DOI: 10.7860/JCDR/2015/10685.5426

Case Report

Ear,Nose and Throat Section

Meningoencephalocoele 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 meningoencephalocoele (ME) which was managed successfully. Two year follow-up revealed no recurrence of the pathology.

Keywords: Meningoencephalocoele, 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.

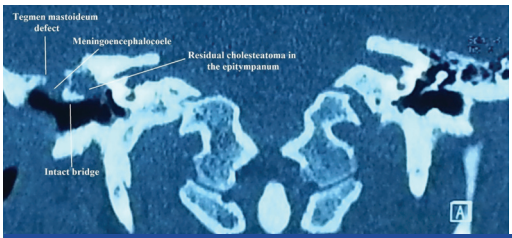
The facial bridge was removed. Residual cholesteatoma and granulation tissue in the middle ear was dissected and excised. Diseased mastoid air cells were exenterated. The malleus and incus were missing. The stapes was present. The facial ridge was lowered; the mastoid cavity margins were saucerized.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

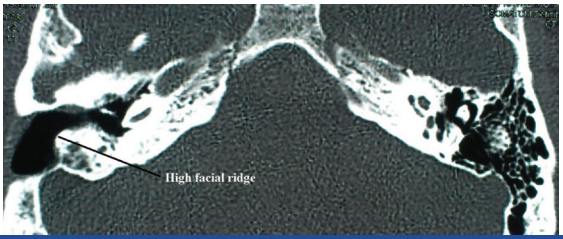
The tegmen defect was repaired using a three layer technique of autologous temporalis fascia, pedicled temporalis muscle flap and conchal cartilage. The repair was supported with gelfoam. A type three tympanoplasty was performed. The temporalis fascia was placed under the tympanomeatal flap and on the 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.

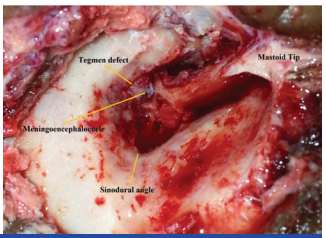
DISCUSSION

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 encephalocoele, brain hernia, brain fungus, brain prolapse, cerebral hernia, dural hernia and endaural encephalocoele 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].

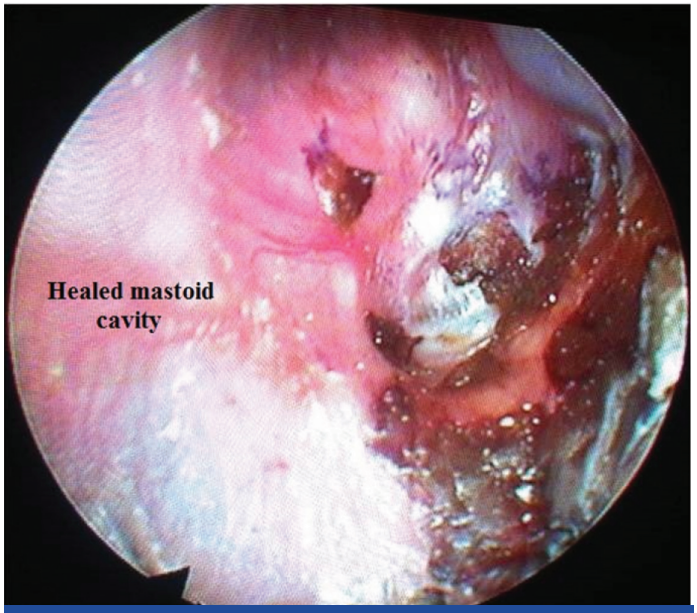
These lesions have multiple aetiologies. Congenital defects of the tegmen plate due to a dehiscence of the petrosquamous suture in the tegmen tympani can cause these lesions [3]. The commonest cause is inadvertent iatrogenic removal of the tegmen plate during mastoidectomy. Tegmen plate removal alone 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







[Table/Fig-1]: Coronal CT revealing the meningoencephalocoele **[Table/Fig-2]:** Axial CT showing a high facial ridge **[Table/Fig-3]:** Operative photograph showing the tegmen defect and meningoencephalocoele



