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Mucoceles of the sphenoidal sinus: a report of four cases and review of the literature

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Abstract. *Mucoceles of the sphenoidal sinus: a report of four cases and review of the literature.* Mucoceles of the sphenoidal sinus represent less than 2% of paranasal sinus mucoceles. The purpose of this paper is to report our experience in patients with mucoceles of the sphenoidal sinus. Four patients with this localization were observed in our department from 1994 to 2005. Symptoms included headache, visual loss, and rhinorrhea. All patients underwent computed tomography (CT) and magnetic resonance imaging (MRI). Surgery was performed using an endonasal endoscopic procedure, in one case a navigational system was used. In light of these observations and a review of literature, the etiopathogenic characteristics, clinical aspects, and therapeutic indications of this localization are discussed.

Introduction

Mucoceles are benign, encapsulated, and expansile masses resulting from the accumulation of products of secretion, desquamation, and inflammation within a paranasal sinus. These lesions are frequent in the frontal and ethmoidal sinuses; the sphenoid sinus is the most rarely affected paranasal sinus. The aim of this article is to report our experience with four cases of sphenoidal mucocele and to discuss the etiopathogenic characteristics, clinical aspects, and therapeutic indications of this localization in light of these observations and a review of literature.

Observations

Case 1

A 20-year-old woman was admitted in January 2005 to the Neurosurgery Department of our hospital for 3 self-resolving episodes of right-eye visual loss that occurred when she was bending forward. Computed tomography (CT) and magnetic resonance imaging (MRI) showed an expansile mass situated in the right sphenoid and ethmoidal sinus and eroding the superior wall of the sphenoidal sinus, with an extension to the orbital apex and the anterior clinoid. A thickening of the bone at the sphenoidal septum and planum sphenoidale related to an associated fibrous dysplasia was also observed (Figures 1,2,3). The patient was then referred to the Ear, Nose, and Throat (ENT) Department.

An evacuation and marsupialization of the mass was carried out by an endoscopic endonasal procedure; the diagnosis of mucocele was then confirmed. The postoperative period was uneventful, and visual acuity returned to normal.

Case 2

A 46-year-old woman with a history of chronic migraine presented in 1994 with postnasal drip. CT and MRI showed a T2-weighted hyperintense expansile mass that was well defined, with only peripheral contrast enhancement, in the sinusal cavities of the sphenoidal and posterior left ethmoidal sinuses.

She underwent surgery by an endonasal endoscopic procedure. During the intervention, a mucocele of the sphenoidal sinus was marsupialized, and polyps of the ethmoidal sinus were discovered and removed. Histological study diagnosed an inverted papilloma of the glandular type.

The papilloma was removed using an external approach (lateral rhinotomy).

The patient underwent a surgical treatment for mucocele of the ethmoidal sinus in 1997, but histological study revealed a recurrence of the inverted papilloma, which was treated later by an external approach (lateral rhinotomy).

No recurrence was observed with an 8 years follow-up.

Case 3

A 37-year-old woman was a victim of a serious road accident in

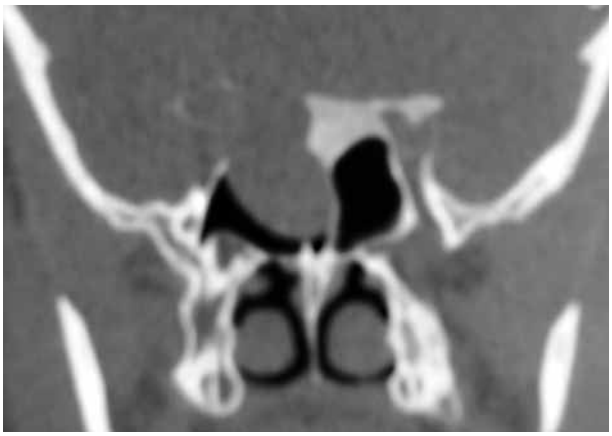

Figure 1
CT scan coronal view showing the mucocele, with erosion of the superior sphenoidal wall, extension to the orbital apex and the anterior clinoid, and the fibrous dysplasia.

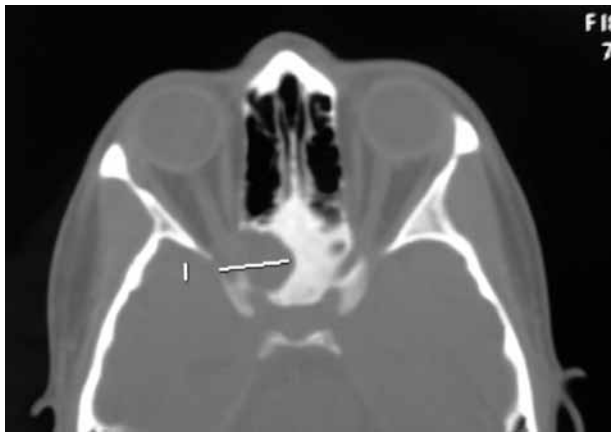

Figure 2
CT scan axial view showing the mucocele with extension to the orbital apex.

