Objective: To describe the clinical presentation and the radiological and surgical findings in patients with spontaneous meningoencephalocele of the temporal bone.

Design: Retrospective case series.

Setting: Academic, tertiary care medical center.

Patients: Fifteen consecutive patients with surgically confirmed meningoencephalocele of the mastoid and middle ear, without a history of trauma, tumor, cholesteatoma, or surgery of the mastoid or cranium, who were treated at our institution between January 1, 1999, and December 31, 2006.

Results: Ten of the 15 patients were women. Ages ranged from 31 to 77 years, with 12 patients 50 years or older. The most common presenting complaint was new-onset hearing loss in 14 patients, followed by aural fullness and headache. Cerebrospinal fluid formed an effusion in the middle ears of 13 patients and was most commonly identified when myringotomy resulted in continuous clear otorrhea. Four subjects had a history of adult-onset recurrent acute otitis media with intermittent otorrhea, which in 1 case was complicated by brain abscess. At least 1 full-thickness defect of the tegmen associated with cortical thinning of the middle fossa floor was identified in all cases on high-resolution computed tomography. At surgery, herniations of meningeal and cerebral tissue were seen through 1 (7 cases) or 2 (8 cases) defects in the middle fossa floor. Obstruction of antral aeration by the meningoencephalocele was present in all 4 cases associated with otitis media.

Conclusions: The onset of otitis media, including middle ear effusions at 40 years or older, warrants the consideration of a meningoencephalocele of the ear. The appearance of tegmental defects and cortical thinning of the middle fossa floor on computed tomography provides a strong indication of the diagnosis and of the need for surgical repair.


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within submucosal collections or flow freely within air-filled spaces, eventually exiting the temporal bone via an opening in the tympanic membrane or via the eustachian tube. Although the term spontaneous CSF otorrhea has been adopted to describe this phenomenon, a more accurate description of this presentation would be spontaneous CSF effusion (SCSFE).

The timely diagnosis of SCSFE requires awareness of this entity and a high degree of clinical suspicion. With the aim of increasing the understanding and awareness of this condition, we present our experience with 15 patients. We highlight the clinical presentation and radiological and surgical findings in this retrospective cohort and draw particular attention to the spectrum of clinical presentation of spontaneous meningoencephalocele of the ear, which includes SCSFE, dormant middle ear mass, and otomastoiditis because of attic block due to herniated meninges.

### METHODS

We performed a retrospective review of patients undergoing evaluation and treatment at The Johns Hopkins Hospital from January 1, 1999, through December 31, 2005. Inclusion criteria were a surgically confirmed tegmental defect with herniation of membranes or brain into temporal bone space, without a history of trauma, tumor, cholesteatoma, or surgery of the mastoid or cranium. After institutional review board approval, data including presenting symptoms and signs, otologic history, results of diagnostic studies, and surgical findings were extracted from patient medical records, and the radiological images were reviewed.

### RESULTS

Fifteen patients fulfilled inclusion criteria (Table 1). The average age at presentation was 58.5 (SD, 14.2) years, with a range of 31 to 77 years. Twelve individuals were 50 years or older, whereas the remaining 3 subjects were aged 31, 38, and 41 years. Most of the patients were female (ratio of women to men, 10:5). There was a slight predilection for right-sided involvement (ratio of right to left sides, 9:6), and a history of bilateral meningoencephalocele was present in 5 patients. The most common presenting symptom was persistent hearing loss in 14 patients, followed by aural fullness and headache, which were present in 5 patients each. These symptoms were usually of rapid or sudden onset, presumably corresponding to the establishment of a CSF fistula producing symptoms due to accumulation in the ear (hearing loss and fullness) and loss from the subarachnoid space (headache). No patients reported rhinorrhea or postnasal drainage. All but 2 individuals were assumed to have serous or chronic otitis media and had been misdiagnosed as having the condition for an average of 1.6 years, with a delay of as long as 10 years before receiving an accurate diagnosis (Table 1). Four patients had a documented history of recurrent or chronic otitis media beginning in adulthood, based on the presence of otalgia, intermittent purulent otorrhea, or otologic findings consistent with inflammatory changes of the tympanic membrane. In 1 patient, the diagnosis was made after presenting with signs and symptoms of meningitis and a temporal lobe abscess.

