Chapter 192: Surgery of the Temporal Bone Neoplasms

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Both benign and malignant lesions involving the middle ear and temporal bone occur in one of the most inaccessible areas of the body. Although a great variety of histologic types of tumor are found in these areas, each is uncommon and clinical experience in their diagnosis and management is thus not easily acquired. Tumors of the middle ear cleft frequently appear early with symptoms of fullness or conductive hearing loss; but massive occult erosion of the petrous apex by tumors of the temporal bone can occur, which is caused by the relative resistance of the bony labyrinth and cranial nerves to erosion by the frequently benign lesions in this area.

Surgical approaches to the skull base are difficult, and the risks of neurologic deficit are significant. The major arteries and veins supplying the brain, intracranial structures, and all the cranial nerves either exit through or are contiguous with the temporal bone. Surgical techniques and approaches have evolved to improve access to lesions of the skull base while preserving residual hearing and avoiding neurologic deficit. Contemporary surgical technique strongly stresses the minimization of further cosmetic or neurologic loss in the treatment of both benign and malignant lesions of the middle ear space and temporal bone. Through the years collaborative approaches to the temporal bone have been developed for benign lesions and are currently being expanded to encompass malignant lesions as well. Nevertheless, considerable knowledge of temporal bone anatomy and temporal bone-drilling laboratory experience are essential in adapting these approaches to the individual case. Surgery has been made more efficient by the use of the operating microscope and the otologic drill with constant water-suction irrigation.

Lesions may be divided into those that arise primarily within the middle ear and mastoid or temporal bone, those that secondarily involve these areas from contiguous neoplastic processes, and those that arise from metastases or systemic involvement from distant sites. Tumors can also be divided into both benign and malignant categories. Many histologic types are rarely encountered, accounting for isolated case reports in the literature. The following is an outline of masses of the middle ear and temporal bone:

I. Primary

    A. Benign

        1. Congenital cholesteatoma
        2. Glomus tympanicum
        3. Facial nerve schwannoma
        4. Adenoma
        5. Intratympanic meningioma
        6. Hemangioma
        7. Glioma
        8. Osteoma
        9. Dermoid cyst
        10. Choristoma (hamartoma)
        11. Cholesterol granuloma
        12. Giant cholesterol cyst
        13. Fibrous dysplasia
        14. Encephalocele
        15. Vascular anomalies

    B. Malignant

        1. Squamous cell carcinoma
        2. Rhabdomyosarcoma
        3. Carcinoma (squamous, adeno-, acinic)
        4. Mesenchymal tumors (chondroma, chondrosarcoma, osteoclastoma)

II. Secondary

    A. Nasopharyngeal carcinoma
    B. Chordoma
    C. Vestibular schwannoma
    D. Schwannoma of cranial nerve IX, X, XI, XII
    E. Meningioma
    F. Sphenoid mucocele
    G. Aneurysm of internal carotid artery

III. Metastatic

    A. Breast carcinoma
    B. Prostatic carcinoma
    C. Renal cell sarcoma
    D. Bronchogenic carcinoma
    E. Lymphoma
    F. Histiocytosis X.

Symptomatology

Middle ear space

Neoplasms of the middle ear space most frequently appear with the insidious onset of conductive hearing loss and aural fullness, although they may also be discovered as an incidental finding on a routine physical examination. Occasionally, lesions appear as recurrent otitis media or refractory otitis media and may fistulize through the tympanic membrane. Some lesions have classic presentations. The glomys tympanicum tumor presents as a reddish pulsatile mass behind the tympanic membrane and frequently is associated with pulsatile tinnitus (Brown, 1953). A facial nerve schwannoma may present with a sudden, a slowly progressive, or a fluctuating facial nerve paralysis and can appear as a gray-white polypoid mass behind the posterior superior tympanic membrane (Pulec, 1972). Dermoid cysts, choristomas, and vascular anomalies may present with malformed or eroded ossicles.

Patients with malignant neoplasms involving the middle ear space and mastoid may have otorrhea, bleeding, pain, swelling, and decreased hearing (Batsakis, 1979a). Although many authors have emphasized the importance of pain as a symptom of carcinoma, this finding is not consistent. Early diagnosis is frequently difficult because the tumor is concealed, but with growth and bony destruction the malignancy may become apparent as it erodes into the external auditory canal or through the mastoid cortex into the skin. Sensorineural hearing loss, vertigo, and facial nerve paralysis may exist in some cases. Further cranial nerve deficits occur as the tumor destroys the skull base. With carcinomas, regional lymphatic spread may occur easily, but distant metastases generally occur late in the course of the disease.

As a general rule, sarcomas occur in the younger age group and carcinoams in the elderly. Embryonal rhabdomyosarcoma is the most frequent middle ear malignant neoplasm in the young, although histiocytosis X also occurs frequently. Again, certain histologic types have a classic presentation. Adenoid cystic carcinomas have pain as an early symptom. Squamous cell carcinomas usually start in an ear with previous otologic disease, including chronic suppurative otitis media, a mastoid cavity, or chronic dermatoses. Adenocystic carcinomas and a wide variety of other carcinomas, including basal cell carcinoma, melanoma, mucoepidermoid carcinoma, and ceruminous gland adenocarcinoma, have been reported (Pulec, 1977).

