CONGENITAL CHOLESTEATOMA - CASE REPORT

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INTRODUCTION

Congenital cholesteatoma is a rare disease, thought to arise from embryonic remnants of squamous epithelial cells in the middle ear. Its typical appearance is a white mass “pearl” behind an intact tympanic membrane in children without history of otitis media or previous otologic procedures.

Although being a benign tumor, it may have expansive and destructive local growth with bone erosion, resulting in hearing loss and potentially fatal complications, emphasizing the need for an early diagnosis.

Computed tomography is needed to assist in preoperative planning. Therapeutic management is surgical removal. Despite adequate care, a 8-18% rate of residual disease and recurrence is described.

CASE REPORT

♂ , 3-year-old referred to our hospital for suspected hearing loss, associated with delayed speech and language development.

No history of recurrent otitis media or previous otologic procedures.

No relevant family history.

No parental suspicion of hearing loss.

Otoscopically, a whitish formation, with a rounded, pearl-like shape is visible behind the tympanic membrane at the anterosuperior quadrant

Hearing assessment

<table>
<thead>
<tr>
<th>Audiology</th>
<th>Tymanometry</th>
<th>Evoked otoacoustic emissions</th>
<th>Auditory brainstem response</th>
</tr>
</thead>
<tbody>
<tr>
<td>✖ non-cooperative</td>
<td>✖ type A bilaterally</td>
<td>✖ Present bilaterally</td>
<td>Normal</td>
</tr>
</tbody>
</table>

CT scan of the middle ear

“...rounded formation in the right middle ear, in contiguity with the tympanic membrane and with the anterior face of the manubrium mallei, with a maximum diameter of 4 mm...”

Surgical Management

Endaural approach followed by removal of an encapsulated lesion.

Anatomopathological report

“...shows exclusively keratin fragment, compatible with cholesteatoma.”

Follow-up

No complications in the postoperative period.

At six-month follow-up, normal postoperative hearing and no signs of disease recurrence.

CONCLUSION

Diagnosis of congenital cholesteatoma is frequently done in advanced stages, with concomitant ossicular destruction. Surgical excision of congenital cholesteatoma should be performed before permanent damage to the ear occurs. High awareness and suspicion is fundamental to early diagnosis and treatment in order to decrease the extent of the disease and its complications. Long-term follow-up is essential.

References


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