Cerebrospinal Fluid Leaks of Temporal Bone Origin: Selection of Surgical Approach

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ABSTRACT

Cerebrospinal fluid leaks of the temporal bone are rare, often occult, and sometimes challenging to localize and repair. This is a retrospective study of eight patients with spontaneous cerebrospinal fluid leak and six patients with cerebrospinal fluid leak or encephalocele discovered during chronic ear surgery who were treated in a tertiary medical center over a 5-year period. All received preoperative temporal bone computed tomography, and six also underwent magnetic resonance imaging, one computed tomography cisternography, and one radionuclide cisternography. All patients initially underwent a transmastoid surgical approach. Additional exposure was necessary in three patients; two underwent middle fossa craniotomy and another required minicraniotomy. Primary surgical repair was successful in six of the eight patients with spontaneous leaks and in all six chronic ear patients. Both recurrences required intradural middle fossa repair. An individualized approach should be taken for repair of temporal bone cerebrospinal fluid leaks. In this series, most were successfully repaired in a single stage using a transmastoid or combined approach. The transmastoid approach provides information about the precise size and location of the dural defect. A primary transcranial approach is needed for defects that are multiple, located in the petrous apex, and in revision cases.

KEYWORDS: Cerebrospinal fluid leak, temporal bone, transmastoid approach, middle fossa craniotomy, encephalocele

Cerebrospinal fluid (CSF) leak of temporal bone origin is an uncommon condition with multiple potential etiologies. It most frequently occurs in the setting of traumatic temporal bone fracture, and less often may arise spontaneously, secondary to erosive chronic otitis media, or as a result of surgical injury.

The incidence of CSF leak involving the temporal bone has not been clearly established, although it is believed to be less common than those involving the anterior skull base. A large series of traumatic temporal bone fractures found the incidence of CSF leak to be 17%,¹ whereas another report noted brain

herniation and/or CSF leak in ~1% of patients undergoing revision surgery for chronic middle ear disease.²

CSF leaks of the temporal bone pose certain diagnostic and management challenges. Symptoms can be subtle and can be present for years prior to diagnosis. In some cases, patients may present acutely with potentially fatal intracranial infection. Once a leak is determined to be present, available imaging methods may not localize the source. There are several options for surgical repair, which include transmastoid, middle fossa, or combined approaches.

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A condition that predisposes to CSF leaks of temporal origin is temporal bone encephalocele, an acquired or developmental herniation of dura and brain tissue through bony defects, usually in the region of the tegmen. This condition may lead to CSF leaks, either spontaneously or iatrogenically during middle ear surgery.

In this paper, the diagnosis and management of spontaneous and acquired encephalocele and otogenic CSF leak are considered. CSF leak should be suspected in patients presenting with persistent serous effusion or meningitis. The rationale for attempting transmastoid surgical repair for most patients is discussed and case reports are presented that illustrate key issues involved in managing this condition.

METHODS

A retrospective chart review examined 14 patients treated in a tertiary medical center over a 5-year period following institutional review board approval. These patients were identified by the senior author (E.S.) as having been treated for CSF leak or encephalocele of temporal bone origin. Eight patients had spontaneous CSF leaks, and six had CSF leaks or encephalocele discovered intraoperatively during chronic ear surgery. Patients who developed temporal bone CSF leaks following acoustic neuroma surgery were excluded from study involvement, as were patients with anterior skull base leaks.

Data were collected from office and hospital charts regarding patient demographics, including age, gender, and comorbid conditions. Also accumulated was data on preoperative diagnostic tests, including laboratory tests, preoperative audiograms, and imaging. Finally, information was recorded on surgical approach, intraoperative findings and method of repair, length of postoperative follow-up, postoperative complications, postoperative audiograms, and on whether recurrence occurred.

RESULTS

Patient Demographics

Fourteen patients underwent 16 operations for CSF leak or encephalocele of temporal bone origin. Nine of the patients were male. The median age at diagnosis was 47, with a range between 13 and 76. Data regarding body mass index were not recorded at the time of clinical evaluation in the patients studied, although one female patient with a spontaneous leak was morbidly obese. No patient had undergone prior surgical repair attempts at outside institutions.

Clinical Presentation

Of the eight patients with spontaneous CSF leaks, four had a history of meningitis. Otorrhea was a symptom in

seven patients, occurring following tympanostomy tube placement or in the presence of a concurrent tympanic membrane perforation. Four patients also complained of rhinorrhea, whereas hearing loss or aural fullness was present in four patients. β -2-Transferrin testing confirmed the presence of a leak in two patients. The average duration of symptoms potentially attributable to spontaneous CSF leak was 7 years. Of the six chronic ear patients with intraoperative temporal bone encephaloceles or active CSF leaks, all presented with chronic conductive hearing loss and one patient with persistent otorrhea.

