Cerebrospinal Fluid Leaks and Encephaloceles of Temporal Bone Origin: Nuances to Diagnosis and Management

Dhruve S. Jeevan[1], D. Ryan Ormond[1], Ana H. Kim[2], Lawrence Z. Meiteles[2], Katrina R. Stidham[2], Christopher Linstrom[2], Raj Murali[1]

Key words
- Cerebrospinal fluid leak
- Cranial fossa
- Encephaloceles
- Hearing
- Middle cranial fossa
- Surgery
- Temporal bone
- Transmastoid

Abbreviations and Acronyms
COM: Chronic otitis media
CSF: Cerebrospinal fluid
CT: Computed tomography
MRI: Magnetic resonance imaging

From the Departments of [1]Neurosurgery and [2]Otolaryngology, New York Medical College, Valhalla, New York, USA

To whom correspondence should be addressed: Dhruve S. Jeevan, B.M. B.Ch. [E-mail: dhruve.jeevan@me.com]

Citation: World Neurosurg. (2015) 83, 4:560-566.
http://dx.doi.org/10.1016/j.wneu.2014.12.011

Journal homepage: www.WORLDNEUROSURGERY.org

Available online: www.sciencedirect.com

OBJECTIVE: **Temporal bone encephalocele has become less common as the incidence of chronic mastoid infection and surgery for this condition has decreased. As a result, the diagnosis is often delayed, and the encephalocele is often an incidental finding. This situation can result in serious neurologic complications with patients presenting with cerebrospinal fluid leak and meningitis. We review the occurrence of, characteristics of, and repair experience with temporal encephaloceles from 2000–2012.**

METHODS: **We conducted a retrospective review of 32 patients undergoing combined mastoidectomy and middle cranial fossa craniotomy for the treatment of temporal encephalocele.**

RESULTS: **The diagnosis of temporal encephalocele was made in all patients using high-resolution temporal bone computed tomography and magnetic resonance imaging. At the time of diagnosis, 12 patients had confirmed cerebrospinal fluid leak; other common presenting symptoms included hearing loss and ear fullness. Tegmen defect was most commonly due to chronic otitis media (n = 14). Of these patients, 8 had undergone prior mastoidectomy, suggesting an iatrogenic cause. Other etiologies included radiation exposure, congenital defects, and spontaneous defects. Additionally, 2 patients presented with meningitis; 1 patient had serious neurologic deficits resulting from venous infarction.**

CONCLUSIONS: **The risk of severe neurologic complications after the herniation of intracranial contents through a tegmen defect necessitates prompt recognition and appropriate management. Computed tomography and magnetic resonance imaging aid in definitive diagnosis. A combined mastoid/middle fossa approach allows for sustainable repair with adequate exposure of defects and support of intracranial contents.**

INTRODUCTION

The herniation of dura mater (meningocele) or brain tissue (encephalocele) into the mastoid or middle ear has become a rare condition. The potential devastating complications, such as meningitis, intracranial abscess, hemorrhage, venous infarction, and seizures, necessitate a high degree of clinical suspicion and prompt surgical evaluation. Before the widespread use of antibiotics, temporal encephaloceles occurred in association with chronic otogenic abscess and sinus thrombophlebitis as well as after surgery for these infections. Antibiotic therapy has greatly reduced the incidence of temporal encephaloceles, and a large series has not been reported in the recent literature.

Meningoceles and meningoencephaloceles of the temporal bone still occur. They may also be seen in congenital defects of the tegmen, after surgery to the skull base, after traumatic fractures, as a result of erosion from intracranial tumors, or after radiation therapy (15). Despite improvements in radiologic imaging of the temporal bone, delayed diagnosis of temporal encephalocele continues to occur. This delay is often due to the nonspecificity of clinical signs and symptoms. The most common presenting symptoms are a conductive or mixed hearing loss with a draining ear or serous otitis media. A retrotympanic pulsating mass occasionally can be found on otologic examination or during surgical drainage of the mastoid. Various surgical techniques have been proposed for the closure of this defect. We review 32 cases of temporal encephalocele at our institution between 2000 and 2012 and discuss some of the nuances of its presentation and management.

MATERIALS AND METHODS

A retrospective chart review was performed examining 32 patients treated from 2000–2012 by a single neurosurgeon. All patients identified had been treated for encephalocele of the temporal bone. In 12 patients, spontaneous CSF leaks occurred, and encephalocele was discovered during mastoid surgery in 3 patients. Patients who developed temporal bone CSF leaks after acoustic neuroma surgery were excluded from the study.

Demographic data were collected from office and hospital charts, including age, gender, and comorbid conditions. Information was also collected regarding preoperative diagnostic tests, including laboratory tests, preoperative audiograms, and imaging. All patients underwent combined mastoid/middle fossa repair with autologous bone graft. Intraoperative findings and the use of lumbar drainage of CSF were also noted. The length of postoperative follow-up, postoperative complications, and audiology were also noted.

