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CONGENITAL CHOLESTEATOMA ISOLATED TO MASTOID PROCESS : A CASE REPORT

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ABSTRACT

Congenital cholesteatoma(CC) accounts for 2-5% of all cholesteatomas[1] most common site being middle ear. We report an extremely rare case of congenital cholesteatoma isolated to the mastoid process, with no middle ear involvement. An 18 year old male presented with ear ache and minimal discharge for 4 months. On examination, external auditory canal was found narrowed with granulations and bony defect present in the postero-inferior part of canal. Computed tomography showed soft tissue contents in the mastoid bone causing full thickness erosion of the same along with attenuation of ear canal.

At surgery, a large cholesteatoma sac was found within the mastoid process completely eroding it and extending to posterior part of external auditory canal. The mastoid antrum and aditus were found normal. Tympanic membrane was intact. CC isolated to the mastoid was diagnosed. Diagnosis of CC isolated to the mastoid should be based on clinical examination and radiological evaluation.

INTRODUCTION

Congenital cholesteatoma is an epidermal cyst arising from congenital remnants of keratinizing squamous epithelium in the temporal bone[2]. CC of the temporal bone can be found extradural in the middle ear or mastoid and intradural, most commonly at the cerebellopontine angle. CC of the mastoid process is the rarest form of presentation, accounting for 3-4% of all CC cases [3] . The symptoms are atypical and in reported cases the most common presentation was an incidental finding during radiologic evaluations. Other symptoms seen are retroauricular pain, dizziness and intermittent ear discharge. CC of mastoid origin could be diagnosed if the patient had intact tympanic membrane, intact skin of external canal, no ossicular erosion and no attic involvement of cholesteatoma in intraoperative and imaging findings. CC of mastoid origin can exist for years in an indolent state, develop to giant size and, in some cases, rapidly cause bone destruction and serious complications[4].

CASE REPORT

An 18 year old male presented with a 4 month history of ear ache and mild ear discharge from the left ear. There was no history of decreased hearing, trauma, prior otorrhea or any otological procedures. On otoscopic examination, there were granulations along the floor of the canal, obscuring the tympanic membrane. Pure tone audiometry revealed a conductive loss of 15 db in the left ear. Topical steroid antibiotics were given for 2 weeks following which examination under microscope was done. It revealed narrowing of the ear canal with minimal granulations along the floor and a bony defect with keratinous debris along the posteroinferior part of the canal. Granulations were biopsied.

Tympanic membrane was found normal. Computed tomography of the temporal bone was done which revealed soft tissue contents within mastoid process with full thickness erosion of the bone, along with involvement of the posterior ECA and causing mild attenuation of the same. The mastoid antrum and aditus were normal. [Figure 1,2] Sinus plate erosion was present. Tympanic cavity , ossicles and tegmen tympani were found to be normal.[Figure3]

Based on the CT findings, the patient was taken up for a left exploratory tympanomastoidectomy. At surgery, a giant cholesteatoma sac was seen within the mastoid process completely eroding it and extending to the bony canal. The sinus plate was found eroded and the fallopian canal was found dehiscent in the vertical segment, which was anomalous in its course, located more anteriorly. The mastoid antrum, aditus and middle ear cavity along with the ossicles were found normal. [Figure 4]

The cholesteatoma sac was removed and and canal wall down mastoidectomy was done. The sac was sent for histopathology. Post surgery the patient is asymptomatic and is currently under follow up for 6 months with no residual disease.

