Temporal bone encephalocele and cerebrospinal fluid fistula repair utilizing the middle cranial fossa or combined mastoid–middle cranial fossa approach

Clinical article

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Object. The goals of this study were to report the clinical presentation, radiographic findings, operative strategy, and outcomes among patients with temporal bone encephaloceles and cerebrospinal fluid fistulas (CSFFs) and to identify clinical variables associated with surgical outcome.

Methods. A retrospective case series including all patients who underwent a middle fossa craniotomy or combined mastoid–middle cranial fossa repair of encephalocele and/or CSFF between 2000 and 2012 was accrued from 2 tertiary academic referral centers.

Results. Eighty-nine consecutive surgeries (86 patients, 59.3% women) were included. The mean age at time of surgery was 52.3 years, and the left side was affected in 53.9% of cases. The mean delay between symptom onset and diagnosis was 35.4 months, and the most common presenting symptoms were hearing loss (92.1%) and persistent ipsilateral otorrhea (73.0%). Few reported a history of intracranial infection (6.7%) or seizures (2.2%).

Thirteen (14.6%) of 89 cases had a history of major head trauma, 23 (25.8%) were associated with chronic ear disease without prior operation, 17 (19.1%) occurred following tympanomastoidectomy, and 1 (1.1%) developed in a patient with a cerebral aqueduct cyst resulting in obstructive hydrocephalus. The remaining 35 cases (39.3%) were considered spontaneous. Among all patients, the mean body mass index (BMI) was 35.3 kg/m2, and 46.4% exhibited empty sella syndrome. Patients with spontaneous lesions were statistically significantly older (p = 0.007) and were more commonly female (p = 0.048) compared with those with nonspontaneous pathology. Additionally, those with spontaneous lesions had a greater BMI than those with nonspontaneous disease (p = 0.102), although this difference did not achieve statistical significance.

Thirty-two surgeries (36.0%) involved a middle fossa craniotomy alone, whereas 57 (64.0%) involved a combined mastoid–middle fossa repair. There were 7 recurrences (7.9%); 2 patients with recurrence developed meningitis. The use of artificial titanium mesh was statistically associated with the development of recurrent CSFF (p = 0.004), postoperative wound infection (p = 0.039), and meningitis (p = 0.014). Also notable, 6 of the 7 cases with recurrence had evidence of intracranial hypertension. When the 11 cases that involved using titanium mesh were excluded, 96.2% of patients whose lesions were reconstructed with an autologous multilayer repair had neither recurrent CSFF nor meningitis at the last follow-up.

Conclusions. Patients with temporal bone encephalocele and CSFF commonly present with persistent otorrhea and conductive hearing loss mimicking chronic middle ear disease, which likely contributes to a delay in diagnosis. There is a high prevalence of obesity among this patient population, which may play a role in the pathogenesis of primary and recurrent disease. A middle fossa craniotomy or a combined mastoid–middle fossa repair incorporating a multilayer autologous tissue technique is a safe and reliable method of repair that may be particularly useful for large or multifocal defects. Defect reconstruction using artificial titanium mesh should generally be avoided given increased risks of recurrence and postoperative meningitis.

Key words: • idiopathic intracranial hypertension • cerebrospinal fluid fistula • cerebrospinal fluid leak • meningoencephalocele • encephalocele • meningitis • temporal bone • middle cranial fossa

Abbreviations used in this paper: BMI = body mass index; CPAP = continuous positive airway pressure; CSFF = cerebrospinal fluid fistula.

This article contains some figures that are displayed in color online but in black-and-white in the print edition.
Temporal bone encephalocele and CSF fistula repair

Surgical repair is recommended to decrease the risk of impending neurological complications.\textsuperscript{3,20,21}

Osseous erosion or bony insufficiency may result from trauma, prior surgery, long-standing infection, or destructive processes such as cholesteatoma or tumor. Over the last 2 decades, there has been a substantial increase in the number of reported “spontaneous” cases, where no discernable cause can be identified.\textsuperscript{2,10,17} Many of these subjects share clinical and radiographic features that overlap with idiopathic intracranial hypertension syndrome. It has been hypothesized that elevated intracranial pressure may contribute to progression of temporal bone encephaloceles and CSFFs, and an increased risk of recurrence following repair.\textsuperscript{1,9,22,26}

