Chapter 126 – Cholesterol Granuloma and Congenital Epidermoid Tumors of the Temporal Bone

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Expansile destruction of the petrous apex may be due to congenital epidermoid tumor or cholesterol granuloma. The etiology, incidence, and management of cholesterol granuloma and congenital epidermoid tumor within the temporal bone are different. Distinguishing between these two processes was difficult before the advent of computed tomography (CT) and magnetic resonance imaging (MRI), especially when the lesion or tumor originated medially and anteriorly within the petrous apex.

Pathology extending in this area challenges the neurotologic team to comprehensively evaluate and effectively manage these lesions. Abnormalities in the petrous apex may be silent for many years because of the depth and isolation of this area. Once the pathology encroaches on surrounding vital structures such as the dura and brain, carotid artery, orbit, 4th through 11th cranial nerves, and otic capsule, symptoms and signs will become apparent. Pathology in the petrous apex is often identified incidentally or when diagnostic imaging is performed for vaguely described symptoms such as lightheadedness; retro-orbital, ear, vertex, or occipital head pain; tinnitus; visual blurriness; or diplopia. Just as the causes and radiologic findings differ, so do the management strategies and surgical approaches.

PERTINENT ANATOMY

The petrous apex is a wedge-shaped structure that occupies the medial and anterior aspect of the temporal bone. The petrous apex is inferior, medial, anterior, and superior to the otic capsule. The superior surface forms the floor of the middle cranial fossa. Along this surface pass the greater and lesser superficial petrosal nerves and is the exit point for the middle meningeal artery through the foramen spinosum. At the tip of the petrous apex lies a depression containing the gasserian ganglion of the trigeminal nerve. The petroclinoid ligament forms the attachment of the tip of the petrous apex to the sphenoid bone. This area is termed *Meckel's cave,* through which passes the fifth cranial nerve. The sixth cranial nerve is located just medially and passes through Dorello's canal. The horizontal portion of the carotid artery lies posteromedial to V3, and its bony covering is often dehiscent. The anterolateral surface of the petrous bone contains the genu of the carotid artery and its horizontal segment. The bony opening of the eustachian tube passes in an anterior, inferior, and medial direction toward the nasopharynx. The jugular bulb and vein exit the inferior surface of the petrous bone through the jugular foramen. The 9th, 10th, and 11th cranial nerves also leave the skull base through the pars nervosa of the jugular fossa. The carotid foramen lies anterior to the jugular bulb on the inferior surface of the petrous bone. The posteromedial surface of the temporal bone faces the posterior fossa. Openings into this surface include the internal auditory canal (IAC), which contains the cochlear, vestibular, facial, and intermedius nerves; the operculum, which contains the endolymphatic duct and sac; and the aperture for the subarcuate artery. The posterior aspect of the temporal bone contains the mastoid cavity and faces the sigmoid sinus and posterior fossa (Fig. 126-1).

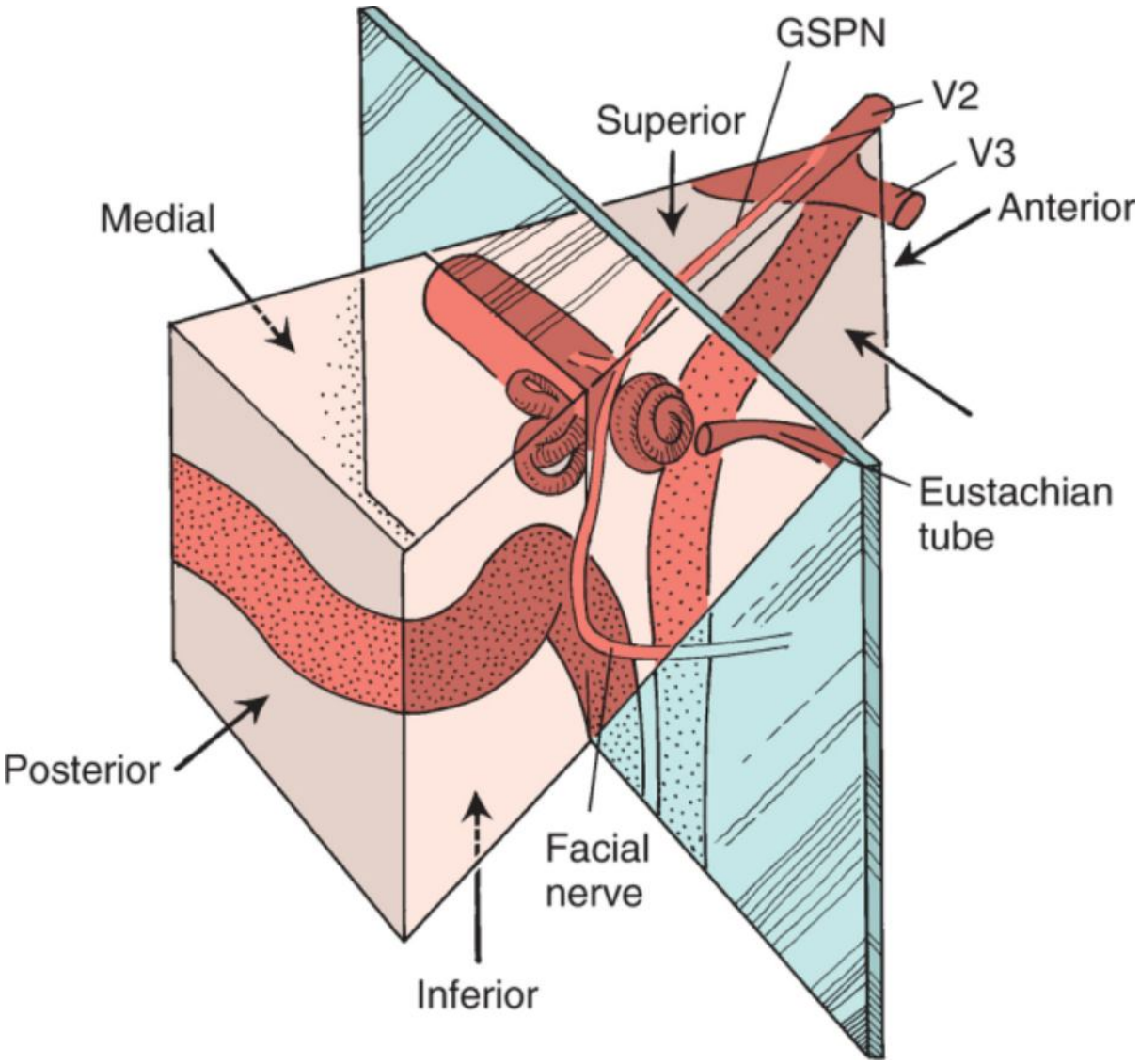

Figure 126-1 Petrous portion of the temporal bone. A coronal plane through the internal auditory canal separates the posterior and anterior petrous apex. The greater superficial petrosal nerve (GSPN) runs along the superior surface of the petrous bone. The gasserian ganglion and the mandibular division of the trigeminal nerve are located in the apex.

The petrous bone can be conceptually and functionally divided into anterior and posterior compartments.[1] This division is in a coronal direction passing through the IAC. Pathology in the posterior compartment is readily accessed through a transmastoid or translabyrinthine approach. Congenital cholesteatomas of the middle ear are frequently accessed via the external auditory meatus. Abnormalities in the anterior compartment, including the petrous apex, require more complex approaches.

Access to the Petrous Apex

Approaches to the petrous apex require an understanding of the anatomy of the temporal bone and surrounding vital structures of the skull base and posterior, middle, anterior, and infratemporal fossae. Access to the petrous apex is limited because of its anatomic location. The position of the otic capsule, facial nerve, and carotid artery obviates direct and complete exposure to this centrally located area.

Before the availability of antibiotics, suppurative disease of the petrous apex was a common sequela of otitis media and was associated with high morbidity and mortality. Historically, various transtemporal approaches have been described to gain access to the relatively isolated area of the petrous apex via the air cell tracts surrounding the otic capsule. These approaches were predominantly used for the management of infectious complications of otitis media causing petrous apicitis (Fig. 126-2). The various routes through the temporal bone include

1. The supralabyrinthine tract inferior to the middle fossa dura
2. The subarcuate tract beneath the superior semicircular canal
3. The air cells of the zygomatic root
4. The small triangular region of bone between the genu of the carotid artery and the cochlea

5. The retrofacial infralabyrinthine tract
6. The infracochlear air cell tract

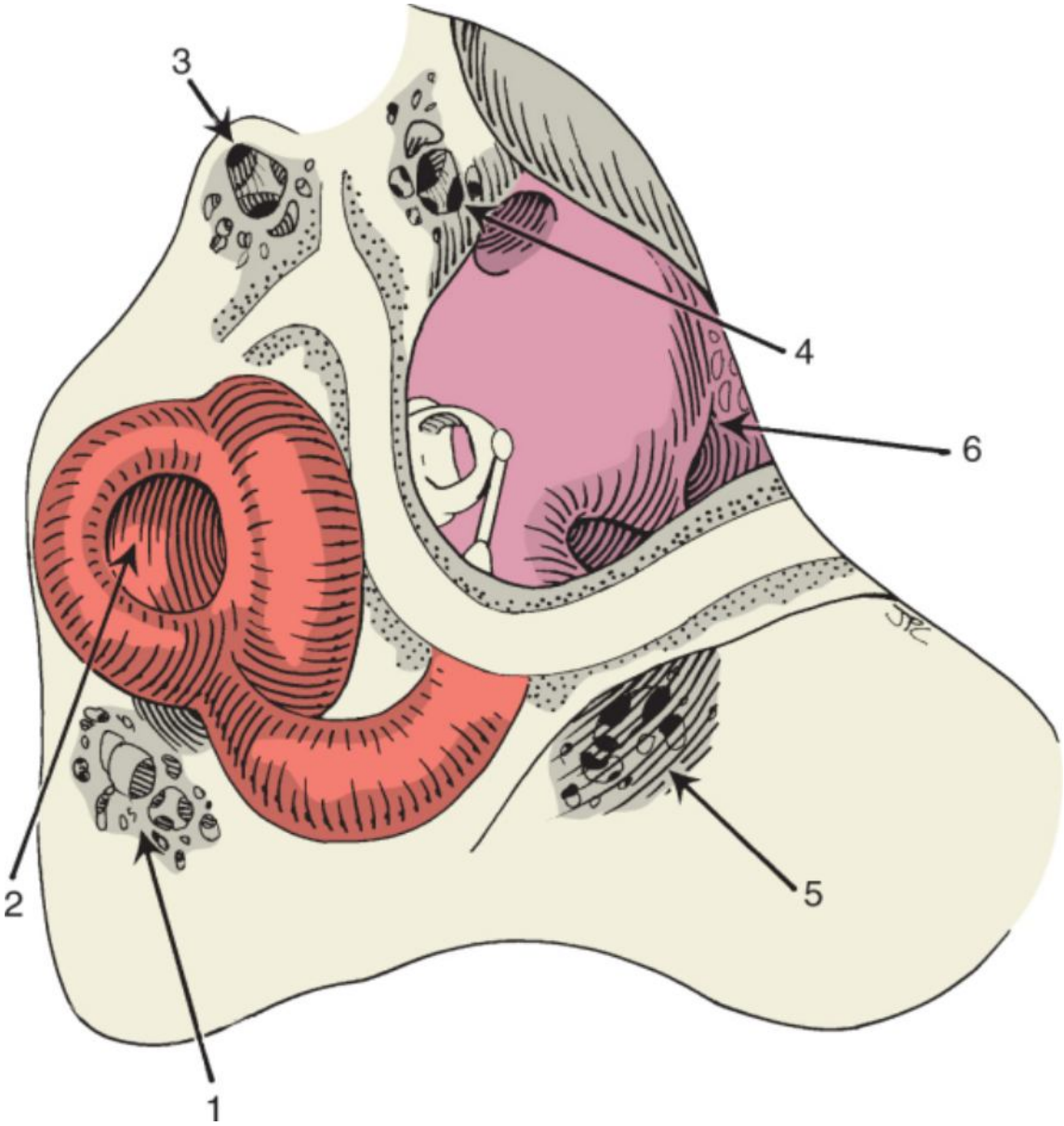

Figure 126-2 Drainage approaches to the petrous apex. 1, Supralabyrinthine tract inferior to the middle fossa dura; 2, subarcuate tract beneath the superior semicircular canal; 3, air cells of the zygomatic root; 4, small triangular region of bone between the genu of the carotid artery and the cochlea; 5, retrofacial infralabyrinthine tract; 6, infracochlear air cell tract.

The first four routes are effective for draining an abscess or suppurative inflammation in the petrous apex but may not adequately provide permanent aeration to this remote area. The latter two routes do provide a more reliable and wider opening to the petrous apex. The infracochlear approach was described by Farrior and then modified by Brackmann and coworkers to adapt the approach for drainage of petrous apex cholesterol granuloma.[2,3]

The development of advanced endoscopic techniques and instrumentation has provided access to the petrous apex though the nose, nasopharynx, sphenoid sinus, and base of the skull. The transnasal approach enters the petrous bone anteriorly and is medial to the carotid artery. The conventional lateral approaches passing superior or inferior to the temporal bone require careful consideration of the carotid artery and otic capsule. Specific indications and limitations must be recognized when planning surgical treatment of pathology in the petrous apex.

Lesions of the Petrous Apex

With more extensive use of high-definition CT and MRI, lesions of the petrous apex are now often identified before they become large and destructive. Most lesions of the petrous apex remain quiescent and grow slowly until they affect neighboring neurovascular structures, including the cranial nerves, brain, orbit, or carotid artery, or extend into the middle ear. Patients suffering from lesions of the petrous apex most often complain of hearing loss,

dizziness, tinnitus, and headache (40% to 60%). Abnormalities of the cranial nerves causing symptoms and signs such as facial twitching or weakness or diplopia are less common (5% to 15%) but raise greater suspicion for pathology in the petrous apex. Extension of disease into the middle ear may result in conductive hearing loss, but perforation of the tympanic membrane with subsequent otorrhea is rare (5%).[4,5] Facial numbness, amaurosis fugax, vertigo, transient ischemic attacks, ataxia, dysmetria, and incoordination can also occur.

Lesions of the petrous apex can be classified into three categories: cystic, solid, and radiographic anomalies. A retrospective study analyzing lesions of the petrous apex determined cystic lesions to be the most common source of pathology.[4] Such lesions include cholesterol granuloma (60%), cholesteatoma (10%), and mucocele (3%). Second in frequency are solid tumors, a heterogeneous group that includes chondrosarcoma (6%), chondroma (3%), cavernous hemangioma (3%), and metastatic carcinoma (2%). The least common category is radiographic anomalies such as retained secretions (6%) and asymmetrical pneumatization (3%). That said, lesions of the petrous apex are quite rare and represent only about 0.5% of pathology found on MRI performed for sudden hearing loss, vestibular symptoms, or tinnitus.[6]

Cholesterol Granuloma

Cholesterol granuloma is a comprehensive term describing a loculated cystic or solid foreign body reactive process directed against cholesterol crystals resulting from the by-products of degraded blood. Terms synonymous with cholesterol granuloma include *cholesterol, chocolate, dark brown,* or *blue dome cysts.* Cholesterol granuloma was originally described by Politzer in 1869 as idiopathic hemotympanum.[7] In 1929, Shambaugh introduced this term to the English literature.[8] Cholesterol granuloma was not reported in the petrous apex until House and Brackmann described the finding in 1982, which was subsequently recognized as a distinct clinical entity in 1985.[9,10] Although these cysts represent the most common lesion of the petrous apex, they remain quite rare, with an incidence of less than 0.6 case per million per year.[11]

Cholesterol granuloma may develop in diverse locations, including the peritoneal cavity, pleura, and paranasal sinus. Cholesterol granuloma originating in the temporal bone can be divided into two categories: cholesterol granuloma of the tympanomastoid compartment and cholesterol granuloma of the petrous apex. Each type has distinctive clinical behavior and pathogenesis. Cholesterol granuloma of the tympanomastoid compartment is typically painless and preceded by chronic otitis, is associated with limited pneumatization in the temporal bones, and rarely erodes bone. Cholesterol granuloma also frequently forms outside the middle ear in patients with well-pneumatized temporal bones who have acute and chronic mastoid inflammation associated with obstruction of the aditus ad antrum. This process is often isolated to one or a few mastoid air cells located toward the mastoid tip or cortex. In contrast, cholesterol granuloma of the petrous apex is often found in patients with dry ears and highly pneumatized temporal bones and is associated with chronic deep otic or retro-orbital pain, as well as neuropathy of cranial nerves V, VI, and occasionally VII. These lesions extensively erode or expand bone.

The pathogenesis of cholesterol granuloma is unknown and somewhat controversial. The inciting event is presumed to be inflammation and hemorrhage in an area of compromised aeration and drainage. Anaerobic breakdown of blood leads to the formation of cholesterol crystals and a subsequent sterile foreign body reaction. The granuloma blocks ventilation, and as a consequence the reaction propagates and results in growth of the cyst. Thus, maintaining good aeration of the cyst is imperative to prevent recurrence after drainage. Curiously, petrous apex cholesterol granuloma develops preferentially in hyperpneumatized temporal bones. Extensive pneumatization is possibly associated with a defect in bone marrow–air cell separation observed on CT. Highly vascular bone marrow may provide a continuous source of bleeding to the growing cyst.[12] The supply of blood may also result from newly described venules located preferentially in the well-pneumatized petrous apex.[13]

The contents of cholesterol granuloma demonstrate a characteristic appearance by both gross observation and histologic examination. The contents of the cyst typically include a yellow-brown, viscous glue-like fluid with glittering crystals detectable by the unaided eye. The brown discoloration is thought to be derived from metabolized hemosiderin. A green-yellow color is a manifestation of its lipid contents, and the glistening refractive properties are attributed to the cholesterol crystals. Histologically, the cyst is characterized by cholesterol crystals surrounded by foreign body reactive giant cells and fibrous connective tissue containing round cells, macrophages, and evidence of neovascularization (Fig. 126-3). Cholesterol granuloma does not contain squamous epithelium or keratin debris. Similar to congenital epidermoid tumor, cholesterol granuloma of the petrous apex is an indolent, slowly growing cystic tumor eroding bone that eventually becomes symptomatic once the lesion has extended to involve the dura and brain, cavernous sinus, carotid artery, orbit, cranial nerves, or otic capsule.

