Mucocele of the sphenoid sinus: A rare entity to keep in mind

J Marrakchi¹, S Mahfoudhi¹, Methnani¹, H Chahed¹, M Jrad², H Mizouni², N Beltaief³, R Zainine¹, G Besbes¹, A charfi³

1 Department of Oto-Rhino-Laryngology and Cervico-Facial Surgery of ‘Rabta’ Hospital Tunisia, Medecine Faculty, El Manar University, Tunisia
2 Department of Radiology of ‘Rabta’ Hospital Tunisia, Medecine Faculty, El Manar University, Tunisia
1 Department of Oto-Rhino-Laryngology and Cervico-Facial Surgery of SIDI Bouzid Hospital Tunisia.

ABSTRACT
Isolated sphenoid sinus mucocele (SSM) is a rare entity that can result in serious sequelae if diagnosis and treatment are inappropriately delayed. Typically, mucoceles are asymptomatic, and they are accidentally identified after computed tomography scan or magnetic resonance imaging of the maxillofacial area performed for other pathologic issues. We report a case of isolated SSM that only presented with headache for over an year, and also review the literature regarding surgical management of such entity.

Keywords: Endoscopic, mucocele, sphenoid sinus, imaging.

BACKGROUND
Mucoceles are benign, encapsulated, expansile, locally destructive masses within cavities, filled with mucous and lined by epithelium. Only 1-2% of all paranasal sinus mucoceles are located in the sphenoid sinus [1]. Typically, mucoceles are asymptomatic, and they are accidentally identified after computed tomography (CT) scan or magnetic resonance imaging (MRI) of the maxillofacial area performed for other pathologic issues. Symptoms, if present, are unspecified and result from mechanical pressure on neighboring structures and/or involvement of nerves in the inflammatory process. The most common symptoms include headache, visual loss and palsies of the III and VI cranial nerve [2].

We report a case of isolated sphenoid sinus mucocele (SSM) that only presented with headache for over an year, treated with the endoscopic technique.

CASE REPORT
A 69-year-old woman was examined by a neurologist for headache over an year. The headache was intermitted, but recently had become more severe in intensity. The neurological examination was normal as well as the ophthalmological examination. Nasal endoscopy did not reveal any anomalies. In order to exclude other symptomatic headaches, brain CT was performed and showed no intracerebral structural lesion. However, it revealed an expansile mass centered in the sphenoid sinus measuring 31*35 mm with erosion of the bony wall over 7 mm (Figure 1).

The MRI study of the maxillofacial area, visualized a cystic mass of the sphenoid sinus that had a high signal intensity on T2-weighted images and on T1-weighted images, without enhancement after injection of contrast medium and with extension to the left parapharyngeal space (Figures 3, 4).

Figure 1 : CT examination (Sagittal view) showing a mass centered in the sphenoid sinus, measuring 31*35 mm with erosion of the bony wall over 7 mm

Corresponding Author : Sana Mahfoudhi
E-mail adress: mahfoudhislana@gmail.com
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**DISCUSSION**

Isolated SSM is a rare entity that can result in serious sequelae if diagnosis and treatment are inappropriately delayed. International literature assesses its incidence between 10% and 17% of sphenoid isolated benign lesions [3, 4]. Etiology is still unknown, but different infective or traumatic factors have been described as possible pathogenetic issues [5]. It commonly occurs in the third and fourth decades of life and is extremely rare in children [6].

Histologically, the mucocele consists of a thin layer of mucus-excreting cells and a large cavity of liquid or semisolid material. Although benign, SSM may involve vital structures, including the dura, pituitary gland, optic chiasm, cavernous sinus, pterygoid canal and nerve, internal carotid artery, and cranial nerves (III, IV, V, and VI) which are vulnerable to injury from the sphenoid lesion. Complications include permanent diplopia, blindness, meningitis, cavernous sinus thrombophlebitis, and the formation of carotid artery aneurysms [7]. Outset symptoms depend on tumor dimensions; more frequently, they are characterized by localized headache, pain, and persistent sinusitis not responding to medical therapy. The most commonly reported symptoms when complication occurs were headache (89% of cases), decreased visual acuity (57%), oculomotor palsy (56%), exophthalmia (25%) and nasal symptoms (12%) [8].

Our patient suffered from severe headache. It has been suggested that headache results from stretching of the dura over planum sphenoidale.

CT imaging of patients complaining of rebel headaches with normal neurological examination can clearly reveal the lesion. CT scan with bone algorithm is nowadays the elective x-ray imaging examination; MRI instead is a complementary study useful in cases with intracranial or intraorbital extension.

Mucoceles may show various imaging features, depending on their protein contents and possible infection. They often have low attenuation on CT, a low signal on T1-weighted MRI and a high signal on T2-weighted MRI, due to their high water content [9]. There is usually no enhancement, or at most marginal enhancement, on CT and T1-weighted MRI, whereas many of the lesions in the differential diagnosis show contrast enhancement.

Differential diagnosis was to be made with chronic sphenoid sinusitis, a fungal ball, benign neoplasms such as inverted papilloma, and rarely with malignant neoplasms [10]. The purpose of surgical treatment of the SSM is to create a large ostium allowing drainage into the sphenethmoidal recess. Surgical approach to sphenoid sinus mucocele could be either traditional or endoscopic. At the present time, the endoscopic approach represents the gold standard for sphenoid sinus mucocele treatment because it allows the best view of the sphenoid sinus and grants a better restoring of respiratory function and a higher compliance from the patient [11]. Endoscopic technique is described in many studies; it can be performed with a direct paraseptal approach with the individuation of natural sphenoid sinus ostium that is positioned 1 to 1.5 cm far from the superior border of the choanae.

The transthyroidal approach was described. In this case, the endoscope passes laterally to the middle turbinate bone, and an anterior and posterior ethmoidectomy is performed. Afterward, the superior turbinate is isolated, and the lower part is excised. Subsequently, inferiorly, and medially, the natural sphenoid sinus ostium can be individuated, and it is then enlarged. The current surgical indication for mucocele treatment is the only marsupialization of sphenoid sinus drainage without excision of the whole mucosa [4,12]. This approach offers lower morbidity and complications and prevents recurrence. Postoperative follow-up for patients who underwent endoscopic surgery must deal with periodic control of mucous membrane healing.

**CONCLUSION**

Mucocele involving the sphenoid sinus is a rare entity. Clinicians should suspect this entity if a patient presents chronic, non-specific symptoms and/or with visual symptoms. The clinical examination may be normal. Imaging is crucial.
to diagnosis. Endoscopic drainage is a safe and effective in eradication of disease.

Competing interests: The authors declare no conflicts of interest.

REFERENCES