Given the rarity of the studied conditions, few large series exist in the literature, and optimal surgical management remains controversial.\textsuperscript{7,14,15,20,25} Treatment options include the transmastoid approach with tegmen repair or tympanomastoid obliteration, subtemporal middle fossa craniotomy, or a combined mastoid–middle fossa approach. Repairs may incorporate intradural or extradural overlay or underlay techniques utilizing a variety of autologous and synthetic substrates. Herein, we report our experience managing temporal bone encephaloceles and CSFFs over the last 12 years utilizing a middle fossa craniotomy or combined mastoid–middle cranial fossa approach. Clinical presentation, radiological findings, operative strategy, and outcomes are reported, and variables associated with outcome are explored.

Methods

Data Collection

After institutional review board approval, a retrospective chart review was conducted, and all patients who underwent surgical repair of a temporal bone encephalocele or CSFF over the last 12 years at 1 of 2 tertiary academic referral centers were identified. Data including clinical presentation, radiographic findings, method of repair, perioperative complications, and recurrence were gathered. Patients were excluded from the study if they had CSFF that was either associated with cochlear or labyrinthine malformations, or occurred following lateral skull base tumor resection (e.g., vestibular schwannoma, meningioma, or malignancy).

Lesions were classified as traumatic, iatrogenic, infectious, or spontaneous, the latter being assigned in the absence of an obvious underlying cause. In patients with a history of chronic otitis media who underwent prior tympanomastoidectomy, the category “infectious” was assigned if middle or posterior fossa dural dehiscence resulting from disease was noted at initial surgery; otherwise, such cases were designated “iatrogenic” by default. The presence of encephalocele with CSFF, encephalocele without CSFF, and CSFF without encephalocele, as well as the number (1, 2, or multiple), laterality (left, right, or bilateral) and size (< 1 cm, 1–2 cm, or > 2 cm) of bony defects were reported. The preoperative BMI was calculated using the formula BMI = weight (kg)/(height [m])\textsuperscript{2}, and subjects were categorized as underweight (< 18.5 kg/m\textsuperscript{2}), normal weight (18.5–24.9 kg/m\textsuperscript{2}), overweight (25–29.9 kg/m\textsuperscript{2}), obesity Class 1 (30.0–34.9 kg/m\textsuperscript{2}), obesity Class 2 (35.0–39.9 kg/m\textsuperscript{2}), or extreme obesity Class 3 (≥ 40 kg/m\textsuperscript{2}) in accordance with the National Institutes of Health guidelines.\textsuperscript{18} Preoperative and the most recent postoperative audiometric data, including word recognition scores, bone conduction thresholds, and pure tone averages (0.5, 1, 2, or 3 kHz), were recorded. When the 3-kHz pure tone threshold was not available, the 2-kHz and 4-kHz average was used as a surrogate.\textsuperscript{11} Postoperative facial nerve function was appraised using the House-Brackmann grading scale.\textsuperscript{15} The distinction of an empty sella turcica was made in cases where the pituitary fossa was largely replaced by CSF, as determined by sagittal T1- or T2-weighted MRI. Specifically, at least two-thirds of the vertical height of the pituitary fossa was occupied by herniating arachnoid of the suprasellar cistern, with resultant compression of the pituitary gland (Fig. 1).\textsuperscript{29}

Data were summarized with means, medians, and ranges, or frequency counts and percentages. Comparisons among categorical data were made using chi-square and Fisher exact tests, whereas group comparisons that involved analyzing continuous data were made using unpaired t-tests; p values of < 0.05 were considered statistically significant.

Surgical Approach

All lesions were accessed through an extradural middle fossa craniotomy or a combined mastoid–middle cranial fossa approach, depending on the size, location, and number of defects. Preoperative imaging was carefully reviewed for any possible superior semicircular canal, ge-
niculate ganglion, or petrous carotid dehiscence (Fig. 2). Bipolar facial electromyography electrodes were placed in the orbicularis oculi and oris for continuous intraoperative neural integrity monitoring. While not routinely performed, intraoperative auditory brainstem response monitoring should be considered in cases of suspected superior semicircular canal dehiscence. Brain relaxation was facilitated using mannitol or an intraoperative lumbar drain, depending on laterality and surgeon preference. Opening pressures may be obtained during lumbar drain placement, with the caveat that normal values do not exclude underlying intracranial hypertension since active high-flow leaks may result in deceptively low pressures until after the diverting CSF is repaired.