Temporal bone

Gacek (1975) describes a progression of symptoms as a lesion expands from the petrous apex to involve temporal bone structures. Clinical features of petrous apex lesions are listed as follows:

Early symptoms
        Headache
        Abducens paresis (CN VI)
        Sensory disturbance, trigeminal nerve V (CN V)
        Middle ear effusion or otorrhea
        Sensorineural hearing loss
        Recurrent meningitis
Posterior extension symptoms
        Facial nerve palsy
        Jugular foramen syndrome (CN IX, X, and XI paresis)
        Hypoglossal paralysis
Anterior extension symptoms
        Diplopia and ophthalmoplegia (CN III, IV, or VI).

At an early stage, traction on the dura of the middle fossa gives rise to parietal or vertex headache. Involvement of the mandibular branch of the trigeminal nerve produces anesthesia or paresthesia over its distribution. Conductive deafness may result from eustachian tube compression and middle ear effusion, or sensorineural deafness may result from labyrinthine involvement. Vertigo may occur, but syncope suggests occlusion of the internal carotid artery as it courses lateral to the petrous apex. Otorrhea may result from invasion of the external canal, and rupture and discharge of the lesion's contents internally into the subarachnoid space may produce aseptic meningitis. As in the middle ear space, facial nerve symptomatology may be sudden in onset, slowly progressive, or fluctuating and recurrent in nature.

Anterior extension of tumors within the temporal bone toward the cavernous sinus produces cranial neuropathies, which result in ophthalmoplegia and diplopia. Posterior spread may involve the contents of the internal auditory canal and jugular foramen. The most extensive lesions ultimately produce a visible mass in the nasopharynx, middle ear, or neck. Solid tumors compress cranial nerves and produce neuropathies more often than the cystic lesions, and malignant tumors more often than benign tumors (Flood and Kemink, 1984).

Diagnostic Evaluation

Accurate diagnosis and evaluation of the extent of tumors of the middle ear space and temporal bone require appropriate physical examination and audiologic, vestibular, and radiologic assessment. Tumors of the middle ear space can frequently be seen on physical examination. The combination of the frequency of certain lesions and their classic presentations can often suggest the tumor type. Tumors of the temporal bone frequently cannot be seen by physical examination, but neurologic deficit caused by the tumor can be detected by physical examination and audiometric and vestibular testing.

When a mass is suspected, radiologic evaluation is needed to confirm the suspicion and delineate the extent of disease. Although plain films can be helpful with large lesions, they seldom provide enough information to perform a thorough imaging evaluation. Complex motion tomography can provide fine bony detail, but very little information about soft tissue is available. Accordingly, such tomography has become nearly obsolete.

Overall, the single most useful imaging evaluation of the temporal bone is provided by computed tomography (CT). Modern CT affords exquisite bony detail as well as fairly good soft tissue information. CT now routinely provides submillimeter resolution at bone-to-soft tissue or bone-to-air interfaces. Soft-tissue-to-soft-tissue discrimination is still limited, due to the very narrow range of electron densities in soft tissues of interest. For example, the entire bony labyrinth can be resolved with precision, but no details of the membranous labyrinth within can be resolved even on the best CT. This limitation does not usually significantly restrict CT evaluation of temporal bone masses as most of these lesions remodel or destroy bone. More soft tissue discrimination is available with CT if iodinated contrast material is administered intravenously. However, such contrast material brings with it the significant risk of idiosyncratic contrast reaction. Unfortunately, CT delivers ionizing radiation at a relatively high dose compared to plain films.

The advent of magnetic resonance imaging (MRI) and its superb soft tissue sensitivity has resulted in fewer indications for iodinated contrast material in CT. Although MRI is able to evaluate alterations in bony structure, it does so only indirectly because cortical bone has few mobile water protons and, hence, nearly no intrinsic signal. For this reason, MRI is inferior to CT in the evaluation of subtle bony changes. Such subtle changes are important findings in temporal bone masses. However, MRI has better soft tissue discrimination than CT. To continue on a previous example, the bony labyrinth can obly be indirectly delineated on MRI by revealing the membranous labyrinth within it. MRI has just begun to resolve individual components of the membranous labyrinth. MRI requires no ionizing radiation. Instead, it uses magnetic fields and radio frequencies to produce images.

The fine soft tissue discrimination of MRI is improved by the intravenous infusion of MRI contrast. Only one MRI contrast agent, gadolinium, is presently approved by the Food and Drug Administration. The incidence of idiosyncratic reactions to this material is dramatically lower, and the rare reaction less severe, than that of the iodinated contrast material used for CT. MRI contrast material is visible at a tissue concentration up to 100 times lower than that of CT contrast material. An example of this is the common visualization of the arteriovenous plexus around the fallopian portion of the facial nerve. This plexus is not visible on CT. As one can conclude, this makes MRI far more sensitive in the evaluation of pathologic contrast enhancement. Practical proof of this superiority is the now already well-known superiority of MRI over CT in the evaluation of vestibulocochlear schwannomas. Another advantage of MRI is its ability to display numerous imaging planes without repositioning the patient, which makes it far easier to acquire coronal, sagittal, and even off-axis oblique images. For these reasons, MRI is superior to CT in defining brain or meningeal effects from temporal bone lesions, including the evaluation of all types of meningoceles as well as direct infiltration of temporal bone masses into the meninges or brain. Unfortunately, MRI is generally more expensive than CT and takes more time to perform. Due to potentially dangerous magnetic effects, MRI cannot be used in patients with certain cardiac pacemakers or valves, ferromagnetic cerebral aneurysm clips, some vena cava filters, and ferromagnetic intraocular foreign bodies.

