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Recurrent *Streptococcus pneumoniae* meningitis and Mondini dysplasia: Association or causation?

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ABSTRACT

Mondini dysplasia is a developmental disorder of the inner ear structures and it is a rare cause of recurrent bacterial meningitis in children. A 10-year-old boy presented with acute febrile encephalopathy and right ear pain. In the past, he had suffered from two distinct episodes of pyogenic meningitis. On examination, he had signs of meningeal irritation and right ear sensorineural deafness. Magnetic resonance imaging of the brain and computerized tomography of the temporal bone was suggestive of Mondini dysplasia in the right ear. Our case highlights the need for (a) screening of hearing loss at the bedside by Rinne and Weber test in case of recurrent bacterial meningitis (b) searching for an underlying inner ear malformation if there is a hearing loss.

Introduction

Recurrent bacterial meningitis is a rare entity in children with an estimated incidence of 1.3% amongst all cases of bacterial meningitis [1]. Mondini dysplasia, a congenital disorder of the inner ear structures, is a surgically amenable cause of recurrent meningitis in children and warrants a high index of suspicion [2]. The clinical hallmarks of Mondini dysplasia are hearing impairment and cerebrospinal fluid (CSF) leaks [3]. We discuss a 10-year-old boy with recurrent bacterial meningitis, unilateral profound sensorineural hearing loss and ipsilateral Mondini dysplasia. Our case highlights the importance of detailed systemic evaluation in children with recurrent meningitis including the otorhinological examination. Screening for hearing loss should be done routinely in such cases at the bedside by Rinne and Weber test and one should search for an underlying inner ear malformation if there is a hearing loss. Pre-existing hearing loss is an important clinical clue in these patients.

Case

A 10-year-old boy developed acute-onset altered sensorium, headache and vomiting following high grade fever and right ear pain for the past 2 days. There were no associated seizures, focal motor deficits, trauma, rash, persistent nasal or ear discharge. In the past, he had suffered from two distinct episodes of pyogenic meningitis. The first episode was at 9 years of age when CSF analysis showed 72 cells/μL (30% polymorphonuclear leukocytes and 70% lymphocytes), glucose 46 mg/dL, proteins 180 mg/dL and absence of organism on microscopy and culture. The second episode occurred at 9.5 years of age when CSF analysis showed 160 cells/μL (65% polymorphonuclear leukocytes and 38% lymphocytes), glucose 46 mg/dL, proteins 180 mg/dL and growth of *Streptococcus pneumoniae* in the CSF culture. Family history was unremarkable.

In the present admission, he had normal mentation, neck stiffness, Kernig's sign, and sensorineural deafness in the right ear detected at the bedside by the Rinne and Weber tests. Rest of the systemic examination was unremarkable. A clinical diagnosis of recurrent bacterial meningitis secondary to an underlying ear malformation was considered. CSF analysis revealed

Abbreviations: CSF, cerebrospinal fluid; CT, computed tomography; MR, magnetic resonance.

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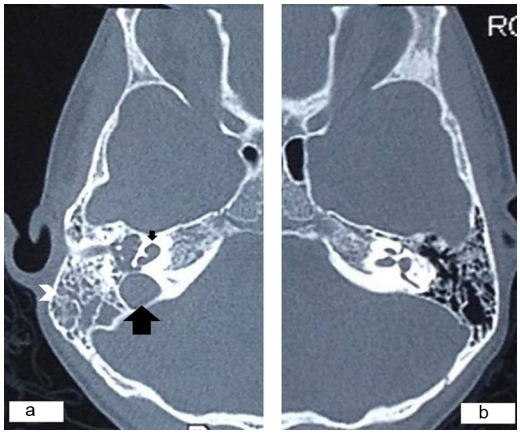

Fig. 1. (A–B) High-resolution computed tomography (axial view) of temporal bone showing (a) one and half turn of right cochlea (small black arrow) with cystic apex (large black arrow) suggestive of Mondini's dysplasia with right sided otomastoiditis (white arrow head) (b) Left sided inner ear structures and mastoid air cells are normal.

1500 cells/μL (81% polymorphonuclear leukocytes), hypoglycorrhachia (18 mg/dL), elevated proteins (480 mg/dL) and growth of *S. pneumoniae* in CSF culture (sensitive to vancomycin and ceftriaxone). Pure tone audiometry confirmed the profound sensorineural hearing loss in the right ear. Serum immunoglobulin, complement levels and nitrobluetetrazolium tests were normal. Serology for human immunodeficiency virus was negative. High-resolution computed tomography (CT) scan of the temporal bones showed a right-sided Mondini dysplasia and otomastoiditis (Fig. 1a and b). Additionally, it showed a high-riding enlarged right jugular bulb with focal dehiscence of its antero-superior wall and a small osteolytic lesion in the left parietal bone adjacent to the sagittal suture suggestive of an epidermoid. Magnetic resonance (MR) cisternography confirmed the inner ear malformation (Fig. 2a and b) and did not reveal any CSF leaks or communication. He was administered intravenous vancomycin and ceftriaxone for 14 days and immunized with *Haemophilus influenzae*, *Pneumococcus* and *Meningococcus* vaccines. As no obvious CSF leak or communication was demonstrated, no surgical intervention was planned and child has been under regular follow-up. Currently, at 8 months follow-up, he is well with no recurrence of symptoms.

