Chapter 127 – Cerebrospinal Fluid Otorrhea and Encephalocele

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Temporal bone cerebrospinal fluid (CSF) otorrhea may occur from a variety of congenital or acquired etiologies (Table 127-1). Leakage of CSF through the temporal bone represents a fistula from the fluid-filled subarachnoid space into the pneumatized temporal bone. Subsequent egress of CSF through a defect in the mastoid cavity, tympanic membrane, or external auditory canal presents as otorrhea. Rhinorrhea represents drainage of CSF through the eustachian tube and may occur with or without otorrhea. An encephalocele is a protrusion of the brain, with or without overlying dura, beyond the confines of the cranial cavity into the mastoid or middle ear. A true encephalocele contains prolapsed brain tissue, whereas a meningoencephalocele contains brain and meninges. The terms *CSF otorrhea* and *encephalocele* both imply an anatomic pathway between the subdural space and the exterior. For this reason, both have equal potential for central nervous system complications and infections.

Gacek demonstrated that aberrant arachnoid granulations may occur in the vicinity of the temporal bone as well as in other areas of the skull base.[1] Unlike classic arachnoid granulations, these aberrant structures do not have an absorptive role associated with CSF. Although arachnoid granulations of the middle or posterior cranial fossa are not routinely encountered during mastoid surgery, the incidence of these structures in temporal bone histopathologic studies of the surface of the middle fossa is as high as 22%. The most common locations of these aberrant arachnoid granulations are lateral to the cribriform plate in the anterior cranial fossa, and on the floor of the middle fossa. Occasionally they may occur in the posterior fossa plate between the sigmoid sinus and the bony labyrinth. Intermittent increases in spinal fluid pressure are hypothesized to cause gradual increase in size of these aberrant arachnoid granulations and erosive changes in the underlying bone, with eventual protrusion through the tegmen or posterior fossa plate. It is thought that these granulations are the largest single cause of spontaneous CSF leaks in adults. The typical presentation of spontaneous CSF leaks in adults is unilateral serous otitis media with no preceding upper respiratory infection, which is refractory to decongestant and antibiotic therapy. Myringotomy and ventilation tube placement usually results in persistent clear watery drainage that does not respond to conventional ototopical therapy. A high index of clinical suspicion is necessary to diagnose this entity.

Perilymphatic fistulas are another potential source of communication between the middle ear and the subarachnoid space. These defects rarely result in any clinically significant accumulation of fluid within the middle ear and therefore typically do not present like other causes of CSF otorrhea or rhinorrhea. Leakage typically occurs from the otic capsule through a dehiscence in the stapes footplate. The diagnosis is usually made in childhood, and typically presents as recurrent meningitis with hearing loss in the affected ear. Perilymphatic fistulas are discussed in detail in Chapter 119.

Spontaneous meningoencephaloceles of the skull base have been reported since early in this century. The most common location for encephaloceles of the temporal bone is along the floor of the middle cranial fossa. The posterior fossa of the temporal bone may also be affected, although rarely. Defects are frequently identified in multiple locations at the time of presentation. Congenital defects in the bone of the skull base predispose to progressive formation of dural and arachnoid herniations. Prolapse into the epitympanum and middle ear may cause conductive hearing loss or present with a mass in the middle ear (Fig. 127-1). Retrograde bacte- rial contamination from the nasopharynx, paranasal sinuses, or middle ear may result in the development of meningitis. Therefore this disorder occurs most frequently in childhood.

Trauma to the temporal bone may occur as a result of blunt head trauma or surgery of the temporal bone. CSF otorrhea secondary to temporal bone fractures is discussed in Chapter 128. Diagnosis is straightforward in these cases, and the majority of these patients respond to conservative management. Bed rest and head elevation results in spontaneous resolution in up to 90% of patients with temporal bone fractures. Most of the remaining cases can be controlled by intermittent or continuous lumbar drainage, leaving only a few patients who require surgical intervention.[2] Iatrogenic trauma to the dura during surgery of the temporal bone can eventually lead to weakening of the dura with resultant meningoencephalocele, encephalocele, and CSF otorrhea. Dural exposure alone, without violation, does not usually result in herniation of brain tissue or CSF leak. If CSF leak is noted at the time of iatrogenic trauma, surgical repair should be performed immediately.

CSF otorrhea and encephaloceles related to chronic otitis media may occur with or without cholesteatoma, usually in patients with an antecedent history of otologic surgery. Cholesteatomas may erode into the otic capsule as well as through the middle or posterior fossa plates. However, chronic otitis media and mastoiditis with granulation tissue alone may also lead to this defect over a long period. The erosive process from cholesteatoma and chronic

inflammation most commonly affects the horizontal semicircular canal, but may extend superiorly to involve the mastoid and middle ear tegmen.

Primary tumors or tumors metastatic to the temporal bone or areas adjacent to it may also cause bone erosion, which may be associated with secondary CSF otorrhea. The most common tumor in children is rhabdomyosarcoma. The most common tumors in adults are epithelial tumors and paragangliomas.

Table 127-1 -- CAUSES OF CEREBROSPINAL FLUID OTORRHEA

Congenital	Acquired	Spontaneous
Meningoencephalocele, secondary to congenital skull base defects	Posttraumatic (temporal bone fractures, surgery)	Arachnoid granulations
Perilymphatic fistulas	Chronic otitis media Neoplasia Postirradiation	

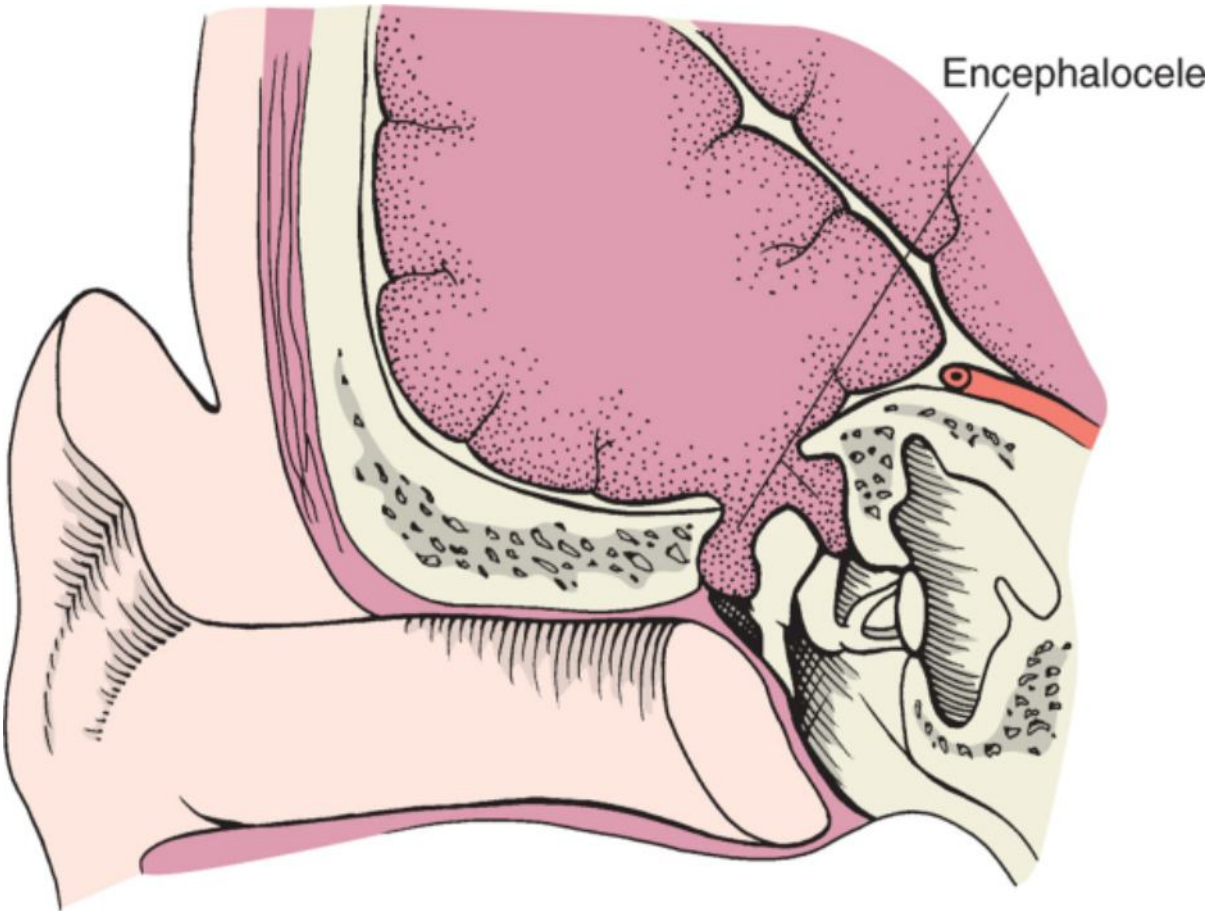

Figure 127-1 Herniation of brain tissue from the middle cranial fossa into the epitympanum.