Accordingly, the usual evaluation of a potential temporal bone mass begins with a noncontrast temporal bone CT. This approach will immediately show large lesions and usually suggest the presence of smaller lesions. Even if this temporal bone CT is negative, MRI might follow if there are strong enough clinical findings to suggest a mass. Like CT, MRI is focused

to the region of interest, instead of just providing thick slices of the entire head. This technique provides greater detail. Temporal bone MRI is almost always performed without and with MRI contrast material. Many practitioners will bypass CT if they feel that the disease is retrocochlear. CT can then be added after the MRI, if precise bone detail is required.

Both CT and MRI are computerized imaging techniques. For this reason, either can be reformatted in any desired digital display. A popular type of reformatted display is a three-dimensional (3-D) image. With CT, this is most commonly an attenuation threshold display where 3-D bone images are provided. Such displays are beautiful to look at and can mimic temporal bone specimens. MRI can be performed in a similar way, but is usually reformatted to show soft tissue. Edge-based algorithms for 3-D reformatting with improved soft tissue discrimination are more complicated and expensive. Regardless of the type, 3-D displays are not required in temporal bone imaging. 3-D displays can assist the surgeon by making it easier to understand the extent of the lesion, but they almost never provide diagnostic information that was not available on the standard axial or coronal slices. Moreover, 3-D displays are heavily postprocessed. Significant image information can be lost in such processing. At the present time, 3-D images are used almost exclusively as supplemental displays as dictated by the tase of a particular practitioner.

Surgical Approach

Small lesions in the middle ear space whose borders are clearly visible through the tympanic membrane can be removed via the external auditory canal. When removing the tumor, the surgeon must clearly differentiate it from vascular anomaly, since injury to the internal carotid artery (ICA) in its intratemporal portion is hazardous and may require carotid artery ligation for control. Tumors whose borders are not visible through the tympanic membrane may be approached through a combined transcanal and facial recess or extended facial recess approach. Occasionally the facial recess is narrow, requiring removal of all or part of the bony posterior external auditory canal wall.

Surgical approaches to lesions deper within the temporal bone must be chosen either to provide adequate exposure for complete excision or to allow easy access to an exteriorized cavity, preserve useful residual hearing if possible, preserve facial nerve and other cranial nerve functions when possible, avoid injury to the brainstem and ICA, and provide for wound closure without cerebrospinal fluid (CSF) leak.

Approaches commonly used include the following:

1. Mastoidectomy
    a. With or without extended facial recess
    b. With or without facial nerve mobilization
2. Radical mastoidectomy
    a. With or without grafting of the middle ear space
3. Lateral temporal bone resection
4. Middle cranial fossa
5. Translabyrinthine
6. Transcochlear
7. Retrolabyrinthine
8. Suboccipital
9. Infratemporal fossa
10. Transsphenoid
11. Total en bloc resection of the temporal bone
12. Various combinations or extensions of these approaches.

Surgical defects can be reconstructed by soft tissue grafts, bone pate, local or regional flaps, or obliteration with abdominal fat.

Mastoidectomy approach

Mastoidectomy with or without a facial recess or an extended facial recess approach is useful for the removal of more extensive lesions of the mastoid and middle ear space. Lesions involving or medial to the facial nerve may require facial nerve grafting or mobilization of the facial nerve for complete exposure and removal. A radical mastoidectomy with or without grafting of the middle ear space can more adequately exteriorize this area. Lateral temporal bone resection can be considered for lesions confined to the external auditory canal or for debulking of large lesions of this area.

Middle cranial fossa approach

In 1959 House used the middle cranial fossa route to expose the internal auditory meatus and search for an otosclerotic focus. His subsequent work established this approach for section of the vestibular nerve, small acoustic tumor removal with preservation of hearing, and intralabyrinthine facial nerve surgery (House, 1963; House al, 1968). Access by this approach is limited, but hearing can be preserved. Currently the middle fossa approach has been used for various tumor removals and exteriorization procedures. This approach has also been used in the treatment of congenital cholesteatomas, repair of spontaneous encephaloceles of the temporal bone, and exteriorization of giant cholesterol cysts of the temporal bone.

Translabyrinthine and transcochlear approaches

The translabyrinthine and transcochlear approaches to the temporal bone as proposed by House (1968) and House and Hitselberger (1976), respectively, sacrifice redisual auditory and vestibular function, but provide excellent exposure to the petrous apex. A complete mastoidectomy with labyrinthectomy is performed, and the facial nerve is mobilized from the

stylomastoid foramen to the internal auditory meatus and rerouted posteriorly. Exenteration of the cochlea allows anterior exposure to the ICA. With this technique the apex, clivus, basilar and vertebral arteries, CN VI to CN XII, and even the opposite internal auditory canal meatus can be visualized. In surgery for apical meningiomas and cholesteatomas, the facial nerve is encountered lateral and posterior to the tumor mass, rather than anteriorly, as in acoustic neuroma surgery. If the facial nerve is sacrificed, end-to-end anastomosis is recommended because mobilization of the facial nerve may gain 1 to 2 cm of length. Tumor access is provided by bone removal rather than retraction of the cerebellum and brainstem. Furthermore, control of the ICA is generally adequate with this approach. The more limited translabyrinthine approach is a standard in acoustic neuroma surgery (Glasscock and Hayes, 1973). It provides adequate access to the posterior temporal bone and the facial nerve in its entire intratemporal course. However, residual labyrinthine function is also sacrificed by this procedure.

Retrolabyrinthine and suboccipital approaches

The retrolabyrinthine and suboccipital (posterior craniotomy) approaches also give limited access to the posterior temporal bone. A suboccipital craniotomy may require more extensive retraction or excision of the lateral third of the cerebellum. This surgical exposure is posterior to the internal auditory meatus, and access to the medial petrous apex and clivus is frequently impossible. The potential advantages of preservation of residual auditory and vestibular function are, however, significant.

