Cerebrospinal fluid (CSF) is produced by the choroid plexus of the lateral third and fourth ventricles at a rate of 0.35 mL/min. The CSF flows into the subarachnoid space and is absorbed by the arachnoid villi in the sagittal sinus. Normal CSF pressure is 5 to 15 cm H₂O. Neurologic symptoms may occur when CSF pressure reaches more than 15 to 20 cm H₂O. Three concomitant factors are needed for CSF leakage: an osseous defect, a meningeal disruption, and a pressure gradient.

Cerebrospinal fluid rhinorrhea may be classified as a function of its site, its cause, and the intensity of intracranial pressure. Nontraumatic fistulas with normal intracranial pressure (4% of all fistulas) constitute a challenge because of their unclear pathophysiologic features. Some authors have used the term spontaneous to include CSF leaks due to a tumor, a delayed CSF leak after head trauma, or CSF leaks associated with congenital malformations of the skull base. Currently, the term spontaneous is associated with CSF leaks without any known cause. For those reasons, it seems preferable to use the term idiopathic spontaneous CSF leak.

Some risk factors of idiopathic spontaneous CSF rhinorrhea in the sphenoid sinus have been described in the literature: skull base malformation; overpneumatized sphenoid sinus, particularly with a lateral extension (present in 16%-27% of adults); empty sella syndrome; and obesity. Obesity is associated with an increase of abdominal and thoracic pressures, leading to cranial venous congestion and permanent benign intracranial hypertension.

The main complications of persistent CSF leakage are meningitis or brain abscess, which are potentially fatal. For those reasons, abnormalities must be repaired surgically. During the last decade, the management of CSF rhinorrhea has been changed by the introduction of endoscopic sinus surgery. However, most of the recent publications on endonasal sinus surgery for CSF rhinorrhea treatment report inconsistent efficacy of the method varying from 85% to 95% for all types of fistulas. The aim of this article is to report 2 exceptional cases of spontaneous idiopathic skull base fistula through the clivus treated by the endoscopic approach.

REPORT OF CASES

CASE 1

An 81-year-old man presented in May 2004 with spontaneous, abundant, and isolated clear fluid discharge from the right nostril that had started 4 months earlier. The patient was not obese, had no contributing medical conditions, and had never had head trauma. The results of neurologic and otorhinolaryngologic examinations were normal except for clear and abundant fluid discharge from the right nostril coming from the posterior part of the nasal cavity (30° endoscope; Karl Storz GmbH & Co KG, Tuttingen, Germany). The glucose content of the discharge was compatible with the presence of CSF (30 mg/mL; to convert to micromoles per liter, multiply by 0.0555).
High-resolution CT (1 mm thick, axial) localized the site of the bone defect at the median part of the clivus. The right sphenoid sinus was filled with fluid. There was no sign of empty sella syndrome (Figure 1). After intrathecal injection via lumbar puncture, CT scan cisternography showed a meningeal abnormality with leakage of the contrast agent from the subarachnoid space in the right sphenoid sinus. Magnetic resonance imaging (MRI; axial T1-weighted sequence with gadolinium) showed an interruption of the sphenoidal mucosal enhancement in front of the bone defect at the medial part of the clivus. The right sphenoid sinus was filled with fluid (Figure 2).

Endoscopic nasal surgery, including a posterior ethmoidectomy, was performed with the patient under general anesthesia. A large sphenoidotomy was performed with a computer-assisted navigator (Digipointeur; Collin ORL, Bagneux, France). The bone abnormality with the CSF leak was found in the middle part of the clivus bone. After removing the mucosa from the edge of the bone abnormality, the fistula was closed using fat harvested from subcutaneous tissue of the patient’s abdomen. The area of the grafted fistula was treated with biological glue (0.5-1 mL of Tissucol Fibrin Glue; Medtronic, Minneapolis, Minnesota). At the end of surgery, 3 pieces of nasal pack Merocel (Medtronic, Minneapolis) were placed in the superior middle meatus and the nasal cavity and were removed 6 days later. The CSF leak immediately stopped. The surgery lasted 70 minutes. Antibiotic injection was given on the first day of surgery, followed by an oral antibiotic for a week. The patient was sent home after 3 days.

Postoperative checkups were performed at 1 month and then every 6 months for 3 years. The MRI checkups were performed 4 months and 1 year after surgery. The abnormality was closed using fat tissue without evidence of leakage. Three years after surgery, the patient had no sign of rhinorrhea.

