CASE REPORT

Sphenoid Cranial Base Defects in Siblings Presenting with Cerebrospinal Fluid Leak

Joseph M. Bernstein, M.D., J. Thomas Roland, Jr., M.D.,
and Mark S. Persky, M.D.

ABSTRACT—Two sisters presented to our medical center with nontraumatic cerebrospinal fluid (CSF) fistulas from left sphenoid sinocranial junction defects. One sister had recurrent meningitis over a 20-year period that prompted a skull base evaluation. Four years later, her younger sister presented with profuse CSF rhinorrhea. Transethmoid sphenoidotomy with sinus obliteration and lumbar–subarachnoid temporary CSF diversion successfully treated one sister, while the other required reoperation and permanent lumbar–peritoneal shunting. In both cases the skull base defect was identically located in the posterolateral left sphenoid sinus. Embryological considerations, evaluation, and management are presented.

Cerebrospinal fluid (CSF) fistulization requires the presence of a multilayer defect. The most resistant obstacle is the cranial base bone, but the fistula needs to establish itself through arachnoid, dura, and nasal mucosa as well. In 1968, Ommaya et al¹ classified CSF rhinorrhea by traumatic and nontraumatic etiologies. They discouraged the prevalent use of the term *spontaneous CSF rhinorrhea*, arguing that it inaccurately related the pathogenesis and natural history of the condition. The traumatic category was subdivided into accidental and iatrogenic causes. In the nontraumatic category a distinction was made between high pressure leaks, as may be due to tumors or hydrocephalus, and normal pressure leaks, with etiologies including osteomyelitic erosion and congenital anomalies.

The following are case reports of two siblings who presented to our institution 4 years apart with identically located cranial base defects and nontraumatic CSF rhinorrhea.

CASE REPORTS

Case 1

A 41-year-old female from Belize presented in 1991 with complaints of an intermittent and profuse

left-sided rhinorrhea over the course of 20 years and frequent headaches. She denied fevers, chills, nausea, vomiting, and visual disturbances. She reported no trauma or previous surgeries. Physical examination revealed a watery left nasal discharge that could not be specifically localized. No nasal masses were noted. Neurological examination was normal. Her medical history is significant for two episodes of meningitis, hypertension, diabetes mellitus, asthma, and obesity.

Glucose testing of the nasal discharge was consistent with CSF. An iopamidol computed tomographic (CT) cysternogram revealed a skull base dehiscence of the left posterolateral sphenoid sinus wall and a left sphenoid mass (Figs. 1, 2). Magnetic resonance imaging (MRI) was suggestive of a temporosphenoidal encephalocele (Figs. 3, 4).

The patient underwent a left external ethmoidectomy approach, via a Lynch incision, for the repair of the sphenoid leak and excision of the sphenoid mass. The intersinus septum was removed, and wide exposure into the sphenoid sinus was obtained, especially of the lateral recess. An active CSF fistula was encountered in the posterolateral aspect of the sinus. Necrotic-appearing tissue was noted within the sinus and was stripped along with all mucosa. The site of the fistula was covered with fascia, which was tucked under the margins of

the bony dehiscence. Abdominal fat was used to obliterate the sinus. A layer of Gelfoam was placed, followed by packing of the nasal cavity with Bacitracin-coated gauze. A lumbar spinal catheter was inserted and remained in place for 5 days.

She had done well for 1 month when recurrent CSF rhinorrhea was noted. A transethmoid re-exploration of the left sphenoid skull base defect was undertaken. The previously placed fat and postoperative fibrosis were debrided. No active leak was visualized. The sinus was again obliterated with fat, and an indwelling lumboperitoneal shunt was inserted to provide for long-term decompression of CSF pressures. The patient has been without further evidence of a CSF leak during 6 years of follow up.

Case 2

The patient is a 40-year-old female, the younger sister of the previous case. She presented in 1995 with complaints of intermittent profuse watery left-sided rhinorrhea for 2 months. She denied fever, chills, headaches, nausea, vomiting, or visual disturbances. There was no history of trauma or previous operations. Her medical history is significant for asthma, hypertension, and obesity. There is no history of meningitis. Physical examination revealed left watery rhinorrhea in upright position with neck flexed. Nasal endoscopic examination failed to localize the source of the leak, but no nasal masses were noted. The patient was noted to be hypertensive and had diffuse wheezing on auscultation of the lung fields.

