

# Cystic cochleovestibular anomaly presenting with congenital deafness and recurrent bacterial meningitis in childhood

Girish Baburao Kulkarni, Roopa S.<sup>1</sup>, Madhu N., Jitender Saini<sup>1</sup>, Ravi Yadav, Veerendrakumar M., Nagaraja D.

Departments of Neurology and <sup>1</sup>Neuroradiology, National Institute of Mental Health and Neurosciences, Bangalore, Karnataka, India

## Abstract

Recurrent bacterial meningitis (RBM) in many instances is associated with identifiable anatomical defects. Presence of congenital deafness with recurrent meningitis should alert clinician for presence of middle and inner ear malformation. These defects can be demonstrated by various neuro imaging techniques and can be surgically corrected. In this case report we describe a child seen at our institute with congenital deafness and recurrent meningitis, discuss the approach to RBM and briefly describe inner ear malformation associated with the same and how to differentiate them.

## Key Words

Congenital deafness, cystic cochleovestibular anomaly, lumbar puncture, recurrent bacterial meningitis

## For correspondence:

Dr. Girish Baburao Kulkarni, Department of Neurology, NIMHANS, Bangalore, Karnataka, India.

E-mail: gbk5000@rediffmail.com

*Ann Indian Acad Neurol* 2012;15:272-5

## Introduction

Bacterial meningitis is a severe, potentially life-threatening infection of the meninges, which carries a significant mortality.<sup>[1,2]</sup> Recurrent bacterial meningitis (RBM) is a relatively rare phenomenon, occurring in about 5–6% of adults and 1.3% of children with community-acquired bacterial meningitis.<sup>[3,4]</sup> The detailed history and directed examination and investigations can help in uncovering the possible underlying abnormality. It is important to identify the cause for recurrence as appropriate treatment prevents further episodes and improves the outcome.

## Case Report

A 5-year-old girl born out of a nonconsanguineous parentage, deaf and dumb from birth with other milestones being normal, was brought to our hospital for evaluation of three episodes of bacterial meningitis.

During the first episode, the child presented to a pediatric general hospital (GH) with high-grade continuous fever and vomiting for 3 days followed by altered sensorium for 2 days. On examination, the child had mild left orbital cellulitis, was drowsy, and meningeal signs were present without features of raised intracranial pressure or focal deficits. Routine investigations were done which showed mild anemia; computerized tomography (CT) of brain showed diffuse cerebral edema with left ethmoidal sinusitis and orbital cellulitis on the left side. Lumbar puncture (LP) revealed turbid cerebrospinal fluid (CSF) with 350 cells (90% polymorphs, 10% lymphocytes), protein of 780 mg/dl and glucose less than 10 mg/dl. Gram stain was negative, but CSF culture showed growth of *Streptococcus pneumoniae* sensitive to penicillin, gentamycin, cefotaxime, ceftriaxone, vancomycin, and ciprofloxacin. She was treated with sensitive antibiotics for 2 weeks and she recovered. She was referred to our institute by the pediatrician who treated her during the first episode, but the patient's father got her discharged within 12 hours because of social problems.

She presented once again to the same pediatric hospital after 4 months with high-grade, continuous fever and headache of 3 days duration. On examination, she was conscious and had meningeal signs with no focal deficits. Routine investigations were normal except for mild anemia. CSF showed 250 cells (100% polymorphs), protein of 65 mg/dl, and glucose less than 20 mg/dl. Gram stain was negative; culture did not grow

### Access this article online

#### Quick Response Code:



#### Website:

www.annalsofian.org

#### DOI:

10.4103/0972-2327.112496

any organism. She received injectable crystalline penicillin and ceftriaxone for 14 days and became fully asymptomatic.

