracranial involvement, although lack of enhancement does not preclude intracranial disease.

Symptoms are related to the sites of invasion and metastasis. Proptosis or impaired vision suggests intracranial extension and are prognosticators of low survival. Unlike sympathetic neuroblastomas, olfactory neuroblastomas rarely result in detectable urinary levels of vanillylmandelic acid (VMA) or homovanillic acid (HVA). The tumor has been reported in patients ranging from 3 to 90 years of age, with peaks of occurrence at 20 and 50 years. There is no predilection for either sex. Macroscopically, these tumors are lobulated soft red-gray masses. They frequently contain areas of necrosis and calcification. Microscopic examination may reveal nests of small cells separated by fibrovascular septae with neurofibrillary intercellular matrix and rosette formations. In some cases, however, only sheets of small round cells are seen, leading to misdiagnosis of olfactory neuroblastomas as anaplastic carcinomas or other tumors. Electron microscopy showing neurosecretory granules or special histochemical stains can help make the diagnosis when light microscopy is inconclusive. Kadish et al (1976) have proposed a staging system in which tumors confined to the nasal cavity are assigned to group A, those extending to the paranasal sinuses to group B, and all tumors extending beyond these limits to group C.

Current treatment protocols use various sequences of surgery, radiation, and chemotherapy depending on the extent of disease. Younger patients tend to have a lower incidence of local recurrence, a higher incidence of metastatic disease, and a more rapid demise. For patients with group C disease, prognosis is poor regardless of treatment.

Neoplasms Involving the Middle Cranial (Infratemporal) Fossa

Although convention divides the floor of the cranium into three separate fossae, this approach is primarily for convenience, as malignant tumors have no respect for anatomic boundaries. Most symptoms are produced by malignant or benign masses invading or distorting the cranial nerves leaving the MCF bone, and this involvement often produces clearly recognizable neurologic signs. Severe, unrelenting preauricular facial pain, sometimes misdiagnosed as trigeminal neuralgia or extracranial migraine, is a relatively common symptom of MCF lesions. Pain from bone or dural involvement is often difficult to control.

Benign neoplasms

Chordoma

Chordomas are invasive and destructive benign tumors that arise in residual or vestigial remnants of the embryonic notochord. They are rare, accounting for 0.2% of nasopharyngeal tumors and less than 1% of tumors of the central nervous system (CNS) (Mills, 1984). About one third arise in the region of the sphenooccipital synchondrosis, producing headache and progressive cranial nerve palsies.

Diplopia from involvement of CN VI is the most common presenting symptmo and is reported by 90% of patients. Visual field cuts and nasal obstruction are frequently seen. Although occasionally appearing as primary nasopharyngeal tumors, most extent downward from the clivus from which they can erode into the sphenoid sinus. Chordomas are found slightly more often in men than women. The average age at presentation is in the third decade. Under gross examination, chordomas are lobulated, mucoid, and partially translucent. Microscopic appearance is variable. Stellate and physaliphorous cells as well as intracellular mucinous matrix may be seen. Hyaline cartilage may be totally absent or may be the predominant element of the tumor. This chondroid variant is seen more frequently in women and occurs in a slightly younger population of patients. Radiographically an expansile osteolytic mass at the clivus is seen on plain radiographs. Calcifications, especially with the chondroid type, may be seen. Angiography demonstrates an avascular mass with superior and posterior displacement of the basilar artery. CT scan can delineate the tumor's superior extent and its extension into the nasopharynx and prevertebral space. MRI may prove useful. As there are not pathognomonic tests for chordoma, biopsy before surgery is necessary and can usually be obtained transorally.

Treatment consists of surgical excision. Postoperative radiation is used as an adjunct, although these tumors tend to be highly radiation resistant. High-energy proton therapy may be more useful for palliation. With less than total excision, recurrence is universal; unfortunately, this outcome is the rule rather than the exception. Overall mean survival is about 7 years but depends highly on histologic type. In one study the average survival of patients with chondroid chordomas was 15.8 years, whereas mean survival for those with typical, nonchondroid tumors was only 4.1 years (Heffelfinger et al, 1973).

Meningioma

A discussion of meningiomas of the MCF is included in the section on benign neoplasias of the ACF (p. 3265).

Schwannoma ("neuroma") and neurofibroma

Neuroam is the common name for tumors that more properly should be called schwannomas, as they differentiate along the lines of Schwann's cells, not nerve cells. Neurofibromas are a second type of tumor that differentiate along Schwann's cell lines. These tumors differ in their microscopic pathology, epidemiology, and malignant potential. Schwannomas and neurofibromas together compose about 6% of all intracranial tumors, and the vast majority of them are found in the posterior cranial fossa (see Chapter 188). These

tumors include the acoustic neuroma and those associated with the jugular foramen.

