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A Spontaneous Middle Ear Encephalocele with Unilateral Chronic Otitis Media

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Abstract

Introduction: Heterotopic Brain and Encephaloceles of middle ear usually present with symptoms of ear discharge and hearing loss. Some patients have additional symptoms of headache, abnormal ringing sensation and vertigo. Radiology may or may not show a communication with central nervous system in encephaloceles and the bony defect may be unnoticeable or attributed to the thinning of bone due to chronic otitis media. On computerised tomography, no distinction may be made between fluid, granulations, cholesteatoma and other space occupying lesions. Morphologically, it may be identified grossly if it presents with its characteristic pink colour and typical convoluted cerebriform pattern (CCP).

Methods: A 26 years female presented to us with unilateral ear discharge since birth and unilateral hearing loss on ipsilateral side for last 18 months. Clinically, radiologically and morphologically it mimicked chronic otitis media with cholesteatoma with no pre-operative and intra-operative distinction from the later. However, the coronal section at the level of anterior tegmen showed a small defect of the tegmen. T2 weighted MRI confirmed presence of brain tissue in middle ear and mastoid cavity.

Results: Combined Middle Cranial Fossa and mastoid approach were employed, unviable brain tissue was excised and sent for histopathology. Viable brain tissue was reduced and tegmen defect was closed with temporalis muscle, conchal cartilage and temporalis fascia. Histopathology revealed glial tissue, ependyma and choroid plexus without any cholesteatoma.

Conclusion: Tympanic encephalocelesare very rare and may closely mimic chronic otitis media. It may become very difficult to differentiate between the two which may lead to unexpected complications andloss.

Keywords: Encephalocele Congenital Middle Ear Heterotopic Brain Cholesteatoma Cranial Fossa Combined Approach Surgery.

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INTRODUCTION

The most frequent space occupying lesions of middle ear are[1] cholesteatoma, teratoma, ganglioma, meningioma, neuroma, scwannoma[2], adenocarcinoma and hamartoma.[3]

Brain tissue in Middle Ear and Mastoid Cavity is a rare condition which may in continuation with Central Nervous System (CNS) termed as encephalocele or may not have a communication with CNS termed as heterotopic brain or choristoma.[4] Herniation of the brain into the mastoid may be acquired or congenital.[5] The acquired results from the trauma to the middle fossa dura and adjacent bone, either from mastoid surgery or from head injury[6], but can also occur following prolonged ear infection.[7,8]

Schurr[6] uses the term "endaural cerebral hernia" and defines it as prolapse of temporal lobe through a defect in the tegmen tympani and dura overlying the middle ear. The prolapse, however, also occurs through a defect in tegmen antri, as well as prolapse of cerebellum through a defect in the bony covering of lateral sinus and the posterior fossa dura.[5]

Neuroglial Choristomas are composed of glial cells and may also contain neural cells, ependyma and choroid plexus.[1] The aetiology is the presence of a small congenital defect in the overlying temporal bone[9], the tegmen tympani, which permitted herniation of the cerebral tissue. The successive closure of this defect would justify the absence of any connection with the CNS. A pedicle directly connecting the neuroglial tissue with the CNS may become detached, eventually absorbed or vestigial.[2,10]

The Heffner[11] mentioned 3 considerations distinguishing ac horistoma and an encephalocele

- ***Embryological considerations:*** In middle ear there does not seem to be a connection in development between ectoderm and neuroectoderm. The middle ear epithelium develops from the first branchial pouch and, during early embryogenesis, this tissue is notably displaced from the developing brain.
- ***Average age of patients:*** The choristomais present in older people. It seems unlikely that a lesion, which does not usually tend to grow significantly, would be symptomatic after decades, especially when middle ears of children are more prone to produce symptoms.
- The tegmen tympani is very thin and, for this reason, encephaloceles are not rare at all. In some cases, there are no surgically or radiologically recognisable bony defects or CSF leaks. The lack of evidence of a connection does not necessarily prove that it does not exist.[12]

AIM

To study management of a middle ear encephalocele.

OBJECTIVES

- To identify a middle ear encephalocele.
- To decide the best surgical approach for the management of middle ear encephalocele.
- To surgically excise the middle ear encephalocele, repair the defect and manage a close follow up of the case.

CASE PRESENTATION

26 years female presented in Otorhinolaryngology Outpatient Department with complaints if Left ear discharge since birth and reduced hearing from left ear since last 18 months. The discharge was initially episodic but continuous for last one month, scanty, white to yellowish in colour, thick, foul smelling, occasionally blood stained not relieved with medication for last one month. She also complained of reduced hearing from left ear for last 18 months which was slow in onset and not progressive. There was no effect on hearing on cleaning the discharge. Patient did not give any history of recurrent headaches, fever, vertigo, tinnitus, earache and aural fullness. Otoscopy showed posterosuperior retraction of left tympanic membrane with attic destruction. Right ear was normal on otoscopy. Rinne's Test was negative on left side and positive on right side. Webber was lateralised to left ear. Pure Tone Audiometry showed mild conductive hearing loss in left ear. HRCT Temporal Bone showed soft tissue density in epitympanum, aditus, antrum, Prussac's space, middle ear and mastoid air cells with partial necrosis of ear ossicles on left side. Right side was normal. After pre operative investigations and evaluation patient was taken for Combined Left Middle Cranial Fossa and mastoid approach and tympanoplasty in General Anaesthesia.