Infratemporal fossa approach

Fisch (1979) has excised lesions of the skull base and petrous apex previously considered unresectable through the infratemporal fossa approach. This route is particularly applicable to lesions of the jugular bulb, including glomus jugulare or jugular foramen neuromas. This procedure may be extended by excision or displacement of the mandibular condyle and by mobilization of the zygoma and lateral orbital rim to gain further access to the infratemporal fossa. A subtotal petrosectomy anterior to the labyrinth exposes the entire intrapetrous course of the ICA. The facial nerve is transposed anteriorly for improved access. The petrous apex may be obliterated, the eustachian tube packed, and the external auditory canal closed laterally. A conductive hearing loss, therefore, results. This approach has been reported in cases involving glomus tumors extending to the petrous apex, meningiomas involving the sphenoid and petrous apex, chordomas of the clivus, congenital cholesteatomas, carcinomas with extensions from the nasopharynx or infratemporal fossa, and ICA aneurysms.

Transsphenoid approach

Montgomery (19770 described the transsphenoid approach to the petrous apex with preservation of cochlear function. Cystic lesions may be exteriorized and drained through the adjacent sphenoid sinus. Exposure may be obtained via an external ethmoidectomy. The patency of the fistula is maintained with a pedicled septal flap and an indwelling Silastic silicone drainage tube. The possibility of ascending infection and limited postoperative access for cavity cleaning still exists.

Total en bloc resection

Campbell et al (1951) introduced total en bloc resection of the temporal bone to the literature. Because of poor surgical results and the difficulties associated with such extensive combined intracranial and extracranial surgery, many otolaryngologist - head and neck surgeons still use partial resection of malignant tumors of the mastoid and temporal bone followed by radiation therapy. The anatomic limitations of the carotid artery have hampered the removal of most en bloc procedures. Graham et al (1984) reported two patients who underwent total en bloc resection of the temporal bone and carotid artery for malignant tumors. More recently, however, surgeons have become more aggressive and resected and grafted the intratemporal carotid artery.

Structures at Risk

To minimize surgical complications and postoperative neurologic deficit, one needs a full understanding of the structure within or adjacent to the temporal bone.

Auditory system

The ossicles within the middle ear space or epitympanum may be easily dislocated or traumatized during removal of lesions within these areas. Should the drill contact an intact and mobile ossicular chain, the vibrations may result in a significant and irreversible sensorineural hearing loss. Similarly, serious loss of cochlear function may result after entry into the perilymphatic spaces of the cochlea by instruments or during removal of areas of bone erosion and fistulization.

Vestibular system

As with the cochlea, tumor or surgical invasion of the semicircular canals or vestibule may produce not only disturbances of balance but also serious sensorineural hearing loss (Fig. 192-1).

Jugular bulb

As Graham (1977) emphasized, the jugular bulb may be at risk in any surgical procedure involving the temporal bone. The jugular bulb may ascend high and medial to the facial nerve, to the level of the posterior semicircular canal or the internal auditory canal. Should this structure be injured during surgery, profuse venous bleeding is inevitable. If bleeding is not readily controllable by extraluminal packing, proximal extradural packing of the sigmoid sinus and distal ligation of the internal jugular vein are necessary. Constant vigilane and use of the diamond drill and suction irrigation when in close proximity to the jugular bulb usually prevent this serious complication.

Sigmoid sinus

Constant awareness of normal variation in the location of the sigmoid sinus ensures against inadvertent damage to this structure. The sigmoid sinus may lie either superficial or deep, or anteriorly or posteriorly within the mastoid cavity. With constant suction irrigation and awareness of these variations, one can usually avoid injury to this structure even while skeletonizing it to allow for deeper entry into the mastoid segment. Should bleeding occur, management is identical with that previously described for control of jugular bulb bleeding. Bleeding associated with small accessory veins from the sigmoid sinus frequently responds to bipolar cautery or packing.

Internal carotid artery

The ICA is at considerable risk in more extensive procedures on the temporal bone. With removal of lesions adjacent to the ICA, the surgeon must recognize that anterior repair within the temporal bone is frought with difficulty due to the lack of substantial adventitial lining of the intratemporal carotid artery and to the difficulty of adequate exposure. Although experience has recently been gained with bypassing the intratemporal portion of the ICA, in most cases substantial injury to the carotid artery within its intratemporal portion requires carotid artery ligation, with the potential risk of devastating neurologic deficit.

Dura

The middle and posterior fossa dura surround the superior and posteromedial aspect of the temporal bone. Even with the use of constant suction irrigation, the cutting drill can tear the dura and may result in CSF fluid leakage or prolapse or herniation of the cerebellum or temporal lobe. Care must be exercised, therefore, in approaching the dura, and injury can usually be avoided with use of the diamond drill and constant suction irrigation. Leakage of CSF may be controlled by direct repair (with suturing of fascial grafts) or by obliteration of the mastoid-epitympanic defect.

Facial nerve

The facial nerve is also at risk in virtually all the surgical approaches for removal of lesions of the middle ear space and temporal bone. During transcanal, transtympanic, or transmastoid approaches, it is at risk in its vertical or horizontal portion. With the middle cranial fossa approach, the facial nerve is at risk during elevation of the dura (if the greater superficial petrosal nerve is tractioned) or when the bone overlying the geniculate ganglion or facial nerve is dehiscent. In some cases the facial nerve must be either transposed or sacrificed. Anterior transposition of the facial nerve from the cochleariform process out into the neck can be accomplished without paralysis or paresis with careful technique. More extensive mobilizations frequently result in facial nerve paralysis with late recovery of facial nerve function. When the facial nerve is sacrificed, all reasonable attempts should be extended to achieve reanastomosis or grafting. Long cable grafts give surprisingly good functional results that are far superior to many other reanimation techniques.