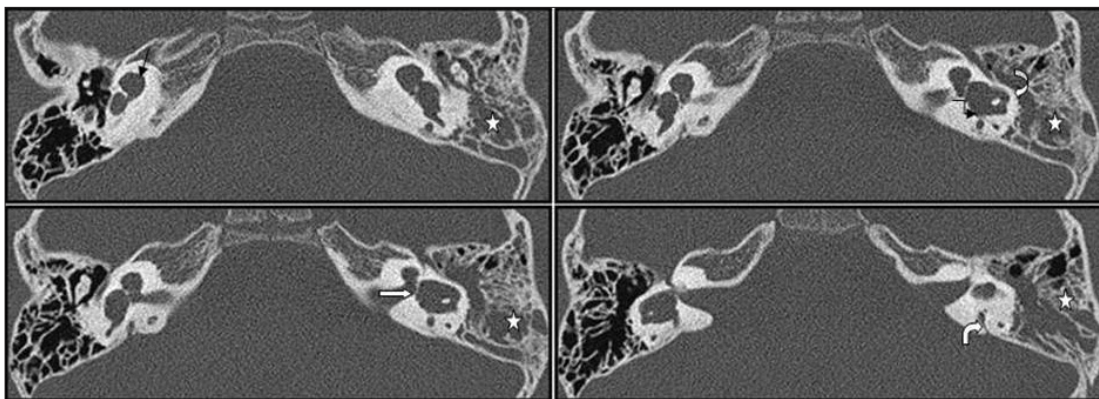
Approximately 1 month later, she once again developed high-grade, continuous fever, vomiting, and altered sensorium of 2 days duration. This time, the child was referred to our hospital for evaluation and treatment, without being admitted at GH. On examination, the child was drowsy and meningeal signs were present without any features of raised intracranial tension or focal deficits. Lumbar puncture revealed turbid CSF with cell count of 2000 (90% polymorphs), protein of 168 mg/dl, and glucose of 23 mg/dl. Pneumococcal antigen was positive in the CSF. Blood and CSF cultures were negative. She was treated with ceftriaxone for 2 weeks in meningitic doses and improved totally. There was no history of any ear discharge, rhinorrhea, head injury, and recurrent respiratory or skin infection. On examination, there was no splenomegaly, sinus, or tuft of hair over spine. Her routine investigation showed mild anemia, leukocytosis, normal biochemistry, and nonreactive HIV test. Brainstem-evoked auditory responses were absent bilaterally with audiometry suggestive of bilateral severe sensory neural hearing loss.

She was evaluated with high-resolution computerized tomography (HRCT) scan of the temporal bone and magnetic resonance imaging (MRI) of the brain. On the left side, the cochlea was showing a featureless single cavity. There was complete absence of modiolus and interscalar septa. The vestibule was grossly dilated. The cystic cochlea and vestibule together gave a

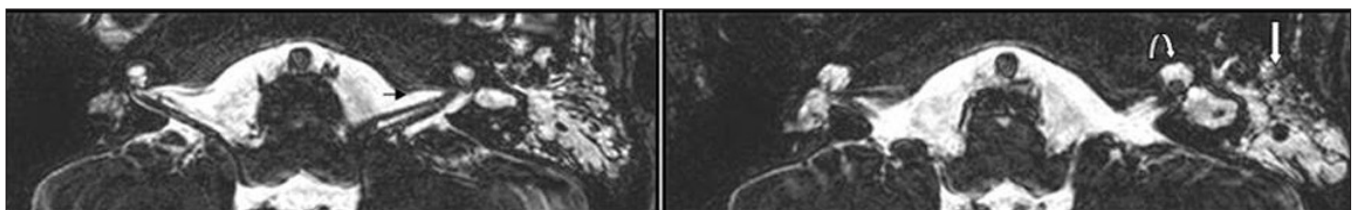
“figure-of-eight” appearance. The lateral semicircular canal was dysplastic and dilated. Superior and posterior semicircular canals were normal. Internal auditory canal (IAC) was dilated and contained normal facial and vestibulocochlear nerves. The vestibular aqueduct was normal. The left tympanum and the mastoid air cells were filled with soft tissue density, indicating either CSF leakage or otitis media with mastoiditis [Figures 1 and 2].

