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# **Case Report**

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# Spontaneous CSF otorrhea in cases of Mondini's dysplasia: a rare cause of recurrent bacterial meningitis

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#### **ABSTRACT**

Inner ear anomaly is rare developmental anomaly with Mondini's dysplasia being the most common. The perilymphatic fistula cause due to high pressure condition in anomalous inner ear result into CSF otorrhea which become a tract for bacteria to reach central nervous system and cause recurrent meningitis. We are presenting 3 cases of Mondini's dyaplasia which had CSF leaking into middle ear cleft which was diagnosed with HRCT of temporal bone with cisternography and MRI cisternography and were managed successfully with surgical repair by transmastoid approach.

**Keywords:** Mondini's dysplasia, Spontaneous CSF otorrhea, Recurrent meningitis

# INTRODUCTION

Mondini's dysplasia is a developmental anomaly of inner ear which is classically described as one and half turns of cochlea resulting in cystic apex and dilated vestibule and vestibular aqueduct. These patients have profound sensori-neural hearing loss since birth. In such anomaly, there are chance of leakage of perilymph from inner ear to middle ear cleft through various fistulised defects which become route of spread of infection directly to meninges, central nervous system resulting into recurrent episodes of meningitis. Hereby, we present 3 cases of Mondini's dysplasia causing spontaneous CSF otorrheas which were the cause for recurrent attacks of meningitis.

#### **CASE REPORT**

#### Case 1

9 year old boy presented with history of 5 episodes of meningitis in last 3 years. He was a diagnosed case of

Wildevanck syndrome (cervico-oculoacoustic syndrome). He had Klippel-Feil syndrome type II (fused two cervical vertrebrae with lumbar hemivertebrae) (Figure 1e), difficulty in abduction in the left eye with indrawing of the eyeball in this movement (Duane's phenomenon) and bilateral profound hearing loss with no language development (Classical triad of Wildervanck syndrome). He had no history of ear ache, discharge or trauma. He had history of undergoing cortical mastoidectomy after three attacks of meningitis in view of suspected coalescent mastoiditis but no glue or granulation were seen. On examination, patient had bilateral intact tympanic membrane with loss of cone of light on right side. Tuning fork test showed no response. There was no abnormality on vestibular examination. HRCT of temporal bone revealed soft tissue density in right side middle ear and mastoid which had Hounsefeild unit suggestive of fluid. There was similar finding on the left side. On both sides, cochlear-vestibular anomaly was present with only one and half turn of cochlea suggestive of Mondini's dysplasia with dilated internal acoustic

meatus (Figure 1a and 1b). The vestibular aqueduct was also dilated on right side (Figure 1c). CT cisternography was done to locate the exact site of leak. It revealed a frank leak on the right side from the oval window in to the middle ear but no evidence of leak on the left side (Figure 1d).

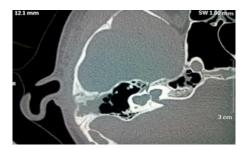


Figure 1a: Mondini's dysplasia with dilated internal acoustic meatus.

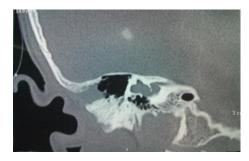


Figure 1b: Mondini's dysplasia with dilated internal acoustic meatus.



Figure 1c: The vestibular aqueduct was dilated on right side.



Figure 1d: It revealed a frank leak on the right side from the oval window in to the middle ear but no evidence of leak on the left side.

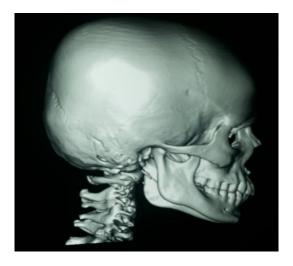


Figure 1e: Klippel-Feil syndrome type II (fused two cervical vertrebrae with lumbar hemivertebrae).

Patient was taken up for exploratory mastoidectomy. Canal wall down mastoidectomy was done and the leak was seen coming out of oval window. The leak was plugged with fat followed by cartilage. Tissue glue was applied followed by insertion of a small stent near round window for it's identification for cochlear implant insertion at a later stage.

