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Clinical Records

Spontaneous cerebrospinal fluid otorrhoea via oval window: an obscure cause of recurrent meningitis

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Abstract
Spontaneous cerebrospinal fluid (CSF) leak via the oval window is uncommon and can result in recurrent bacterial meningitis. Current understanding of spontaneous CSF otorrhoea is reviewed and a diagnostic algorithm is presented.

A seven-year-old boy presented with bilateral congenital deafness and recurrent meningitis. High-resolution computed tomography (HRCT) of the temporal bone showed a labyrinthine deformity and communication between the internal auditory canal (IAC) and the cochlea. Subtotal petrosectomy with closure of the external acoustic meatus and eustachian tube was performed. Post-operatively, the child had no further episodes of meningitis.

This rare and obscure cause of recurrent childhood meningitis requires a high index of suspicion and the use of diagnostic tools, especially HRCT.

Key words: Deafness; Meningitis; Cerebrospinal Fluid Otorrhoea; Petrous Bone, Surgery

Introduction

Bacterial meningitis in the paediatric age group is not uncommon, even in the present antibiotic era. It is known to be associated with significant mortality; the risk of death ranges between 6.3 and 25 per cent, as reported by Thomas[1] and Paul and Alan.[2] Among the documented associated morbidities which may arise are neurological and functional impairment such as sensorineural hearing impairment (8.3 per cent), recurrent seizures (2.8 per cent), motor (7 per cent), blindness (2.8 per cent), obstructive hydrocephalus (2.8 per cent), hyperactivity (4.2 per cent), speech delay (7 per cent), learning difficulties (12.7 per cent), and mental retardation.[1,2]

Management of bacterial meningitis includes aggressive broad-spectrum antibiotics which are effective against common organisms such as *Pneumococcus*, *Haemophilus*, *Escherichia* and *Meningococcus*.[2] It is equally important that the underlying cause be identified, for example, prior neurosurgical interventions, otitis media, sinusitis or birth abnormalities.

In a small proportion of those patients who present with more than one episode of bacterial meningitis, anatomical abnormality or functional impairment could be the culprit. The diagnostician should be aware and vigilant about unusual functional causes such as humoral or complement immunodeficiency syndromes, hypoimmunoglobulinopathies, complement component deficiencies, and asplenia conditions.[3–5] At the same time, anatomical defects which result in ascending infection from the paranasal sinuses or middle ear[5,6] must be excluded in the diagnostic evaluation. The latter cause is more probable if the patient has concomitant deafness, as this signifies a possible structural abnormality in the middle or inner ear. Once the ear is suspected as the primary source of infection, radiological investigations in the form of high-resolution computed tomography (HRCT) of the temporal bone remain the best diagnostic tool.

Spontaneous cerebrospinal fluid (CSF) leak via the oval window is an obscure cause of recurrent bacterial meningitis. As such it is a diagnostic challenge to the clinician, and a high index of suspicion is mandatory. Successful management hinges on a strong clinical suspicion, an accurate diagnostic evaluation and appropriate surgical intervention. Current understanding of spontaneous CSF otorrhoea is being reviewed, and a diagnostic algorithm is presented below. We illustrate this with a case presentation of a child with bilateral hearing impairment and a history of four episodes of meningitis.

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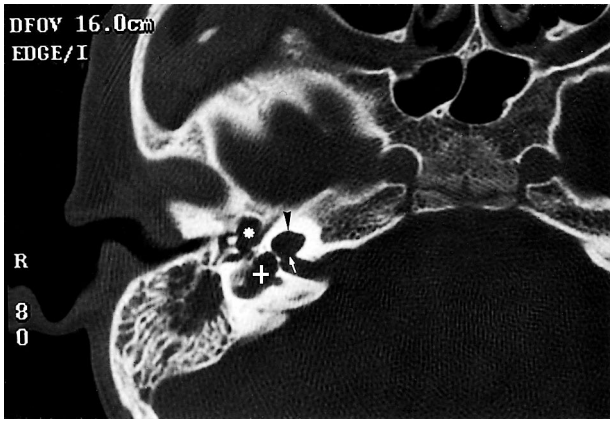

FIG. 1

Axial high-resolution computer tomographic scan of the temporal bone shows the dilated right cochlea (black arrowhead) due to the lack of septation between the apical and middle turns. The vestibule is also dilated (white cross). The white arrow points to the enlarged canal of the cochlear nerve and the absence of the modiolus of the cochlea. Note the opacification of the middle ear cavity (white asterisk).

