CASE REPORT

Recurrent meningitis due to spontaneous cerebrospinal fluid otorrhea in adults

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Spontaneous cerebrospinal fluid (CSF) fistula in congenital malformations of the inner ear is rare, and it is found most commonly in infants and children with impaired hearing. Spontaneous CSF otorrhea in adults is often due to defects in the tegmen and the bone overlying the posterior fossa in an otherwise normal ear. In the literature, there are few reports of adult-onset spontaneous CSF otorrhea in congenital inner ear malformations. In this article, we report on an adult case with pediatric-pattern presentation.

A 27-year-old patient with congenital hearing loss was referred to our center because of recurrent meningitis. Computed tomography scan revealed an inner ear anomaly, and surgical exploration found an oval window. The fistula was successfully closed using a minimally invasive nonablative technique with a muscle plug that did not violate inner ear structures.

Congenital inner ear disorders are developmental abnormalities that can either remain asymptomatic or present as congenital deafness, progressive hearing loss, vestibular disorders, or developmental motor deficits. About 20% of patients have anomalies in the bony labyrinth, detectable by computed tomography (CT) scan. These patients may have leakage of cerebrospinal fluid (CSF) to the middle ear and, sometimes, to the ear canal or nasopharynx, a patent path for infection that can result in meningitis.[1] This clinical picture (congenital hearing loss, inner ear anomaly, and recurrent meningitis) is primarily seen in children and infants.[2]

Adults with spontaneous CSF otorrhea are mostly overweight or obese, middle-aged or elderly women.[3] They have bony defects in tegmen tympani or tegmen mastoideum. These defects result from CSF pulsation over years. Patients usually present with fluid in the middle ear and conductive hearing loss.[4] We report on a young adult with pediatric-pattern spontaneous CSF otorrhea, which is rare in the medical literature.

CASE REPORT

A 27-year-old woman with history of congenital hearing loss and recurrent meningitis was referred to our center. She had had 3 episodes of meningitis at one year, one month, and two weeks prior to referral, all of which required hospitalization and treatment with parenteral antibiotics. During the past year, she had had intermittent watery discharge in her left ear that didn't change with Valsalva maneuver or positional adjustments. She had no postnasal secretion or rhinorrhea. The only significant finding in her medical history was congenital bilateral hearing loss; there was no family history of hearing loss. She was taking no medication other than recently prescribed antibiotics.

Although she was wearing a hearing aid in her left ear, she couldn't recognize words. She read lips, and the hearing aid just helped her with sound detection. Otoscopy revealed fluid behind the intact drum in the left side. Audiometry disclosed only faint air-conduction responses at 100 to 110 db symmetrically. A speech reception threshold test detected no response. Tympanometry of the right ear was type A and of the left ear, type B. Overall, hearing in the left ear seemed subjectively slightly better than the right ear, although, again, only in detecting sound source not in recognizing words.

High resolution CT scan (HRCT) showed anomalous cochlea, dysplastic semicircular canals, and a bony defect at the lateral end of internal auditory canal (IAC) that was more significant in the left ear. Opacity due to fluid accumulation was seen in the left middle ear and mastoid air cells (Figure 1).

We suspected CSF fistula and explored the left middle ear under general anesthesia. We elevated the tympanomeatal flap transcanally, which immediately triggered a large flow of clear fluid. The tympanic membrane was very thin at some points. Temporary microperforations as a result of intermittent increases in the pressure from middle ear fluid were the apparent explanation for the on-and-off watery discharge that the patient had been experiencing from her ear. After suctioning, the source of fluid outflow was found to be at the central part of the oval window. Other areas in the middle ear were carefully inspected but showed no evidence of leakage. Because the patient wanted to retain her minimal hearing in this ear, we preserved the stapedius.

After stripping the mucosa from the oval window and surrounding area, the defect in the central part was sealed with a muscle graft harvested from the temporalis muscle. The muscle graft was packed between, superior, and inferior to the stapedial crura. The flow stopped. The patient was placed in the Trendelenburg position, and positive pressure ventilation was applied. No leak occurred. Believing that the round window could be another site of leakage, we decided to seal it with muscle to prevent future problems.

