CASE REPORT

Recurrent meningitis and Mondini dysplasia: A case report

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Mondini dysplasia represents a developmental arrest during the embryogenesis of the inner ear. The malformation, commonly associated with deafness, is frequently heralded by meningitis secondary to communication of the middle ear with the subarachnoid space. Early recognition and successful repair of the cerebrospinal fluid leak is important to avoid the sequelae or repeated episodes of meningitis.

We report a case of a 5-year-old girl in whom Mondini dysplasia presented initially with recurrent pyogenic bacterial meningitis. This case is being reported for its rarity and to increase awareness among physicians regarding the possibility of Mondini dysplasia in children with recurrent pyogenic meningitis and sensorineural deafness.

Recurrent pyogenic meningitis can occur because of cerebrospinal fluid (CSF) leaks or immune deficiencies (eg, complement deficiency, asplenia, hypogammaglobuinanemia). CSF leaks can either be traumatic (cranial fractures or cranial surgery) or, in children, can occur via various malformations of the labyrinth (including Mondini dysplasia).[2,3] Mondini dysplasia often remains undiagnosed until radiologic investigations for deafness or recurrent meningitis are undertaken.[1]

CASE REPORT

A 5-year-old girl, deaf since infancy, was referred to the Ear, Nose, and Throat (ENT) clinic after her fourth attack of pneumococcal meningitis. She had suffered from 4 similar episodes in the past 3 years and had received treatment elsewhere. At each episode, pyogenic meningitis was diagnosed on CSF analysis and treated with parenteral antibiotics, resulting in complete recovery. However, investigations for testing hearing ability were not undertaken until she presented to us.

At the age of 2 years, she had developed the first attack of pneumococcal meningitis, with otitis media on the right side. No evidence of CSF otorrhea or rhinorrhea was revealed at that time. Six months afterward, the child suffered her second attack of pneumococcal meningitis. One year after the second attack, she developed her third attack, and the child was referred to a neurosurgery clinic for a possible intracranial closure of the CSF leak.

Investigations for rhinorrhea with imaging and physical examination showed no abnormality. The patient underwent duraplasty and fistula reconstruction by bicoronal approach in the neurosurgery clinic. Six months after the operation, the patient developed her fourth case of pneumococcal meningitis and was treated with parenteral antibiotics resulting in complete recovery. The child was referred this time to the ENT clinic. General examination revealed a child of normal height and weight and normal vital parameters. Bicoronal incision could be seen in the frontal region of the patient's face.

Neurologic examination on admission revealed minimal meningeal signs. There were no focal neurologic deficits. CSF did not show any organism on smear or culture. Workup for immunologic abnormalities including serum immunoglobulin levels; complement levels were normal. Her serologic test for HIV antibodies was negative. Otorhinolaryngologic evaluation revealed profound sensorineural hearing loss in both ears on brainstem auditory evoked response (BAER) testing and audiometry. Petrous high-resolution computer tomography (CT) demonstrated the characteristic features of Mondini dysplasia in both ears. The apical turns of the cochlea were dysplastic, with 1.5 turns. The vestibule was large and bulbous, and the semicircular canals were wide and short (Figure). The internal auditory canal was normal.

The middle ear was explored after an intrathecal injection of a prepared solution (1 cc fluorescein 10% mixed with 8 cc sterile isotonic solution). The dye was subsequently found to be leaking 30 minutes after the intrathecal injection. Both tympanic membranes were intact. The CSF in the middle ear was seen trickling through the Eustachian-tube orifice on nasopharyngoscopy, manifesting as CSF rhinorrhea with profuse leakage. A diagnostic myringotomy revealed that CSF was leaking into both tympanic cavities. The patient's CSF otorrhea was persistent after the procedure, and continuous spinal drainage was done. She subsequently developed another attack of hemolytic streptococcal meningitis.

Once the meningitis was brought under control, both middle ears were explored. Upon surgery, a CSF fistula was noted at the oval window leaking through a defect in both ears. The stapes was removed, and the vestibule of both ears was filled with fascia and muscle. A lumboperitoneal shunting procedure was done and left in for 7 days. Antibiotherapy was administered, and immobilization was performed.

Surgical closure of the defect (at the footplate of the stapes at the oval window causing the CSF-perilymph leak) was carried out successfully and the child has had no further episodes of pyogenic meningitis on a follow-up of 6 months.

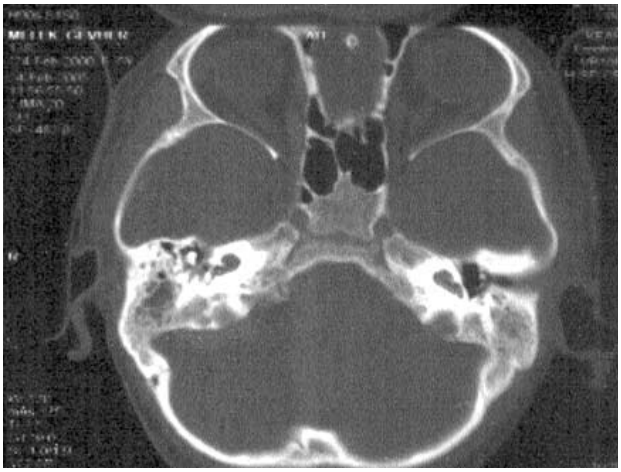

Figure: Petrous high-resolution computer tomography demonstrated the characteristic features of Mondini dysplasia in both ears

DISCUSSION

Mondini described postmortem anatomic findings of the inner ear in a congenitally deaf 8-year-old boy in 1791. In the Mondini deformity, the cochlea has but 1 to 1.5 turns instead of the normal 2.5 to 2.75 turns. Only the basal turn is fully developed.[4-6] (Valvassori and colleagues noted that nearly one-half of the abnormal cochlea in their patients with Mondini deformity were reduced in size rather than actually reduced in number of turns.7) The vestibule is bulbous while the semicircular canals may be foreshortened, enlarged, or missing. The modiolus and osseous spiral lamina are hypoplastic, and the cochlear duct is shortened and broad.8 A defect in the oval window or the stapes footplate is usually present and provides the route for CSF from the inner to middle ear. Since the external and middle ear are derived from first and second branchial arch mesoderm, unlike the stapes footplate which develops from neurectoderm, there are no external auricular clues that an anomaly is present.

