Case Report

Management of Meningoencephalocele Herniation of Temporal Bone with Five-Layer Repair in a Tertiary Care Centre

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BACKGROUND: Meningoencephaloceles of the temporal bone are rare and often misdiagnosed, posing a diagnostic and a surgical challenge for the otorhinolaryngologist. Etiologies like chronic otitis media, head trauma or previous surgical procedures involving the temporal bone can lead to it. Timely surgical interventions can prevent life threatening complications like meningitis, encephalitis, and cerebral abscesses.

CASE PRESENTATION: In a pediatric case discussed in this report, the tegmen breach was iatrogenically caused in a prior surgery. Management involved reduction and repair in 5 layers using trans-mastoid approach. The patient improved clinically and post-operative computerized tomography showed repaired and intact tegmen and pure tone audiogram showed improvement in hearing on subsequent follow ups.

CONCLUSION: A holistic approach with timely diagnosis, appropriate radiological assessment to visualise the defect and early surgical intervention are key factors in improving the outcome in patients with meningoencephalocele.

KEYWORDS: Lateral skull base, Meningitis, Meningoencephalocele, Temporal bone.

INTRODUCTION

Meningoencephalocele, fungus cerebri or herniation of brain tissue is through defects in the skull base secondary to congenital causes, cholesteatoma or post otological surgeries.¹

Congenital meningoencephalocele through temporal bone occurs due to the failure in the embryologic closure of the neural tube or defects in skull base which have been scarcely reported in literature.²

The presentation and diagnosis of a meningoencephalocele poses a challenge for otologists because of its overlap with other benign conditions of the ear in the otorhinolaryngology practice like aural polyps, granulation tissue, malignancies.³

CASE REPORT

A 9-year-old boy, fully immunised for his age came to our tertiary care hospital with complaints of watery discharge from his left ear for a year along with a known mass in his left ear for 6 months. The child had undergone modified radical mastoidectomy for atticoantral disease in June 2019 in another hospital.

The child had been asymptomatic for a year after which he developed watery discharge from his left ear, which was followed by a grey coloured mass in the left external auditory canal (EAC) (Fig. 1).

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Fig. 1: Grey coloured mass in the left external auditory canal on presentation.

There was no history of ringing sensation in the ear, facial deviation, headache, double vision, seizures, vomiting or similar complaints of the right ear.

Examination of the left ear revealed a grey coloured mass in the left EAC with watery discharge surrounding it. Hearing tests done with Weber's showed lateralisation to the left and facial nerve was clinically intact.

Pre-operative pure tone audiogram (PTA) revealed left sided moderate conductive hearing loss (55dB loss). and pre-operative computerized tomography (CT) done revealed herniation of the temporal brain parenchyma, measuring 1.8*1.7*1.4 cm, (Fig. 2)

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with intact meningeal covering through a 1.2 cm defect in the tegmen tympani, suggestive of meningoencephalocele.

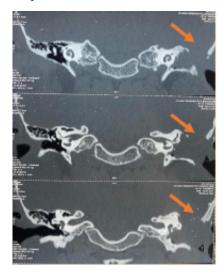


Fig. 2: Pre-operative CT done to reveal the temporal bone defect with the herniated meningoencephalocele.

Pre-operatively, the patient was put on age appropriate coverage of aminoglycoside and cephalosporin betalactam antibiotic along with mannitol and was closely monitored by the pediatrics team. The renal function was taken into consideration since we did not want aminoglycoside induced ototoxicity which could worsen hearing loss.

Operative technique

After taking informed consent, patient underwent left tegmen tympani defect repair using a post aural approach which started with an excision of the herniated brain tissue as it was non-viable. This was followed by five layer closure using fascia lata, bone dust, conchal cartilage, fibrin glue and thigh fat (in this sequence) to repair the bony defect. The conchal cartilage was put for better anchoring. A five layer repair approach was undertaken because this was a pediatric case and intraoperatively two defects were visualised and a layered closure was done to make sure that there is no recurrence in the future. The attic was also partially obliterated with fat and bone dust and tympanomeatal flap was put back and the mastoid was obliterated with thigh fat.

The patient tolerated the procedure well and there were no intraoperative complications. A mastoid dressing was put post operatively and patient was discharged on 2 weeks of post-operative antibiotics.

The tissue sent for histopathology was found consistent with meningoencephalocele. Post-operative PTA showed an improvement in the air bone gap from 55dB to 25dB (Fig. 3).