Because the leakage stopped and CT images revealed no evidence of semicircular canal fistula, we didn’t explore the mastoid cavity and semicircular canals. Imaging of the right ear, however, showed deformities similar to those in the left ear, so we continued surgical exploration. The right middle ear

was normal without evidence of abnormality in the foot plate or other areas. We prophylactically sealed the round and oval windows in the right ear. The patient was discharged after an uneventful 4 days in the hospital on bed rest and acetazolamide. She was followed for 6 months with no significant problem.

DISCUSSION

CSF otorrhea is classified as pediatric or adult. Children present mostly with congenital hearing loss, inner ear anomaly, and recurrent meningitis.[5] Adult CSF otorrhea can be further divided into three types: otorrhea following trauma, otorrhea with congenital defects in the temporal bone overlying the posterior fossa or tegmen, and otorrhea with inner ear anomalies and recurrent meningitis. Our patient fell into this last group.

Otorrhea following trauma is the most common form and usually results from temporal bone fractures or skull base surgeries. Some temporal bone diseases, such as tumor or cholesteatoma, can cause CSF leakage per se or subsequent to surgical resection.

Throughout the lives of patients with otorrhea and congenital defects in the temporal bone overlying the posterior fossa or tegmen, constant pulsation of CSF and intermittent physiologic pressure changes gradually cause enlargement of bony defect, herniation of the dura with or without brain prolapse, and, ultimately, small perforations of the dura and onset of leakage.[6] A survey of 10 patients with this type of CSF otorrhea revealed that most were women in their 60s, who presented with serous otitis; conductive hearing loss; and, less common, otorrhea, rhinorrhea, and meningitis.[3]

Otorrhea in patients with inner ear anomalies and recurrent meningitis is similar to pediatric otorrhea, but, for reasons unknown, it remains asymptomatic until adulthood. This form is quite rare, and few cases have so far been reported. Gundense and Hayes reported one case of this type with 36 attacks of meningitis at the age of sixty one .[7]

For CSF otorrhea to occur, abnormal paths between the subarachnoid space and inner ear and between the middle and inner ear must be present.[8] The interconnections between the subarachnoid space and inner ear include the IAC fundus, the cochlear aqueduct, and the perineural passages, with abnormality in the IAC fundus the most common. Evidence of cochlear aqueduct involvement, however, has been disputed.[9]

Patients with IAC fundus abnormality usually have a defect in the bony plate that separates the IAC from the inner ear, which can be seen on axial CT scan. Semicanal anomalies are a common cofinding in these patients. A common cavity and its variants may also be present.

The connections between the middle and inner ear include the oval window, round window, and promontorium. Oval window connection is the most common. Sometimes fistulas are located around this window, mostly anterior to it. In a study of more than 331 temporal bones, microfissure in the promontorium was found in 25% of patients.[10]

CT scan can reveal the bony defects, and T2-weighted magnetic resonance imaging (MRI) can precisely demonstrate leakage and its route.(11,12) Treatment depends on hearing status. In those with serviceable hearing, some advocate a short-term conservative approach including bed rest and even lumbar drainage.[13] Some otologists proceed directly to the next step, surgical intervention.

If the conservative treatment fails in an ear with good hearing, the best choice would be middle ear exploration and sealing of the detected site of leakage, using muscle and other connective tissue. If this also fails, some suggest posterior fossa craniotomy and sealing the IAC,[9] although re-exploring the middle ear and a second attempt to seal the leakage seems to be a wise option. Using a lumbar drain after the second trial is quite reasonable. If hearing is not serviceable, stapedectomy and obliteration of the vestibule with an oversized muscle plug is another option.[13] Another technique—translabyrinthine IAC closure—has also been proposed with an acceptable success rate.(8) A lumbar drain may be used postoperatively in all procedures.

CONCLUSION

We recommend that every patient with unilateral fluid in the middle ear space, especially with recurrent bacterial meningitis, be evaluated by an otolaryngologist for skull base CSF leakage and parameningeal sources of infection. The most common sources of these infections are the ears and paranasal sinuses. If the initial examination reveals no source of infection, imaging including CT scan, MRI, and even CT cisternography in search of skull base CSF leakage is recommended.[14] Most of the time leakage can be successfully managed with minimally invasive techniques.

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