ORIGINAL RESEARCH PAPER

Volume-7 | Issue-9 | September-2018 | PRINT ISSN No 2277 - 8179

INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

CONGENITAL CHOLESTEATOMA; AN ISOLATED RARE CASE OF MASTOID ORIGIN: A CASE REPORT

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ABSTRACT

We report a rare case of congenital cholesteatoma arising from the mastoid . A 45 year old female presented with blocking and ringing sensation along with decreased hearing in the right ear for last 6 months with no history of otorrhoea , tympanic membrane perforation or any otological surgery. Case was evaluated and surgical exploration done .

KEYWORDS

Congenital Cholesteatoma In Mastoid , Mastoid ,assam Medical College

INTRODUCTION

Congenital cholesteatoma is mostly found in the middle ear cavity but the mastoid is the rarest site for the onset of congenital cholesteatoma . A high grade suspicion is required to diagnose it .The symptoms are typical and minimal with evidencing no prior history of otorrhoea, tympanic perforation, or previous otologic procedures ; a normal pars flaccida and pars tensa ; and a pearly white mass medial to the tympanic membrane.

CASE REPORT : A 45 year old female presented in outpatient department of Otorhinolaryngology , Assam Medical College and Hospital , Dibrugarh with history of blocking and ringing sensation in the right ear along with decreased hearing in the right ear for last 6 months with no history of otorrhoea , previous tympanic membrane perforation or any prior otological surgery. She also complained of dull aching headache on the right side but there was no vision error or associated with nausea and vomiting. There was no any suggestive allergic history or any other systemic involvement.

On Speculum examination , she had bilateral normal external ear , external auditory canal , intact tympanic membrane . On Otoscopic examination , she had intact bilateral ear and tympanic membrane with no discharge . On performing RINNE'S test she had bone conduction > air conduction on diseased ear and WEBER test lateralised to the right ear . On PURE TONE AUDIOMETRY , right ear had moderate conductive hearing loss of 40 decibal and on left ear hearing was within normal limit. The HRCT of temperomastoid region showed a non- enhancing soft tissue density lesion at the right sided posterior inferior mastoid air cells with bony wall dehiscence at posterior wall, sigmoid fossa and jugular fossa. No extramastoid extension was noted. Tympanic membrane and ossicular chains were intact. Vestibular apparatus , semicircular canals, cochlea , internal auditory meatus were intact.

Mastoid exploration of the right ear was done under general anaesthesia on 11/07/14. The soft tissue mass was seen to occupy in the posterior inferior portion of the mastoid eroding its posterior wall with exposure of adjacent dura , sigmoid sinus , and jugular fossa and also extended to the petrous apex. The mass was fully resected. Peri and postoperative period was uneventful . The mass was sent for histopathological examination which showed homogenous cartilage material with foci of calcification and inflammatory cells at places suggestive of choleasteatoma.

DISCUSSION :

Kojima et al[1] reported congenital cholesteotoma in mastoid was only 4%.

Congenital cholesteatomas have historically been considered a rare disorder. It was first reported by HOUSE et al [2] (1953) that classically presents as a white mass situated in the anterior superior quadrant of the middle ear behind an intact tympanic membrane associated with ipsilateral conductive or sensorineural hearing loss.

Derlacki and Clemis et al[3] established the diagnostic criteria of congenital cholesteotoma . These are (1) pearl white mass medial to the tympanic membrane.

(2) Normal pars flaccida and pars tensa

(3) no prior history of otorrhoea , tympanic perforation, or previous otologic procedure Besides the migration of epithelial cells through the intact tympanic membrane , the reflux of amniotic cells , the metaplasia theory and aberrant embryonic cell rests . It has been suggested that congenital cholesteotomas may develop from “epidermoid formations” that persist from the embryonic period. The most common sites of presentation are the anterior-superior and posterior-superior quadrants of the tympanic membrane . Conductive hearing loss is the most common presenting symptom . **Derlacki and Clemis** et al[3] were the first to classify congenital cholesteatoma. They classified congenital cholesteatoma into

1. Petrous pyramid cholesteatoma
2. Cholesteatoma involving the mastoid cavity
3. Cholesteatoma involving the middle ear cavity

Isolated congenital cholesteatoma of the mastoid process is the rarest form of presentation in the temporal bone , with few such cases described in the literature . The only studies found in the literature similar to the present case were conducted by Derlacki & Clemis et al[3] in 1965, Luntz et al[4] in 1997 , and Movie et al[5] in 2002. These authors reported the appearance of cholesteatoma of the mastoid without progression to other regions of the temporal bone , removing all doubts as to their origin.