Glomus Tumors of the Temporal Bone

The glomus tumor of the temporal bone is the most frequently encountered neoplasm after the acoustic neuroma. Alford and Guilford (1962) thoroughly described the signs, symptoms, and classification of glomus tumors of the temporal bone. They divided glomus tumors into two categories, based on their site of origin: the glomus tympanicum and glomus jugulare.

The glomus tympanicum tumor most frequently appears with the insidious onset of pulsatile tinnitus and a conductive hearing loss, although it may also be discovered as an incidental finding on routine physical examination (House and Glasscock, 1968). The glomus jugulare tumor arises within the dome of the jugular bulb and commonly appears late, after considerable growth and bony destruction. It may cause a neurologic defect in the function of cranial nerves passing through the jugular bulb (CN IX to CN XII), facial nerve paresis caused by tumor extension into the mastoid, or sensorineural hearing loss caused by bony erosion of the labyrinth. Occasionally either a glomus tympanicum or a glomus jugulare tumor may erode the tympanic membrane and appear as a bleeding mass in the external auditory canal.

Diagnostic evaluation

Accurate diagnosis and evaluation of the extent of a glomus tumor of the temporal bone requires appropriate physical examination and radiologic assessment (Glasscock et al, 1974). The physical examination infrequently differentiates between the two kinds of glomus tumors. Only if all borders of the tumor are visible through the tympanic membrane can one assume that it is a glomus tympanicum tumor. If all of the border are not visible behind the tympanic membrane, the tumor may be either a large glomus tympanicum tumor or a much larger glomus jugulare tumor, with a superior border that extends from the jugular bulb into the hypotympanum. If cranial nerve abnormalities are associated with a lesion, it is most likely of a glomus jugulare tumore.

Although physical examination helps deduce the extent of a paraganglioma, radiologic evaluation is required to precisely define the lesion. Both CT and MRI are usually used for this evaluation (Figs. 192-2, 3). The findings from these studies can be so distinctive that surgery may directly follow. However, paragangliomas are generally very vascular neoplasms. Hence, many practitioners prefer arteriography before approaching these lesions. Arteriography is required if preoperative embolization of the lesions is desired. Such embolization takes place through specialized intraarterial catheters and can lead to a marked reduction of the vascularity of these masses, which reduces intraoperative blood loss and hemorrhage-related complications. Such embolization is seldom considered definitive and is most often used as an adjunct to surgery. With the advent of high-quality CT, MRI, and on-line arteriographic digital subtraction, retrograde venography is now seldom performed. This technique was formerly used to determine the extent of glomus jugulare-type paragangliomas into the internal jugular vein.

As most paragangliomas are named by their topography, imaging is ideal to classify them. Lesions along the middle ear would be considered glomus tympanicum paragangliomas. These usually present early and, hence, show little bone erosion. The glomus jugulare

paraganglioma, the most common of these tumors, causes somewhat selective erosion of the pars vasculosa portion of the jugulare foramen. It also frequently destroys the bony septum between the ICA and internal jugular vein. This is a very distinctive structure whose absence is easily seen. Confounding lesions include schwannomas of the ninth, tenth, or eleventh cranial nerves as these also destroy the jugular foramen (Fig. 192-4). Fortunately, these lesions usually destroy the more medial portion of the pars vasculosa or, if originating from the ninth cranial nerve, the pars nervosa. At times, the lesion presents only after becoming very large and eroding nearly all of the jugular foramen. Simple differentiation by the localization of bone erosion is then not possible. MRI is very helpful in defining these lesions, but must be interpreted carefully. The complex flow characteristics of venous blood cause many signal artifacts in and about the jugular vein. Although experienced practitioners have become used to these artifacts, errors can still be made, and careful attention to the precontrast- and postcontrast-enhanced MRI or flow-specific MRI is necessary so that one does not interpret flow-related signal as a vascular lesion. Some of these tumors can produce vasoactive peptides, and appropriate studies should be ordered to evaluate their presence.

Surgical approach

Only small glomus tympanicum tumors whose borders are clearly visible through the tympanic membrane can be removed via the external auditory canal. In radiologically proven glomus tympanicum tumors whose borders are not entirely visible through the tympanic membrane, the extended facial recess approach is recommended (House, 1968). Occasionally the facial recess is narrow, requiring removal of all or part of the bony posterior external auditory canal.

The removal of glomus jugulare tumors is a major surgical task (Hilding and Greenberg, 1971; Kinney, 1979). Most commonly a glomus jugulare tumor is removed by the infratemporal approach or a modification thereof. It requires identification in the neck of the cranial nerves of the jugular foramen, transposition of the facial nerve, and removal of all the bone lateral to the tumor and jugular bulb.

A wide mastoidectomy is performed, and the facial nerve is identified from the cochleariform process into the parotid gland and transposed anteriorly. The sigmoid sinus is packed off and opened, and the jugular vein is ligated and divided in the neck. The jugular vein is then elevated superiorly up to the jugular bulb, with care being taken to visualize and preserve the cranial nerves on its anteromedial surface.

The tumor mass is then dissected free working the tumor margin in a circumferential fashion. As the tumor bleeds it is packed off with Surgicel, and a new area is dissected. Anteriorly, care must be taken to avoid injury to the carotid artery. Identification of the carotid artery remote from the tumor may be helpful in the development of this plane, but conservative resection is indicated in this area unless carotid artery ligation or reconstruction is anticipated. Resection of posterior fossa dura, facial nerve, cochlea, and semicircular canals may be required in larger tumors. Any dural defects that exist are repaired.