Whereas conductive hearing loss was easily confirmed with tuning fork testing and audiometry, the otoscopic presentation of CSF effusion was very subtle. The presence of fluid in the middle ear was initially missed in many cases. The examination results were usually thought to be normal because the effusion was colorless and the tympanic membrane was not altered by erythema, retraction, or other findings (Figure 1). The lack of color also made it difficult to discern bubbles or an air-fluid level, particularly with a monococular otoscope. However, careful observation of the tympanic membrane with binocular microscopy revealed an accentu-

### Table 1. Clinical Presentation of Patients With Spontaneous Meningoencephalocele of the Temporal Bone

<table>
<thead>
<tr>
<th>Patient No./Sex/Age, y</th>
<th>Side</th>
<th>Hearing Loss</th>
<th>Aural Fullness</th>
<th>Headache</th>
<th>Otoscopic Findings</th>
<th>Previous Diagnosis</th>
<th>Time to Diagnosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 (M/72)</td>
<td>Left</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>OWM</td>
<td>No</td>
<td>COM</td>
</tr>
<tr>
<td>2 (F/41)</td>
<td>Right</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>OWM</td>
<td>No</td>
<td>SOM</td>
</tr>
<tr>
<td>3 (M/55)</td>
<td>Right</td>
<td>Yes</td>
<td>Yes</td>
<td>No</td>
<td>Translucent, bulging TM</td>
<td>No</td>
<td>SOM</td>
</tr>
<tr>
<td>4 (F/55)</td>
<td>Right</td>
<td>Yes</td>
<td>Yes</td>
<td>No</td>
<td>Translucent, bulging TM</td>
<td>No</td>
<td>SOM</td>
</tr>
<tr>
<td>5 (F/77)</td>
<td>Right</td>
<td>Yes</td>
<td>No</td>
<td>Yes</td>
<td>OWM</td>
<td>No</td>
<td>COM</td>
</tr>
<tr>
<td>6 (F/38)</td>
<td>Right</td>
<td>Yes</td>
<td>Yes</td>
<td>No</td>
<td>OWM</td>
<td>No</td>
<td>SOM</td>
</tr>
<tr>
<td>7 (M/77)</td>
<td>Left</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
<td>OWM, inflamed TM</td>
<td>No</td>
<td>COM</td>
</tr>
<tr>
<td>8 (F/31)</td>
<td>Right</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Bulging, hyperemic TM</td>
<td>No</td>
<td>SOM</td>
</tr>
<tr>
<td>9 (F/57)</td>
<td>Left</td>
<td>Yes</td>
<td>No</td>
<td>Yes</td>
<td>OWM</td>
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<td>Unknown</td>
</tr>
<tr>
<td>10 (F/51)</td>
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<td>No</td>
<td>Yes</td>
<td>OWM</td>
<td>No</td>
<td>SOM</td>
</tr>
<tr>
<td>11 (F/64)</td>
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<td>Yes</td>
<td>No</td>
<td>OWM</td>
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<td>SOM</td>
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<tr>
<td>12 (F/66)</td>
<td>Left</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>OWM, mucopurulence, hyperemic TM</td>
<td>No</td>
<td>COM</td>
</tr>
<tr>
<td>13 (M/54)</td>
<td>Right</td>
<td>Yes</td>
<td>Yes</td>
<td>No</td>
<td>Middle ear mass</td>
<td>No</td>
<td>Meningoencephalocele</td>
</tr>
<tr>
<td>14 (F/70)</td>
<td>Left</td>
<td>Yes</td>
<td>No</td>
<td>Yes</td>
<td>Normal TM after resolved AOM</td>
<td>Yes</td>
<td>COM</td>
</tr>
<tr>
<td>15 (M/70)</td>
<td>Right</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>OWM</td>
<td>No</td>
<td>SOM</td>
</tr>
</tbody>
</table>

Abbreviations: AOM, acute otitis media; COM, chronic otitis media; NA, not applicable; OWM, otorrhea with myringotomy; SOM, serous otitis media; TM, tympanic membrane.
ated convexity of an otherwise normal tympanic membrane, particularly of the pars flaccida, in 2 patients without myringotomies. The diagnosis of a middle ear effusion was usually made after empirical myringotomy was performed, or else it was suspected on observing fluid density opacifying the middle ear and mastoid on computed tomographic (CT) images. Persistent clear otorrhea after myringotomy was observed in 10 patients, making it the most common physical finding leading to the diagnosis of SCSFE. Three patients had evidence of otitis media on presentation, indicated by thickened and hyperemic tympanic membranes. One patient presented after repair of a meningoencephalocele in the contralateral ear with a pale pink mass in the posterosuperior quadrant of the middle ear and a history of intermittent aural fullness (described in the “Case 3” subsection of the “Report of Cases” section). For 10 patients with available audiometric results, the mean preoperative air-bone gap was 25.4 (range, 8.3-50.0; SD, 12.1) dB.

