**Tegmen tympani defect**


**Neurosurgery Department, University General Hospital of Alicante, Foundation for the Promotion of Health and Biomedical Research in the Valencian Region (FISABIO), Alicante, Spain**

**Epidemiology**

Brain tissue herniation through tegmen tympani defect is rare. Although it mostly depends on congenital skull base defects, trauma, infection, and tumours, idiopathic and iatrogenic cases are also encountered\(^1\)\(^2\)\(^3\)\(^4\). They are usually observed in patients who underwent mastoid cavity surgery due to chronic otitis media with or without cholesteatoma.

The incidence of encephalocele developed due to tegmen defect is decreased with the use of broad-spectrum antibiotics and technological developments in ear surgeries.

**Etiology**

Congenital defects of the tegmen tympani develop ventral to the geniculate ganglion and may be due to incomplete ossification of the tegmental process of the otic capsule.\(^5\).

Inadequate closure of the petrosquamous suture may be a factor as well\(^6\).

Defects in the tegmen tympani may be accompanied by defects in the overlying dura, with resultant egress of cerebrospinal fluid (CSF) or cortical tissue forming an encephalocele. Trauma or infection involving the middle ear and temporal bone are likely considerations, but often the otorrhea accompanying a tegmen tympani defect is a spontaneous cerebrospinal fluid otorrhea.\(^7\).

Characteristically, the disorder occurs in otherwise healthy ears and is the consequence of embryogenic faults in the dura mater and adjacent Tegmen tympani. After years of exposure to
physiologically normal CSF pressures, these faults may fistulize into the tympanomastoid compartment and predispose to the development of an encephalocele.

**Types**

Clinical types of tegmen plate defects:

Primary (spontaneous)

Secondary:

Cholesteatoma

Necrotizing inflammations of the temporal bone (diabetes mellitus)

Traumatic

Surgical (iatrogenic)

Neoplastic

Accidently discovered

**Clinical Features**

The onset may be at any age, but is more common after age 40. About 20% of cases have a history of one or more bouts of meningitis.

Spontaneous cerebrospinal fluid otorrhea due to tegmen tympani defects can result in hearing impairment and predispose to meningitis. Seizures or neurological deficits are additional risks, particularly when associated with an encephalocele.

Clinical presentations:

Unilateral persistent OME

Aural fullness+/− hearing loss

CSF otorhea - otorhinorhea

Life threatening events (meningitis)

Pulsating external and/or middle ear masses

History of chronic ear infection

History of ear trauma/surgery
Diagnosis

The site of the leak is characterized by one or more defects measuring 2-5 mm in the dura mater and adjacent bony plate, usually in the area of the petrous ridge. About 25% of defects are associated with small meningoceles or meningoencephaloceles.

Computed tomographic and magnetic resonance imaging, as well as testing with fluorescein dye, provide confirming diagnostic data.

Treatment

Tegmen tympani defect treatment.

Copeland Technique

For repair of spontaneous, postsurgical, or posttraumatic middle ear and mastoid meningoencephaloceles extending through the middle fossa floor, or when performing repair of superior semicircular canal dehiscence, Copeland et al. prefer a standard middle fossa approach with intraoperative facial nerve monitoring.

A curvilinear incision is made beginning in front of the tragus of the ear and ending at the temporoparietal junction.

During dissection, the scalp and pericranium are separated from the temporalis fascia which is harvested for later use. The muscle is then released from its bony origin and turned inferiorly to expose the squamous temporal bone, root of the zygoma, and external auditory canal.
1. The posterior root of the zygoma

2. Asterion.

A craniotomy is performed anteriorly and posteriorly to the external auditory meatus and attempts are made to keep the inferior margin flush to the floor of the middle fossa.

1. External auditory meatus

The temporal dura is dissected off the skull base in a posterior to anterior direction to avoid injuring the greater superficial petrosal nerve or dehiscent geniculate ganglion and facial nerve.
* Defect

** Greater superficial petrosal nerve

Intraoperative mannitol (0.5 mg/kg IV) is given to assist with atraumatic elevation of the temporal lobe.

However, the option of a lumbar external drainage system for cerebrospinal fluid release may lessen the degree of temporal lobe retraction and can be utilized very effectively.

As appropriate to the operative indication, encephaloceles are amputated and breaches in the dura are repaired with 5–0 or 6–0 Prolene suture (Ethicon, Somerville, NJ).

