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# Congenital cholesteatoma
## (A case report)

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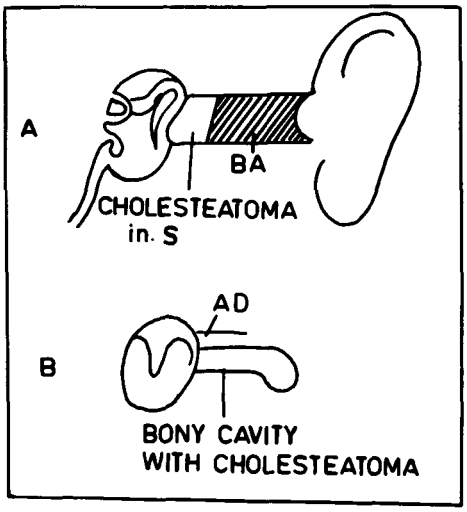
### Introduction

Aural cholesteatoma is usually associated with an attic or a postero-superior marginal perforation and accompanied by a history of malodorous discharge, often with hearing impairment. It may also present itself without any perforation in the tympanic membrane, otorrhoea or radiological evidence of bone destruction. This type of lesion is assumed to be of congenital origin, although this is difficult to prove (House and Sheehy, 1980). Cushing (1922) first speculated on the possibility of a congenital origin of aural cholesteatoma, but the first case of congenital cholesteatoma of the middle ear behind an intact tympanic membrane was published by House in 1953. Since then 47 patients with congenital middle ear cholesteatoma have been described in the literature of which 12 were seen in children (Schwartz *et al.*, 1983). We report here another case of congenital cholesteatoma in a 9-year-old male child who presented with recurrent meningitis.

### Case report

A nine-year-old boy attended the ENT OPD with complaints of deafness and slight pain in the left ear. Examination revealed compete atresia of the left external auditory meatus with microtia. The right ear, nose and pharynx were normal. The Rinne was negative on the left side and positive on the right side, with lateralization of the Weber to the left side. There was no other congenital abnormality in the body.

Audiometry showed a conductive deafness with an A-B gap of about 60 dB. X-rays of the mastoids showed hypocellularity on the left side. There was a significant history of repeated upper respiratory tract infections though, at the time of examination, the nose and throat were essentially normal. The patient was submitted to surgery under GA. Through a post-auricular incision, the mastoid antrum was reached without difficulty. The external auditory canal was found to have a bony atresia which was drilled down under the operating microscope. The canal was widened to about double the normal size. There was a small space between the tympanic membrane and the atretic plate, and a huge cholesteatomatous mass found in this space (Fig. 1), which extended posteriorly below the level of the aditus ad antrum. The cholesteatomatous cavity did not have any communication with the middle ear. The mass was sent for histopathology and this showed cholesteatoma. The middle ear was entered, working anteriorly from the antrum, along the aditus. The tympanic membrane was reflected anteriorly from the sulcus. The incus was found to be normal except for the fact that its lenticular process was smaller than normal; the stapes was represented only by its footplate, the crura and head being absent. The footplate was found to be fixed. A small hole was made in the centre of the footplate with straight and right-angled picks and there was some oozing of perilymph. A teflon piston was looped around the long process of the incus and retained with a fat graft taken from the lobule. The chorda tympani nerve was not seen. The canal wall was lined with a free skin graft taken from the post-auricular region and, after placing some medicated gelfoam pieces in the external auditory canal and on to the tympanic membrane, the canal was packed with a medicated tampon as after stapedectomy. The post-auricular wound was closed after putting a drain into the antrum. Ten days after the operation, the packing was removed and the hearing improved, post-operative audiometry showing a residual conductive hearing loss of about 30 dB. The patient was advised to keep a mould in the ear for six months, and to have it changed and cleaned every day. The




BA – BONY ATRESIA
S – SPACE BETWEEN BA AND TYMPANIC MEMBRANE
AD – ADITUS

FIG. 1

Diagrammatic representation of the cholesteatoma cavity, in the coronal plane and in the lateral view.
(A) Shows the cholesteatoma cavity.
(B) Bony cavity with cholesteatoma extending laterally and posteriorly below the aditus.

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mould was made of dental stent with a hole in the centre, thereby minimizing the chance of stagnation in the ear canal while the mould was *in situ*. The canal was patent and improved hearing was maintained at subsequent follow-up visits for about six months.

## Discussion

Congenital aural cholesteatoma is a rare pseudo-tumour seen in patients without any evidence of middle ear disease. It may be located within the temporal bone, in the region of the petrous apex, in the middle ear cleft or enclosed within the lawyers of the tympanic membrane. In a review of literature, Curtis (1979) found only ten cases of congenital cholesteatoma in children.

Schwartz *et al.* (1983) described two other cases in infants aged five weeks and three months, the youngest recorded in the literature. Three cases of bilateral cholesteatoma (congenital) have also been described, one of which was seen in a two-year-old child (Curtis, 1979; Wingert *et al.*, 1976).

In our own case the cholesteatomatous cavity did not show any communication with the middle ear cleft. Since it was located in the space between the atretic plate and the tympanic membrane, it is conceivable that it developed from the epidermoid tissue present there. Perhaps a CT Scan would have shown the cholesteatoma cavity. Hitherto, to our knowledge, a cholesteatomatous lesion in this location has not been mentioned in the literature. It therefore appears that it is not the sequestration variety but rather a retention one, i.e. it must have developed from the retained epidermal tissue caught in the pocket.

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