Preoperative computed tomography (CT) was obtained in all patients. All underwent high-resolution noncontrast temporal bone CT in the axial and coronal planes using 1-mm contiguous sections, with bone algorithm to enhance resolution. Temporal bone magnetic resonance imaging with and without gadolinium contrast was also performed in five patients. Two patients underwent radionuclide cisternography using intrathecal technetium 99m-DTPA, and high-resolution temporal bone CT cisternography using iohexol was performed in one patient. Correlation with preoperative CT and intraoperative defect localization was present in six of eight patients with spontaneous leaks. Of the chronic ear patients, preoperative CT suggested a bony skull base defect in two patients. In the period between diagnosis and repair, prophylactic antibiotics were not employed in any of the patients.

Surgical Approach, Intraoperative Findings, and Repair Materials

Selection of surgical approach and materials chosen for repair were based on several factors including the location of the temporal bone defect, presence of a recurrent leak, and surgeon preference. All patients underwent a transmastoid surgical approach during initial repair (Table 1). Two patients also underwent a transcranial approach at the time of primary repair, and one patient underwent minicraniotomy.

Of the patients with spontaneous leaks, tegmen defects were identified intraoperatively in six patients (Table 2). Six patients had a posterior fossa defect just inferior to the superior petrosal sinus, and six patients had a defect in the petrous apex. Encephaloceles were visualized intraoperatively in four spontaneous leak patients, and multiple tegmen defects were found at the time of primary repair in one patient.

Table 1 Approach to Repair

Initial Approach	Number
Transmastoid	14
+Middle fossa craniotomy	2
+Minicraniotomy	1

Table 2 Intraoperative Findings

Intraoperative Findings	Number
Tegmen defects	12
Petrous apex defect	1
Posterior fossa defect	1
Encephalocele	7

Of the chronic ear patients, all bony defects were localized to the tegmen mastoid/tympani. Three had intraoperative CSF leaks alone, two had encephaloceles and no active leak, and one had both an active CSF leak and encephalocele.

Three patients required additional exposure via either middle fossa approach or minicraniotomy, as noted previously. In two cases, brain herniation through a tegmen defect was visualized via transmastoid exposure, but the encephalocele could not be reduced without supplemental access. In one other patient, a transmastoid approach did not visualize any defect and middle fossa craniotomy was necessary to localize a bony dehiscence in the petrous apex.

All the defects were repaired using multilayered closure with a combination of autologous, allogeneic, and synthetic materials. Autologous materials included abdominal fat, temporalis muscle/fascia, conchal cartilage/perichondrium, and calvarial bone. Synthetic materials included fibrin glue, hydroxyapatite bone cement, and vicryl mesh. Acellular dermis (Alloderm, LifeCell, Branchburg, NJ) was used in two patients.

Hearing Status

Efforts to preserve middle ear structures for patients with residual hearing were attempted in most patients. Middle ear obliteration was performed in two patients undergoing transmastoid repair. One patient had profound preoperative hearing loss, and the other was the patient described above in which transmastoid exposure alone could not localize a defect.

Postoperative Course

Average follow-up was 13 months. Lumbar drains were used postoperatively in four patients following primary repair and in both patients undergoing secondary repair. Lumbar puncture without lumbar drain was performed in one patient postoperatively. No postoperative infections occurred. Pre- and postoperative audiograms were available for five of eight patients with spontaneous leaks; conductive hearing thresholds were found to improve in three of these patients. Of the chronic ear patients, pre- and postoperative hearing thresholds remained unchanged.

Recurrent leaks occurred in two patients. One patient with a recurrent leak initially underwent transmastoid approach and minicraniotomy for a tegmen mastoid defect. On secondary repair, the patient was found to have displaced bone/cartilage graft; repair was successfully accomplished with a combined transmastoid/middle cranial fossa approach. Another patient with a recurrent leak had a history of skull base irradiation during childhood. As initial repair involved middle ear obliteration, secondary repair was performed via middle fossa craniotomy alone, where she was found to have multiple dehiscences in both the tegmen and petrous apex.

Case Examples

The following cases highlight specific issues:

CASE 1

A 49-year-old man with insulin-dependent diabetes presented to an outside hospital emergency department in coma secondary to resistant pneumococcal meningitis. During a prolonged hospital course, he underwent bilateral tympanostomy tube placement for serous effusion. Following resolution of his intracranial infection, the patient complained of persistent left clear otorrhea. He underwent noncontrast temporal bone CT (Fig. 1A), showing a bony defect of the tegmen mastoid. CT

Figure 1 (A) Noncontrast temporal bone CT showing tegmen defect. (B) CT cisternography showing pooling of contrast in the mastoid. CT, computed tomography.