DISCUSSION

In 1953, Howard House was the first to describe a cholesteatoma behind an intact membrane[5]. In 1965, Derlacki and Clemis described 6 cases of CC and established the clinical criteria for the diagnosis. It includes a pearly white mass medial to an intact tympanic membrane, a normal pars tensa and flaccid and no history of otorrhea, perforation or previous otologic procedure[6] . Luntz et al. in 1997, were the first to describe the imaging features of an isolated congenital cholesteatoma of the mastoid, and defined three aspects characteristic of this disease: 1) pain in the upper neck; 2) CT scan revealing a lytic and expansive lesion affecting the mastoid process, without compro¬mise to the middle ear; 3) MR examination revealing hyperintensity on T2-weighted images, with no significant postcontrast enhancement[7]. These 3 criteria may not be present in all patients, and other symptoms may coexist in this disease. High-resolution CT scan demonstrates a nonspecific, nonenhancing delineated soft tissue mass within the temporal bone. MRI can be used to distinguish cholesteatoma from other soft tissue masses. The specific site and rarity of the lesion should involve careful differential diagnosis from all other possible tumors: tympanojugular paragangliomas, tumors of the endolymphatic sac, meningiomas, cholesterol granulomas, an anomalous sigmoid sinus, and neoplasmas [8]. Warren et al described 9 cases of CC, and in that series, the most common presentation was an incidental finding during radiologic evaluations [9].

In the literature there are a few case reports about congenital cholesteatoma of mastoid origin [4,10,11,12]. The symptoms are atypical and include ear ache, retroauricular swelling and pain and dizziness[10] or patients can be asymptomatic and present with narrowing of the ear canal. In our case the patient presented with ear ache and discharge.

Erosion of the mastoid cortex could be the reason of pain because the disease involves the periosteum. Intermittent ear discharge was a symptom which can be due to inflammation of intact meatal skin[13]. Surgical management of these lesions is primarily directed by intraoperative and imaging findings. If the lesion is gigantic and where posterior canal wall is destroyed, a canal wall down mastoidectomy should be done and for limited disease a canal wall up surgery should suffice.

CONCLUSION

We have presented a case of congenital cholesteatoma of the rarest variety presenting with minimal clinical symptoms, which was diagnosed with the help of radiological examination and managed with surgical excision. Although congenital cholesteatoma of mastoid origin has a congenital origin, it tends to present in adulthood probably because of minimal symptoms and delay in seeking medical advice. Otologists should maintain a high index of suspicion in cases of normal otomicroscopic examination, ear pain, mild ear discharge and dizziness. Prompt imaging and diagnosis of congenital cholesteatoma is most important to prevent serious complications.