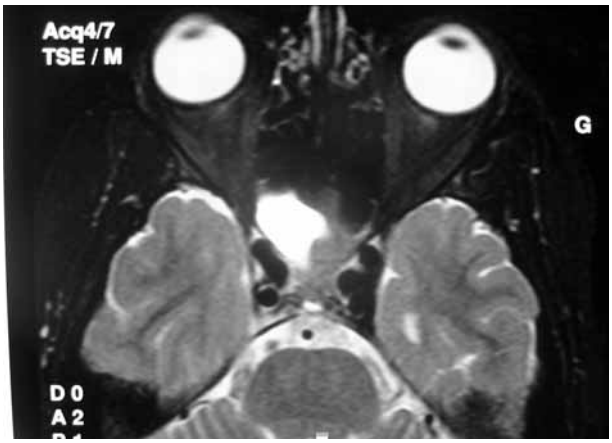

Figure 3
MRI axial image of the mucocele showing compression of the optical nerve.

1984, with skull base trauma, blindness of the left eye, hemianopia of the right eye, ptosis, and ethmoido-sphenoidal fracture.

She presented in 1997 with meningitis and cerebrospinal rhinorrhea after a minimal cranial trauma. She underwent surgery in the neurosurgery department.

The patient presented in 2005 with a recurrence of the cerebrospinal rhinorrhea and chronic headache. CT and MRI diagnosed a mucocele of the sphenoidal sinus and a meningocele in the posterior left ethmoidal sinus. An endoscopic, endonasal procedure with marsupialization of the mucocele was performed. The meningocele was not opened, but repulsed and isolated from paranasal structures by a fragment of abdominal fat. No complications were observed in the postoperative period.

Case 4

A 69-year-old man presented in 1995 with characteristic clinical symptoms of acromegaly. CT scan revealed a pituitary adenoma; he underwent successful surgery through a transnasal, trans-sphenoidal approach. No postoperative complications were observed, and plasma GH levels returned to normal.

In 2002 the patient presented with chronic headache and rhinorrhea. CT and MRI showed a mucocele of the sphenoidal sinus. Drainage was performed by an endoscopic approach using a navigation system. The postoperative period was uneventful, and no complications were observed with a 3 years follow-up.

Discussion

Paranasal sinus mucoceles are cystic lesions containing thickened mucus due to obstruction of the sinus ostium caused by an obstacle to drainage of any of the sinuses, leading to retention of their secretions. Obstruction can be due to congenital abnormalities, allergy, infection, trauma (our third case being an example), neoplasm, or surgical intervention.[1] According to Kessler *et al.*,[2] a mucocele of the sphenoidal sinus can develop 15–25 years after the primary surgery for sinusitis. In our fourth case, the mucocele developed only 7 years after surgery.

Only eight cases of a sphenoid mucocele occurring after transsphenoidal hypophysectomy have been previously reported in the world literature[2-4]; our fourth case is the ninth. Postoperative changes, such as narrowing due to cicatrization, adhesion, and bone hyperplasia, are strongly related to mucocele formation.[2]

In a review of the literature from 1965 to 2003, Kosling *et al*.[1] found nine case reports of a tumour occluding the sphenoidal ostium, leading to development of a mucocele. These tumours and tumour-like lesions were carcinoma (two cases), fibrous dysplasia (two cases), osteoma (two cases), ossifying fibroma (one case), fibromyxoma (one case), and Paget's disease (one case). Our first case is the third reported case of an association of a sphenoidal mucocele with a fibrous dysplasia. Fibrous dysplasia is a tumour -like disease of young patients (under 30 years of age). Cranial localizations are frequent, and the skull base is involved in 50% of cases, mainly in the sphenoidal region. Radiology, especially CT, is sufficient for diagnosis.[5] In the condensing variant (the most frequent), the bone is deformed, expanded, and thickened. Anterior clinoid localizations of mucocele are exceptional: seven cases have been previously reported in the literature.[6] Our second case is apparently the first case of an association of inverted papilloma with sphenoidal mucocele.

Other etiological hypotheses have been proposed, such as dilatation of glandular cystic structures, development from embryonal epithelial residues, and even an atypical form of craniopharyngioma.[7]

Most cases of paranasal sinus mucocele are found in adults aged from 30 to 60. They are localised in the frontal sinus (65%), anterior ethmoid (30%), and maxillary sinus (3-10%); posterior ethmoidal and especially sphenoidal localizations are very rare (1%).[7] However, it is a well-recognized entity in sinus pathology that is believed to represent 15 to 29 per cent of all cases of isolated sphenoid disease.[8] In contrast to mucoceles of other paranasal sinuses, descriptions of mucoceles of the sphenoidal sinus have been reported more frequently as case reports.[1] Since the first report of a case of mucocele of the sphenoidal sinus by Berg in 1889, nearly 140 symptomatic cases have been documented, with 53% in males and 47% in females, and an age range of 8 to 83 years – 48% of all cases were 30 to 60 years old. The interval between the first symptoms and diagnosis varied from 3 days to 38 years, with an average of 3.7 years. The aeration of paranasal sinuses begins between 12 and 24 months of age; mucoceles of the sphenoidal sinus and its orbital manifestations almost never occur before 3 years of age.[7] Mucoceles develop progressively and result in resorption and even erosion of the bony walls of the sinus. The sphenoid sinus is localised near vital anatomical structures vulnerable to compression: the dura mater; pituitary gland; optic nerve and chiasm; cavernous sinus; internal carotid artery; cranial nerves III, IV, V1, V2, and VI; sphenopalatine ganglion; sphenopalatine artery; and pterygoid canal.