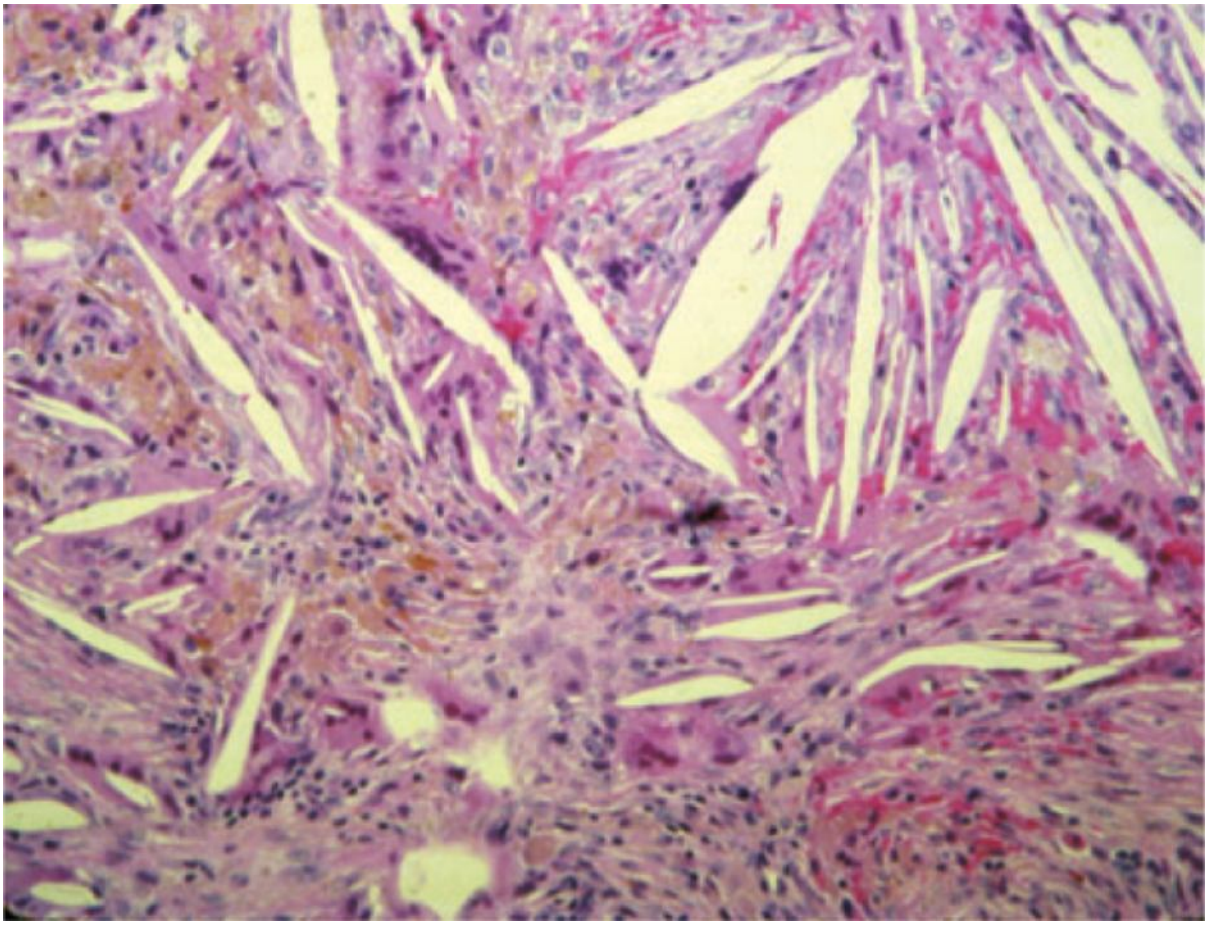

Figure 126-3 Histopathology of cholesterol granuloma. Note the large cholesterol crystal surrounded by a foreign body reaction with giant cells, macrophages, and fibrous connective tissue.

Congenital Epidermoid Tumors

Congenital epidermoid tumors of the temporal bone can have their origin in the middle ear, geniculate ganglion area, mastoid process, squamous epithelium, petrous apex, jugular foramen, or posterior fossa. Congenital epidermoid tumors are also termed *congenital cholesteatoma, keratosis, primary keratoma, inclusion cyst,* and *pseudomucocele,* especially when they are located within the middle ear.[14] The histopathology of congenital cholesteatoma or epidermoid tumor demonstrates a squamous epithelium–lined cyst filled with keratin debris. There are no additional dermal elements other than a surrounding outer layer of connective tissue that may be moderately vascular. The theory that these tumors may originally develop from congenital epithelial cell rests was proposed by Von Remak in 1854.[15]

According to Schuknecht, congenital epidermoid tumors occur in five sites in the temporal bone: (1) the petrous apex, (2) the middle ear, (3) the mastoid, (4) the cerebellopontine angle (CPA), and (5) the external auditory canal (EAC).[14]

Congenital epidermoid tumors of the petrous apex are believed to originate in the foramen lacerum during cephalic flexure of the head in early embryonic development with trapping of epithelial components. The cyst grows slowly. Similar to cholesterol granuloma, the symptoms relate to the space-occupying nature of the mass. These lesions become symptomatic by progressively compressing the eustachian tube, the trigeminal nerve, or the IAC and its contents or by stretching the dura. Congenital epidermoid tumors of the petrous apex may extend into the middle ear space. These tumors remain quiescent until they affect the cranial nerves, brain, orbit, or carotid artery or extend into the middle ear and cause further hearing loss or otorrhea. Hearing loss is the most common initial symptom.[16] Signs and symptoms of facial nerve involvement characterized by paresis, synkinesis, twitching, and paralysis are the second most common manifestation of congenital epidermoid tumors of the petrous apex.[17]

Establishing that a cholesteatoma of the middle ear is of congenital origin can be controversial. It is argued that eustachian tube dysfunction, retraction of the tympanic membrane, middle ear infections, and epithelial migration from insertion of a myringotomy tube are the forces that trap epithelium in the middle ear space. However, Michaels clearly demonstrated an epidermoid formation in the anterior superior middle ear mucosa in 54% of embryos examined at 10 weeks of gestation.[18] With normal development of the temporal bone, the epidermoid

formation should undergo regression by 33 weeks of gestational age. The cause of congenital cholesteatoma is considered to be lack of regression and continued growth of the epidermoid formation.

The differential diagnosis of a white mass within or medial to the tympanic membrane also includes congenital cholesteatoma of the petrous apex extending to the middle ear, osteoma of the tympanic ring or scutum, tympanosclerosis, or an inclusion cyst originating within the tympanic membrane. It is presumed that congenital epidermoid tumors are present from birth and continue to grow. Congenital cholesteatoma of the middle ear is most commonly identified at 4 years of age, but the age at diagnosis may range from infancy to the second decade of life.[19] Congenital cholesteatoma of the middle ear is often diagnosed on routine physical examination or during evaluation of a child with unilateral or bilateral hearing loss or otorrhea. When detected early, the epithelial pearl is typically located in the anterosuperior quadrant of the middle ear space (Fig. 126-4). Progression of disease with breakdown of the cyst laterally through the tympanic membrane results in bacterial colonization and otorrhea. Breakdown within the middle ear permits extension of disease throughout the middle ear, mastoid, and temporal bone and results in chronic otitis media and mastoiditis. Once recurrent otorrhea and middle ear involvement are established, it is almost impossible to distinguish congenital cholesteatoma of the middle ear from acquired cholesteatoma. Congenital middle ear epidermoid or cholesteatoma may extend into the petrous apex, but they are not usually as extensive as primary lesions originating in this area. Furthermore, growth of a congenital middle ear epidermoid generally results in early identification because of signs and symptoms of conductive hearing loss and otorrhea.

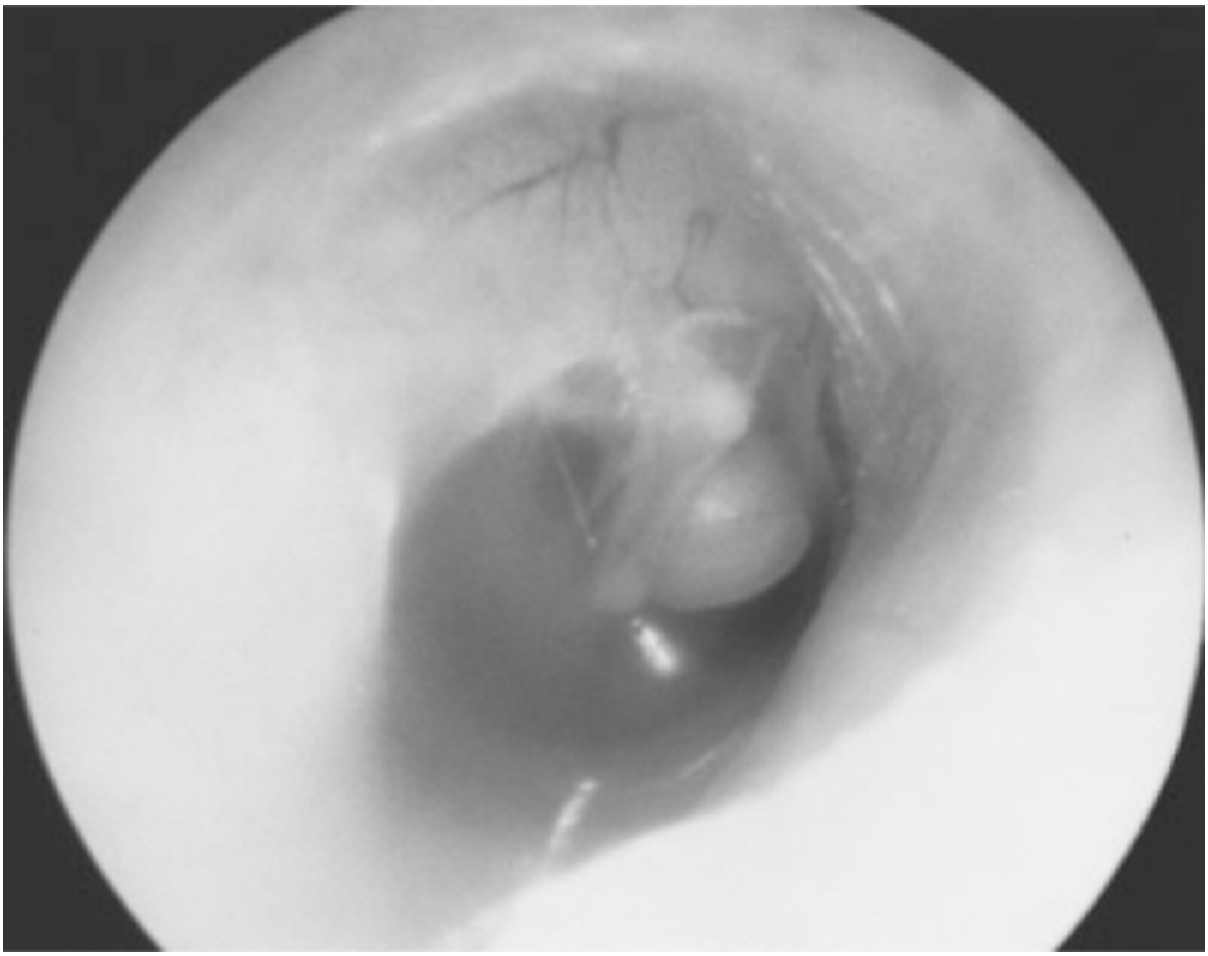

Figure 126-4 Congenital cholesteatoma located in the anterosuperior quadrant of the right ear behind an intact tympanic membrane.

Congenital epidermoid tumors of the mastoid are also proposed to be of embryologic origin. They develop in patients lacking a history of recurrent otitis media, eustachian tube dysfunction, and attic retraction. Frequently, epidermoid tumor of the mastoid is manifested as hearing loss associated with destruction of the ossicular chain or perforation of the tympanic membrane. As it progressively expands into the middle ear, epidermoid tumors of the mastoid may be difficult to differentiate from middle ear cholesteatoma.

Intracranial epidermoid cysts are most commonly located in the CPA. Congenital epidermoid tumors account for 6% to 7% of all tumors found in the CPA. It has been proposed that epidermoid tumors in the CPA arise from either multipotential embryonic cells or displaced cells of the otic capsule. They usually become manifested during the third and fourth decades of life, with a slight increased predilection in males.[20] Because of the close proximity

of the cranial nerves, congenital epidermoid tumors of the CPA are often accompanied by symptoms of hearing loss, tinnitus, or vertigo. Facial nerve signs of twitching and progressive weakness also occur. Infiltration of the dura by the tumor triggers the trigeminal nerve and causes headaches. As the tumor expands, cerebellar compression becomes manifested by postural instability, ataxia, and dysarthria. Extension anterosuperiorly and inferiorly will affect the 5th through 12th cranial nerves and create increased intracranial pressure, thereby causing headaches, nausea, vomiting, and potential loss of consciousness.[21] Because of their propensity for causing central neurologic symptoms and signs, congenital (primary) epidermoid tumors of the posterior fossa are seen more frequently by neurosurgeons.

Radiologic Imaging

Contemporary radiologic imaging modalities have introduced a new era into the diagnosis and management of temporal bone pathology. MRI can accurately define the soft tissue characteristics of these lesions and detect intracranial extension and involvement of the carotid artery and sigmoid sinus. CT demonstrates not only the specific pattern of rearrangement and destruction induced by the lesion but also the anatomic relationships of the tumor to the otic capsule, middle ear, carotid artery, and posterior and middle fossae. Despite the concern for cost containment in contemporary medicine, this is one area in which obtaining both imaging formats greatly assists the surgeon in formulating the diagnosis and planning the operative approach. Differentiation of lesions of the petrous apex is achieved by observing characteristic findings on both MRI and CT (Table 127-1).

Data obtained from the variable stimulating and capture techniques used with MRI (T1 versus T2 weighting) help determine the nature of the temporal bone pathology. A bright or hyperintense signal is observed on unenhanced (non–contrast-enhanced) T1-weighted images as a result of the presence of proteinaceous fluid, blood, or fat. A bright or hyperintense signal on conventional spin-echo T2-weighted sequences is indicative of fluid. Because cholesterol granuloma is composed of both methemoglobin breakdown products with paramagnetic properties and serous fluid, it is characteristically bright on both T1- and T2-weighted images (Fig. 126-5). The use of intravenous contrast agents such as gadolinium does not provide significant additional information for this purpose. Although the capsule of the cyst may demonstrate slight enhancement with gadolinium because of localized neovascularization, it is not usually evident because of the bright signal already present on the non–contrast-enhanced T1-weighted image. In addition to establishing the diagnosis, MRI is useful to track recurrence of cholesterol granuloma after surgical drainage. When faced with the presence of an opacified surgical bed, MRI effectively differentiates fluid retention from accumulation of cystic contents because only recurrence appears bright on T1-weighted images. Similarly, epidermoid cysts also have a characteristic appearance on MRI. An epidermoid tumor is usually isodense relative to brain and appears hypointense on T1-weighted imaging. However, similar to cholesterol granuloma, epidermoid tumors are bright on T2-weighted images. Also similar to cholesterol granuloma, epidermoid cysts do not significantly enhance with gadolinium, but slight peripheral enhancement corresponding to the capsule may be seen.[22] However, if the cyst is infected or infiltrated with granulation tissue, it will demonstrate frank contrast enhancement. Arachnoid cysts may be difficult to differentiate from epidermoid; both are filled with fluid and demonstrate low T1 and high T2 signal. In this case, fluid-attenuated inversion recovery (FLAIR) sequence will help differentiate these two pathologies because epidermoid cysts are bright on FLAIR se-quences and arachnoid cysts are not. Chondrosarcoma; metastasis from breast, kidney, and prostate cancer; and mucocele are other pathologies of the petrous apex that are bright on T2-weighted images but can be distinguished from epidermoid by differential examination of CT and MRI (see Table 126-1).

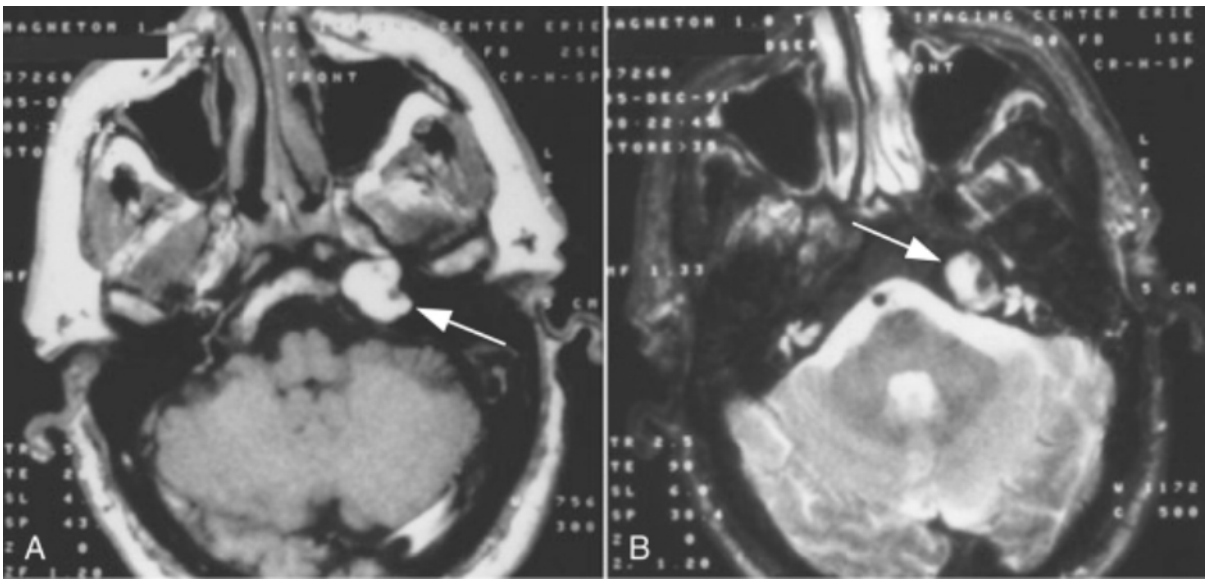

Figure 126-5 Petrous apex cholesterol granuloma, preoperative views. **A,** T1-weighted magnetic resonance image (MRI), without contrast enhancement, demonstrating a bright left petrous apex lesion *(arrow)* that is isointense with fat. **B,** T2-weighted MRI showing a bright left petrous apex lesion *(arrow)* that is isointense with cerebrospinal fluid and perilymph.

Table 126-1 -- CHARACTERISTIC RADIOLOGIC FINDINGS OF CHOLESTEROL GRANULOMA AND CONGENITAL EPIDERMOID TUMOR

	MRI			*CT*	
Lesion	T1-Weighted	Contrast Enhancement	T2-Weighted	Bone-Eroded Margin	Contrast Enhancement
Cholesterol granuloma	Hyperintense	No	Hyperintense	Smooth	Occasional rim enhancement
Congenital epidermoid tumor	Hypointense	No	Hyperintense	Smooth	No

CT, computed tomography; MRI, magnetic resonance imaging.

Newer MRI pulse sequences have been developed that further help differentiate pathology in petrous bone. FLAIR sequences appear similar to T2-weighted sequences except that free fluid such as cerebrospinal fluid (CSF) is suppressed. Proteinaceous fluid, as seen in most air cell pathology, remains bright. Most tumors, including epidermoid tumors, will be bright on FLAIR sequences, just as they are on T2-weighted sequences. Diffusion-weighted imaging is a method of evaluating the brownian motion of water molecules. Restricted water diffusion, thought to be due to the waxy consistency of epidermoid tumors, results in high signal intensity (Fig. 126-6).