In the MCF these lesions arise on the mandibular or maxillary division of CN V and are usually associated with generalized neurofibromatosis (von Recklinghausen's disease). With involvement of CN V hypesthesia of the face in the distribution of the affected nerve division is the most common symptom. Schwannomas usually present in adults in their fifth or sixth decades, whereas neurofibromatosis may affect young children. On gross appearance both tumor types are white to gray, encapsulated, and usually firm. Vascularity is variable and they may contain cystic spaces. Microscopically schwannomas are characterized by areas of high and low cellularity called Antoni A and B areas, respectively. In Antoni A areas palisaded nuclei called Verocay bodies may be seen. Neurofibromas lack these features and ara characterized by interlacing bands of spindle cells. Malignant transformation can occur with either schwannoma or neurofibroma, but is much less frequent with the former.

MRI and CT are complementary in evaluating these lesions. The soft tissue component of the tumor is best visualized on MRI, whereas CT is superior in demonstrating enlargement of the involved bony foramina. Smooth enlargement of the foramen rotundum may be seen if the maxillary division of CN V is involved; enlargement of the foramen ovale may be seen with involvement of the mandibular division. On contrast CT, enhancement is variable, although any intracranial component shows a well-circumscribed lesion despite poor vascularization (Fig. 187-11). Identification of a soft tissue component allows differentiation from other causes of smooth enlargement such as congenital variants.

Treatment consists of surgical excision of the entire lesion. Function of the involved cranial nerve is often lost. Complete surgical excision constitutes an effective cure. Unfortunately, persons with MCF lesions often have generalized neurofibromatosis in which synchronous and metachronous lesions are common.

Paraganglioma ("glomus tumor")

Paragangliomas are rare tumors that arise from the chemoreceptor systems located in the carotid, vagal, and jugulotympanic bodies. Like schwannomas, paragangliomas most frequently involve the posterior cranial fossa (see Chapter 188). However, they may extend to the MCF and so are mentioned here for completeness. Commonly referred to as glomus tumors, paragangliomas have little in common with true glomus tumors, which are neoplasms of the neurovascular shunting apparatus and arise most frequently under the fingernails and toenails. That true glomus tumors can occasionally occur in the head and neck only adds to the confusion.

Initial symptoms are often pulsatile tinnitus progressing to conductive hearing loss. As they slowly grow, paragangliomas often involve the structures passing through the jugular foramen in the posterior cranial fossa. In the MCF they may extend to the internal carotid artery. Women with paragangliomas outnumber men 3:1. The tumor occurs most frequently in persons between the ages of 30 and 60 years. On gross appearance they are firm, tan-red, and usually encapsulated. Despite this encapsulation they are often densely adherent to adjacent vessels and difficult to excise. Paragangliomas are usually composed microscopically of well-differentiated epithelioid cells that contain neurosecretory granules. Thus paragangliomas belong to the broader class of apudomas and some secrete vasoactive

catecholamines. Routine screening for urinary catecholamines is suggested for all patients suspected of having paragangliomas. Schwaber et al (1984) recommend selective venous catheterization studies when screening values are elevated, there is a family history of similar tumor, or multiple tumors are suspected. Venous sampling can be used to determine tumor location. With larger tumors CT scan reveals irregular destruction of bone at the jugular foramen and MRI may show stasis in the sigmoid sinus. Carotid and vertebral arteriography can determine if these vessels send branches to the tumor. Retrograde jugular venography can be dangerous and is usually not indicated as CT, MRI, and the venous phase of arteriography can be used to determine involvement of the jugular system.

Treatment of these tumors is by surgical excision. Preoperative embolization at the time of angiography can reduce intraoperative blood loss but runs a definite risk of causing permanent paralysis and other effects related to CNS infarction. Recurrence rates range from 10% for tumors of the carotid body to 60% for those arising elsewhere. Malignant degeneration with widespread metastasis leading to death is seen in about 10% of patients with paragangliomas.

Epidermoid cyst

Epidermoid cysts are identical histologically to congenital cholesteatoma. They arise from epithelial remnants trapped during fetal development and become manifest within the bones of the skull and the spinal canal. The petrous portion of the temporal bone is the most common site for intracranial epidermoid cysts; at least 40% are located there.

The lesions may be extensive despite minimal neurologic symptoms. When symptoms do occur they include slowly progressive dysfunction of CN VII and CN VIII and are manifested as facial tic or paresis, sensorineural hearing loss, and imbalance with diminution or loss of caloric response. Pathologically lesions consist of a bone-eroding cavity lined with skin and filled with desquamative squamous epithelium. Plane films reveal a sharply defined lucency with smooth margins, which has been said to resemble a large trephine opening (Glasscock et al, 1990). On CT scan they appear as homogeneous areas of low density in contrast to the high density surrounding bone they have eroded. Calcification occasionally occurs in the capsule; this can also be seen in meningiomas (Mafee et al, 1984). When the capsule is thick, a ring of contrast enhancement may be seen. The contents of the cyst are avascular keratin debris and do not enhance.

Treatment consists of surgical excision of the lesion within its capsule, which constitutes a cure. Failure to remove all capsular remnants leads to recurrence. Because of the frequency of recurrence after removal of extensive disease, diligent follow-up is necessary and second-look operations have been advocated.

