International Journal of Contemporary Pediatrics
Saravanamuthu T et al. Int J Contemp Pediatr. 2026 Jul;13(7):1223-1225
https://www.ijpediatrics.com
pISSN 2349-3283 | eISSN 2349-3291

DOI: https://dx.doi.org/10.18203/2349-3291.ijcp20261917

Original Research Article

Congenital cochlear aplasia presenting as recurrent meningitis and cerebrospinal fluid rhinorrhea: a diagnostic challenge

Thirunavukkarasu Saravanamuthu[1], Vivek Balaji[1]*, Janani Sankar[2], Padmapriya Kaveri Haribabu[3], Gopinathan Kathirvelu[4]

[1]Department of Pediatric Otorhinolaryngology and Airway Surgery, Kanchi Kamakoti Childs Trust Hospital, Nageswara Road, Nungambakkam, Chennai, Tamil Nadu, India
[2]Kanchi Kamakoti Childs Trust Hospital, Nageswara Road, Nungambakkam, Chennai, Tamil Nadu, India
[3]Department of Pediatric Medicine, Kanchi Kamakoti Childs Trust Hospital, Nageswara Road, Nungambakkam, Chennai, Tamil Nadu, India
[4]Scans World, Rama Street, Valluvarkotam High Road, Nungambakkam, Chennai, Tamil Nadu, India

Received: 03 April 2026
Accepted: 16 June 2026

***Correspondence:**
Dr. Vivek Balaji,
E-mail: viv.cool18@gmail.com

Copyright: © the author(s), publisher and licensee Medip Academy. This is an open-access article distributed under the terms of the Creative Commons Attribution Non-Commercial License, which permits unrestricted non-commercial use, distribution, and reproduction in any medium, provided the original work is properly cited.

ABSTRACT

Cerebrospinal fluid (CSF) otorrhea presenting as "pseudo-rhinorrhea" via the Eustachian tube is a rare and deceptive diagnostic entity. In pediatric patients, congenital inner ear malformations can cause this phenomenon, often leading to misdiagnosis, treatment delays, and life-threatening recurrent meningitis. This case highlights the importance of distinguishing otogenic sources in patients presenting with apparent rhinorrhea. A 3-year-old boy presented with persistent clear watery nasal discharge and a history of recurrent bacterial meningitis. Initial brain MRI failed to identify an anatomical defect or leak source. However, a focused history revealed prior ear pain, prompting targeted imaging. High-resolution CT (HRCT) and CT cisternography identified a complex right-sided congenital malformation, including cochlear aplasia, a cystic vestibule, and an absent stapes footplate (Type I leak). The diagnosis of CSF otorrhea manifesting as rhinorrhea was confirmed as contrast tracked from the inner ear through the middle ear and down the Eustachian tube. The patient underwent definitive surgical repair via a post-auricular approach, involving a canal wall down mastoidectomy, oval window plugging, Eustachian tube obliteration, and blind sac closure of the external auditory canal. The patient remained symptom-free with no recurrence of meningitis at the 6-month follow-up. Clinicians must consider otogenic sources for apparent CSF rhinorrhea in children with recurrent meningitis. Standard brain MRI may overlook subtle otic capsule defects; therefore, HRCT of the temporal bone and a high index of suspicion for inner ear malformations are essential for accurate diagnosis.

Keywords: CSF otorrhea, Meningitis, CSF rhinorrhea, CT cisternography

INTRODUCTION

Cerebrospinal fluid (CSF) leaks presenting as rhinorrhea are well-recognized, but CSF otorrhea presenting as nasal discharge via the Eustachian tube ("pseudo-rhinorrhea") is a rare diagnostic entity.[1] This condition is particularly challenging to diagnose in children with congenital inner ear malformations, often leading to delays in treatment and recurrent meningitis.

CASE REPORT

A 3-and-a-half-year-old boy presented with recurrent episodes of bacterial meningitis and suspected CSF rhinorrhea. He was normal until 2 years and 9 months of

age, after which he developed his first episode of acute bacterial meningitis (*Pneumococcus* was cultured). He was treated with IV antibiotics and anti-epileptic drugs, recovering after a 27-day hospital stay. Within a week of discharge, he developed another fever with elevated CRP, and the child began to show watery nasal discharge. CSF rhinorrhea was suspected, but MR cisternography was inconclusive for any anterior anatomical defect. The child was stable for 8 months before being admitted again with fever and refractory status epilepticus. CSF PCR was positive for Cytomegalovirus. He continued to have persistent, clear watery nasal discharge. The child was stabilized and referred to our ENT OPD for evaluation of the persistent discharge. Detailed history taking revealed one prior episode of pain and swelling behind the right ear. This finding raised the suspicion of CSF otorrhea manifesting as rhinorrhea.

