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Congenital transsphenoidal meningocele: case report and review of the literature

Catherine Blaivie[a], Thomas Lequeux[a], Stelios Kampouridis[b], Stephanie Louryan[c], Sven Saussez[c,d,*]

[a]Department of Otorhinolaryngology, Head and Neck Surgery, CHU Saint-Pierre, Brussels, Belgium
[b]Department of Medical Imaging, Head and Neck Surgery, CHU Saint-Pierre, Brussels, Belgium
[c]Laboratory of Anatomy and Embryology, Faculty of Medicine, Université Libre de Bruxelles, Brussels, Belgium
[d]Laboratory of Anatomy, Faculty of Medicine and Pharmacy, University of Mons-Hainaut, Mons, Belgium

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Abstract Intrasphenoidal meningoencephalocele is a rare clinical entity. Its origin can be congenital, traumatic, tumoral, or spontaneous. Presenting as cerebrospinal fluid fistula with rhinorrhea, the diagnosis and treatment of this uncommon disease is a real challenge for the otorhinolaryngologist. We report a case of sphenoidal meningocele treated using an endoscopic procedure and review the literature regarding its congenital origin.

1. Case report

A 73-year-old woman with a history of spontaneous left rhinorrhea was referred to the ENT department of Saint-Pierre Hospital. The patient was obese and had considerable cardiovascular comorbidity. No history of previous craniofacial trauma or surgery was established. Physical examination showed no particularities, and the nasal endoscopy revealed clear fluid pulsing from the left sphenoid ostium. Computed tomography (CT) demonstrated a paramedial left lacuna in the roof of the sphenoid sinus, with abnormal material in the sinus (Fig. 1). T2-weighted magnetic resonance imaging pointed out a left sphenoid sinus filled with liquid and presenting abnormal hydrostatic features in its upper part. Sagittal sequences suggested a dural defect in the posterosuperior wall of the sinus with a communication between it and the suprasellar cistern (Fig. 2). The patient developed a bacterial meningitis that required intravenous antibiotherapy (ceftriaxone and metronidazole). She remained confused after normalization of the infectious parameters. A cerebral CT revealed 2 right subdural hematomas of chronic onset with acute emphasis.

The sphenoid sinus was exposed using a transsphenoidal endoscopic approach. The ruptured meningocele appeared through a bony defect in the posterosuperior wall of the sinus. A duraplasty was performed after first applying a layer of septal cartilage. Afterward, the sinus mucosa was completely removed to prevent the onset of an iatrogenic mucocele and then the sinus was filled with abdominal fat. Finally, a second layer of septal cartilage was put in place before sealing the

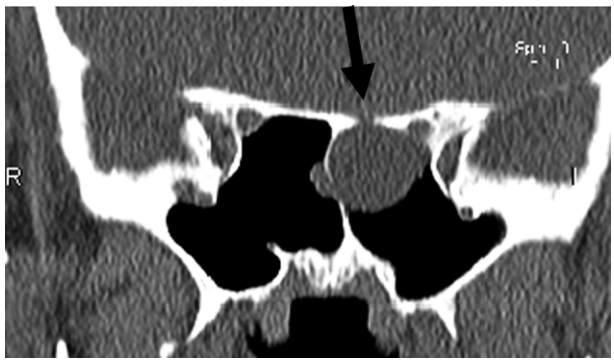

Fig. 1. Coronal CT scan section, with demonstration of a narrow lacuna (arrow) at the roof of the sphenoid sinus, with associated round-shaped material in the cavity.

* Corresponding author. Department of Anatomy, Faculty of Medicine, University of Mons-Hainaut, Pentagone 1B-Avenue du Champ de Mars, 6, B-7000 Mons, Belgium. Tel.: +32 65 37 35 62; fax: +32 65 37 35 57.
E-mail address: sven.saussez@umh.ac.be (S. Saussez).

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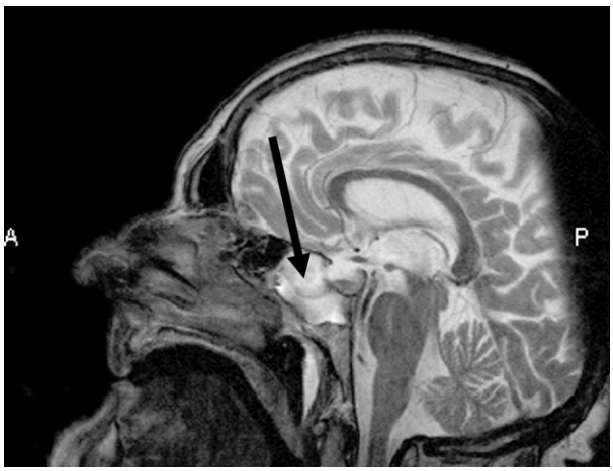

Fig. 2. Sagittal T2 MR section. The material found in the sphenoid sinus is hyperintense (arrow) and corresponds to liquid.

reconstruction with biologic glue (Tissucol, Baxter, Vienna, Austria). No lumbar drainage was used and the patient was discharged after 10 days without evidence of recurrent rhinorrhea. Four-month follow-up did not reveal any sign of transsphenoidal cerebrospinal fluid (CSF) leak but, unfortunately, the patient died a few weeks later of pulmonary complications associated with chronic heart failure.

Although the follow-up period in our case may be considered too short, the convincing rationale of the technique leads us to conclude that the result would have lasted.