RESULTS

Patient Demographics

There were 32 patients who underwent 34 operations for temporal encephalocele; 15 of the patients were male. The median age at diagnosis was 51 years (range, 5–74 years). Of patients, 2 had undergone prior repair—1 patient via a transmastoid repair alone and 1 patient with use of only fascia for skull base repair.

Clinical Presentation

Of the 12 patients with spontaneous CSF leaks, 2 had a history of meningitis or presented with meningitis confirmed by lumbar puncture. Otorrhea was a symptom in 10 patients. In 3 patients, otorrhea occurred after tympanostomy tube placement, and in the other patients, otorrhea occurred in the presence of a concurrent tympanic membrane perforation. Beta-2-transferrin testing confirmed the presence of leak in 2 patients. The average duration of symptoms potentially attributable to CSF leak was 5 years. Cholesteatoma had been diagnosed in 3 patients, and 14 patients had a history of chronic otitis media (COM), of whom 8 had undergone prior mastoidectomy. In 3 patients, encephaloceles were found incidentally on surgical mastoid exploration for presumed chronic infection. One patient (5 years old) was found to have bilateral congenital encephaloceles. One patient (47 years old) had received radiation therapy for leukemia at a young age and now presented with meningitis, CSF leak, and bilateral temporal encephaloceles. In 2 patients in our series, there was erosion of the tegmen from intracranial tumors; 1 patient had an epidermoid tumor, and the second patient had a middle cranial fossa meningioma treated with 2 prior resections and radiation therapy. The excision of the tumor and repair of the bony defect were identical to brain herniation, and CSF leak would have resulted without adequate repair. One patient had an acquired encephalocele following trauma, resulting from a gunshot wound associated with extensive skull base fracturing. Presenting symptoms and physical findings are summarized in Table 1.

Preoperative high-resolution temporal bone computed tomography (CT) was performed in all patients with coronal reconstructions using 1-mm contiguous sections. Magnetic resonance imaging (MRI) with and without contrast enhancement was also performed in all patients. All patients were found to have tegmen defects on imaging, with MRI confirming encephalocele.

Surgical Technique

All procedures were completed under general anesthesia. After induction, 7 patients underwent placement of a lumbar drain at the discretion of the operating surgeon. This decision was made based on the presence of CSF leak at presentation, size of the defect, and brain volume status on imaging. In the patients who had a lumbar drain placed, it was maintained for 48–72 hours postoperatively. Patients were positioned supine with the head turned to the contralateral side. Lateral positioning is used for very obese patients. A Mayfield 3-pin head holder is used for patients with poor cervical rotation; otherwise, a donut pillow was used with surgical taping. Facial nerve monitoring was used routinely. A reverse S-shaped retroauricular incision was marked (Figure 1A) starting at (or below) the mastoid tip and extending superiorly over the ear to the temporal line. The temporalis fascia was harvested at this time for later dural repair. The temporalis was divided and retracted using a self-retaining retractor. A mastoidectomy was then performed (Figure 1B).

A temporal craniotomy was performed aided by CSF drainage from the lumbar drain if used. The craniotomy, which rarely needs to measure more than 3 cm × 2 cm, was started just posterior to the root of the zygomatic arch (Figure 1C). Surgery was kept extradural as much as possible. During the craniotomy, care was taken to preserve the lateral edge of the bone to support the bone plate that was inserted to fill the bony defect. An extradural exploration was performed along the tegmen mastoideum and tegmen tympani, exposing the cranial base defect and any encephalocele encountered (Figure 1D). Dura mater was stripped far enough medially and anteriorly to ensure that a second defect in the bone was not missed. Careful review of preoperative CT scan of the temporal bone also helped in detecting additional defects in the bone. At this point, the encephalocele was excised from below. The small opening in the dura mater was repaired with sutures, if possible, or covered with temporalis fascia and fibrin glue. In rare instances, the dural defect could not be fully delineated extradurally, or the encephalocele could not be seen to allow excision, and intradural exploration was performed. In these cases, a dural substitute was used for repair. The bony defect was dissected circumferentially, and the middle ear ossicles, when involved, were identified. A portion of the temporal craniotomy bone flap was cut and appropriately shaped for concave repair of the cranial base with coverage of the entire defect. A sheet of temporalis fascia was placed over the repair and covered with a small amount of fibrin glue. Attention was paid to avoid dripping of fibrin glue onto the ossicular chain. The temporal craniotomy defect was covered with heavy-weight titanium mesh.

The mastoid cavity was packed with fat. The temporalis fascia and galea were closed in separate layers with interrupted sutures, and the skin was closed with running nylon suture. A sterile mastoid dressing was applied. The lumbar drain, if placed, was used to drain 5–10 mL/hour postoperatively and usually removed on postoperative day 3 after a 24-hour period of clamping.

Hearing Status

Preoperative and postoperative audiometric data were available for 25 patients (Table 1). Preoperative testing was not done in 7 patients. A mixed hearing loss was most commonly reported. No patients lost hearing after the repair. One patient sustained additional sensorineural hearing loss secondary to total petrosectomy. Hearing improved postoperatively in 5 patients.

Postoperative Course

Average follow-up was 4.9 years. One patient had a postoperative infection requiring