

Case Report

In Patients with Unexplained Non-Epidemic Recurrent Meningitis a Direct Communication between the Subarachnoid Space and the Outside Must Be Sought

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Cerebrospinal fluid (CSF) rhinorrhea is not an uncommon condition, and it may lead to recurrent attacks of meningitis. The detection of the leakage site is an essential part of the investigations performed for the patient. In some cases, an anterior skull base site cannot be detected despite continuous leakage. In these cases, a more distal site must be sought. Furthermore, in some cases, the leakage occurs in the temporal bone. In such patients, a history of hearing loss or previous ear complaints indicates a hidden CSF otorhinorrhea. We present two adult cases of congenital hearing loss due to unilateral inner ear anomalies who presented later in life with repeated meningitis attacks and CSF otorhinorrhea.

KEYWORDS: CSF rhinorrhea, meningitis, CSF otorrhinorrhea

INTRODUCTION

In patients with unexplained non-epidemic recurrent meningitis a direct communication between the subarachnoid space and the outside must be sought. The commonest pathway is usually through the nose and paranasal sinuses. A diligent search for the leakage site must be performed to enable the surgeon to effectively seal the defect. However, in some patients, the leakage site is not obvious. In such cases, a distal site must be investigated; leakage from the temporal bone, causing cerebrospinal fluid (CSF) otorhinorrhea, is commonest among such cases ^[1-3]. CSF otorrhea usually occurs due to trauma or complicated chronic ear disease, or it may be congenital. Congenital CSF otorrhea can occur through an abnormal labyrinth, a tegmen defect, or developmental defects [the petromastoid canal, a wide cochlear aqueduct, the facial canal, or the tympanomeningeal (Hyrtl's) fissure] ^[4-5].

In some patients, the leak is not readily apparent, and they remain undiagnosed for a long time. In some other patients, CSF otorrhea is misdiagnosed as serous otitis media, and they are treated for this condition until a myringotomy is performed and clear CSF becomes evident ^[5, 6].

We report two cases of adult patients presenting with recurrent meningitis and CSF rhinorrhea.

CASE PRESENTATION

Informed consent was obtained from the patients for the publication of their blinded data (IRB registry CR-ENT 741/17).

Case 1

A 24-year-old woman presented with recurrent meningitis attacks and had a history of CSF rhinorrhea. She was previously investigated for CSF rhinorrhea and underwent a trial of endoscopic repair. However, the treating surgeon informed her that he had failed to localize the leak and that she might need a transcranial approach. The leakage persisted postoperatively, and she ex-

perience another meningitis attack. On recovery, she was referred for a possible endoscopic revision. A new set of radiologic images was obtained; however, a clear leakage site could not be ascertained. On reviewing her radiologic films, fluid density was noticed in the mastoid air cell system. The patient informed that she was deaf on the left ear since birth. Dedicated temporal bone studies revealed an anomalous inner ear with fluid filling the middle ear and mastoid air cells. This was assumed to be the source of CSF rhinorrhea; thus, an exploratory tympanotomy was performed. The middle ear was full

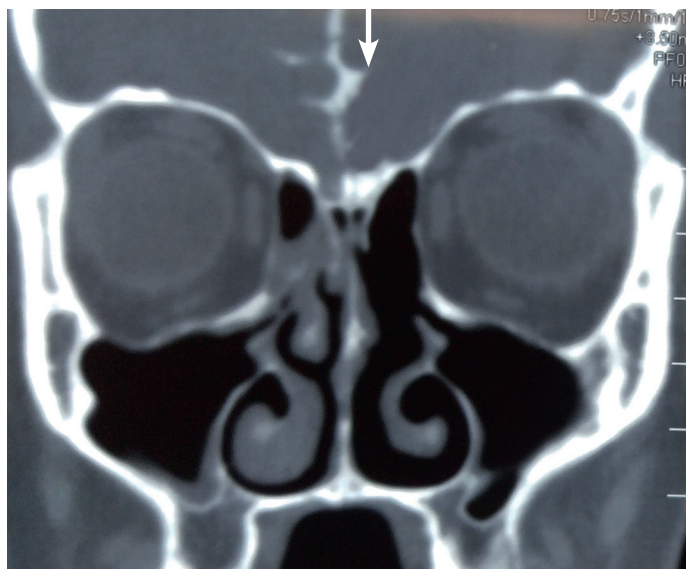


Figure 1. Metrizamide computed tomography of the paranasal sinuses showing evidence of a previous surgery and no apparent cerebrospinal fluid leakage.