Developmental arrest at the sixth or seventh week of embryonic life is believed to be the cause of the defect.1 Rarely, the defect may be bilateral. The vestibular structures may be abnormal as well.[5] At times, some sensory epithelium may still be present, and this gives the hope of some preservation of hearing. Our patient had bilateral Mondini deformity with bilateral total sensorineural hearing loss.

Evaluation includes analysis of auditory function and definition of the anatomy of the middle and inner ear. Audiograms and/or BAERs as well as high resolution CT scanning of the petrous bones should be obtained. Metrizamide-enhanced CT scans or radioisotope tracer studies are not necessary if the temporal bone studies are diagnostic.

Mondini Dysplasia may occur in association with Klippel-Feil syndrome, Pendred syndrome, trisomy syndrome, or DiGeorge's syndrome, or it may occur in isolation.[6] Recently, microdeletion at the locus DFN3 on chromosome X was shown to be present in familial Mondini dysplasia.[9] In recent years, this condition has been diagnosed more frequently because of polytomographic studies and has been recognized as one of the most common causes of congenital deafness.[5]

In the normal structure of the inner ear, a cribriform plate of bone exists between the fundus of the internal auditory canal and the vestibule.[10] This bony plate provides passage for the fibers of the vestibular nerve. As demonstrated in study using Pantopaque, patients with Mondini dysplasia may harbor a defect in this bony plate through which the CSF enters the vestibule from the subarachnoid space referans.

If another defect exists in the stapedial footplate or the oval or round window, the CSF then leaks from the vestibule through the defect to the middle ear cavity, resulting in CSF rhinorrhea or otorrhea.

A search of the literature revealed detailed descriptions of 33 cases in which bacterial meningitis preceded the diagnosis of spontaneous CSF otorrhea.[2,6,11,12] Most cases have been reported in the otolaryngology literature, and descriptions of this condition in the neuroscience literature are few.

All except 2 cases occurred in children, the majority of whom were under the age of 10 years at the time of initial presentation. The presence of unilateral or bilateral hearing impairment before an attack of meningitis was common. In all except 1 case, the CSF leak was not sufficiently profuse to be noted clinically. Streptococcus pneumoniae was the most common organism to be isolated. In at least half of the cases, there were inner ear anomalies suggestive of Mondini

dysplasia, such as a dysplastic cochlea or an abnormally large vestibule.

Mondini dysplasia should be considered a possible cause of recurrent and unexplained attacks of meningitis in a child who suffers congenital deafness. Audiometry and polytomography of the petrous bone are recommended as initial investigations. If Mondini dysplasia is demonstrated on the polytomograms, myringotomy or exploratory tympanotomy should follow to confirm the presence of the CSF in the tympanic cavity. The intrathecal injection of fluorescein 10% immediately before the procedure will facilitate recognition of CSF otorrhea. Once the CSF otorrhea has been confirmed by myringotomy or tympanotomy, further investigations such as isotope cisternography, metrizamide CT, or a Pantopaque study of the posterior fossa may not be necessary, as in our case.

Surgical correction of spontaneous CSF otorrhea when associated with congenital anomalies of the ear is particularly difficult. In 33 cases reported in the literature, the otolaryngologist first performed tympanotomy and attempted to close the defects. However, 61% of the patients subsequently required repeated surgical procedures before the CSF fistula was obliterated permanently.

We recommend a tympanotomy with obliteration of the defect and the middle ear cavity as an initial procedure when attempting to close a CSF fistula associated with congenital anomalies of the ear. CSF leak through a defect of the fallopian canal or Hyrtl's fissure and a widely patent cochlear aqueduct can be arrested successfully by an otolaryngologist using this approach.

In cases where an extracranial attempt has failed, an intracranial procedure may allow successful closure of the CSF leak, exposed through a suboccipital craniectomy.

An intracranial closure of the CSF leak in patients with Mondini dysplasia may present a more difficult problem because of the absence of an identifiable dural defect. CSF leak occurs through a defect in the fundus of the internal auditory meatus, it is essential to plug the internal auditory meatus with muscle or fascia through a suboccipital craniectomy.[6,13-16]

A lumboperitoneal shunting procedure has proved to be effective in controlling the CSF leak, carry little risk of suctioning bacteria into the intracranial space.[17] It may also be used as a primary or adjunctive procedure for treatment of the spontaneous CSF otorrhea secondary to congenital anomalies of the ear. However, severe low-pressure headaches can follow shunting, especially in patients who have normal CSF absorption. Our patient tolerated these drains without difficulty, and we have experienced no complications associated with this procedure.

In summary, congenital anomalies of the cochleovestibular system may be associated with recurrent episodes of meningitis. The infection gains entry to the subarachnoid compartment through a fistula at the stapes footplate in association with a defect between the inner ear and subarachnoid space. Successful management first depends on recognition of the entity which may be occult in the pediatric age group. Otosurgical obliteration of the middle-to-inner ear communication and concurrent use of a lumbar drain to allow the tissue seal to heal has resulted in early success with low morbidity.

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