CASE REPORT

Mondini Dysplasia Presenting as Otorrhea without Meningitis

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Key Words

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Mondini dysplasia is a rare inner ear malformation that is usually only diagnosed after recurrent meningitis. Surgical intervention is mandatory. This report highlights the case of a patient with Mondini dysplasia who presented with hearing impairment and otorrhea and was diagnosed and treated before the occurrence of meningitis, thus preventing morbidity and neurologic sequelae. Hearing impairment may be the only manifestation of Mondini dysplasia, and the benefit of hearing screening is emphasized. Temporal bone computed tomography should be considered in children with unilateral sensorineural or mixed-type hearing impairment.

1. Introduction

As a rare congenital inner ear malformation, the Mondini malformation is characterized by cochlear malformation with dilatation of the vestibule, aqueduct, and ampulla, and incomplete partition of the cochlea.1 A perilymph fistula with cerebrospinal fluid (CSF) leakage is relatively common, and CSF rhinorrhea or otorrhea may develop.2 Recurrent meningitis is easily complicated and leads to a diagnosis of Mondini dysplasia.3 Hearing impairment is often associated with, but seldom the first clinical presentation of, Mondini dysplasia. We present the case of a girl with Mondini dysplasia who was diagnosed on the basis of hearing impairment and otorrhea before meningitis had occurred.

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2. Case Report

A 10-month-old girl who was delivered at term was referred to our outpatient clinic due to abnormal findings during hearing screening. There were no perinatal insults, and her growth and development were within normal limits. No other discomfort was noted, and no significant abnormal findings were disclosed by physical examination. Several tests were arranged to evaluate the patient’s hearing impairment. Type A (left ear) and type B (right ear) tympanograms were identified. Steady-state evoked potential analysis revealed hearing threshold levels of 30 dB for the left ear and 90 dB for the right ear. Further visual reinforcement audiometry was normal for the left ear and a mixed-type hearing impairment was identified for the right ear.

The patient was regularly followed-up at our outpatient clinics, and bilateral otitis media developed when she was 15 months old. Oral amoxicillin/clavulanate was prescribed, but her effusion persisted. Bilateral ventilation tubes (grommets) were implanted 3 months later, and then the patient developed right otorrhea. The discharge was clear, without any smell. No fever, rhinorrhea, vomiting, cough, or other discomfort was associated with the otorrhea. Physical examination was normal except for identification of bilateral ventilation tubes in situ and right otorrhea. The white blood cell count was $8.5 \times 10^9/L$. CSF leakage was suspected and proven by analysis of the discharge (glucose 65 mg/dL, protein 365 mg/dL). Temporal bone multi-slice computed tomography revealed accumulation of fluid in the right middle ear and mastoid area (Figure 1). A right inner ear anomaly with dilatation of the right vestibule, right lateral semicircular canal, and lack of the spiral laminar septum in the apical turn were noted; there were only 1.5 cochlear turns (Figure 2). Mondini dysplasia of the right ear was diagnosed, and surgical intervention was performed. A perilymph fistula between the facial nerve and oval window was found during the operation. Excision of the fistula and repair of the defect were successful. Culture of the CSF otorrhea yielded *Haemophilus influenzae* non-type b. The postoperative course was uncomplicated and no cochlear implantation was performed.

3. Discussion

Mondini dysplasia is a prevalent inner ear malformation associated with CSF leakage.\(^1\)\(^2\) It is thought to be the result of arrest of neural tube development during the seventh week of gestational age, and has been associated with meningocele.\(^5\) Some gene loci have been identified, such as DFN3 and SLC26A4, and several congenital syndromes have been associated with the disorder, including Klippel-Feil syndrome, Pendred syndrome, DiGeorge syndrome, and chromosomal trisomies.\(^6\) A relatively high prevalence of the SLC26A4 mutation spectrum has been noted in Taiwan, with IVS7-2A>G being the most common mutation.\(^7\) Genetic counseling was performed for our patient, but no compatible syndrome was identified and so no further gene analyses were performed.

A perilymph fistula communicating between the subarachnoid space and the middle ear is common in patients with Mondini dysplasia.\(^2\) CSF otorrhea and rhinorrhea may occur in such patients, but there is no CSF otorrhea in those with an intact tympanic membrane. CSF otorrhea may occur when the membrane ruptures, as was the case with the present patient. Retrograde meningitis is relatively common in such cases, and recurrent meningitis is the diagnostic hint.\(^3\)\(^8\) Often, Mondini dysplasia remains undiagnosed until recurrent meningitis occurs. Meningitis is associated with high mortality and morbidity, and neurologic sequelae are common. Early diagnosis and treatment of meningitis can improve outcomes and reduce permanent...
damage. Fortunately, no episode of meningitis developed in our patient, and early surgical repair could have prevented its occurrence.

Recurrent meningitis is very rare. Anatomical defects and immunological disorders may contribute to recurrent meningitis, and Mondini dysplasia is often diagnosed during pursuit of its underlying etiology. However, the presentation of Mondini dysplasia may mimic other disease entities, such as otosclerosis. Although hearing impairment is relatively common in patients with Mondini dysplasia, it can easily be missed in children due to difficulties associated with evaluation. Children cannot cooperate with physicians and express their hearing impairment, leading to delays in diagnosis. The diagnosis of Mondini dysplasia before the occurrence of meningitis is rare. Recurrent meningitis can guide us to such a diagnosis, but early diagnosis before the occurrence of meningitis would be preferable.

Hearing impairment is not rare, with the prevalence in the adult population being about 11.5–17.1% in western countries and 21.4% in Taiwan.11 Careful evaluation is required since many etiologies are associated with hearing impairment. All external, middle, and inner ear diseases as well as nervous system disorders can result in hearing impairment. The type of hearing impairment in Mondini dysplasia is mainly sensorineural, but it may be accompanied by conductive hearing impairment once CSF leakage or another disorder develops. Multi-slice temporal bone computed tomography is indicated in children with unilateral sensorineural hearing impairment.12 However, when computed tomography is not easily accessible, children with hearing impairment should at least be evaluated carefully and be followed-up in the long term.

Hearing impairment is easily missed in children due to its occult presentation; universal hearing screening is beneficial for these patients.13 Our patient was referred due to an abnormal finding identified during a hearing screen; thus, the benefit of hearing screening is clear. Although otoacoustic emission and automated auditory brainstem response tests are widespread, universal hearing screening is not yet performed in Taiwan. We strongly suggest that it be included in the national health insurance program.

The common pathogens of meningitis in Mondini dysplasia are similar to those found in otitis media, among which Streptococcus pneumoniae is the leading cause.14 The culture result for our patient, Haemophilus influenzae non-type b, is also a common pathogen. The increasing common use of immunization with the conjugate pneumococcal vaccine may change the epidemiology. Neither Haemophilus influenzae type b nor the pneumococcal vaccine was administered to our patient before admission. Vaccination against these pathogens was administered after the current episode.

In conclusion, Mondini dysplasia is a rare cause of hearing impairment, and delayed diagnosis may lead to recurrent meningitis and cause neurologic sequelae. Meningitis or CSF leakage does not always occur early in patients with Mondini dysplasia. Universal hearing screening should be promoted since hearing impairment can be the only manifestation in children. For children with unilateral sensorineural or mixed-type hearing impairments, temporal bone computed tomography should be arranged before the occurrence of CSF rhinorrhea, CSF otorrhea, or recurrent meningitis. Early detection and treatment are valuable, since these can prevent the occurrence of meningitis, as was the case with our patient.

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References