Thin-cut coronal CT of the temporal bone demonstrated some consistent findings in all cases of spontaneous meningoencephalocele. In all 15 cases, at least 1 defect of the tegmental cortex created direct communication between the middle ear and antrum or attic, including the subsegmental layer of air cells (Figure 2 and Table 2). Cortical thinning of the surrounding middle fossa floor (Figures 2, 3, and 4) was also a prominent finding, leaving the upper layer of mastoid air cells open to the intracranial compartment. Opacification of the mastoid and middle ear by tissue or fluid density was present in all cases. Underlying mastoid development was normal in all but 1 case. Cortical loss involving the middle fossa floor was present on the contralateral side in some cases without tegmental defect. Bilateral meningoencephaloceles were identified in 3 patients. Varying degrees of otic capsule thinning overlying the superior semicircular canal was also observed, but no patients presented with the Tullio phenomenon or stigmata of the superior semicircular canal dehiscence syndrome. The bony architecture of the labyrinth was otherwise normal.

The presence of tegmental defects on CT, particularly in the context of clear otorrhea, prompted the diagnosis of spontaneous CSF leak and a recommendation for surgical repair. Further testing, including evaluation for β2-transferrin levels and magnetic resonance imaging (MRI), was not performed in most patients. Patients were counseled of the possible need for middle fossa craniotomy to repair bony and dural defects, for which informed consent was acquired before surgery. Surgical planning also included scheduling of a monitored bed for the night of surgery, where frequent neurological assessments would be conducted. All patients first underwent mastoidectomy to confirm the presence and location of herniated meningeal tissue and an associated CSF leak.

A minicraniotomy of the middle fossa was then placed for optimal access to the tegmental defect and meningoencephalocele located through the mastoid cavity. The inferior edge of a 3 × 4-cm craniotomy was placed within 5 to 10 mm of the tegmen to optimize visualization of

![Figure 1. Otoscopic appearance of the right ear with spontaneous cerebrospinal fluid effusion. A, View demonstrating the normal appearance of the par tensa. There is evidence of a clear-colored effusion in the middle ear based on the presence of an air-fluid level in the protympanum and a bubble in the posterior inferior mesotympanum. B, A more superior view of the same ear showing the pars flaccida. Retraction is absent and a possible slight convexity is supported by the diminished prominence of the lateral process of the malleus. This finding is in contradistinction to the retraction and prominent lateral process evident in patients with middle ear effusions resulting from eustachian tube dysfunction. Inset, ABL indicates air bubble; AFL, air-fluid level; LP, lateral process of the malleus; MBM, manubrium; and PF, pars flaccida.](http://archotol.jamanetwork.com/pdfaccess.ashx?url=/data/journals/otol/9810/)
the lateral middle fossa floor with minimal to no temporal lobe retraction. Patients were administered a second- or third-generation cephalosporin and intravenous corticosteroids at the beginning of the procedure and 20 to 40 g of intravenous mannitol approximately 1 hour before the middle fossa craniotomy to reduce the need for temporal lobe retraction. The patient underwent hyperventilation to maintain a PCO₂ of 25 to 30 mm Hg. In 2 cases, attempts to repair tegmental defects via the mastoid without middle fossa craniotomy failed.

The middle fossa floor had a “moth-eaten” appearance consisting of deficient cortical lining and unroofed mastoid air cells. All patients were found to have meningeal and cerebral tissue herniating through 1 (7 cases) or 2 (8 cases) defects in the middle fossa floor into the attic or antrum (Table 3). Of 23 such herniations, the tegmental defect was located primarily above the attic (tegmen tympani) in 8 cases, above the antrum (tegmen mastoideum) in 12 cases, and spanning both areas in 2 cases. These defects ranged in diameter from 5 to 40 (mean, 13.8; SD, 10.9) mm. For those cases in which herniation into the attic was observed, the ossicular chain remained intact but was often encased in adhesions. The herniated dural tissue was elevated from the ossicular chain, other surfaces in the ear, and the rim of the tegmental defect. Any incarcerated cerebral tissue within a defect was cauterized with bipolar instrumentation at its stalk and amputated. In the case of a dural defect, fine sutures were used to form a watertight closure using primary closure and/or temporalis fascia graft. The tegmental defect was closed using split calvarial bone harvested from the edge of the mastoidectomy and shaped to form a stable covering. For 8 patients with available audiometric results, the mean postoperative air-bone gap in the affected ear was 16.5 (range, 1.7-41.7; SD, 12.9) dB. The mean time to postoperative audiometry was 7.5 (range, 1-30; SD, 8.8) months.