The cribriform bony plate between the cochlea and IAC was deficient, producing an open fundus communicating with the cochlea. This could be a probable site of communication between the subarachnoid space and inner ear. CT cisternography, which reveals the abnormal transotic CSF leak, was not performed in our case. However, the oval and round windows were wider. These could be the probable sites of communication between the inner ear and middle ear. The middle ear ossicles (malleus, incus, and stapes) and tympanic membrane were normal. The right inner ear also revealed similar abnormality. However, the tympanum and mastoid were normal.

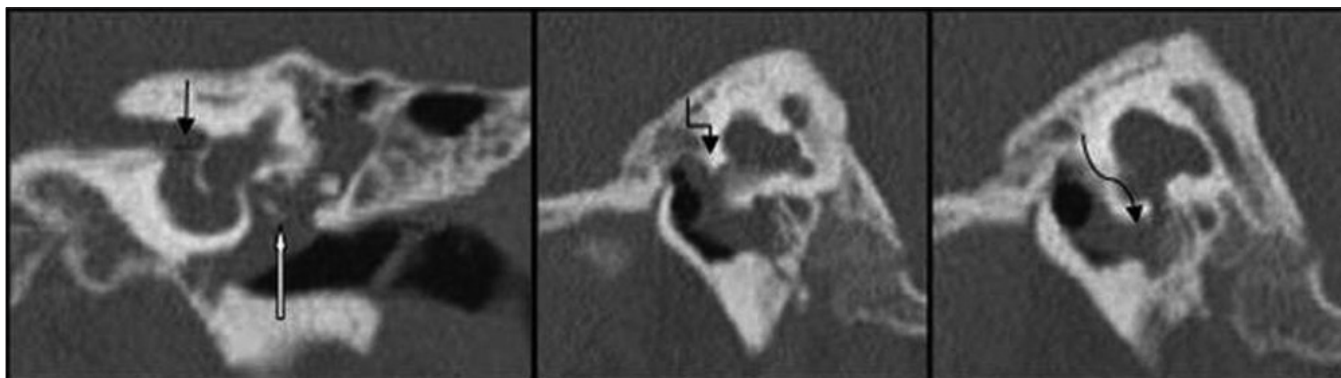
With these features, the patient was diagnosed as a case of congenital deaf-mutism with RBM, cystic vestibulocochlear anomaly, and probable infection arising from left middle ear cavity. She was evaluated by the ENT surgeon, and after 1 month of the last episode, she underwent left middle ear exploration and a communication at the stapes foot plate with inner ear was identified and closed. The patient has had no



**Figure 1:** HRCT of temporal bone. The cystic cochlea and vestibule together give a “figure-of-eight” appearance. The cochlea (line arrow) has a featureless single cavity due to complete absence of modiolus and interscalar septa. The vestibule (zigzag arrow) is grossly dilated. The lateral semicircular canal (curved block arrow) is dysplastic and dilated. The cribriform plate (straight block arrow) between the cochlea and internal auditory canal is deficient and the IAC is dilated. However, the vestibular aqueduct (right angled block arrow) is normal. The left tympanum and the mastoid air cells (star) are filled with soft tissue density, indicating CSF leakage. The right tympanum and mastoid are normal (air density)



**Figure 2:** MRI CISS constructive interference in steady state 3D of temporal bone . Normal facial and vestibulocochlear nerves (line arrow) are seen in the dilated internal auditory canal. Cystic cochlea (curved arrow), dilated vestibule, and dysplastic lateral semicircular canal are evident. Left mastoid air cells (straight block arrow) show T2 hyperintensity suggestive of fluid. Right mastoid air cells are normal



**Figure 3: Probable site of translabyrinthine fistulae. HRCT of the temporal bone. An open fundus (straight arrow) of the IAC, communicating with the cochlea due to absent cribriform plate, which could be a probable site of communication between the subarachnoid space and inner ear. Wide oval window (zigzag arrow) and round window (curved arrow) – probable sites of communication between the inner ear and middle ear. Stapes is indicated with straight block arrow**

recurrences from the last 1½ years.