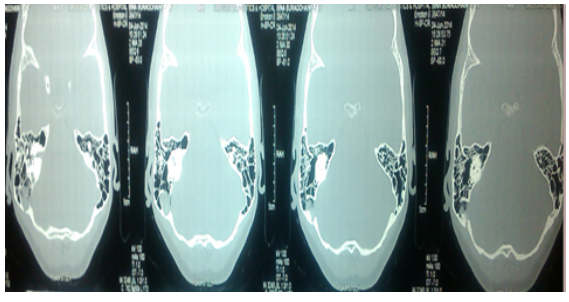
Treatment of congenital cholesteatoma is still surgical. Postoperative hearing results are associated with the status of the ossicular chain preoperatively. In the case reported cholesteatoma mass was detected in the posterior inferior portion of mastoid region, excised mass was 2 cm in size and as per DERLICKS criteria this case falls under stage 2.

CONCLUSION:

Here ,we described a rare case of isolated congenital cholesteatoma in the mastoid process reported in AMCH , DIBRUGARH , ASSAM . High grade of clinical suspicion is required to diagnose such a rare case . Diagnosis of such a rare case is done primarily on the basis of the typical clinical and imaging features as described above . However , this suspicion requires subsequent histological confirmation . Management of this extremely rare condition is done by surgical treatment with full lesion resection and periodic postoperative follow-up.

PHOTOS RELATED TO THE REPORT :

1.



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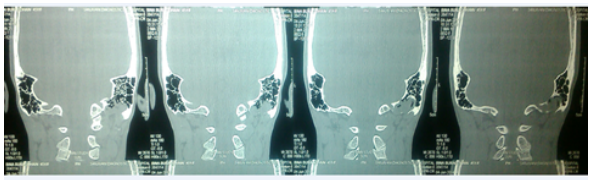

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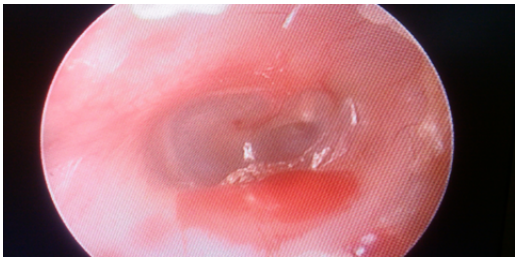

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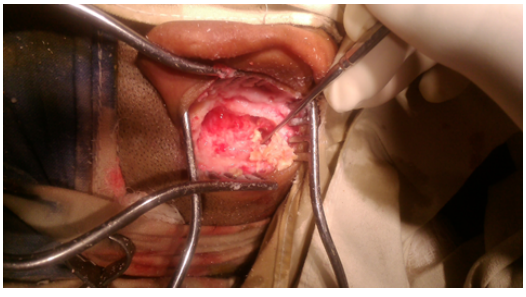

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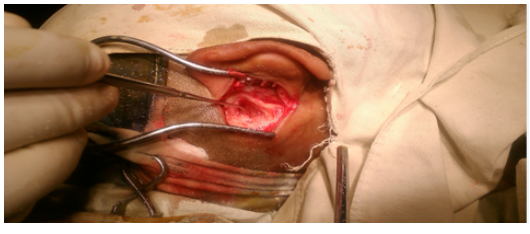

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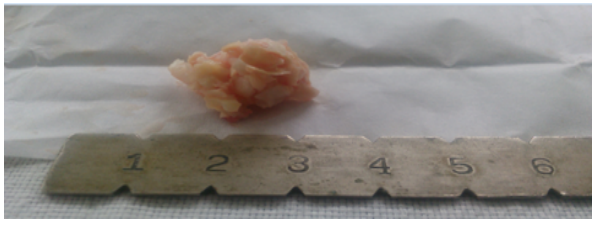

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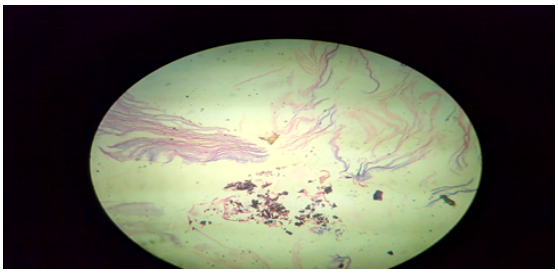

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