Figure 2 (A) Transmastoid approach shows dural defect in tegmen mastoid. (B) Conchal cartilage placed into dural defect.

cisternography was also performed showing pooling of contrast in the mastoid (Fig. 1B). The patient underwent surgical repair via a transmastoid approach, which revealed an active CSF leak through a dural defect in the tegmen mastoideum (Fig. 2A). The defect was repaired through the mastoid using conchal cartilage, temporalis fascia, and fibrin glue, followed by fat obliteration of the mastoid (Fig. 2B). The leak did not recur after 3 years of follow-up.

This case illustrates the devastating complications that can result from an occult CSF leak. The patient in this example was fortunate to have survived a life-threatening episode of meningitis prior to diagnosis and repair of the CSF leak. He reported retrospectively that he had unilateral hearing loss for a few years, suggesting a middle ear CSF effusion that was not previously recognized or treated.

CASE 2

A 34-year-old man complained of worsening hearing in his left ear, his better hearing ear. He had a history of bilateral cholesteatomas and had previously undergone bilateral canal wall down mastoidectomies. Physical

examination revealed a mass in the left mastoid bowl covering the tympanic membrane, originating from the tegmen. CT showed a tegmen defect with soft tissue in the middle ear. At surgery, an encephalocele was identified (Fig. 3A), along with a small recurrent cholesteatoma. The dural edges were freed, and the encephalocele was amputated, causing a brief intraoperative CSF leak. The tegmen defect was repaired using conchal cartilage, hydroxyapatite bone cement, and fibrin glue (Fig. 3B). Because of the open mastoid cavity and the desire to maintain hearing, fat obliteration was not performed. No recurrence was observed, but the patient did develop partial exposure and desiccation of the bone cement layer, necessitating shave excision of the cement and local soft tissue flap coverage 18 months later.

Encephalocele occurring in an open mastoid cavity poses a higher risk of CSF leak and meningitis. Ordinarily, the safest way to deal with this situation is to obliterate the cavity with fat and oversew the meatus. However, in this case, we wanted to avoid overclosure for the sake of hearing preservation and cholesteatoma surveillance; hence, the tegmen defect

Figure 3 (A) Intraoperative finding of encephalocele in open mastoid cavity. (B) Repair using cartilage, bone cement, and fibrin glue.

was reconstructed with hydroxyapatite bone cement. This material provides a secure seal but requires adequate soft tissue coverage, which necessitated a secondary procedure.

CASE 3

A 40-year-old woman, who recently recovered from an episode of meningitis, developed persistent left clear otorrhea after tympanostomy tube placement for presumed serous effusion. CT showed opacification of the left middle ear/mastoid space and a possible posterior fossa defect in the region of the sigmoid sinus (Fig. 4). Operative approach was initially via transmastoid exposure alone, which did not identify a posterior or middle fossa defect. Middle fossa craniotomy was then performed, which revealed a dural defect above the petrous apex. She underwent intradural repair bolstered by temporalis muscle/fascia and mastoid fat obliteration. A lumbar drain was used postoperatively, and there was no recurrence.

The site of leakage can be difficult to ascertain by any imaging modality, and the presence of a bony dehiscence on CT is not definitive. Transmastoid exploration of the tegmen and posterior fossa plate usually identifies the site of the leak and provides access for repair. In this case, however, the leak was over the petrous apex and could not be exposed by this route, so a middle fossa procedure was necessary.

Figure 4 Suspected bony defect on preoperative CT. CT, computed tomography.

DISCUSSION

CSF leaks of temporal bone origin have multiple potential causes. Trauma is most common, whether related to temporal bone fracture or following surgery of the posterior fossa or mastoid. Spontaneous leaks can also rarely occur in the absence of trauma. In children, these are usually due to congenital otic capsule abnormalities, such as Mondini deformities. In adults, spontaneous leaks are less well understood. Theories include bony erosion by arachnoid granulations,³ or CSF pulsations from undiagnosed intracranial hypertension, especially in obese individuals.^{4,5}

Occult CSF leaks should be suspected in cases of refractory unilateral serous otitis media. Although this condition is relatively rare, CSF otorrhea must be included in the differential diagnosis of a nonresolving effusion. It is often first discovered after tympanostomy results in profuse clear drainage. Rhinorrhea (via the eustachian tube) may not occur or may go undetected. One of our patients reported that her chronic cough resolved after CSF leak repair; in retrospect, this was caused by occult CSF rhinorrhea causing a relentless postnasal drip that was present for years. As noted previously, sometimes the initial presentation of an occult CSF leak may be life-threatening meningitis.

