

CASE REPORT

Mondini Dysplasia Presenting with Recurrent Meningitis

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Mondini dysplasia (MD) is the most common congenital malformation of the cochlea associated with sensorineural hearing loss in variable degrees and recurrent meningitis. It occurs due impairment of the embryonic development of the inner ear during the seventh week of fetal life. Congenital anomalies of temporal bone may cause fistulisation between the middle ear and subarachnoid space. MD is often associated with spontaneous cerebrospinal fluid leakage that leads to recurrent bacterial meningitis. MD generally remains undiagnosed until the radiological investigation for hearing loss and recurrent meningitis. Computed tomography of the temporal bone is the best method of MD diagnosis. Immediate surgical closure of the defect is the treatment of choice to prevent further episodes of meningitis. In this article, we presented a case of Mondini dysplasia in a 5-year-old patient with recurrent meningitis as an initial symptom and discussed the pathogenesis of meningitis and treatment methods. We also reviewed pediatric MD cases with recurrent meningitis since 1990.

Submitted : 22 November 2009

Revised : 22 July 2010

Accepted : 24 August 2010

Introduction

Mondini dysplasia (MD) is a malformation of cochlea associated with large vestibular aqueduct and dilatation of semicircular canal ampulla^[1]. It is the most common type of cochlear anomalies (about 50% of cases)^[2]. Mondini described this abnormality after the postmortem examination of the 8-year-old child with severe hearing loss in 1791.1 Cochlea has 1 ½ turns instead of normal 2 ½ turns. Classic MD has three components; cystic apex, dilated vestibule and large vestibular aqueduct. Only the basal turn is developed; the upper turns form a common cavity³. According to Jackler et al. it is correspond to incomplete partition of cochlea^[2]. Hearing loss may change mild to profound deafness because of the number of spiral ganglion cells are decreased. If the malformation is unilateral, child's family may not recognize hearing loss^[1,3].

Congenital anomalies of temporal bone may cause fistulisation between the middle ear and subarachnoid space. These patients can be presented with sensorineural hearing loss, otorrhea and rarely recurrent bacterial meningitis^[1]. MD is often associated with spontaneous cerebrospinal fluid (CSF) leakage that leads to recurrent bacterial meningitis. Meningitis is especially developed during episodes of bacterial otitis media. Recurrent meningitis can occur due to

immune deficiency or CSF leakage. CSF leakage may occur due to the cranial trauma or temporal bone malformations. MD may remain undiagnosed until the radiological examination of temporal bone for recurrent meningitis and hearing loss^[1,4,5].

We presented a 5-year-old girl with MD whose initial symptom was recurrent bacterial meningitis. We also discussed the pathogenesis of meningitis and surgical treatment modalities.

Case Report

A 5-year-old girl was consulted from the pediatrics infectious diseases department for the evaluation of the underlying cause of her recurrent meningitis. She had five bacterial meningitis attacks previously. *S. pneumoniae* was isolated from both CSF and blood during last meningitis episode. She had no neurologic sequel of meningitis and family was unaware of her hearing loss. A subtotal tympanic membrane perforation and pulsated mucopurulent discharge from right ear was detected on otoscopic and otomicroscopical examination. The hearing threshold of the left ear was in normal range, and total deafness was detected in the right ear with brainstem evoked response audiometry (BERA). The other physical examination was normal. The laboratory findings were

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also in normal limits. The apical cochlear turns, the vestibule and the lateral semicircular canal were dilated without normal septation on temporal bone computer tomography (CT) (Figure 1). There was a thin membranous barrier between brain and middle ear through fundus of the internal acoustic canal. Additionally, the tympanomastoid cavity was filled with soft tissue and aditus ad antrum was blocked by inflammatory tissue (Figure 2).

Tympanomastoidectomy was planned to eradicate chronic ear infection and close the tympanic membrane perforation. The mastoid cellulae were filled with granulation tissue and aditus ad antrum, epitympanic recess and oval window occupied by a translucent sac filled with clear fluid. The malleus and incus were intact, but stapes was not observed. The sac pressed down gently for the maintaining of attic-antral passage after the completing the mastoidectomy. During this maneuver, the sac was ruptured and cerebrospinal fluid gusher was developed. CSF leakage was repaired by grafting with temporal fascia and fibrin glue. Tympanic membrane perforation was repaired by using tragal cartilage island graft. The postoperative period was unremarkable. She was discharged at the seventh postoperative day. We didn't encounter any meningitis attack after the surgery during the two years follow-up.

Discussion

Mondini dysplasia is a congenital disorder of the inner ear affects the membranous and osseous labyrinth. The inner ear develops during the 4th–8th week of

intraembryonal life. The cochlea has only 1 ½ turns at seventh week of gestation. MD may result from arrested development during fetal life or from abnormal development due to genetic defects or teratogenic agent^[6]. It may be rarely bilateral. Some patients have an abnormal stapes. There may be a defect in the footplate or occasionally, the stapes is totally absent. Although associated syndromes like Klippel-Feil syndrome, Penred's syndrome, Di-George syndromes may be present with MD, it can be seen as an isolated finding^[5]. Recently, a microdeletion at the locus DFN3 on chromosome X was shown in a familial MD^[7]. Sensorineural hearing loss occurs due to anomalies of Corti organ as well as altered pressure in the perilymphatic space. Mixed hearing loss may occur due to ossicular pathology or CSF otorrhea. The cochlea is seen smaller than the normal size and partially or completely lacks interscalar septum in radiological evaluation. When there is CSF fistula, connection of subarachnoid space and inner ear is mostly through the fundus of internal acoustic canal^[1]. In our case, connection of subarachnoid space and inner ear is also through the fundus of internal acoustic canal.