Management and outcome

The child underwent HRCT of the temporal bone and a CT cisternogram. Brain stem evoked response audiometry (BERA) showed profound sensorineural hearing loss in the right ear and normal hearing in the left. This unilateral SNHL was a key clinical finding. The HRCT showed a right cochlear aplasia, cystic dilatation of the vestibule with dysplastic lateral semicircular canal, and stapes deformity, categorized as an Incomplete partition 1 malformation. The rest of the semicircular canals were well formed. The CT cisternogram was diagnostic, showing the right middle ear and mastoid cavities filled with contrast. The contrast-filled CSF was clearly seen escaping from the inner ear into the right middle ear cavity, with subsequent excretion extending into the choana. The left ear was normal.

The child underwent mastoidectomy, plugging of the defect and eustachian tube along with a blind sac closure of the right ear via a post auricular approach of the right ear via a post-auricular approach. The procedure successfully achieved three key aims:

Oval window plugging

A canal wall down mastoidectomy was performed, middle ear mucosa removed, and the oval window defect was plugged with post-auricular muscle and soft tissue to seal the inner ear communication.

Eustachian tube obliteration

The orifice of the Eustachian tube was sealed using post-auricular muscle and soft tissue to eliminate the CSF conduit.

Blind sac closure

This final step was done to prevent chronic discharge and potential ascending infection. The post-operative period was uneventful. At the six months follow-up, the child remains well, with no recurrence of meningitis or CSF leakage.

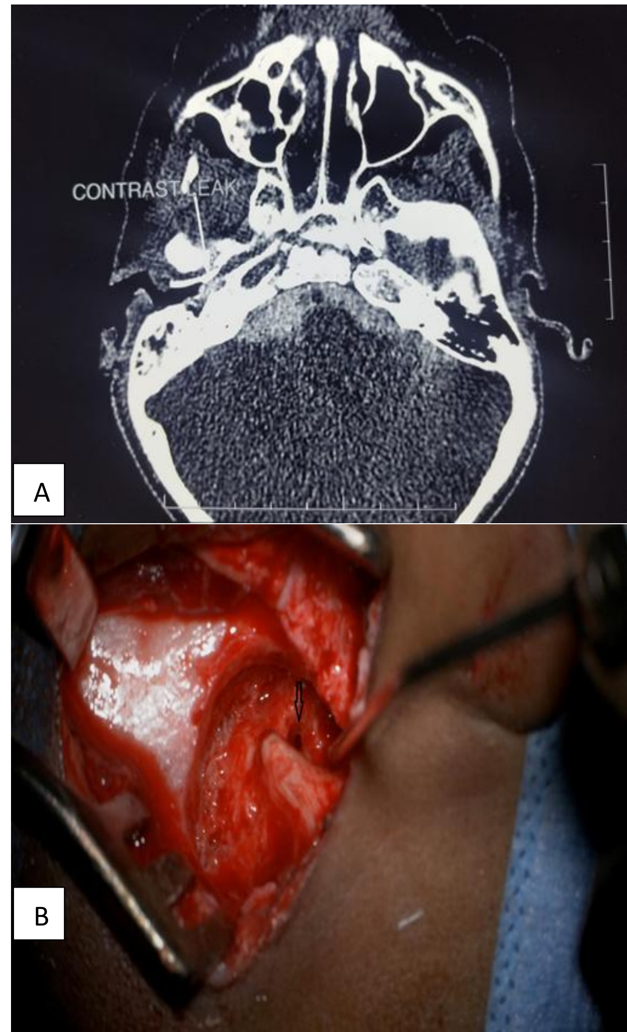

Figure (A and B): A) showing the CT cisternogram picture where the dye leak is seen in the middle ear and B) showing the intra operative defect in the oval window (black arrow).

DISCUSSION

CSF leaks in children, spontaneous or congenital, are typically associated with inner ear malformations that create abnormal communications between the subarachnoid space and the middle ear.[1] These defects predispose patients to recurrent meningitis due to the proximity of the perilymphatic spaces to the subarachnoid space. The most common presentations are usually meningitis, hearing loss or CSF otorrhea or rhinorrhea.[2] Congenital anomalies, resulting from arrested labyrinthine development, often involve a defect at the oval window, round window, or internal auditory canal. CSF otorrhea usually drains externally, but with an intact tympanic membrane, the CSF is forced through the Eustachian tube into the nasopharynx, manifesting as the rare CSF rhinorrhea (pseudo-rhinorrhea). Recurrent