If a combined mastoid–middle cranial fossa approach was planned, a lazy-S incision was marked approximately 0.5 cm behind the postauricular sulcus extending antero-superior, to provide adequate exposure for a subtemporal craniotomy. A temporalis fascia graft was harvested, pressed, dried, and set aside for later use. The temporalis muscle was then divided and retracted forward, leaving a small cuff of muscle along the superior temporal line to facilitate muscle approximation during wound closure. It has been our experience that, even in revision cases, adequate autologous graft material can be harvested from the surgical site; loose areolar or true temporalis fascia, conchal, or tragal cartilage, and perichondrium, mastoid cortex, and split-thickness calvarial bone from the craniotomy plate, are excellent materials for reconstruction.

A mastoidectomy was commonly employed to confirm pathology, assist with dissection of encephalocele from either the ossicles or the tympanic segment of the facial nerve, optimize craniotomy placement, and provide the option of additional reinforcement from the mastoid side using bone pâté or fascia. A cortical mastoidectomy was performed, and the tegmen was skeletonized without creating new areas of dural dehiscence. All bony defects were identified, and any devitalized herniating tissue was bipolar-coagulated and amputated. We found that anterosuperior exposure, toward the zygomatic root, was particularly critical for accessing the middle ear space for treatment of tympanic encephalocele.

Following mastoidectomy, a middle fossa craniotomy was planned according to the location and size of the defect(s) identified. In the case of a single large defect, a limited craniotomy centered over the dehiscence was performed (Fig. 3), whereas in the case of multifocal and/or medial defects, a “standard” craniotomy was helpful (Fig. 4). Care was taken to avoid incidental durotomy, and the inferior bone cut was flush with the floor of the middle fossa to improve visualization and minimize temporal lobe retraction. Under microscope magnification, the dura was elevated from the middle cranial fossa floor in a posterolateral to anteromedial direction until the full extent of the defect(s) was visualized. We rarely found the need to divide the middle meningeal artery since most defects are located posterior and lateral to foramen spinosum. Dural defects that could be primarily repaired were then closed using an interrupted or running 6-0 monofilament suture. However, often the dural defects were numerous and the dura quality very poor such that primary suture closure was not possible. In such cases, fascia, followed by cartilage or bone, was generously draped over any bony defects, and finally a layer of fascia or a

![Fig. 2. A–C: Coronal temporal bone CT scans demonstrating a left-sided superior semicircular canal dehiscence (white arrows) in a patient with a tympanic cavity encephalocele. D: Coronal CT scan demonstrating coincident left-sided encephalocele and labyrinthine segment facial nerve dehiscence (black arrow). Note the thin bony covering of the contralateral facial nerve (white arrow) for comparison.](image)
collagen-based dural substitute was interposed between the repair and the dura. Sequential placement of multiple unfurled graft layers, particularly over medial defects, can be tedious even when using a middle fossa retractor. In many instances, we found that creating a “sandwich” of fascia, cartilage, or bone, and dural substitute ex situ, and placing the entire repair at once as a single unit was quite helpful. The medial leading edge of the assembly was sutured in several places before placement to ensure that the fascia did not fold over on itself when the repair was advanced medially. Finally, the bone plate was secured, and the wound was closed in anatomical layers.

Results

Baseline Population Data

Between 2000 and 2012, 86 patients (89 ears) underwent an extradural middle fossa craniotomy or combined mastoid–middle fossa encephalocele and/or CSFF repair. Fifty-one patients (59.3%) were women. The mean age at time of surgery was 52.3 years (median 55.0; range 9.3–75.9), and the left side was affected in 48 (53.9%) of 89 cases (Table 1). Sixty-eight (76.4%) of 89 cases involved concomitant encephalocele and CSFF, 17 (19.1%) involved encephalocele without CSF leak, and 4 (4.5%) involved CSFF without associated meningeal herniation (Table 2). Thirteen (15%) of 86 patients had bilateral disease; 3 of these patients underwent sequential bilateral repair during the study period. Ten patients were evaluated for recurrence after prior unsuccessful repair. Of these, 8 had initially undergone mastoidectomy alone, while 2 had originally been managed with a middle cranial fossa approach performed elsewhere.