Malignant neoplasms

Nasopharyngeal malignancies (nasopharyngeal carcinoma)

The term *nasopharyngeal malignancies* refers to all cancers arising in the nasopharynx. The term *nasopharyngeal carcinoma (NPC)* is confusing, as it has been used historically to imply a variety of histologically different lesions, some of which are not even carcinomas. The World Health Organization restricts the term NPC to those carcinomas that arise from the surface epithelium of the nasopharynx (International Histological Classification of Tumors, 1978), and this is the definition used in this discussion. Simply stated, NPC is squamous cell carcinoma of the nasopharynx. Almost all malignant tumors arising in the nasopharynx are NPC. Rarely malignant lymphoma, salivary gland malignancies, and sarcomas are also seen.

The nasopharynx lies primarily beneath the body of the sphenoid bone. It is adjacent to the foramen ovale, the foramen spinosum, the carotid canal, the jugular foramen, and the hypoglossal canal. Nasopharyngeal malignancies commonly spread into the MCF either through the foramen lacerum and carotid canal or through the foramen ovale. Having arrived in the MCF via these routes, they are close to the cavernous sinus and thus CN II, III, IV, and VI.

NPC is uncommon among whites, representing no more than 0.5% of all malignancies; but it has a high incidence in other ethnic groups including souther Chinese, Maltese, and Kenyans. Males are affected more often than females and peak incidence occurs in the fourth and fifth decades. In high-risk groups, early diagnosis is helped by recognition of the significance of nasal obstruction and epistaxis. Unfortunately, in more than half of all cases the presenting sign is a painless lump in the neck, representing metastatic disease. Metastases to the cervical nodes are found in up to 70% of patients (Lederman, 1961).

On gross examination NPC appears as the ulcerating fungating growth typical of squamous cell carcinomas elsewhere. The microscopic appearance of NPC ranges from undifferentiated carcinoma to well-differentiated squamous cell carcinoma.

Lateral radiographs of the nasopharynx may reveal a soft tissue mass, but when large enough to be detected by this means, the tumor has usually invaded the cranial cavity and orbit. In addition to osteolytic bone changes, neoplastic invasion may provoke a sclerotic reaction in the basisphenoid and floor of the sphenoid sinus. CT scan demonstrates the soft tissue mass as well as the bone destruction. Enhancement by intravenous contrast is often greatest for the intracranial part of the lesion.

Treatment and prognosis depend on the stage at presentation. Extension to the skull base or cranial nerve involvement places patients in the T4 classification (Wei, 1984). Because these tumors often present at a late stage in which they are not resectable, radiation has been the mainstay of treatment. The possibility of surgery to the clivus and parapharyngeal space (Chapter 191) emphasizes the importance of pretreatment assessment. With recent advances in radiation treatment overall 5-year survival has been about 50%.

Orbital rhabdomyosarcoma

Rhabdomyosarcomas are malignant tumors having muscular differentiation. They account for about 4% of all malignancies in children under 15 years of age, and are the most common soft tissue malignancies of the head and neck in this age group. About one third of all head and neck rhabdomyosarcomas arise in the orbit from which they freqently extend posteriorly into the MCF. The principal finding is rapidly progressive unilateral proptosis. The peak incidence of rhabdomyosarcoma is around 5 years of age, and there is a slight predominance of males. White children are affected three times more often than those of other races. The term *rhabdomyosarcoma* refers to a histologically heterogeneous group of tumors. The embryonal variety accounts for the majority in children and consists of densely packed lymphocyte-sized cells with hyperchromatic nuclei and stellate or bipolar cell processes.

CT demonstrates an enhancing, noncalcified mass. Invasion and destruction of adjacent structures including direct extension into the MCF are common. The diagnosis is made by biopsy. Chest CT, bone scan, liver scan, bone marrow aspirate and CSF cytology may all be included in the search for metastatic disease. Orbital rhabdomyosarcoma has a more favorable prognosis than when the same tumor is found in other locations, perhaps because there are few lymphatics in the orbit and because the confined space of the orbit leads to proptosis and relatively early diagnosis.

Treatment consists of biopsy and, in some cases, resection or debulking if this can be accomplished without sacrifice of vital structures. Surgery is followed by multiple agent chemotherapy and radiation. This combined therapy has produced cure rates of up to 80% (Maurer et al, 1988). Recurrences, however, are almost universally fatal (Anderson et al, 1990).

Metastatic neoplasm

Most metastatic lesions involving the skull base develop from carcinomas of the prostate, lung, and breast. Prostatic carcinomas produce hyperostotic bone, whereas the other two show lytic changes, although it is doubtful if radiologic changes are specific enough to justify more than suspected metastasis. In many instances skull base metastases are only part of more widespread disease. A solitary metastasis from a previously unsuspected renal carcinoma is not uncommon, however, and may carry a more favorable prognosis than metastases from most other tumors. In inaccessible sites such as the infratemporal fossa, Shapsay et al (1979) suggest percutaneous core needle biopsy for patients with occult tumor and severe facial pain. This technique is recommended only after a complete clinical and radiologic evaluation, and then only under radiographic monitoring. In most patients, age and clinical history suggest to the surgeon that the skull base lesion may have a metastatic origin.