Mucocelo esfenoidal: "Doença-camaleão"

Sphenoid mucocele: "A chameleon-like disease"

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ABSTRACT

Paranasal Sinus Mucocele is an entity caused by the accumulation of mucinous content in a pseudoencapsulated form at the level of paranasal sinuses. Its clinical presentation is variable, can produce intracranial and/or vascular compromise but it can also be discovered as a casual finding. Diagnosis is based on clinical suspicion and imaging tests, to identify the nature of the lesion. The treatment of choice is the endoscopic drainage to avoid future recurrences. In this article we present three cases of patients diagnosed with mucocele with different clinical manifestations that, thanks to a suspected diagnosis, therapeutic management was performed promptly, avoiding complications and recurrences on follow-up.

Keywords: Mucocele, sphenoid, ophthalmoplegia, endoscopy, sinusitis.

INTRODUCTION

Mucoceles are cystic masses located in the paranasal sinuses, with mucinous content covered by a columnar pseudostratified epithelium^[1]. Its evolution is variable and sometimes can reach a high expansive potential and produce erosion of adjacent bony walls.

Its physiopathology isn't yet fully identified, however, one of the most accepted hypotheses is the cystic degeneration of the respiratory epithelium in the mucosa, due to obstruction or insufficient drainage. This could be caused by the presence of embryonic elements,

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previous aggressive surgeries or recurrent episodes of rhino-sinusitis^[2].

In patients with sphenoid mucocele, the most frequent symptoms are headache and ophthalmological symptoms: periorbital pain (39%), exophthalmos (24%), decrease in visual acuity (17%), diplopia (13%), and alteration of the extraocular musculature (5%)^[2]. However, it can be asymptomatic and discovered as an incidental finding (through CT or MRI).

We describe three clinical cases of sphenoid mucoceles diagnosed, in our third level hospital center, explaining the management and treatment applied. We also present a short review of the literature about this topic.

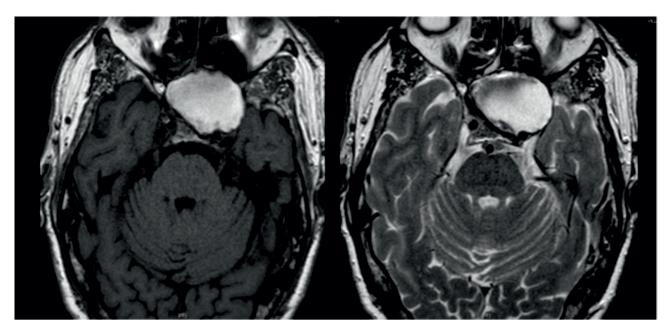
Case number one:

A 29-year-old woman with a history of Raynaud's disease and occasional rhino-sinusitis was followed by the Neurosurgery service due to a left hemifacial pain of several weeks. The physical examination provided data suggestive of trigeminal neuralgia. A nuclear magnetic resonance (MRI) was requested, identifying the presence of a lesion at the level of the left sphenoid recess invading the pterygoid fossa and the infratemporal region. Due to these findings, she was referred to our otorhinolaryngology service in order to assess a possible surgical treatment (fig 1). It was decided to perform an endoscopic approach. During the surgery, the entire anterior osteomeatal complex was opened, facilitating the sphenoid sinus exposure. After opening, a cystic mass of mucoid content was observed, occupying, practically the entire sinus, reaching the internal carotid and clinoid processes. During its postoperative evolution, a complete remission of its symptoms was observed after 2 weeks of follow-up.

Case number two:

A 53-year-old woman with a history of bilateral massive nasosinusal polyposis, who required endoscopic surgical treatment 3 years before, was referred from a regional hospital for presenting symptoms of progressive diplopia associated with ptosis in the right eye and frontal headache. Clinical examination identified paresis of extraocular musculature with preservation of sight. Fiber optic nasolaryngoscopy showed edema of the nasal mucosa and postsurgical changes due to previous

FIGURA 1
Magnetic resonance image (MRI) showing a lesion with hyperintense mucoid content in T1 and T2 in left sphenoid sinus suggestive of mucocele



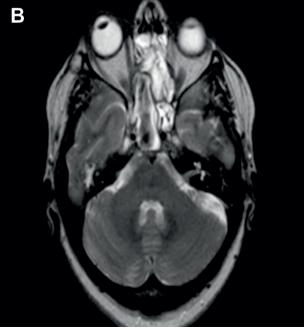
surgery. CT scan and MRI showed an inflammatory process occupying the right sphenoid sinus with a possible involvement of the third cranial nerve, without infiltration (fig 2). Initially, it was decided to treat the acute episode with third-generation cephalosporins and intravenous corticosteroids, deferring surgical treatment. Patient evolution didn't show any clinical improvement and so surgery was performed. During the endoscopic

procedure, a mucocele was identified with significant involvement of the right sphenoid sinus eroding its posterior bony wall. During the opening of the mucocele, a protrusion of the optic nerve was observed, which was intact. Sellar structures and the internal carotid artery were also intact. Subsequently, the patient had an adequate postoperative evolution within a month of surgery.