In all 4 patients with otomastoiditis, meningoencephaloceles were located in the antrum and middle ear, where dense adhesions to the ossicles were observed to block the aditus. In 2 of these, meningoencephalocele and CSF leak were encountered unexpectedly during mastoid surgery for management of chronic otitis media. Although modest amounts of granulation tissue were encountered, there was no cholesteatoma found in these cases. These patients had no history of ear surgery. Another patient was simultaneously diagnosed as having a congenital epidermoid located within the occipital bone just posterior to the mastoid and extending into the posterior fossa but distant from the tegmental defect and meningoencephalocele.

Table 2. Radiological Findings in Patients With Spontaneous Meningoencephalocele of Temporal Bone

<table>
<thead>
<tr>
<th>Patient No.</th>
<th>Opacification of Middle Ear or MAC</th>
<th>Thinned Cortical Lining of MF Floor</th>
<th>Tegmental Dehiscence</th>
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</thead>
<tbody>
<tr>
<td>1</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
</tr>
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<td>5</td>
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<td>7</td>
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<td>10</td>
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<td>No</td>
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<td>11</td>
<td>No</td>
<td>Yes</td>
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<tr>
<td>12</td>
<td>Yes</td>
<td>Unknown</td>
<td>Unknown</td>
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<tr>
<td>13</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
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<tr>
<td>14</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>15</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
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</tbody>
</table>

Abbreviations: MAC, mastoid air cells; MF, middle fossa.
Cephalocele. An additional patient was diagnosed as having Wegener granulomatosis that spared the temporal bone. The following case reports highlight the clinical and radiological nuances of this disease entity.

**REPORT OF CASES**

**CASE 1**

A woman aged 41 years (patient 2) presented with a presumed diagnosis of chronic eustachian tube dysfunction. Two years before presentation at The Johns Hopkins Outpatient Clinic, a pressure equalization (PE) tube was placed in the right ear, after which she experienced

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**Figure 3.** Coronal computed tomography series demonstrating a diffusely thinned and perforated cortical floor of the left middle fossa floor of patient 7 (described in the “Case 2” subsection of the “Report of Cases” section). A large continuous defect is evident in the roof of the attic (D-F), below which soft tissue and fluid density fill the mastoid and middle ear spaces.

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**Figure 4.** Otoscopic and radiological findings in a case of meningoencephalocele of the right ear of patient 3 (described in the “Case 3” subsection of the “Report of Cases” section). A, Otoscopic appearance of the right eardrum showing a pink mass deep to the posterosuperior quadrant of the tympanic membrane and an associated protrusion of the eardrum. B, Coronal computed tomographic image of the right temporal bone demonstrates loss of the cortical lining of the middle fossa floor and multiple defects producing a communication between the attic and the intracranial compartment. Soft tissue density fills the attic but, as suggested by the otoscopic examination findings, the remainder of the middle ear space is aerated, consistent with encephalocele without cerebrospinal fluid effusion. C, T1-weighted magnetic resonance image of the brain with gadolinium contrast shows soft tissue that is isointense with brain tissue herniating into the mastoid region, consistent with meningoencephalocele.
daily drainage of clear fluid. The PE tube was removed and she subsequently began to experience increased auricular pressure and hearing loss. A second myringotomy and PE tube placement relieved these symptoms but clear otorrhea resumed. This fluid was collected and found to be positive for β2-transferrin, leading to the diagnosis of a CSF leak. She denied headaches, fevers, vertigo, otalgia, and tinnitus and she had no history of meningitis, trauma, or prior cranial surgery.

On otoscopic examination, the right side showed an intact tympanic membrane with a metal PE grommet from which clear fluid was draining. The remainder of the canal was grossly normal with no evidence of purulence or infection. Preoperative CT revealed cortical deficiency of the right middle fossa floor compared with the left side and 2 tegmental dehiscences (Figure 2D-F). She underwent mastoidectomy and middle fossa craniotomy and was found to have an 8-mm dehiscence in the tegmen mastoideum and a 1-cm meningoencephalocele within the antrum. She had an uneventful postoperative course. At the 6-month follow-up, otoscopy revealed a dry ear and aerated middle ear space. She reported normal hearing, which was confirmed by an air-bone gap of 3.4 dB.

