Spontaneous Cerebrospinal Fluid Leak through the Posterior Aspect of the Petrous Bone*

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J Neurol Surg B 2012;73:71–75.

Abstract

Spontaneous cerebrospinal fluid (CSF) leak through the posterior fossa (PF) aspect of the petrous bone is exceedingly rare. A case series allows analysis of etiologies and how they may differ from the more common middle fossa (MF) route of leakage. The design was a retrospective case series. The setting was a tertiary care institution. A series of three patients with PF spontaneous CSF leaks was identified. High-resolution imaging (CT and MRI) and intraoperative observations were evaluated. Both in this series and in previously reported cases, patients share the demographics typically found in the MF leak population. In our series, two patterns of PF CSF leak were identified: (1) large unilateral with cerebellar encephalocele and (2) small punctate defects just lateral to the endolymphatic sac. Two presented with simultaneous MF and PF leaks suggesting a shared etiology, at least in some cases, with a role for increased intracranial pressure. In spontaneous CSF leaks, it is important to evaluate the posterior petrous bone along with the tegmen. The concomitant appearance of MF with PF leaks points out the risk that repair via MF craniotomy could fail to identify a leakage site in the vicinity of the endolymphatic sac.

Keywords

- cerebrospinal fluid leak
- arachnoid granulation
- petrous bone
- posterior fossa

Spontaneous temporal bone cerebrospinal fluid (CSF) leak is defined as a communication between the subarachnoid space and the pneumatized space of the temporal bone, and is distinguishable from leaks caused by trauma, neoplasm, or infection.^{1–4} This rare phenomenon is often due to a defect in the tegmen of the middle ear or mastoid, and rarely in the posterior fossa (PF) plate of the petrous bone. A disproportionate number of patients with middle fossa (MF) leaks are female, aged 40 to 60 with a body mass index (BMI) greater than 30.^{3–8} Patients typically present with conductive hearing loss, aural fullness, and cerebrospinal otorrhea.^{1,6,9,10} Due to the rarity of PF leaks, the demographics and clinical presentation are less well defined.

The etiology of the defects in the dura and underlying temporal bone is subject to considerable debate in the literature. The arachnoid granulation (AG) theory has become widely cited as causative for both MF and PF leaks. The proponents of this theory describe aberrant AGs sitting against bone causing slow erosion over time. The MF is more prone to dehiscence and CSF leak due to a combination of a thinner bony plate and a higher proportion of AGs compared with the posterior petrous face.^{11–13} Recently in the last 5 to 10 years, associations between temporal bone CSF leak, obesity,^{6–8,14,15} and idiopathic intracranial hypertension (IIH)^{3,5,6,14–16} have been noted. IIH interestingly shares the similar demographics of age 40 to 60, female gender, and BMI greater than 30. Some authors have incorporated these findings into the AG theory, designating the AG's as an arrowhead leading a process of erosion due to unexplained increased intracranial pressure.^{6,10,13} Others simply identify

*This article was originally published online in *Skull Base* on November 30, 2011 (DOI:10.1055/s-0031-1296040)

increased intracranial pressure as the central etiologic factor for spontaneous CSF leak.¹⁴

The purpose of the present study is to add our series to the limited number of previously published cases, to compare the PF variety of transtemporal CSF leakage with the more common MF route in terms of demographics, clinical presentation, imaging characteristics, and potential etiologies.

Methods

We reviewed all the spontaneous temporal bone CSF leaks cases that have presented to our tertiary medical center in the last 8 years and identified three patients with leaks in the posterior aspect of the petrous bone. Clinical presentation, high-resolution imaging, and intraoperative observation were examined and compared with what has previously been described in the literature. The Institutional Review Board (IRB) at our institution approved this study (IRB-16879).

Results

Case 1

A 63-year-old female presented with a lifetime history of rhinorrhea and right otitis media that progressed to meningitis. She had no history of otologic disease or trauma. Her BMI was 29. A right middle ear effusion was noted on exam. A CT scan showed multiple dehiscences of the right tegmen and numerous scalloped defects of the posterior petrous face bilaterally just lateral to the posterior semicircular canal, in the vicinity of the endolymphatic sac and its

operculum (►Fig. 1A). The excavated shape of the pits suggested erosion from the intracranial aspect. A MRI showed irregularity of the posterior petrous face (as outlined by CSF), but no obvious encephalocele (►Fig. 1B). During right MF craniotomy and mastoidectomy, punctuate dural and osseous defects were found on both the MF floor (2×2 mm) and PF (5×5 mm) immediately superior to the endolymphatic sac. A small PF encephalocele was noted and resected. The MF floor was repaired with temporalis muscle and the PF with fascia and a muscle plug in the mastoid cavity. She remains asymptomatic 3 years postoperatively.

