Posterior Fossa CSF Leak and Encephalocele Secondary to Arachnoid Granulations: A Case Report and Review of the Literature

Juan C. Yanez-Siller, MD, MPH[1]; Tasfia Hussain, MHS[2]; Shubham Patel, MD[1]; Rustin G. Kashani, MD[1]
[1]Department of Otolaryngology–Head & Neck Surgery, Emory University
[2]Emory University School of Medicine

Background

Spontaneous posterior fossa (PF) osseous defects, cerebrospinal fluid (CSF) leaks and associated encephaloceles are rare entities with an incompletely understood pathogenesis representing a rare subset of lateral skull base disease[1, 2]. Although most spontaneous skull base defects are attributed to idiopathic intracranial hypertension or prior surgical intervention, arachnoid granulations (AGs)—specifically aberrant AGs, which fail to communicate with venous sinuses and instead remain in direct contact with the PF plate—have emerged as an alternative or co-occurring etiologic mechanism[1-5]. Chronic pulsatile CSF forces at this interface are thought to drive progressive osseous erosion and eventual dural compromise.

Objective

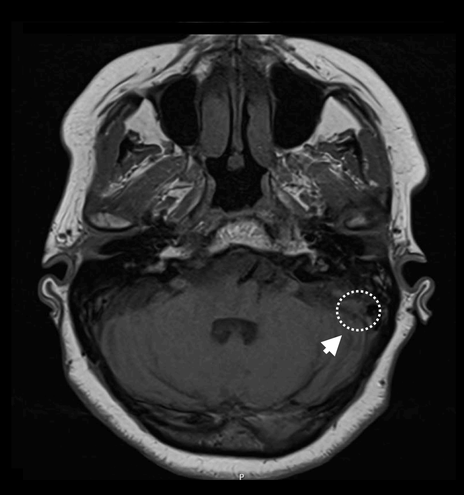
To describe a rare case of a spontaneous PF CSF leak and encephalocele associated with progressive bony erosion likely mediated by AGs, in the absence of traditional risk factors, and to highlight the diagnostic and surgical considerations associated with this entity.

Patient

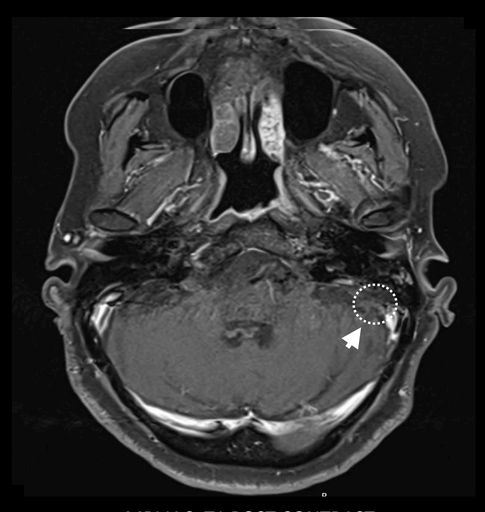
66-year-old female with a prior hospitalization for bacterial meningitis was incidentally noted on imaging to have otomastoiditis and a focal punctate bony defect anterior to the sigmoid sinus. Serial imaging demonstrated progressive enlargement of the defect over time. The patient had no history of prior skull base surgery, no clinical or radiographic evidence of idiopathic intracranial hypertension, and no elevated body mass index.

Intervention

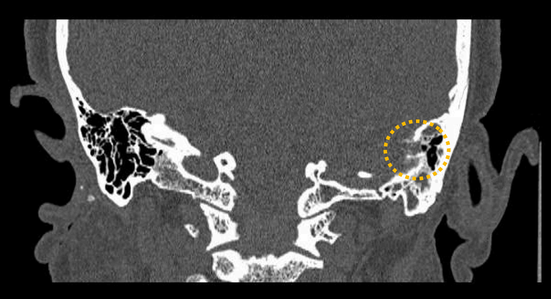
The patient underwent mastoidectomy for direct evaluation of the PF plate. Intraoperatively, irregular bony dehiscence of the PF and sigmoid sinus plates was identified, with herniation of arachnoid and glial tissue consistent with an encephalocele. Abnormal soft tissue overlying the expected location of the ELS was encountered and biopsied, demonstrating no evidence of neoplasia. The ELS was directly exposed and appeared grossly normal. The PF defect was repaired using a multilayered reconstruction technique with no evidence of recurrent leak postoperatively.