## Discussion

Our patient presented with three episodes of bacterial meningitis, of which on two occasions pneumococcus was the etiological agent. Even though the patient had deafness from childhood, this clue was not considered and routine CT scan did not help in the identification of the inner and middle ear abnormalities in our patient during the initial evaluation. Subsequent history examination and investigations done at our institute helped us to diagnose the condition and it was followed by definitive surgery which has prevented the recurrences.

RBM is defined as two separate episodes of meningitis separated by a period of convalescence and full recovery; to be more precise, two or more episodes which are caused by different bacterial organisms or by the same organism separated by an interval of 3 weeks after the completion of the therapy of the initial episode.<sup>[5]</sup> The predisposing causes for RBM are broadly classified into congenital and acquired causes and which may be due to anatomical defects, immunodeficiencies, and parameningeal infections. 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.<sup>[5,6]</sup>

Children who present with meningitis and hearing disturbances should be evaluated for developmental malformations of the inner ear. The most common organism associated with meningitis is *S. pneumoniae*, followed by *Haemophilus influenzae* and *Neisseria meningitidis*.<sup>[7]</sup> The inner ear malformations present with first episode in early childhood. Although there are many case reports or small series of RBM associated with inner ear malformations, in majority of them, the underlying defect is Mondini dysplasia or the abnormality is unspecified.<sup>[5]</sup> Our patient had cystic vestibulocochlear anomaly; it is also called as incomplete partition type I or

unpartitioned cochlea. It is a rare inner ear abnormality comprising less than 2% of all labyrinthine anomalies. The defect occurs due to arrest in otic capsule development during the 5<sup>th</sup> week of intrauterine life. It is characterized by a cystic featureless cochlea due to complete absence of modiolus and interscalar septa. There will be gross vestibular dilatation. The cystic cochlea and vestibule together give a “figure-of-eight” configuration. The cribriform plate between the cochlea and IAC will be defective. The IAC will be dilated. Semicircular canals can also be dysplastic. However, the vestibular aqueduct will be normal.<sup>[8]</sup> This is a severe anomaly compared to Mondini deformity. Mondini deformity, also called as incomplete partition type II or incompletely partitioned cochlea, occurs due to arrest during the 7<sup>th</sup> week of intrauterine life. It is a more common inner ear anomaly. It has three components: (1) cochlea with lesser number of turns (2–1.5), a normal basal turn and a cystic apex caused by fusion of middle and apical turns; (2) an enlarged vestibule; and (3) a large vestibular aqueduct containing a dilated endolymphatic duct and sac. Modiolus is present. Although both anomalies have dilated vestibules, it is greater in incomplete partition type I.<sup>[8]</sup>

Inner ear malformations are associated with recurrent meningitis due to CSF otorrhea occurring through abnormal connections between the subarachnoid space and middle ear, which could be either translabyrinthine or perilyabyrinthine.<sup>[9,10]</sup> In the commoner translabyrinthine type, there is abnormal connection between subarachnoid space and inner ear, and between inner ear and middle ear. The most common pathologic interconnection between the subarachnoid space and inner ear is through the fundus of the IAC due to absence of cribriform plate which separates the distal IAC from labyrinthine lumen. Contrast CT cisternography can reveal leak of contrast into the inner ear from subarachnoid space.<sup>[11,12]</sup> Leak can also occur around the facial nerve or through the cochlear aqueduct. The abnormal connections between the inner and middle ears can be through oval window, round window, or a fissure on the promontory.<sup>[10,11]</sup> The middle ear can get infected by the spread of infection from nasopharynx through eustachian tube. Perilymphatic fistula, an abnormal connection between the inner ear and tympanic cavity, can also be associated with inner ear malformations. However, clinically, they present with sudden or progressive

hearing loss and vertigo instead of recurrent meningitis.