The mean delay between symptom onset and diagnosis was 35.4 months (median 12; range 0.5–240). The most common presenting symptoms were hearing loss (82 [92.1%] of 89 cases) and persistent ipsilateral otitis media (65 cases [73.0%]), while few reported ipsilateral rhinitis (8 cases [9.0%]), seizures (2 cases [2.2%]), or a history of intracranial infection, such as meningitis (5 cases [5.6%]) or temporal lobe abscess (1 case [1.1%]) (Fig. 5). Four lesions (4.5%) were discovered incidentally following imaging for seemingly unrelated complaints (Table 1). Of the 71 ears with preoperative audiometry available for review, 52 (73.2%) had more than a 10-dB air-bone gap.

Associations Between Etiology, Body Mass Index, and Empty Sella Syndrome

Thirteen (14.6%) of 89 cases had a history of major trauma, 23 (25.8%) were associated with chronic ear disease without prior operation, 17 (19.1%) occurred following uncomplicated tympanomastoidectomy, and 1 CSFF (1.1%) developed in a patient with a cerebral aqueduct cyst resulting in obstructive hydrocephalus. The remaining 35 cases (39.3%) occurred without antecedent trauma, chronic ear disease, or surgery, and were considered spontaneous. There were no statistically significant associations between presenting symptoms and underlying cause of disease. Among all patients, the mean BMI was 35.3 kg/m² (median 34.8; range 19.0–67.3). Categorically 6.0% of patients were normal weight; 19.3%, overweight; 44.6%, obese (Class I or II); and 30.1%, extremely obese (Class III).

Of the 56 patients with preoperative MRI available for review, 26 (46.4%) exhibited empty sella syndrome. Elevated BMI (p = 0.004) and female sex (p = 0.049) were statistically associated with the finding of an empty sella. Patients with spontaneous lesions were statistically significantly older (mean age 57.4 vs 49.0 years; p = 0.007) and
Notable Intraoperative Findings and Surgical Strategy

...were more commonly female (71.4% vs 48.1%; \( p = 0.048 \)) compared with those with nonspontaneous pathology. Additionally, those with spontaneous lesions had a greater mean BMI compared with those with nonspontaneous disease (37.0 versus 34.1 kg/m\(^2\); \( p = 0.102 \)), although this difference did not achieve statistical significance (Table 2).

**Notable Intraoperative Findings and Surgical Strategy**

Nearly half (35 of 72; 48.6%) of cases had defects that were intermediate in size (1–2 cm) or large (> 2 cm), and 46 (51.7%) had 2 or more areas of dehiscence. Eight temporal bones (9.0%) had concurrent superior semicircular canal dehiscence, and 10 (11.2%) had significant dehiscence of the geniculate ganglion on the middle fossa floor. Bony defect(s) involved both the mastoid and middle ear in 49 ears (55.1%), while isolated tegmen tympani involvement occurred in 35 ears (39.3%) and mastoidectomy dehiscence in 5 ears (5.6%) (Table 3).

Thirty-two (36.0%) of 89 temporal bones underwent a middle fossa craniotomy alone, while 57 (64.0%) underwent a combined mastoid–middle fossa approach. A multilayer repair was used in all but 1 case. Temporalis fascia or a fascia lata graft was used in 88 ears (98.9%), tissue adhesive in 83 (93.3%), calvarial bone in 64 (71.9%), collagen-based dural graft substitute in 42 (47.2%), bone pâte in 24 (27.0%), titanium mesh in 11 (12.4%), conchal or tragal cartilage in 6 (6.7%), and muscle in 5 (5.6%). In 7 ears, an autologous bone graft was rigidly fixed over the bony defect using titanium miniplates and screws to prevent migration (Fig. 6). An intraoperative lumbar drain was used in 32 surgeries (36.0%) to facilitate temporal lobe relaxation.

