Surgical management of temporal meningoencephaloceles, cerebrospinal fluid leaks, and intracranial hypertension: treatment paradigm and outcomes

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Objective. Thinning of the tegmen tympani and mastoideum components of the temporal bone may predispose to the development of meningoencephaloceles and spontaneous CSF leaks. Surgical repair of these bony defects and associated meningoencephaloceles aids in the prevention of progression and meningitis. Intracranial hypertension may be a contributing factor to this disorder and must be fully evaluated and treated when present. The purpose of this study was to establish a treatment paradigm for tegmen defects and elucidate causative factors.

Methods. The authors conducted a retrospective review of 23 patients undergoing a combined mastoidectomy and middle cranial fossa craniotomy for the treatment of a tegmen defect.

Results. The average body mass index (BMI) among all patients was 33.2 ± 7.2 kg/m². Sixty-five percent of the patients (15 of 23) were obese (BMI > 30 kg/m²). Preoperative intracranial pressures (ICPs) averaged 21.8 ± 6.0 cm H2O, with 10 patients (43%) demonstrating an ICP > 20 cm H2O. Twenty-two patients (96%) had associated encephaloceles. Five patients underwent postoperative ventriculoperitoneal shunting. Twenty-two CSF leaks (96%) were successfully repaired at the first attempt (average follow-up 10.4 months).

Conclusions. Among all etiologies for CSF leaks, those occurring spontaneously have the highest rate of recurrence. The surgical treatment of temporal bone defects, as well as the recognition and treatment of accompanying intracranial hypertension, provides the greatest success rate in preventing recurrence. After tegmen dehiscence repair, ventriculoperitoneal shunting should be considered for patients with any combination of the following high-risk factors for recurrence: spontaneous CSF leak not caused by another predisposing condition (that is, trauma, chronic infections, or prior surgery), high-volume leaks, CSF opening pressure > 20 cm H2O, BMI > 30 kg/m², preoperative imaging demonstrating additional cranial base cortical defects (that is, contralateral tegmen or anterior cranial base) and/or an empty sella turcica, and any history of an event that leads to inflammation of the arachnoid granulations and impairment of CSF absorption (that is, meningitis, intracranial hemorrhage, significant closed head injury, and so forth).

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Key Words • cerebrospinal fluid • otorrhea • otorhinorrhea • tegmen repair • temporal meningoencephalocele • mastoidectomy • intracranial hypertension

Several mechanisms have been proposed for thinning of the tegmen cortex of the temporal bone and the resultant CSF leaks. Potential causes include congenital defects, trauma, infection, and intracranial hypertension.6-7,10-12,15 As the middle fossa cranial base develops progressively enlarging defects, CSF pulsations contribute to dehiscence and can lead to meningoencephalocele development and eventual dural disruption.4,8,12,16 This sequence of events results in effusions in the middle ear and mastoid air cells that can manifest as CSF fistulas, through a disrupted tympanic membrane or via the eustachian tube.1-3,5,11,12,15

Given that intracranial hypertension has been implicated as a significant factor in temporal encephalocele formation,2,10-12,15 we hoped to clarify this correlation by reviewing a series of tegmen defects repaired surgically and the associated ICP measurements in these patients. Additionally, we aimed to determine the rate of and indications for VP shunt placement.

Methods

Patient Characteristics

We conducted a retrospective chart review of 23 consecutive patients undergoing a combined mastoidectomy and middle cranial fossa craniotomy for the treatment of a tegmen defect. These patients were all treated at a single institution over a 66-month period (March 2006–September 2011). This review received approval from our local institutional review board.
Surgical Technique

After inducing general anesthesia, a lumbar puncture is performed to determine OP and for placement of a lumbar drainage catheter. The lumbar drain is used intraoperatively and maintained for 48–72 hours postoperatively to aid with decompression of the dural repair. Patients are placed supine with the head turned to the contralateral side (Fig. 1). Lateral positioning is used for very obese patients or those with very limited cervical spine rotation. A Mayfield 3-pin head holder is used for head fixation. The postauricular and inferior temporal area are clipped, prepared, and draped in the usual fashion. A C-shaped retroauricular incision is marked starting 3–4 cm behind the ear, beginning at (or below) the mastoid tip, extending superiorly over the ear, and curving down toward the root of the zygomatic process (Fig. 2). The scalp is incised down to and through the galea. A pedicle of pericranium is created from the superior portion of the incision posterior to the temporalis muscle and is used during closure and reconstruction. The temporalis muscle is elevated in the subperiosteal plane and reflected as one layer with the skin flap. A self-retaining retractor is placed, and a mastoidectomy is performed.