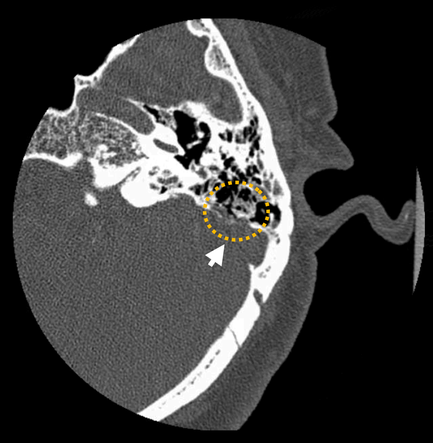


MRI IAC, T1



MRI IAC, T1 POST CONTRAST





Discussion

This case supports AGs as an independent pathogenic mechanism for spontaneous PF defects, even in the absence of elevated intracranial pressure or other recognized risk factors. The lack of established predisposing conditions, including prior skull base surgery, idiopathic intracranial hypertension, and elevated body mass index, further supports an AG-mediated erosive process. A review of the published literature of cases of PF CSF leak was carried out (Table 1)[1-13]. **This represents the 23rd case of PF CSF leak reported in the literature.**

Spontaneous PF CSF leaks are uncommon and often underdiagnosed, as clinical presentations may mimic middle fossa defects and radiographic findings can be subtle. Most reported series identify obesity and idiopathic intracranial hypertension as the predominant risk factors[3] however, AGs may represent a distinct and potentially independent mechanism, particularly in patients without these traditional factors[4, 5]. Careful longitudinal imaging review and deliberate intraoperative inspection of the PF plate are therefore essential, especially when progressive focal bony erosion is observed.

Encephaloceles, meningoencephaloceles and AGs are the principal tissue associations in PF CSF leaks. In this case, surgical exploration revealed irregular dehiscence of the PF and sigmoid sinus plates with herniation of arachnoid and glial tissue, confirming an encephalocele and supporting a chronic erosive mechanism. AGs exert sustained pulsatile stress on thin or pneumatized bone, leading to gradual osseous erosion and dural compromise[3-5]. Importantly, AGs may mimic meningoencephalocele, meningioma, or endolymphatic sac tumors, complicating diagnosis[4] . Additional factors—such as reduced bone density from infectious or nutritional influences, congenital thinning or dehiscence of the PF cortex, and elevated intracranial pressure—may modulate the rate and extent of AG-mediated erosion, influencing progression to active CSF leak or meningoencephalocele formation[4, 5] .

Clinical presentation may be complicated by meningitis, which has been reported with increased frequency in PF CSF leaks[1, 2, 4] and was also observed in our patient. This underscores the importance of maintaining a high index of suspicion and pursuing definitive surgical management.

Histopathologic temporal bone studies show that AGs frequently penetrate the dura and erode adjacent bone, with age-related progression that may culminate in meningoencephaloceles and spontaneous CSF leakage[5]. These observations underscore that AG-associated PF erosion is likely underrecognized in clinical practice.

From a surgical perspective, wide exposure of the PF plate is critical to avoid missed pathology and failed repair. This is most reliably achieved via mastoidectomy, which should be regarded as the preferred operative corridor for definitive diagnosis and durable repair[2] . Meticulous intraoperative assessment, defect-specific surgical planning, and multilayered reconstruction are essential for achieving durable outcomes in this rare entity.