Discussion

Recurrent meningitis is defined as two or more episodes of meningitis with a period of complete resolution of symptoms, signs and laboratory findings in between [4]. Recurrent bacterial meningitis is not a common disease and needs evaluation for an underlying etiology such as CSF leaks secondary to anatomic, developmental and traumatic defects, or an underlying immunodeficiency. Of the 363 cases of recurrent meningitis, anatomical defects (59%), immunodeficiency (36%) and parameningeal infections (5%) were the most common causes in children [4].

Congenital anatomic defects are an important but an often missed risk factor for recurrent meningitis. Mondini dysplasia is a rare inner ear malformation which occurs due to an arrest in the development of cochlea in the seventh week of gestation. Individuals with Mondini dysplasia have one and a half coils of the cochlea in association with a dilated vestibule and an enlarged vestibular aqueduct. It usually occurs as an isolated anomaly but may have syndromic association with Pendred, Klippel–Feil and Di-George syndromes [5]. Often, an associated deficient stapes foot plate or oval window results into a fistulous CSF connection with the middle ear, which may cause recurrent bacterial meningitis [6] and variable sensorineural hearing loss. The presence of hearing loss, thus, provides an important clinical clue towards an underlying inner ear malformation and can be easily confirmed at the bedside by the Rinne and Weber tests.

High-resolution CT is the best imaging modality for evaluation of temporal bone defects. MR imaging aids in the identification of defects in the membranous structures and delineation of CSF leaks. MR cisternography in the index case did not reveal any CSF leak or communication between the intra and extra cranial structures as has also been previously reported by Anandi et al. [2]. The probable reason for this is unknown. Sensitivities of high-resolution CT and MR cisternography in detecting CSF fistulas range from 50 to 100% and 55 to 76% respectively [7]. Hence, in the presence of this known risk-factor for recurrent bacterial meningitis in children, it is hard to refute the possible causation in the index case. Alternatively, there may be a small fistulous connection in between the subarachnoid space and the middle ear cavity which the imaging modalities fail to detect [4]. At the same time, the role of additional anatomic abnormalities in the index patient such as a high riding enlarged right jugular bulb with focal dehiscence of its antero-superior wall and a small osteolytic defect in the left parietal bone is not clear in the causation of recurrent meningitis.

Surgical repair of the defect is typically necessary to prevent recurrent episodes of meningitis. Amplification aids or cochlear

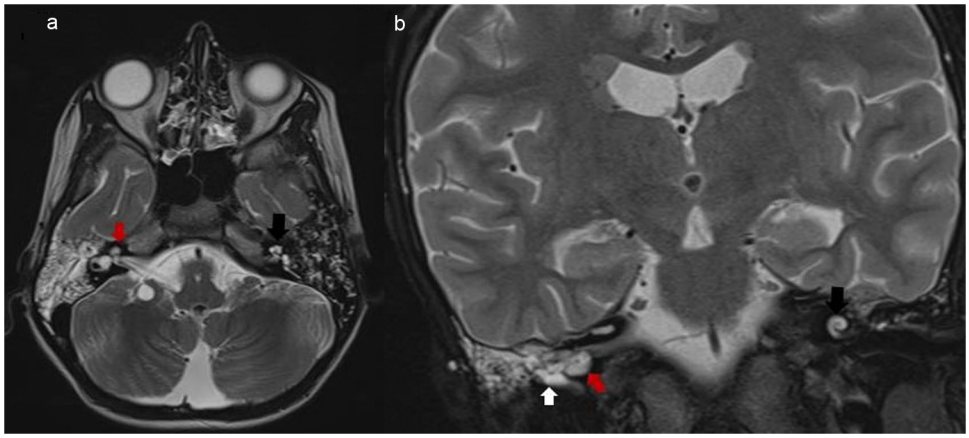

Fig. 2. (A–B) Magnetic resonance imaging of brain (a) T2-weighted axial and (b) coronal sections showing one and half turn of the cochlea (red arrow) with cystic apex (white arrow) on the right side. The normal inner ear on the left side is shown in the same images for comparison (black arrow).

implants may be required for associated hearing impairment [8]. However, the management of children with no demonstrable CSF fistulous connection on neuro-imaging is less clear. Regular follow-up, prompt antimicrobials for any clinical infection and repeated neuroimaging may be needed in symptomatic children. In such children, a low threshold for antibiotics administration to treat the middle ear infections is advisable. Children should be vaccinated for *Pneumococcus*, *Meningococcus* and *H. influenzae* type b. Timely vaccination helps in reducing the colonization of bacteria in the affected individuals and prevents further episodesof meningitis [9].

Conclusions

Recurrent bacterial meningitis with sensorineural hearing loss in a child should raise a suspicion of an inner ear malformation such as Mondini dysplasia. Management of children with no demonstrable CSF fistulous connection is less clear. Regular follow-up, prompt antimicrobials for clinical infection and repeat neuroimaging may be needed in symptomatic children. Vaccination is important to prevent recurrences.

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Competing interests

None declared.

Ethical approval

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