**Short- and Long-Term Clinical Outcomes**

The median postoperative hospital stay was 2 days (mean 3.0 days; range 1–14 days). Following surgery, 1 patient developed simple partial seizures manifesting with intermittent episodes of speech arrest. This 73-year-old female was successfully managed with levetiracetam and continued to be free of seizure activity as of the most recent follow-up. The 2 patients who presented with preoperative seizures did not experience any further episodes following surgery. One patient with geniculate ganglion dehiscence experienced a mild transient postoperative facial nerve paralysis that normalized within the month following surgery. Of the 45 ears with both pre- and postoperative audiometry available for review, the pure tone average was preserved within 10 dB of preoperative thresholds in 26 (57.8%), improved in 11 (24.4%), and worse in 8 (17.8%). None of the patients experienced profound sensorineural hearing loss following surgery. One patient endured a significant perioperative myocardial infarction and died of cardiopulmonary complications. None of the patients experienced postoperative aphasia, memory impairment, or stroke. Additionally, none of the patients developed postoperative tension pneumocephalus, which is important since over 70% of patients were obese, with a high proportion having sleep apnea using nocturnal CPAP therapy. It has been our preference to withhold postoperative CPAP use for at least 3 weeks unless absolutely necessary, understanding that CPAP generates supraphysiological middle ear pressures that theoretically place early strain on the repair and increase the risk of tension pneumocephalus (Thom JJ, Carlson ML, Driscoll CL, et al, paper submitted to the American Academy of Otolaryngology-Head and Neck Surgery Annual Meeting & OTO EXPO, 2013).

At a mean follow-up of 14.5 months (median 6.7 months; range 0–89.8 months), 7 ears (7.9%) experienced recurrent CSF leak; 2 patients with recurrence developed meningitis. The use of artificial mesh to reconstruct the middle fossa floor was statistically associated with the development of recurrent CSF leak (\( p = 0.004 \)), postoperative wound infection (\( p = 0.039 \)), and meningitis (\( p = 0.014 \)). Also notable, 6 of the 7 cases with recurrence had evidence of intracranial hypertension, determined by...
elevated opening pressures of ≥ 30 cm of water (n = 3) and/or empty sella syndrome (n = 4). The 3 patients with elevated opening pressures were successfully managed with ventriculoperitoneal shunt placement. One patient underwent successful revision surgery using a combined mastoid–middle cranial fossa repair with postoperative lumbar drain, and 3 cases spontaneously resolved after a course of observation ranging from 5 days to 18 months. When the 11 cases that involved using titanium mesh were excluded, 96.2% of patients who underwent an autologous multilayer technique had durable results without recurrent leak or meningitis at the last follow-up.

**Discussion**

Over the last 12 years, 89 temporal bone encephaloceles and/or CSFF in 86 patients were repaired using a middle fossa craniotomy or a combined mastoid–middle fossa approach. We found that a multilayer technique using autologous tissue was a safe and reliable method of repair.

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**TABLE 2: Comparison of variables associated with spontaneous and nonspontaneous temporal bone encephalocele and/or CSFF**