Case report

Presentation

The patient was a seven-year-old Malaysian boy who was diagnosed with bilateral sensorineural hearing loss at the age of three years. He had previous myringotomies with ventilation tube insertions for suspected serous otitis media.

His first episode of meningitis occurred when he was five years of age. Lumbar puncture was performed and *Pneumococcus* was evidently cultured from his CSF. He was successfully treated and discharged. He went on to have two further episodes of pneumococcal meningitis that year. In the second episode, he presented with meningism and had a positive blood culture of *Pneumococcus*, although the CSF was sterile. In the third episode, he again presented with meningism and CSF smears revealed *Pneumococcus*. This was followed by a fourth episode the following year, when he presented with meningism. However, both blood and CSF cultures were negative. Neutrophils were seen in the CSF but no organism was isolated.

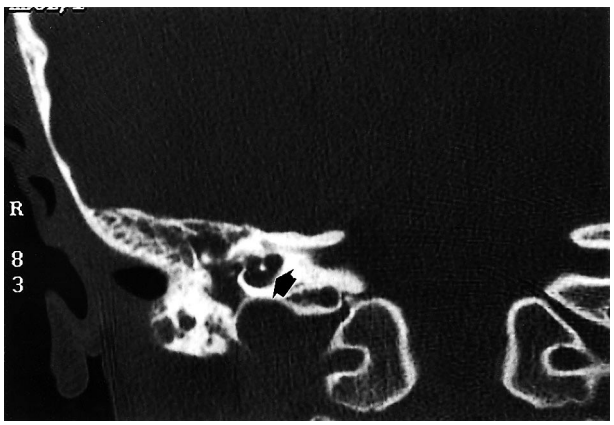

FIG. 2

Coronal high-resolution computed tomograph of the temporal bone. The broad arrow points to the dilated basal turn of the cochlea.

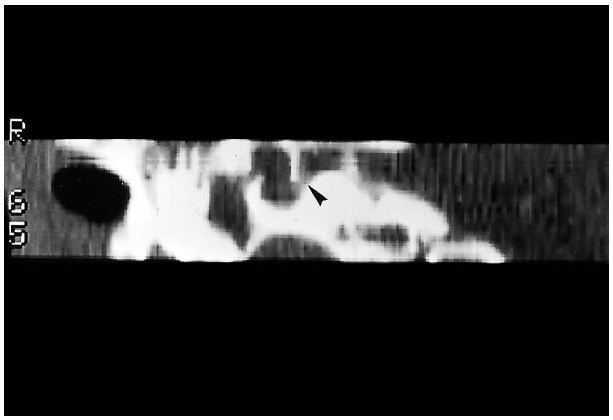

FIG. 3

Oblique coronal reconstruction of the right temporal bone along the plane of the canal of the cochlear nerve. This reconstructed image best demonstrates the wide communication between the internal auditory canal and the cochlea (arrowhead).

The patient was treated with ceftriaxone, a third-generation cephalosporin, and responded well to treatment during all episodes.

A full immune work-up was conducted to exclude immune deficiencies. Investigations included tests for T-cell and B-cell function, complement levels (namely, C3 and C4), and serum immunoglobulin (Ig) levels (Ig G, Ig M, Ig A and Ig E). In all these investigations, the results were normal. A CT scan of the brain incidentally revealed an opacification in the right mastoid air cells. In view of this abnormal radiological finding, an ENT opinion was sought.

Clinical findings and investigations

General examination showed a thriving child developed appropriately for his age, with no dysmorphic features. Otoscopy revealed a bulging right tympanic membrane. The rest of the examination was unremarkable.