Case 2

A 37-year-old female presented with multiple episodes of meningitis. She had no history of otological disease or trauma. Her BMI was 28. A CT scan showed extensive loss of the right bony posterior petrous plate from the sigmoid sinus laterally to the posterior semicircular canal medially (►Fig. 2A). MRI demonstrated cerebellar herniation through the defect into the mastoid air cells. (►Fig. 2B). Of note, the contralateral cerebellum approximated the posterior aspect of the petrous pyramid without the usual intervening CSF space of the cerebellopontine angle. At mastoidectomy, the encephalocele was observed to be ~20 mm in diameter and protruded 10 to 15 mm into the mastoid. The dural and osseous breach spanned from the sigmoid sinus to the posterior semicircular canal. The encephalocele was resected and the defect was closed with temporalis muscle and fascia. Postoperative imaging showed an effective repair and she remains asymptomatic 3 years since surgery.

Figure 1 Noncontrast fine-cut axial computed tomography scan (A) demonstrating bilateral small posterior fossa defects (black arrows). The white arrow marks what was found to be a small encephalocele during the surgery. Axial T2 magnetic resonance image (B) shows fluid in the mastoid air cell system and a PF defect (black arrow), but no obvious encephalocele.

Figure 2 Noncontrast axial computed tomography scan (A) demonstrating a right large posterior petrous defect spanning from sigmoid sinus to the labyrinth (black arrow). On T2-weighted magnetic resonance image (B) a broad-based encephalocele is identified (black arrow). Note the contralateral cerebellum approximates the posterior aspect of the petrous pyramid without the usual intervening cerebellopontine fluid space of the cerebellopontine angle (white arrow).

Case 3

A 68-year-old male presented with a history of rhinorrhea and right aural fullness. He had no relevant otologic or traumatic history. A tympanostomy tube showed β -transferin-positive watery otorrhea. Exam revealed a BMI of 30 and a right serous middle ear effusion. A CT showed multiple dehiscences of the right tegmen and possible PF bony erosion just lateral to the endolymphatic sac operculum (**Fig. 3a**). However, this PF abnormality could not be visualized on MRI (**Fig. 3b**). At MF craniotomy, multiple dural dehiscences within a large osseous defect of the tegmen were found. The patient's postoperative course was complicated by partial complex seizures and hydrocephalus. At placement of a ventriculo-peritoneal shunt, a lumbar puncture opening pressure was 21. Despite shunt placement, he continued to have symptoms of rhinorrhea and aural fullness. Three months from his initial surgery, he underwent a transmastoid exploration during which an occult area of PF dura dehiscence between the sigmoid sinus and posterior semicircular canal was identified. Then thin bone of the PF was repaired with hydroxyapatite cement. He has been asymptomatic since 2 years.

Discussion

A review of the English language literature since 1966 (MEDLINE) revealed 10 previous cases of spontaneous CSF leak emanating from the posterior aspect of the petrous pyramid (**Table 1**).^{3,7,11,17-22} Some of these cases were published before the era of high-resolution imaging and in others the defect was poorly characterized. We report a series of three patients with high-resolution imaging corroborating their clinical history and precise surgical observation.

Figure 3 Noncontrast fine-cut axial computed tomography scan (A) showing possible small defect in the posterior fossa plate of the petrous bone (white arrow). This finding was not visualized on T2-weighted magnetic resonance imaging (B).

On evaluation of all cases, including our own, PF leak patients do appear to share the demographics of MF leaks. Of the 12 PF cases, 58% were female compared with the 60 to 90% incidence reported in published MF leak cases.^{1,3,5-8,10} Interestingly, only 5 of the 12 cases fell in the characteristic age range of 40 to 60 years, however the average age was still 59. Not enough information was available with regards to obesity and IIH. It should be noted that all three of our patients were overweight, but none had a BMI greater than 30. Clinical presentation was similar though meningitis was a more common presentation in PF leak patients.

Prior cases confirm that punctate lesions, solitary or multiple, predominate. In our series, two PF CSF leaks were of narrow diameter whereas one had an extensive defect in the posterior petrous face. In the earlier literature, 2 of the 10 cases had bilateral defects in the posterior aspect of the petrous bone. We recognized a bilateral deformity in our case 1, but actual leakage on one side.