[Table/Fig-4]: Postoperative photograph showing a healed mastoid cavity

with cholesteatoma can erode the tegmen plate resulting in dural exposure. The exposed dura is weakened due to enzymatic lysis and chronic low grade infection resulting in a ME subsequently. ME`s have also been reported following trauma to the temporal bone. The middle cranial fossa dura is tightly adherent to the temporal bone. Fractures of the temporal bone or penetrating trauma in the form of gunshot wounds can breach the tegmen plate and dura resulting in a herniation [6]. Transmastoid drainage of otogenic brain abscesses may also cause ME`s. Spontaneous occurrence of ME`s in adults without a history of ear surgery, trauma or chronic otitis media has been reported [7]. These have been attributed to tegmen defects as a result of chronic CSF pulsations on a thin tegmen plate. Spontaneous ME`s are more common in the elderly and in patients with increased intracranial pressure. Rare causes include cranial irradiation [3], vitamin D deficiency with rickets [8] and ectopic arachnoid granulations in the temporal bone [9].

These patients may present with CSF otorrhoea, CSF otorhinorrhoea, and conductive hearing loss due to CSF in the middle ear or the ME impinging on the ossicular chain. Patients may also present with recurrent meningitis symptoms. Rare presentations are temporal lobe epilepsy, facial paralysis, aphasia, tension pneumocephalus [10] and subcutaneous postauricular mass [11]. Examination may reveal clear fluid in the middle ear. A bluish grey mass may be seen in the ear canal, middle ear or mastoid cavity. The mass is soft to touch, pulsatile and increases in size on performing the valsalva manoeuvre.

CT and MRI play an important role in the management of these patients. High resolution CT of the temporal bones in both axial and coronal planes will clearly reveal the tegmen defect. CT with intrathecal water soluble contrast (omnipaque) helps in identifying possible CSF fistulae. MRI helps in identifying the nature of pathology. It differentiates fluid from soft tissue masses like cholesteatoma, ME and cholesterol granuloma [12].

The treatment of ME`s 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 temporalis 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

[1] Von Recklinghausen FD. Uber multiple hirnhernien. Sitzungsberichte der wurzburger physik med gesellsch. N.F.11,1872.
[2] Ramanikanth TV, Smith MC, Ramamoorthy R, Ramalingam KK. Postauricular cerebellar encephalocoele secondary to chronic suppurative otitis media and mastoid surgery. *J Laryngol otol.* 1990;104(12): 982-85.
[3] Lalwani AK, Jackler RK, Harsh GRiv, Butt FYS. Bilateral temporal bone encephalocoeles following cranial irradiation. *J neurosurgery.* 1993;79: 596-99.
[4] Ramsden RT, Latif A, Lye RH, Dutton JE. Endaural cerebral hernia. *J laryngol otol.* 1985;99(7): 643-51.
[5] Dedo HH, Sooy FA. Endaural brain hernia(encephalocoele): Diagnosis and treatment. *Laryngoscope.* 1970;80(7):1090-99.
[6] Hasso AN, Ledington JA. Traumatic injuries of the temporal bone. *Otolaryngol clin N Am.* 1988;21(2):295-316.
[7] Iurato S, et al. Histopathology of spontaneous brain herniations into the middle ear. *Acta otolaryngol (stockh).* 1992;112:328-33.
[8] Ruttin E. Zur klinik urd pathologie der Hernhernien im bereiche des schlefenbeines. *Monatsschr ohrenheilkd.* 1938;72:197.
[9] Barron SH. Herniation of the brain into the mastoid cavity. Post surgical, Post infectional or Congenital. *Arch otolaryngol.* 1969;90(6): 779-85.
[10] Cartwright MJ, Eisenberg M. Tension pneumocephalus associated with rupture of a middle fossa encephalocoele. *J Neurosurg.* 1992;76:292-95.
[11] Thompson JW. Transmastoid encephalocoeles. A Case report. *Int J pediatric otorhinolaryngol.* 1988;15(2):179-84.
[12] Keseff LG, et al. Magnetic resonance imaging of brain herniation into the middle ear. *Amer J otol.* 1992;13:74-77.
[13] Lalwani AK. Temporal bone encephalocoele. In: Jackler RK, Brackmann DE (eds.) Neurotology. Missouri: Mosby Publishers; 1994.
[14] Sanna M, Fois P, Russo A, Falcioni M. Management of meningoencephalic herniation of the temporal bone: personal experience and literature review. *Laryngoscope.* 2009;119(8):1579–85.

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FINANCIAL OR OTHER COMPETING INTERESTS: None.

Date of Submission: **Aug 24, 2014**
Date of Peer Review: **Oct 10, 2014**
Date of Acceptance: **Nov 21, 2014**
Date of Publishing: **Jan 01, 2015**