The surgical defect is packed with an abdominal fat graft. The external auditory canal skin may be oversewn after removal of the deep external auditory canal skin.

Radiation therapy

Although radiation therapy is not the treatment of choice for surgically resectable glomus tumors of the temporal bone, tumor reductions after radiation therapy have been recorded (Brackmann et al, 1972; Silverstone, 1973; Spector et al, 1974). Glomus tumors have been shown to have nonuniform responses, and tumor sterilization by radiation therapy is infrequent. Radiation therapy is useful for management of recurrences and unresectable lesions.

Facial Nerve Schwannomas

Schwannomas may involve the facial nerve along its intracranial, intratemporal, or extratemporal course. Altmann (1935) presented the first comprehensive description of a facial nerve schwannoma and felt that its symptomatology was sufficiently pathognomonic for the facial nerve schwannoma to be diagnosed clinically. These reported cases were generally diagnosed late and presented with long-standing facial nerve paralysis and obstruction of the external auditory canal with tumor. More recently, with the increased awareness of this clinical entity and improved neurotologic diagnostic methods, earlier diagnosis is possible.

Diagnostic evaluation

Facial weakness has been the most common presenting symptom in a number of series (Neely and Alford, 1974; Pulec, 1972). The classic history is slowly progressive facial paralysis over months to years, not days to weeks, although sudden onset of paralysis, fluctuating paresis, and facial tic may occur (Bailey and Graham, 1983). Hearing loss is the second most common presenting symptom (Horn et al, 1981). Conductive hearing loss may occur with middle ear involvement caused by tumor expansion and pressure on or disruption of the ossicular chain. In these cases a gray-white polypoid mass frequently exists behind the posterosuperior tympanic membrane. Sensorineural hearing loss may result from erosion into the labyrinth or tumor compression on the cochlear nerve.

An intratemporal facial nerve schwannoma generally extends along the course of the facial nerve for a greater distance than clinically or radiologically anticipated. The schwannoma visualized in the middle ear may extend intracranially into the internal auditory canal and cerebellopontine angle or extratemporally into the parotid gland. Preoperative radiologic assessment addresses this potential spread, but the surgeon must anticipate a more extensive nerve involvement than clinically or radiologically suspected.

Topographic testing of facial nerve function, including tearing, stapedius reflex, taste, and salivation, may be helpful but is not definitive in the evaluation of the extent of facial nerve neuroma.

Definitive assessment of facial canal alterations is made with axial and coronal temporal bone CT. The entire bony path of the facial nerve can be traced with certainty. In the normal structure, there is seldom much variation in this path from side to side. Hence, it is simple to compare right and left to determine abnormality. The most common abnormality seen in facial schwannomas is simple enlargement of the fallopian canal by a soft tissue mass. These lesions usually exactly follow the path of the facial nerve. One great mimic of facial

schwannomas is the facial nerve hemangioma. This uncommon lesion probably arises from the arteriovenous plexus around the fallopian portion of the facial nerve. Differential diagnostic imaging characteristics include calcification-based higher CT attenuation in a hemangioma as compared to a schwannoma.

MRI is superior to CT in delineating a soft tissue mass along the facial nerve, especially when MRI contrast material is used. This material provides such sensitivity that the arteriovenous plexus along the fallopian portion of the facial nerve is visualized. This plexus can normally enhance in a spotty fashion, making it possible for inexperienced practitioners to idetify this abnormality. In addition, inflammatory lesions of the facial nerve will also result in contrast enhancement. Accordingly, facial nerve enhancement on MRI should not be considered indicative of neoplasm unless there is a soft tissue mass. One must correlate the clinical findings, CT, or MRI before diagnosing a facial nerve neoplasm. If this correlation is not definite, later imaging follow-up after a clinically appropriate amount of time has passed should resolve the issue.

In some cases of facial paralysis, radiologic evidence for tumor is lacking. Surgical exploration of all progressive facial palsies that occur over an extended period of time or facial palsies that show no return of tone or function after 6 to 12 months has been recommended, and facial schwannomas are occasionally discovered.

Surgical approach

The treatment goals of facial nerve neuroma surgery are complete removal of the tumor with preservation of hearing and restoration of facial nerve function. Most commonly, the transmastoid and middle cranial fossa approaches are adequate for exposure of neuromas of the intralabyrinthine facial nerve segments. In patients with nonserviceable hearing, a translabyrinthine exposure of the facial nerve may be indicated.

Surgical removal and cable grafting of the facial nerve with frozen section histologic control is indicated in most cases. The nerve graft is usually placed in the fallopian canal, and its ends are approximated and stabilized with packing. Approximation of the cable graft to the facial nerve stump in the internal auditory canal is difficult. When the involved segment of nerve is limited to less than 1 cm, a facial nerve transposition and primary anastomosis can sometimes be performed. If primary anastomosis or cable grafting fail, hypoglossal-facial anastomosis may be necessary. Recently, we have elected to decompress a facial nerve shwannoma in several patients with normal facial nerve function. This approach may be particularly useful in the elderly patient.

Congenital Cholesteatoma

Congenital cholesteatoma of the temporal bone may be divided into four anatomic groups: middle ear, perigeniculate area, petrous apex, and cerebellopontine angle. These cholesteatomas arise from epithelial rests and should be distinguished from acquired cholesteatomas and from congenital auditory canal cholesteatomas associated with group I abnormalities in congenital atresia of the external auditory canal.