Any obvious encephalocele is carefully removed from the middle ear taking great care not to disarticulate the ossicles. If dehiscent, the superior semicircular canal is occluded with bone dust and bone wax (Angiotech, Vancouver, BC). The middle fossa floor is then resurfaced using the fascia-bone-fascia technique.

A generous piece of temporalis fascia is first laid over the tegmen defect(s). A split-thickness bone graft at least three times the size of the tegmen defect is next harvested from the craniotomy flap.

Rather than simply placing this over the fascia, however, Copeland et al. modified the technique to include fixing the graft to the inferior edge of the craniotomy margin using a single straight plate and two screws from a craniotomy fixation system (OsteoMed, Addison, TX).

Finally, the remaining temporalis fascia is laid over the secured bone graft, against the native subtemporal dura, completing the fascia-bone-fascia sandwich. The entire construct is then covered with fibrin sealant (Baxter, Deerfield, IL). The craniotomy and wound are closed in typical fashion.

Complications

Spontaneous cerebrospinal fluid otorrhea due to tegmen tympani defects can result in hearing impairment and predispose to meningitis. Seizures or neurological deficits are additional risks, particularly when associated with an encephalocele.
Case series

2017

Twenty-six patients the mean age at surgery was 60±14 years and 65% of patients were female. The majority of defects involved both the tegmen mastoideum and tympani (69%); multiple defects were present in 11 patients. Small craniotomy (2 × 3 cm) was performed and defects were repaired using composite grafts constructed with fascia, bone, and/or cartilage, and dural substitute affixed with suture. The suture tail was left long and passed from the middle fossa through the defect into the mastoid. At average follow-up of 8.3 months, no patients of recurrent CSF leak were noted. Significant improvements in both mean pure-tone average and air-bone gap were noted for the entire cohort (p = 0.04 and p = 0.02, respectively).

A combined transmastoid-middle cranial fossa for the repair of lateral skull base CSF fistula and encephaloceles using the suture “pull-through” technique is efficacious and the complication profile is favorable. This method facilitates reliable placement of a composite graft in the center of lateral skull base defects through a small craniotomy that minimizes temporal lobe retraction

2014

Twenty-two patients who underwent surgical repair of tegmen defects associated with cerebrospinal fluid (CSF) leakage and/or meningocele/meningoencephalocele by a combined transmastoid/minicraniotomic approach.

A retrospective review of videos of surgery and charts of patients with tegmen tympani or tegmen antri defects and CSF leakage, temporal lobe encephalocele, and/or meningoencephalocele.

All patients underwent the combined approach and had their defects closed, without significant intraoperative or postoperative complications.

Mastoidectomy with temporal minicraniotomy represents an effective approach in patients with tegmen tympani dehiscence; the advantages of this technique are the control of the floor of the middle cranial fossa and the possibility to reach bony defects located anteriorly without manipulation of the ossicular chain and temporal lobe.

2013

8 individuals who presented with CSF otorrhea and MCF encephaloceles associated with conductive hearing loss. Defects in the tegmen tympani were noted in all patients on preoperative cranial imaging, and six of the eight patients had an associated encephalocele. The average age was 57 years (range 26 to 67) with a male:female ratio of 7:1. Most defects occurred on the left side (6 left/2 right). A standard MCF approach and repair of the dural defect with an autologous dural graft (Durepair or DuraGen, Medtronic, Minneapolis, Minnesota, USA) and a synthetic polymer glue (DuraSeal, Covidien, Mansfield, Massachusetts) was performed in each case with universal success. Resolution of the CSF otorrhea was noted in all cases. All cases but one exhibited an improvement in hearing. One patient developed a delayed methicillin-resistant Staphylococcus aureus meningitis 3 months after surgery that resolved with surgical re-exploration and antibiotic therapy. Facial nerve
monitoring was standard. All patients exhibited normal facial function postoperatively. Prophylactic lumbar drain placement was only utilized in the first three patients. The MCF approach is an excellent route to effectively repair CSF leaks and encephaloceles due to tegmen tympani and dural defects.

2005

11 defects of the tegmen tympani or the mastoidal roof. Routing symptom was in all cases at least one previous meningitis. Radiological diagnostics included high-resolution computed tomography (CT) and magnetic resonance imaging (MRI) as well as CT- or MR-cisternography. The transtemporal approach was also performed in large defects of the tegmen tympani and mastoidal roof as well as in recurrences.

1994

Two clinical cases, the temporal bone histopathological findings in a third case suffering fatal meningitis.

References


