UNILATERAL MEMBRANOUS ATRESIA WITH OSSICULAR MALROTATION AND CONGENITAL CHOLESTEATOMA

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ABSTRACT

INTRODUCTION

Congenital cholesteatoma is a rare entity, arising from aberrant epithelial remnants left at the time of closure of the neural groove between the third and fifth week of fetal life with incidence ranging from 4 to 24%. Congenital membranous atresia is more common on right side and unilateral presentation is a rarity in females which is seen in our case. Here we report a rare case of 44 year old female with bilateral hard of hearing which is more on right side with ear discharge and was diagnosed by CT scan to have congenital unilateral membranous atresia with rare finding of ossicular malrotation associated with congenital cholesteatoma which are extremely rare combination of findings in a single patient. Knowing such rarity may help in appropriate surgical approach when confronted with such cases in clinical practice.

KEYWORDS

Congenital Cholesteatoma, Unilateral, Membranous Atresia, Ossicular Malrotation, Congenital Atresia.

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INTRODUCTION: Congenital cholesteatoma of the middle ear was first described by Howard House in 1953. Congenital cholesteatoma arises from aberrant ectodermal remnants that are trapped within the temporal bone during embryogenesis. Its incidence ranges from 4 to 24%.[1] Congenital membranous atresia occurs in about 1 in 10-15,000 births and up to 50% is associated with some craniofacial syndromes like Treacher Collins, Nager, Crouzon's syndrome but they usually present as bilateral cases. The pathogenesis of cholesteatoma growth is still poorly understood as evidenced by the multiple theories currently found in the literature. The most accepted theory is the epithelial cell rest theory. Rarely, they present as unilateral cases with associated congenital cholesteatoma in it. They are more commonly seen on right side and show male predominance.

CASE REPORT: A 44year old female came with the complaints of right ear block and ear discharge since childhood on left side with pain and hard of hearing on both sides (right >left) since 1 year. No significant trauma history, tinnitus, headache or giddiness.

On examination, vitals were stable and ENT examination showed normal external auditory canal on left side while it was found to be atretic on right side. Tympanic membrane was not visualized on right side and left showed subtotal perforation.

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Rinne test showed bone conduction >air conduction bilaterally, Weber was lateralized to left. Audiogram showed moderately severe mixed hearing loss on right side (58.3dB) and moderate conductive hearing loss on left side (45dB). Fig. 1.

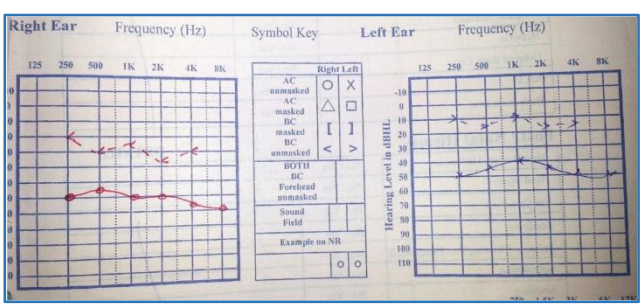
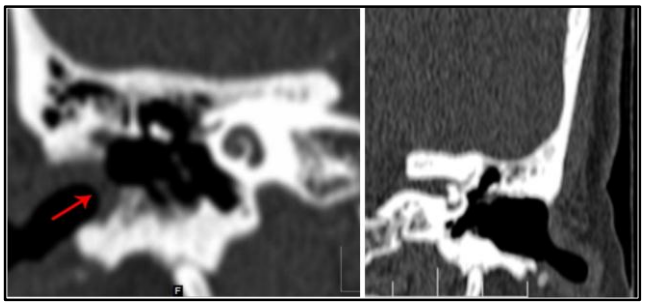


Fig. 1: Audiogram of both ears showing right moderate mixed hearing loss (58.3dB) and left moderate conductive hearing loss (45dB)

CT IMAGING: Right temporal bone:

- Soft tissue density noted in the right external auditory canal with non-communication of the entire auditory canal with middle ear-suggestive of membranous atresia. Fig. 2, 3.
- Right malleus is hypoplastic with non-visualization of handle of malleus with right incudo-stapedial joint being anteriorly and inferiorly located compared to left side and both anterior and posterior crux of stapes visualized. Fig. 4, 5, 6.
- Inner ear structures-cochlea, vestibule, semi-circular canals, internal auditory canal and vestibular aqueduct are normal. Right mastoid air cells are normal.

