Porencephalic Cyst: A Review of the Literature and Management of a Rare Cause of Cerebrospinal Fluid Otorrhea

*John M. Ryzenman, ††Vanessa S. Rothholtz, and †§Richard J. Wiet

*The Ohio Ear Institute, LLC, Columbus, Ohio; †Department of Otolaryngology–Head and Neck Surgery, Northwestern University, Feinberg School of Medicine, Hinsdale, Illinois; ‡Department of Otolaryngology–Head and Neck Surgery, University of California, Irvine, California; and §Ear Institute of Chicago, LLC, Hinsdale, Illinois, U.S.A.

Objective: To discuss the first reported case of spontaneous cerebrospinal fluid (CSF) otorrhea caused by a massive CSF-containing porencephalic cyst.


Setting: A tertiary neurotologic referral center.

Patient: A 65-year-old woman with congenital hemiplegia presented with left-sided spontaneous CSF otorrhea of 4-month duration. An audiogram revealed a mixed hearing loss. High-resolution computed tomography revealed a thinning of the tegmen and epitympanum without an obvious defect. Magnetic resonance imaging revealed a massive porencephalic cyst essentially replacing the entire left cerebral hemisphere.

Intervention: A transmastoid approach with three-layered closure was used to successfully repair the sieve-like defects that were discovered in her tegmen.

Results: The patient remains free of drainage, and the conductive hearing loss has resolved.

Conclusion: Spontaneous CSF otorrhea caused by a porencephalic cyst is an uncommon cause of conductive hearing loss that has never been reported before. Only a few cases of traumatic CSF otorrhea/rhinorrhea associated with a porencephalic cyst have been reported. A high level of suspicion, a β2-transferrin assay, and appropriate radiographic imaging are required for diagnosis in adults without a history of trauma, meningitis, chronic ear disease, or previous ear surgery.

Key Words: Cerebrospinal fluid—Encephalocele—Otorrhea—Porencephaly.


A porencephalic cyst is a fluid-filled intracranial lesion that may communicate directly with the ventricular system or is separated from it by a thin layer of brain tissue and is covered exterioirly by arachnoid. These lesions are uncommon but can occur congenitally or arise postnatally as a result of trauma, infection, or hemorrhage. Rarely does a porencephalic cyst lead to or present with cerebrospinal fluid (CSF) rhinorrhea or otorrhea. Untreated CSF otorrhea can result in meningitis, and surgical intervention is typically warranted.

To our knowledge, this is the first known case of spontaneous CSF otorrhea caused by a massive porencephalic cyst, which initially presented as an effusion with conductive hearing loss.

CASE REPORT

A 65-year-old woman with congenital hemiplegia attributed to a neonatal polio infection presented to her primary care physician with complaints of hearing loss. The patient was referred for audiometric and clinical evaluation. The patient was noted to have bilateral mild sensorineural hearing loss and a unilateral left conductive hearing loss with a flat tympanogram. Examination revealed a noninfected middle ear effusion. After failing conservative management of the effusion, a tympanostomy tube was placed with some relief of pressure and slight improvement in hearing. However, the tympanostomy tube was removed and for the next 4 months, the patient had persistent clear otorrhea that subsequently was diagnosed definitively as CSF with a β2-transferrin assay.

The patient was referred to the senior author (R. J. W.) for management. Examination revealed an anterior
inferior perforation with spontaneous flow of clear fluid despite repeated suctioning. The patient denied any occurrence of salty rhinorrhea or meningismus symptoms.

A high-resolution computed tomography (CT) of the temporal bone revealed thinning of the tegmen and epitympanum but without an obvious defect or mass lesion in the temporal bone (Fig. 1). Given the known CSF leak, a high index of suspicion was present for an encephalocele or tumor as a possible cause for the otorrhea. Magnetic resonance imaging with gadolinium enhancement was obtained, which revealed a previously undiagnosed massive porencephalic cyst encompassing the entire left hemisphere (Fig. 2). No dural enhancement of the middle fossa, suggestive of infection or inflammation, was noted. There was no herniation of the brain content into the temporal bone.

