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Clinical Records

Recurrent meningitis due to a congenital fistula of the stapedial footplate

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Abstract

A rare case of a congenital fistula of the stapes footplate, in a 10-year-old girl, resulting in recurrent meningitis, is reported. A full ENT examination and a high index of suspicion, is essential if the diagnosis is to be made.

Exploratory tympanotomy should be considered on clinical grounds, even if a high resolution CT scan of the temporal bones does not show any anomalies, in order to ascertain the definite diagnosis of a fistula and to seal it permanently.

Key words: Stapes; Fistula, congenital; Meningitis

Introduction

Despite modern antibacterial agents, meningitis remains a serious and potentially life-threatening condition. Recurrent meningitis in children often involves repeated hospital admissions for treatment and investigations, in order to find the underlying cause. This in turn has long psychological and emotional sequelae for both parents and child.

Congenital fistula of the stapes footplate is a rare, but well documented cause of recurrent meningitis. However, the diagnosis of such fistulae is usually difficult and often delayed. A close liaison between the paediatrician, the ENT surgeon and neuroradiologist, is important to establish the definite diagnosis and subsequent management.

Case report

A 10-year-old girl was referred for an ENT opinion, after having three episodes of meningitis. She had been known to be profoundly deaf since her first episode of meningitis at the age of seven months and had been subsequently wearing binaural hearing aids. The second episode was at the age of nine years and was preceded by left-sided otitis media, as was her third attack at the age of 10 years. On each occasion *Streptococcus pneumoniae* meningitis was treated successfully with the appropriate antibiotics.

A full ENT examination, two weeks after the third episode, showed a rather dull-looking left tympanic membrane. A pure tone audiogram revealed bilateral severe sensorineural hearing loss (Figure 1). Tympanometry showed normal compliance on the right, and low compliance on the left. A CT scan of the temporal bones did not show any abnormalities.

In view of her history, it was decided to explore the left ear to exclude a fistula between the inner and the middle ears.

A left tympanotomy, under general anaesthesia, revealed fine granulation tissue around the stapes footplate. After careful dissection of this granulation tissue, cerebrospinal fluid (CSF) was found to be leaking through a large defect in the centre of the

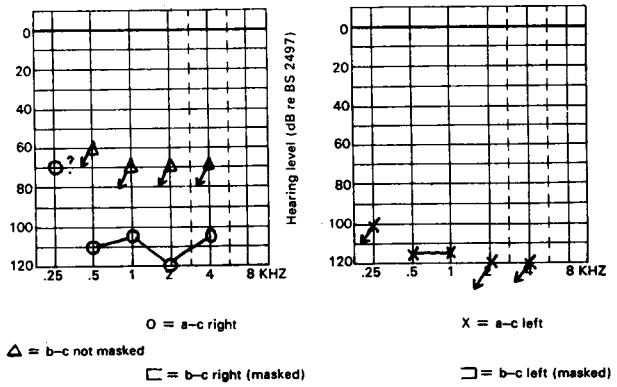

FIG. 1
Audiogram showing bilateral severe sensorineural loss.

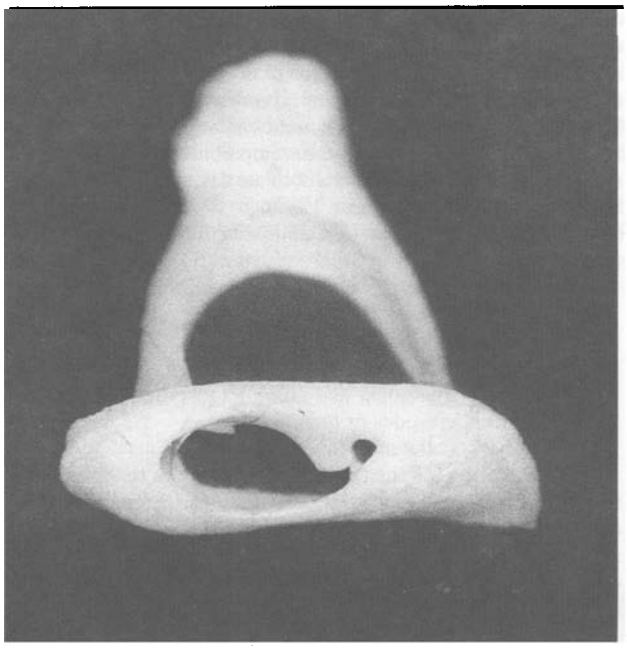

FIG. 2
Stapes showing a large defect in the footplate.