Glucose testing of the nasal discharge was consistent with CSF. An iopamidol CT cisternogram was obtained that demonstrated a skull base dehiscence in the left posterior sphenoid sinocranial junction with con-

trast material filling the left sphenoid sinus, suggesting an encephalocele or meningoencephalocele (Figs. 5, 6).

Medical consultation was obtained, and the patient's asthma and hypertension therapy were optimized.

The patient underwent a left external ethmoidectomy approach, via a Lynch incision, for the repair of a sphenoid sinus CSF fistula. A posterolateral dehiscence was identified with fiberoptic nasal endoscopes. No active leak was noted at the time of surgery. The sinus mucosa was stripped, fascia was tucked into the defect, and the sphenoid sinus was obliterated with abdominal fat. A layer of Gelfoam was placed, and the nasal cavity was packed with Bacitracin-coated gauze.

Given the lack of an active leak intraoperatively our neurosurgical colleagues decided against the placement of an indwelling lumboperitoneal shunt. A lumbar spinal drain was inserted and remained in place for 5 days. The patient remains without further CSF rhinorrhea after 2 years of follow up.

DISCUSSION

In 1977, Kaufman et al² reported on five cases of middle fossa CSF fistulas and proposed that coincident anatomical and physiological factors may explain the pathogenesis of acquired nontraumatic normal pressure CSF leaks. Pneumatization of the sinuses along the skull base in conjunction with pulsatile and intermittently increased CSF and central nervous system pressures may result in slow erosion of the thin bone at the sinocranial junction and lead to a spontaneous CSF fistula over time. They also pointed out that the floor of the middle cranial fossa contains arachnoid villi, in association with the middle meningeal veins, which may enlarge over several decades into bony dehiscences secondary to constant pulsatile

Figure 1. Axial CT scan demonstrating the left posterolateral sphenoid defect with soft tissue filling the sinus.

Figure 2. Coronal iopamidol CT reveals a left cranial base defect with enhancement within the left sphenoid sinus.

CSF pressures. These dehiscences in the sinocranial junction may lead to dural herniation and the formation of encephaloceles and meningoencephaloceles. As these expand the dural cover thins and may lead to CSF fistulization.

In a study of 138 adult sphenoid bones, Hooper³ noted 27 defects that could predispose to CSF fistulas. The defects were surrounded by very thin bone and appeared to have been gradually enlarged by pressure erosion. A congenital etiology, however, could not be ruled out. Van Alyea,⁴ in a study of 100 sphenoid bones, noted no dehiscences, making the incidence of sphenoid defects difficult to determine.

The evaluation of patients presenting with nontraumatic CSF rhinorrhea centers on localization of the fistula. Physical examination alone is inadequate for precise localization even in the presence of an obvious unilateral leak. CT scan infrequently reveals abnormalities other than tumor or hydrocephalus. Radionuclide cisternography with pledget scanning is rarely better at localizing a leak than clinical examination, but it is advocated for confirmation of the diagnosis of CSF rhinorrhea.⁵ CT cisternography with metrizamide or iopamidol is described as the most specific modality for localization of an active CSF fistula.⁶ Intrathecal in-

jected fluorescein is most helpful in intraoperatively localizing the leak.⁷

Treatment of nontraumatic CSF leaks is surgical, with conservative medical management rarely being warranted in established fistulas. Lumboperitoneal CSF shunts as the primary therapeutic modality are advocated by Bret et al⁸ for patients who are elderly, have medical conditions that contraindicate a more extensive procedure, have impaired CSF dynamics, or have an unknown leakage site. Shunts are also indicated for recurrent or persistent CSF leakage.

