Meningoencephalocele in a Mastoid Cavity: A Case Report and Review of Literature

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ABSTRACT
A young adult male presented to us with a discharging mastoid cavity. Clinical and radiological examination revealed cholesteatoma recidivism along with a meningoencephalocele (ME) which was managed successfully. Two year follow-up revealed no recurrence of the pathology.

Keywords: Meningoencephalocele, Mastoid, Temporal bone

CASE REPORT
A 28-year-old male presented to us with a history of right sided scanty, purulent and malodorous otorrhoea which was present since childhood. This was associated with a hearing loss. The patient had undergone two canal wall down procedures previously.

Otologic examination revealed a narrow meatoplasty. The facial ridge was high. A large pink mass was seen in the ear canal. Cholesteatoma flakes were seen below the mass. Tuning fork tests revealed a conductive deafness in the right ear. The left ear, nose and throat were normal. A clinical diagnosis of discharging mastoid cavity with cholesteatoma recidivism was made.

Pure tone audiometry confirmed the presence of a moderate conductive hearing loss with an air bone gap of 35 decibels. High resolution computerized tomography (CT) of the temporal bones showed a tegmen mastoideum defect with herniation of intracranial soft tissues into the mastoid resembling a tear drop which suggested either a ME or a meningocoele. A soft tissue mass in the attic was also seen [Table/Fig-1]. The facial bridge was intact. It was not excised by the earlier surgeon. Axial cuts revealed a high facial ridge [Table/Fig-2]. The patient could not afford magnetic resonance imaging (MRI).

A revision mastoidectomy was done under general anaesthesia. A pink soft tissue mass was seen in the mastoid cavity which was herniating through a 1.5 cm tegmen defect [Table/Fig-3]. This mass was debulked and excised using a bipolar cautery and sent for histopathological analysis which revealed neuroglial tissue. A cerebrospinal fluid (CSF) leak occurred during the excision which was sealed using a piece of free temporalis muscle.

The facial bridge was removed. Residual cholesteatoma and granulation tissue in the middle ear was dissected and excised. Diseased mastoid air cells were exenterated. The malleus and incus were missing. The stapes was present. The facial ridge was lowered; the mastoid cavity margins were saucerized. The mastoid bowl was smoothened using a diamond burr. The mastoid was washed with normal saline followed by povidone iodine.

The tegmen defect was repaired using a three layer technique of autologous temporalis fascia, pedicled temporalis muscle flap and conchal cartilage. The repair was supported with gelfoam. A type three tympanoplasty was performed. The temporalis fascia was placed under the tympanomeatal flap and on the stapes head. The cavity was packed with ribbon gauze impregnated with antibiotic ointment. The postoperative course was uneventful. The ear healed completely after eight weeks [Table/Fig-4]. The patients hearing also improved with closure of the air-bone gap by 15 decibels.

DISCUSSION
Temporal bone ME are herniations of brain and meningeal tissue into the temporal bone. These rare lesions of the temporal bone were first observed by Von Recklinghausen [1]. Various other names like encephalocele, brain hernia, brain fungus, brain prolapse, cerebral hernia, dural hernia and endaural encephalocele have been given to these lesions. The basic prerequisite for these lesions to occur is a defect in the bony plates separating the middle ear and mastoid from the middle or posterior cranial fossae. Herniations into the middle ear and mastoid from the middle cranial fossa through tegmen plate defects are more common. Posterior cranial fossa herniations are extremely rare, but they have been reported [2].

These lesions have multiple aetiologies. Congenital defects of the tegmen plate due to a dehiscence of the petrosquamous suture in the tegmen tympani can cause these lesions [3]. The commonest cause is inadvertent iatrogenic removal of the tegmen plate during mastoidectomy. Tegmen plate removal alone does not cause a herniation. There should be an associated dural tear for herniations to occur [4,5]. The tear in the dura and a possible small CSF leak may go unnoticed during surgery. Many months or years later, these patients may develop a ME. Chronic otitis media...
The treatment of MEs depends on the size of the defect and whether the ossicular chain is involved or not [13]. Small defects of the tegmen plate and cerebellar plate are treated using the transmastoid approach. A cortical mastoidectomy is performed, the meningoencephalocoele is excised using a bipolar cautery and the defect is repaired using autologous bone or composite cartilage perichondrium graft. The mastoid is obliterated using either a temporalis muscle or Palva flap.

In large defects without ossicular chain involvement, the middle cranial fossa approach is used. The dural fistula is closed using autologous connective tissue like temporalis fascia, perichondrium or fascia lata. The bone defect is repaired using a bone or cartilage graft. A temporals muscle flap is placed between the connective tissue and bone/cartilage graft thus repairing the defect in three layers.

In large defects with ossicular chain involvement, a combined middle cranial fossa and transmastoid approach is used [14]. The ME from the middle ear is excised, the tegmen defect is reconstructed and a tympanoplasty is performed.

CONCLUSION

Soft tissue masses in the middle ear or mastoid which are continuous with intracranial soft tissue structures through a tegmen defect must be suspected as ME and managed as described above. This case also demonstrates the importance of not damaging the dura during otologic surgery.

REFERENCES