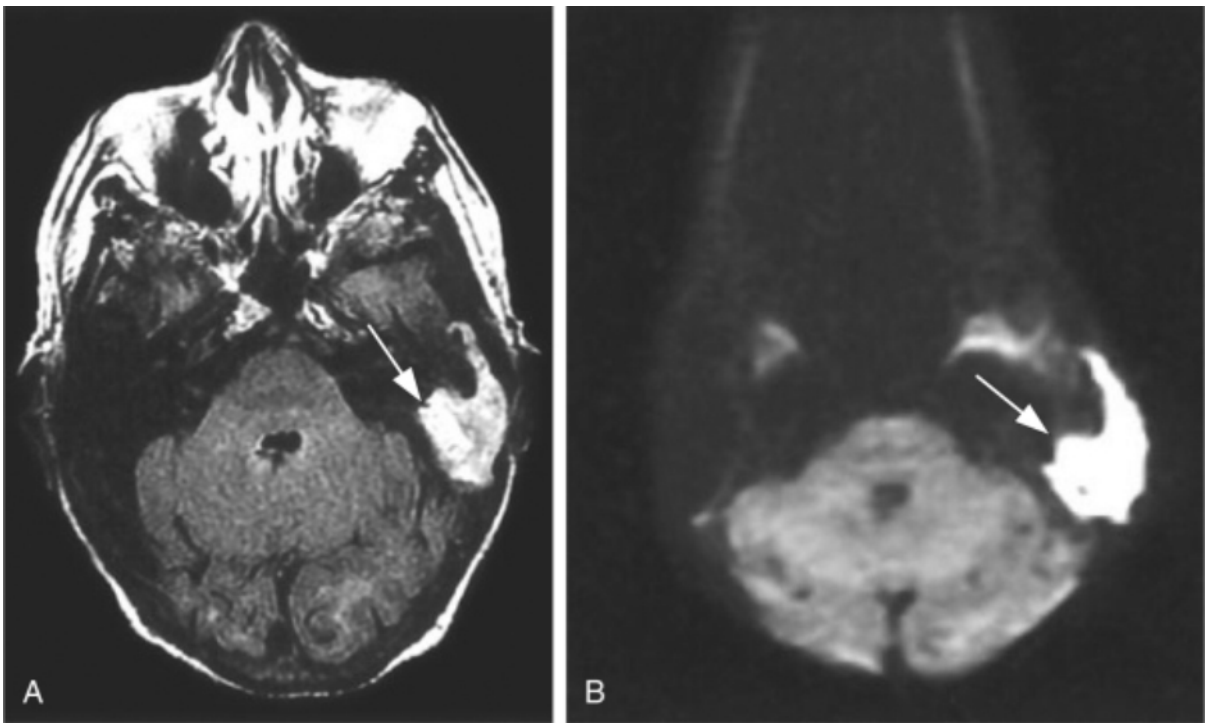

Figure 126-6 Magnetic resonance images of left petrous epidermoid tumor. **A,** Fluid-attenuated inversion recovery sequence showing a bright signal in the petrous and lateral temporal bone *(arrow)*. This excludes arachnoid cyst from the differential diagnosis. **B,** Diffusion-weighted image showing a bright signal in the same location, which strongly suggests it to be consistent with an epidermoid tumor *(arrow)*.

CT scanning provides significant additional information regarding the characteristics of petrous apex lesions. Cholesterol granuloma and cholesteatoma both appear as a smoothly marginated expanding mass. They appear isointense or mildly hypointense relative to nearby brain on soft tissue windowing. MRI with intravenous contrast does not show significant enhancement or provide additional information unless the cyst is infected or infiltrated by granulation tissue. However, data obtained from a bone window algorithm are most useful. The presence of temporal bone pneumatization suggests that the lesion is probably a cholesterol granuloma. Congenital

cholesteatoma is thought to inhibit normal temporal bone aeration, development, and thus pneumatization. The edges of cholesteatoma are sclerosed, eroded, and scalloped. Capsular calcification is occasionally seen. The size and location of the expan- sile lesion are clearly demonstrated. The relationship of the lesion or tumor to the otic capsule, ossicles, facial nerve, IAC, internal carotid artery (ICA), jugular bulb, and posterior fossa can be defined (Fig. 126-7). A bone-windowed CT scan in combination with MRI and MR angiography provides a road map for planning the operative approach.

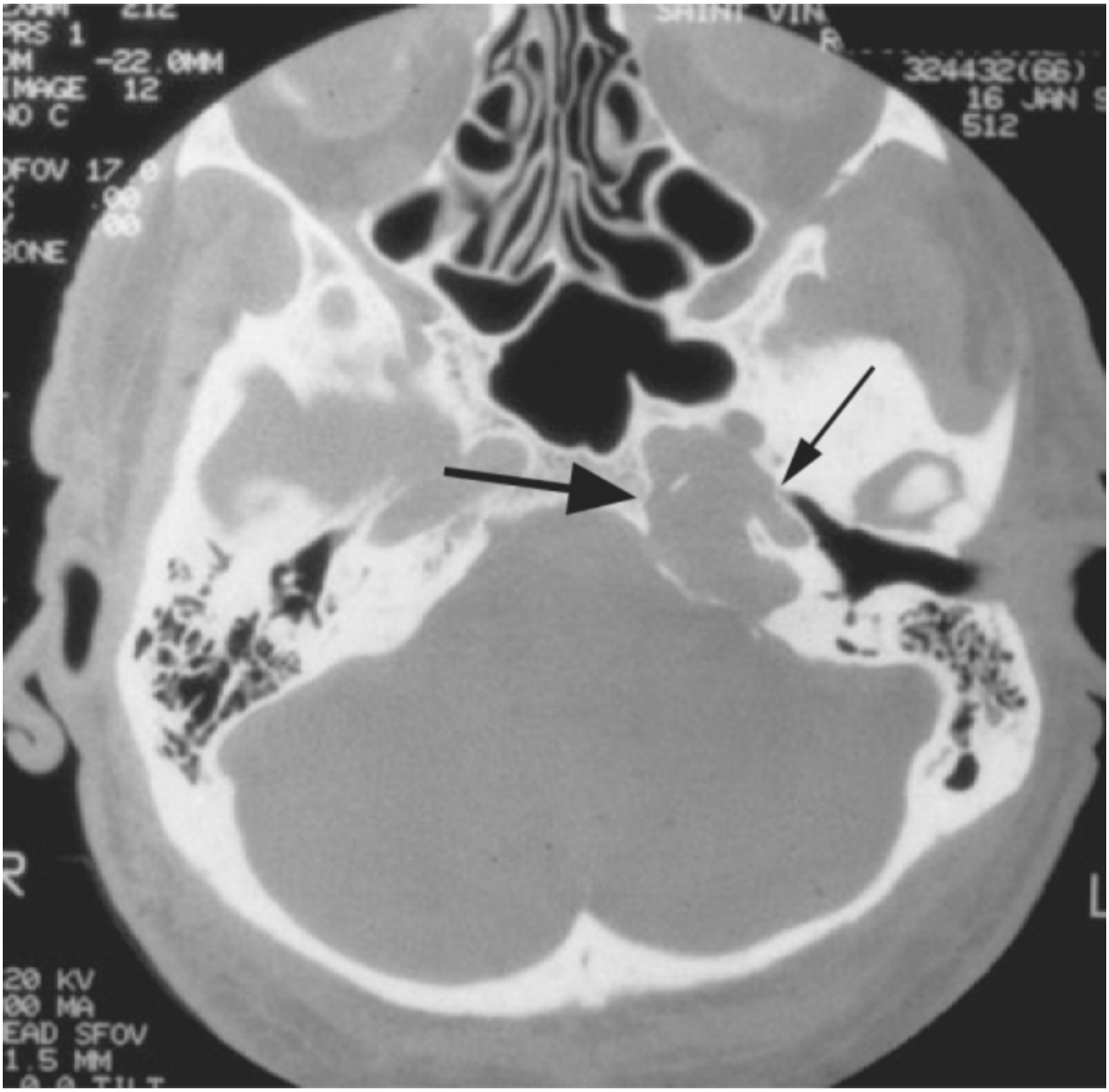

Figure 126-7 Bone-windowed computed tomography scan revealing an expansile left petrous apex lesion with smooth margins and isointense with brain *(large arrow),* consistent with cholesterol granuloma. Note the relationship to the carotid artery *(small arrow).*

PREOPERATIVE EVALUATION

Patients with congenital cholesteatoma or cholesterol granuloma will have had a presumptive diagnosis made that was based on initial signs, symptoms, and radiologic findings. Patients are questioned regarding their otologic history and previous surgical procedures. It should be determined whether they have had otorrhea, hearing loss, tinnitus, vestibular symptoms, facial paresis or twitching, diplopia, amaurosis fugax, or transient ischemic attacks. They are also asked about the location and severity of pain.

A complete neuro-otologic and head and neck physical examination is performed. The status of the tympanic membrane and middle ear space is evaluated. Facial and corneal sensations are tested for evidence of trigeminal nerve involvement. The full range of extraocular motion should be confirmed. Facial nerve function is carefully examined to look for evidence of fasciculation, weakness, or synkinesis.

A complete assessment of hearing consisting of air and bone conduction and speech audiometry is obtained. It is

important to evaluate auditory function in both the involved and uninvolved ears. Operative management may necessitate closing the ear canal, which results in maximal conductive hearing loss. On rare occasion, cochlear function may have to be sacrificed to completely remove an invasive cholesteatoma or congenital epidermoid tumor. Therefore, the contralateral ear should have normal hearing or be suitable for aural rehabilitation with amplification.

Radiologic imaging is critical in diagnosing and planning treatment of cholesterol granuloma and congenital epidermoid tumors. These two pathologic processes affecting the temporal bone are similar in that both CT and MRI are necessary to formulate the diagnosis and treatment plan. MRI defines the differential diagnosis based on the characteristic signals generated by the soft tissue and fluid content of the tumor or cyst. The tumor or cyst and the pneumatization tracts of the temporal bone are identified by bone-windowed CT. A well-developed pneumatized temporal bone is more amenable to exteriorization of epidermoid tumor and to drainage and aeration of cholesterol granuloma than a sclerotic temporal bone is. The location of the pathology in the petrous apex in relation to the sphenoid sinus should be examined. Expansion anteriorly toward a large sphenoid sinus allows the surgeon to consider a transnasal-transsphenoidal approach for management of the lesion. The relationship of the jugular bulb to the facial nerve and otic capsule is critical when a drainage procedure inferior to the otic capsule is likely. A high and lateral jugular bulb obstructs retrofacial infralabyrinthine access to the petrous apex. In this situation, axial and coronal images are carefully reviewed to determine whether the aeration and anatomy will permit a transcanal infracochlear approach.

The surgeon may also obtain a CT scan with fiducial markers in anticipation of performing surgery with image guidance technology. Intraoperative localization from an anterior approach is fairly accurate because the skull base contains numerous bony landmarks that allow registration of known anatomic sites. However, despite proposed intraoperative accuracy to less than 2 mm,[23] image guidance is not widely used in lateral skull base approaches because of technical problems with accurate registration and the fact that a 2-mm error may be catastrophic in the small operative win-dow afforded by the infracochlear or infralabyrinthine approaches.

Large tumors within the petrous apex that approximate or encase the carotid artery warrant further evaluation. MR angiography is useful in preoperative planning to evaluate the anatomic relationship with the vessel. It must be determined whether blood flow from the contralateral side through the circle of Willis is adequate if it is expected that the ipsilateral carotid artery may be retracted, temporarily occluded, or resected during the procedure. Balloon test occlusion of the ipsilateral ICA provides reliable data for predicting the patient's tolerance to ICA manipulation or sacrifice; however, 15% of patients with normal collateral supply may still experience a stroke if the artery is sacrificed.[24]

It is important to reach a presumptive preoperative diagnosis based on physical examination and radiologic imaging findings to provide the surgeon with the ability to plan both the surgical approach and optimal management of the lesion. The specific surgical approach depends on the presumed pathology; the location of the tumor or cyst; the status of hearing, balance, and facial nerve function; the likelihood of encountering CSF; and the integrity of the carotid artery and the middle and posterior fossa dura. Depending on the nature, location, and extent of the pathology, the likelihood of encountering CSF, the patient's hearing status, and facial nerve involvement, the surgeon can anticipate whether total resection or exteriorization is advisable. Meticulous preoperative planning allows more aggressive and complete resection of recurrent tumors, thereby decreasing the need for salvage surgery.

The need for intraoperative facial nerve monitoring is dictated by the proximity of the pathology to the facial nerve and the likelihood that dissection, drainage, or resection of the tumor or cyst may place the facial nerve in jeopardy. As with most otologic procedures performed with the patient under general anesthesia, paralytic agents are avoided. General anesthesia is maintained with inhalation agents, narcotics, and hypnotics. Blood is typed and cross-matched for transfusion or collected for autotransfusion if the carotid artery is to be manipulated. The goals and risks of the procedure along with the limited alternatives are reviewed with the patient and family.

SURGICAL MANAGEMENT

Cholesterol Granuloma

Management of cholesterol granuloma and epidermoid tumor is different. In contrast to a true cyst, the walls of a cholesterol granuloma lack an epithelial lining. It is therefore unnecessary to remove the capsule entirely to successfully prevent recurrence or expansion of the granuloma. As long as adequate drainage and ventilation are maintained, the cyst should not recur. That said, surgery is not always warranted. Because of the indolent clinical course of cholesterol granuloma, small, asymptomatic, or inaccessible cysts with tolerable symptoms can be managed expectantly with frequent clinical follow-up and serial radiologic imaging. Several published series include long-term follow-up of patients managed expectantly, and quite often the clinical symptoms and cyst dimensions remain stable.[11,25]

The specific surgical treatment of cholesterol granuloma of the petrous apex depends on both the patient's hearing status at the time of surgery and the anatomic relationship of the tumor with surrounding neurovascular structures. For patients with nonserviceable hearing, good surgical exposure becomes more important than preservation of residual cochleovestibular function (Table 126-2). Thus, when sensorineural hearing loss is greater than 70 dB and word discrimination is less than 30%, cholesterol granuloma is drained via a translabyrinthine or combined translabyrinthine-transcochlear approach. The translabyrinthine ap-proach provides greater exposure and drainage than the retrofacial infralabyrinthine approach alone does. Dissection through the temporal bone superior and inferior to the IAC increases ventilation to the petrous apex. If additional exposure is needed, the cochlea is sacrificed.

Table 126-2 -- DECISION MAKING FOR MANAGEMENT OF CHOLESTEROL GRANULOMA AND EPIDERMOID TUMOR

Cholesterol Granuloma			
Good Hearing		Poor Hearing	Asymptomatic/Tolerable Symptoms with Difficult Access
Complete Excision	***Drainage***		
No Stent	**With Stent**	**With or without Stent**	
Middle + posterior fossa Middle fossa + transpetrosal	Infracochlear Infralabyrinthine Middle fossa Transsphenoidal	Translabyrinthine Transcochlear	Observe Serial magnetic resonance imaging or computed tomography
Epidermoid Tumor			
Preservation of Hearing		**Sacrifice of Hearing**	
Canal wall down mastoidectomy Preauricular subtemporal + infratemporal Middle fossa + occipital		Translabyrinthine Transcochlear Transotic	

There are two management philosophies for treatment of patients with serviceable hearing. Some surgeons argue that total excision of the cyst wall best prevents reaccumulation of the cyst's contents. The approach can involve a combination of middle, posterior, or infratemporal fossa techniques for access. Recurrence rates are low, and stenting is not necessary when the entire cyst is removed.[25] The drawback is the possibility of entry through the dura with subsequent potential complications, including CSF leak and chemical meningitis. However, most authors, including ourselves, favor continuous drainage of the cyst into the pneumatized mastoid or middle ear cavities with placement of a permanent stent. Such drainage can be accomplished through the nose into the sphenoid sinus and nasopharynx. The transtemporal approaches of choice are the transmastoid infralabyrinthine and the transcanal infracochlear. Since 1990 the transcanal infracochlear approach has been supplanting the infralabyrinthine approach because the infracochlear approach for drainage of the cyst is achievable even in the presence of a high jugular bulb.[26] The success and complication rates of the two surgeries are similar. Overall, one can expect symptomatic relief in 80% of patients with minimal complications.[27] The drawback of long-term drainage with permanent stenting is a high recurrence rate of 15% to 60%, usually associated with obstruction of the stent.[28] The older literature described the highest recurrence rate of 60% in patients drained via a transethmoid, transsphenoid approach.[28] A recent retrospective series by Brackmann and Toh is very encouraging for transtemporal drainage procedures; a recurrence rate of only 3% was reported for cases in which a stent was placed.[27]

A middle fossa or keyhole middle fossa approach may produce adequate exposure but is not recommended because it is technically difficult to maintain cyst ventilation/drainage into the mastoid cavity or eustachian tube via stenting.[29] However, if establishment of drainage is successful, recurrence rates are similar to those of the infralabyrinthine approach.[27] A middle fossa approach with stenting is a valid option.

Transsphenoid drainage is an alternative to consider in cases in which the cholesterol granuloma abuts the posterior wall of the sphenoid sinus. This technique, originally described by Montgomery in 1977, was performed through a contralateral external ethmoidectomy approach.[30] Knowledge of the surgical anatomy plus the use of contemporary instrumentation such as endoscopic sinus telescopes permits endonasal access to the petrous apex tip directly through the sphenoid sinus. However, inability to maintain a patent fistula through this approach may result in recurrence of the cystic lesion. It is possible to enhance drainage of the cyst when intraoperative

navigation is used in conjunction with endoscopy; more aggressive drainage may be achieved while avoiding damage to vital structures lying behind the sinus wall.[31]

The development of minimally invasive endoscopic neurosurgery may provide a valid option in the near future for resection of petroclival tumors. The expanded endonasal approach provides excellent access to petrous apex tumors and has been applied to drainage procedures for cholesterol granuloma and other tumors of the anterior petrous bone. At this juncture the lack of sufficient follow-up makes it difficult to predict the appropriateness of this type of approach for tumors of the petrous apex. The procedure includes bilateral sphenoidotomy and posterior maxillary antrostomy with drilling of the petrous bone encasing the ICA. Lateral mobilization of the carotid artery can be performed to gain greater access to the petrous apex. Marsupialization of the granuloma into the sphenoid is performed. Kassam and colleagues, experienced in resection and debulking of petroclival tumors via the expended endonasal approach, propose to use this method for resection of cholesteatoma in addition to cholesterol granuloma.[32] It remains to be seen whether the newer techniques of endoscopic nasal surgery will provide more permanent drainage and lower the incidence of recurrence.

Epidermoid of the Petrous Apex

In contrast to cholesterol granulomas, epidermoid tumors must be excised completely; if tumor is left behind, the cyst will recur. However, the tumor is often tightly adherent to surrounding neurovascular structures and dura. It is believed that microrupture of the capsule may occur and induce an inflammatory reaction that binds the tumor to surrounding structures. Complete resection bears considerable risk because epidermoid tumors frequently encase vital structures. At the time of initial examination by an otolaryngologist, cranial nerves are involved in the majority of patients (85%). Hearing loss, vestibular deficits, and facial weakness are the most common, followed by deficits in function of cranial nerves V and VI. In addition to affecting nerves, tumors often involve major vessels (80%), including the carotid artery in 50% of cases.[33]

It is important to completely excise the tumor despite its intimate involvement with neurovascular structures. Recurrence is likely when the surgeon tries to preserve cranial nerve function and leaves epidermoid lining in these areas. Ultimately, cranial nerve deficits are not spared because salvage procedures will probably result in additional morbidity and sacrifice. In summary, the goal of resection of epidermoid tumors is safe but complete removal of tumor, which is facilitated by thorough preoperative planning and excellent surgical exposure.