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stapes footplate (Figure 2). The stapes was completely removed, the vestibule was plugged with temporalis fascia and muscle. The middle ear was also packed with temporalis muscle. Prophylactic penicillin-V was given post-operatively for two weeks. She made an uneventful recovery and was discharged home five days after her operation.

Four months later, when she was reviewed in the outpatient clinic, she was doing well and had had no further episodes of meningitis. An offer of right tympanotomy, to exclude similar pathology affecting the right ear, was turned down by the parents.

Discussion

Cerebrospinal fluid (CSF) otorrhoea is due to pathological communication between the subarachnoid space and the tympanic cavity. The common causes are fractures involving the temporal bone, surgical trauma, neoplasms and infections of the temporal bone. A rare cause of CSF otorrhoea is congenital abnormality with defects of both the inner and the middle ears. This was first described by Escat in 1897.

The majority of CSF leaks in previously reported cases, as in our case, occurred through defects of the stapes footplate (Quiney *et al.*, 1989), the annulus of the stapes footplate (Skolnik and Ferrer, 1959) or the round window (Bauer, 1962). Other rare sites were also reported: the promontory (Nenzelius, 1951), the roof of the eustachian tube (Stool *et al.*, 1967) and the tegmen of the antrum (Kaufman *et al.*, 1977).

The site of communication between the subarachnoid space and perilymphatic spaces of the inner ear is controversial. Some authors believe it occurs through defects of the lamina cribrosa at the fundus of the internal auditory meatus (Harrington and Birck, 1967; McNab-Jones and Fairburn, 1977). An abnormally wide and patent cochlear aqueduct has also been proposed as a possible site of communication (Schuknecht, 1974). Congenital CSF fistula has been reported in some cases in association with other congenital anomalies; Mondini dysplasia of the labyrinth (Hicks *et al.*, 1980; Guindi, 1981), Klippel–Feil syndrome (Richards and Gibbin, 1977).

The pathogenesis of the defect in the stapes footplate is unknown. There are two theories, one of which suggests that the defect is a developmental abnormality of the stapes footplate (Hipskind *et al.*, 1976), and the second one postulates that the defect is due to mechanical erosion of the footplate secondary to raised intralabyrinthine pressure (Gunderson and Haye, 1970; Parisier and Birken, 1976). The association of the fistula with Mondini dysplasia favours the developmental concept, as it is now accepted that the stapedial footplate develops at least in part from the otic capsule (Wright, 1987).

The diagnosis of congenital CSF fistula of the inner ear is difficult, as it may present in different ways:

(1) CSF rhinorrhoea if the tympanic membrane is intact and CSF passes down the eustachian tube.
(2) Middle ear effusion.
(3) CSF otorrhoea if the tympanic membrane is perforated, or during myringotomy and grommet insertion.
(4) Recurrent episodes of meningitis.
(5) Deafness, unilateral or bilateral.

High resolution CT scan of the temporal bones to identify the site of the fistula and any associated inner ear anomalies, is the main investigation carried out (Phelps, 1986). However, normal scan findings, as in the present case, do not exclude the presence of fistula. A high index of clinical suspicion is of paramount importance for diagnosis and subsequent treatment.

Exploratory tympanotomy is the treatment of choice. Infiltration of the skin of the ear canal with adrenalin should be avoided, as it could be mistaken for CSF if it tracks into the middle ear cavity. The oval and round windows should be carefully and clearly seen under high magnification. Any mucosal strands should be divided to view the whole footplate of the stapes. If a CSF leak is found at the oval window, the stapes should be removed, provided the ear is dead, and the vestibule plugged and packed firmly with temporalis fasica and muscle. The middle ear should also be packed with the same muscle to provide a second barrier.

Post-operatively, the patient should be nursed in the semirecumbent position, should be given prophylactic antibiotics and advised against any straining, coughing, sneezing or nose blowing.

We agree with Phillipps (1986) that bilateral tympanotomy should be considered in cases with severe bilateral deafness to exclude the possibility of bilateral CSF fistulae.

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