**CASE 2**

A man aged 77 years (patient 7) with no history of otological disease presented to The Johns Hopkins Hospital with left-sided frontal pain. Physical examination results showed a middle ear effusion on the left side. A PE tube was placed, with egress of clear fluid. He returned in 6 months reporting continued frontal headaches and clear otorrhea. Otoscopy revealed a hyperemic tympanic membrane with an extruded PE tube and clear fluid emanating from the myringotomy defect. High-resolution CT showed opacification of the left mastoid air cells and middle ear. Computed tomography demonstrated opacification of the right mastoid and middle ear and dehiscence of the middle fossa cortex through which a meningoencephalocele protruded into the antrum and middle ear. After repair via a middle fossa craniotomy, the patient had an uneventful postoperative course and was free of otorrhea and frontal headache at his 6-month follow-up.

**CASE 3**

A man aged 55 years (patient 3) underwent repair of a left temporal bone encephalocele with CSF leak in 2003. He returned 2 years later reporting intermittent aural fullness and hearing loss in the right ear. Right-sided otoscopy showed a pale pink mass in the posterosuperior quadrant of the middle ear (Figure 4A). Computed tomography demonstrated opacification of the right mastoid and middle ear and dehiscence of the middle fossa cortex through which soft tissue density was seen to extend from the temporal lobe into the ear on MRIs (Figure 4C). A meningoencephalocele was diagnosed. The patient consented to undergo repair of a tegmental defect via the mastoid and middle fossa approaches. At surgery, the cortical floor of the middle fossa was found to be absent, replaced by multiple moth-eaten defects. An 8 × 4-mm defect contained an encephalocele extending to the attic and posterior mesotympanum. A 2 × 3-mm defect was present at the posterior limit of the tegmen mastoideum, through which passed a small encephalocele located within the sinodural angle. The patient had an uneventful postoperative course and experienced closure of the air-bone gap in his right ear at his 6-month follow-up.

**CASE 4**

A woman aged 66 years (patient 12) presented with a 1-year history of intermittent left purulent otorrhea and otalgia. Exacerbations of otitis media were easily cleared using topical and systemic antibiotics but soon returned after completion of treatment. A PE tube did not resolve the symptoms. She had no other otologic procedures and no history of trauma. On otoscopy, the left ear canal was found to contain a significant amount of mucopurulent material. The eardrum appeared hyperemic and opaque. A patent PE tube was present surrounded by actively pooling purulent drainage. Computed tomography showed severe sclerosis of the left mastoid with opacification of the antrum and attic with soft tissue density. A simple mastoidectomy was planned to manage what was thought to be chronic otomastoiditis. At surgery, a small tegmental dehiscence was found containing a meningoencephalocele that extended to the mesotympanum, embedding the middle ear ossicles and blocking the aditus. A middle fossa craniotomy was subsequently performed with repair of the tegmen. At her 6-month follow-up visit, the patient had no more otorrhea or otalgia.

**CASE 5**

A woman aged 70 years (patient 14) with a 6-month history of recurrent left otitis media presented to an outside hospital with acute onset of fever, headache, and left-sided otalgia and increasing confusion. Sampling of CSF
via lumbar puncture was performed, and the results were found to be consistent with meningitis. A brain MRI demonstrated an enhancing pattern consistent with a multiloculated abscess of the left temporal lobe (Figure 5A). At outpatient consultation, after recovery from her acute illness, results of the otoscopic examination were unremarkable. The CT images demonstrated erosion of the left tegmen tympani and soft tissue density within the adjacent attic and antrum, consistent with an encephalocele (Figure 5B). At mastoidectomy, 2 meningoencephaloceles associated with tegmental defects in the antrum and at the sinodural angle, each measuring 5 mm, were observed. Necrotic brain tissue was removed after bipolar cauterization. A split calvarial bone graft was harvested from adjacent occipital bone and was placed intracranially between elevated dura and the middle fossa floor via the mastoid without performing a middle fossa craniotomy and without encountering a CSF leak. The tegmen was then recontoured using hydroxyapatite paste or carbonated calcium phosphate paste (Norian Corp, Cupertino, California). The patient had an uneventful postoperative course without symptoms and signs of CSF leak or recurrent ear infections at a 4-month follow-up examination. A second CT image was obtained after the recurrence of aural fullness and hearing loss in the left ear. These images showed displacement of the bone graft used to reconstruct the tegmental defect and opacification of the adjacent antrum with soft tissue density.

