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Congenital stapes footplate fixation associated with duplication of lobule

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Abstract
We report the case of an 18-year-old male patient who presented to us with unilateral conductive hearing loss with duplication of the lobule. An exploratory tympanotomy revealed stapes footplate fixation. A stapedotomy with insertion of a Teflon piston was performed with improvement in his hearing. We believe this is the first reported case of such an abnormality.

Key words: Stapes; External Ear; Surgery; Hearing Loss, Conductive

Introduction

Congenital anomalies of the middle ear are rare, sporadic and mostly non-familial. However these anomalies may present as a part of a syndrome such as Pfeiffer's, brancho-oto-renal, cervico-oculoacoustic, proximal symphalangia, Kleippel-Feil, frontometaphyseal dysplasia, or first and second branchial arch syndromes.[1,2]

Duplication of a part or whole of the pinna is also a rare anomaly, with a few cases reported in the literature.[3] Again, such lesions may be sporadic or may arise as a part of a congenital syndrome. In our search of the medical literature, we did not find any case report of a congenital footplate fixation associated with duplication of the lobule, and the extreme rarity of such a case has prompted us to present this report.

Case report

An 18-year-old male patient presented to us with a non-progressive hearing loss and a mass behind the right ear. The mass had been present since birth and the hearing loss had been noticeable since childhood. The patient gave a history of two to three episodes of otalgia and otorrhoea involving both ears in childhood: in each incidence this responded to medication. There was no history of any otological abnormalities in the family.

On examination, a well-formed duplicated lobule was present in the right post-aural region (Figures 1 and 2). The tympanic membranes were normal in appearance but tuning fork tests revealed a conductive hearing loss in the right ear and a pure tone audiogram revealed a conductive loss in the right ear with a mean air-bone gap of 45 dB in the three frequencies 256 Hz, 512 Hz and 1024 Hz. Impedence audiometry revealed reduced compliance and normal middle-ear pressure (Figure 3), and high-resolution computed tomography (CT) showed no ossicular, facial nerve or inner-ear anomalies.

An exploratory tympanotomy was performed under local anaesthesia, and the only abnormality found was an absence of the round window reflex, suggesting the diagnosis of congenital stapes footplate fixation. No other anomaly of the middle-ear conductive apparatus, such as ossicular dislocation, ossicular necrosis or tympanosclerosis, was identifiable. The stapes footplate appeared to be blended with the surrounding otic capsule and no discrete focus of otosclerosis was identifiable. A small fenestra stapedotomy with insertion of a 0.4 mm piston was performed. The procedure was uneventful and the patient reported an immediate gain in hearing. Since a small fenestra stapedotomy was performed, the footplate could not be evaluated histopathologically. Three months post-operatively, the air-bone gap was recorded to be 10 dB (i.e. a 35 dB gain).

Discussion

Congenital ossicular anomalies are very rare, with an incidence of less than one per 15 000 births.[4] In most cases, patients presenting with a non-progressive hearing loss with no history suggestive of recurrent ear infections show no abnormality on otoscopy. Our patient presented in a similar manner.

Most investigations, including an audiogram and impedence audiometry, are usually non-contributory in reaching an exact diagnosis.[5] A high-resolution CT scan may demonstrate anomalies such as bony bars in the middle ear, lateral malleolar fixation, absent or disconnected ossicles (as well as congenital cholesteatoma and vascular lesions), but the yield from such an investigation is normally low.[5] In our patient the scan revealed no abnormality, although impedence audiometry showed reduced compliance, suggesting the possibility of an ossicular fixation. The diagnosis was finally confirmed on an exploratory tympanotomy. The lack of progression of hearing loss, absence of any other middle-ear pathology, lack of a round window reflex, absence of any obvious otosclerotic focus, and a marked immediate hearing improvement with stapedotomy, point towards a congenital fixation of the stapes footplate.

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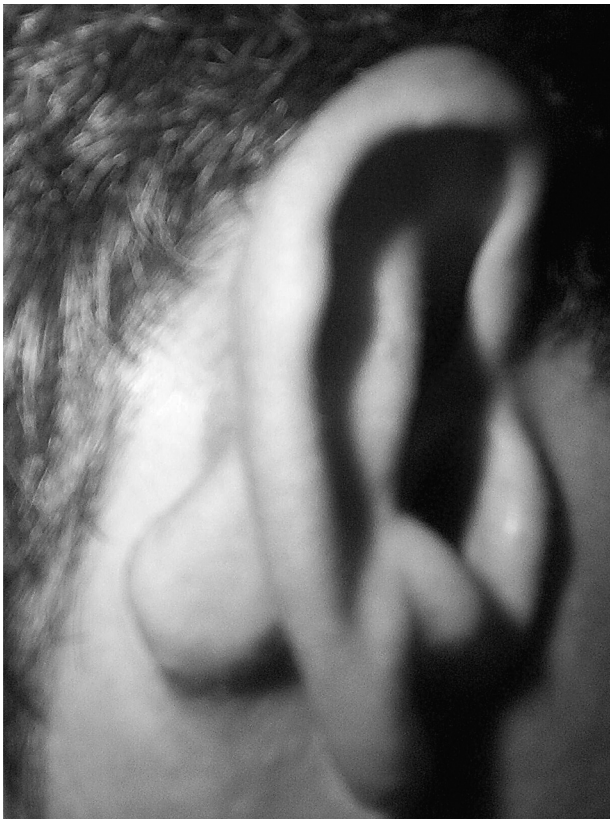

FIG. 1
Duplication of pinna on the right side.