For wide exposure of the petrous apex, we recommend the translabyrinthine approach. If the surgeon needs good exposure of the carotid artery, conversion to a transcochlear approach is possible. In the transotic procedure described by Fisch, the EAC is closed, the tympanic membrane and ossicles are removed, and the facial nerve is transposed anteriorly.[34] In the transcochlear approach originally described by House and colleagues, the facial nerve is displaced posteriorly.[35] The drawbacks of these approaches are sacrifice of cochlear and vestibular function, risk of CSF leak, and facial nerve palsy secondary to mobilization of the facial nerve to improve access, which may result in facial weakness in up to 75% of cases.[36] To avoid such complications, some surgeons advocate a middle fossa approach.

In the presence of intact cochlear function, we prefer to manage congenital epidermoid tumors with a canal wall down procedure if adequate exposure and exteriorization of disease can be achieved. This requires a well-pneumatized temporal bone with a high tegmen tympani. Unfortunately, congenital epidermoid tumors often originate in poorly pneumatized, contracted temporal bones, thus making it difficult to adequately exteriorize the tumor with the cochlea intact. Exposure is enhanced by sacrificing the cochlea and therefore hearing.

Occasionally, wide marsupialization of an epidermoid tumor is inadequate or unsafe. Total resection becomes necessary when exteriorization results in a CSF leak, when the dura or the carotid artery is exposed and at risk for desiccation in an open cavity, or when the opening of the marsupialized epidermoid cavity results in a very narrow, circuitous tract that makes postoperative examination and débridement difficult. In this situation, the entire tumor and its epithelial lining are removed and the wound is obliterated.

Surgical Techniques

Tympanomeatal Approach for Congenital Middle Ear Cholesteatoma

This approach is directed at isolated congenital middle ear cholesteatoma characteristically located in the anterosuperior quadrant of the middle ear. The patient is positioned supine on the operating room table with the head turned toward the contralateral ear. Using the operating microscope, the ear canal is injected in four-quadrant fashion with 2% lidocaine (Xylocaine) with 1:100,000 epinephrine. In young children, the anesthetic solution is diluted by half. After adequate local anesthesia and vasoconstriction have been achieved, a tympanomeatal flap based inferiorly is elevated. An incision is made lateral to the annulus from the 3- to the 9-o'clock positions. The tympanomeatal flap is elevated to enter the middle ear in the posterosuperior quadrant. The tympanic membrane, including the pars flaccida, is carefully dissected off the scutum and over the short

process of the malleus. A sharp sickle knife or curved pick is used to incise the thin layer of mucoperiosteum along the posterior aspect of the malleus handle. The tympanic membrane is retracted inferiorly toward the umbo to expose the epithelial tumor (Fig. 126-8). The extent of inferior dissection of the tympanic membrane is dictated by the size and location of the mass. Using suction, right-angled picks, and curved excavators, the tumor is carefully freed from its attachments to bone and ossicles and removed (Fig. 126-9). Great care is taken to remove the epithelial pearl intact and avoid spillage of keratin in the middle ear and mastoid. Once the pearl is removed, the ossicular chain is examined for integrity and mobility, and the tympanic membrane is carefully inspected for a tear or perforation. If a defect is identified, it is repaired with a small piece of wet Gelfoam or an underlay connective tissue graft placed medial to the area. The tympanic membrane is returned to its anatomic position. A silk sleeve is placed over the tympanic membrane and supported by cotton balls with ointment. The ear canal may also be packed with Gelfoam moistened with antibiotic ear drops.

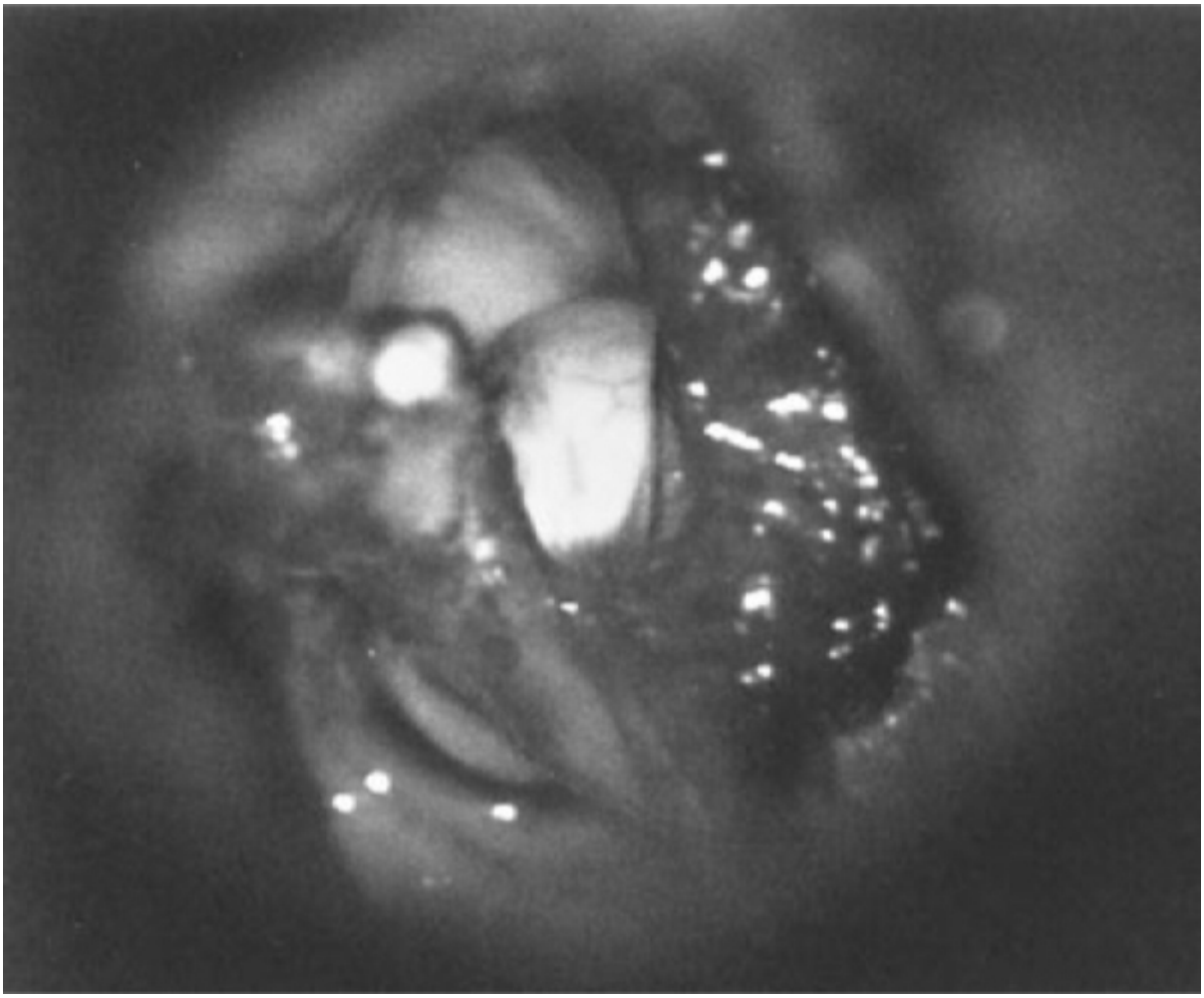

Figure 126-8 An inferiorly based tympanomeatal flap attached to the umbo exposes the encapsulated keratin pearl.

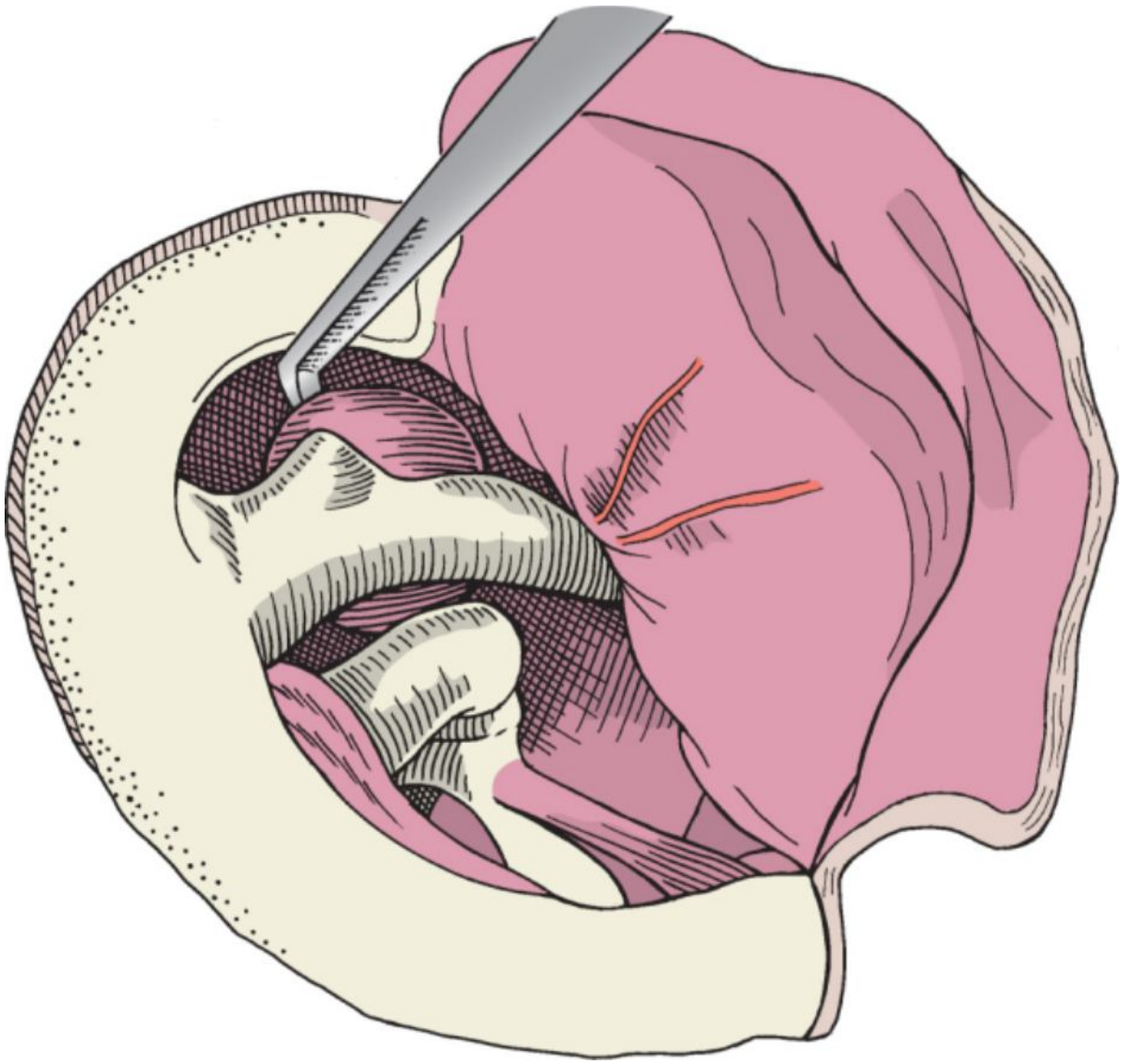

Figure 126-9 Right-angled picks and excavators are used to remove the keratin pearl intact.

More extensive primary cholesteatoma occupying the epitympanum may require additional exposure via a cortical mastoidectomy. Drilling is continued anteriorly through the root of the zygoma to expose the body of the incus and head of the malleus. Tumor located in this area frequently creates a conductive hearing loss. Removal of the incus and possibly the head of the malleus may be necessary to obtain access to the anterior epitympanum. The canal wall may have to be taken down to exteriorize any epitympanic disease that cannot be completely removed. Reconstruction of the ossicular chain is reviewed in Chapter 114. When the epithelial lining of the cyst ruptures into either the middle ear or the EAC, primary cholesteatoma of the middle ear becomes difficult to distinguish from acquired chronic otitis media with cholesteatoma. Surgical management is dictated by the extent and location of the cholesteatoma matrix within the middle ear and mastoid. This subject is reviewed in Chapter 115.

Transcanal Infracochlear Approach

Because of the limited opening into the petrous apex, this drainage procedure is better suited for treatment of cholesterol granuloma than epidermoid tumor. This approach is preferred when a high jugular bulb inhibits adequate exposure via the retrofacial infralabyrinthine approach. It has the advantage that problems arising from blockage of a Silastic shunt tube can be addressed through an inferior myringotomy.[3]

After induction of general anesthesia, the head is turned toward the contralateral ear. The facial nerve is electrically monitored throughout the procedure. Under the operating microscope, 2% lidocaine with 1:100,000 epinephrine is injected into the four quadrants of the EAC, as well as the postauricular skin. After adequate hemostasis is obtained, 12- and 6-o'clock incision are made from the mid–bony canal medially toward the external meatus laterally. The inferior incision stops at the conchal cartilage. These two incisions are connected vertically to create a long conchal flap. A postauricular incision is made, the posterior canal skin is elevated to the level of the endaural incisions, tracheostomy tape is placed through the meatus, and the auricle is reflected anteriorly.

Tympanomeatal incisions are made at the 10-o'clock position (right ear) and brought laterally and obliquely from the annulus. The lateral and inferior aspect of the tympanomeatal flap is incised at the prominence of the bulge in the canal wall (Fig. 126-10). Elevation of the meatal skin flap is taken down to the level of the annulus. The middle ear is entered posteriorly, and the annulus is elevated to create a flap that is based superiorly and pedicled on the umbo of the malleus. The anterior, posterior, and inferior bony canal walls are enlarged toward the hypotympanum. When the posterior canal wall is drilled, care must be taken to avoid injury to the vertical portion of the facial nerve. If the chorda tympani nerve is used as the posterior limit of dissection, injury from the drill can be avoided (Fig. 126-11). The inferior tympanic ring is drilled to expose the hypotympanic air cells. Drilling is continued inferiorly to expose the genu of the carotid artery and jugular bulb. The posterosuperior limit of dissection is the round window. A triangle consisting of the carotid artery anteriorly, the jugular bulb posteriorly, and the basal turn of the cochlea superiorly forms the area for continued dissection. From measurements in cadavers, the size of the operating window averages 9.4 × 7.3 mm. Sometimes the space between the great vessels is small, less than 5 mm, thus restricting the usefulness of this approach for biopsy and drainage only rather than resection of cholesteatoma.[26] Cutting and then diamond burrs are used to continue the dissection in a medial and slightly superior direction (Fig. 126-12). This brings the dissection inferior to the cochlea and anteroinferior to the IAC.

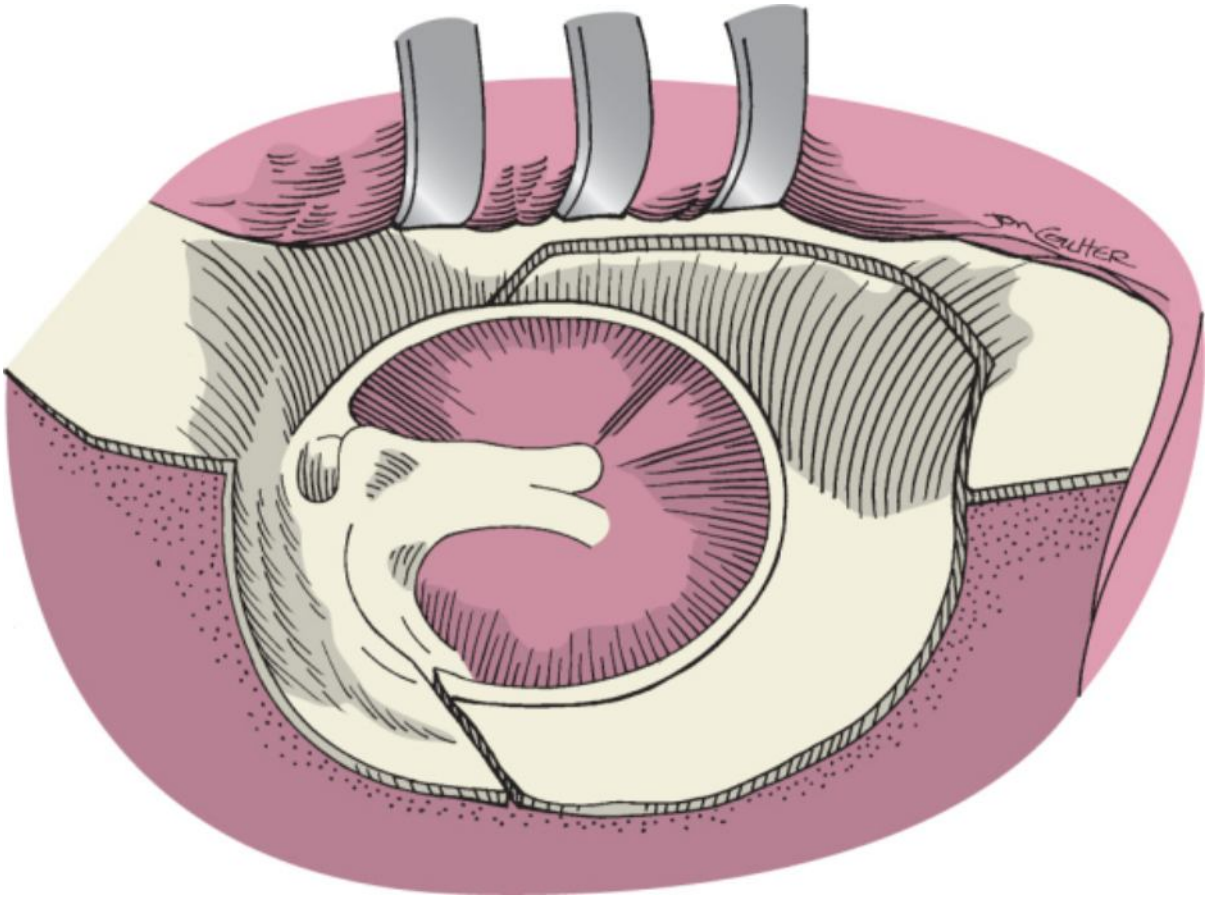

Figure 126-10 The superiorly based tympanomeatal flap is incised. The prominent anteroinferior bony canal wall will be removed.