Left Temporal Bone: Soft tissue opacification noted in mastoid air cells, aditus ad antrum, Prussack space with sclerosis of mastoid air cells–suggestive of left mastoiditis. No erosion of ossicles noted. Fig. 7, 8.



A B

Fig. 2A: Coronal section of right temporal bone shows soft tissue density noted in the external auditory canal– suggestive of membranous atresia. 2B-Normal external auditory canal on left side.

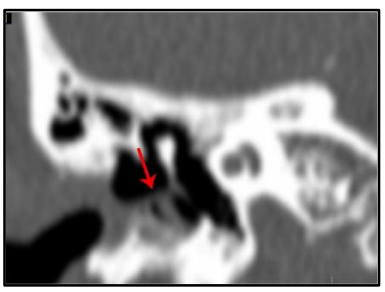
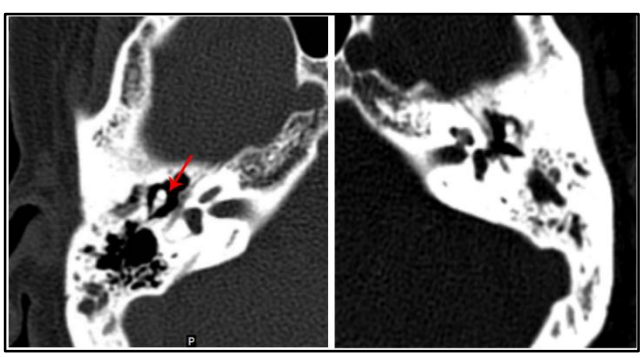


Fig. 3: Coronal section shows soft tissue opacification in the right hypotympanum with few air pockets and no bony erosion-most likely congenital cholesteatoma.



A B

Fig. 4A: head of malleus and body of incus showing normal ice cream cone appearance on right side. 4B-Left side shows normal head and handle of malleus with body of incus.

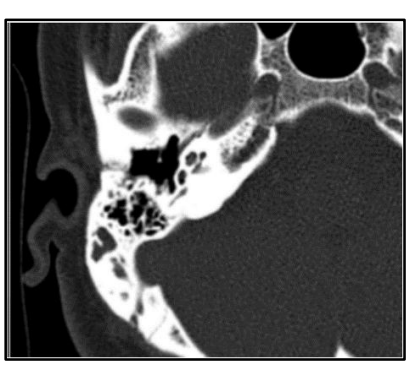


Fig. 5: CT right temporal bone axial section at the level of mesotympanum shows only long process of incus with absence of handle of malleus and appears smaller in size compared to left.

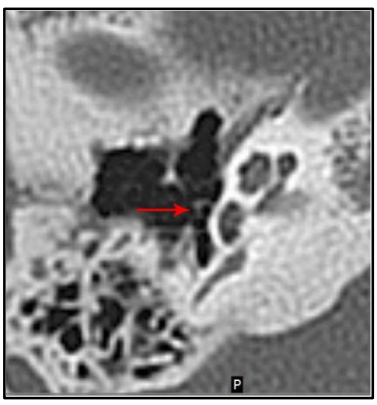
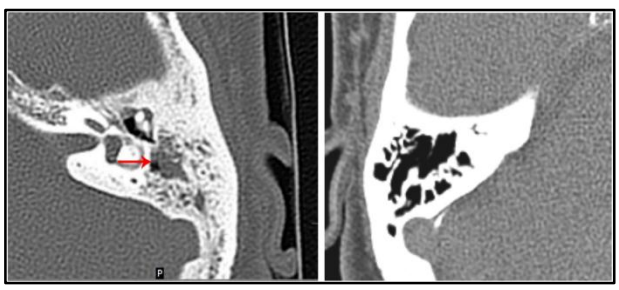
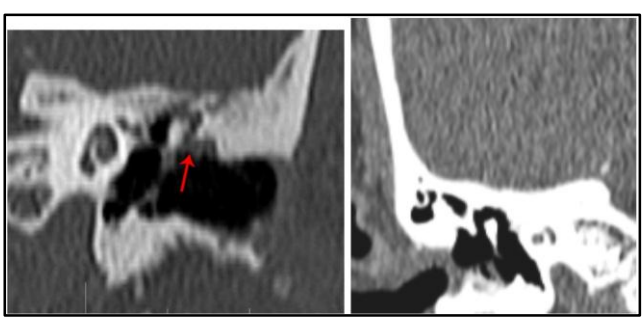


Fig. 6: CT coronal section shows normal foot plate of stapes on right side but right incudo-stapedial joint is in lower position and mild anterior displacement compared to left side.