Extracranial approaches have been established as having low morbidity and mortality and a high success rate. They are the preferred initial treatment of CSF fistulas in patients without intracranial injuries or tumors. The sphenoid sinus may be approached transeptal via a transnasal or sublabial incision, transthemoid via a Lynch incision or lateral rhinotomy, and endoscopically.^{5-7,9} The preferred route may depend on the underlying etiology of the fistula, its location within the sinus, and the surgeon's expertise with the various techniques.

The two sisters presented in this report had no history of accidental or iatrogenic trauma. CT cisternography and surgical exploration revealed identically located left posterolateral sphenoid defects without evidence of associated dehiscences. Given the lack of evidence for a traumatic etiology and the similarities between the sisters, we propose a congenital predisposition for their development of a CSF fistula. Despite the presence of a congenital skull base defect the sisters did not present with CSF leaks until adulthood. This, in part, may be due to the cumulative effect of the physiological CSF forces previously described. Obesity, with its association with increased intracranial pressures,^{10,11} was likely contributory in both cases, and intermittent rises in intracranial pressure due to poorly controlled hypertension and asthma are hypothesized to have been a significant factor in patient 2. Both patients were treated successfully via extracranial approaches, although patient 1 required an indwelling CSF shunt to address her altered CSF dynamics.

CONCLUSIONS

1. Nontraumatic CSF leaks are a rare entity, and their presentation in siblings with identical cranial base defects is strongly suggestive of a congenital etiology.
2. Constant pulsatile CSF forces superimposed on intracranial pressure fluctuations due to concurrent medical conditions may predispose patients with congenital cranial base defects to CSF fistula formation.

Figures 3, 4. Corresponding axial MRI T₁ and T₂ images reveal an anteromedial middle fossa defect with herniation of soft tissue and CSF into the sphenoid sinus.

Figure 5. Coronal iopamidol CT demonstrating the left posterolateral sphenoid sinus defect.

Figure 6. Coronal iopamidol CT demonstrating enhancement within the left sphenoid sinus.

REFERENCES

1. Ommaya AK, Di Chiro G, Baldwin M, Pennybacker JB: Non-traumatic cerebrospinal fluid rhinorrhea. *J Neurol Neurosurg Psychiatry* 31:214–225, 1968
2. Kaufman B, Nulsen FE, Weiss MH, et al: Acquired spontaneous, nontraumatic normal-pressure cerebrospinal fluid fistulas originating from the middle fossa. *Neuroradiology* 122:379–387, 1977
3. Hooper AC: Sphenoidal defects—A possible cause of cerebrospinal fluid rhinorrhea. *J Neurol Neurosurg Psychiatry* 34:739–742, 1971
4. Van Alyea OE: Sphenoid sinus. Anatomic study, with consideration of the clinical significance of the structural characteristics of the sphenoid sinus. *Arch Otolaryngol* 34:225–253, 1941
5. Hubbard JL, McDonald TJ, Pearson BW, Laws ER Jr: Spontaneous cerebrospinal fluid rhinorrhea: Evolving concepts in diagnosis and surgical management based on the Mayo Clinic experience from 1970 through 1981. *Neurosurgery* 16:314–321, 1985
6. McCormack B, Cooper PR, Persky M, Rothstein S: Extracranial repair of cerebrospinal fluid fistulas: Technique and results in 37 patients. *Neurosurgery* 27:412–417, 1990
7. Persky MS, Rothstein SG, Breda SD, et al: Extracranial repair of cerebrospinal fluid otorhinorrhea. *Laryngoscope* 101:134–136, 1991
8. Bret P, Hor F, Huppert J, et al: Treatment of cerebrospinal fluid rhinorrhea by percutaneous lumboperitoneal shunting: Review of 15 cases. *Neurosurgery* 16:44–47, 1985
9. Papay FA, Maggiano H, Dominquez S, et al: Rigid endoscopic repair of paranasal sinus cerebrospinal fluid fistulas. *Laryngoscope* 99:1195–1201, 1989
10. Hannerz J, Greitz D, Ericson K: Is there a relationship between obesity and intracranial hypertension? *Int J Obesity Rel Metab Disord* 19:240–244, 1995
11. Wall M: Idiopathic intracranial hypertension. *Neurol Clin* 9: 73–95, 1991