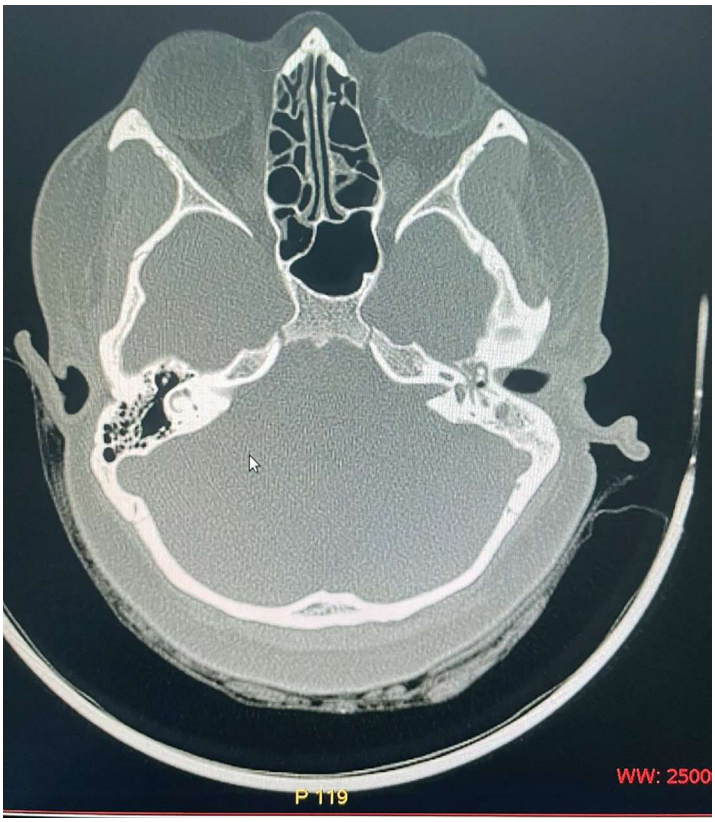

Fig. 1: HRCT Temporal Bone axial section of epitympanum from the console showing a small defect of epitympanum, presence of soft tissue density in leftepitympanum and mastoid air cells with partial necrosis of body of incus. Medial part of external auditory canal is opacified

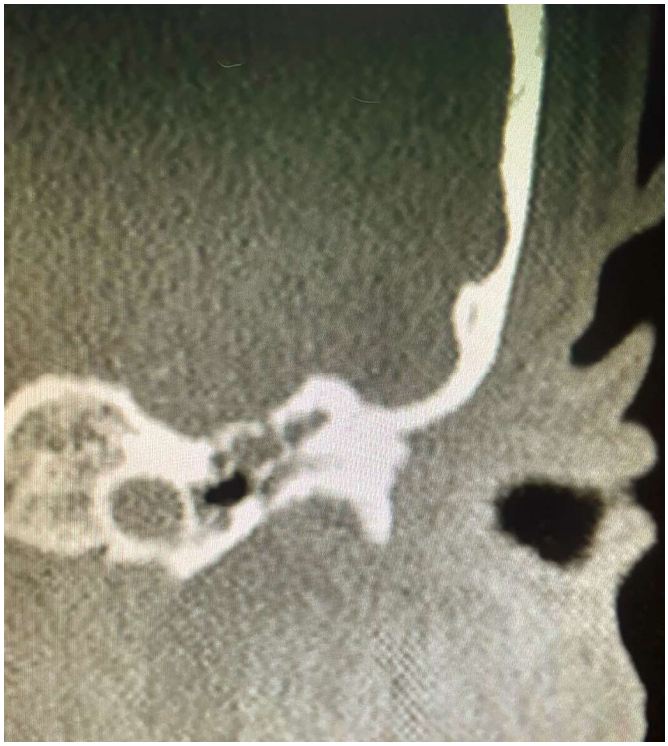

Fig. 2: Coronal Section HRCT Temporal Bone from the console at the level of anterior tegmen showing a defect of tegmen tympani and soft tissue density in middle ear