<table>
<thead>
<tr>
<th>Variable</th>
<th>Spontaneous Lesions (n = 35)</th>
<th>Nonspontaneous Lesions (n = 54)</th>
<th>p Value</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>basic</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>age (yrs)</td>
<td>57.4 (56.6; 33.7–74.1)</td>
<td>49.0 (51.2; 9.3–75.9)</td>
<td>0.007</td>
</tr>
<tr>
<td>female sex</td>
<td>25 (71.4)</td>
<td>26 (48.1)</td>
<td>0.048</td>
</tr>
<tr>
<td>BMI (kg/m²)</td>
<td>37.0 (37; 19–67.3)</td>
<td>34.1 (33; 21–50)</td>
<td>0.102</td>
</tr>
<tr>
<td>time to diagnosis (mos)</td>
<td>27.9 (12; 1–144)</td>
<td>40.5 (12; 0.5–240)</td>
<td>0.261</td>
</tr>
<tr>
<td><strong>diagnosis</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>isolated encephalocele</td>
<td>7 (20.0)</td>
<td>10 (18.5)</td>
<td>1.000</td>
</tr>
<tr>
<td>isolated CSFF</td>
<td>1 (2.9)</td>
<td>3 (5.6)</td>
<td>1.000</td>
</tr>
<tr>
<td>concurrent encephalocele &amp; CSFF</td>
<td>27 (77.1)</td>
<td>41 (75.9)</td>
<td>1.000</td>
</tr>
<tr>
<td><strong>laterality of lesion</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>left</td>
<td>17 (48.6)</td>
<td>31 (57.4)</td>
<td>0.515</td>
</tr>
<tr>
<td>bilateral</td>
<td>4 (11.4)</td>
<td>9 (16.7)</td>
<td>0.555</td>
</tr>
<tr>
<td>defect size &gt;1 cm</td>
<td>13 (43.3) of 30</td>
<td>22 (52.4) of 42</td>
<td>0.482</td>
</tr>
<tr>
<td>2 or more defects</td>
<td>17 (48.6)</td>
<td>26 (48.1)</td>
<td>1.000</td>
</tr>
<tr>
<td><strong>coincident finding</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>empty sella syndrome</td>
<td>15 (53.6) of 28</td>
<td>11 (39.3) of 28</td>
<td>0.422</td>
</tr>
<tr>
<td>significant dehiscence of geniculate ganglion</td>
<td>3 (8.6)</td>
<td>7 (13.0)</td>
<td>0.734</td>
</tr>
<tr>
<td>superior semicircular canal dehiscence</td>
<td>4 (11.4)</td>
<td>4 (7.4)</td>
<td>0.707</td>
</tr>
<tr>
<td>preceding intracranial infection</td>
<td>5 (14.3)</td>
<td>1 (1.9)</td>
<td>0.033</td>
</tr>
<tr>
<td>recurrence</td>
<td>4 (11.4)</td>
<td>3 (5.6)</td>
<td>0.427</td>
</tr>
</tbody>
</table>

* Values are expressed as the number of cases (%) or as the mean (median; range). Percentages were calculated from 35 cases for spontaneous lesions and 54 cases for nonspontaneous lesions, unless otherwise specified. Boldfaced values indicate statistical significance.
TABLE 3: Notable anatomical data for all 89 cases (86 patients), determined from radiological imaging and intraoperative findings

<table>
<thead>
<tr>
<th>Defect size†</th>
<th>No. of Cases (%)*</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt;1 cm</td>
<td>37 (51.4)</td>
</tr>
<tr>
<td>1–2 cm</td>
<td>19 (26.4)</td>
</tr>
<tr>
<td>&gt;2 cm</td>
<td>16 (22.2)</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Defect location(s)</th>
<th>No. of Cases (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Combined tegmen tympani &amp; mastoid process</td>
<td>49 (55.1)</td>
</tr>
<tr>
<td>Tegmen tympani alone</td>
<td>35 (39.3)</td>
</tr>
<tr>
<td>Tegmen mastoid process alone</td>
<td>5 (5.6)</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Ipsilateral defects</th>
<th>No. of Cases (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>43 (48.3)</td>
</tr>
<tr>
<td>2</td>
<td>14 (15.7)</td>
</tr>
<tr>
<td>Multiple (≥3)</td>
<td>32 (36.0)</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Coincident finding</th>
<th>No. of Cases (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Empty sella syndrome</td>
<td>26 (46.4) of 56</td>
</tr>
<tr>
<td>Bilateral lesions</td>
<td>13 (14.6)</td>
</tr>
<tr>
<td>Significant dehiscence of geniculate ganglion</td>
<td>10 (11.2)</td>
</tr>
<tr>
<td>Superior semicircular canal dehiscence</td>
<td>8 (9.0)</td>
</tr>
</tbody>
</table>

* Percentages were calculated from 89 cases, unless otherwise specified.
† Defect size was calculated using a denominator of 72 cases.