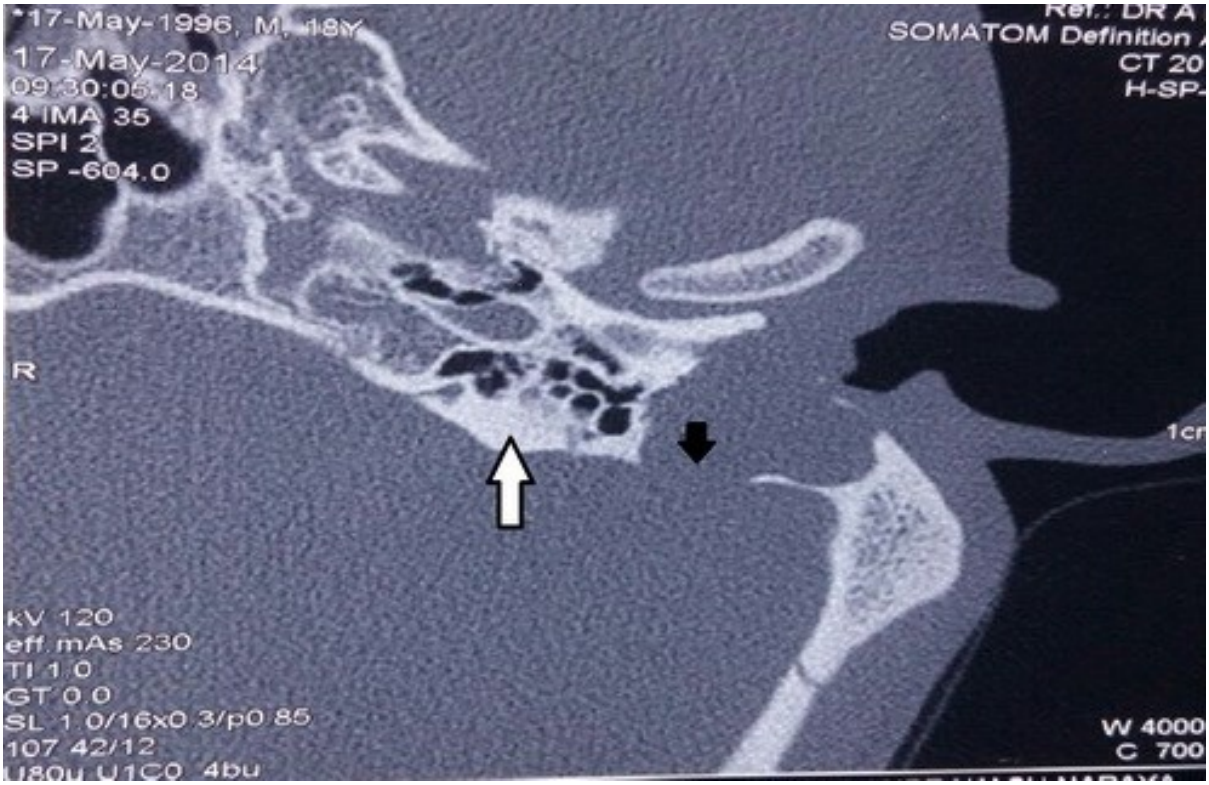

Fig 1: Showing normal septate air cells of mastoid antrum (white arrow) and sinus

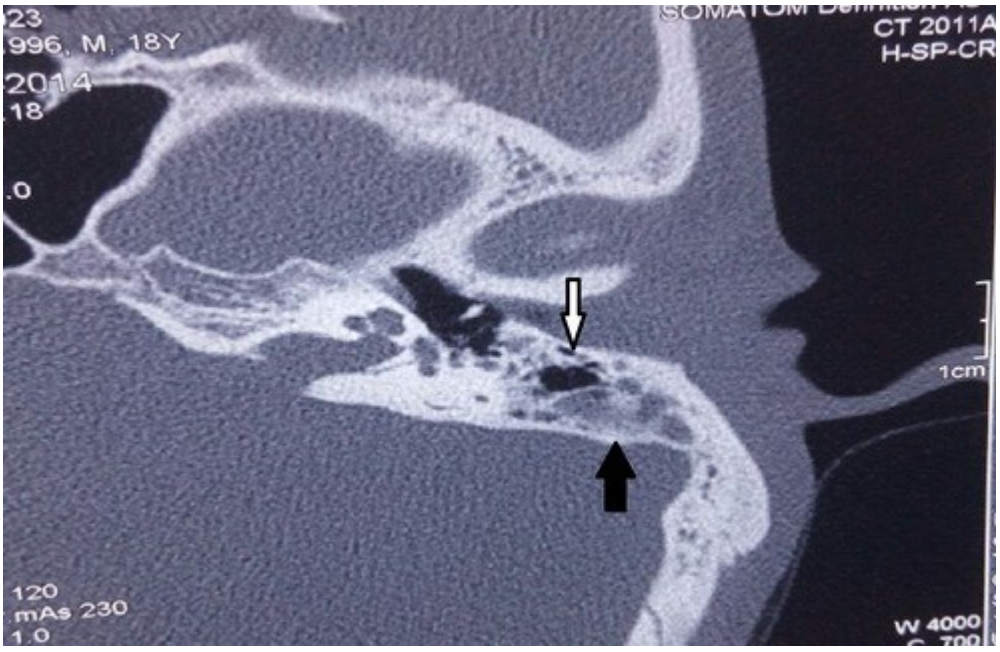

Fig 2: Erosion of mastoid process leading to cavity formation (white arrow) and soft tissue contents due to cholesteatoma (black arrow)