After careful and detailed counseling of the patient and her family, a transmastoid approach was chosen. Consent was obtained for possible overclosure of the external ear canal. With neurosurgical consultation, a middle fossa approach was discouraged because of possible rupture of the cyst with temporal lobe retraction (which was in direct continuity with the ventricular system). A lumbar drain was placed with gentle egress of CSF not only to prevent excessive contact pressure on the cyst wall but also to provide decreased flow of CSF through the suspected tegmen defect and thus facilitate healing of the repair.

Intraoperatively, the middle ear was found to be free of disease, and several adhesions were noted in the region of the eustachian tube that were left undisturbed. A complete canal wall-up tympanomastoidectomy was performed, with special attention to the tegmen and epitympanum. After meticulous removal of the epitympanic air cells, several sieve-like defects of less than 1 mm in diameter were identified.

A layer of Gelfoam (Pharmacia & Upjohn Company, Kalamazoo, MI) covered with fascia was placed to protect the ossicular chain. A three-layer closure was performed with hydroxyapatite paste applied to the bony defects in the epitympanum and mastoid tegmen followed by a muscle plug, temporalis fascia, and fibrin glue.

FIG. 1. Preoperative noncontrast CT of the temporal bone. Axial image 1 (A), axial image 2 (B), coronal image 1 (C), and coronal image 2 (D).
Postoperatively, the patient was maintained on perioperative antibiotics, bed rest, and stool softeners. The lumbar drain was removed on postoperative Day 3, and the patient was discharged the following day. The patient has remained free of CSF otorrhea or rhinorrhea, with resolution of her conductive hearing loss in the ensuing 8 months of follow-up.

DISCUSSION

Porencephaly—Definition/Pathogenesis

Porencephaly is defined as a fluid-filled “defect communicating with ventricles or separated from them by a thin layer of brain tissue, and covered on the outside by arachnoid” (1). Postnatally, a porencephalic cyst may result from trauma, infection, or hemorrhage. Hypoperfusion leads to focal encephalomalacia, focal necrosis of both the gray and white matter, and then cystic degeneration (2). Porencephalic cysts that develop antenatally are divided into two types. Typically lined by a cicatricial glial membrane, Type I cysts originate from the unilateral destruction of cerebral tissue caused by a central nervous system infection or intracerebral hemorrhage, with the extent of the cyst depending on the size of the hemorrhage. Type II cysts develop because of a developmental deficit in neuronal migration, thereby creating bilateral symmetric cysts that are lined by ependymal tissue (3). Cerebral injury may occur during delivery or with

FIG. 2. Preoperative magnetic resonance imaging of the temporal bone. Coronal image of porencephalic cyst abutting the temporal bone (tegmen) at the site of CSF leak (A), T1-weighted image (B), and T2-weighted image (C).
Complications From Porencephalic Cysts

Infants with porencephaly exhibit hemiplegia contralateral to the lesion, with some developmental delay (3,5). Other complications include distal hypertonicity, spasticity, and truncal ataxia (3). Rarely does a porencephaly cyst lead to or present with CSF rhinorrhea or otorrhea (6,7). The prognosis of porencephaly is not dependent on the cause but rather on the location and extent of the cyst (2).

Multiple Defects of Tegmen—Pathogenesis

Impeding the spread of infection and the leaking of CSF, the tegmen forms the floor of the middle cranial fossa and overlies the tympanic and mastoid cavities, creating a barrier between the middle fossa and the temporal bone. Single or few tegmen defects are common, with autopsy studies reporting an occurrence in 15 to 34% of cadavers. In less than 1% of temporal bones, multiple spontaneous tegmental defects (more than five) occur, thereby producing a sieve-like appearance to the bony floor (8,9).