PATIENT SELECTION

CSF leak of otogenic origin may present in several ways. If the tympanic membrane is violated, clear otorrhea is noted. With an intact tympanic membrane, the patient may complain of clear watery drainage from the nose, which usually occurs when bending over or straining. If leakage is minimal or intermittent, CSF may drain directly into the pharynx and esophagus without being noticed by the patient. The second clinical presentation occurring in individuals with an intact tympanic membrane, is hearing impairment and aural fullness as a result of middle ear effusion. Even in the absence of an antecedent upper respiratory infection, the diagnosis is presumed to be serous otitis media, and a myringotomy is often performed to drain the fluid. Following this procedure, the patient has continuous clear otorrhea, which persists despite antibiotic, antihistamine, and decongestant therapy. The latter presentation is typical of spontaneous otogenic CSF leaks in adults and a high index of suspicion is critical in making a timely diagnosis. The incidence of bilaterality in spontaneous otogenic CSF leaks may be up to 22%.[3]

Meningitis (single or repeated episodes) may be the initial presentation of a CSF leak. This is more common in

children when they have been unaware of any hearing loss or aural fullness in the affected ear. Meningitis often leads to a profound sensorineural hearing impairment, which may often be the presenting problem in children. Any patient presenting with meningitis must be carefully evaluated for a potential otogenic source.

Patients complaining of hearing impairment occasionally will have a mass discovered within the middle ear, with or without hearing loss (Fig. 127-2). High-resolution computed tomography (CT) of the temporal bone usually identifies the origin of the soft tissue mass and points to any defects in the bony plates of the middle or posterior fossa. Anyone with previous tympanomastoid surgery presenting with otorrhea, a middle ear effusion, or rhinorrhea should be suspected of having a defect in the dura as a result of the original procedure. A history of prior trauma to the temporal bone should alert the examiner to the potential diagnosis of encephalocele or meningoencephalocele.

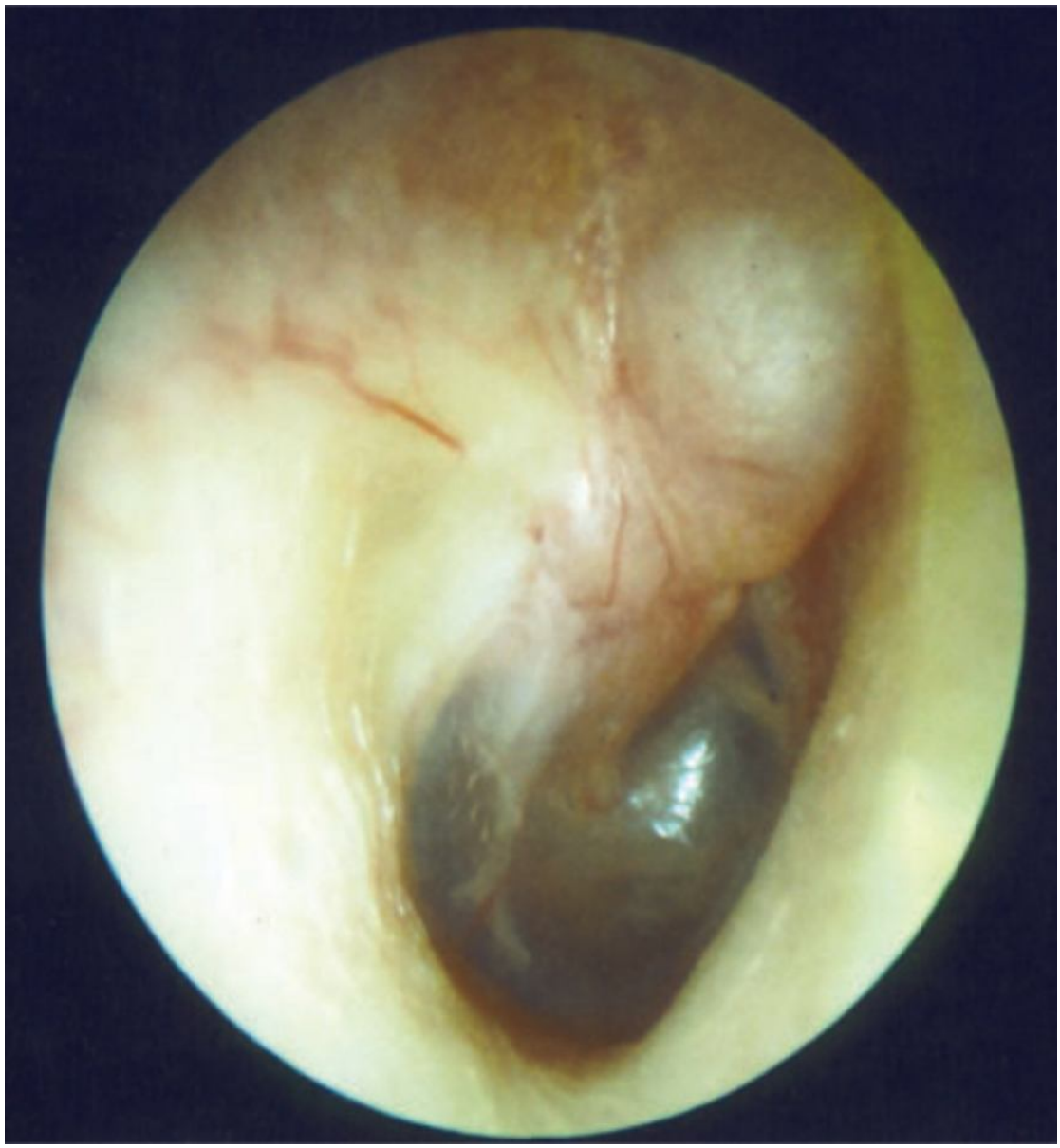

Figure 127-2 Middle ear mass visualized on otoscopy.

PREOPERATIVE PLANNING

Once CSF otorrhea, rhinorrhea, or encephalocele is suspected, efforts must proceed quickly to confirm the presence of a dural defect and furthermore to locate the defect precisely. The surgeon should attempt to elicit the symptoms of rhinorrhea. This can often be accomplished by having the patient bend forward and place the head between the knees for several minutes. It may also be accomplished by having the patient lie in a head-down position and then turn to the prone position to encourage active drainage. When CSF is observed clinically, there is usually no question about its origin. It appears as clear as water and is not cloudy or sanguinous. Difficulty in identification may be encountered only in trauma cases in which there is concomitant bleeding. In these cases, a

telltale ring of CSF may be seen outside the stain left by blood and serum on a dressing or bed linens (halo sign), which may alert the physician to this problem.

Testing fluid for the presence of glucose, usually with a test strip, is notoriously unreliable and usually requires large amounts of fluid to produce consistent results. It is therefore no longer used as a primary diagnostic modality for confirmation of CSF. When the origin or nature of the drainage is in question, we prefer the technique of identifying β_2-transferrin within the fluid using immunoelectrophoresis. This testing requires remarkably scant quantities of CSF and may be submitted to the laboratory as free fluid or on a carrier material such as Gelfoam or Merocel. β_2-transferrin is present only in CSF and perilymph, and is not found in blood or serum. The sensitivity and specificity of this test is approximately 100% and 95%, respectively, and is not influenced by contamination with blood or other body fluids.[4]

High-resolution CT imaging of the temporal bone using a bone algorithm is the cornerstone of diagnosing and localizing CSF leaks in the temporal bone. Coronal cuts usually enable the examiner to identify the location and number of bony defects in the petrous temporal bone. CT imaging also helps in identifying soft tissue within the middle ear and mastoid, erosion of the bone of the otic capsule, or structural anomalies of the inner ear (Fig. 127-3). Magnetic resonance imaging of the brain plays a complementary role in determining the nature and origin of soft tissue in the middle ear and mastoid when a large defect in the tegmen is identified on CT imaging (Fig. 127-4). Neither radionuclide cisternography or CT cisternography provides any additional diagnostic information in the evaluation of CSF leaks.[5]

