Double Skull Base Defects with Primary Spontaneous Cerebrospinal Fluid Leaks in a Single Patient: Temporal and Sphenoid Bones

**Benjamin Schmitt, M.D.,¹ Jean-Michel Badet, M.D.,¹
Jean-Claude Chobaut, M.D., Ph.D.,^{1,2} and Laurent Tavernier, M.D.^{1,2}**

ABSTRACT

Primary spontaneous cerebrospinal fluid leaks occurring in the absence of any apparent cause are rare. Patients may present such leaks simultaneously, or successively, in multiple locations. We report here the sixth case of anterior and lateral skull base defects presented in a single patient. Although rhinoliquorrhea and otoliquorrhea were reported separately in the literature, we postulated that the pathophysiology is the same for the whole skull base; obesity seems to contribute to the development of such primary spontaneous cerebrospinal fluid leak. Consequently, management is similar and regular follow-up is very important.

KEYWORDS: Skull base defect, primary, spontaneous, CSF leak

Cerebrospinal fluid (CSF) rhinorrhea and otorrhea can be classified as traumatic or spontaneous. Traumatic leaks result from skull trauma and surgical intervention, while spontaneous CSF leaks can be classified into primary and secondary leaks. Spontaneous secondary leaks result from malformation (Mondini, Sternberg's canal, etc.), infection or neoplastic invasion; whereas primary spontaneous leaks occur in the absence of any apparent triggering factor. Primary spontaneous leaks are estimated to represent between 6%¹ and 40%² of all leaks. We report a case of primary spontaneous CSF leaks of tegmen tympani and lateral wall of sphenoid sinus in an obese female patient. To our knowledge, this is the sixth case of anterior and lateral skull base defects in a single patient. Review of literature suggests that obesity contributes to the development of primary spontaneous CSF leak.^{3–6} Although rhinoliquorrhea and

otoliquorrhea were reported separately in the literature, we postulated that the pathophysiology is the same for the whole skull base. Consequently, management is the same and regular follow-up is very important.

CASE REPORT

A 59-year-old woman presented with a 1-year history of intermittent clear right rhinorrhea; she was obese, with a body mass index (BMI) of 33 kg/m². There was no history of either head trauma or nasal otologic surgery. The patient had been treated for allergic rhinitis for 6 months. After a positive glucose oxydase test strip, the patient was referred to our institution for suspicion of CSF rhinorrhea.

A computed tomography (CT) scan showed complete opacification of the right sphenoid sinus and bony

¹Department of ENT and Head and Neck Surgery, University Hospital of Besançon; ²EA4268, I4S, IFR133, University of Franche-Comté, Besançon cedex, France.

Address for correspondence and reprint requests: Laurent Tavernier, M.D., Department of ENT and Head and Neck Surgery, University Hospital of Besançon, Service d'ORL et de chirurgie cervico-faciale, Centre Hospitalier Universitaire Jean Minjoz, 3, boulevard Fleming, 25030 Besançon cedex, France (e-mail: ltavernier@

chu-besancon.fr).

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Figure 1 Sagittal CT scan localizes a defect of lateral right sphenoid sinus with effusion.

defects on the lateral wall (Fig. 1). Magnetic resonance imaging (MRI) (Fig. 2) revealed an encephalocele in the lateral portion of right sphenoid sinus, and an empty sella. Based on these findings, the patient underwent surgical repair of the right sphenoid for the ongoing CSF leakage. The patient was treated by endoscopic repair whereby the encephalocele was removed and the sphenoid sinus was obliterated with adipose tissue and fibrin glue; packing over the graft area included Alginate calcium. The patient required a second repair 12 months after surgery following a recurrence of CSF rhinorrhea; this was undertaken with endoscopic surgery combined with external ventricular derivation.

Three years after the onset of rhinoliquorrhea, the patient experienced aural blockage and hearing loss of the right ear for several weeks. She was diagnosed with serous otitis media and underwent myringotomy which resulted in a pulsating watery discharge. CT scan (Fig. 3) and MRI revealed tegmen tympani defects and middle ear opacification and an audiogram indicated a conductive hearing loss of 30 dB in the right ear and normal hearing in the left ear. A right mastoidectomy was performed and a small tegmen

Figure 2 Sagittal, unenhanced T1-weighted MRI of empty sella.

tympani defect was discovered then repaired with a perichondrium graft.

During hospitalization, the patient presented visual disturbances due to intracranial hypertension; a polysomnographic exam was performed on the basis of the patient's snoring and obesity, and concluded obstructive sleep apnea.

One year after the surgery there was no evidence of recurrence.

DISCUSSION

The incidence of primary spontaneous CSF leaks is low and the description of double location—on the anterior as well as on the lateral skull base—in a single patient has rarely been reported in the literature. To our knowledge, there are only five other cases to date (Table 1).⁷⁻¹¹

Figure 3 Coronal CT scan demonstrates a bony defect of right tegmen tympani and middle ear CSF effusion.