The audiogram revealed bilateral profound hearing loss of 90 dB., with a type B tympanogram in the right ear.

High-resolution computed tomography of the temporal bone was done with a 1-mm thick contiguous scan, providing axial and coronal planes with oblique coronal reconstruction. Bone algorithm, scanning from the level of the internal auditory canal (IAC) to the round window in the axial plane and from the level of the head of malleus to the round window in the coronal plane, without contrast, was performed.

This showed incomplete septation between the apical and middle turns of the cochlea, with dilatation of the basal turn in both ears. The vestibules on both sides were also dilated, with a short lateral semicircular canal (Figure 1). A wide communication between the IAC and the cochlea was demonstrated on the right side due to an enlarged canal for the cochlear nerve (Figures 2 and 3). The modiolus was absent, resulting in no bony partition between the IAC and the cochlea. The middle ear cavity and the mastoid air cells on the right side were opacified. The canal of the cochlear nerve was also slightly widened on the left side and the modiolus was present, although it was dysplastic. A thin bony partition could still be appreciated separating the IAC from the cochlea. The middle ear cavity and mastoid air cells on the left side were clear of any fluid or debris.

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Based on the imaging findings, an abnormal middle ear–perilymph–CSF communication on the right side was suspected.

Treatment

The middle ear–perilymph–CSF communication was confirmed during exploratory tympanotomy of the right ear, during which CSF from the superior region of the annulus of the oval window was noted. The defect was plugged with muscle harvested from the temporalis muscle. A right subtotal petrosectomy with blind sac closure of the external acoustic meatus and closure of the eustachian tube was performed.

Discussion

When a young patient presents with recurrent meningitis, a fistulous communication between the CSF space and the ear must be sought. In the presence of long-standing deafness, a congenital CSF fistula of the petrous temporal bone is the most likely site of communication.

Recurrent meningitis with deafness is not an unreported entity. Parisier and Birken[6] reported that meningitis was the presenting complaint in 14 out of 15 patients with CSF otorrhoea. These authors believed that initial otitis media spread to involve the labyrinth before extending upwards intracranially, presenting with meningism. Noticeably, these patients did not have vertigo due to a non-functioning vestibular system.[6]

Pneumococcus is the commonest organism isolated in most studies, although the Australian experience showed *Haemophilus influenza* type B to be the case in 75 per cent of cases.[1,2,5,6]

Potential sites of cerebrospinal leak in the inner ear include the oval window, stapes footplate, eustachian tube and promontory fistula.[5–10] The leaks are mostly due to congenital temporal bone dysplasia with fistulous connections between the subarachnoid space and the middle ear cavity.

Congenital CSF fistulae of the petrous temporal bone can be of two types: perilabyrinthine and translabyrinthine. In perilabyrinthine fistulae, the communication could be through a defect in the tegmen tympani via the Hyrtl's fissure, via the petromastoid canal or via the first part of the facial nerve canal. Patients with perilabyrinthine fistulae usually do not present with congenital deafness, although labyrinthitis ossificans, a consequence of meningitis, can subsequently lead to deafness.

In contrast, patients with translabyrinthine fistulae usually have congenital deafness. Imaging of these patients usually reveals severe labyrinthine deformity, particularly, cystic deformity of the cochlea and vestibule.[5]

Leakage of the CSF commonly occurs through the oval window. Thirteen out of 15 cases in Parisier and Birken's review and five out of seven cases in the review of Quiney *et al*. had leaks via the oval window.[5,6] Leaks from the round window or through defects in the promontory are uncommon. The CSF leakage through the oval window could be due to an associated faulty development of the stapes, such as a hole in the footplate, or due to an increase in the vestibular perilymph pressure which displaces the footplate, leading to defects in the annulus.[6,8] The latter was the case in our patient.