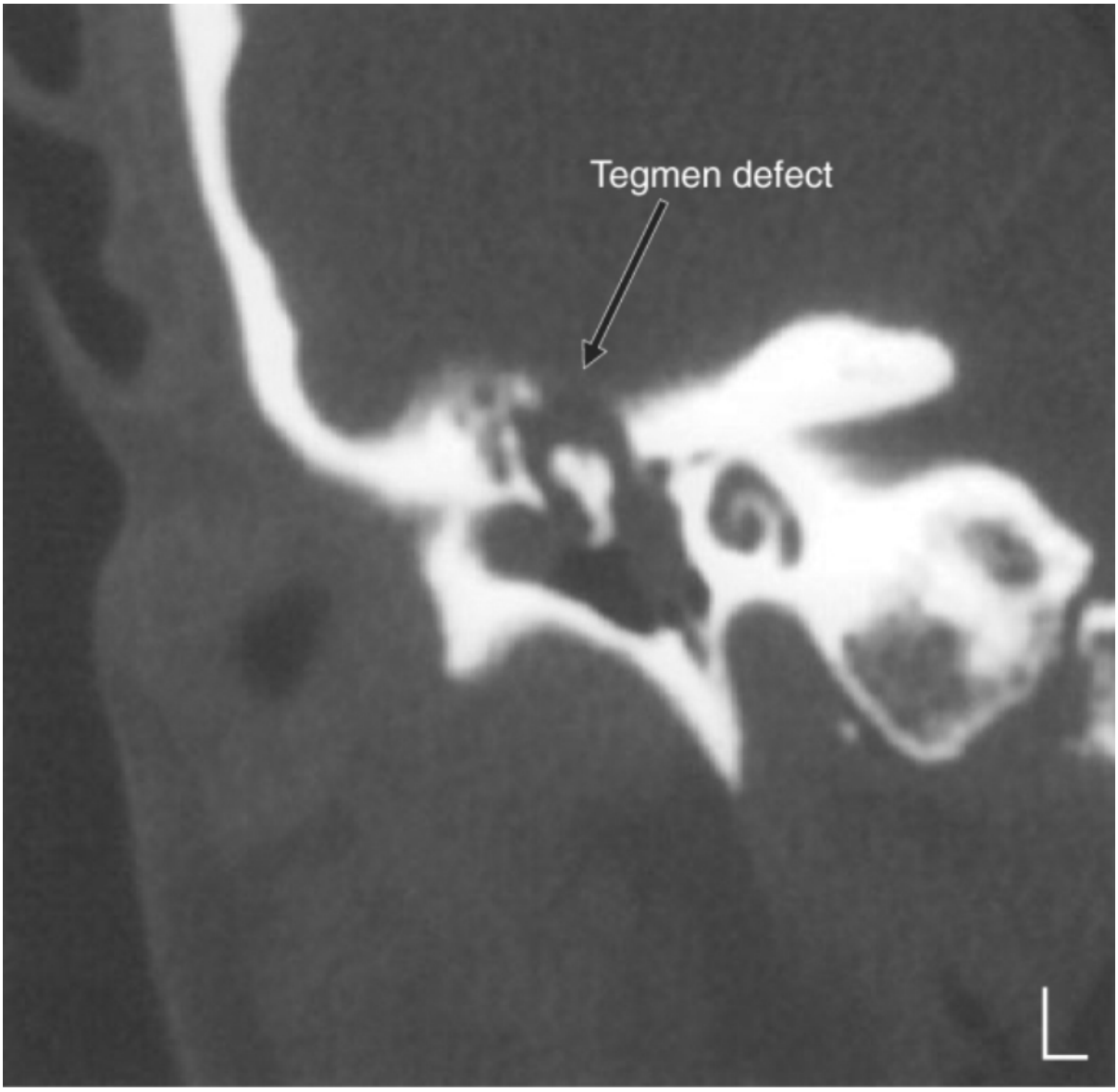

Figure 127-3 High-resolution coronal computed tomography scan of temporal bone showing the defect in the bone in the tegmen tympani and soft tissue in the epitympanum and middle ear *(arrow)*.

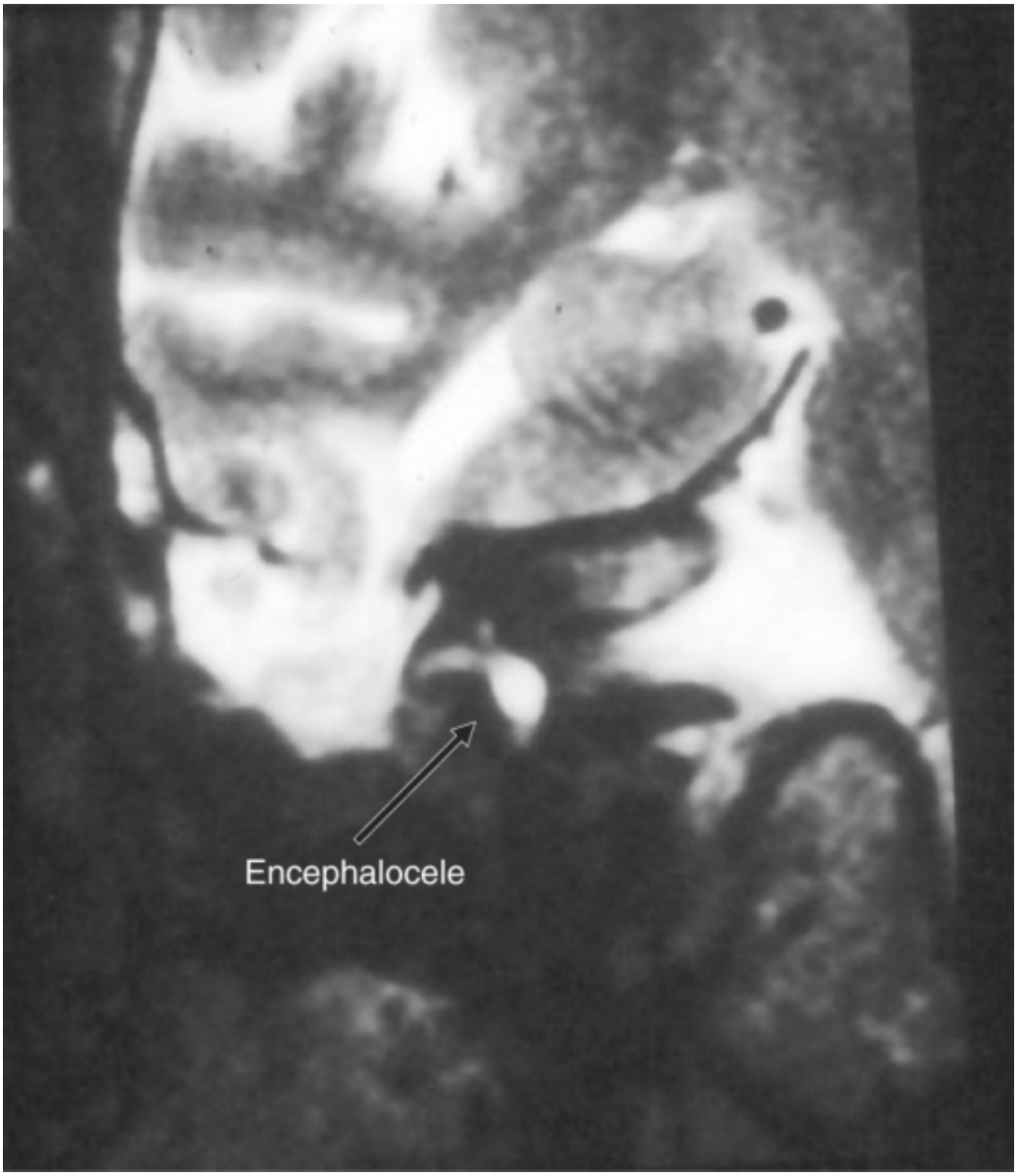

Figure 127-4 T2-weighted magnetic resonance imaging (MRI) of brain in coronal plane confirming soft tissue seen in Figure 127-3 as encephalocele *(arrow)*.

An audiogram should be obtained to evaluate the presence and extent of hearing impairment in the affected ear before surgical management. A hearing preservation approach is preferred in the presence of any aidable residual hearing in the operated ear.

SURGICAL APPROACHES

The surgical approach is dictated by the size, number, and location of bony defects, as well as the status of the hearing in the affected ear. Dehiscence of the posterior fossa plate, as well as of the posterior and lateral portions of the middle fossa plate, can almost always be repaired through a transmastoid approach (Fig. 127-5). Medial and anterior defects involving the tegmen tympani and petrous apex, respectively, require a middle cranial fossa approach for repair. Large or multiple tegmen defects usually require a combined transmastoid and middle cranial fossa approach. Patients with chronic otitis media and defects eroding the otic capsule may require a canal wall-down mastoidectomy in order to obtain adequate exposure for surgical repair and exteriorization of disease if extensive cholesteatoma is identified.