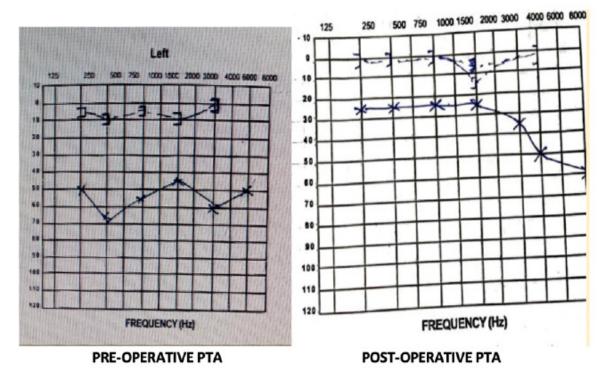


Fig 3: Post-operative pure tone audiogram showed an improvement in the air bone gap from 55dB to 25dB.

Post-surgical high resolution computed tomography of temporal bone done after 3 month showed the repaired site of tegmen tympani with no evidence of brain tissue herniation (Fig. 4).

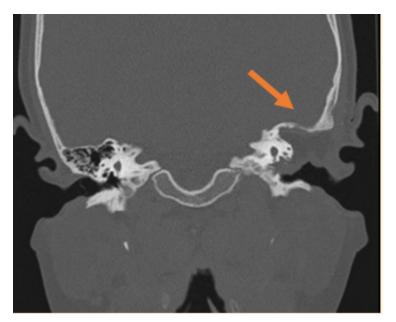


Fig 4: Post-surgical high resolution computerized tomography (HRCT) temporal bone done after 3 months showed the repaired site of tegmen tympani.

This case report has been approved by the institutional review board (IRB) of the Ethical Committee, Kasturba Medical College, Manipal, Manipal University. We have also documented the informed, written consent of the patient's caregivers before surgery and publishing and taken utmost measures to maintain patient anonymity.

DISCUSSION

Meningoencephalocele, fungus cerebri or herniation of brain tissue is through defects in the skull base secondary to congenital causes, cholesteatoma or post otological surgeries.¹

Congenital meningoencephalocele through temporal bone occurs due to the failure in the embryologic closure of the neural tube or defects in skull base which have been scarcely reported in literature.²

The presentation and diagnosis of a mening oencephalocele poses a challenge for otologists because of its overlap with other benign conditions of the ear in the otorhinolaryngology practice like aural polyps, granulation tissue, malignancies.³

The etiology can be attributed to chronic otitis mediasquamosal type, acute otitis media and most commonly after mastoidectomy.⁴

In a case series analysed by Ramalingam et al., patients with meningoencephalocele were analysed and found that in all cases the tegmen was breached as a result of previous mastoid surgery and a trans-mastoid and minicraniotomy approach was used to repair the defect.⁵

An advantage for the prognosis, particularly in this instance, was the ability to distinguish encephaloceles from cholesteatoma using pre- and post-gadolinium weighted magnetic resonance imaging (MRI) which also forms the investigation of choice for accurate and timely diagnosis.⁶

In 15 patients studied retrospectively by Nahas et al., the tegmental defect was located above the attic (tegmen tympani) in 8 cases and above the antrum (tegmen mastoideum) in 12 cases, while in our study the herniated brain tissue along with the meninges was intraoperatively found covering both.⁷

The main form of treatment is surgery through transmastoid approach, the middle fossa craniotomy approach, and the transmastoid and middle fossa combination technique. All repairs must be customised to the type and size of defect and there is no universal agreement on the most appropriate approach. While there is a possibility of a less secure repair in the minimally invasive transmastoid extracranial approach, the middle cranial fossa approach carries a high potential of surgical and perioperative morbidity owing to the need for craniotomy and temporal lobe retraction.

Recent years have showed an increasing trend in the minimally invasive transmastoid approach with a multilayer closure to ensure tight closure and prevent recurrence as also reviewed by Gupta et al in 16 such cases showing improvements in both post-operative scans and audiograms.¹⁰

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CONCLUSION

Prompt diagnosis temporal bone meningo -encephaloceles is diagnostic challenge otolaryngology practice since it mimics common otological symptoms and requires high degree of suspicion. Timely radiological investigation is of utmost important to diagnose the defect in middle cranial fossa. Transmastoid approach is ideal in such cases to reduce the herniated mass and five layer repair was done along with mastoid obliteration. Post-operative audiogram showed improvement in air bone conduction and high resolution computerized tomography (HRCT) showed repaired and intact tegmen.

List of Abbreviations

CT: Computerized tomography. EAC: External auditory canal.

HRCT: High resolution computed tomography.

IRB: Institutional review board. MRI: Magnetic resonance imaging.

PTA: Pure tone audiogram.

Informed Consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal.

Disclosure

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

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