A difference between the cases we provide compared with previously published PF leak cases was that two of the three patients presented with coexisting MF and PF leaks. For one case, not recognizing the PF dehiscence resulted in continued leak following a MF repair necessitating a second surgery. The concomitant MF and PF leak as well as shared demographics point to a possible similar etiology for tegmen and posterior plate defects.

It has been proposed that raised intracranial pressure may be the central etiologic factor for transtemporal CSF leaks. Long-term pressure may lead to erosion of often thin dural plates on the faces of the petrous pyramid, the MF being more susceptible than the PF.¹⁴ The bilateral scalloping of the posterior petrous face in case 1 suggests underlying high pressure and perhaps erosion by prominent AGs.^{12,13}

Alternatively, preformed pathways could provide an avenue for dural herniation into the pneumatic spaces of the temporal bone. Schuknecht suggests that "embryogenic faults" in the dura and bone can slowly break down with normal CSF pressure over time.²³ One possible site is at the endolymphatic sac and operculum where the cartilage of the embryonic otic capsule fuses with the temporal bone.²⁴ The narrow opening in the vicinity of the endolymphatic sac in case 3 suggests such a mechanism. The large focused posterior defect with cerebellar herniation in case 2 may also suggest such a mechanism; however, the contralateral cerebellum abutting the posterior petrous face also implicates a role for increased intracranial pressure.

It is imperative that when evaluating a patient for a transtemporal CSF leak the PF plate be carefully evaluated on preoperative imaging. If irregularity or defect is noted, exploration via mastoidectomy is warranted. It should be cautioned that antrotomy alone is insufficient and could easily overlook a subtle posterior surface deficiency. The entire petrous face from sigmoid sinus laterally to posterior semicircular canal medially should be inspected. Special scrutiny should be given to the area of the endolymphatic sac and its operculum.

Table 1 Summary of Published PF CSF Leak Cases to Date Including Our Case Series

Study Author	Year	Age	Gender	Side	Presentation	Posterior Fossa Defect
Schuknecht et al ¹⁷	1982	53	Female	Left	Meningitis	Defect between superior petrosal and sigmoid venous sinus
Briant et al ¹⁸	1982	76	Male	Left	Otalgia, hearing loss, meningitis	Defect anterior to sigmoid sinus
Wetmore et al ²⁰	1987	53	Female	Right	Hearing loss, tinnitus, rhinorrhea	Three tiny defects between sigmoid sinus and posterior semicircular canal
Gacek ¹⁹	1990	64	Male	Right	Aural fullness, hearing loss, otorrhea	Defect with AG anterior to sigmoid sinus
Gacek ¹¹	1992	53	Female	Right	Serous otitis media	Bilateral defects anterior and medial to sigmoid sinus
Welge-Luessen & Probst ²²	2004	64	Male	Right	Otalgia, meningitis	Three 3 × 4 mm defects
Welge-Luessen & Probst ²²	2004	57	Female	Left	Meningitis	1 × 1 cm defect in Trautmann's area
Rao et al ³	2005	75	Female	Right	Meningitis	Defect medial to sigmoid sinus
Lee et al ²¹	2008	41	Male	Left	Otorrhea, meningitis	Bilateral defects, left greater than right
Nadaraja et al	2011	68	Female	Right	Rhinorrhea, aural fullness, otitis media, meningitis	Defect superior to the endolymphatic sac
Nadaraja et al	2011	37	Female	Right	Meningitis	Large defect from sigmoid sinus to posterior semicircular canal
Nadaraja et al	2011	68	Male	Right	Rhinorrhea, aural fullness, hearing loss	Occult defect between sigmoid sinus and posterior semicircular canal

AG, arachnoid granulation; CSF, cerebrospinal fluid; PF, posterior fossa.

Conclusion

We present three well-documented patients with spontaneous CSF leak from the posterior aspect of the petrous bone. Patients with PF leaks have similar demographics to the MF leak population in that they tend to be age 40 to 60, female, and with a BMI greater than 30. Two of the three patients had both MF and PF leaks suggesting a similar etiology, with increased intracranial hypertension playing a central role. AG and congenital temporal bone defects may also contribute. It is important to examine the posterior face of the petrous bone along with the tegmen both on imaging and intraoperatively when treating a patient with spontaneous CSF leak of the temporal bone.

Note

Institutional approvals: This study has approval of the IRB for the Stanford School of Medicine (IRB-16979).

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