**THE CLINICAL SPECTRUM OF SPONTANEOUS MENINGOENCEPHALOCELE OF THE EAR**

This study presents some relevant clinical findings that can aid in the timely diagnosis of spontaneous meningoencephaloceles of the temporal bone. This entity is likely to remain asymptomatic for many years until (1) it is appreciated as a middle ear mass on otoscopy; (2) it produces symptoms of poor mastoid aeration because of attic block; (3) it generates an intermittent or continuous leak of CSF; or (4) it becomes a pathway for the intracranial spread of infection.

Spontaneous CSF effusion and otorrhea are the most common and well-recognized presentations of spontaneous meningoencephaloceles. New-onset aural fullness and hearing loss found to be conductive on results of tuning fork testing or audiometry and the presence of middle ear fluid should prompt consideration of a differential diagnosis that includes nasopharyngeal carcinoma, other primary skull base neoplasms, metastatic disease in the temporal bone, or a meningoencephalocele with CSF leak. Otoscopic examination findings in cases of CSF effusion are often underwhelming because the fluid is colorless and because the eardrum is neither retracted nor hyperemic in most cases. Despite the absence of the hallmark straw-colored effusion and tympanic membrane retractions, patients are often given a diagnosis of serous otitis media for many years until continuous clear drainage after myringotomy prompts consideration of CSF leak. In patients with intermittent or low-volume leak, the diagnosis may never be made unless septic intracranial complications occur or unless the diagnosis is considered and imaging is performed. In our study, high-resolution CT of the temporal bone demonstrated characteristic changes to the middle fossa floor, including cortical erosion and defects associated with soft tissue density in the middle ear or mastoid. In all cases of confirmed meningoencephaloceles, these radiological findings were present. The onset of ear symptoms on average in the fifth decade of life, including aural fullness, hearing loss, or otorrhea with myringotomy, are common themes in the published studies of 199 patients with this condition since 1913 (Table 4).

The steady increase in the number of reported cases of SCSFE and otorrhea argues that this entity may be more common than was once believed. A review of the literature shows that the first case of adult SCSFE was published in 1913. In a review of the literature by Ferguson et al, 29 cases were reported by 1986. The mean age of presentation was 48 years, and aural fullness with middle ear effusion or clear otorrhea constituted the most common initial symptoms. In 1995, May et al described a series of 12 patients diagnosed as having SCSFE. The mean age at onset was 52 years. Most of these patients also presented with hearing loss and aural fullness. A provisional diagnosis of SCSFE was made in 11 of 12 patients when myringotomy resulted in profuse clear otorrhea. Within the next few years, Lundy et al and Gacek et al further bolstered these observations in their reports of 11 and 21 patients, respectively. In the past 10 years, more than 150 cases have been reported, all with similar findings (Table 4). Most recently, Leonetti et al described the findings in 51 patients in whom the most common presenting symptoms were aural fullness and hearing loss, whereas the most common physical find-
ings were clear middle ear effusion and clear tympanos-
tomy tube otorrhea. Although the prevalence has likely
remained unchanged, this apparent increase is likely owing
to greater awareness and recognition of the condi-
tion, itself a result of increased reporting in the litera-
ture. The diagnostic value of tegmental dehiscence on high-
resolution CT has been previously discussed, but
the additional feature of generalized cortical loss of the
middle fossa floor, which was recognized in all cases in
this study, should alert the clinician to associated me-
ningoencephalocele.

PROPOSED PATHOLOGICAL MECHANISMS

Arachnoid granulations, responsible for the resorption of
CSF into the venous system and the maintenance of
normal intracranial pressure, are normally located within
the lumen of the venous sinuses. Occasionally during
development they come to lie in direct contact with bone
rather than within the dural venous sinuses. The pres-
sure of CSF pulsating directly against the bone results
in bony erosions, which over time can culminate in ob-
sure of CSF pulsating directly against the bone results
rather than within the dural venous sinuses. The pres-
sure most commonly occurring at 50 years or older. In-
creased intracranial pressure, particularly in obese pa-
tients, may also facilitate the spontaneous development of
meningoencephaloceles by this mechanism. Because the tegmen is usually thinner than the posterior fossa surface of the petrous pyramid, with larger spaces into which intracranial tissue can herniate, it is not surprising that meningoencephaloceles of the temporal bone more commonly arise from the middle fossa.