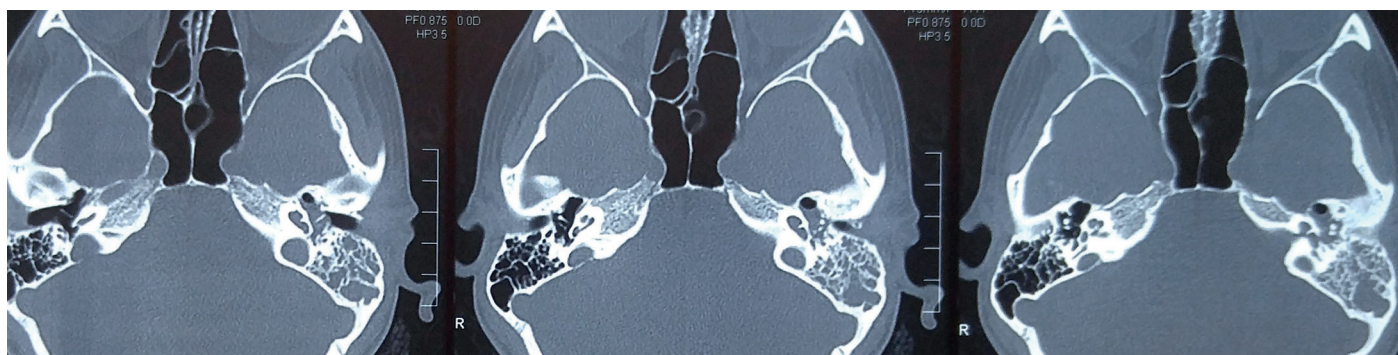


Figure 2. Axial computed tomography image of the temporal bone showing fluid in the middle ear and metrizamide droplets. The inner ear anomaly is also apparent [IP2].

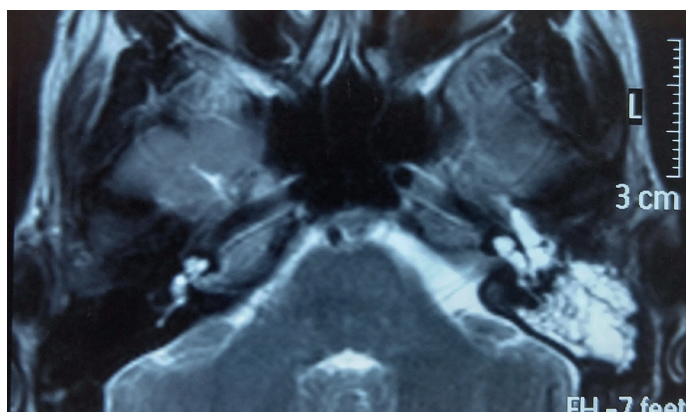


Figure 3. Axial T2-weighted magnetic resonance imaging showing the fluid in the middle ear and mastoid air cells and the anomalous inner ear.

of CSF, and a defect was found in the stapedial footplate with a CSF gusher. The defect was packed with muscle and tissue glue. The leakage stopped, and the patient is now free of CSF rhinorrhea for three years (Figures 1-4).