meningitis in a child is a strong indicator of a perilymphatic fistula or CSF leak, a diagnosis often delayed.[3,4] This delay highlights how Eustachian tube-mediated rhinorrhea can be misattributed to a more common anterior skull base defect. The underlying pathology in our patient-cochlear aplasia and an absent stapes footplate represents a severe end of the congenital inner ear malformation spectrum.[5] The vulnerability created by the absent stapes footplate at the oval window allowed for a low-resistance path for CSF egress. Neely classified this condition, where the leak occurs through an abnormal dehiscence in the osseous labyrinth, as a type I leak, the same like in our patient.[6] Furthermore, Lee et al reported a case of congenital oval window atresia with cochlear aplasia that presented similarly with recurrent meningitis, underscoring that these specific malformations carry a high risk of dural communication.[7] The accompanying profound ipsilateral SNHL is a crucial clinical clue, strongly correlated with this severe structural defect and its high risk of dural communication. The diagnostic process highlights the limitation of initial MRI, which is excellent for brain parenchyma but often misses subtle inner ear anomalies without targeted imaging. Defects involving the stapes footplate are subtle and require high-resolution studies.[8] The key to correct diagnosis relies on a high index of suspicion from the detailed history (prior ear pain) and the objective finding of profound unilateral SNHL. The CT cisternography was essential for definitive diagnosis. This technique dynamically visualizes the CSF tracking, confirming the leak originated from the malformed inner ear structures and travelled precisely through the Eustachian tube, establishing the anatomical pathway of this misleading presentation. Surgical repair must achieve permanent and complete closure. Given the patient's profound, non-serviceable SNHL, an obliterative procedure was the preferred management. Our technique of oval window plugging combined with Eustachian tube obliteration aligns with protocols for high-flow congenital leaks in an ear with non-serviceable hearing.[9,10] The obliteration of the Eustachian tube orifice is critical in these rhinorrhea cases, as it eliminates the CSF conduit and, more importantly, prevents the ascending infection route from the nasopharynx to the meninges. The successful outcome and absence of meningitis recurrence at follow-up supports the efficacy of this definitive closure strategy.

CONCLUSION

Congenital CSF otorrhea manifesting as rhinorrhea is a rare and frequently misdiagnosed cause of recurrent meningitis in children. This case of cochlear aplasia and absent stapes footplate confirms that a high index of suspicion, a thorough history, and targeted imaging (HRCT temporal bone and CT cisternography) are non-negotiable for accurate diagnosis. Definitive surgical management with oval window plugging and Eustachian tube obliteration provides a permanent resolution, preventing potentially fatal recurrences.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Zwierz A, Masna K, Burduk P. Recurrent meningitis in congenital inner ear malformation. Ear Nose Throat J. 2021;100(1):38S-41S.
2. Wang B, Dai WJ, Cheng XT, Liuyang WY, Yuan YS, Dai CF, et al. Cerebrospinal fluid otorrhea secondary to congenital inner ear dysplasia: diagnosis and management of 18 cases. J Zhejiang University-Sci B. 2019;20(2):156-63.
3. Sipahimalani M, Wiet RJ. Cerebrospinal fluid fistula and recurrent meningitis in children with congenital inner ear malformations. Otol Neurotol. 2004;25(4):618-623.
4. Kuczkowski J, Niemczyk K, Stankiewicz C, Pilarska E, Szurowska E, Plichta Ł. Lateral petrosectomy with obliteration cavity for spontaneous cerebrospinal otorrhea in children. Am J Otolaryngol. 2014;35(5):651-4.
5. Ghorayeb BY, Jahrsdoerfer RA. Congenital inner ear malformations and associated cerebrospinal fluid fistulae. Otolaryngol Head Neck Surg. 1987;96(3):213-219.
6. Neely JG. Classification of spontaneous cerebrospinal fluid middle ear effusion: Review of forty-nine cases. Otolaryngol Head Neck Surg. 1985;93(5):625-34.
7. Lee D, Chin O, Oh JH, Lee JG. Congenital oval window atresia with cochlear aplasia presenting as recurrent meningitis. Am J Otolaryngol. 2012;33(5):610-613.
8. Weber PC, Gantz BJ, Smith RJ. Congenital defects of the stapes footplate. Am J Otol. 1993;14(1):34-38.
9. Jang CH, Kim SH, Lee YH, Cho YB. A rare case of congenital labyrinthine fistula presenting as CSF rhinorrhea in a 3-year-old child. Acta Otolaryngol. 2018;138(3):285-288.
10. Kimitsuki T, Inamitsu M, Komune S, Komiyama S. Congenital malformation of the inner ear associated with recurrent meningitis. Europ Arch Oto-rhino-laryngol. 1999;256(1):S11-4.

Cite this article as: Saravanamuthu T, Balaji V, Sankar J, Haribabu PK, Kathirvelu G. Congenital cochlear aplasia presenting as recurrent meningitis and cerebrospinal fluid rhinorrhea: a diagnostic challenge. Int J Contemp Pediatr 2026;13:1223-5.