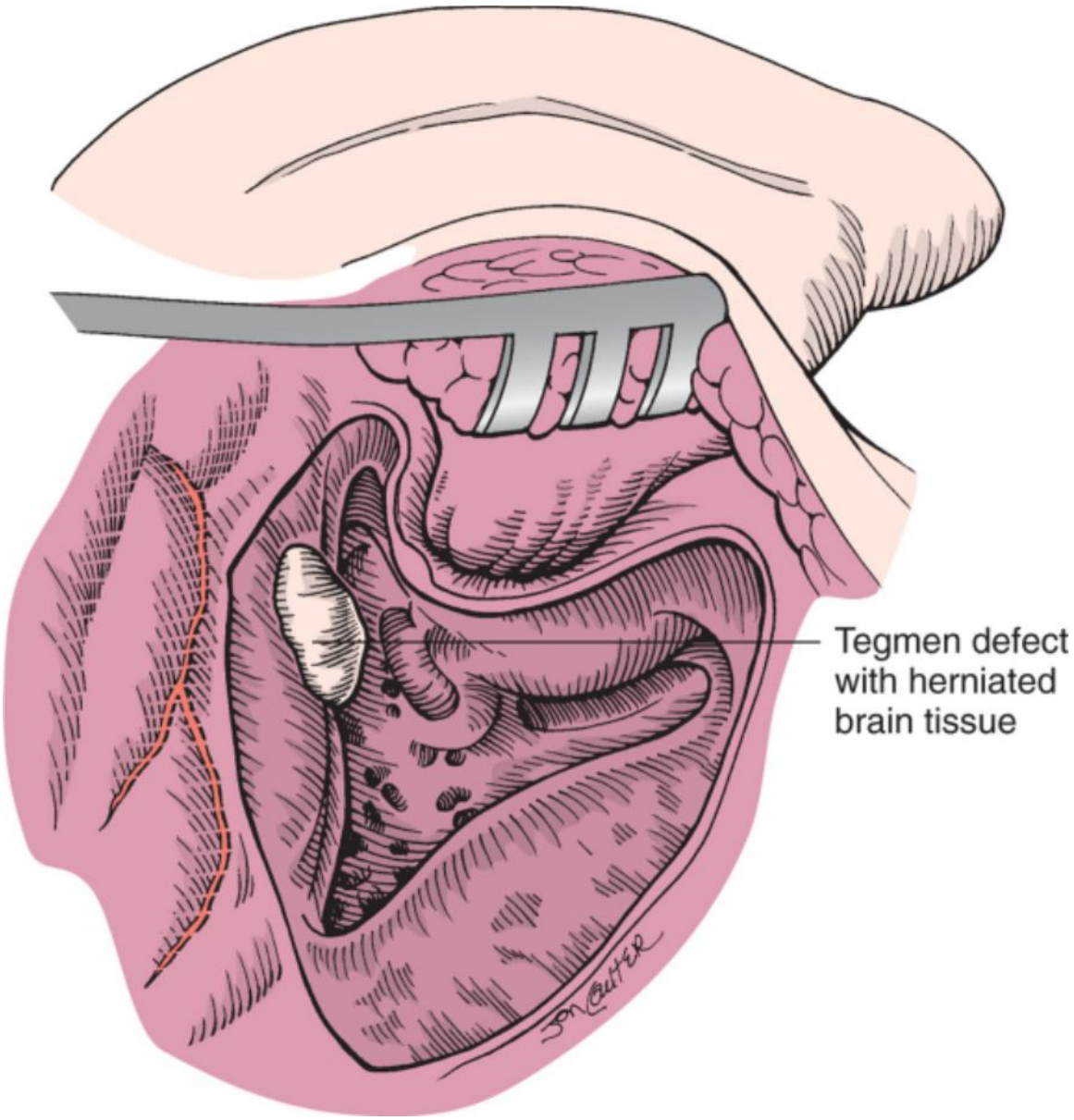

Figure 127-5 The transmastoid approach for repair of an encephalocele (right ear).

Details of the techniques for each of these approaches are described in Chapters 115 and 124. The transmastoid approach provides the distinct advantage of extracranial visualization of the defect without temporal lobe retraction. However, bony defects exceeding 2 cm in diameter are more reliably repaired from above using a supplemental middle fossa approach. In these cases, the postauricular incision is extended superiorly and anteriorly, and a limited craniotomy is performed, centered around the location of the defect identified from below. Similarly, in a patient with previous canal wall-down mastoid cavity with residual hearing, the defect in the tegmen and the encephalocele should be repaired from above using the middle fossa approach.[6]

When CSF otorrhea is copious, a lumbar drain is placed at the time of surgery. During surgery, the drain can be clamped to aid localization of the site of the leak. The Valsalva maneuver, performed by the anesthetist, may also be helpful, when necessary.

When brain herniation is encountered, there is uniform agreement that this tissue is nonfunctional and may be removed. This lack of function is assumed to be the result of tissue strangulation, ischemia, and resultant edema of this tissue. In general, broad-based encephaloceles herniating through large bony defects are considered viable and may be reduced back into the cranial vault. Bipolar electrocautery is used to remove nonviable tissue until the margins of the bony defect through which it has extruded is well defined.

Many methods of repair of these defects have been advocated over the years. A multilayer closure using a combination of free grafts has remained the most reliable technique to repair the defects. A large variety of materials including both biologic tissues and synthetic materials have been used to repair the dura. Autologous temporalis fascia, pericranium, and fascia lata are preferred when available because they are easy to handle,

nontoxic, inexpensive, and have favorable biologic behavior. Fascia lata may be obtained if temporalis fascia is lacking due to previous surgery, but obtaining fascia lata requires additional surgery, operating time, and donor site morbidity. The use of synthetic materials is no longer widely used because of local tissue reactions, excessive scar formation, meningitis, and hemorrhage. Solvent-preserved, gamma-sterilized Tutoplast bovine pericardium has also been widely used for dural substitution with favorable clinical outcomes.[7,8] The use of fibrin glue with primary closure alone does not appear to have any additional benefit.[2] Hydroxyapatite cement cranioplasty has been used in conjunction with autologous fat for prevention of CSF leak following vestibular schwannoma surgery and for primary repair of small defects in the tegmen in spontaneous CSF leaks.[9]

Two layers of temporalis fascia are used with a transmastoid approach, one on either side of the bony defect. Following removal of all soft tissue around the bony defect in the posterior or middle fossa, the intracranial dura is dissected with a duckbill or hockey-stick elevator until 1 cm of elevation is obtained circumferentially around the bony defect. At this point, dried temporalis fascia is placed within the bony defect and spread out over the intracranial surface between the dura and the tegmen (Fig. 127-6). Reexpansion of the brain contents ensures the retention of the fascial graft. The bony defect is then closed using a cancellous bone graft taken from the mastoid process, retrosigmoid area, or squamous portion of the bone, or using a conchal cartilage graft. This provides additional support for the primary graft material. When small bone defects are present, the bone graft or cartilage graft can be contoured to fit tightly into the exact shape of the space it is filling (Fig. 127-7). A tightly fitting graft in a small defect requires nothing further to keep it in place. A second layer of temporalis fascia is then placed over the bony defect on the mastoid side (Fig. 127-8), buttressed in place by means of temporalis muscle or abdominal fat.

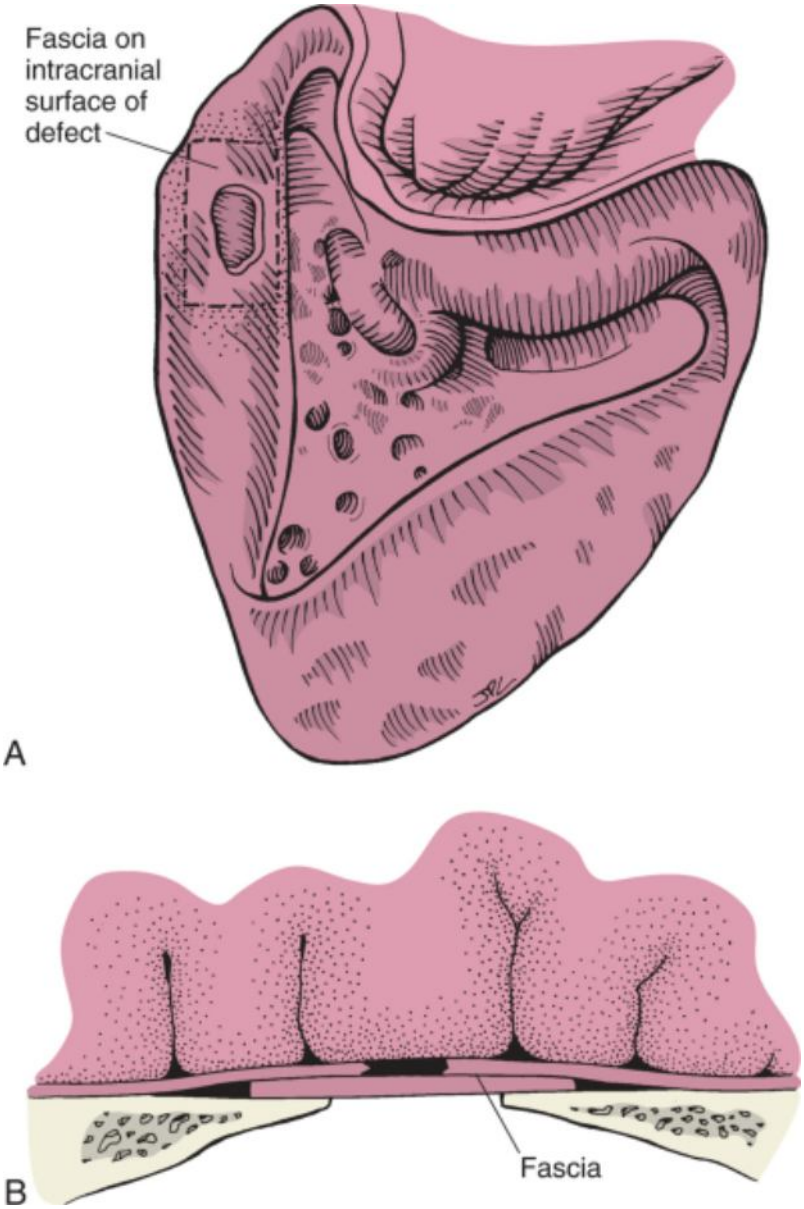

Figure 127-6 Use of fascia intracranially to form the first layer of repair of a dural defect.