Case 2

A 34-year-old woman presented with a sudden onset of disturbed consciousness and fever. She was diagnosed with meningoencephalitis with venous sinus thrombosis. She was admitted to the intensive care unit and treated accordingly. On recovery, she reported the drainage of a clear fluid from her nose. The fluid proved to be CSF, and the source of leakage was sought. Due to her venous sinus thrombosis, temporal bone imaging was ordered, and a fluid-filled middle ear and mastoid were observed. This patient also reported that she was deaf on the left ear since birth and that she was not aware that this might be related to her condition. An exploratory tympanotomy was performed that revealed a massive CSF leakage from the round window. The round window was sealed with fascio-adipose plug and glue. The leakage stopped, and the patient has remained leakage-free for 6 months (Figures 5-8).

DISCUSSION

Causing cerebrospinal fluid (CSF) leakage through the paranasal sinuses or the temporal bone is usually silent and remains misdiagnosed for extended periods [4-6]. In some cases, the course is indolent and does not cause significant problems. CSF leakage may be a sign of increased intracranial pressure or may lead to recurrent meningitis attacks with its disabling, or sometimes, fatal sequelae [3, 7, 8]. Even if the morbidity is minimal, it is always important to detect and document the leakage



Figure 4. Intraoperative image showing the hole in the stapedial footplate.

site in order to effectively control the leakage. Various techniques can be used singly or in combination to detect this leakage [6, 9]. In cases with intermittent leakage, patients must be investigated at different times to detect the site. Although the usual sites involve the nose, paranasal sinuses, or anterior skull base, in cases with persistent leakage or negative exploration, other sites must be sought especially through the temporal bone [1, 2, 3, 5]. The presence of hearing loss should also alert the surgeon to the possibility of CSF otorhinorrhea [2].

In the above two cases, recurrent leakage and failed repairs as well as a history of unilateral congenital sensorineural hearing loss directed our attention to a possible temporal bone source. Both patients had congenital inner ear anomalies that were the cause of the leakage. In one case, the hearing loss was attributed to unidentified meningitis during childhood and did not incite any further investigation.

Although it is unusual for congenital CSF leakage secondary to an inner ear anomaly to manifest later in life [8, 10], in cases of CSF rhinorrhea without an apparent leakage site on the anterior skull base, the whole skull base including the temporal bone should be investigated. In such cases, there is usually a history of hearing loss, whether

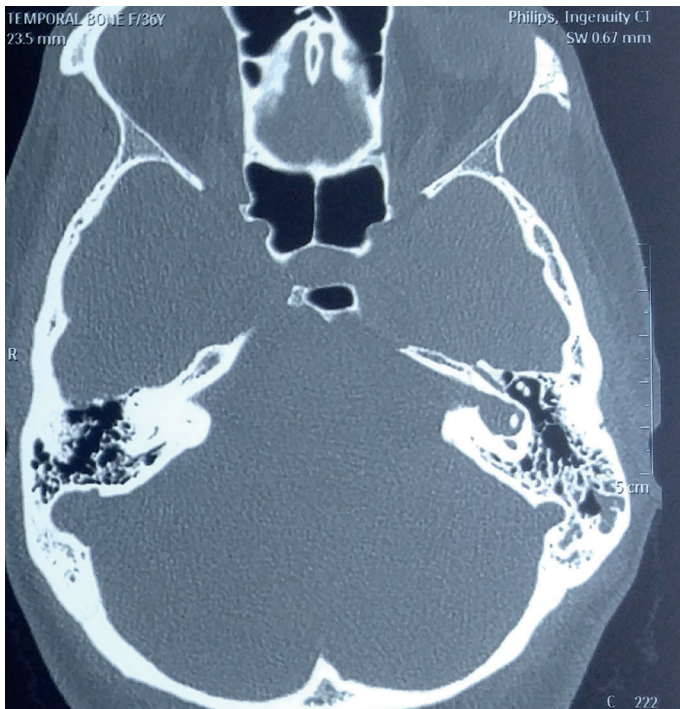


Figure 5. Axial temporal bone computed tomography showing the anomalous inner ear [common cavity].