Table 1. Reported Cases of Spontaneous PF CSF Leaks Over Time, Including the Present Case

Author	Year	Number of Spontaneous PF CSF Leaks
Schuknecht et al[6]	1982	1
Briant et al[7]	1982	1
Wetmore et al[8]	1987	1
Mostafa[9]	1997	2
Gacek et al[3]	1999	2
Welge-Luessen et al [10]	2004	2
Rao et al[11]	2005	1
Lee et al[12]	2008	1
Nadaraja et al[13]	2011	3
Wick et al[2]	2016	5
Cooper et al[1]	2020	3
Present case	2025	1
Total	—	**23**

Conclusion

AGs should be considered in the differential diagnosis of spontaneous PF CSF leaks and encephaloceles, particularly in patients without traditional risk factors. While obesity and idiopathic intracranial hypertension remain the most commonly associated conditions, AG-mediated erosion may be coexisting or represent a distinct and potentially independent pathogenic mechanism. Progressive focal bony erosion on serial imaging is an important diagnostic clue. Accurate diagnosis and durable management rely on longitudinal imaging surveillance, meticulous intraoperative evaluation, and multilayered skull base reconstruction.

Contact

Juan C. Yanez-SIller, MD, MPH
Dept of Otolaryngology-HNS
Emory University
jyanezs@emory.edu

References

1. Cooper T, Choy MH, Gardner PA, Hirsch BE, McCall AA. Comparison of Spontaneous Temporal Bone Cerebrospinal Fluid Leaks From the Middle and Posterior Fossa. Otol Neurotol. 2020;41(2):e232-e237. doi:10.1097/MAO.0000000000000247
2. Wick CC, Killeen DE, Clark M, Kutz JW, Isaacson B. Posterior Fossa Spontaneous Cerebrospinal Fluid Leaks. Otol Neurotol. 2017;38(1):66-72. doi:10.1097/MAO.0000000000001261
3. Gacek RR. Arachnoid granulation cerebrospinal fluid otorrhea. Ann Otol Rhinol Laryngol. 1990;99(11):854-862. doi:10.1177/000348949009901102
4. Mafee MF, Bradley WG, Almutairi A, Zawaideh M, Patel K. Arachnoid granulations and spontaneous cerebrospinal fluid otorrhea: Role of imaging. Operative Techniques in Otolaryngology-Head and Neck Surgery. 2014;25(1):74-86. doi:10.1016/j.otot.2013.11.01
5. Yew M, Dubbs B, Tong O, et al. Arachnoid granulations of the temporal bone: a histologic study of dural and osseous penetration. Otol Neurotol. 2011;32(4):602-609. doi:10.1097/MAO.0b013e318212902
6. Schuknecht HF, Zaytoun GM, Moon CN Jr. Adult-onset fluid in the tympanomastoid compartment. Diagnosis and management. Arch Otolaryngol. 1982;108(12):759-765. doi:10.1001/archotol.1982.00790600003002
7. Briant TD, Bird R. Extracranial repair of cerebrospinal fluid fistulae. J Otolaryngol. 1982;11(3):191-197
8. Wetmore SJ, Herrmann P, Fisch U. Spontaneous cerebrospinal fluid otorrhea. Am J Otol. 1987;8(2):96-102.
9. Mostafa BE. Spontaneous CSF Leaks From the Temporal Bone. Skull Base Surg. 1997;7(3):139-144. doi:10.1055/s-2008-1058605.
10. Welge-Luessen A, Probst R. Spontaneous cerebrospinal fluid otorrhea in the posterior fossa as a rare cause of adult bacterial meningitis. Otolaryngol Head Neck Surg. 2004;130(3):375–377. doi: 10.1016/j.otohns.2003.07.001.
11. Rao AK, Merenda DM, Wetmore SJ. Diagnosis and management of spontaneous cerebrospinal fluid otorrhea. Otol Neurotol. 2005;26(6):1171-1175. doi:10.1097/01.mao.0000179526.17285.cc.
12. Lee M H, Kim H J, Lee I H, Kim S T, Jeon P, Kim K H. Prevalence and appearance of the posterior wall defects of the temporal bone caused by presumed arachnoid granulations and their clinical significance: CT findings. AJNR Am J Neuroradiol. 2008;29(9):1704–1707. doi: 10.3174/ajnr.A1214
13. Nadaraja GS, Monfared A, Jackler RK. Spontaneous Cerebrospinal Fluid Leak through the Posterior Aspect of the Petrous Bone. J Neurol Surg B Skull Base. 2012;73(1):71-75. doi:10.1055/s-0032-1304560.