Temporalis fascia was draped over the middle ear and mastoid and wide meatoplasty was done. Immediate post-operative, lumbar drainage of 50 cc of CSF was done to reduce the CSF pressure for seal to settle. On 6 months of follow up, patient was doing well with no recent episodes of meningitis.

# Case 2

6 year old girl presented 3 episodes of meningitis in last 1 year. Patient had profound hearing loss since birth. She had no history of ear discharge or trauma but she had complaint of left ear ache preceding every episode of meningitis. She had underwent cortical mastoidectomy in view of mastoiditis which concluded with no positive finding.

On examination, bilateral pars tensa was intact with presence of tympano-sclerotic plaque in left side. HRCT of temporal bone depicted presence of fluid in the middle ear on the left side with bony defect in lateral bone of mastoid secondary to mastoidectomy but the site of leak could not be localized. HRCT of temporal bone also showed presence of Mondini's dysplasia with normal vestibular and cochlear aqueduct.

Patient had a recent episode of meningitis and hence, CT cisternography was deferred to avoid risk of chemical meningitis due to injection of contrast (Figure 2a and b). MRI cisterography revealed presence of fluid in middle ear in continuation with oval and round window.



Figure 2a: Patient had a recent episode of meningitis and hence, CT cisternography was deferred to avoid risk of chemical meningitis due to injection of contrast.



Figure 2b: Patient had a recent episode of meningitis and hence, CT cisternography was deferred to avoid risk of chemical meningitis due to injection of contrast.

Patient underwent radical mastoidectomy with otorrhea repair in both oval and round window. Leak was repaired in fat, cartilage and temporalis fascia augmented with tissue glue. The mastoid cavity was obliterated with pedicled temporalis muscle flap. On 6 months of follow up, patient was doing well with no recent episodes of meningitis.

## Case 3

13 year old female with congenital profound sensorineural hearing loss presented with history of 4 episodes of meningitis within last 3 years. She was a known hypothyroid and was taking 50 micrograms of thyroxine since last two years. She had no history of ear discharge or trauma but complained of right ear ache before last two episodes. On examination, patient had wax granuloma in right ear and intact pars tensa. HRCT temporal bone showed bilateral Mondini's dysplasia with soft tissue density in middle ear cleft on right side (Figure 3a). MRI brain confirmed the finding (Figure 3b-3d). CT cisternography was done and the contrast leaked out instantly from the right nostril suggestive of presence of contrast in the middle ear which leaked into nasal cavity through Eustachian tube. The site of leak was localized to oval window.

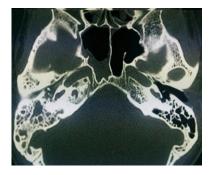


Figure 3a: Bilateral Mondini's dysplasia with soft tissue density in middle ear cleft on right side.

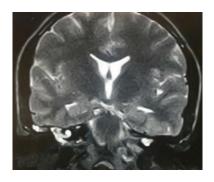


Figure 3b: MRI brain confirmed the finding.

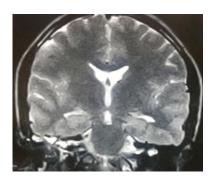


Figure 3c: MRI brain confirmed the finding.

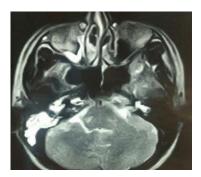


Figure 3d: MRI brain confirmed the finding.

Patient was operated for canal wall down mastoidectomy with multiple layer leak repair. On follow up, patient was doing well without any complication. Patient was subjected to perchlorate discharge test which was positive suggestive Pendred syndrome.

