Spontaneous transtemporal CSF leakage: A study of 51 cases

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Abstract
We conducted a retrospective study of 51 cases of spontaneous transtemporal cerebrospinal fluid (CSF) leakage in 48 adults who had presented to our tertiary care academic referral center between July 1, 1988, and June 30, 2002. All patients had undergone high-resolution temporal bone computed tomography, and 26 patients had undergone magnetic resonance imaging. All patients were treated with a middle fossa craniotomy to repair the CSF fistulae. During a mean follow-up of 4.9 years, 46 of the 48 patients (95.8%) had experienced a complete cessation of CSF leakage (49 of 51 cases [96.1%]). The 2 patients whose leakage recurred were successfully managed with a subtotal petrosectomy with occlusion of the eustachian tube and obliteration of the middle ear and mastoid. No patient developed meningitis.

Introduction
Cerebrospinal fluid (CSF) leakage through the temporal bone can be either acquired, congenital, or spontaneous. Acquired CSF leakage can be caused by skull trauma, surgical intervention (iatrogenic leak), infection, or neoplastic invasion of the temporal bone. Congenital leakage is most often associated with the development of anomalous transcranial pathways. Spontaneous leakage occurs in the absence of any apparent precipitating event or explainable congenital anomaly. Some authors who have addressed the topic of spontaneous CSF fistulae believe that it is rare.

In this article, we describe our experience in diagnosing and managing 51 episodes of spontaneous CSF leakage in 48 patients. Our aim is to raise awareness of this condition in the hope that this diagnosis will be considered prior to myringotomy and tympanostomy tube placement.

Patients and methods
We undertook a retrospective review to identify cases of spontaneous CSF leakage that had been treated at our tertiary care academic referral center. We reviewed the records of all patients who had been diagnosed with CSF leakage between July 1, 1988, and June 30, 2002. After excluding from our study all cases of traumatic, surgically iatrogenic, neoplastic, infectious, and congenital leakage, we identified 51 cases of spontaneous leak in 48 patients. We further researched these cases by examining outpatient, inpatient, and surgical records to obtain data on presenting signs and symptoms, radiographic findings, intraoperative findings, and long-term results.

Results
The study group was made up of 28 women and 20 men, aged 28 to 92 years (mean: 45.7). The CSF fistulae were located on the right in 24 patients, on the left in 21 patients, and bilaterally in 3 patients.

Presenting manifestations. The most common presenting symptoms were aural fullness and hearing loss, while the most common physical findings were clear middle ear effusion and clear tympanostomy tube otorrhea (table). The duration of symptoms ranged from 6 months to 25 years (mean: 5.8 yr). Three patients (6.3%) had a history of unexplained meningitis.

Radiographic findings. All patients underwent high-resolution temporal bone computed tomography (CT), and 26 of the 51 leaks were evaluated by magnetic resonance imaging (MRI). Coronal CT revealed discrete, single tegmen defects, multiple pinhole tegmen fistulae, or a combination of these findings (figure 1). No additional posterior fossa defects were identified on CT. MRI detected middle ear and mastoid fluid that was consistent with CSF (figure 2).

Other diagnostic studies. All patients had undergone routine audiometric evaluation. Most patients had an ipsilateral conductive hearing loss or a mixed hearing loss.
Radionucleotide and B-2 transferrin testing had not been used in making the diagnosis of CSF leakage.

Surgical technique. All 51 cases of transtemporal CSF leakage were surgically repaired via a middle fossa craniotomy approach. An inferiorly based, upside-down, U-shaped incision was made through the skin and subcutaneous tissue, centered on the external auditory canal (figure 3, A). A thick temporalis fascia graft was harvested for dural defect repair, and the temporalis muscle was reflected anteroinferiorly (figure 3, B). A rectangular middle fossa bone flap was elevated, and a corner of the flap was used to repair the bone defect on the middle fossa floor.

When encountered, brain herniations were bipolar-coagulated, and pinhole dural defects were closed with 6-0 Neurilon (figure 4). Larger dural defects were repaired with temporalis fascia graft patching. The floor of the middle fossa and posterior fossa was flattened with diamond-stone drilling, and larger tegmen defects were bridged with free bone grafts (figure 5). The bone flap was secured, the wound was closed in layers, a suction drain was inserted, and a mastoid-type compressive dressing was used for 2 or 3 days. Lumbar drainage was not performed intraoperatively or postoperatively, which allowed for early identification of any possible inadequate defect repairs.

Postoperative care. All patients were placed in a neurosurgical intermediate care unit overnight; thereafter, they remained hospitalized for an additional 2 to 4 days. Upon discharge, patients were instructed to refrain from heavy lifting, nose-blowing, exercising, straining, and airplane travel for 1 month. Sutures were removed 7 days postoperatively. Standard postoperative follow-up visits were scheduled for 1, 3, and 6 months and annually thereafter for 5 years.

Complications. Minor wound complications occurred in 4 patients—3 seromas and 1 hematoma. All 4 cases were managed with fluid aspiration and a compressive dressing.

Table. Presenting manifestations in the 51 cases of spontaneous CSF leak

<table>
<thead>
<tr>
<th>Symptoms</th>
<th>n</th>
<th>Signs</th>
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<tbody>
<tr>
<td>Aural fullness</td>
<td>46</td>
<td>Clear effusion</td>
<td>33</td>
</tr>
<tr>
<td>Hearing loss</td>
<td>42</td>
<td>Otorrhea</td>
<td>18</td>
</tr>
<tr>
<td>Tinnitus</td>
<td>8</td>
<td>Pulsatile TM*</td>
<td>3</td>
</tr>
<tr>
<td>Imbalance</td>
<td>6</td>
<td>Rhinorrhea</td>
<td>3</td>
</tr>
<tr>
<td>Headache</td>
<td>2</td>
<td>Perforated TM*</td>
<td>1</td>
</tr>
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* TM = tympanic membrane.
One patient developed a delayed-onset incomplete facial palsy, which resolved completely over 2 months. Two patients experienced prolonged postoperative headache that required referral to our pain management service and long-term narcotic therapy.