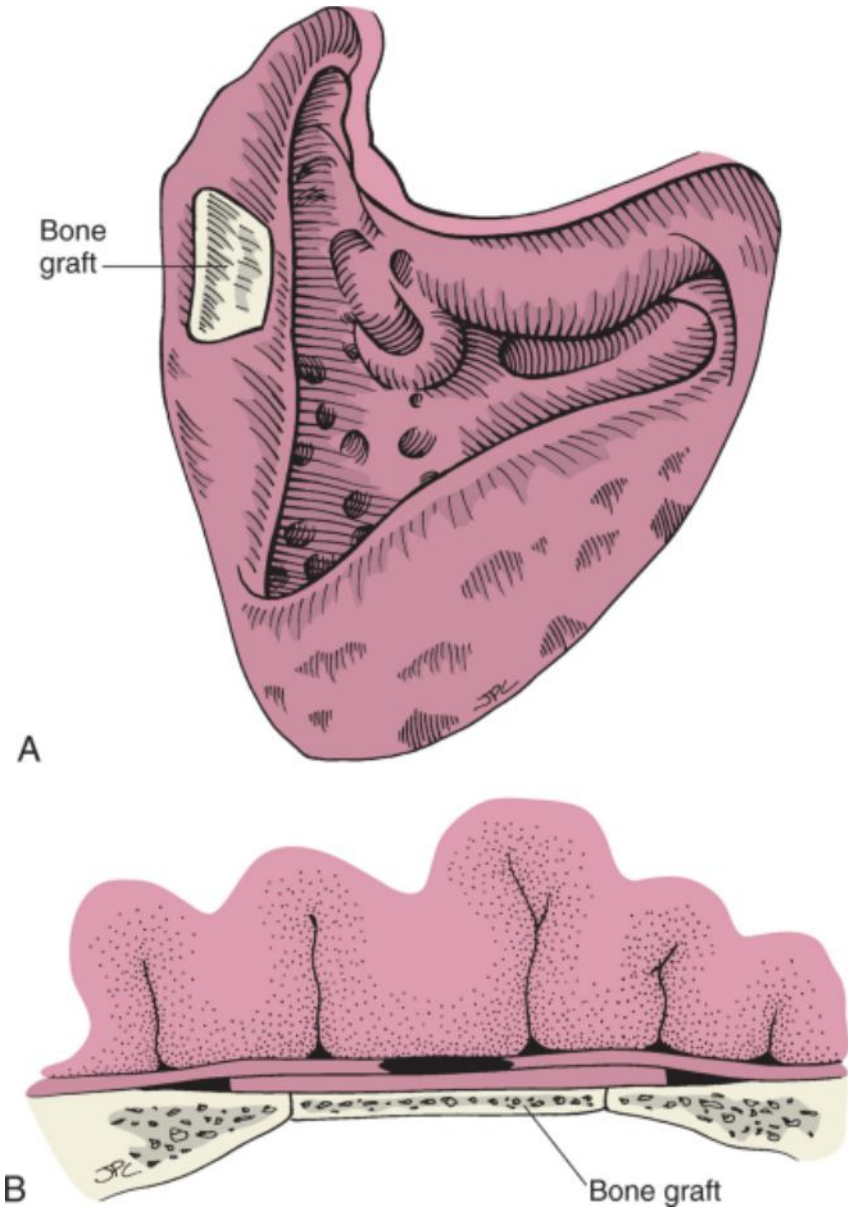

Figure 127-7 A shaped bone graft placed into the tegmen defect.

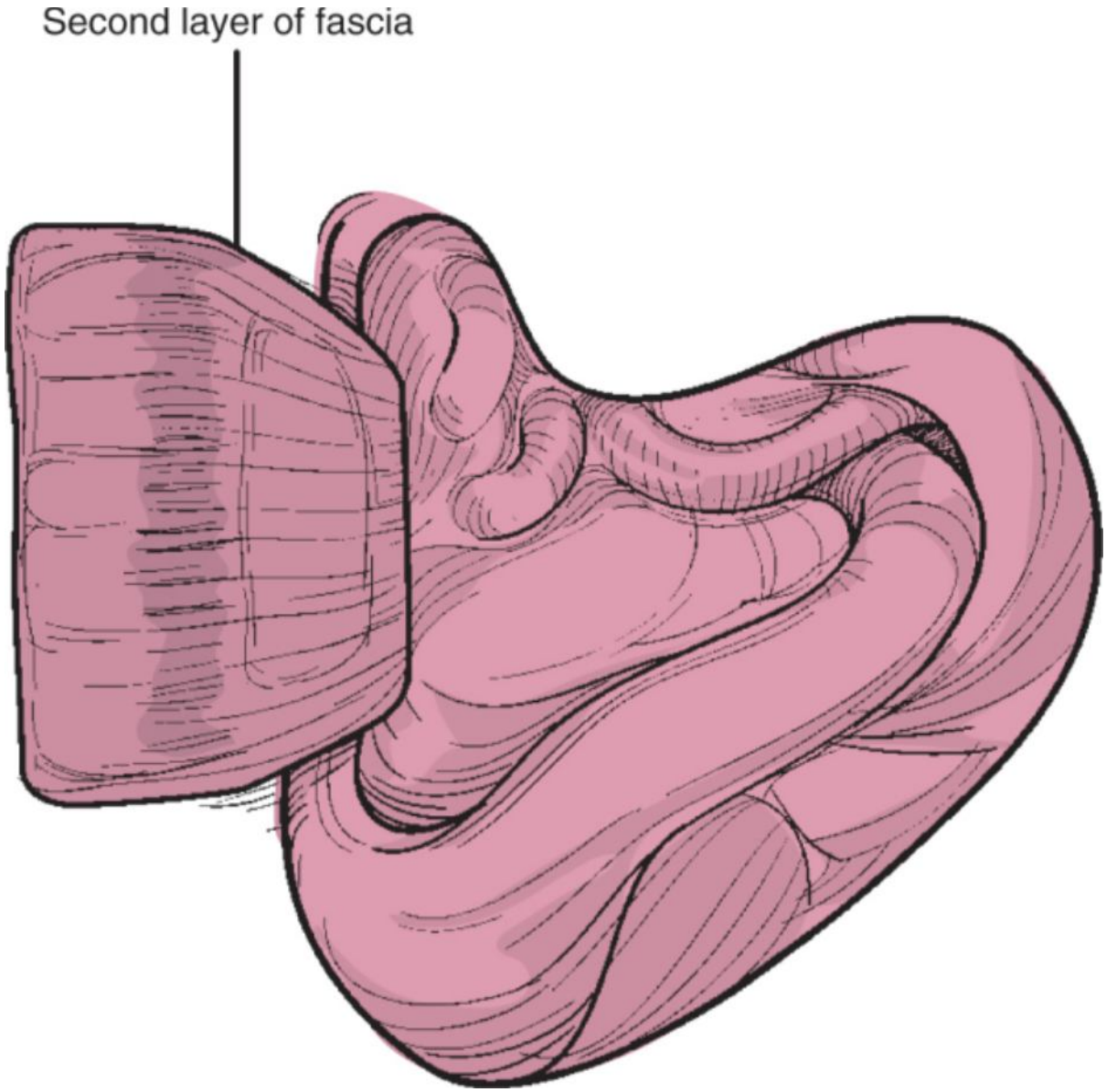

Figure 127-8 A second layer of fascia placed lateral to the bone graft.

A middle cranial fossa approach may be used as the primary approach for repair when anterior, medial, or multiple defects are present. Following extradural elevation of the temporal lobe, soft tissue is once again carefully removed from the bony defect (Fig. 127-9). A large piece of dried temporalis fascia is next placed across the floor of the middle cranial fossa (Fig. 127-10), and the temporal lobe is allowed to reexpand and hold this tissue in place. Larger defects should be supported using an additional bone or cartilage graft placed over the fascial graft. In addition, an attempt is made to close the dural defect responsible for the leak. When this defect is small, it can be closed primarily by suturing. When the defect is larger, a small piece of temporalis muscle is oversewn with 6-0 Nurolon sutures (Fig. 127-11). Alternatively, the entire repair can be done by an intradural technique.[10] Opening the dura on the lateral surface of the temporal lobe permits elevation of the brain from the underlying dura. This allows for close inspection of the herniated contents as well as the adjacent brain. Larger broad-based encephaloceles in uninfected fields may potentially be recranialized using this technique. The graft is placed on the dura, and the expanded brain then secures its fixation. In the rare case of a large defect with high-volume CSF drainage, vascularized tissue such as a pedicled temporalis muscle flap or pericranial flap should be rotated intracranially and secured to dura medial to the defect.

