Presenting a Case of A Congenital Cholesteatoma of the Mastoid in Adult

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INTRODUCTION

- Congenital cholesteatoma (CC) is a benign destructive epithelial cyst resulting from embryonic epithelial rests with an estimated incidence of 1-5% of all cases of all cholesteatomas[1].
- CCs can originate from the middle ear (most common), petrous apex, external canal, and other temporal bone regions[1,3,5].
- Unlike other types of CC, CC isolated to the mastoid are extremely rare.
- Due to their location within the mastoid and nonspecific symptoms, congenital cholesteatomas isolated to the mastoid may remain undetected for an extended period.

DIFFERENTIAL DIAGNOSIS

- Mastoiditis
- Encephalocele
  Mastoid Mucocele
- Glomus Tumors
- Vascular Diverticulum
  Other vascular lesions

CLINICAL IMAGES

Figure 1

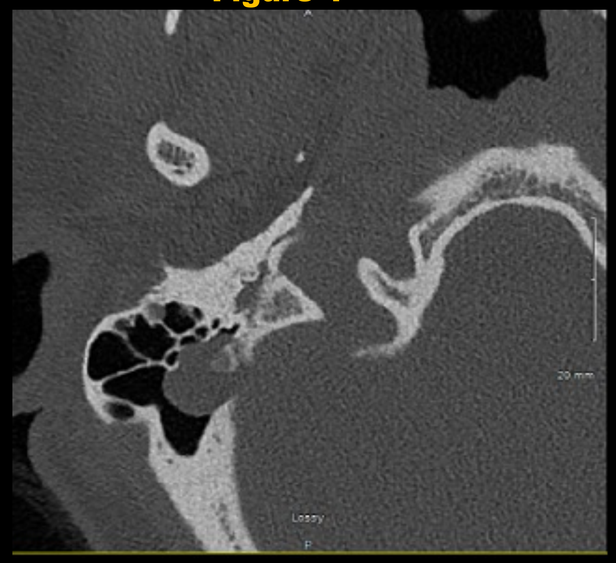

Figure 1: CT Temporal bone (right) axial view showing a soft-tissue mass within the right mastoid with erosion of the skull base into the posterior fossa.

Figure 2

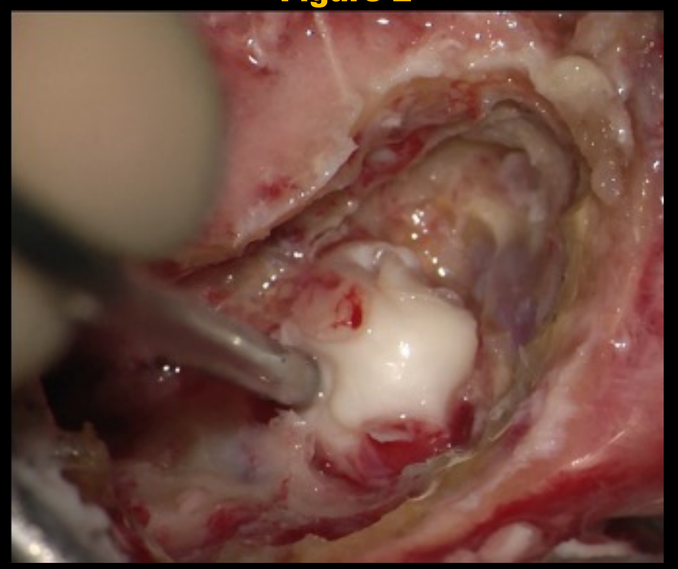

Figure 2: Intraoperative photo of CC isolated to the mastoid and associated skull base defect.

CASE REPORT

- 61-year-old female referred to our clinic for complaints of left sided “muffled hearing” with a previous history of chronic otitis media and a left sided tympanic membrane perforation.
- Audiometry confirmed LEFT moderate conductive hearing loss and normal hearing in the RIGHT ear.
- Imaging revealed a soft tissue mass with a defect of the posterior fossa was found within the RIGHT mastoid cavity abutting the sigmoid sinus (Figure 1).
- MRI demonstrated a well circumscribed, T1 isointense, T2 hyperintense, non-enhancing lesion. DWI findings were consistent with cholesteatoma.
- The patient underwent a right mastoidectomy with complete lesion resection. Intraoperatively, the patient had a well-aerated mastoid and was found to have a pearly white lesion within the mastoid cavity at the skull base.
- After complete lesion resection fistulization into the posterior fossa and exposed, yet intact, sigmoid sinus were identified . (Figure 2).
- The dura, jugular bulb and facial nerve and external auditory canal were uninvolved.
- The skull base defect was reconstructed following resection of the lesion.
- Pathology was consistent with cholesteatoma.
- The patient has been followed up periodically without evidence of recurrence.

DISCUSSION

- Congenital cholesteatoma isolated to the mastoid is the rarest subtype of all congenital cholesteatomas and, overall, represents an exceptionally rare finding.
- Its diagnosis requires a high index of suspicion, given its nonspecific presentation and the broad differential diagnosis.
- Our patient presented with a mastoid isolated CC with asymptomatic erosion of the skull base found incidentally.
- The lesion was removed and the skull base reconstructed without complications.

CONCLUSION

- Congenital cholesteatoma isolated to the mastoid is an extremely rare finding, and also represents the rarest form of all congenital cholesteatomas.
- The indolent course of combined with its typically late presentation and hidden location, presents significant challenges in both diagnosis and management.
- Optimal management involves surgical intervention with complete lesion resection, followed by regular follow-up to monitor for any recurrence or complications.

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