Diagnostic evaluation

The clinical presentation of a congenital cholesteatoma varies, depending on its location. Middle ear cholesteatomas were originally reported by House (1953) and have been reviewed recently by House and Sheehy (1980) and Curtis (1979). Conductive hearing loss and a bulging whitish mass behind an intact tympanic membrane are common findings in middle ear/mastoid congenital cholesteatomas (Peron and Schuknecht, 1975). Half of all cases appear with current otitis media and may fistulize through the tympanic membrane. They may also be discovered incidentally at the time of a myringotomy or routine physical examination in the pediatric population.

Perigeniculate and petrous apex cholesteatomas usually present with insidious or rapidly progressive facial nerve paralysis (Fisch, 1978; Nager, 1975). Sensorineural hearing loss from labyrinthine or internal auditory canal erosion is common, but a conductive hearing may also result from cholesteatoma extension into the middle ear or blockage of the eustachian tube. Facial twitching may occur in the presence of congenital cholesteatomas as well as with facial nerve neuromas. Vestibular dysfunction may complete the symptom complex. On occasion the cholesteatoma may erode into the middle or posterior fossa and expand markedly before producing symptoms. Preservation of hearing despite extensive destruction of the labyrinth has been reported (Bumsted et al, 1977; Thomsen et al, 1980).

Radiologic evaluation of the temporal bone is essential in the evaluation of a congenital cholesteatoma (Fig. 192-5). Only the largest lesions will show on plain film. High resolution, focused CT of the temporal bone is ideal in this evaluation as any alteration in normal bony structure can be seen. At times, the normal asymmetry in aeration, inflammatory, or passive effusion and bone marrow deposition in the petrous apices can cause diagnostic difficulties. These difficulties are usually resolved by clinical correlation. If still problematic, however, MRI will provide further definition, as congenital cholesteatomas will almost always have a slightly higher signal than spinal fluid on T1-weighted images with a moderately high signal on T2-weighted images. In contradistinction, bone marrow fat will have a very high signal on T1-weighted images and then go on to fade dramatically on T2-weighted images. Typical effusions will have low signal on T1-weighted MRI with high signal on T2-weighted MRI. Giant cholesterol cysts can be difficult to separate from congenital cholesteatomas of the petrous apex (Fig. 192-6). However, giant cholesterol cysts, but not congenital cholesteatomas, tend to show capsular enhancement.

Surgical approach

Surgical management of a congenital cholesteatoma requires either complete removal of the cholesteatoma matrix or permanent exteriorization. The isolated middle ear cholesteatoma may be removed transtympanically. The surgeon must be prepared to visualize clearly the complete extent of the cholesteatoma in the middle ear particularly on the medial aspect of the tympanic membrane. Routine middle ear reconstructive techniques may be used if ossicles are eroded or removed or if the tympanic membrane is sacrificed.

Removal of congenital cholesteatomas of the perigeniculate area or petrous apex may be accomplished by the transmastoid middle cranial fossa approach, the transsphenoidal approach, or by a combination of these procedures. Frequently a cholesteatoma must be

dissected from the facial nerve, and occasionally the intralabyrinthine portion of the nerve must be mobilized to remove completely a perigeniculate cholesteatoma. This maneuver may require sectioning of the greater superficial petrosal nerve and transposition of the facial nerve posteriorly at the geniculate ganglion. The cholesteatoma may also insinuate itself between the dura and the middle fossa floor and may extend for a considerable distance along this plane. Petrous apex cholesteatomas may act similarly and erode the bony labyrinth or internal auditory canal. Only by careful and often tedious dissection can one prevent injury to these structures.

Cholesteatomas of the petrous apex may be exteriorized into the mastoid cavity or the sphenoid sinus; however, frequent cleansing of debris from the cholesteatoma cavity is required to prevent inspissation and subsequent infection (Gacek, 1980).

Malignancy

A wide variety of malignant tumors may involve the middle ear and mastoid. Squamous cell carcinomas are the most frequent histologic type in this area (Batsakis, 1979a). Although the high incidence of squamous cell carcinoma of the mastoid is well documented in radium-dial painters, the inciting cause in the majority of middle ear carcinomas is unknown (Beal et al, 1965). Although ultraviolet light is considered to cause squamous cell carcinoma of the pinna, chronic eczematoid external otitis and chronic suppurative otitis have been proposed as causing squamous cell carcinomas of the external auditory canal and middle ear. Adenoid cystic carcinoma and a wide variety of other carcinomas, including basal cell carcinoma, melanoma, mucoepidermoid carcinoma, and ceruminous gland adenocarcinoma, have been reported in this location (Batsakis, 1979b; Pulec, 1977).

A rhabdomyosarcoma is the most common childhood middle ear and mastoid malignancy. A recent spectacular improvement in 5-year survival rates in this disease has been achieved by several continued refinements of combined therapy: more accurate staging of parameningeal disease, adjustments of chemotherapeutic agents and dosage schedules, and advances in radiotherapy techniques (Feldman, 1982; Raney et al, 1983). Although total tumor removal is the surgical goal when possible, surgical debulking is an important concept in this disease. Gross total removal with preservation of the dura, otic capsule, and neurologic and vascular structures, with exteriorization of the mastoid middle ear cavity, is performed.

Diagnostic evaluation

In carcinoma of the mastoid and temporal bone, early diagnosis is difficult because the tumor is often hidden from view. With growth and destruction of the bone, headache and pain may occur. The carcinoma may become apparent as it erodes into the external auditory canal or through the mastoid cortex into the skin. Cranial nerve deficits occur as the tumor destroys the skull base. Cervical metastases may occur early, but distant metastases usually occur late in the course of disease. Death from local extension is the rule.