The mastoidectomy allows for closure of the tegmen mastoideum as well as the tegmen tympani and aids in functional preservation of the incus and malleus. After the sigmoid sinus and tegmen are skeletonized, the aditus is identified and opened widely so that the incus and the head of the malleus can be skeletonized in the epitympanum. Careful exploration along the tegmen, usually in the region of the epitympanum, will often allow for identification of the (meningo)encephalocele(s), which may be in contact with the ossicles. The encephaloceles can then be reduced toward the middle cranial fossa.

Aided by the judicious removal of CSF via lumbar drain, the temporal craniotomy is started. A bur hole is created in the temporal region above the tegmen tympani, and the dura mater is dissected away from the inner table of the skull. Because of chronic epidural inflammation, the dura can be extremely adherent, and multiple bur holes may be required to prevent dural violation and injury to the underlying brain during the exposure. A temporal craniotomy is made with a craniotome, and the inferior temporal dura is carefully dissected from the temporal floor. An extradural dissection is performed along the tegmen mastoideum and tegmen tympani, exposing the cranial base defect and any associated meningoencephaloceles. The bony defect is then dissected circumferentially, and when involved, the middle ear ossicles are identified. This dissection allows full exposure of the dural defect(s) (Fig. 3).

The lateral temporal dura is incised and opened. Dural dissection allows for mobilization of the encephalocele, which can be cauterized. An appropriately sized sheet of synthetic collagen-based dural inlay substitute is placed along the intradural surface of the dural defect, and the dural incision is closed with a running 4-0 Nurolon suture (Ethicon, Inc.). Once the dura is reinforced, the osseous defect must be repaired. If the middle ear ossicles are visible through the defect, they must be protected to prevent the dura from contacting them directly and restricting their movement. In this case, a portion of the temporal craniotomy bone flap is cut and appropriately shaped for concave repair of the tegmen tympani. Autologous bone repair of the cranial base defect is also used when there is a large defect (> 1 cm in diameter). A sheet of pericranium (or temporalis fascia if pericranium is unavailable) is dissected free from the scalp flap, placed over the tegmen repair, and covered with a small amount of fibrin glue (Fig. 4). The temporal craniotomy defect is covered with heavyweight titanium mesh (Fig. 5).

For smaller cranial base defects, bone is not used for the repair. In these cases, only the inlay synthetic dural graft and extradural pericranium or temporalis fascia are used, and the temporal craniotomy bone is secured back in position using titanium microplates (Fig. 6). The temporalis fascia and galea are closed in separate layers with interrupted sutures, and the skin is closed with a running absorbable monofilament suture. A sterile mastoid dressing is then applied.

The lumbar drain is used to drain 5–10 ml/hour postoperatively and is usually removed on postoperative Day 3 after a 24-hour period of clamping to ensure there is no recurrent otorhinorrhea. Prior to the lumbar drain’s removal, a determination is made regarding the need for a VP shunt. This decision is based on an assessment of an individual patient’s risk factors for recurrence, including etiology, preoperative OP measurement, and body habitus.

Follow-Up Evaluation

Both radiological and clinical follow-up are required to evaluate the efficacy of the repair and to allow for early recognition of any recurrence of CSF otorrhea. In the immediate postoperative period, a cranial noncontrast CT with fine-cut and reconstructed images of the temporal
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bones is obtained. Following discharge, the patient is seen in the outpatient clinic at 2 weeks and 3 months postoperatively by both the neurosurgeon and the otolaryngologist to evaluate wound healing and the patient’s clinical condition. At 1 year postoperatively, the patient is again seen with a new temporal bone CT to assess the tegmen repair, including osseous integration of any autologous bone graft that may have been used. It is also important to ensure that the middle ear ossicles are free and have not become impacted by the graft. A final CT is obtained and reviewed at the 2-year postoperative outpatient visit. Additional radiological studies are obtained for any new symptoms, including an MRI for the evaluation of any neurological concerns.

Clinical Analysis

Preoperative demographics reviewed for each patient included age, sex, BMI, etiology, history of meningitis, presence of hearing loss, presenting symptoms, history of myringotomy tube placement, laterality of cranial base defect, and presence of an empty sella or contralateral cranial base defects on preoperative MRI. The intraoperative parameters collected were preoperative OP, use of intraoperative lumbar drain, and type of intradural and extradural repair. The need for VP shunting was recorded, as was the recurrence of a CSF leak and associated symptoms. All complications were reviewed.

Statistical Analysis

Standard descriptive methods were used to summarize the data. Frequencies and percentages were used for nominal variables, and means and standard deviations or medians and ranges were used for continuous variables.
Results

Patient Demographics

Twenty-three patients underwent surgical repair of tegmen defects during the study period. A summary of the preoperative demographics is shown in Table 1. The average age of the patients was 55.1 ± 12.5 years. There was no sex predilection, with nearly equal distribution between males (11) and females (12). The patients reviewed were generally overweight, with an average BMI of 33.2 ± 7.2 kg/m². In fact, only 4 patients (17%) had a BMI < 25 kg/m², and 65% (15 of 23) were obese (BMI > 30 kg/m²).