High-resolution computed tomography of the temporal bone revealed an enlarged canal of the cochlear nerve and absent modiolus on the right side (Figures 1–3). This allowed the CSF pressure, which is normally higher than the perilymphatic pressure, to be transmitted to the fluid within the cochlea. There was also an abnormal, wide communication between the basal turn of the cochlea and the vestibule. This in turn allowed the high pressure to be transmitted from the cochlea to the vestibular perilymph, resulting in a leakage around the stapes footplate. The increased vestibular perilymphatic pressure in our patient was clearly shown by the perilymph gusher experienced during the stapedectomy.

Phelps and King showed that the degree of hearing loss depends on the presence of the basal turn and the calibre of the cochlear.[11] Hence, some patients may have normal auditory and vestibular function, while the more severe cases, like our patient, have profound hearing loss and a non-functioning vestibular system. Cochlear dysplasia may also occur as part of a syndrome in trisomies, Klippel Feil, Pendred or Di George.[5,12]

Interestingly, our patient had bilateral profound deafness but only the right cochlear was seen to have no bony partition with the IAC, hence providing a portal of entry. On the left side, the dysplastic modiolus provided a thin bony partition separating the IAC from the cochlea.

- **This case study reviews the literature on recurrent meningitis as a result of congenital temporal bone abnormalities**
- **The use of high-resolution computed tomography is recommended**

In this case, radiology provided us with the most vital pre-operative clue by successfully identifying the source of the CSF leak, thereby enabling more precise surgery to be performed. Prior to the advent of HRCT, surgery was less focused and, as a result, patients frequently required repeated surgery to stop the leak. Intra-operatively, the surgeon has to decide where the leak is coming from, which is difficult because of the welling of CSF in the small middle ear cavity. As a result, multiple surgical procedures have previously been required for CSF otorrhoea.[6] Thirty-six surgical procedures were performed on 15 patients in Parisier and Birken's study, a testimony to the difficulty of localizing the leak.[6]

In retrospect, our patient probably had an ongoing CSF leak into the middle ear, causing a bulging tympanic membrane. The unsuspecting surgeon performed a myringotomy and tube insertion for what was thought to be a simple case of serous otitis media. Hence, the condition went undiagnosed until the patient presented with meningitis.

Quiney *et al*. offered five reasons for the frequent misdiagnosis of CSF fistula secondary to inner ear congenital abnormalities:[5]

(1) CSF rhinorrhoea may be difficult to differentiate from normal nasal discharge.
(2) Middle ear effusion of CSF may not be common because of leakage down the Eustachian tube. Persistent otorrhoea after myringotomy may be treated as otitis media.
(3) CSF flowing down the Eustachian tube may be swallowed rather than present as rhinorrhoea.
(4) Children presenting with meningitis may be investigated with CT scans of the head, but not of sufficient resolution or cut size to demonstrate abnormalities of the vestibulo-cochlear system.
(5) Unilateral deafness in a child may be difficult to diagnose.

Hence, there must be a high index of suspicion for CSF otorrhoea in patients with persistent rhinorrhoea and otorrhoea presenting with meningitis. Bulging tympanic membrane in the presence of meningitis should alert one to

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possible CSF otorrhoea. The fluid should be sent for biochemistry to determine whether it is CSF. When either of the above is present or if the patient has a history of hearing impairment, an HRCT of the temporal bones should be performed with the proper bone settings to look for vestibulo-cochlear abnormalities. This also helps identify the cochlear of interest in the event of bilateral cochlear dysplasia, as in our patient. Phelps and King recommended surgery for any patient with cochlear dysplasia presenting with their first episode of meningitis.[11,13]

In some countries, having an effective screening programme for childhood deafness could identify children with cochlear dysplasia or even CSF otorrhoea. Recommendations could be made regarding injury avoidance and risks in contact sports.[13]

At present, the onus is on the clinician to identify those patients who may benefit from surgery, to prevent subsequent episodes of meningitis and the associated long-term morbidity.

Conclusion

This case highlights an obscure cause of recurrent childhood meningitis. A high index of clinical suspicion and appropriate use of diagnostic tools, especially HRCT, can contribute significantly to the establishment of this diagnosis.

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