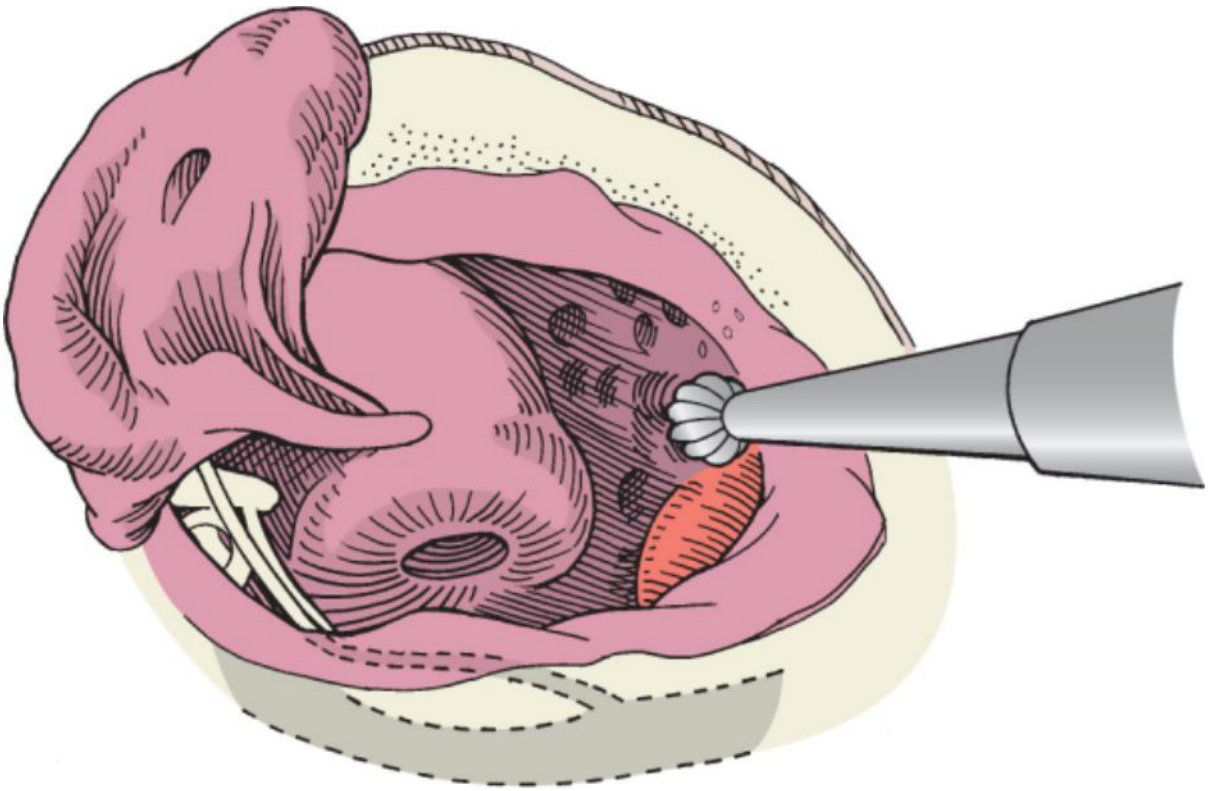

Figure 126-11 The superiorly based tympanomeatal flap is elevated to expose the mesotympanum and hypotympanum. A cutting burr is used to remove the prominence of the lateral canal wall and the inferior tympanic ring for further exposure of the hypotympanum. Care is taken posteriorly to avoid the vertical portion of the facial nerve.

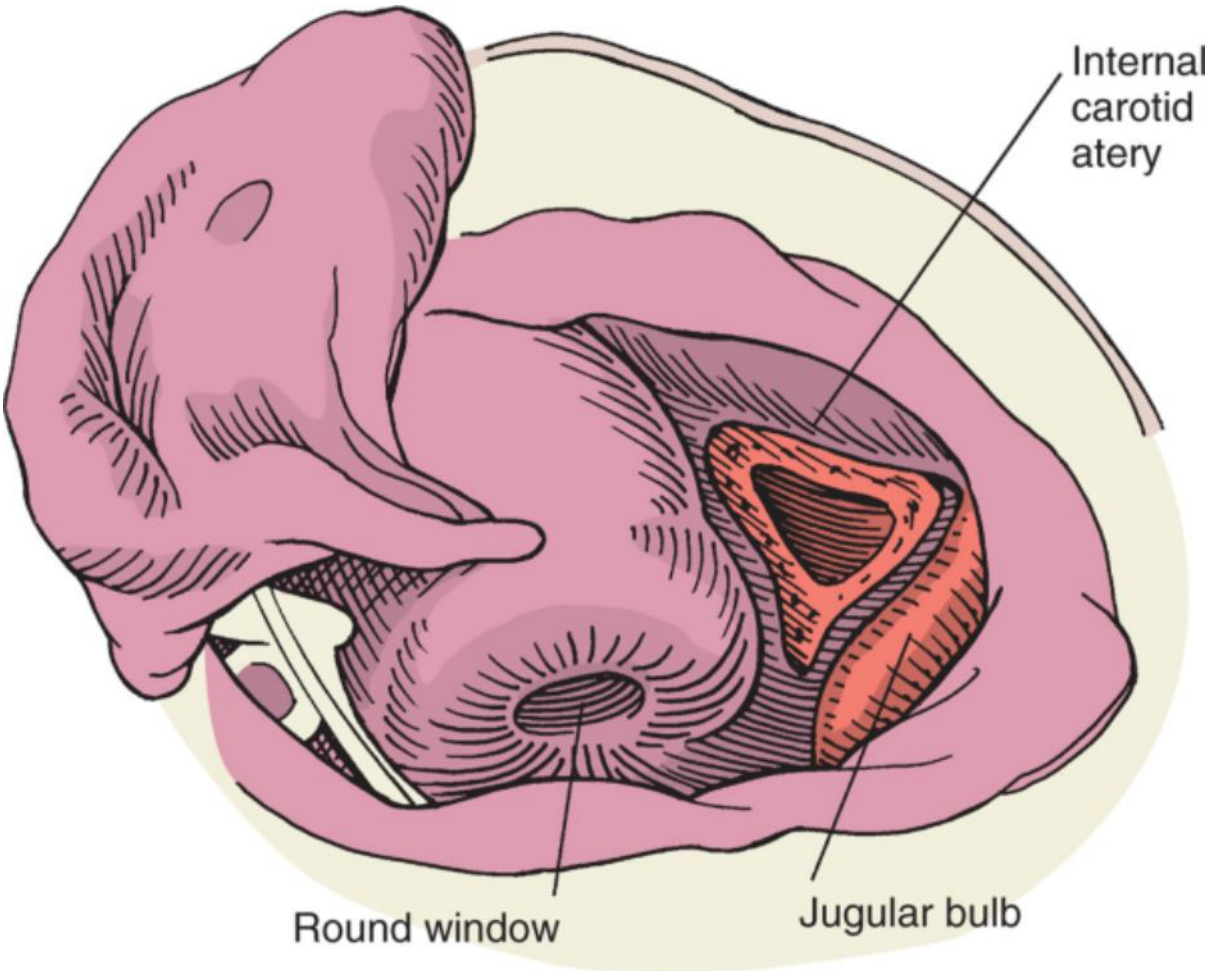

Figure 126-12 A diamond burr is used to remove the hypotympanic air cells. The limits of dissection are the internal carotid artery anteriorly, the jugular vein posteroinferiorly, and the basal turn of the cochlea superiorly.

The air cells lateral to the cholesterol granuloma often appear dark. Once the cyst is entered, the characteristic thick, yellow-brown fluid and cholesterol crystals are encountered. The fistula opening into the cyst is enlarged while keeping the carotid artery and jugular vein in view. Cup forceps are used to remove some of the lateral inferior lining of the cyst wall. The cavity is evacuated of its fluid contents. The size of the opening is maximized by drilling to the limits of the carotid artery, jugular bulb, and basal turn of the cochlea. The cavity is irrigated with bacitracin-saline solution. If the cyst is multiloculated and the surgical approach gives limited exposure, endoscopy may be helpful. A sinus (2.7 mm) or otologic (1.9 mm) endoscope may facilitate visualization of hidden parts of the cystic lesion. In combination with suction and irrigation, endoscopy facilitates débridement and efficient drainage of septated cholesterol granuloma or eradication of cholesteatoma limited to the inferior petrous apex.[37]

The area is stented and drained with silicone or Silastic tubing. If not available, Silastic sheeting (0.02 inch) is cut in a rectangular shape and rolled to make a cylinder (Fig. 126-13). The inferior and lateral aspect of the Silastic tubing is trimmed so that it is contained completely within the middle ear and hypotympanic spaces. Gelfoam impregnated with an antibiotic suspension is placed in the newly created hypotympanum and along the medial floor of the new bony EAC. The inferior aspect of the tympanomeatal flap is returned to its anatomic position. A small graft of fascia positioned medial to the tympanomeatal skin flap and extended laterally toward the ear canal may be needed to cover a residual defect. Gelfoam with antibiotic suspension is placed lateral to the tympanic membrane and along the flap of the meatal skin. The auricle is returned to its anatomic position while ensuring that the posterior conchal skin flap is laid flat against the posterior bony canal wall. The lateral cartilaginous canal is packed with lateral rosebud packing, Gelfoam, or a Merocel sponge. The postauricular incision is closed with 4-0 resorbable suture braided subcutaneously and 5-0 fast-absorbing suture in the skin and covered with Steri-Strips, and a sterile mastoid dressing is applied.

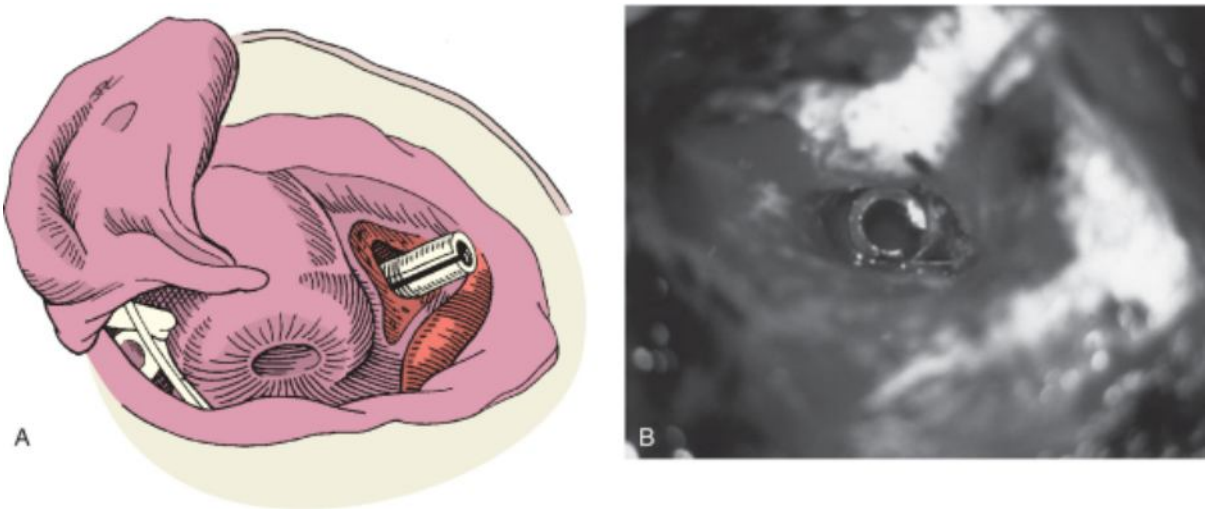

Figure 126-13 **A,** A rolled Silastic sheet is placed in the opening of the fistula from the cystic cavity to the hypotympanum. **B,** The Silastic sheeting in place.

The transcanal infracochlear approach requires exposure of the sigmoid sinus and drilling under the cochlea, which can potentially lead to complications. A modification was proposed in which only exposure of the ICA is required and a surgical window is drilled anterior to the cochlea, thereby presumably reducing the risk of injury to the jugular bulb and round window. The tradeoff for increased safety is a smaller working space averaging 4.7 × 3.2 mm instead of 7.3 × 9.4 mm with a traditional infracochlear approach.[38]

Retrofacial Infralabyrinthine Approach

The patient is positioned supine on the table with the head turned toward the contralateral ear. One centimeter of postauricular hair is shaved, and the ear is prepared in usual fashion. Two percent lidocaine with 1:100,000 epinephrine is infiltrated into the skin 2 cm posterior to the postauricular sulcus. A curvilinear incision is made through the skin. Fibroperiosteal tissue is elevated from the mastoid cortex, and self-retaining retractors are placed.

A complete transcortical mastoidectomy is performed. The sigmoid sinus is decompressed, and 1 cm of bone

posterior to the edge of the sinus is removed. The horizontal semicircular canal is delineated. The aditus ad antrum is opened as widely as possible to maximize ventilation into the epitympanic and middle ear spaces. Further ventilation through the middle ear is accomplished by opening the facial recess if communication between the aditus ad antrum of the mastoid and the epitympanum of the middle ear is insufficient. The posterior semicircular canal is outlined with a diamond drill. The vertical portion of the facial nerve is defined, and a thin covering of bone is left for protection. Additional bone is removed to expose the mastoid tip and digastric ridge.

Using a cutting burr, drilling is continued through the air cells posterior to the vertical portion of the facial nerve. Dissection is carried in an anteromedial direction. The continuation of the sigmoid sinus into the jugular bulb must be defined. To obtain adequate access through this route, a high jugular bulb must not be present. This anatomic variation would have been identified on preoperative bone-windowed CT scanning. In patients with a high jugular bulb, the procedure can be converted into an infracochlear approach.[26] Posterior compression of the sigmoid sinus enhances the exposure and angle necessary to gain access to the petrous apex cyst. Once it is opened, the contents of the cyst are evacuated. Cup forceps are used to completely remove the lining of the posterolateral cyst wall. This opening is maximized through the confines of the area limited by the facial nerve anteriorly, the ampullated end of the posterior semicircular canal superiorly, the posterior fossa dura posteriorly, and the jugular bulb inferiorly.

The cavity is irrigated with bacitracin solution. A Silastic tube or sheeting rolled into a cylinder is placed in the cavity and trimmed to an appropriate length so that it lies within the inferior aspect of the mastoidectomy (Fig. 126-14). The postauricular incision is closed with 4-0 resorbable braided suture, and a sterile mastoid dressing is applied.

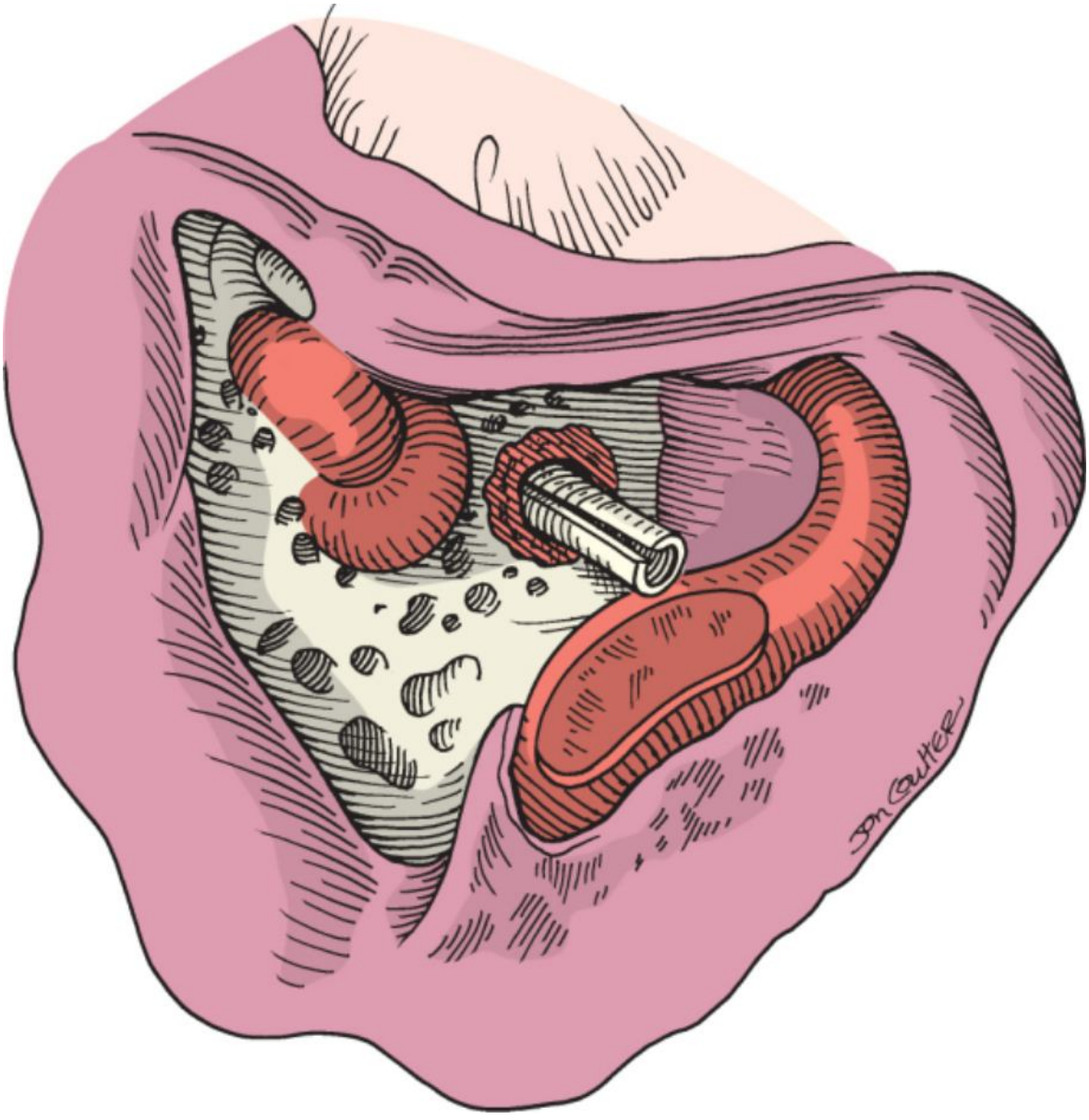

Figure 126-14 Retrofacial infralabyrinthine approach. A transcortical mastoidectomy is performed to identify the antrum, posterior semicircular canal, facial nerve, digastric ridge, and jugular bulb. A Silastic tube from the mastoid into the cyst provides drainage and aeration.

Translabyrinthine Approach

The translabyrinthine approach for drainage of the petrous apex is identical to that for resection of an acoustic neuroma (Chapter 124). This approach is undertaken if hearing is significantly compromised or absent. Bone is removed 1 cm posterior to the sigmoid sinus, and the middle fossa tegmen is thinned. A complete labyrinthectomy is performed. The IAC is carefully outlined while taking care to not enter the dura of the IAC, the posterior fossa, or the middle fossa.

The incus is removed, and the fossa incudis is drilled to maximize communication between the mastoid and the epitympanic space. A trough is created superior to the IAC if the bony anatomy permits. Entrance into the petrous apex can be achieved through suprameatal air cells, if present. Care must be taken to avoid injury to the facial nerve in its labyrinthine segment.

The bone inferior to the IAC is removed with a diamond burr (Fig. 126-15). The location of the jugular bulb is noted. Caution should be exercised when working in the area superior to the jugular bulb and inferior to the IAC. The cochlear aqueduct enters the vestibule in this area, and entry into the cochlear aqueduct results in leakage of CSF into the wound. This is difficult to suture and may need to be plugged with fat or muscle. If it cannot be occluded in watertight fashion and sealed from potential communication with the cyst, the cholesterol granuloma drainage procedure must be terminated. Abdominal fat is packed into the cavity, and muscle is plugged into the epitympanum.