In our patient, the probable site of translabyrinthine fistulae was identified on imaging [Figure 3]. An open fundus of the IAC communicated with the cochlea due to absent cribriform plate, which could be a probable site of communication between the subarachnoid space and inner ear. Wide oval window and round window were the probable sites of communication between the inner ear and middle ear. This finding was confirmed during surgery and it was closed which prevented recurrences. We have not evaluated the immunoglobulin and complement levels in our patient because these diagnoses are unlikely, considering her clinical profile.

## Conclusion

In case of RBM, early diagnosis of the underlying pathology is imperative to prevent further episodes and improve the overall outcome. The age of the patient, the organism isolated, and clinical features usually point to the predisposing condition. Children who present with RBM and sensorineural hearing loss should be evaluated for developmental malformations of the inner ear. High-resolution CT or MRI provides excellent images that confirm the diagnosis.

## References

- Durand ML, Calderwood SB, Weber DJ, Miller SI, Southwick FS, Caviness VS Jr, et al. Acute bacterial meningitis in adults. A review of 493 episodes. *N Engl J Med* 1993;328:21-8.
- Sigurdardottir B, Bjornsson OM, Jonsdottir KE, Erlendsdottir H, Gudmundsson S. Acute bacterial meningitis in adults. A 20-year overview. *Arch Intern Med* 1997;157:425-30.
- Adriani KS, van de Beek D, Brouwer MC, Spanjaard L, de Gans J. Community-acquired recurrent bacterial meningitis in adults. *Clin Infect Dis* 2007;45:e46-51.
- Drummond DS, de Jong AL, Giannoni C, Sulek M, Friedman EM. Recurrent meningitis in the paediatric patient-the otolaryngologist's role. *Int J Pediatr Otorhinolaryngol* 1999;48:199-208.
- Tebuegge M, Curtis N. Epidemiology, Etiology Pathogenesis, and Diagnosis of Recurrent Bacterial Meningitis. *Clin Microbiol Rev* 2008;21:519-37.
- Kline MW. Review of recurrent bacterial meningitis. *Pediatr Infect Dis J* 1989;8:630-4.
- Ohlms LA, Edwards MS, Mason EO, Igarashi M, Alford BE, Smith RJ. Recurrent meningitis and Mondini dysplasia. *Arch Otolaryngol. Head Neck Surg* 1990;116:608-12.
- Sennaroglu L, Saatci I. Unpartitioned versus incompletely partitioned cochleae: Radiologic differentiation. *Otol Neurotol* 2004;25:520-9.
- Phelps PD. Congenital cerebrospinal fluid fistulae of the petrous temporal bone. *Clin Otolaryngol Allied Sci* 1986;11:79-92.
- Quiney RE, Mitchell DB, Djazeri B, Evans JN. Recurrent meningitis in children due to inner ear abnormalities. *J Laryngol Otol* 1989;103:473-9.
- Barcz DV, Wood RP, Stears J, Jafek BW, Shields M. Subarachnoid space: Middle ear pathways and recurrent meningitis. *Am J Otol* 1985;6:157-63.
- Burton EM, Keith JW, Linden BE, Lazar RH. CSF fistula in a patient with Mondini deformity: Demonstration by CT cisternography. *Am J Neuroradiol* 1990;11:205-7.

**How to cite this article:** Kulkarni GB, Roopa S, Madhu N, Saini J, Yadav R, Veerendrakumar M, et al. Cystic cochleovestibular anomaly presenting with congenital deafness and recurrent bacterial meningitis in childhood. *Ann Indian Acad Neurol* 2013;16:272-5.

**Received:** 07-01-12, **Revised:** 26-01-12, **Accepted:** 30-01-12

**Source of Support:** Nil, **Conflict of Interest:** Nil