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From Diarrhea to CSF Leak: A Case Report of a Deaf Child

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Abstract
Parenteral diarrhea due to infections outside the gastrointestinal tract is a well-known entity in pediatrics. We report a case where a child with invasive pneumococcal infection presented with parenteral diarrhea and later CSF rhinorrhea, revealing the underlying inner ear malformation. A 15-month-old male child with isolated language delay due to congenital hearing loss presented with pneumococcal septic shock following acute gastroenteritis. Two days later, CSF rhinorrhoea was noted. The Child was stabilised with intravenous fluids and antibiotic therapy. Computed tomography imaging revealed a congenital bony inner ear malformation (IEM). Recurrent meningitis can occur due to communication between IEM and the subarachnoid space. CSF otorrhoea presenting as rhinorrhoea can occur due to meningitis with raised intracranial pressure. Children presenting with invasive pneumococcal infection and sensorineural hearing loss should be evaluated for predisposing factors, such as cerebrospinal fluid leaks, which can unmask underlying inner ear abnormality.
Keywords: Cerebrospinal fluid rhinorrhea; Congenital inner ear malformation; Paediatric meningitis.

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Introduction

The concept of "parenteral diarrhea," where infections outside the gastrointestinal tract manifest as diarrhea, has been recognized for over a century and remains clinically relevant in pediatrics.[1,2] This phenomenon is often linked to conditions such as otitis media, urinary tract infections, or systemic infections like sepsis.[3] Though uncommon, parenteral diarrhea highlights the importance of considering extra-gastrointestinal causes in children presenting with acute diarrheal illness

Additionally, Cerebrospinal fluid (CSF) rhinorrhea is a rare condition in the pediatric population, often signifying an underlying structural defect, infection, or trauma. Congenital causes of CSF rhinorrhea include persistent nasopharyngeal canals, nasal encephaloceles or meningoencephaloceles, and anatomical defects involving the skull base, temporal bone, or dura mater.[1,4] Among these, inner ear malformations (IEM) that create abnormal communication between the subarachnoid space and the middle ear or nasal cavity are particularly concerning, as they predispose to CSF leaks and increase the risk of recurrent meningitis.[4]

This report presents the case of a 15-month-old male child with bilateral congenital profound sensorineural hearing loss and severe acute malnutrition, who was admitted with acute gastroenteritis and septic shock secondary to invasive pneumococcal infection. During hospitalization, an unexpected finding of CSF rhinorrhea prompted further investigations, which revealed a bony inner-ear malformation characterized by cochlear aplasia and a cystic cochleovestibular malformation. This case underscores the need for thorough evaluation in children with congenital hearing loss and atypical clinical presentations, as it may reveal structural anomalies with significant implications for management and outcomes.

Case Report

A 15-month-old male child, the second-born to non-consanguineously married parents, presented with acute gastroenteritis and compensated shock. His antenatal and perinatal history was uneventful. The child had a significant language delay with a language development quotient of 40% whereas other domains were normal. He had not received the pneumococcal conjugate vaccine (PCV), despite being immunized according to the National Immunization Schedule.

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Anthropometric evaluation revealed severe acute malnutrition (SAM), with weight-for-height less than -3 standard deviations. Previously, the child had been evaluated for language delay, which revealed bilateral congenital sensorineural hearing loss. However, further evaluation for this condition had not been conducted.

The child was managed with fluid boluses and intravenous antibiotics. Initial investigations revealed normal blood counts, metabolic acidosis with compensated respiratory alkalosis, and elevated erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP). Serum electrolytes, blood sugars, liver, and renal function tests were within normal limits. Blood culture identified *Streptococcus pneumoniae* sensitive to ceftriaxone.

On the third day of admission, the child was transferred from the ICU to the ward, where continuous watery nasal discharge from the dependent nostril was observed. An otorhinolaryngologist made a spot diagnosis of cerebrospinal fluid (CSF) rhinorrhea, which was confirmed by nasal fluid analysis with glucose levels of 51 mg/dL (approximately two-thirds of the patient's blood glucose level), white blood cell counts of 75 cells/mm^3, and no microbial growth on culture.

Considering the possibility of meningitis leading to CSF rhinorrhea, as well as primary nasal or otological causes, a detailed history review revealed bilateral congenital profound hearing loss diagnosed via brainstem evoked response audiometry (BERA). The child was on the waiting list for cochlear implantation at another hospital, but imaging of the temporal bones and inner ears had not yet been performed.

The history of profound hearing loss raised suspicion of an inner ear malformation (IEM) as the source of meningitis. Bilateral high-resolution computed tomography (HRCT) of the temporal bones revealed bony IEM, with findings suggestive of cochlear aplasia and cystic cochleovestibular malformation.

Given the diagnosis of pneumococcal sepsis with suspected meningitis, ceftriaxone was continued for 14 days. CSF rhinorrhea was managed conservatively and gradually resolved within a few days. The child remained clinically stable and was scheduled for further imaging and evaluation to assess the potential for surgical intervention of the malformed inner ear as part of hearing rehabilitation.