Notably, a large number of patients in the current series presented with spontaneous lesions, and the majority of patients were obese, regardless of underlying etiology. While only 11 patients underwent reconstruction of the middle fossa floor using an artificial titanium mesh, we found a statistically significant increased risk of postoperative CSFF, wound infection, and meningitis within this group. The current series corroborates earlier studies demonstrating that the majority of patients with temporal bone encephaloceles and CSFF present with persistent otorrhea and hearing loss, mimicking common otitis media. Consequently, findings are often nonspecific since incidental tegmen defects are a ubiquitous finding in the general population and the radiological appearance of encephalocele and CSF effusion replicate that of chronic otitis media, contributing to the potential for diagnostic delay. Since there is a perceived risk of intracranial infection with encephalocele and CSFF, it is generally agreed that persistent lesions mandate surgical repair. In the current study, the mean delay between symptom onset and diagnosis was nearly 3 years, and over 10% of patients had symptoms for over a decade before being referred for treatment.

A history of persistent clear unilateral middle ear effusion or posttympanostomy otorrhea following major head trauma, prior mastoidectomy, or chronic ear disease should raise suspicion for CSFF. An absence of rhinorrhea with provocative maneuvers, such as valsalva with chin tuck, in a patient with a clear middle ear effusion does not rule out disease, as evidenced by the fact that only 9% of patients in the current series had a history of ipsilateral clear nasal drainage. On otomicroscopy, middle ear encephalocele may present as a superiorly based retrosigmoidic pink or blue mass, and posttympanostomy otorrhea is frequently pulsatile. If the diagnosis remains uncertain, then every attempt should be made to collect a fluid sample for beta-2 transferrin analysis. Additionally, we recommend obtaining MR images in all patients with suspected encephaloceles and CSFFs, as it provides invaluable information for differentiating encephalocele from other pathologies, including cholesteatoma, and may assist in evaluating for indirect signs of elevated intracranial pressure and ruling out other causative intracranial processes. Temporal bone encephaloceles can be well visualized on coronal and sagittal T2-weighted sequences where herniating meninges demonstrate low signal intensity in contrast to surrounding high-intensity CSF. Additionally, T1 postcontrast imaging may show meningeal enhancement in the region of dural herniation.

In the preantibiotic era, temporal bone encephaloceles and CSFs were most commonly associated with chronic otitis media, either resulting from long-standing disease or from mastoid exenteration procedures that involved unmagnified mallet and gouge techniques. Within the last half-century, we have seen a growing proportion of traumatic lesions, likely coinciding with the increasing availability of motor vehicles, and high-impact blunt head injury. A comparison of our current series to earlier publications demonstrates a significant increase in the number of spontaneous lesions managed within our practices. The reasons for this are not entirely clear, but are likely related to a growing awareness of disease, improved diagnostic capabilities, and the rising incidence of obesity within the US.

Several studies have demonstrated a strong link between obesity, empty sella syndrome, and elevated intracranial pressure. We found that the mean BMI and the prevalence of empty sella syndrome were higher in patients with spontaneous lesions than in those with...
Temporal bone encephalocele and CSF fistula repair

nonspontaneous lesions; however, these differences did not reach statistical significance. Our data reveal a female preponderance among patients with spontaneous CSFFs, similar to that observed among patients with idiopathic intracranial hypertension syndrome. Congenital defects of the middle fossa floor, and iatrogenic tegmen dehiscence following mastoidectomy, occur at a much higher prevalence than do temporal bone encephalocele and CSFF. Therefore, it is very likely that additional factors contribute to the development of meningeal herniation and subarachnoid fistulization.³⁰ The prevalence of obesity among the 86 study patients was more than twice that of the general US population and nearly 3 times that of our residing state estimates.⁴,⁵ The fact that a large proportion of patients in all groups was obese and had radiographically empty sellae suggests that elevated intracranial pressure may contribute to disease progression, regardless of disease classification.

Temporal bone encephaloceles and CSFFs may be managed through a transmastoid approach with tegmen reconstruction or middle ear obliteration, a subtemporal middle fossa craniotomy, or combined mastoid–middle fossa surgery.⁴,⁵,¹²,¹³,¹⁹ Proponents of the transmastoid approach cite minimized morbidity, the option of concurrent ossiculoplasty, access to tegmen and posterior fossa plate without brain retraction, and earlier hospital discharge. However, a contracted mastoid, anterior sigmoid, low-lying tegmen, or large encephalocele may limit access to the mastoid and tympanic space. Additionally, managing medial, large, multifocal, or epitympanic defects with an intact ossicular chain may be very challenging. We consider middle ear obliteration with blind sac closure of the external auditory canal a last resort in patients with multiply recurrent disease or unreconstructable defects. While highly effective, this approach results in maximum conductive hearing loss and does not permit in-office disease surveillance in patients with a history of cholesteatoma.