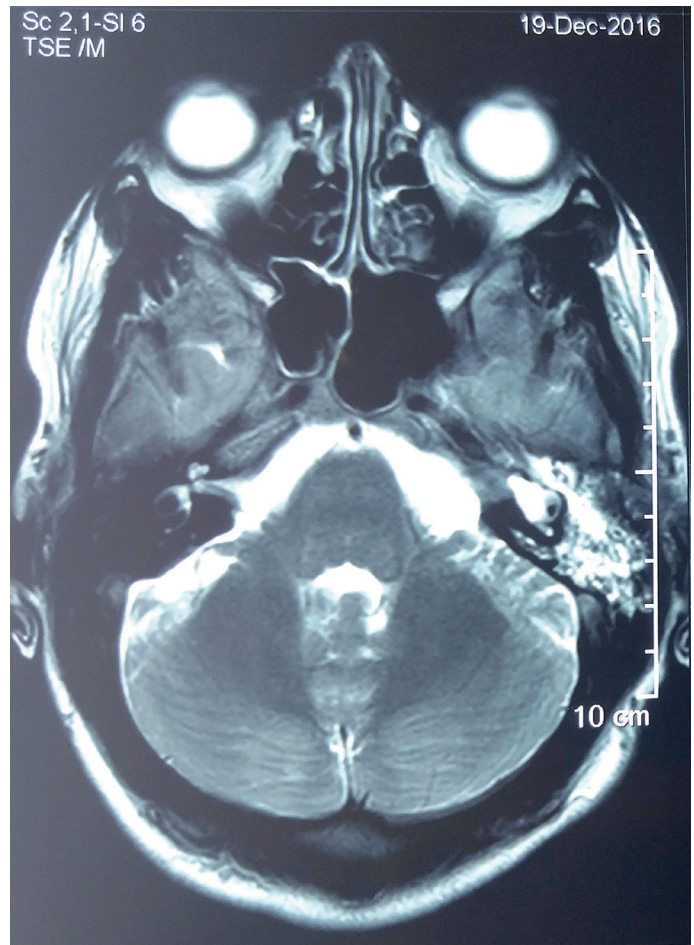


Figure 6. Axial T2-weighted magnetic resonance imaging showing the fluid in the middle ear and mastoid.

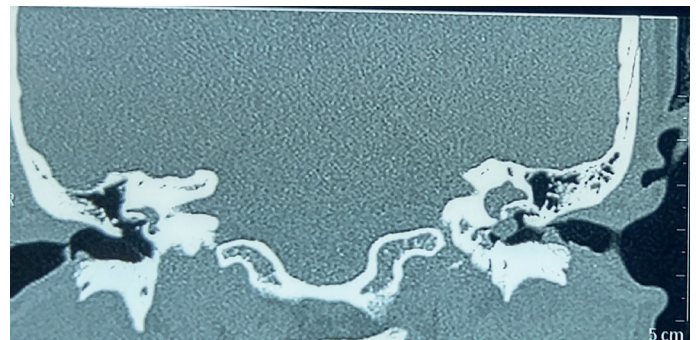


Figure 7. Coronal computed tomography image of the temporal bone showing the "perilymphoceles" around the round window niche.



Figure 8. a-c. Intraoperative views of case 2. a) Perilymphocoele obscuring the round window niche. b) Defect in the round window membrane seeping perilymph/cerebrospinal fluid. c) Fascio-adipose plug sealing the round window defect.

conductive or sensorineural, and the leakage is always posterior, not positional. Stooping forward usually increases the leakage, and at times, it can mimic a bilateral leakage. The leakage in the temporal bone is either through a defect in the windows, a patent fissure, or the tegmen. Usually, repair is best performed using a fat or muscle plug with biological glue. In some cases, CSF leakage occurs through the whole otic capsule; in such cases, the best treatment would be subtotal petrosectomy and fat obliteration of the middle ear cleft [3, 5, 7].

CONCLUSION

In all cases with recurrent unexplained meningitis and a possible CSF rhinorrhea with negative radiological findings, the ear should also be investigated. The presence of a long standing hearing loss and/or an apparent congenital anomaly of the inner ear are important signs to be considered and may point to the ear as a source of the CSF leak.

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