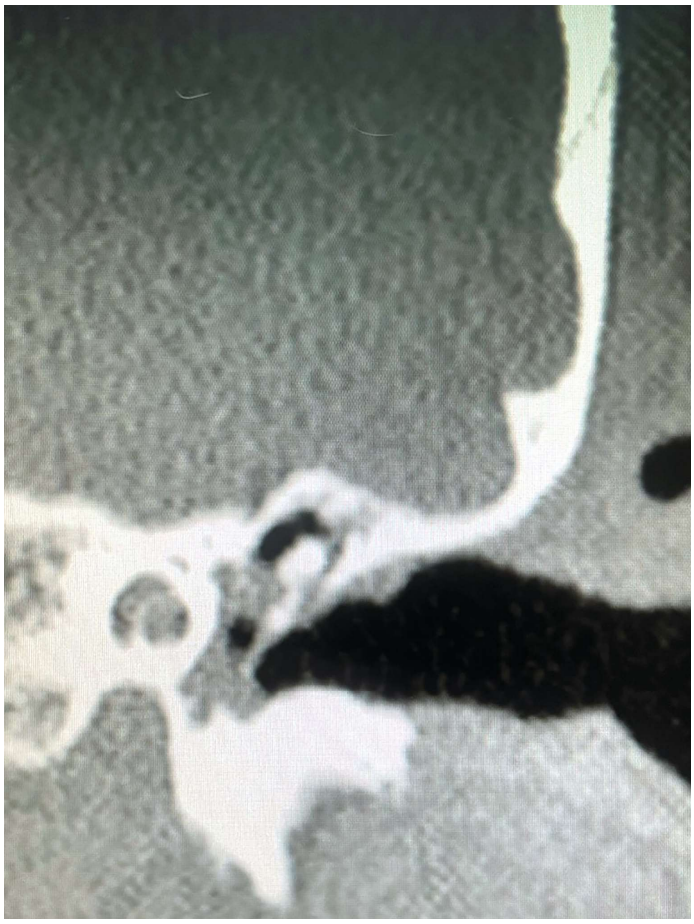

Fig. 3: The coronal view of HRCT Temporal Bone at the level of malleus showing a soft tissue density in middle ear, antrum and mastoid air cells

Intraoperatively, Craniotomy was done and meninges were traced to the inferolateral most portion of the left temporal lobe, Epitympanic defect and brain herniation were identified. The herniating part of the meninges and brain was found to be necrosed. It was excised and defect was closed with surgicell, conchal cartilage and temporalis fascia. Now Wilde's post-aural incision was given on left side and mastoid access gained.Tympanic membrane was retracted posterosuperiorly. On raising tympanomeatal flaps alongwith the tympanic membrane greyish white mass was seen involving whole of middle year and epitympanum. No ear ossicles were seen. Drilling could be just started over Mc Evan's triangle when large greyish white mass was seen touching the outer cortex of mastoid bone. It was freed from its attachments using a ball probe. Posteriorly it was touching the sigmoid sinus. Superiorly it was filling whole of the attic. Inferiorly it was extending till the mastoid tip cells. Anteriorly external auditory canal was preserved. Medially, it was occupying whole of antrum and aditus. No ear ossicles were seen. Slight bleeding was seen while freeing the posterior surface of the mass from the sigmoid sinus for which surgicell wasused and it stopped immediately. The greyish white mass was removed in pieces and sent for histopathology. Type 4 tympanoplasty was done using conchal cartilage and temporalis fascia graft. Epitympanic closure was reinforced with cartilage and fascia. External auditory canal was packed with gel foam soaked in antibiotic ear drops. External auditory opening was packed with Framycetin soaked cotton plug. There was no bleeding or flow of clear fluid at the end of the procedure. Vitals were well maintained throughout the procedure. Incision was closed and mastoid dressings were done. Intraoperative and postoperative period was uneventful.

DISCUSSION

A High-Resolution Computerised Tomography Temporal Boneis a pre-requisite for surgical

planning of mastoid surgery. Imaging slices of 0.5 mm should be obtained in axial, coronal and sagittal view including all sections of the temporal bone to visualize small, fine structures of temporal bone. The three-dimensional (3D) multiplanar reformatted/3D volume rendered CT images are essence of time.[12] Coronal sections of thickness 0.5 mm or less are best to visualize any defects of tegmen tympani, tegmen antri and tegmen mastoid. Instead of the CT/MRI films, all cuts should reach the surgeon on the desktop in his Office using a Pax.

In present sections, we could visualize a defect of Tegmen tympani pre-operatively. I always write in my prescription HRCT Temporal Bone 0.5 mm coronal, axial sagittal all cuts. But 2 mm cuts are never sufficient for HRCT Temporal bone. Neither we nor other otologists suspectedan encephalocele thorough clinical history and examination. It clearly seemed a case of chronic otitis media with cholesteatoma. But the radiologist could find a hidden encephalocele. The surgery was done combinedly by a Neurosurgeon and an Otologist. The Otologist here has an experience of over 5000 mastoid surgeries.

We saw the huge greyish white mass touching the outer mastoid cortex just at the beginning of mastoid drilling. So, practically no drilling was required. The mass was separated carefully from the surrounding anatomical structures and sent for histopathology. There were no intra-operative and immediate post-operative complications.Histopathology revealed presence of central nervous tissue alone. No cholesteatoma was seen in histopathology

Utilizing intraoperative video recording in synergy with a written operative report also proved to be feasible and furthermore, superior to the classic narrative operative report alone[13,14]. We hope that with better pre-operative, intra-operative and post-operative workup, vigilance and facilities such surgeries may be undertaken successfully.

CONCLUSION

However, every disease has a natural history and most of the times it behaves in the confines of the same pattern but still exceptions are possible. In diseases with rare findings, only continued vigilance and suspicion from every discipline for each and every patient and best of diagnostic and operative facilities in Departments well equipped with trained manpower leads to successful outcome.

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