FIGURA 2

- (A). Computed Tomography (CT) showing a non expansive lesion remodeling its bony limits suspicious of a sphenoid mucocele.
- (B). MRI show a hyperintense lesion in T2 compatible of sphenoid mucocele

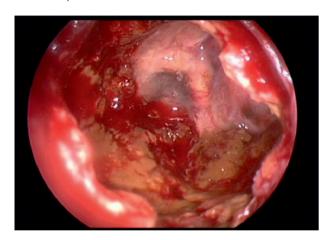




Case number three

A 72-year-old male patient with a history of carotid atheromatosis was seen in outpatient clinics for several months due to a tinnitus and progressive hearing loss. The audiometric tests evidenced a symmetrical bilateral sensorineural hearing loss, without any additional data. MRI was requested showing an incidental finding of a hyperintense lesion in T1 with gadolinium and hypointense in T2 of a probable proteinaceous material at the level of the left sphenoid sinus. It wasn't identified affectation of others neighboring structures. Do to the clinical and potential local aggressiveness of the mass, surgical management was considered. During the endonasal endoscopic surgery, after opening the anterior wall of the sphenoid sinus, it was obtained a mucopurulent material. It was possible to observe the erosion of all bony walls of the sphenoid sinus, exposing the internal carotid artery, parasellar structures and the optic nerve (fig 3). Samples were taken for both microbiology and pathological analysis, with the final result of a mucocele. The postoperative evolution was satisfactory.

FIGURA 3Anterior sphenoidotomy. Mucinous content, internal carotid artery and sella exposed.



DISCUSSION

The anatomist Rouge documented the first sphenoid mucocele in the literature in 1872. Afterwards, Rolland and Berg completed this description in 1889^[2]. Despite being an infrequent pathology, 1% of the cases are located in the sphenoid sinus, and around 200 cases were published in recent years^[2]. Its incidence according to location is, in descending order, frontal (65%), ethmoidal (25%), maxillary (10%) and sphenoid (1%)^[3].

This pathology is associated with chronic rhinosinusitis, allergic rhinitis, trauma and aggressive sinus interventions. However, just one of our patients had history of nasal polyposis. The age of presentation goes between 30-60 years with a 1:1 gender ratio.

The sphenoid sinus is in proximity of various intracranial structures such as the internal carotid artery, pituitary gland, clivus, orbit or cavernous sinus (including the course of cranial nerves that cross it), making any growth in this cavity able to produce a severe outcome.

Initially this pathology produces nonspecific symptoms such as headache (70-90%) or occasional rhinorrhea. After some time, it can grow and increase the intrasinusal pressure causing erosion, remodeling and expanding through the bony walls and progressively cause periorbital inflammation (39%), diplopia (13%), exophthalmos (24%) and decrease visual acuity (17%) etc^[3]. Our patients present a big variety of different symptoms^[4].

The fundamental pillar for the diagnosis, in spite of the clinical suspicion, is radiological techniques with an axial and coronal paranasal sinus CT. This allows making an assessment of the intracranial and bony extension of the pathology.

The use of MRI provides additional information, about the nature of the lesion, since mucoceles vary their protein concentration during their evolution, altering the viscosity of their content. This translates into greater hyperintensity in T1 in the short term and more in T2 at long term^[4].

The definitive treatment is undoubtedly the endoscopic surgery, generating an endonasal drainage path by performing an anterior transnasal sphenoidotomy^[5]. Endoscopic approaches have shown better results than open surgery (transeptal sphenoidectomy and ethmoidectomy) being described between 0.9-2.2% of recurrences and better aesthetic results^[5].

CONCLUSION

The mucocele is an entity with "chameleon-like" symptoms manifesting itself according to the affected paranasal sinus and the structures that it can compromise. Therefore, it should be suspected, principally in patients with previous rhinologic interventions or with history of chronic rhino-sinusitis, and allergic rhinitis, using the adequate imaging algorithms for their study and subsequent endoscopic drainage.

Protecção de pessoas e animais

Os autores declaram que os procedimentos seguidos estavam de acordo com os regulamentos estabelecidos pelos responsáveis da Comissão de Investigação Clínica e Ética e de acordo com a Declaração de Helsínquia da Associação Médica Mundial.

Confidencialidade dos dados

Os autores declaram ter seguido os protocolos do seu centro de trabalho acerca da publicação dos dados de doentes.

Conflito de interesses

Os autores declaram não ter nenhum confito de interesses relativamente ao presente artigo.

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