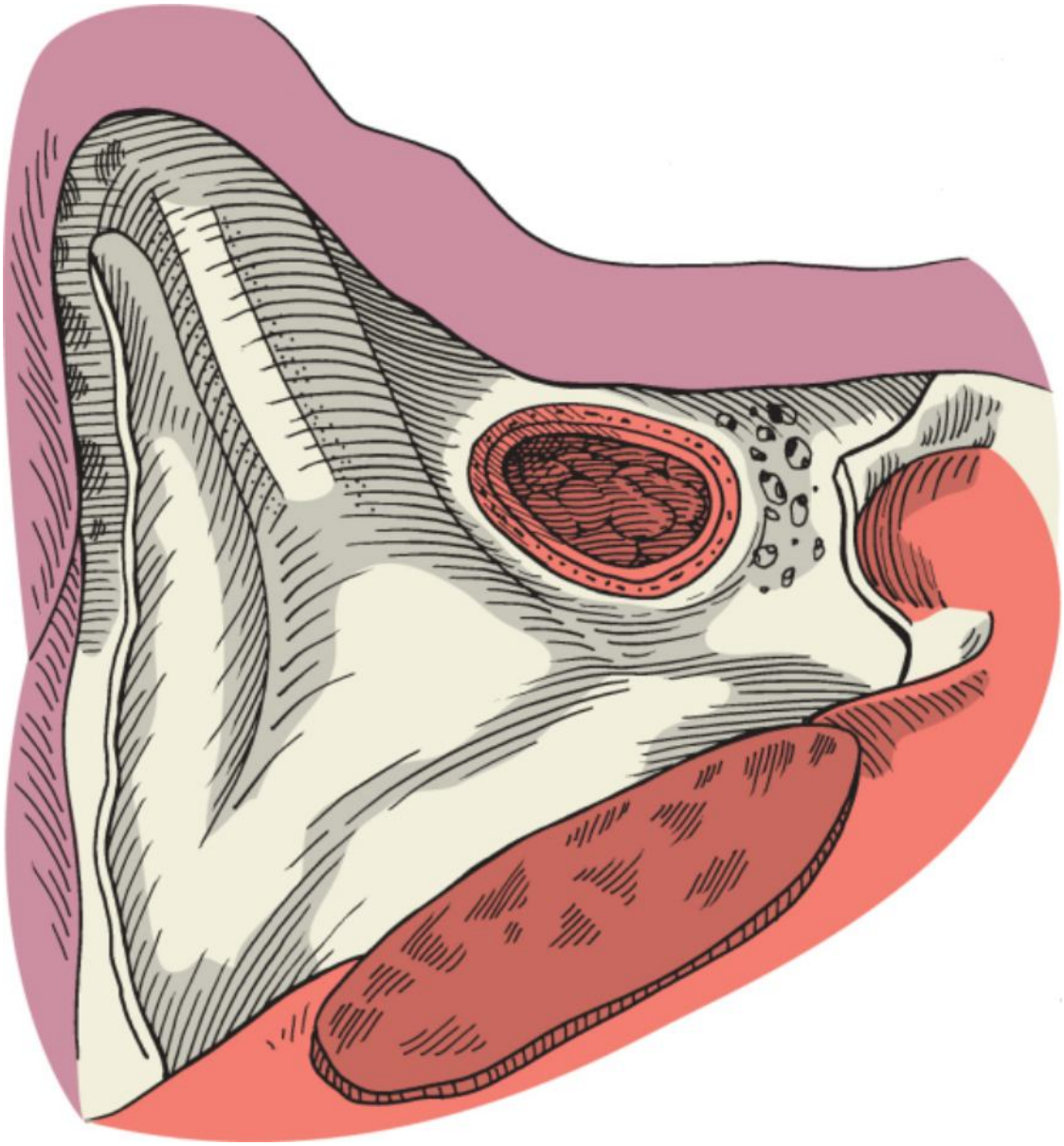

Figure 126-15 Translabyrinthine approach to the petrous apex. The IAC is skeletonized to create an inferior and superior trough around the porus acusticus. Care is taken to avoid the cochlear aqueduct and jugular bulb. The labyrinthine segment of the facial nerve is noted.

The inframeatal exposure of the petrous apex should drain and ventilate the cyst adequately. More aggressive removal of cyst lining is possible with wider exposure. Drainage with a catheter or Silastic sheeting is unnecessary if a large opening in the fistula is created. The wound is copiously irrigated with bacitracin solution. The skin is closed in two layers, and a sterile mastoid dressing is applied.

Transcochlear Approach

The transcochlear approach is reserved for patients who have a large tumor of the petroclival region with preexisting hearing loss. It is a modification of the translabyrinthine approach in which the cartilaginous EAC is closed in a cul-de-sac. A canal wall down mastoidectomy is then performed with unroofing of the sigmoid sinus and exposure of the dura of the posterior and middle fossae. The IAC is identified and the facial nerve is decompressed from the stylomastoid foramen to the IAC. The facial nerve is dissected free from the fallopian canal and transposed posteriorly. This requires identification and sacrifice of the greater superficial petrosal nerve (GSPN). After liberation of the geniculate ganglion, the labyrinthine, tympanic, and vertical portions of the facial nerve are successively released and transposed posteriorly toward the dura of the posterior fossa. The nerve is immobilized with fibrin glue. The cochlea and fallopian canal are then drilled to expose the vertical portion of the ICA. Additional anterior exposure requires drilling the anterior wall of the EAC and displacing the mandibular condyle anteriorly. The horizontal portion of the IAC is then exposed. At the end of the procedure, the eustachian tube is plugged with muscle, Surgicel, and fat harvested from the abdomen, which is then covered with a temporalis myofascial flap.

Although this approach is fairly safe and allows good exposure, it is frequently complicated by cranial nerve palsy. For instance, in a review of 66 cases, every patient in whom the facial nerve was transposed had facial nerve weakness, with only 5% of patients recovering normal facial nerve function 1 year after the procedure. Most of the patients (60%) recovered to House-Brackmann grade III function. Other postoperative deficits included weakness of cranial nerves IV (6%) and VI (9%).[39]

Tympanomastoid Exteriorization

A petrous apex epidermoid tumor or cholesterol granuloma can be marsupialized or drained directly into the tympanic space. This approach almost always entails a canal wall down mastoidectomy. Whether the ossicular chain can be reconstructed depends on the location of the disease, the status of the hearing preoperatively, and the procedure that was performed.

The patient is positioned supine on the operating table with the head turned toward the contralateral ear. The supra-auricular and postauricular hair is shaved, and the ear is prepared and draped in the usual sterile fashion. A longer Koerner flap is made in the EAC in anticipation of a canal wall down mastoidectomy. A postauricular incision is made, and dissection is continued into the EAC. The posterior canal skin flap is elevated and retracted into the ear canal with tracheotomy tape. The auricle is retracted anteriorly. A transcortical mastoidectomy is performed. Because the approach is directed toward the anterior epitympanic area, creation of a large mastoid cavity is to be avoided. Therefore, bone from the mastoid tip and sinodural angle does not have to be extensively removed unless the temporal bone is pneumatized and mucosal disease is present.

The aditus ad antrum, horizontal semicircular canal, and incus are identified. Attention is then turned to the EAC. A tympanomeatal flap is elevated to identify and disarticulate the incudostapedial joint. The incus is removed and saved if it is not involved with cholesteatoma. The posterior bony canal wall is taken down, the anterior and posterior buttress is removed, and the facial ridge is lowered to the level of the EAC. Using a malleus nipper through the neck of the malleus, the head of the malleus is removed. Drilling and dissection are continued anteriorly through the zygomatic route. The geniculate ganglion is noted, and dissection is continued along the GSPN. If greater exposure is necessary inferiorly, the malleus and tensor tympani tendon are removed as well. Although it may permit drainage of a cholesterol cyst, the tympanomastoid approach is predominantly used to marsupialize cholesteatoma located in this area (Fig. 126-16).

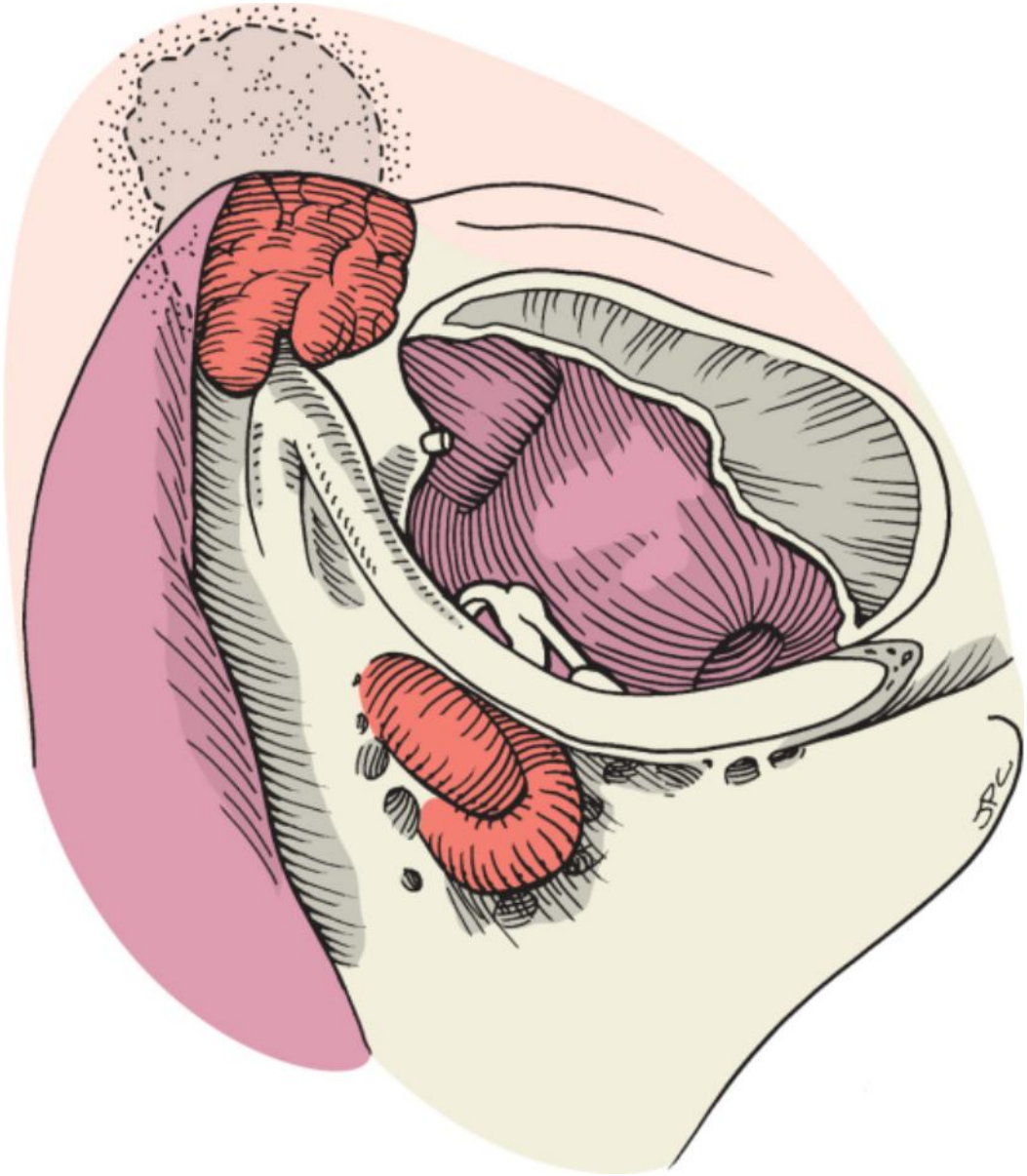

Figure 126-16 Tympanomastoid exteriorization. A canal wall down mastoidectomy is used to expose the anterior epitympanum and the cholesteatoma in the petrous apex. Dissection along the greater superficial petrosal nerve opens the petrous apex through the air cells of the zygomatic route.

The invasive and expansive nature of petrous apex epidermoid tumors is likely to cause sensorineural hearing loss. If cochlear function has been compromised preoperatively (speech reception threshold >70 dB, discrimination <30%), greater exposure is achieved by continuing the dissection through the cochlea. The inferior limits of this opening are the eustachian tube and genu of the carotid artery. Drilling is continued through and parallel to the plane of the cochlea. The proximal modiolus and lateral fundus of the IAC should be avoided to prevent a CSF leak. The contents of the petrous apex cholesteatoma are evacuated with blunt instruments such as suction, cup forceps, curettes, and smooth elevators. The epithelial matrix is usually left intact (Fig. 126-17). The cavity is copiously irrigated with bacitracin solution.

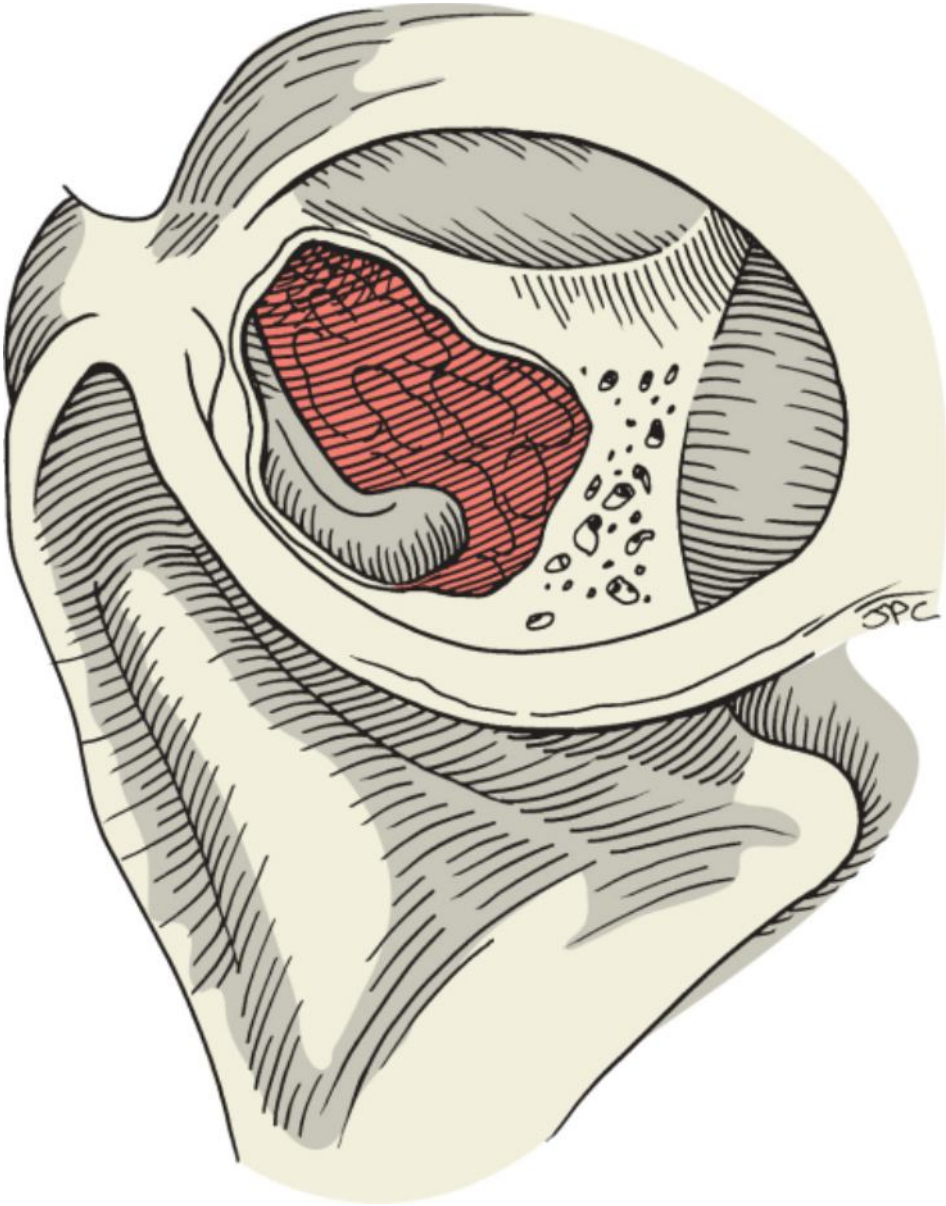

Figure 126-17 Transcochlear approach to the petrous apex. The contents of the epidermoid tumor are removed while leaving the matrix lining intact.

Despite the use of meticulous surgical technique while removing or exteriorizing an epidermoid tumor or cholesterol granuloma, CSF leakage may occur as a result of attenuation of the dura. Another situation in which a transepitympanic petrous apex exteriorization approach may need to be converted to obliteration is when the carotid artery becomes significantly exposed. Concern for desiccation of the adventitia of the carotid artery in a marsupialized cavity demands soft tissue protection and coverage. This routinely entails obliteration of the cavity with a temporalis muscle flap or abdominal fat. If the cavity must be obliterated because of a CSF leak, wide exposure of the carotid artery, or excessive exposure of the dura, great effort is taken to completely remove the wall of the granuloma or the epidermal matrix lining. The external auditory meatus is closed in a two-layer technique (see Chapter 125). An abdominal fat graft is most effective in sealing the CSF leak and obliterating the cavity. The posterior portion of the temporalis muscle is divided, rotated inferiorly, and sutured to the sternocleidomastoid muscle and subcutaneous tissue of the posterior skin.

If cochlear function was not sacrificed, reconstruction of the ossicular chain is undertaken. These techniques are reviewed in Chapter 114. When performing an exteriorization procedure, the epidermoid cavity is packed with continuous gauze impregnated with antibiotic ointment. The packing is placed through the external auditory meatus and positioned in the depth of the epidermoid cavity. The packing is continued through the epitympanic space and mastoid cavity to secure the Koerner flap posteriorly. The postauricular wound is closed with 4-0 resorbable braided suture subcutaneously and 5-0 fast-absorbing suture in the skin supported by Steri-Strips, and an external mastoid dressing is applied.

Preauricular Subtemporal Infratemporal Approach

This approach is used when complete excision of cholesterol granuloma or epidermoid tumor of the petrous apex is necessary. If normal cochlear function is present preoperatively, a hearing conservation procedure is warranted. The patient is positioned supine on the table with the head turned toward the contralateral ear. Should lateral neck rotation be limited, an ipsilateral shoulder roll is placed to provide additional rotation. A strip of hair is shaved from the lateral side of the scalp. The head is placed in three-point fixation with pins using a Mayfield head holder. The preauricular incision is outlined in curvilinear fashion toward the hairline of the forehead (Fig. 126-18). A solution of 1% lidocaine with 1 : 100,000 epinephrine is injected into the planned incision. The skin is incised, and the dissection is taken down to the level of the temporal fascia. Anterior and posterior skin flaps are elevated. Anteriorly, the frontal or temporal branch of the facial nerve must be preserved by incising the deep temporalis fascia in the area just superior to the temporal fat pad. Dissection is continued anteroinferiorly beneath the level of the deep temporalis fascia to protect the frontalis branch within the anterior skin flap from transection, although it is still subject to being stretched (Fig. 126-19). The root and arch of the zygoma are palpated and isolated by incising the periosteal attachments superior and inferior to the arch. Using a Freer or Adson elevator, the medial surface of the zygomatic arch is freed of its muscular attachments.