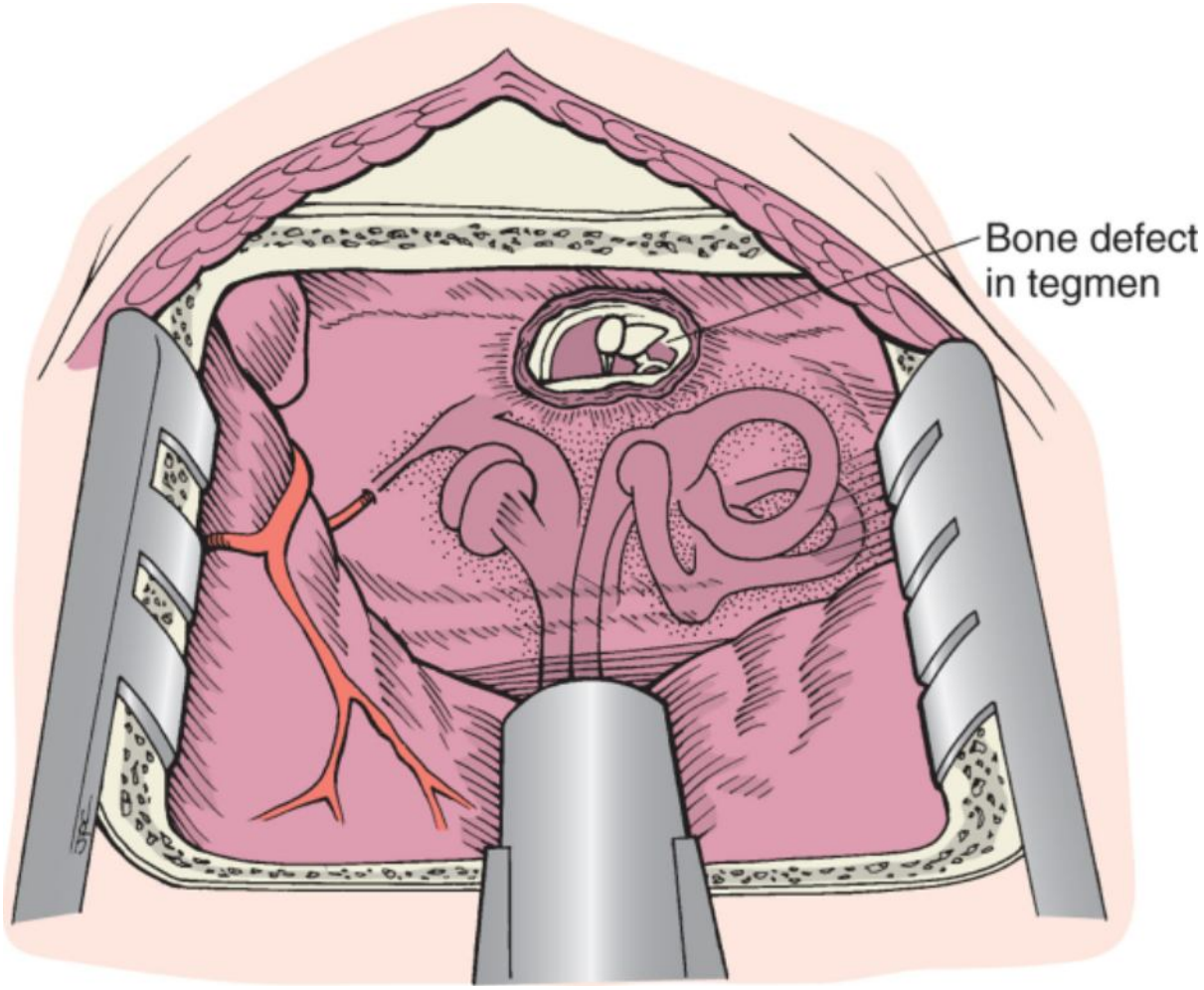

Figure 127-9 The middle fossa approach to repair showing the defect in the bone after removal of herniated tissue.

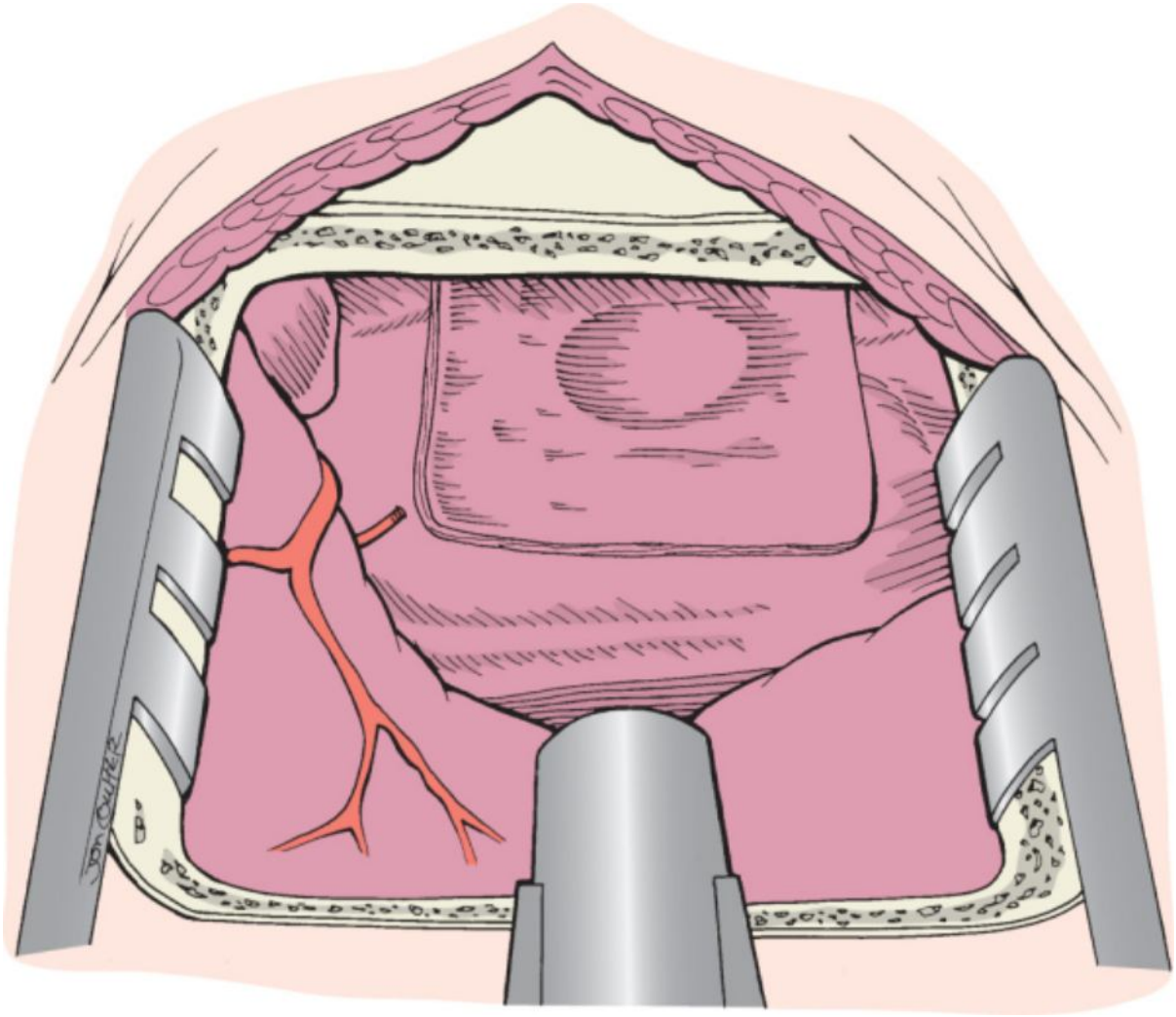

Figure 127-10 A large piece of dried temporalis fascia is used to cover the defect in the bone.