A few theories on the pathogenesis of multiple tegmen defects have been postulated. When multiple defects occur in the absence of other otologic or intracranial diseases, the dehiscences are thought to be caused by excessive bone resorption at the time of initial pneumatization of the temporal bone or by defects in embryogenesis (10). It has also been proposed that the defects could be age-related because of progressive dural thinning and chronic exposure to CSF pulsations (11). Aging may also lead to enlargement of arachnoid granulation–associated dural defects. These arachnoid granulations do not reach a venous lumen and are aberrantly distributed among the cranial fossae. Cerebrospinal fluid pressure variations, increasing age, and enlargement of these defects may cause the bone to erode through the fibrous covering of the arachnoid granulation, leading to CSF leak (12-14).

CSF Otorrhea—Pathogenesis

The most common causes of otorrhea are trauma and chronic ear disease; it may be iatrogenic (i.e., tumor removal) or congenital (11,15). However, for otorrhea to be considered “spontaneous,” none of the aforementioned can be attributed as a cause. As stated previously, CSF pressure variations, increasing age, and enlargement of defects caused by trauma, ear disease, or congenital malformations may lead to discharge of CSF into the middle ear. Congenital malformations that may lead to otorrhea include an enlarged petrosal fallopian canal, widened cochlear aqueduct, patent tympanomeningeal (Hyrtl) fissure and Mondini malformation (16,17). A spontaneous abnormal connection between the perilymphatic and subarachnoid spaces may occur between the vestibule and the internal auditory canal, the cochlear aqueduct, the vestibular aqueduct, and the subarcuate fossa (13,15). A defect in a pneumatized tegmen and a compromised dura must both exist for a CSF leak to occur (18). The classification of spontaneous CSF otorrhea has been described in three encompassing abnormal connections: Type 1, through the otic capsule; Type 2, adjacent to the otic capsule; and Type 3, distant from the otic capsule (19). Cerebrospinal fluid middle ear effusion/otorrhea can develop in adults without an apparent cause or concomitant ailment such as a history of meningitis of head trauma (20). Ferguson et al. (11) analyzed that patients who develop spontaneous CSF otorrhea from tegmen defects are usually older (mean age, 48 yr; range, 8 mo to 80 yr) and present with aural fullness, middle ear effusion, or clear otorrhea.

Diagnosis of CSF Otorrhea

Although the concentration of glucose and protein of ear fluid can be measured to detect CSF otorrhea, the β2-transferrin assay is more commonly used because it is both highly sensitive and specific for CSF (21). Glucose testing of concentration of 30 mg/dl is considered confirmatory if the patient has normal blood glucose levels; however, its use in the detection of CSF otorrhea has diminished because of its high false-positive rate caused by various confounding factors such as meningitis and sample contamination.

With the advent of modern medical imaging and with so many imaging techniques at our disposal, the question arises with regard to which is best used as a diagnostic modality for CSF otorrhea associated with tegmen defects. It has been found that high-resolution CT, although not absolute in its ability to detect the minute defects in the tegmen, identifies defects in up to 71% of the patients with CSF leak and can also be used for operative planning (22). Magnetic resonance imaging in the coronal plane is useful for examining the middle cranial fossa to effectively identify if the bony defects contain fluid, meninges, or brain tissue (18).

Repair of CSF Leak (Otorrhea/Rhorrhea)

Many techniques with a variety of materials have been proposed for the successful management of CSF otorrhea (20,23). The leak may be identified intraoperatively by injecting fluorescein or Ringer solution through a preoperatively placed lumbar drain. The treatment of the tegmen defect depends on its size and location (18). Exposure via the transmastoid approach, middle cranial fossa approach, and craniotomy is frequently used to gain maximal access in repairing the CSF leak (17,23–25). The transmastoid approach avoids a craniotomy and temporal lobe retraction, although still allowing both the middle and posterior fossa to be assessed and repaired (17,20). Some authors note that the size, number, and location of bony tegmen defects can be assessed with a transmastoid approach (17,23). However, studies have shown that a transmastoid

approach alone is insufficient for managing large (>1.5 cm) multiple or anterior tegmen defects and may lead to multiple procedures for correction (20). If the defects cannot be closed with a transmastoid approach, an intracranial approach may be necessary (11,24). Many authors recommend a combined temporal craniotomy and transmastoid repair of the defects (18).