Major and minor congenital anomalies of the ear may occur.[6] Major anomalies involve the abnormalities of the pinna and external auditory canal, as well as the tympanic cavity, and include various degrees of microtia, anotia and atresia. Minor anomalies involve fixation and defects of the ossicular chain and anomalies of the oval and round windows. Our case does not fall into any of these classes and we can find no such anomaly of the external ear associated with an ossicular abnormality in a literature search on Medline and PubMed. The key words used for the search were 'stapes', 'external ear', 'stapedectomy' and 'hearing loss'.

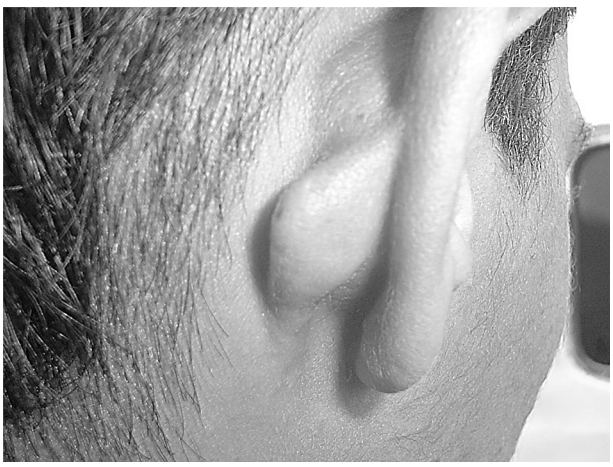

FIG. 2
Duplication of pinna on the right side.

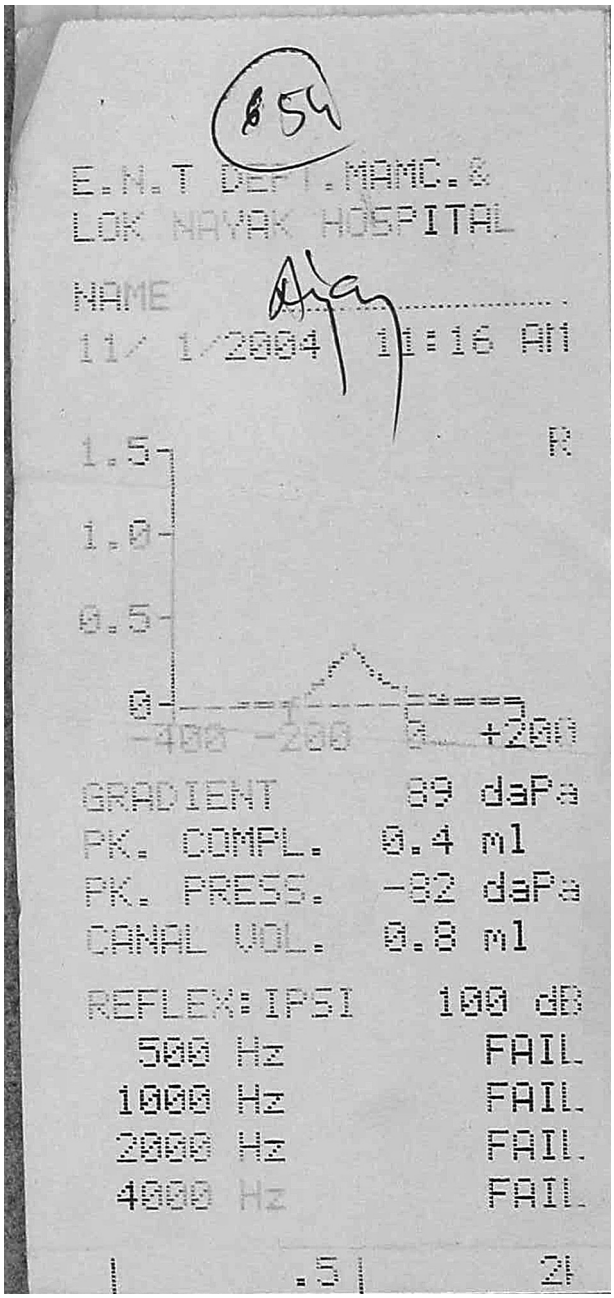

FIG. 3
Reduced compliance on the tympanogram.

Here we briefly discuss the possible embryological cause for the particular abnormality in our patient. The stapes has two developmental origins. The suprastructure and the tympanic part of the footplate develop from the second branchial arch, whereas the vestibular surface of the footplate is derived from the otic capsule.[6] Differentiation and condensation of a part of the cartilagenous otic capsule gives rise to lamina stapedialis.[7] The secondary differentiation of this laminar tissue is responsible for the formation of the annular ligament that enables the stapes to become a separate mobile structure. Thus, the failure of the lamina stapedialis to differentiate and separate from the otic capsule is responsible for the fixation of footplate. During the third week of fetal life, six hillocks develop from the first and second branchial arches (three anterior hillocks from the first and three posterior

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hillocks from the second arch).[8] The lobule is derived from the sixth hillock (second arch). Failure of fusion of these hillocks has been blamed for the development of polyotia. However, the exact point of fault in embryological development responsible for duplication of a part or whole of the pinna is not known. Since the embryological basis of congenital fixation of the stapes footplate and duplication of pinna are different, the occurrence of both conditions in the same patient cannot be explained on the basis of a single fault in the process of embryological development, and this is indeed an extremely rare occurrence.

- **Congenital anomalies of the middle ear are rare and may be familial or non-familial**
- **Duplication of a part or whole of the pinna is a rare anomaly**
- **The embryological basis for the development of congenital stapes footplate fixation and duplication of lobule appear to be different**
- **The different developmental basis for the two rare anomalies makes it extremely unusual for our patient to present with both these anomalies together**
- **Our patient responded well to a small fenestra stapedotomy, with an immediate improvement in hearing**

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Dr A Sethi takes responsibility for the integrity of the content of the paper.
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