#### **DISCUSSION**

Recurrent bacterial meningitis is defined as two or more episodes of meningitis that are separated by a period of convalescence and the complete resolution of all signs, symptoms, and laboratory findings.<sup>1-3</sup> It constitutes less than one percent of total incidence of bacterial meningitis. The challenge in such cases is to diagnose the underlying cause. The most common causes of recurrent meningitis are anatomic bacterial abnormalities (congenital or acquired) and conditions resulting in immunodeficiency or immunosuppresion. Presence of anomalous tract in paranasal sinuses or inner ear are most common routes of spread of bacterial infection to central nervous system. The age of the patient and the organism isolated give clue to the underlying defect. Recurrent pneumococcal meningitis is seen in head injury with basal fracture, congenital skull defects, meningocele, neurenteric cyst, inner ear dysplasia, asplenia, x-linked agammaglobulinemia, IgG subclass deficiency, early complement deficiency, HIV infection, and chronic otitis media/mastoiditis.2

Mondini's dysplasia is one of the most common developmental anomaly of inner ear. Mondini's defect which was first described by Mondini in 1791, is a developmental anomaly of otic capsules with an arrest in the development of the cochlear modiolus in the seventh foetal week. This leads to development of only one and half turns of cochlea instead of normal two and half turns of cochlea. The cochlea is an amorphous sac lacking a modiolus or central bony spiral without the normal basal turn. There is absence of the interscalar septum in the upper part of the cochlea, with the formation of a common scala (scala communis).<sup>4</sup> dysplasia or aplasia of stapes may be seen in patients with Mondini's dysplasia. Although associated syndromes like Klippel-Feil syndrome, Pendred's syndrome, Di-George syndromes may be present with Mondini's dysplasia, it can be seen as an isolated finding.<sup>5</sup> Recently, a micro-deletion at the locus DFN3 on chromosome X was shown in a familial Mondini's dysplasia.6 In our series ,one patient had Klippel-Feil syndrome as a part of Wildervank syndrome (cervico-oculo-acoustic syndrome) and one patient was diagnosed with Pendred syndrome as she was having subclinical hypothyroidism and positive perchlorate discharge test.

In patients with Mondini's dysplasia, CSF leaks occur when CSF enters the middle ear by transgressing perilymphatic space where it enters through the subarachanoid space. The site of leak is most commonly is the defect in stapes footplate or oval window. But rare sites like fallopian canal, round window, promontory and Eustachian tube area have also been reported in literature. CSF fistula can also be present when internal auditory canal is dilated and is directly communicating with inner ear and subarachanoid space. In our two patients unilateral CSF fistula was present and in all the three

patient, leak was from the region of oval window and in one patient also from the round window.

The radiological investigation of choice in such patients is CT cisternography. MR cisternography can also be done in cases who allergic to contrast or have contraindication for doing lumbar puncture or have a higher chance of risk of precipitation of chemical meningitis. The advantage of HRCT temporal bone cisternography is, in addition of demonstration of leak, it also gives detail of bony configuration of temporal bone and relation of site of leak with important structure like facial nerve, promontory and jugular bulb.

The most common organisms reported are streptococcus pneumonia, Haemophilus inflenzae and Staphylococcus aureus. In two of our patients, the causative organism was pneumococcus and one had Acinetobacter.

The line of management is control of meningitis followed by surgical repair of site of the leak. Surgical repair is done by transmatoid approach with obliteration of vestibule with fascia or muscle. In all our cases, we did stapedectomy with obliteration of oval window with fat, fascia and cartilage augmented with tissue glue. Generally, cul de sac closure is not required in such patients. Hearing rehabilitation should be done by either use of hearing aids or cochlear implants. If cochlear implantation is planned, round window niche can be preserved and marked by using small stent for identification during subsequent surgery. In our patients, one patient did not want any implant and two have been posted for implant after three months from date of completion.

# **CONCLUSION**

Spontaneous CSF otorrhea secondary to inner ear anomaly should strongly be suspected in patients with profound hearing loss since birth presenting with recurrent meningitis. HRCT temporal bone cisternography is gold standard for localizing the leak. Surgical closure of the leak by obliteration with fascia, cartilage and free muscle graft is the preferred line of management.<sup>7,8</sup>

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