Although previous studies looking at SCSFE may have
eliminated patients with a history of middle ear inflam-
mation from their series in the belief that a history of in-
flammation caused the bony defect, we believe that the reverse may be true—that the soft tissue herniation may have predisposed the patient to varying degrees of attic block and the development of secondary otomastoid-
itis. Involvement of the attic by meningoencephalocele of the temporal bone is relatively common (9 of 15 cases) and is likely to obstruct normal airflow to the mastoid
in at least some cases. Consequently, we propose that oto-
mastoiditis is part of the spectrum of adult-onset ear dis-
ease that should prompt consideration of spontaneous
meningoencephalocele of the temporal bone. Rather than
small punctate dehiscences of the tegmen observed in
spontaneous cases, tegmental defects resulting from
chronic otitis media and previous surgery are signifi-
cantly larger with well-preserved surrounding middle fossa cortex.

Compared with congenital forms of CSF fistulae, me-
ingitis is less commonly encountered in adults with spon-
taneous SCSFE. When all of the 199 cases reported in
the literature are combined, including those in our se-
ries (Table 4), 28 (14.1%) had an episode of meningitis.

Table 4. Published Reports of SCSFE Cases

<table>
<thead>
<tr>
<th>Source</th>
<th>No. of Patients</th>
<th>Average Age, y</th>
<th>Hearing Loss</th>
<th>Aural Fullness</th>
<th>Otorrhea With Myringotomy</th>
<th>Meningitis</th>
</tr>
</thead>
<tbody>
<tr>
<td>28 Reports with &lt; 5 cases each; 1913-2004</td>
<td>36</td>
<td>56.5</td>
<td>17</td>
<td>6</td>
<td>22</td>
<td>11</td>
</tr>
<tr>
<td>Kemink et al, 1986</td>
<td>6</td>
<td>54.5</td>
<td>3</td>
<td>1</td>
<td>Unknown</td>
<td>0</td>
</tr>
<tr>
<td>Pappas et al, 1992</td>
<td>8</td>
<td>61</td>
<td>6</td>
<td>5</td>
<td>7</td>
<td>2</td>
</tr>
<tr>
<td>May et al, 1995</td>
<td>12</td>
<td>52</td>
<td>2</td>
<td>3</td>
<td>11</td>
<td>1</td>
</tr>
<tr>
<td>Lundy et al, 1996</td>
<td>11</td>
<td>57</td>
<td>Unknown</td>
<td>Unknown</td>
<td>11</td>
<td>0</td>
</tr>
<tr>
<td>Gacek et al, 1999</td>
<td>21</td>
<td>63</td>
<td>Unknown</td>
<td>Unknown</td>
<td>14</td>
<td>1</td>
</tr>
<tr>
<td>Patel et al, 2000</td>
<td>7</td>
<td>55</td>
<td>6</td>
<td>0</td>
<td>6</td>
<td>3</td>
</tr>
<tr>
<td>Savva et al, 2003</td>
<td>8</td>
<td>Unknown</td>
<td>5</td>
<td>Unknown</td>
<td>8</td>
<td>3</td>
</tr>
<tr>
<td>Mayeno et al, 2004</td>
<td>5</td>
<td>59</td>
<td>5</td>
<td>0</td>
<td>3</td>
<td>1</td>
</tr>
<tr>
<td>Brown et al, 2004</td>
<td>9</td>
<td>57.8</td>
<td>2</td>
<td>5</td>
<td>5</td>
<td>0</td>
</tr>
<tr>
<td>Rao et al, 2005</td>
<td>10</td>
<td>58</td>
<td>Unknown</td>
<td>Unknown</td>
<td>6</td>
<td>2</td>
</tr>
<tr>
<td>Leonetti et al, 2005</td>
<td>51</td>
<td>45.7</td>
<td>42</td>
<td>46</td>
<td>18</td>
<td>3</td>
</tr>
<tr>
<td>Present study</td>
<td>15</td>
<td>58.5</td>
<td>14</td>
<td>5</td>
<td>10</td>
<td>1</td>
</tr>
<tr>
<td>All studies</td>
<td>199</td>
<td>56.5</td>
<td>102/157 (65.0)</td>
<td>71/149 (47.7)</td>
<td>132/193 (68.4)</td>
<td>28/199 (14.1)</td>
</tr>
</tbody>
</table>

Abbreviation: SCSFE, spontaneous cerebrospinal fluid effusion.