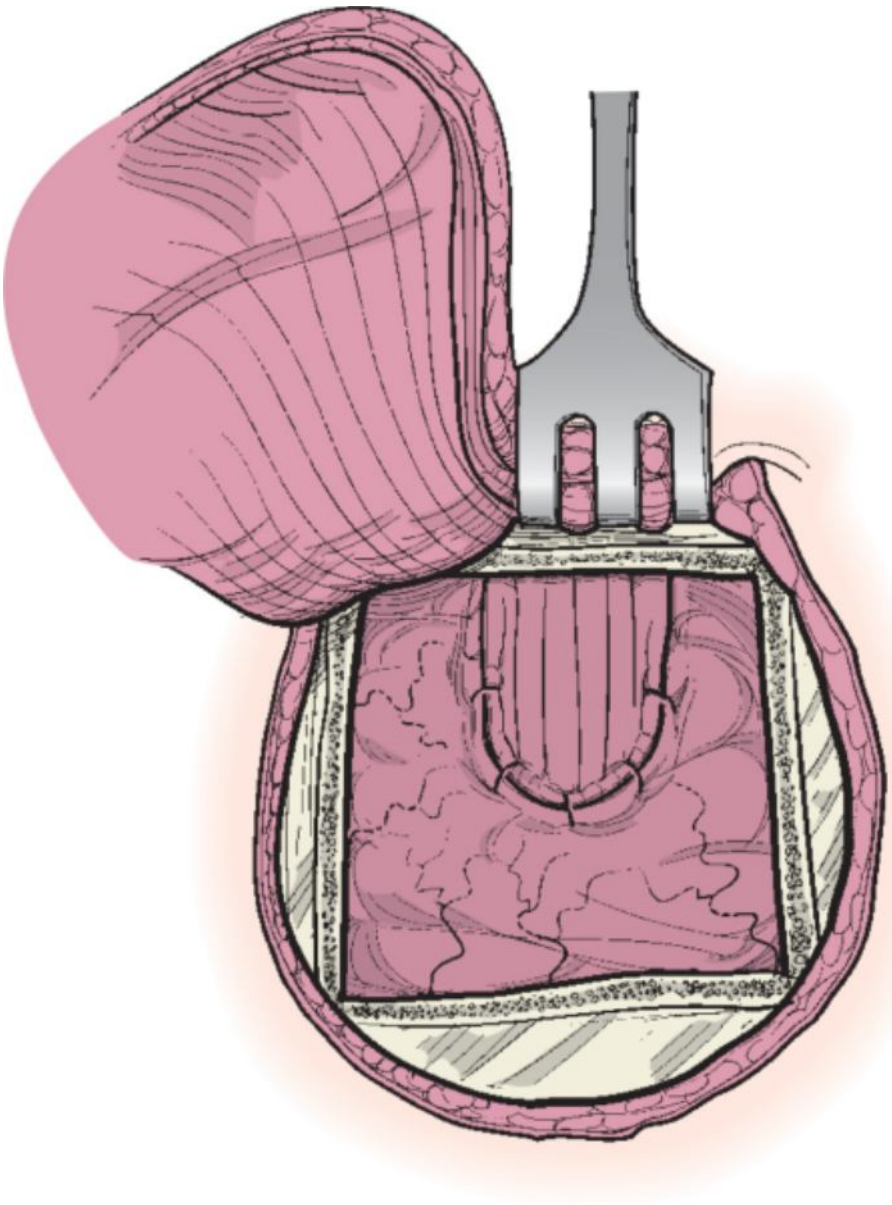

Figure 127-11 The temporalis muscle oversewn on the dura to repair the dural defect.

Defects in the otic capsule or tegmen plate associated with chronic middle ear disease are approached in the standard tympanomastoid fashion. In the absence of any useful hearing in the affected ear, the eustachian tube, mastoid, and middle ear may be obliterated and the ear canal closed. The malleus, incus, tympanic membrane, and skin of the medial external auditory canal are removed. It is also necessary to carefully seal the eustachian tube using bone and temporalis muscle (Fig. 127-12). The external auditory meatus is oversewn and closed in a layered fashion by means of absorbable Dexon sutures medially and nylon sutures laterally (Fig. 127-13). Temporalis muscle and abdominal fat are used to fill the bony defects (Fig. 127-14).

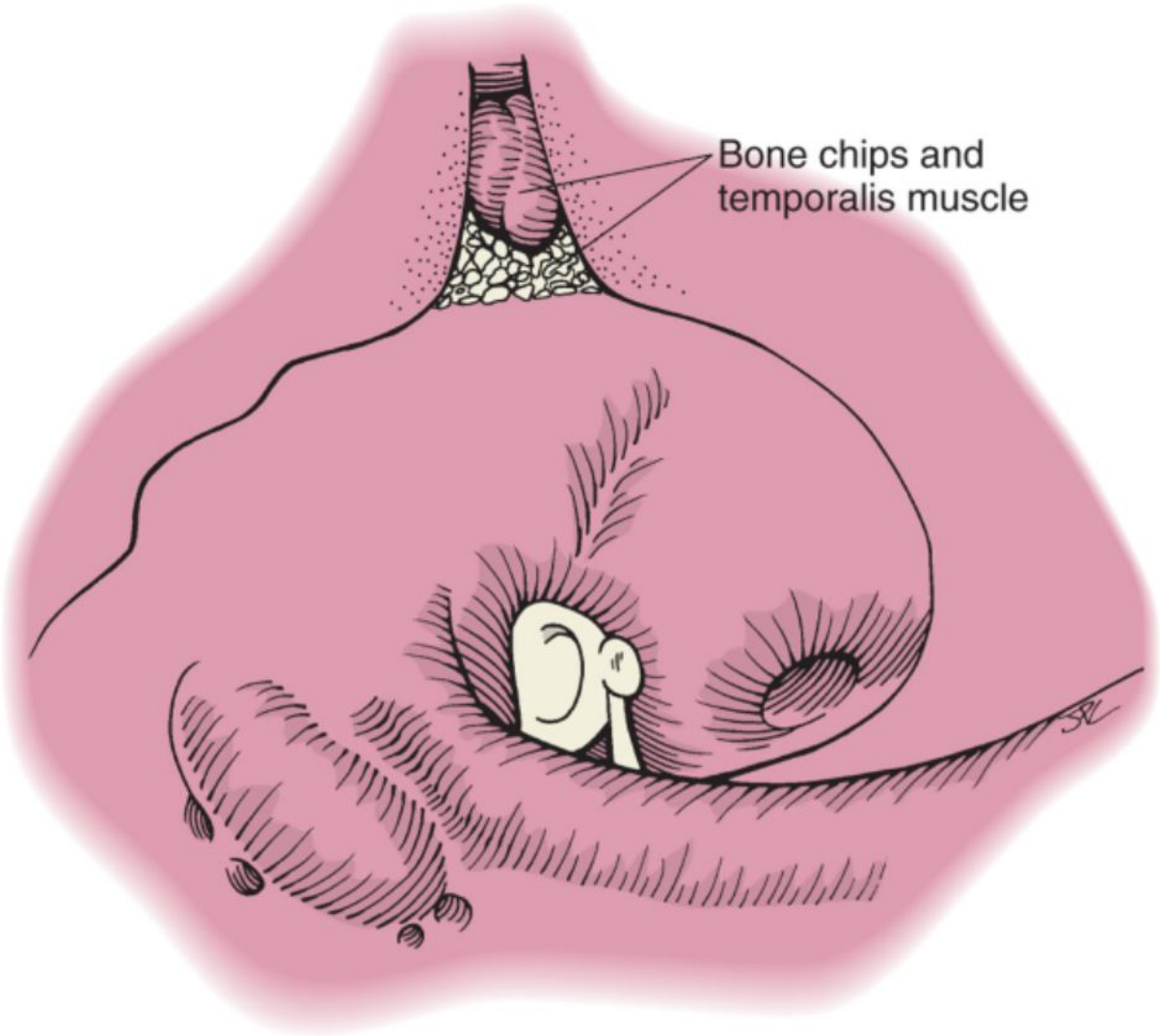

Figure 127-12 Obliteration of the eustachian tube with bone chips and temporalis muscle.