Testing of β -2-transferrin in samples of otorrhea/rhinorrhea fluid is highly sensitive and specific for identifying CSF.⁶ Collecting a sufficient aliquot remains a problem, however. Another more recently utilized marker, β -trace protein, has higher specificity than β -2-transferrin and potentially allows for earlier result interpretation due to more rapid laboratory processing.⁷

Imaging studies are essential for determining the site of the CSF leak in the temporal bone. High-resolution noncontrast CT (HRCT) is the initial diagnostic study of choice in all cases and may be the only imaging necessary. HRCT will usually identify the bony defect of the temporal bone but will not demonstrate the site of the dural tear. Other imaging studies are potentially helpful. Magnetic resonance imaging can distinguish middle ear/mastoid encephalocele from chronic ear disease, as the CSF signal will be bright on T-2 weighted images, and an encephalocele will appear contiguous with brain. CT cisternography will demonstrate the leak but can be falsely negative if the leak is intermittent or of low volume. This study can be valuable in cases where more than one potential defect exists on HRCT,⁸ although (as seen in case 1) it may not add new diagnostic information or alter management strategy for patients with isolated defects. Radionuclide cisternography is sometimes employed when a questionable leak exists. This study is very sensitive for detecting occult CSF otorhinorrhea—when radioactive tracer injected intrathecally appears on cottonoid strips placed in the ear canal and nasal cavities, it is fairly certain that a leak

exists—although the study is limited in its ability to localize the leak because of its low resolution.

Certain principles of management of CSF leaks of the temporal bone are generally agreed upon. Traumatic CSF leaks often close with conservative management, but spontaneous or iatrogenic CSF leaks will almost always require surgical repair. The risk of meningitis increases with traumatic leaks that persist beyond 1 week,¹ so surgical intervention should be performed early. Regarding closure techniques, the highest likelihood of successful repair has been shown to be with multilayered closure, usually using a combination of autologous and synthetic materials.⁹

Selection of surgical approach for repair of middle fossa defects remains a matter of individual judgment. No prospective randomized trial exists comparing the success of various approaches for surgical exposure and repair; hence, the available literature reflects individual surgeon experience. A retrospective series of 55 patients managed surgically for temporal bone CSF leaks found no significant differences in recurrence based on approach to repair,⁹ although interpretation of results may be limited by selection bias.

The transmastoid approach has the advantage of allowing localization of the dural defect in the temporal bone while avoiding brain retraction and potential neurological complications. Our case series and the experience of others suggest that a transmastoid approach alone may be adequate to treat most dural defects.^{2,9–11} Cases 1 and 2 illustrate situations in which this approach is sufficient—where an isolated tegmen defect can be exposed in entirety and repaired from below using a multilayered closure. Potential disadvantages of the transmastoid approach include decreased exposure of the defect, lower initial success rate and durability of the repair, and increased risk of hearing loss.

The middle fossa craniotomy has long been documented as an effective treatment for this condition.^{3,12–17} Its advantages include a direct route and improved exposure of the entire tegmen plate and petrous roof for a secure intradural repair with little chance of recurrence. Additionally, hearing is preserved with this approach in the great majority of cases. Potential disadvantages of craniotomy include the risk of neurological complications such as venous infarction related to the vein of Labbé and seizure.

Our experience has led us to favor a transmastoid approach as the initial route of access to CSF leaks of the temporal bone in most cases. In many instances this provides sufficient exposure to precisely define the defect and repair it. An intracranial approach is needed in cases of large, multiple, or medial defects or revision procedures for recurrent CSF leaks. We often utilize a combined transmastoid/transcranial approach to bolster the security of our repair, and this technique has a low demonstrated risk of failure.

Through transmastoid exposure, we situate cartilage or bone in the tegmen defect, place a fat graft in the mastoid, and occlude the eustachian tube. Formal temporal craniotomy then allows for intradural repair with an oversized graft of fascia or acellular dermis (Alloderm) placed between the middle fossa floor and the temporal lobe.

Whatever approach is chosen for the initial repair, we have found that preoperative collaborative planning by experienced skull base otolaryngologists and neurosurgeons is a key component of managing patients with CSF leaks of temporal origin and temporal encephaloceles.

CONCLUSION

CSF leaks of temporal bone origin are rare and should be considered in patients with nonresolving middle ear effusion or meningitis. Temporal bone CT represents the most valuable imaging modality for diagnosis and preoperative surgical planning. Isolated dural defects of the tegmen may be repaired through transmastoid approach alone, whereas multiple defects, defects in the petrous apex, and revision cases usually require an intracranial approach.

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