Conductive hearing deficits were restored to within 10 dB throughout the speech frequencies after 46 of the 51 operations (90.2%). Conductive hearing loss worsened in 1 patient, and 2 patients experienced a worsening of sensorineural hearing loss (SNHL).

Recurrence. Two patients developed recurrent CSF leaks, as evidenced by clear middle ear effusion; one recurrence manifested 28 months postoperatively and the other at 32 months. Both recurrences were attributed to a prolapse of the dural repair into the epitympanum. Both patients were elderly, and both had ipsilateral moderate-to-severe SNHL. These 2 patients underwent subtotal petrosectomy, oversew of the cartilaginous ear canal, occlusion of the eustachian tube, and obliteration of the middle ear and mastoid. One of these patients remained leak-free at 4 years of follow-up and the other at 2 years.

Discussion
Spontaneous CSF temporal bone leakage might occur as a result of an encephalocele or secondary to the development of arachnoid granulations along the tegmen of the middle ear or mastoid. Histologic temporal bone studies suggest that 6% of the population has some degree of bony defect in this area. Thinning of the dura with age or as a result of brain pulsation may result in dural tear and ensuing CSF leakage into the middle ear and/or mastoid. Intermittent plugging with brain tissue may explain the fluctuating symptoms and infrequent otorhinorrhea seen in these patients.

The duration of symptoms in our series, as in other published reports, varied widely. Aural fullness and hearing loss, the most common presenting symptoms in our series, had been present for as long as 25 years (mean:

Figure 3. A middle fossa scalp flap (A) and a temporalis fascia graft (B) are used to repair bony and dural defects.

Figure 4. Intraoperative views show a bony defect in the middle cranial fossa floor (A, arrow) and a small brain herniation (B, asterisk).
5.8). A history of meningitis was documented in 3 of the 48 patients (6.3%).

Making a diagnosis of CSF middle ear effusion on the basis of otoscopic findings can be difficult because a middle ear filled with clear fluid may appear to be normal. Microscopic otoscopy should facilitate the differentiation between serous (straw-colored) and CSF (clear) effusion. Myringotomy, with or without tympanostomy tube placement, will result in the development of clear and continuous otorrhea, which will increase with straining (figure 6).

When CSF middle ear effusion is suspected, coronal CT of the temporal bone should be obtained prior to office-based myringotomy. In our series, CT correlated well with the identification of single or multiple tegmen defects. Keeping the system sealed may reduce the risk of otitic meningitis. MRI added little to the diagnosis or treatment planning in our series.

A variety of transtemporal approaches can be employed in the surgical management of CSF middle ear or mastoid effusion. We prefer a subtotal petrosectomy with oversew of the ear canal, direct eustachian tube occlusion, and middle ear and mastoid obliteration in patients with severe-to-profound SNHL or a history of recurrent meningitis.

We used the middle fossa approach for all 51 operations. This approach allows for extradural elevation of the temporal lobe along the entire floor of the middle fossa, identification of all dural defects, and direct repair of the dura and bony defects. Repair from above rather than below (transmastoid) offers the best chance for long-term cessation of CSF leakage because it negates the gravitational effects of constant brain pulsation over repaired tegmen defects. This approach was successful in halting CSF leakage after 49 of the 51 operations (96.1%) over a mean follow-up of 4.9 years.

Recurrent CSF otorrhea following middle fossa repair may occur as a result of unrecognized or inadequately repaired defects. Subtotal petrosectomy with oversew of the external auditory canal allows for direct occlusion of the eustachian tube and obliteration of all potential transtemporal routes of CSF leakage. This approach is also used in patients with significant hearing loss or multiple episodes of otitic meningitis following the development of temporal CSF fistulae.

In conclusion, spontaneous CSF leakage can occur as a result of bony defects over the tegmen tympani or tegmen mastoideum. Clear middle ear effusion or otorrhea should alert the physician to this possibility. High-resolution coronal CT will demonstrate the site or sites of the leakage. In our series, middle fossa craniotomy allowed for hearing preservation and cessation of CSF leakage in 96.1% of cases.

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presentation. Patients with liposarcoma generally present with a painless mass that has caught their attention only after it has achieved significant size.

On gross pathologic examination, liposarcomas are typically well encapsulated, although microscopic examination can demonstrate infiltrative margins. Tumors are represented histologically by moderately sized malignant lipoblasts set in a myxoid background with a rich capillary network. There are five histologic subtypes: myxoid, well-differentiated, round-cell, pleomorphic, and dedifferentiated. Dedifferentiated liposarcomas have the appearance of well-differentiated liposarcomas except for areas of dedifferentiation with a nonlipoblastic sarcomatous component. Regional metastasis is rare. Distant metastasis via hematogenous spread is more common in the poorly differentiated forms; Zagars et al reported a rate of 37%.3

The treatment of choice is wide local excision. The efficacy of radiation as a primary or adjuvant therapy is debatable, but it has a definite role in treating partially resected or unresectable tumors. Chemotherapy may play a limited role in the myxoid type of tumor. Zagars et al reported a 10-year survival rate of 39% for poorly differentiated forms.3

The case described herein illustrates that clinical suspicion should be high for malignancy when atypical symptoms are associated with a seemingly benign lesion.

References