Five patients (22%) had a history of cranial trauma that was suspicious for an inciting event for the development of a tegmen defect and CSF fistula. In 4 patients (17%), there was a history of meningitis. All 23 patients presented with hearing loss in the ear ipsilateral to the tegmen defect to be repaired. The presenting symptom in 19 patients (83%) was CSF otorrhea. In all but 2 of these patients, the CSF otorrhea occurred after placement of the myringotomy tubes for middle ear fullness and fluid. Other presenting symptoms were temporal meningoencephalocele (discovered intraoperatively during a prior otological procedure, 2 patients), an episode of acute otitis media and mastoiditis (1 patient), and pulsatile tinnitus (1 patient). The tegmen defect repairs occurred nearly equally in the right (10 patients) and left (13 patients) temporal bones. However, there were bilateral defects in 10 of the patients (43%). An empty sella was noted on preoperative MRI in 4 patients (17%).

Operative Details

The intraoperative findings are summarized in Table 2. The average preoperative OP obtained during lumbar puncture under general anesthesia was 21.8 ± 6.0 cm H₂O. A lumbar drain was placed preoperatively in every patient, with the exception of 2 in whom difficulty with passing the catheter prevented its use. Both of these patients had had previous lumbar spinal fusion procedures. The lumbar drain was maintained throughout the surgical procedure and was usually removed on postoperative Day 3. In 22 patients (96%), a synthetic dural inlay was used for intradural repair. The one exception was a patient with significant intradural adhesions due to chronic inflammation and very prominent temporal veins. An extradural graft was not placed in this case given concerns that venous drainage might be disrupted. Extradurally, autologous tissue, either pericranium or temporalis fascia, was used to cover the osseous cranial base defect in 20 patients (87%). In 11 of them (48%), a portion of the temporal craniotomy bone was used to cover exposed middle ear ossicles prior to the placement of autologous graft. In 2 patients (9%), allograft was used both intradurally and extradurally. A single patient had only autologous bone used for extradural repair since a significant dural defect was not identified, and this patient only presented with pulsatile tinnitus and not otorrhea.

Intracranial Hypertension Management

Our experience with intracranial hypertension management in this population is summarized in Table 3. Ten patients in the study had a preoperative OP ≥ 20 cm H₂O, with 7 patients demonstrating an OP ≥ 25 cm H₂O. Long-term CSF diversion in the form of VP shunting was suggested to 8 patients. Three of them refused shunts. A

**Fig. 5.** Postoperative coronal CT of the temporal bone demonstrating the autologous bone graft spanning the tegmen defect and the titanium mesh used to repair the craniotomy site.

**Fig. 6.** Illustration of the middle fossa repair of a smaller tegmen defect when the middle ear ossicles are not exposed. An epidural layer of local pericranium (pink layer) is placed, and an intradural collagen graft (gray layer) is used prior to closure of the dura. Printed with the permission of Paul Schiffmacher, 2012.
recurrent leak developed in 1 patient within a week but was managed with aggressive lumbar drainage; despite the recurrence, the patient continues to refuse a VP shunt and remains under close observation. One of the patients who underwent shunting had an OP of 12 cm H2O, but because of the presence of a high-volume leak and a BMI of 40.4 kg/m2 and Type 2 diabetes mellitus.

**Patient Outcomes**

During an average follow-up of 10.4 ± 6.4 months (range 3–26 months), operative repair prevented further CSF leaks or recurrent symptoms in 22 patients (96%). The 1 patient with transient residual postoperative CSF otorrhea was successfully treated with the replacement of a lumbar drain and aggressive drainage for 7 days. With 19 months of follow-up, that patient continues to be free of any further symptoms of CSF leakage. Unfortunately, he did require ossicular chain reconstruction 5 months postoperatively because the tegmen repair was contacting the middle ear structures despite being covered with a segment of the temporal craniotomy bone. In this case, the head of the incus became fixed to the overlying bone graft. Additional complications (Table 4) included 1 case each of local wound infection, meningitis, deep vein thrombosis, and postoperative seizures. These last 3 complications all occurred in 1 patient who had a number of preoperative medical comorbidities, including a BMI of 40.4 kg/m2 and Type 2 diabetes mellitus.

