Recurrent Bacterial Meningitis Associated With Mondini Dysplasia

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We reported two cases of recurrent meningitis and both of them had Mondini dysplasia, which provides a link between the brain and inner ear and is associated with cerebrospinal fluid, otorrhea/rhinorrhea, hearing impairment, and recurrent meningitis. Patients who have hearing impairment and recurrent meningitis should be evaluated for the possibility of this congenital dysplasia, and early diagnosis and prompt surgical intervention may prevent further episodes. Copyright © 2011, Taiwan Pediatric Association. Published by Elsevier Taiwan LLC. All rights reserved.

1. Introduction

Recurrent bacterial meningitis represents a rare but serious problem. Even under prompt and appropriate antibiotic therapy, its mortality rate is still high, and severe neurodevelopmental sequelae may occur in 10–20% of patients recovering from bacterial meningitis, too. Therefore, trying to find the cause of recurrent meningitis and prevent further episodes can improve personal health and reduce social cost. We report here two cases of recurrent meningitis plus unilateral hearing loss that evolved into Mondini dysplasia. Details of diagnosis methods and the treatment course are presented and discussed further.

2. Case 1

A 5-year-old boy had a fever for 4 days, and antibiotics with amoxicillin/clavunate was prescribed to him for 1 week under the assumption that it was acute otitis media. However, after vomiting more than 10 times, having a fever up to 40°C, severe headaches, and delirium, he was brought to the emergency department of National Taiwan University Hospital...
Hospital for treatment. His Glasgow Coma Scale was E3M4V2, and his Brudzinski sign showed positive. He was transferred to the pediatric intensive care unit for further care.

Tracing back his past history, he was admitted for bronchopneumonia when he was 3 years old. Pneumococcal meningitis developed 2 days after that period of hospitalization. He recovered under adequate treatment, but his brainstem auditory revealed potential hearing impairment in his right ear. He followed up regularly at our ear, nose, and throat clinics. Rhinorrhea was intermittently noted for 1 year and treated as allergic rhinitis.

This time at pediatric intensive care unit, endotracheal tubes with ventilator support were given initially, as well as a fluid challenge (?) for hypotension. His hemogram showed leukocytosis with left shift (white blood cell count 17,590/μL with 92.2% neutrophils). Cerebrospinal fluid (CSF) study showed pleocytosis with higher protein and lower glucose level (white cell count 435/μL with 13.1% lymphocyte and 86.9% neutrophils, and the glucose level <1 mg/dL). A CSF latex test showing positive pneumococcal antigen and bacterial culture revealed pneumococcus with a penicillin minimum inhibition concentration of 1.5 μg/mL, vacomycin 0.5 μg/mL, and cefotaxime 0.5 μg/mL. Brain computerized tomography (CT) showed brain swelling and right-sided mastoiditis. High-resolution CT (HRCT) showed that the right cochlea was enlarged and defective at least by one turn, which was compatible with Mondini dysplasia (Figure 1). His serum IgG, IgM, IgA, C3, and C4 levels were all within normal ranges. Vancomycin, cefotaxime, and mannitol were given for penicillin-resistant pneumococcus. A seizure occurred on the 2nd day of hospitalization, so phenytoin plus phenobarbital were given. His fever gradually subsided, and his consciousness improved. An extubation was done, and antiepileptic drugs were tapered down. After complete antibiotics therapy, he was discharged on the 21st day of hospitalization.

One month later he received surgical intervention, and a transcanal incision was performed. After removing the stapes, CSF gush-out was noted. Gentamicin-rinsed gel foams were used to obliterate the vestibule via an oval window. After the operation, there was no new episode of meningitis during the following year. CSF rhinorrhea, which was previously presented as the picture of allergic rhinitis, also was completely resolved.

3. Case 2

A 10-year-old boy had a fever accompanied by headaches and vomiting on the morning of June 25, 2006. He took some medicine from a local clinic. However, as the headache progressed, he went to a medical center. There the pediatrician found that he had positive meningeal signs and an injected right eardrum. CSF study revealed a white cell count of 3816/μL, protein count of 185.4 mg/dL, and a glucose count of 38 mg/dL. He was then hospitalized under the assumption that it was bacterial meningitis.

Tracing his medical history, he had one previous hospitalization for meningitis of unknown pathogens in the same medical center in November 2005.