A biopsy of the lesion leads to the histologic diagnosis. Unusual varieties of both benign and malignant tumors in this area may require special histologic or electron microscopic study for diagnosis.

Radiologic evaluation of tumor extent and surgical resectability many times begins and ends with contrast enhanced CT (Fig. 192-7 to 192-12), because many of these lesions are large and show obvious bone destruction at the time of presentation. In addition, cervical adenopathy, so important in staging these lesions, can be evaluated efficiently with contrast enhanced CT. MRI is usually added only if there is suspicion of involvement of the brain, ICA or internal jugular vein. Although these vessels could be imaged with arteriography, MRI is less invasive and affords nearly equivalent evaluation of neoplastic involvement of such large vasculature. Contrast-enhanced CT is also able to provide information about these vessels, but is less sensitive than MRI in this regard.

Surgical approach

Lateral temporal bone resection preserves the facial nerve with minimal cosmetic and functional disabilities when squamous cell carcinoma is restricted to the external auditory canal (Parsons and Lewis, 1954). With carcinomas extending into the middle ear or mastoid, more extensive surgical procedures with greater cosmetic and functional disabilities may be necessary - either subtotal resection lateral and posterior to the carotid artery (Arena, 1974; Gacek and Goodman, 1977; Neely and Forrester, 1982) or total resection after planned ICA ligation (Graham et al, 1984). Whether these more extensive surgical procedures improve long-term prognosis is not known. Postoperative radiation therapy should be strongly considered with extensive lesions.

Encephalocele and CSF Leaks

Postsurgical and postinfectious encephaloceles through the tegmen tympani and tegmen mastoideum are well-recognized complications in otology (Baron, 1969; Paparella et al, 1978; Glasscock et al, 1979). Temporal bone herniation of the mastoid cavity is secondary to loss of both bony and dural support. Subsequent management must deal with CSF leakage, herniation of the brain into the mastoid cavity, and skin and cholesteatoma matrix in the middle ear and mastoid. Less frequently recognized is a spontaneous congenital encephalocele associated with CSF otorrhea or rhinorrhea (Blatt, 1963; Dysart, 1959; Jahrsdoerfer et al, 1982; Kamerer and Caparosa, 1982; Koch, 1950; Neely et al, 1982).

Diagnostic evaluation

The appearance of postsurgical encephaloceles may occur early or late in the postoperative course. Early occurrence are generally associated with maceration of exposed brain and CSF leakage. Late occurrences are associated with pulsatile, progressive prolapse of skin-covered brain into the mastoid cavity.

Spontaneous congenital encephalocele may appear as serous otitis media, CSF rhinorrhea, or CSF otorrhea through a tympanic membrane perforation or pressure-equalizing tube. Occasionally, prolapsed brain can be present as a mass behind the tympanic membrane.

As small and often multiple bony defects can be involed, carefully focused CT in both the axial and coronal planes is needed to find these fenestrations (Figs. 192-13 and 192-14). If necessary, the subarachnoid space can be opacified with myelographic contrast material, and the actual flow of the contrast-opacified spinal fluid through the fenestrations can be

observed. 3-D displays of such fenestrations can be dramatic, but are generally considered only supplemental to standard axial and coronal sections.

Surgical approach

Temporal bone encephaloceles and CSF leaks may be surgically approached either transmastoidally or through the middle cranial fossa intradurally or extradurally. Although transmastoid repair of the defect is frequently successful, repair through the middle cranial fossa allows greater visualization. Techniques for repair of the tegment defect via the mastoid approach have included intracranial placement of reconstructive material (that is, fascia, cartilage, or prosthetics) from the mastoid combined with obliteration of the mastoid with flaps or free tissue (Glasscock et al, 1979; Jahrsdoerfer et al, 1981; Jahn, 1981; Kamerer and Caparosa, 1982; Neely et al, 1982). A middle cranial fossa approach for repair provides greater visualization and can be either extradural or intradural (Graham, 1982; Spetzler and Wilson, 1978). A variety of closure techniques, including primary dural repair and fascia-bone-fascia, bone, cartilage, or syntehtic supports, have been used.

The bony and dural defects in spontaneous encephaloceles are routinely small and multiple (Kemink et al, 1986). Frequently an encephalocele exists in the anterior epitympanum, where visualization may be inadequate and surgical repair via the transmastoid approach is difficult without ossicular disruption. Although we routinely explore these cases transmastoidally to define the defect, repair is accomplished through the middle cranial fossa. The bony defects are usually small and do not require repair. The dural defects can be sealed by wide coverage of the middle cranial fossa with temporalis fascia and bone or cartilage as necessary.

Chordoma

Cranial chordomas arise from the clivus and progressively destroy the skull base (Blatt, 1963; Jahrsdoerfer et al, 1981; Koch, 1950) (Figs. 192-15, 192-16). They may extend ventrally into the nasopharynx, nose, or sinuses and cause obstruction. Headache, loss of vision, and cranial nerve deficits with involvement of the abducens, trigeminal, facial, and acoustic nerves are common presenting complaints. Because of the inaccessibility of the tumor site, surgical exposure and tumor removal may be difficult. Surgical debulking of the tumor followed by high-dose radiation therapy is the treatment of choice and may afford good palliative results.

More recently skull base surgeons have become more aggressive "tailoring" surgical approaches to individual lesions in efforts to ensure complete removal. Because of the difficult accessibility, surgical control of chordoma is a unique challenge to the skull base surgeon and lends itself to creative surgical approaches (see Chapter 190).