Discussion

CSF rhinorrhea occurs due to a defect in the anterior skull base, resulting in an aberrant communication between the subarachnoid space and the nasal cavity, causing cerebrospinal fluid leakage.[5] Congenital hearing loss associated with inner ear malformations (IEM) may arise from either membranous (80%) or bony (20%) abnormalities. Membranous abnormalities involve defects in the inner ear hair cells. They are typically characterized by normal bony structures on high-resolution computed tomography (HRCT) and normal eighth cranial nerve findings on magnetic resonance imaging (MRI). In such cases, cochlear implantation (CI) is the preferred surgical approach for hearing rehabilitation.

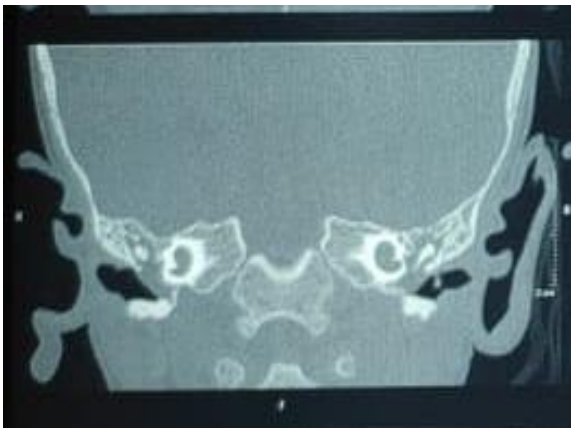

Figure 1. HRCT showing incomplete cochlea and cystic cochleovestibular malformation (original).

In contrast, bony abnormalities of the bony labyrinth can be radiologically detected with HRCT and MRI. These malformations present unique challenges in surgical planning and decision-making. Options for hearing rehabilitation in these cases include hearing aids, CI, or auditory brainstem implants (ABI), depending on the extent of the malformation, the involvement of the subarachnoid space, and the presence of the eighth cranial nerve. Bony abnormalities with direct communication to the subarachnoid space are associated with a significantly increased risk of meningitis.[4]

Congenital bony IEMs are broadly classified into several types, including complete labyrinthine aplasia (Michel's deformity), rudimentary otocyst, cochlear aplasia, common cavity deformity, cochlear hypoplasia (incomplete partitions I, II, and III), enlarged vestibular aqueduct, and cochlear aperture abnormalities.[4] In our patient, cochlear aplasia with a dilated vestibule (CADV) or common cavity (CC) deformity is suspected. Further imaging with HRCT and MRI is essential to differentiate between these two conditions by examining the membranous labyrinth, eighth cranial nerve, and the extent of subarachnoid communication.[4]

In CADV, the vestibule and semicircular canals are dilated, the internal auditory canal is underdeveloped, and functional stimulation is not feasible, making CI contraindicated; ABI is the preferred option. In CC deformity, the internal auditory canal is typically well developed, and the single cavity representing both the cochlea and vestibule may contain functional neural

structures, allowing the possibility of CI.[4] However, distinguishing these malformations can be challenging, even with imaging, and hearing outcomes may remain uncertain until after surgery. Family counseling regarding the potential need for contralateral ABI is essential.

Inner ear abnormalities have also been associated with genetic conditions, such as trisomies 13, 18, and 21, intrauterine exposure to infections like rubella, or teratogenic drugs such as thalidomide. Non-syndromic familial cases have also been reported.[6] In this case, there was no history of intrauterine infection, drug exposure, or dysmorphic features indicative of trisomies.

Our patient, who had congenital bilateral sensorineural hearing loss, presented with acute diarrhea, fever, septic shock, and Streptococcus pneumoniae bacteremia. Although lumbar puncture was deferred due to the patient's critical condition, nasal fluid analysis revealed glucose consistent with CSF, even though beta-2 transferrin, the gold standard test, [7] was unavailable. Although the nasal fluid culture was sterile, the patient completed an antibiotic regimen for meningitis.

Imaging plays a crucial role in diagnosing and managing congenital IEMs. [4-8] In children with sensorineural hearing loss and invasive pneumococcal infections, clinicians should consider the possibility of congenital bony IEMs, which may lead to meningitis and CSF leaks manifesting as oto-rhinorrhea. CSF leaks may be overlooked by caregivers and missed clinically if not actively investigated.

To prevent recurrent invasive pneumococcal infections, surgical intervention tailored to the specific malformation is critical, in addition to pneumococcal vaccination. For this patient, further HRCT and MRI are necessary to identify bony defects, evaluate the membranous labyrinth, and confirm the presence of the eighth cranial nerve. If the nerve is present, CI with selective electrode placement may be possible. In its absence, ABI becomes the only option. However, ABI feasibility depends on cost, surgical expertise, and postoperative hearing rehabilitation, which may pose additional challenges.

Lessons Learnt

Acute CSF rhinorrhea in a child with congenital hearing loss can be a rare but critical presentation of congenital bony inner ear abnormalities.

Preventive strategies, including pneumococcal vaccination and prompt management of upper respiratory tract infections, are crucial to reducing the risk of recurrent infections while awaiting definitive surgical intervention for hearing rehabilitation.

Conclusion

In this case, a child with isolated language delay due to congenital hearing loss presented with parenteral diarrhea and CSF rhinorrhea as the primary manifestations, which ultimately led to the diagnosis of an underlying inner ear malformation (IEM). Early recognition of such presentations and timely diagnosis of underlying IEMs are essential for preventing further invasive infections and enabling earlier hearing rehabilitation.

Acknowledgments

None.

Competing Interests

The authors declare no conflict of interest.

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