During this hospitalization, he had received vancomycin, ceftriaxone, and mannitol. His serum IgG, IgA, and IgM levels were within normal range. CSF culture yielded penicillin-resistant pneumococcus. HRCT showed abnormally enlarged cochlea with only two turns and an absent horizontal semicircular canal in the right inner ear and right mastoiditis (Figure 2). Pure-tone hearing test revealed right hearing loss. The glucose level of his nasal drip was 37 mg/dL, which supported the possibility of CSF rhinorrhea.

He was then transferred to our hospital for surgical repair of the CSF leakage after the completion of antibiotic treatment. He received surgical intervention, and a defect in the stapes footplate with CSF leakage was found. Soft tissue graft was harvested to eliminate the defect between the stapes footplate and oval window. The postoperative course was smooth. During 6 months of follow-up, no new episode of meningitis occurred, and he also got rid of CSF rhinorrhea.

4. Discussion

Predisposing factors of recurrent meningitis include congenital CSF fistula in 55% of cases, traumatic or surgical CSF fistula in 17%, immunodeficiency in 21%, and unknown causes in 6%. Complement deficiency in C5-C9 is sometimes associated with recurrent meningococcal meningitis, but levels of C3, C4, IgA, IgM, and IgG in our patients were within normal range. Because our patients got pneumococcal meningitis rather than meningococcal meningitis, complement or humoral immunodeficiency was unlikely. In our cases, the obvious predisposing factor is CSF fistula resulting from inner ear malformation because there were neither previous traumatic nor surgical histories in our patients.

The type of organisms found in recurrent bacterial meningitis also strongly correlate with the site of defect. Escherichia coli and Staphylococcus aureus are the common pathogens when the primary defect is in the lumbar sacral region. In contrast, pneumococcus was the most common pathogen in cases with the intracranial defects. Episodes of pneumococcal meningitis resulting from intracranial defect in our patients endorse the above conclusions in the literature.

The Mondini inner ear malformation was first described by Carlo Mondini in 1791, and it is the result of arrested labyrinth development during the 7th week of fetal life. Typically, the cochlear capsule is flattened with an
underdeveloped bony structure in the apical portion of the cochlear, reducing the number of cochlear turns. It means that the true Mondini dysplasia has a cochlea with a normal basal turn and a deficient interscalar septum for the distal one-and-a-half turns. In cases of Mondini dysplasia, the cochlea characteristically has 1–1.5 turns rather than the 2.5–2.75 turns seen in normal individuals. It may be associated with hypoplastic modiolus, large vestibule, and wide semicircular canals. The auditory and vestibular sense organs and nerves may be immature or normal, and these are attributable to varying degrees of hearing impairment or vestibular dysfunction clinically.

The most common route of CSF leakage in cochlear dysplasia is a fistula of the modiolus connecting the subarachnoid space via the internal auditory canal. In our patients, after removal of the stapes, CSF gushed out. Our surgeons used gentamicin-rinsed gel foam to obliterate vestibule via oval window directly in Case 1 and an autologous-harvested soft tissue graft to obliterate the defect in Case 2. Although we do not know the exact site of the fistula, we have closed the bridge of the subarachnoid space and the middle ear. No more CSF leakage or new episode of meningitis occurred at the follow-up.

Clinical presentations of cochlear dysplasia include varying degrees of sensorineuronal hearing loss, CSF otorrhea or rhinorrhea, and recurrent meningitis. If a vestibule gets involved, vestibular vertigo may be present. However, most patients may not have obvious symptoms and signs. In such cases, their otorrhea or rhinorrhea is difficult for us to discriminate between CSF leakage, otitis, and allergic rhinitis, and so recurrent bacterial meningitis may be the first warning. Our cases have given us such valuable lessons, and we have two suggestions for the clinicians. First, children who get the first episode of meningitis should receive a hearing examination. Although hearing impairment is the most common sequel to meningitis, abnormal hearing test results not only show us the postmeningitis functional status of patients but also alert us whether inner ear malformations do exist. Second, persistent otorrhea and rhinorrhea should be checked to see if they are of CSF origin or not. If CSF otorrhea or rhinorrhea or unilateral hearing impairment is present, an image study should be performed in case of inner ear malformation. HRCT provides a good diagnostic tool for inner ear malformation. It has been proven that prophylactic antibiotics are not effective in prevention of recurrent meningitis, but surgical intervention is established to avoid further complications.

In conclusion, Mondini dysplasia or CSF fistula should be suspected in cases with recurrent bacterial meningitis. HRCT is a good diagnostic tool, and surgical intervention is mandatory to get rid of CSF leakage and to prevent new episodes of bacterial meningitis.

References