CASE 2

A 34-year-old woman was admitted to the Infectious Disease Department of the University of Paris—
Descartes with a diagnosis of meningitis. The cerebral CT scan was normal, but opacity of the right sphenoid sinus was reported. The CSF was sterile. The results of neurologic and otorhinolaryngologic examinations were normal. The CT revealed a right sphenoiditis. An endoscopic sphenoidotomy was performed with the patient under general anesthesia, and all symptoms stopped after the surgery. Six months later, the same symptoms recurred, and the patient was admitted to our Department of Otorhinolaryngology. For the first time, the nasal endoscopic examination revealed a clear fluid discharge from the right nostril. The glucose content was compatible with the presence of CSF (30 mg/mL). A new radiologic evaluation (CT and MRI) (Figure 3) was performed, including CT scan cisternography, which showed an abnormal clivus ossification filled with fluid. A revision endoscopic nasal surgery with a computer-assisted navigator was performed. The bone abnormality was revealed at the middle part of the clivus bone (posterior wall of the right sphenoid sinus). The defect was closed using fat harvested from the subcutaneous abdominal tissue of the patient. Biological tissue glue and nasal packing were applied around the fat graft. Postoperative imaging (Figure 4) performed 4 months later showed no evidence of leakage. Throughout the follow-up period (every 3 months for a total period of 1 1/2 years), there was no evidence of leak recurrence. Eighteen months after surgery, the patient had no signs of rhinorrhea or episodes of meningitis.

**COMMENT**

In the current literature, the prevalence of idiopathic spontaneous CSF rhinorrhea represented between 3% and 4% of all cases of CSF leaks. Recently, this prevalence has increased to between 14% and 46%. The reason for this increase in the recent literature is unclear, although better recognition of this
The cause of these 2 reported cases of idiopathic spontaneous leakages through the clivus is unclear. Our 2 patients were not obese, had no manifestation or radiologic evidence of increased intracranial pressure, and did not have empty sella syndrome. These clinical findings disagree with most of the presentations described in the literature. Mirza et al.\textsuperscript{14} found that 88% of spontaneous CSF leaks had intracranial pressure augmentation and mainly occurred in obese women. This high intracranial pressure could explain the rate of recurrence after surgery (ie, 50% recurrence rate).\textsuperscript{11}

The transcranial approach has been highly successful in repairing high-pressure leaks and, in some cases, extreme lateral recess of the pneumatized sphenoid sinus.\textsuperscript{13} However, endonasal endoscopic techniques are more commonly indicated and better established than the transcranial approach because they are less invasive and decrease morbidity. In addition, endonasal endoscopic techniques decrease the risk of alteration to the patient’s sense of smell and decrease the duration of hospital stay. These approaches also give a better exposure of the sphenoid than a transcranial approach would. In our 2 patients, the limited endoscopic approach was used with success and no morbidity. We also used an image-assisted navigator system during exploration of the skull-base abnormality. This system provided significant assistance for the localization of the defect. Intrathecal fluorescein dye is frequently used at the time of surgery for endoscopic CSF leakage repair. However, in our 2 patients, the CSF leakage was easily located, and this intrathecal injection was not necessary.

Various grafting materials were used to eliminate CSF leakages (eg, abdominal fat, fascia lata, rotation middle turbinate flap, nasal septum mucosal flap, septal cartilage, and free muscle flap). Synthetic materials, such as hydroxyapatite cement, which is a biocompatible material that slowly replaces itself with bone, have been recently used in sphenoid fistula repair. Nevertheless, the application of hydroxyapatite in the ethmoid sinuses for repair of CSF leakages was condemned in this article. Therefore, hydroxyapatite cement should not be listed as a viable first-line approach for endoscopic CSF leakage repair.\textsuperscript{15} The choice of grafting material depends on the surgeon’s experience, the site and size of the fistula, and the intensity of intracranial tension. The material used in our patients was a combination of fat and biologic tissue glue, which turned out to be satisfactory, safe, and cost effective. In addition, these materials, unlike polymers, did not present hazards of extrusion or infection.

Cerebrospinal fluid leakages through the posterior wall of the clivus are rare. Fewer than 5 cases are described in the literature. A recent integrated radiologic diagnostic workup with a logic algorithmic pattern is mandatory, in addition to clinical and biochemical diagnosis, for accurate and precise identification of the location of the fistula. Endonasal endoscopic surgery with use of an image-assisted navigator is a simple approach. The use of fat from the subcutaneous tissue of the patient and biologic tissue glue created an optimal graft for closing these fistulae.

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