While we have not commonly employed local or regional vascularized flaps in lateral skull base encephalocele repair, several reports have demonstrated promising results.⁴,⁶,¹⁹ The temporalis muscle rotation flap and the temporoparietal fascia flap provide local in-field options for repairing defects involving the mastoid and middle cranial fossa floor. Since these flaps receive their blood supply from the deep and superficial temporal arteries, respectively, it is critical to carefully plan temporal incisions to avoid loss of the anteriorly based vascular pedicles. Regional flaps and vascularized free-tissue transfer afford tremendous versatility and can be used to reconstruct large compos-ite defects but carry several notable drawbacks, including increased donor site morbidity, added surgical time, excessive bulk, and the potential need for extensive undermining.⁴,⁶,¹²,¹⁹ For these reasons, such options are generally reserved for patients with poor healing capacity, complex or large defects following tumor resection, and in cases where external beam radiation is anticipated.

Over the last decade, it has been our preference to perform an extradural middle fossa craniotomy or combined mastoid–middle fossa repair. These approaches offer broad exposure of the defect(s) and permit an extradural overlay repair using bone and fascia. Furthermore, intracranial bleeding can be directly controlled, and the epitympanum can be accessed while preserving an intact ossicular chain. Analogous to repairing a leaking roof “from above,” we feel that utilizing a middle fossa craniotomy and a multilayer bone and fascia overlay provides the best chance of long-term repair while carrying minimal morbidity. This approach may be particularly suited for obese patients with spontaneous encephaloceles and CSFFs whose risk of recurrence may be increased.

Overall, 7 cases (7.9%) recurred following surgery; 2 patients with recurrence developed meningitis. Notably, titanium mesh was used in 11 repairs, accounting for 4 recurrences, 2 of 3 wound infections, and both cases of meningitis. Of the 78 ears that were reconstructed using an autologous multilayer technique without titanium mesh, only 3 (3.8%) had recurrence. None of the patients whose ears were reconstructed using this method developed postoperative meningitis. Given these findings, we now avoid the use of artificial graft material, even something as inert as titanium mesh, in defect reconstruction, and instead use at least 1 layer of autologous bone or cartilage and 1 or more layers of fascia.

We wish to acknowledge several limitations of this study. Inherent to a retrospective design, we were dependent on the availability and accuracy of data within the medical record. Given our preference to perform a middle fossa craniotomy or combined mastoid–middle cranial fossa approach, we did not include a comparison population of mastoid-only repairs and therefore cannot prove superiority of one approach over another. However, it is notable that of the 10 patients that initially presented with recurrence, 8 had previously received a mastoidectomy alone for repair. Of these 10, only 1 patient recurred following revision surgery at our institution; this single patient had been initially repaired using titanium mesh. Additionally, there was a large array of substrates used for defect repair in varying combinations, making subgroup comparisons difficult. Based on the available data, at minimum, a combination of autologous fascia and bone should be used, and synthetic titanium mesh should be avoided. Finally, it remains possible that the high prevalence of obesity observed within our study population was exaggerated by referral pattern biases associated with large tertiary care centers. While we cannot completely discount this possibility, anecdotally, the frequency of obesity among other patients referred for neurosurgical or neurotological care is substantially lower than what we have observed within the study population. Moreover, the fact that the prevalence of obesity among the 86 subjects was nearly 3 times that of our residing states further supports a strong association between obesity and the development of temporal bone encephalocele and CSFF.⁴

Conclusions

Patients with temporal bone encephalocele and CSFF commonly present with persistent otorrhea and conductive hearing loss mimicking chronic middle ear disease. Among this patient population, there is a high prevalence of obesity, which may play a role in the pathogenesis of primary and recurrent disease. A middle fossa craniotomy or a combined mastoid–middle cranial fossa approach incorporating a multilayer autologous tissue technique provides a safe and reliable method of repair that may be
particularly useful for large or multifocal defects. Artificial titanium mesh should generally be avoided given an increased risk of recurrence, postoperative wound infection, and meningitis.

 Disclosure

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