Symptoms are often nonspecific, resulting in diagnostic delay. In 87% of cases, the principal symptom was pain (often unilateral and fronto-orbital headache). Other presenting signs were ophthalmic symptoms (85%), amaurosis (58%), oculomotor palsies (55%), ENT symptoms (anosmia, nasal obstruction, hypoacusis, and nasal discharge in, 38%), endocrine disorders (3%), and panhypopituitarism (0.8%).[7] The IIIrd cranial nerve is the most commonly affected (31%), while the IVth cranial nerve is rarely involved (5%).[9] The presence of cranial nerve deficits accelerates the diagnosis.

Mucoceles may have variable densities on CT and variable signal intensities on MRI, according to their water and protein content, density, and possible infection, and do not show contrast enhancement.[1,6]

On imaging, the differentiation from simple fluid retention that is found more often than mucocele relies on the expanding character of the mucocele, because the encapsulation is only detectable in cases with rim enhancement.[10] Toriumi reports that MRI imaging may be less reliable than CT scan because inspissated mucus in a mucocele can be hypointense in both T1- and T2-weighted images and by then can be mistaken for an aerated cavity.[11] However, such signal intensity from this kind of lesion is very rare and is different from that of an aerated cavity with a-signal area. CT scan may be superior to MRI imaging in order to study the structure of bony walls. Also, CT scanning can be performed in an emergency situation. An experienced otolaryngologist can easily perform a diagnosis relying on a CT scan, but supportive MRI imaging allows the differentiation of mucocele from

some other lesions, such as tumours. If the lesion stretches to the nasal cavities, an endoscopic examination can be a good aid to diagnosis.[9]

Misdiagnosis of sphenoidal mucocele as chordoma has been reported.[1] Other differential diagnoses of such lesions include necrotic primary adenoma in cases with significant intrasellar extension, craniopharyngioma, chordoma, plasmacytoma, osteoma, osteoblastoma, basal cell and squamous cell carcinoma, rhinolitis, polyps, and fibrous dysplasia.[7] The contents of mucoceles are usually sterile, although there may be a bad odour, suggesting pus. The contents may also be infected (mucopyoceles): in those cases, the most frequently isolated germs in mucoceles are Gram-positive organisms (*Staphylococcus aureus*, *Staphylococcus epidermidis*, and streptococci) followed by anaerobic bacteria species, gram-negative bacteria (*Hemophilus influenza*, *Escherichia coli*, *Pseudomonas aeruginosa*, *Proteus*), and fungi (aspergillus, chlamydia).[7] In such cases, antibacterial therapy should be based on the organisms most likely to be encountered. In case of failure to isolate the bacteria, the spectrum of antimicrobial drugs should cover as many causative microorganisms as possible, based on the predisposing factors.

Surgical treatment is absolutely indicated in mucoceles. In cases of acute visual loss it must be performed as soon as possible to make recovery more likely.[7,12]

The purpose of surgical treatment of the sphenoid sinus mucocele is to create a large ostium that will allow drainage into the sphenoethmoidal recess. An endonasal sphenoid sinus marsupialization that attempts to totally exteriorize the sphenoid sinus cavity with sufficient removal of the anterior and inferior wall of the sphenoid sinus and drainage of the mucocele is believed to be the therapy of choice.[1,13] Intranasal marsupialization of mucocele was reported as early as 1921 by Horwath.[13] However, this procedure may be insufficient in tumour-associated cases (our second case being an example), thus imposing an external approach.[10]

Recurrence was not observed during follow-up periods of 10 years in any of the 47 patients in Moryiani's study[12] and 4 years in any of the 12 patients in Har's study.[14] It was prevented in 95.8% of cases by an exclusively endoscopic treatment in ethmoidofrontal, maxillary, sphenoidal, and ethmoidal sites, in the Venail study.[15] However, because the mucocele develops 15-25 years after the initial surgery, a long-term postoperative follow-up is necessary in order to determine the real risk of recurrence.[16]

Conclusion

Mucoceles involving the sphenoidal sinus are rare lesions. They are quite diverse in their clinical and radiological presentation. Special care should be taken with regard to differential diagnoses and tumour-associated cases. In case of visual loss, the immediate surgical drainage of mucocele of the sphenoidal sinus with decompression of the orbit and optic nerve is necessary and usually results in the resolution of ophthalmic symptoms and recovery of visual acuity. The endonasal procedure is simple, effective, and safe.

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