*For presenting conditions, data are expressed as the proportion of cases for which the presence or absence of the presenting condition was reported.
Taken together, these results indicate that, although less common, meningitis in the adult population is a real possibility, thus prompting the need for immediate recognition and treatment to avoid this potentially life-threatening complication. Antipneumococcal immunization may be considered to lower the risk of meningitis in these patients.

**RADIOLOGICAL FINDINGS**

Whereas clinical suspicion and judgment are critical to the diagnosis of meningoencephalocele and associated SCSFE, radiological and laboratory studies are valuable adjuncts. In our experience, coronal CT images of the temporal bone most accurately predict these findings at surgery. This high degree of accuracy is consistent with other reports in the literature, in which CT is considered the gold standard for diagnosing SCSFE and associated defects of the temporal bone.37 We have observed that the absence of normal cortical tegmental bone is a common finding that predicts the surgical observation of a moth-eaten middle fossa floor within which larger defects are seen to contain meningoencephaloceles. The identification of this cortical deficiency is particularly helpful when tegmental defects are too small or CT sections are too thick to detect them.

Other diagnostic modalities, including MRI, radionuclide cisternography, nuclear medicine studies, and β₂-transferrin studies, provide additional useful information, but alone they are less useful than CT in guiding therapy. Magnetic resonance imaging of the brain should be considered, however, in the setting of large skull base defects and meningoencephaloceles in which white matter may accompany gray matter. In these cases, reduction of the encephalocele when possible is preferable to amputation. In our series, 2 patients underwent MRI and 3 had β₂-transferrin studies. The results, however, did not change the management plan, which relied on CT findings. Stone et al49 have also reported that high-resolution CT identified significantly more patients with SCSFE than did radionuclide cisternography and CT-cisternography. Although nuclear medicine studies, using for example, indium In 111, have the advantage of being able to localize the leak to the ear and confirm that it is indeed CSF, these procedures are invasive, requiring a lumbar puncture.30 A less invasive means of confirming the presence of CSF is the use of a β₂-transferrin study, which has a sensitivity of 84% and specificity of 100%.51 It is our impression that the clinical and radiological presentation are sufficient to diagnose SCSFE, and that β₂-transferrin studies may be reserved for cases that are clinically less straightforward.

**PRINCIPLES OF SURGICAL MANAGEMENT**

The surgical approach for the repair of tegmental defects has been well established in the literature.1,40 We have found that the combined transmastoid–middle fossa approach has had a success rate of 100% in our series. Our surgical experience with these patients has demonstrated the importance of paying particular attention to exploring the tegmen via the mastoidectomy so as to appropriately place a middle fossa craniotomy for optimal access to the tegmental defect. The repair of both dural and bony defects is needed. Without a secure bony reconstruction, dural herniation and/or fistulization recur. Hydroxyapatite paste is a useful adjunct with bone grafting to fill small cortical defects.32 Dural defects can be closed primarily and/or bolstered using free autologous tissue, dural substitutes, or pedicled flaps such as the temporoparietal flap.

Lumbar drains were not used in any patient within this series without apparent compromise of the repair, even in cases of active CSF drainage. Unlike spontaneous anterior cranial leaks, the role of intracranial hypertension in the pathophysiologic mechanism of temporal bone meningoencephaloceles is poorly characterized.53 Morbidity associated with underlying intracranial hypertension should nevertheless be considered and managed in these cases. A recent report by Prichard et al39 of obesity in 7 patients and radiological evidence of empty sella in 5 of 8 patients with spontaneous CSF otorrhea provides preliminary evidence of an increased risk of pseudotumor cerebri in these patients. Diagnostic lumbar puncture and neuro-ophthalmologic examination of the fundi should be considered to rule out this diagnosis. Appropriate management may benefit the long-term integrity of the tegmental repair, may prevent the development of new defects, and will reduce risks to vision.

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**Author Contributions:** Drs Nahas, Tatlipinar, and Francis had full access to all the data in the study and took responsibility for the integrity of the data and the accuracy of the data analysis. **Study concept and design:** Nahas and Francis. **Acquisition of data:** Nahas, Tatlipinar, Limb, and Francis. **Analysis and interpretation of data:** Nahas, Limb, and Francis. **Drafting of the manuscript:** Nahas, Tatlipinar, Limb, and Francis. **Critical revision of the manuscript for important intellectual content:** Nahas and Francis. **Statistical analysis:** Nahas and Francis. **Administrative, technical, and material support:** Tatlipinar and Francis. **Study supervision:** Limb and Francis.

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**REFERENCES**