**Impact and Management of Intracranial Hypertension**

Although thinning of the tegmen cortex appears to...
be fairly common in autopsy studies, ranging from 15% to 34%, the occurrence of CSF otorrhea is fairly infrequent.1,4,16 The predisposing factor to the formation of cranial base (meningo)encephaloceles is thought by many to be intracranial hypertension.2,7,11,15 It is believed that the pathogenesis of tegmen thinning shares many characteristics with benign intracranial hypertension, or pseudotumor cerebri.12 Although not our experience, a female preponderance in middle fossa CSF leaks has been reported by many groups.2 Additionally, a patient’s body habitus appears to play a significant role, as these patients tend to be obese (BMI > 30 kg/m²).2,7 Other signs of intracranial hypertension may also be evident, such as the radiographic finding of an empty sella, which was noted in 17% of the patients in our series, a higher rate than the 5%–6% seen in the normal population.7,10 Furthermore, it is likely that the rate of intracranial hypertension is underestimated in this population if the ICP is measured while CSF is actively leaking or at least before the defect(s) is fully repaired. Unrecognized intracranial hypertension may be a reason for recurrent CSF leakage, either from the same site or from a remote cranial base defect, after surgical repair.

Thus, our treatment of this disorder involves not only repair of the disrupted dura and reinforcement of the middle fossa floor but also an assessment of intracranial hypertension and CSF diversion, if present. We use an intraoperative lumbar drain to aid temporal lobe relaxation during evaluation of the tegmen cortex. The drain is then left in place for 3 days postoperatively to allow for decompression of the dural closure during early healing of the repair. The drain is always placed immediately preoperatively under general anesthesia to record the most accurate OP. In our review, the measured preoperative ICPs averaged 21.8 ± 6.0 cm H₂O, with 10 patients (43%) demonstrating ICP > 20 cm H₂O.

Although a CSF fistula develops through a single site in these patients, the disease process more likely represents a global intracranial problem. We found that 43% of our patients had bilateral tegmen defects and thus were at risk for a recurrent CSF leak through the same site on the contralateral side. For this reason, we have selectively used long-term CSF diversion via VP shunting in patients with significant risk factors for recurrence. We propose that those risk factors include a spontaneous CSF leak not caused by a predisposing condition (that is, trauma, chronic infections, or prior surgery), high-volume leaks, CSF OP > 20 cm H₂O, BMI > 30 kg/m², preoperative imaging demonstrating multiple cranial base cortical defects and/or an empty sella turcica, and any history of an event that leads to inflammation of the arachnoid granulations and impairment of CSF absorption (that is, meningitis, intracranial hemorrhage, significant closed head injury, and so forth).

Utilizing these criteria, we suggested VP shunt placement to 8 of the 10 patients with an ICP > 20 cm H₂O, 3 of whom refused this treatment. A recurrent leak developed in 1 of these patients within 1 week of surgical repair. Another patient with a normal OP received a shunt given the presence of other risk factors for recurrence. In general, the decision for VP shunt placement is made on an individual basis. Although we are aggressive with long-term CSF diversion in this group of patients, we believe that the prevention of recurrent or additional leaks and the associated risk of meningitis outweigh the risks related to VP shunts.

**Critique of Current Study**

The limitations of this study include its retrospective nature and limited size. The duration of postoperative follow-up was also variable, with 1 patient not returning...
at all postoperatively and 6 patients lost to follow-up after only 5 months of monitoring. This follow-up may be long enough, however, to identify most recurrent CSF leaks, despite their insidious nature. The 1 recurrence in our series occurred in a patient within a week of his initial tegmen repair, and other reports have indicated that relapses occur most often within a few months of surgery.9 Note, however, that rare cases of recurrent CSF leakage have been reported up to 2–4 years after surgery.2,9 The development of new CSF fistulas at remote sites of the cranial base likely occurs years, or even decades, after an initial leak, and further follow-up of this group of patients is needed.

Relying on an OP measurement at the time of an active CSF leak to determine intracranial hypertension may not always provide an accurate assessment. Some patients with a normal pressure may demonstrate recurrent CSF leakage at the repair site or elsewhere in the cranial base as a result of continued unrecognized intracranial hypertension. In those patients for whom this may be a concern, it may be prudent to perform a lumbar puncture for OP measurement at a later postoperative date.

Conclusions

The occurrence of thinning or dehiscence of the tegmen cortex is fairly common, and a portion of patients with this finding will demonstrate meningoencephaloceles and CSF skull base fistulas. Surgical repair of these defects should address the osseous and dural defects as well as any underlying intracranial hypertension. Assessing the latter and determining which patients will require long-term CSF diversion are difficult in this setting. We have suggested some risk factors that should be considered in making these decisions.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Kenning, Evans. Acquisition of data: Kenning. Analysis and interpretation of data: Kenning. Drafting the article: Kenning. Critically revising the article: Willcox, Artz, Farrell, Evans. Reviewed submitted version of manuscript: Evans. Approved the final version of the manuscript on behalf of all authors: Kenning. Statistical analysis: Kenning. Administrative/technical/material support: Kenning, Schiffmacher. Study supervision: Evans.

References

7. Prichard CN, Isaacson B, Oghalai JS, Coker NJ, Vrabec JT:


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