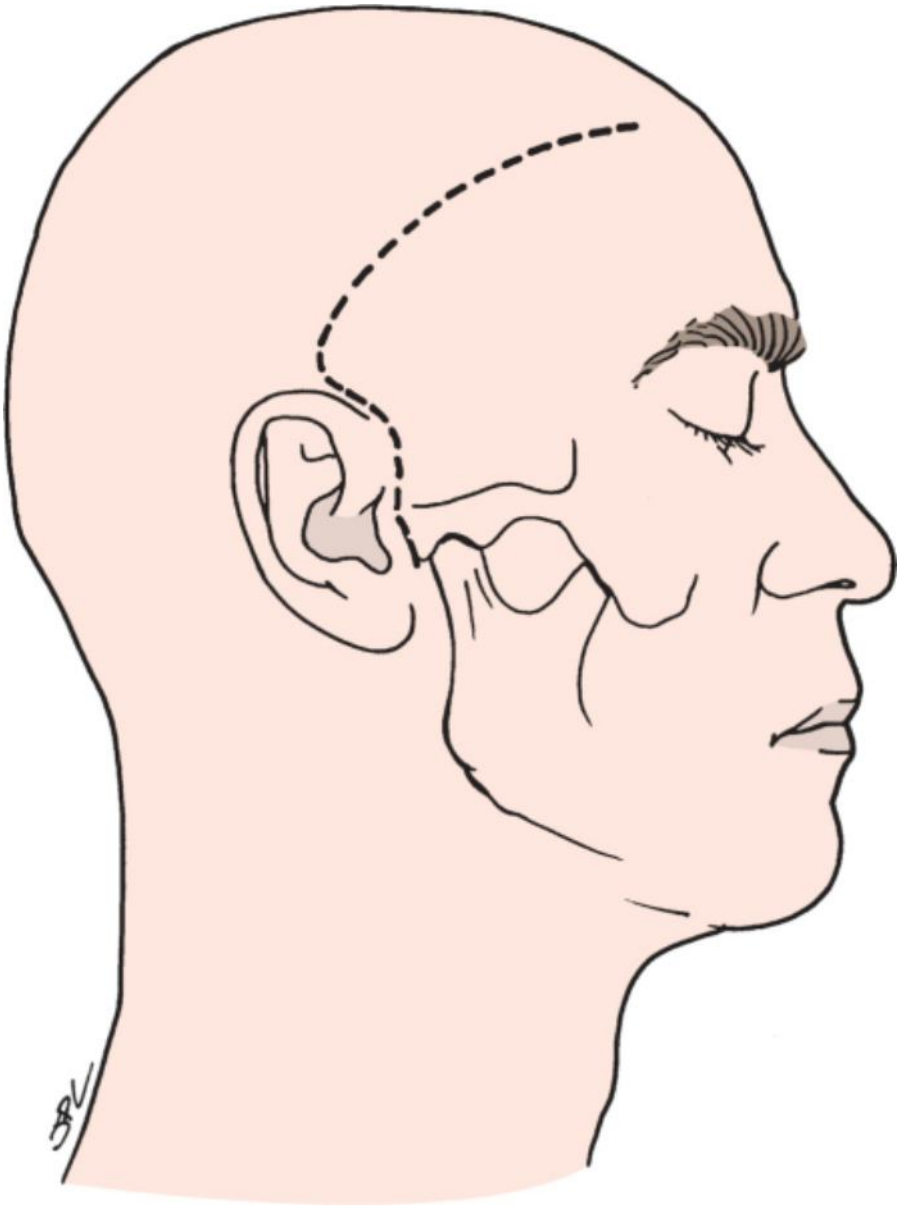

Figure 126-18 Incision for a preauricular subtemporal infratemporal fossa approach.

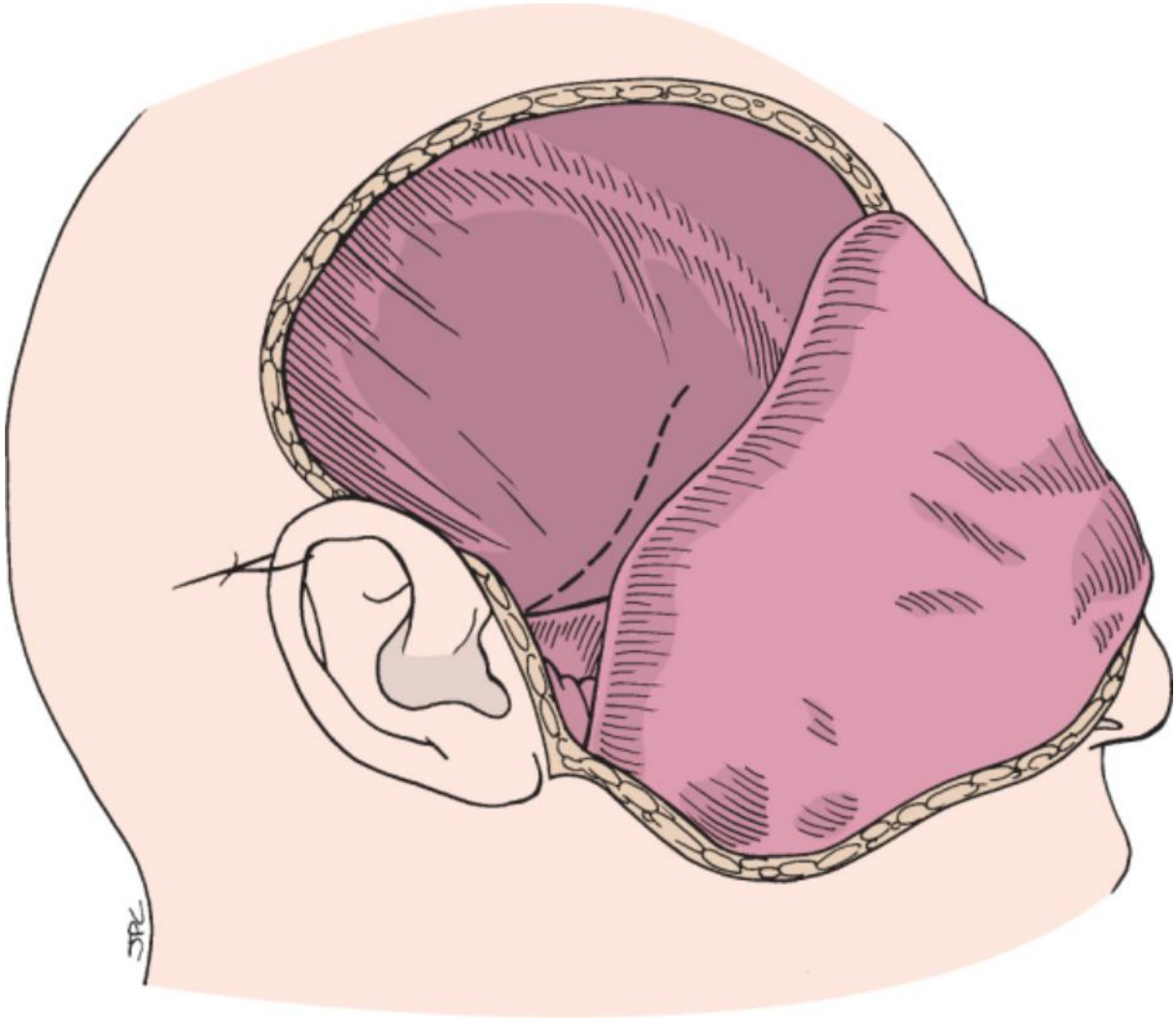

Figure 126-19 The anterior flap is elevated. The frontal branch of the facial nerve is preserved by incising the superficial and deep layers of the temporalis fascia superior to the level of the temporal fat pad.

The attachment of the temporalis muscle is incised along its borders of insertion. A margin of the fibrous attachment along the superior temporal line is preserved to facilitate subsequent reapproximation and wound closure. The temporalis muscle is incised posteriorly, superiorly, and anteriorly and elevated from the temporal fossa. The inferiorly based temporalis muscle flap is dissected well into the infratemporal fascia.

The zygomatic arch containing the lateral glenoid fossa and part of the body of the zygoma are removed as one unit. This permits further retraction of the inferiorly pedicled temporalis muscle flap and provides greater exposure of the infratemporal fossa (Fig. 126-20).

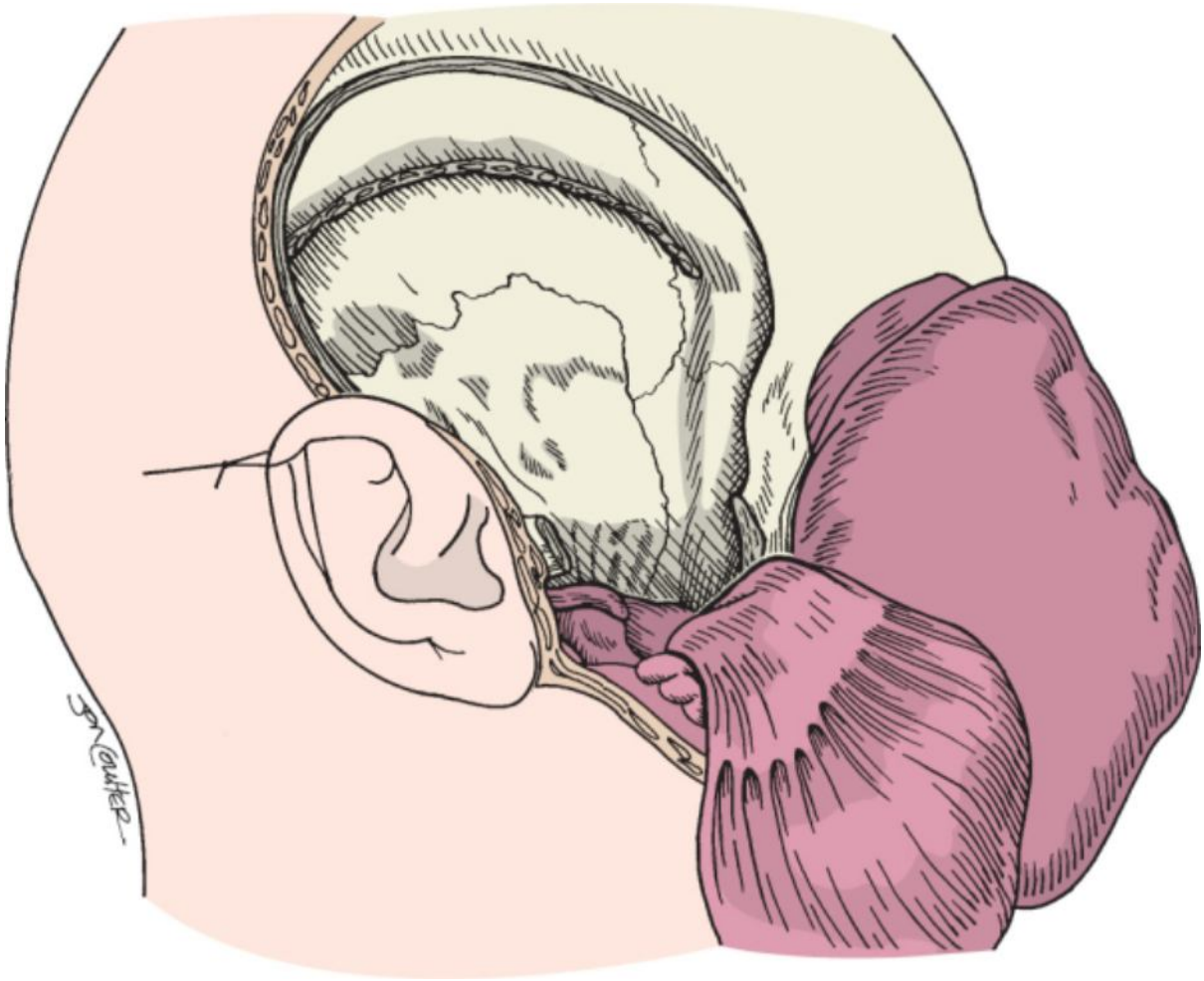

Figure 126-20 The zygomatic arch and glenoid fossa are removed. The temporalis muscle, pedicled on its origin, is retracted inferiorly.

If access to the vertical portion and genu of the carotid artery is necessary, resection of the mandibular condyle is undertaken. Soft tissue surrounding the mandibular condyle is elevated. Venous bleeding is often encountered but is controlled with bipolar coagulation. The neck of the condyle is freed of its periosteum, and the bone is cut with a reciprocating or sagittal saw. If proximal exposure of the carotid artery is not necessary, the capsule and meniscus of the temporomandibular joint are dissected from the glenoid fossa, and the condyle is retracted inferiorly.

The subtemporal approach is continued by performing a temporal craniotomy centered over the infratemporal fossa. The inferior aspect of the craniotomy should be made as low as possible (Fig. 126-21). The base of the skull consisting of the inferior squamous portion, part of the petrous portion, and the posterior extent of the sphenoid bone is further removed with a cutting burr. The middle fossa dura is elevated medially in a posterior-to-anterior direction to the level of the superior petrosal sinus. The temporal lobe is gently elevated and retracted superiorly with a Greenberg retractor. The middle meningeal artery is identified and divided via bipolar coagulation. The third or mandibular division of the trigeminal nerve is identified. Along the floor of the middle cranial fossa, the GSPN is identified and provides a landmark for the geniculate ganglion. The geniculate ganglion also serves as a superior landmark for the cochlea. The GSPN is then transected to avoid traction injury to the facial nerve. Further drilling in this area with cutting and then diamond burrs provides initial exposure of the carotid artery.

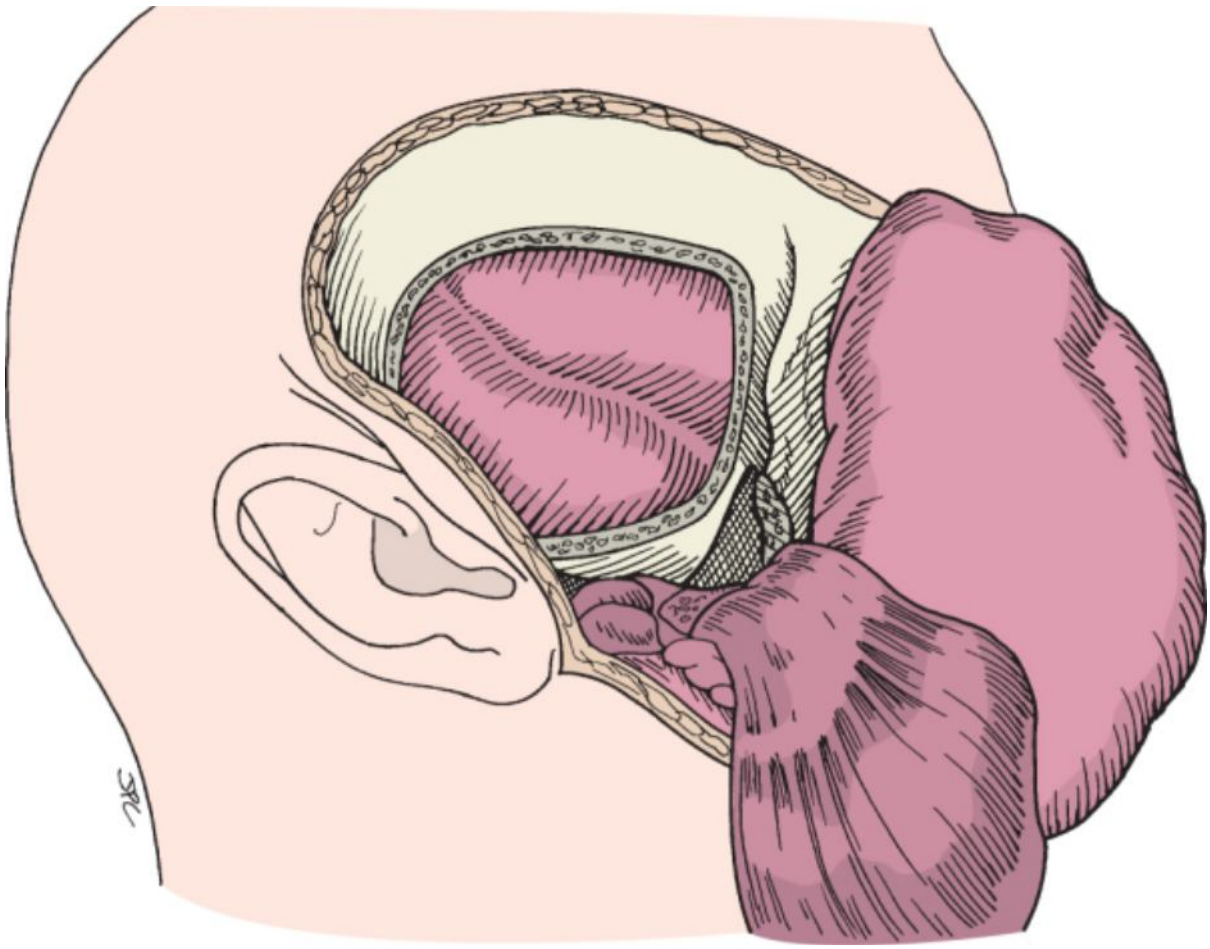

Figure 126-21 A frontotemporal craniotomy is performed to provide access to the middle (subtemporal) and infratemporal fossae.

The horizontal portion of the carotid artery must be dissected for identification, retraction, or possibly temporary clipping. Bone between the facial hiatus and the foramen spinosum is removed with a drill and rongeurs to unroof the carotid artery. Dissection lateral to the artery exposes the tensor tympani muscle and the eustachian tube. The eustachian tube is transected, and its cartilaginous portion is suture-ligated if the carotid artery is mobilized anterolaterally.

Drilling is continued in the central aspect of the petrous apex. An expansile tumor or cyst should be readily apparent at this time (Fig. 126-22). The area is entered, and its contents are evacuated. Once a cavity has been created, meticulous dissection of the epidermal matrix or cyst wall is performed. Care must be taken when the cyst lining is removed from the carotid artery because of possible attenuation of the adventitia and wall of the carotid artery. Attention is also given to the dura from which the cyst wall lining was removed. The integrity of the dura must be verified to prevent postoperative CSF leakage. When the entire cyst wall has been removed, the cavity is copiously irrigated with bacitracin solution. The residual cavity defect warrants soft tissue obliteration with abdominal fat. The lateral craniotomy bone flap is returned to its anatomic position and secured with miniplates or 2-0 Vicryl sutures, the temporalis muscle is reattached to its insertion with multiple 2-0 Vicryl sutures, and the zygomatic arch is secured in its original position with miniplates or sutures. The skin is closed in three-layer fashion, and a sterile pressure dressing is applied.