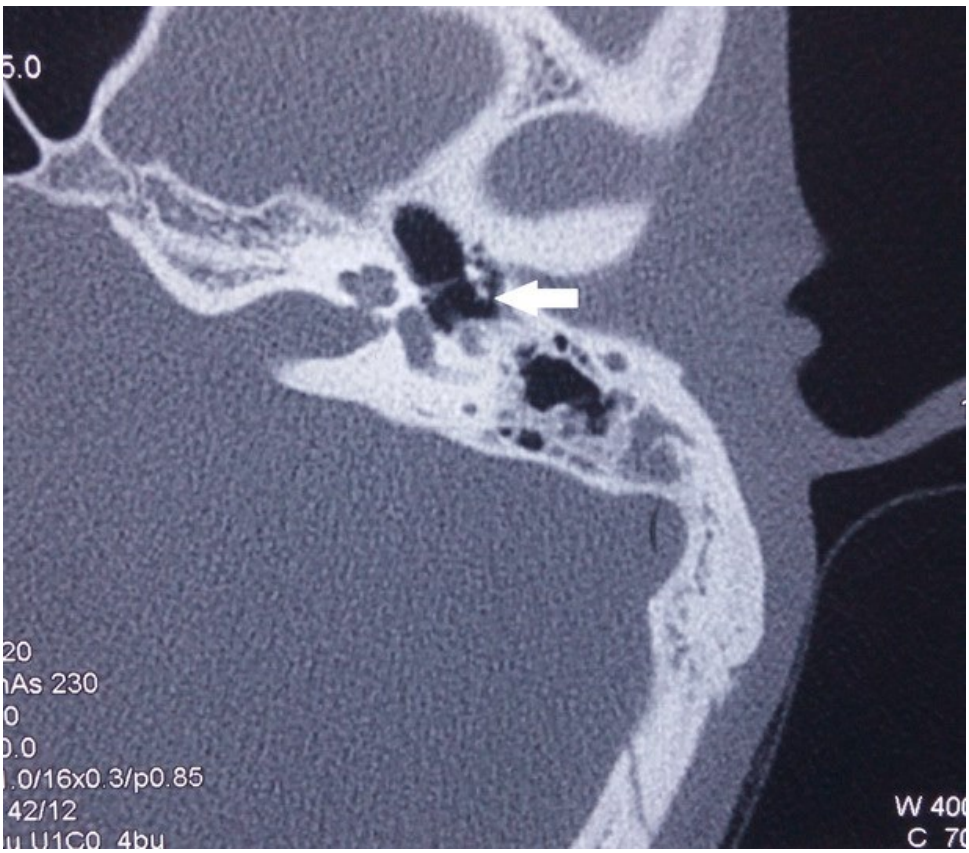

Fig 3: Normal middle ear cavity with intact ossicles

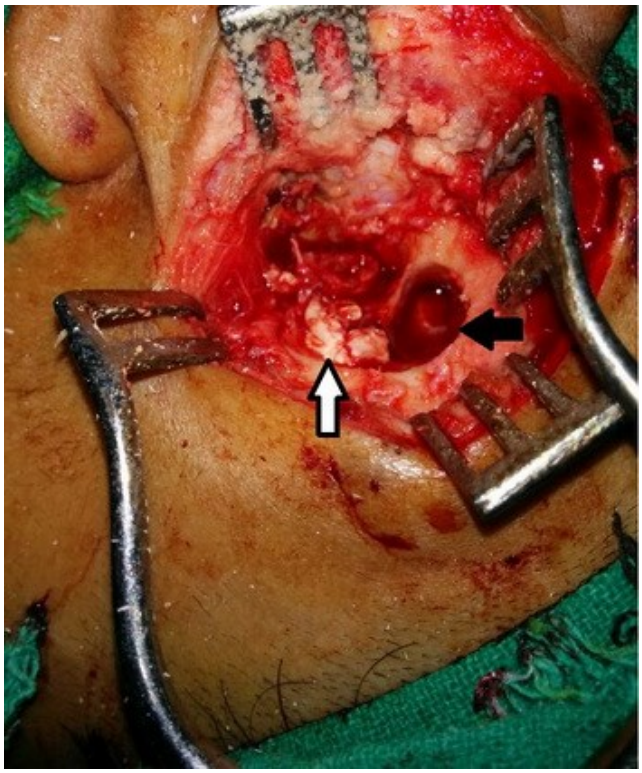

Fig 4: Cholesteatoma arising from mastoid tip and eroding it (white arrow) and normal antrum (black arrow)

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