Table 1 Cases of Double Skull Base Defects with Primary Spontaneous CSF Leak

Author	Year	Sex	Age	Anterior Skull Base Defect		Lateral Skull Base Defect		Time*
				Location	Surg. Approach	Location	Surg. Approach	
Rotillio ⁵	1982	M	64	R sphenoid s.	Transphenoidal	L tegmen tympani	Transmastoid	2 years
May ⁶	1995	F	62	L ethmoid s.	Neurosurgical	L mastoid tegmen	Transmastoid	3.5 years
Pappas ⁷	1996	F	57	R sphenoid s.	Fronto-temporal	R tegmen tympani	Transmastoid and middle fossa	4 years
Raghavan ⁸	2002	F	72	R cribrif. pl.	Endoscopic	L tegmen tympani	Transmastoid	1 month
González-García ⁹	2006	F	62	R cribrif. pl.	Endoscopic	R tegmen antri	Transmastoid	Concomitant
Case report	2008	F	59	R sphenoid s.	Endoscopic	R tegmen tympani	Transmastoid	3 years

*Period between anterior and lateral skull base defect.
cribrif. pl., cribriform plate; L, left; R, right; s., sinus; Surg., surgical.

The exact etiology of these lesions is not well understood. Different theories explain temporal and anterior skull base's CSF leaks.

For the temporal location, one of the first theories suggested the presence of aberrant arachnoid granulations which cause bony defects.¹² Another theory suggests that incomplete and abnormal embryologic development results in tiny congenital openings that evolve into defects with CSF pulsations.¹³ Dynamic factors have been implicated in this development which include age (spontaneous temporal CSF leak is more common in patients in their seventies)¹⁴ and obesity (association between morbid obesity and temporal lobe encephaloceles has been demonstrated).⁶ Indeed, a recent study confirms a correlation between radiographic empty sella, increased BMI and spontaneous CSF otorrhea.⁵ Empty sella syndrome is a common radiologic finding in patients with spontaneous CSF otorrhea, and, in the same study,⁵ seven out of eight patients were obese. The majority of primary spontaneous CSF leaks of the lateral skull base occurred through the middle fossa tegmen.¹⁴ The choice of treatment of these leaks—whether middle fossa craniotomy, transmastoidectomy or a combined approach—largely depends of the defect location. However, wherever possible, a standard mastoidectomy remains the preferred surgical approach for the treatment of tegmen defects as it is minimally invasive.

In 1968, Ommaya et al¹⁵ explained the cribriform plate leaks by focal atrophy in the anterior skull base. Later, some authors¹⁶ developed a congenital theory which described congenital anterior skull base defects. At the same time, Rotilio et al⁷ links CSF rhinorrhea with empty sella syndrome and increased intracranial pressure; and continuous intracranial pressure (ICP) monitoring at the lumbar level revealed raised ICP with intermittent high waves. More recently, one study³ clearly describes this association. Thus, elevated CSF pressures play an important role in spontaneous rhinorrhea pathophysiology.

The clinical association between obese patients and spontaneous CSF leaks has also been described^{4,17}; indeed, patients with a BMI higher than 30 are at risk of developing primary spontaneous CSF rhinorrhea. A correlation between (female) gender and spontaneous rhinorrhea was also suggested¹⁷; this could be a consequence of osteoporosis after menopause. The most common sites of anterior skull base's leaks are at the floor of the anterior cranial fossa and sphenoid sinus, the thinnest bone walls of the area. For therapeutic treatment, neurosurgical approach (anterior craniotomy, etc.) is no longer the standard⁷⁻⁹; rather, endonasal endoscopic procedures have become the preferred treatment of anterior skull base defects.^{10,11}

For both temporal and anterior skull base, theoretical and acquired factors are described to explain spontaneous CSF leaks; in fact, it is most likely a combination of those factors that are responsible for spontaneous leaks.

Dynamic factors, as reported in the literature, have an important place in the development of temporal and anterior skull base CSF leaks. They include obesity (BMI higher than 30), gender (female), age (middle aged) and bone density (particularly evidence of osteoporosis). Empty sella likely represents a sign of elevated intracranial pressure, which is more common in obese females and similar to benign intracranial hypertension (BIH). This supports the thesis that intracranial hypertension plays a role in the pathogenesis of spontaneous CSF rhinorrhea and otorrhea.

The long-term success of endoscopic repair depends on the CSF leak etiology. Primary spontaneous CSF leaks have the highest recurrence rate of all etiologies,¹⁸ with BIH providing a reasonable explanation. For this reason, clinical follow-up is very important to detect and treat any such recurrence of CSF leaks.

CONCLUSION

With the double localization of CSF leaks in an obese middle aged female patient presenting clinical symptoms

of BIH, we postulated that the pathophysiology of spontaneous CSF leaks is the same for all bones of the skull base and, consequently, requires similar management. Recurrent leaks often occur with some delay and could present at a different site, it is therefore important to institute regular follow-ups. At present, we suggest treatment by minimally invasive surgical procedures: endoscopic repair for anterior skull base's defect, and a transmastoid approach for temporal location.

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