2. Discussion

2.1. Etiopathology of CSF leaks

The etiologies of CSF rhinorrhea can be subdivided into traumatic and nontraumatic causes (Table 1). The first group includes cranial traumas, which represent the most frequent causes (80%) of CSF rhinorrhea [1]. Surgery on the skull base accounts for 16% of cases. Postoperative rhinorrhea is most frequently found after resection of the skull base near the cribriform plate, transsphenoidal hypophysectomy, and ethmoidectomy [2].

Nontraumatic CSF fistulae are a rare entity (3-4%), mostly secondary to the presence of an intra- or extracranial process [3,4]. High-pressure leaks are associated with tumors and hydrocephalus. Tumoral expansion induces extra- or intracranial bone erosion, but also contributes to increased hydrocephalus due to greater CSF pressure caused by mass effect or obstruction of normal CSF flow [5,6]. On the other hand, congenital and acquired defects (osteitis, excessive aeration, focal erosion, and encephalocele) are characterized by low- or normal-pressure fistulae [5-8].

Knowing the complex ontogeny of the sphenoid bone is crucial to understand the pathogenesis of the nontraumatic acquired and congenital forms of skull base defects. The sphenoid bone develops from numerous ossification centers, which are initially separated by synchondroses [9]. The coalescence of these centers is a complex process, which may result in localized lack of bone tissue. The anterior sphenoid bone, the lesser wings, the posterior sphenoid bone, the greater wings, and the lateral pterygoid processes originate as independent cartilaginous precursors (only parts of the medial pterygoid process are built up by membranous ossification). After their ossification (at the third month of fetal life), the union of these several parts gives rise to the complex sphenoid bone (at the time of birth and during the neonatal period). When posterior fusion is incomplete (more precisely on the posterolateral side of the sphenoid sinus), a lateral craniopharyngeal canal is created, called Sternberg's canal (incidence of 4% in adults). This congenital bony defect can communicate with the sphenoid sinus after the necessary pneumatization has taken place. Indeed, the fusion plates offer special resistance to pneumatization; therefore, sphenoidal cranial base defects at fusion plates are more likely to be of congenital origin than to have been

Table 1
Etiologies of CSF leaks

Traumatic	Nontraumatic	Intracranial	High pressure	Olfactory neuroblastoma
Cranial trauma (skull base fractures, pneumocephalus)				Meningioma
Skull base surgery, surgical trauma (sphenoethmoidectomy, transsphenoidal hypophysectomy, etc)				Clival chordoma
				Craniopharyngioma
				Glioma
				Pituitary tumors (prolactinoma, Rathke's cyst, adenoma)
			Low or normal pressure	Encephalocele
				Meningocele
			High pressure	Angiofibroma
		Extracranial		Ethmoid osteoma
			Low pressure	Previous radiation therapy to the head and neck region
				Chronic suppurative sinus disease
				Syphilis
				Leprosy
				Influenza

acquired [10-12]. Moreover, Lanza et al [6] pointed out that the floor of the middle cranial fossa harbors small perforations in the bone, or so-called pit holes, of unknown origin. Normal intracranial pressure dynamics may cause enlarged dural/bone defect. This can also explain how defects appearing over the underlying lateral extension of the sphenoidal sinus (16-27% of adults have well-developed lateral recesses of the sphenoidal sinus [13]) may induce brain herniation and lead to CSF leak via the sinuses. Caution should be used in distinguishing this type of fistula from the more common medial varieties, originating at the perisellar and ethmoidal regions [8].

Encephaloceles are uncommon and are defined as herniations of brain parenchyma in the paranasal cavities. However, many skull base defects occur after prior trauma or surgery, so otorhinolaryngologists must be aware of the potential for spontaneous encephaloceles.

According to Buchfelder et al [14] and Daniilidis et al [15], only 16 cases of intrasphenoidal encephalocele have been mentioned in the international literature. For most of them, the bony defect is localized in the lateral or posterolateral walls of the sphenoidal sinus, involving mainly women, with an average age between 40 and 67 years. The clinical features consist mainly of spontaneous CSF rhinorrhea, recurring meningitis, and headaches [14]. Up to now, only 1 case of congenital intrasphenoidal meningocele has been described [11]. The origin of the meningocele was localized in the right parasellar region and was confirmed surgically. It appeared to be due to the persistence of the lateral craniopharyngeal canal (Sternberg's canal) and was then considered as congenital intrasphenoidal meningocele.

In our case report, we could not find any history of trauma, tumor, intracranial infection, or a surgical procedure involving the sellar region or the paranasal sinuses. Therefore, we propose that the intrasphenoidal meningocele observed through a small bone defect localized in the sphenoid roof could be a persistent Sternberg's canal. To our knowledge, our case report constitutes only the second case of congenital intrasphenoidal meningocele.

The cerebral CT of our patient revealed the existence of 2 subdural hematomas. Among the 16 previously published cases of intrasphenoidal encephalocele, only one has been described with subdural hematoma [16].

3. Conclusion

We have related a case of intrasphenoidal meningocele revealed by a spontaneous CSF rhinorrhea. The sinus CT allowed us to locate a bony defect in the sphenoid roof. Based on knowledge of the complex ontogeny of the sphenoid bone and on the lack of previous trauma, we propose that this skull base defect was congenital and corresponds to the persistence of a Sternberg's canal.

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