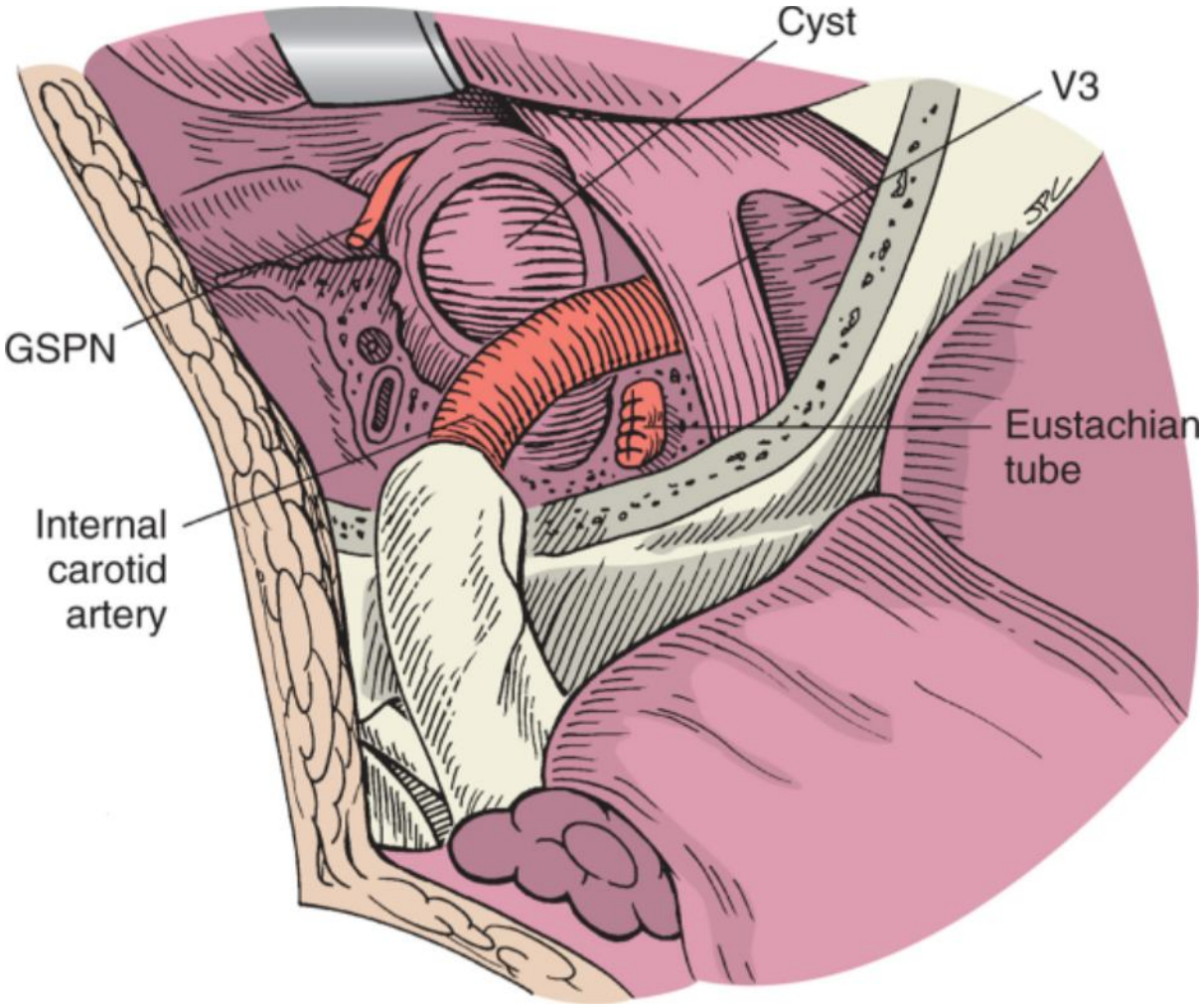

Figure 126-22 Bone separating the subtemporal dura from the infratemporal fossa is removed to expose V3. The greater superficial petrosal nerve (GSPN) has been divided to permit decompression of the internal carotid artery. This exposure allows excision of a petrous apex cholesterol granuloma if a drainage procedure is unsuccessful or resection of an isolated epidermoid tumor if hearing is intact.

Expanded Endoscopic Transnasal Approach

Since writing of the first edition of this textbook, surgeons at the University of Pittsburgh Medical Center have developed and mastered transnasal endoscopic procedures directed to the pituitary gland, anterior skull base, anterior cranial fossa, infratemporal fossa, and posterior fossa. These surgical techniques have been adapted to gain access to the petrous bone from an anterior approach despite development of the sphenoid sinus.[32] The dissection through the sphenoid bone is medial to the carotid artery. The petrous apex is the point of entry for access and drainage of cholesterol granuloma.

POSTOPERATIVE MANAGEMENT

The intensity of immediate postoperative care is determined by the extent of the operative procedure undertaken and whether the cranial nerves, the carotid artery, the dura, or the otic capsule were affected or compromised by the procedure. Patients undergoing a drainage procedure for cholesterol granuloma via the infracochlear or infralabyrinthine route can be discharged from the hospital on the same or the following day, depending on their tolerance for the procedure and reaction to general anesthesia. Patients are seen 7 to 10 days postoperatively for ear canal packing and removal of the postauricular Steri-Strips. If the medial external canal covering the tympanomeatal flap was filled with Gelfoam, antibiotic suspension drops are prescribed twice daily. The patient is seen every 2 to 3 weeks to suction out the remaining Gelfoam and ensure that healing of the inferior canal wall is taking place. After a few weeks, the middle ear can be assessed. It takes 4 to 6 weeks for the inferior canal and hypotympanum to heal. The patient's middle ear is observed in the interim to monitor for resolution of postoperative serous effusion. Concern for compromised middle ear aeration via the eustachian tube is managed by placement of a myringotomy tube in the anterosuperior quadrant. An audiogram is obtained after complete healing to assess the status of postoperative hearing.

A pressure dressing is maintained over wounds that were obliterated with fat or a transposed tem-poralis muscle.

Drains are not placed within the operative site routinely. However, if an abdominal fat graft was obtained and a Penrose drain was placed in the abdominal wound, the drain is removed once drainage diminishes. In patients with congenital epidermoid tumor who are able to have the disease exteriorized by marsupialization, the mastoid dressing is removed the day after surgery. A cotton ball placed in the external auditory meatus is subject to serosanguineous drainage and is changed frequently. When the patient is seen 7 to 10 days postoperatively, the postauricular Steri-Strips and part of the continuous-strip gauze packing are removed. The packing is withdrawn until there is significant resistance to removal or mild bleeding is encountered. Topical antibiotic ear drops are prescribed, and the remainder of the packing is removed the following week. Oral antibiotics are not prescribed, although oral narcotic analgesics are provided for pain relief.

Patients requiring extensive dissection or resection are at risk for CSF leak and injury to the cranial nerves, otic capsule, carotid artery, dura, and brain. Patients are closely monitored for symptoms or signs related to these neurovascular structures. The patient's neurologic status is monitored frequently when a CSF leak is encountered or a subtemporal approach was performed. Patients are evaluated for signs of mental status change or evidence of meningitis. Those requiring a transpetrous or subtemporal infratemporal fossa approach for total resection of cholesterol granuloma or epidermoid tumor are usually hospitalized for a few days. The wound and patient are observed for infection, subdural hematoma, and CSF accumulation, otorrhea, or rhinorrhea. Further details regarding skull base approaches are reviewed in Chapter 102.

The presence or development of facial nerve dysfunction requires supportive care. Particular attention is devoted to protection of the cornea. Weakness of the orbicularis oculi with symptomatic lagophthalmos is managed by artificial tears during the day and ointment at night. A clear moisture chamber is provided if topical drops alone do not adequately relieve the dryness and irritation. Placement of a gold weight within the upper eyelid is recommended for assisted lid closure and corneal protection if it is anticipated that facial paralysis will be present for more than a brief time (see Chapter 121). Persistent erythema with eye irritation warrants ophthalmic consultation.

Inadvertent or intentional entry into the otic capsule usually results in vertigo if vestibular function was present preoperatively. Supportive measures consisting of antiemetic medications such as phenothiazine or droperidol should be taken. Assisted ambulation will enhance central vestibular recovery. The physical therapy department is consulted to initiate a rehabilitation treatment plan. Physiologically young patients will adapt centrally to the unilateral vestibular loss. At our institution, patients who continue to exhibit uncompensated disequilibrium are subsequently referred to the vestibular rehabilitation physical therapist for ongoing outpatient care. Coping strategies and vestibular retraining exercises are the focus of their program.

If a hearing preservation procedure was performed, a postoperative audiogram is obtained at approximately 1 month. Unless the procedure consisted of marsupialization into a large mastoid cavity, it is necessary to monitor for recurrence with radiologic imaging. Given that the tumor or cystic lesion demonstrated bone expansion with smooth remodeling, a CT scan can be used to effectively evaluate changes in the cavity. Regardless of whether the procedure entailed drainage and ventilation or resection with obliteration, a CT scan obtained 3 months postoperatively will provide the baseline for future comparisons. MRI does have a role in postoperative monitoring (Fig. 126-23). If there is concern for recurrence of pathology despite a stable CT scan, MRI readily differentiates changes in tissue density. If the cavity was obliterated with fat, fat suppression MRI techniques can be used to critically assess the contents of the bony cavity. A subsequent scan is obtained 1 year after the baseline scan. If the petrous apex is found to contain air, adequate ventilation has been achieved. If the cavity was obliterated and the patient shows no new clinical signs or symptoms, a CT scan is obtained 3 years later.

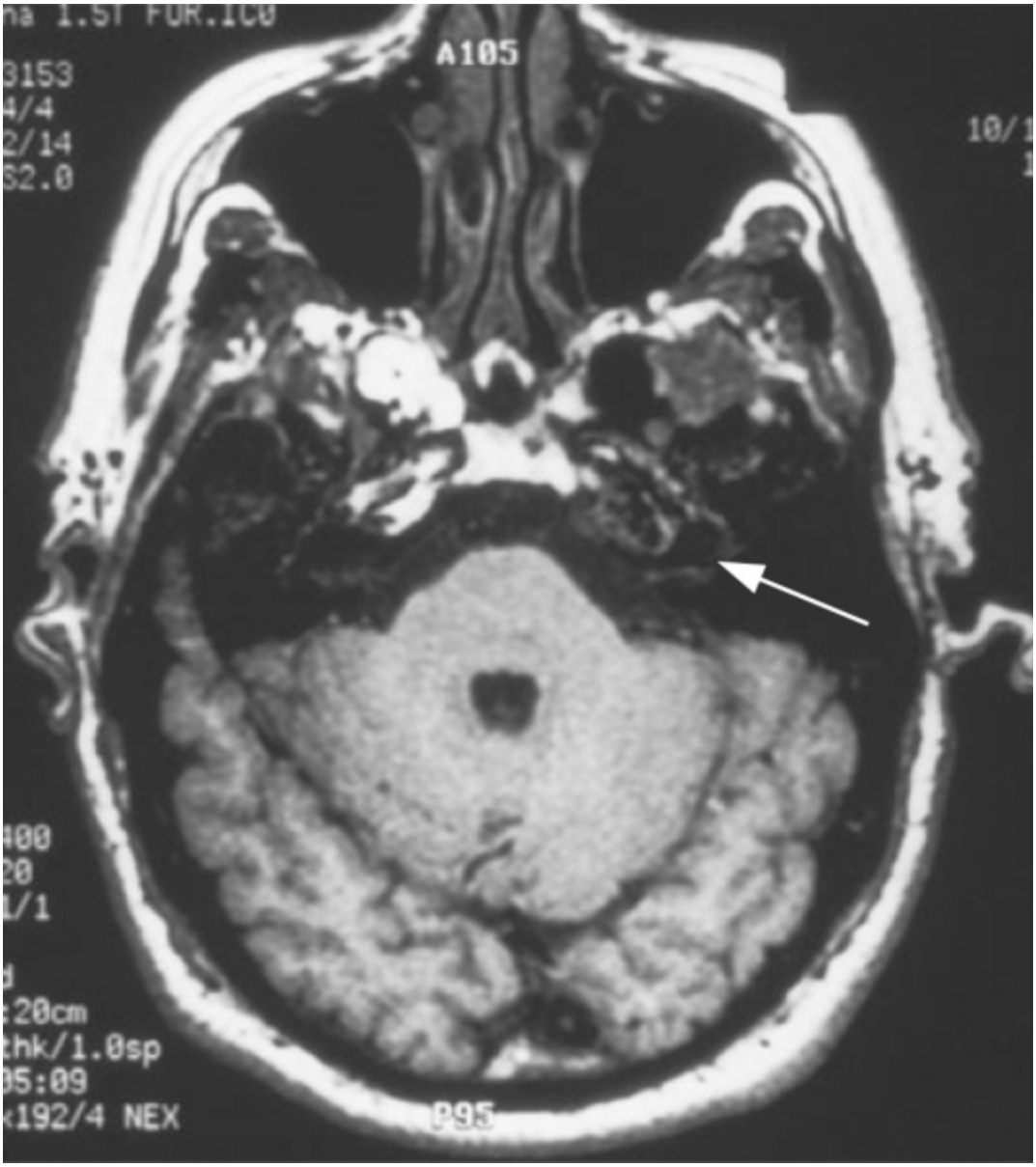

Figure 126-23 Postoperative magnetic resonance image of the cholesterol granuloma shown in Figure 121-5. This non–contrast-enhanced T1-weighted image demonstrates lack of signal in the petrous apex *(arrow)*, indicative of excellent aeration and resolution of the cyst.

COMPLICATIONS

The problems and complications that arise after management of cholesterol granuloma and large epidermoid tumors depend on the surgical approach and completeness of the procedure. In addition to the problem of persistent or recurrent disease, complica-tions can be grouped by cranial nerve deficits, central nervous system sequelae, vascular injury and bleeding, and problems related to the wound.

A preexisting conductive hearing loss may remain or increase in magnitude, depending on the operative management of the ossicular chain and tympanic membrane. The first goal of the procedure is to effectively eradicate or exteriorize the pathologic process for which the procedure was performed. This may require radical mastoidectomy, which by definition may maximize a conductive hearing loss. Under this circumstance, the conductive hearing loss is an accepted outcome from the procedure rather than a risk or complication. However, if the ossicular chain was reconstructed and a conductive hearing loss persists, subsequent aural rehabilitation can be provided by revision surgery or hearing aid amplification.

Despite normal cochlear function, the procedure may occasionally require sacrifice of the otic capsule to eliminate the cyst or tumor. Again, this would be not a complication but rather a planned outcome. However, if it was unplanned, sensorineural hearing loss may result from inadvertent injury to the otic capsule or ossicular chain. Planned or unintended entry into the inner ear typically results in vestibular signs of nystagmus, vomiting, and postural instability and symptoms of nausea, vertigo, and disequilibrium.

Middle ear effusion develops when the eustachian tube is sacrificed during the subtemporal infratemporal fossa approach. The resultant conductive hearing loss is treated with a ventilation tube. It is best to wait approximately 4 to 6 weeks before tube insertion to allow complete healing of any potential dural defects.

Resection of large tumors of the petrous apex extending toward the cavernous sinus may also put the third, fourth, fifth, and sixth cranial nerves at risk. Patients with petrous apex involvement occasionally have preoperative symptoms of facial hypoesthesia. Care should be taken to minimize further trauma to the trigeminal nerve. Similarly, the immediate proximity of the sixth nerve to Meckel's cave requires that it be identified during intradural procedures to avoid excessive manipulation. Injury to this nerve results in paresis of the lateral rectus muscle. If the nerve was stretched during the procedure, return of function can be expected within a few months. Sacrifice of the sixth cranial nerve or paralysis that does not resolve warrants consultation with an ophthalmologist for corrective muscle surgery.

Injury to the seventh cranial nerve may result in paresis or paralysis of the facial musculature. If it is known that dissection around the facial nerve left the nerve anatomically intact, supportive care is provided as described previously under “Postoperative Management.” Sacrifice of the seventh nerve is corrected by reconstruction with a hypoglossal-to-facial nerve anastomosis if primary or cable graft repair could not be performed at the time of the procedure.

The lower cranial nerves may also be affected by large epidermoid cysts or cholesterol granuloma of the petrous apex. However, direct invasion is relatively rare. Intraoperative injury to these nerves results in dysphagia, aspiration, hoarseness, and inanition. Temporary dysfunction is managed by injection of Gelfoam into the vocal cord and enteral supplementation via nasogastric feeding. Permanent injury to the vagus nerve resulting in hoarseness, dysphagia, and aspiration is managed by a type I thyroplasty and cricopharyngeal myotomy.

Infralabyrinthine or infracochlear drainage procedures require intimate dissection around the jugular bulb. Intraoperative extradural venous bleeding can be quite bothersome, but it is usually controlled with Surgicel, bone wax, or bipolar cautery. Dissection of the petrous apex requires careful attention to the carotid artery. Preoperative testing with balloon occlusion provides an indication of how the patient will tolerate temporary intraoperative occlusion to repair injury to the ICA. It is best to avoid a potential injury to the carotid artery by meticulous dissection and protection of the vessel. If the dura into the posterior fossa is opened, similar respect is given to the intracranial vessels. Repair of these small vessels is technically most difficult, and clipping or bipolar coagulation may be necessary. This may result in serious central nervous system deficits in the cerebellum, midbrain, and brain stem.

Procedures in which resection or drainage is attempted in an extradural fashion may still be complicated by CSF otorrhea and rhinorrhea. Leakage around the otic capsule into a closed middle ear space will be manifested as CSF rhinorrhea. Canal wall down procedures (radical mastoid cavity) in which tympanic membrane reconstruction was not undertaken will have CSF leaking directly through the aural wound. The risk for meningitis demands repair of the fistula. Lumbar drainage of CSF is rarely effective in controlling the leak once a fistula has developed in an open cavity. Operative intervention requiring soft tissue obliteration with wound compression is usually necessary for effective control.

Wound infection is a rare complication of otologic surgery. Soft tissue infection and cellulitis are treated with systemic antibiotics. Formation of an abscess such as in the epidural space requires drainage, culture for an organism, and appropriate parenteral antibiotics. Drainage procedures for cholesterol granuloma provide a potential pathway for retrograde infection into the cystic cavity. If the expansion process had thinned the wall of the cavity and exposed the carotid artery, carotid hemorrhage may result. A bacterial or fungal infection may become established in the cancellous bone, with the subsequent development of temporal bone osteomyelitis.[18]

Excessive retraction of or direct injury to the temporal lobe may occur from procedures directed at the petrous apex via the subtemporal infratemporal fossa or direct transtemporal approaches. Injury to the left temporal lobe may result in expressive or receptive aphasia. Though rare, ischemia and infarction may produce temporary or permanent hemiparesis and hypoesthesia.

PEARLS

- Both CT and MRI are necessary for determining the type of pathology affecting the petrous apex.
- Transnasal/transpetrous approaches to pathology of the petrous apex should be considered when reviewing images showing the anatomy of related structures.
- Sacrifice of the eustachian tube results in middle ear effusion when the tympanic membrane remains intact or is reconstructed. Delayed ventilation of the middle ear should be planned after extensive procedures to avoid the risk of CSF otorrhea.
- Temporary or permanent injury to the facial nerve requires early aggressive eye care to protect the cornea from desiccation or further injury.

- Placement of a ventilation tube in the tympanic membrane may obviate early occlusion of a Silastic drainage tube positioned between the petrous apex and the hypotympanum after an infracochlear approach.

PITFALLS

- Underestimating the exposure necessary for complete removal or creating suboptimal drainage of pathologic processes of the petrous apex will result in persistence or recurrence of disease.
- CSF otorrhea or rhinorrhea after approaches to the petrous apex may not respond to lumbar drainage; operative intervention will probably be necessary.
- Subtotal removal of the epithelial matrix of an epidermoid cyst may be temporizing and represents the first stage of subsequent procedures.
- Performing a procedure that drains the petrous apex and creation of a radical mastoid cavity without coverage of an exposed carotid artery may result in desiccation and rupture.
- A middle fossa approach to cholesterol granuloma of the petrous apex may not afford adequate drainage into the middle ear. The entire cyst and its lining will need to be removed.