Closure of the defect is frequently and most definitively accomplished using a multilayer technique. The rate of successful closure by Savva et al. (23) was 81% at 2 years when using multiple layers of autologous materials alone and 100% at 2 years when using artificial materials (Gelfoam, Oxycel [Becton and Dickinson, Franklin Lakes, NJ] cotton, and bone wax) combined with multiple layers of autologous material. Materials used include calvarial bone; pericranium; bone wax; Oxycel cotton; Marlex (CR Bard, Covington, GA) mesh; fibrin glue; abdominal, sternocleidomastoid, or temporals muscle; temporals fascia; auricular cartilage; or abdominal fat (15,17,20,23,24). A mixture of bone paste and fibrin glue covered with temporal fascia may also be used to close a CSF leak via the middle fossa approach (16). Another successful technique to achieve CSF leak closure via the transmastoid approach is the use of hydroxyapatite cement. As with any biomaterial, certain precautions must be taken to avoid complications. Hydroxyapatite must not be used in the presence of an active infection. In addition, the cement must be meticulously applied and completely dried before skin closure to minimize the chance of the material’s migration onto the ossicles, which may lead to hearing loss. With the aforementioned precautions considered, the material’s ability to maintain bone conduction to integrate itself within the bone by stimulating osteogenesis and its malleability that allows the material to fill microscopic defects makes hydroxyapatite an excellent choice in tegmen repair (26).

The Author's Choice

For appropriate-sized defects, a transmastoid repair begins with adequate exposure of all tegmen defects and dissection of all epitympanic air cells, with removal and replacement of the posterior segment of the external auditory canal if needed. A temporalis muscle plug is placed after reduction of cranial tissue through the defect or its resection if this is not possible. OtoMimix (Lorenz Surgical, Jacksonville, FL) (hydroxyapatite paste) is applied to all defects, and after hardening, fascia and fibrin glue are used to complete the repair. Postoperative care for successful maintenance of CSF leak closure includes antiemetics and stool softeners. A lumbar drain is rarely required. We administer a short course of prophylactic antibiotics to prevent meningitis; however, this practice is still controversial.

Porencephaly and CSF Otorrhea

Porencephaly presenting with otologic involvement and CSF leak is a rare entity and has been previously reported after trauma to the temporal bone. Abrunhosa et al. (2) reported a 20-year-old man with a porencephalic cyst secondary to two separate incidences of trauma at the ages of 3 and 14 years. Although the initial trauma produced a CSF leak, it spontaneously resolved, and the patient was asymptomatic until he acquired meningitis secondary to a middle ear infection and cholesteatoma. Upon computed tomographic imaging, a porencephalic cyst was revealed. Another case was reported by Jenkins et al. (6). After being struck by an automobile 2½ years before admission, a 10-year-old boy presented with meningitis and CSF otorrhea. Further workup revealed a posttraumatic porencephalic cyst of the mastoid. The tegmen defect that allowed displacement of the cyst into the mesotympanum and antrum was repaired with temporal fascia, a bone plug, and a rotating sternocleidomastoid muscle flap (7). Porencephaly may also present with CSF rhinorrhea especially if the defect is in the paranasal sinuses. Tokoro et al. (7) presented two patients with frontal lobe atrophy and porencephaly after trauma. Both incidences of rhinorrhea were repaired using a vascularized free rectus abdominus flap.

CONCLUSION

Spontaneous CSF otorrhea caused by a porencephalic cyst is an uncommon cause of conductive hearing loss, which has never before been reported. Only a few cases of traumatic CSF otorrhea/rhinorrhea associated with a porencephalic cyst have been reported. A high level of suspicion, a β₂-transferrin assay, and appropriate radiographic imaging are required for diagnosis in adults without history of trauma, meningitis, chronic ear disease, or previous ear surgery. Careful surgical planning and intervention can result in resolution of the otorrhea while preserving normal hearing.

REFERENCES

4. Naef RW. Clinical features of porencephaly; a review of thirty-two cases. AMA Arch Neurol Psychiatry 1958;80:133–47.