Patients with MD are predisposed to developing CSF leakage. CSF can flow from the subarachnoid space into the perilymphatic space. CSF can leak through a defect in the otic capsule into the middle ear. Although defects have been reported in the round window, eustachian tube, promontory, fallopian canal and hypotimpanium, the most common site of leakage is the oval window or stapes footplate. A CSF effusion



Figure 1. The apical cochlear turns; the vestibule and the lateral semicircular canal were dilated without normal septation were seen on temporal bone CT. (Black arrow)

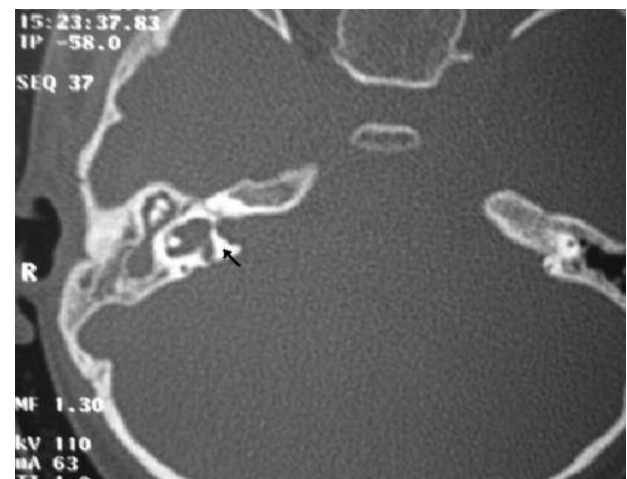


Figure 2. The apical cochlear turns; the vestibule and the lateral semicircular canal were dilated without normal septation were seen on temporal bone CT. There was a thin membranous barrier between brain and middle ear through fundus of internal acoustic canal. (Black arrow)

in the middle ear cavity may cause otorrhea or rhinorrhea. The effusion is mostly asymptomatic. When the CSF communicates with the middle ear, bacterial contamination of CSF and retrograde meningeal infection may occur through the defect in the otic capsule^[8-10]. Ohlms and colleagues reported three new MD cases with recurrent meningitis and reviewed literature. They reviewed 39 cases which are published from 1967 to 1990 in the literature. In these cases two patients had 20 episodes of meningitis prior to diagnosis^[1]. We also reviewed the English literature and seven pediatric cases were reported since 1990. Table 1 reviews the MD patients with recurrent meningitis described in the literature since 1990.

S. pneumoniae is the most common infecting organism in the patients with recurrent meningitis, followed by *H. influenzae* and *S. aureus*^[1]. In our case *S. pneumoniae* was isolated from CSF and blood during the last meningitis episode. The most common isolated microorganism is *S. pneumoniae*.

Radiological evaluation and BERA verified the diagnosis. High resolution CT is the gold standard procedure^[11]. Intrathecal fluorescein injection can be helpful for the detection of CSF leakage during surgery. CT cisternography which is an invasive technique can be difficult to perform in young children^[12,13].

The treatment of CSF leak is the Mondini deformity includes a thorough examination of the middle ear and targeted approach to the area of the leak. The method of closure of the fistula depends on the functional status of the ear. The leakage mostly originates from the oval window. Stapedectomy with obliteration of the vestibule by a free muscle and fascial grafts were recommended for surgical closure of the defect. Blind obliteration of the middle ear cavity is not recommended. If the patient had residual hearing, rehabilitation with hearing devices can be used. Cochlear implantation has been also shown to be useful for the hearing restoration. The diagnosis and treatment of CSF leak in a patient with bilateral MD can be problematic^[1,2,6].

As a conclusion; all children with recurrent meningitis without an obvious cause should undergo CT of the temporal bone to exclude inner ear malformation. Normal hearing history does not exclude inner ear anomaly because of family may not recognize unilateral hearing loss. Early diagnosis and surgical closure of the CSF leakage is vital to prevent complications of recurrent meningitis.

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Table 1. Review of some pediatric MD patients with recurrent meningitis described in the literature. (BERA: Brainstem Evoked Response Audiometry, y: year, F: female, M: male)

Authors	Years	Age, Sex	Attacks of meningitis	Clinical features	Audiometry, BERA	Region of fistula
Present case	2009	5y,F	5	Recurrent meningitis	Right hearing loss	Oval window
Ciftdogan et al ⁸	2009	16y,M	2	Recurrent meningitis	Right hearing loss	Not determined
Liu et al ¹⁰	2008	4y,F	4	Recurrent meningitis	Right hearing loss	Not determined
Tullu et al ⁹	2004	11y, F	5	Recurrent meningitis	Right hearing loss	Oval window
Kitazawa et al ⁴	2004	2,F	2	Recurrent meningitis	Right hearing loss	Promontorium
Lue and Manolidis ¹³	2004	1,5y, M	Multiple episodes	Recurrent meningitis	Bilateral hearing loss	Not determined
Rupa et al ⁶	2000	12y, F	3	Recurrent meningitis	Right hearing loss	Oval window
Muranjan et al ¹²	1999	4y, M	2	Recurrent meningitis	Left hearing loss	Oval window

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