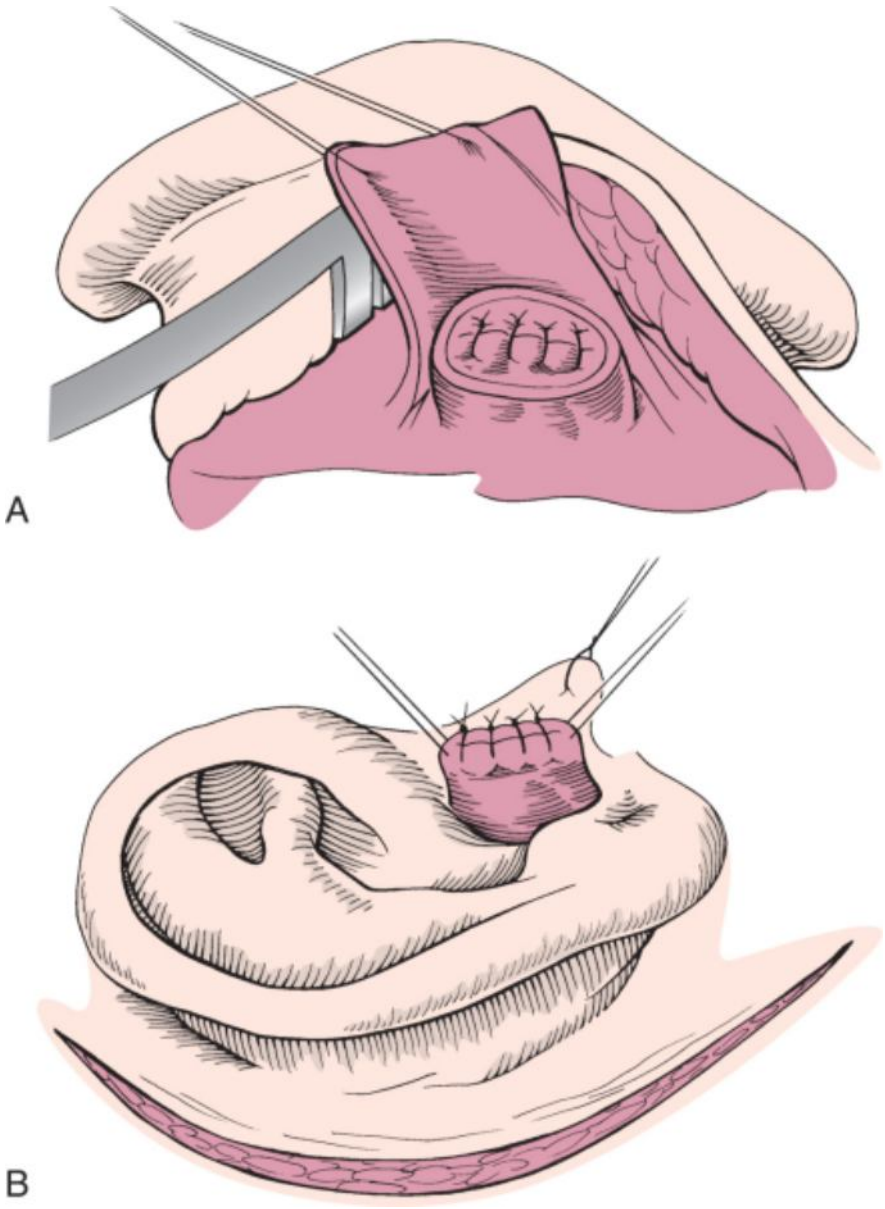

Figure 127-13 **A,** Sealing of the external meatus by sewing a fibrofascial flap to its medial surface. **B,** Completion of the meatal closure with everting sutures placed.

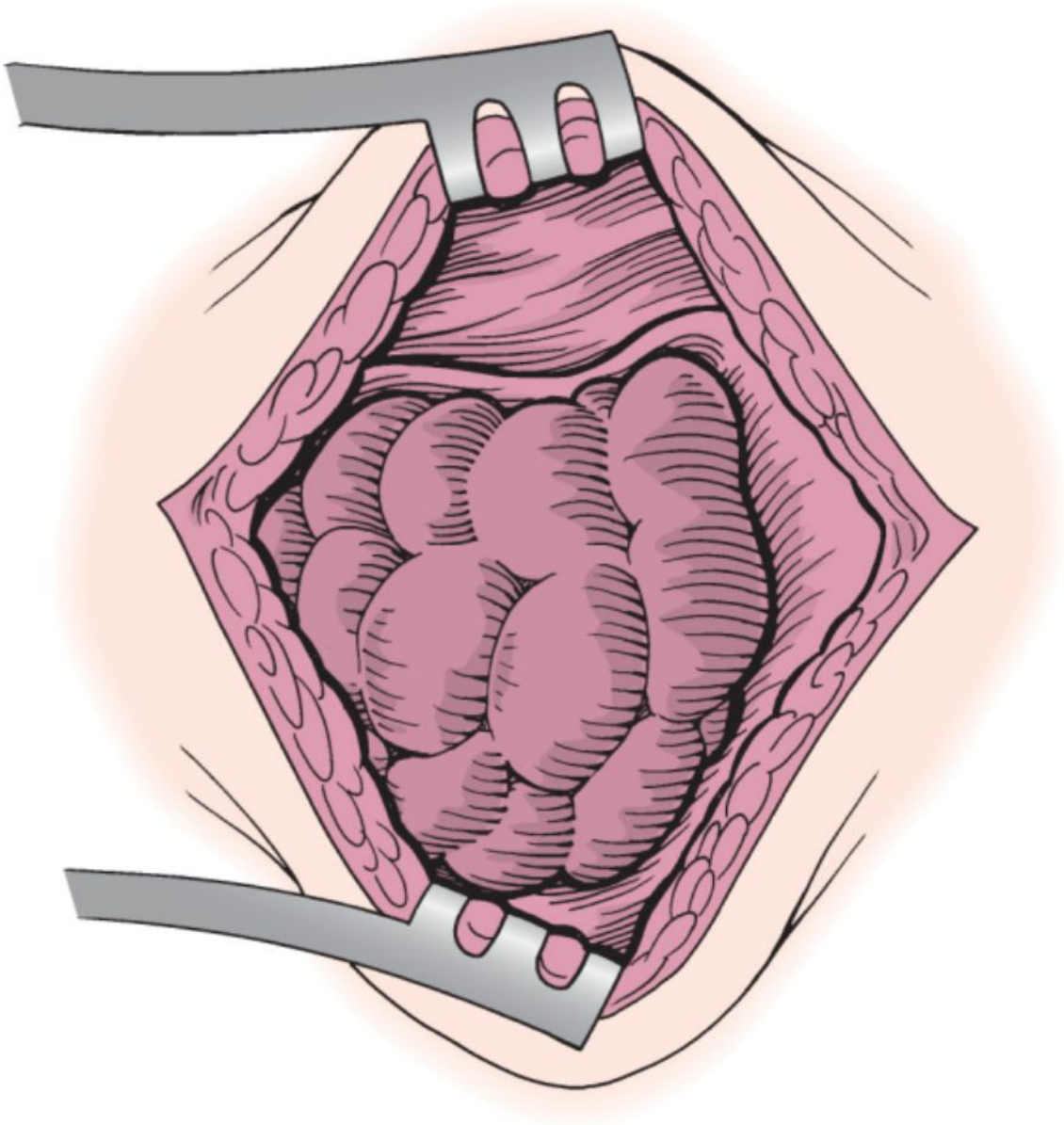

Figure 127-14 Obliteration of the mastoid with a fat graft taken from the abdominal wall.

Following completion of repair and before wound closure, the head is elevated 45 degrees; this is continued in the recovery room and throughout the hospitalization. Because of the potential for intracranial infection, a bolus of intravenous antibiotic is always given perioperatively. We use third-generation cephalosporins with broad-spectrum coverage. When purulent exudate is encountered at the time of surgery, cultures help determine the choice of antibiotic.

POSTOPERATIVE MANAGEMENT

Postoperatively, the head is elevated 45 degrees during the hospital stay and patients are encouraged to continue this for 7 to 10 days at home. Patients who have undergone craniotomy for repair of CSF leaks should be observed in the hospital for at least 3 to 4 days. Those who have undergone transmastoid surgery may generally be discharged home in 1 to 2 days. Antibiotic coverage is also continued intravenously until the patient is discharged, and then a similar agent is given orally for 7 days. When lumbar drainage has been placed preoperatively, it is continued for 24 to 72 hours postoperatively, depending on the extent of CSF leakage and size of defect repaired. The subarachnoid drain is then clamped for an additional 24 hours to confirm the absence of any further CSF leak, and then removed.

Persistent CSF leak is the most common complication following surgery. Because of the blood within the middle ear postoperatively, visualization of the tympanic membrane is not a good guide to successful cessation of CSF leakage in the immediate postoperative period. However, within 3 to 4 weeks, the middle ear should be pneumatized and clearly visualized. The patient is also questioned repeatedly and asked to report any incidence of

nasal or posterior pharyngeal drainage, which might indicate continued CSF otorrhea. Restraints against lifting, straining, and exercise are encouraged for 2 weeks following surgery.

Although temporary aphasia and seizures are potential postoperative complications from temporal lobe resection or retraction, our patients have had no problems related to the excision of nonviable brain tissue. Neurologic deficits are not expected postoperatively, and headache has not been a significant problem. Meningitis and brain abscess are potential complications, but these have usually been seen preoperatively rather than postoperatively.

PEARLS

- The vast majority of traumatic CSF leaks resolve with conservative management alone.
- Dural exposure alone without violation at the time of otologic surgery does not lead to CSF leak.
- Due to a significant incidence of multiple tegmen defects in spontaneous CSF leaks, the middle fossa approach allows for thorough inspection of the middle fossa floor before surgical repair.
- The best results are obtained with multilayer closure techniques using autologous tissue.
- In a nonhearing ear, obliteration of the middle ear and mastoid with blind sac closure of the external auditory canal offers the most effective and reliable repair for CSF leaks and large encephaloceles.

PITFALLS

- Failure to recognize multiple defects on preoperative CT imaging may lead to incomplete repair and persistent postoperative CSF leak.
- Failure to circumferentially elevate dura around the bony defect will not allow for reliable graft placement and repair.
- Primary dural closure alone is usually not adequate for repair of CSF leaks.
- The use of fixatives, such as fibrin glue, does not improve surgical success rates.
- With large tegmen defects, the weight of the temporal lobe alone, without the use of a supporting bone or cartilage graft, is not adequate for supporting a soft tissue graft over the defect.