A B

Fig. 7A: CT Left temporal bone axial section shows soft tissue opacification of the mastoid air cells and in Prussack space. 7B–Normal mastoid air cells on right side.



A B

Fig. 8A: Left middle ear coronal section shows soft tissue opacification in Prussack space and epitympanum– suggestive of chronic suppurative otitis media (CSOM). 8B–Normal right Prussack space

Otoscopic findings confirmed the same. Intraoperatively, membranous atresia with absent handle of malleus and abnormally positioned head of malleus, body of incus confirmed the imaging findings. Cholesteatoma was noted on the right side, hence underwent right external auditory canal atrerioplasty with myringostapediopexy using autologous malleus was done. Postoperative period was uneventful.

DISCUSSION: External ear develops earlier than middle ear and so atresia imply an arrest in the development at any stage. External ear which is an ectodermal derivative arises from 1st branchial cleft. A solid core of epithelium migrates inward towards the 1st branchial pouch (endoderm) and re-

canalizes by 6th month by hollowing out from medial to lateral.

So atresia implies an arrest of recanalization process leading to various deformities. External auditory canal atresia is of 2 types–bony and membranous atresia.

It occurs bilaterally in most of the cases and of which 61% are males. When unilateral, it most commonly occurs on right side (58%).[2] Colman classified into 3 types–minor aplasia (partial recanalization)–incomplete recanalization, moderate aplasia–tympanic bone has developed but has failed to recanalize, severe atresia–complete absence of the external canal. There are numerous possible ossicular chain abnormalities associated with this disorder ranging from bony fusion of middle ear ossicles to hypoplasia or disruption.[3] For unilateral atresia, surgical repair is usually postponed due to normally functioning opposite ear but surveillance of the affected ear after teenage is necessary for the presence of congenital cholesteatoma or any involvement of middle or inner ear which warrants immediate intervention to prevent further damage.[4]

In our case, there was fully developed tympanic membrane and bony canal with a stenotic membranous canal leading to canal cholesteatoma causing ossicular chain malrotation on right side.

Congenital external auditory canal (EAC) cholesteatoma is a rare entity, arising from aberrant ectodermal remnants left at the time of closure of the neural groove between the third and fifth week of foetal life.[5] The annual incidence of cholesteatoma is reported as 3 per 100 000 in children and 9.2 per 100 000 in adults with a male predominance of 1.4:1.[6] Typical congenital cholesteatoma is found in the anterior mesotympanum or in the peri-eustachian tube region. They are identified most commonly in early childhood.[7,8] In adults, EAC cholesteatoma occurs mostly in 40-70 years.

Conductive hearing loss is the most common presenting symptom. There is a lack of uniformity of reporting and classifying congenital cholesteatomas.[9] It is differentiated from acquired cause according to clinical criteria: intact tympanic membrane, no history of aural infection and no history of tympanic membrane perforation or surgery.[8,10] In our case, it was a congenital variant with intact tympanic membrane and no aural infection.

CONCLUSION: Management of unilateral atresia can be challenging due to its rarity in clinical practice and its variable presentation with associated syndromes or cholesteatoma or with ossicular malformation. Hence proper follow up after teenage is necessary with High Resolution CT in unilateral cases and surgical management has to be tailored to individual patient and their clinical presentation.

CLINICAL SIGNIFICANCE: Unilateral membranous atresia with associated congenital cholesteatoma is a rare entity. A high index of suspicion during clinical and radiological evaluation would go a long way in diagnosing and managing such a rare condition at the earliest. Possibility of ossicular malrotation prior to surgery should be borne in mind which alters the approach of intervention.

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