

Bilateral Sphenoid Mucocele Presenting with Decreased Visual Acuity

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ABSTRACT

Introduction: Naso-sinus mucoceles are benign, inflammatory pseudocystic lesions that most commonly occur in the anterior fronto-ethmoidal region. Sphenoid involvement is rare, and bilateral cases are exceptional. We report a case of bilateral sphenoid mucocele that presented with decreased visual acuity.

Observation: The patient was a 45-year-old woman with a history of chronic headaches persisting for over six years treated with standard analgesics who presented with progressive distance vision loss that had been evolving for nearly six years, unaccompanied by ocular pain, diplopia, or other neuro-ophthalmological symptoms. Ophthalmological examination of the right eye revealed a corrected visual acuity of no light perception, exotropia, dry eye, and an relative afferent pupillary defect (RAPD); fundus examination showed optic disc pallor and temporal peripapillary atrophy. In the left eye, corrected distance visual acuity was 10/10, and dry eye, a normal pupillary light reflex, and a normal fundus were noted. The otolaryngological examination revealed congestion of the oropharynx with mild enlargement of the palatine tonsils. The OCT showed, in the right eye, very severe thinning of the RNFL and GCL with preservation of the inferonasal sector, and in the left eye, thickening of the RNFL in the inferonasal sector with a sectoral defect in the GCL in the superior sector. The MRI showed bilateral ballooning of the sphenoid cells with mixed fluid and blood content in some areas; the fluid component appeared as a T1-hyposignal and T2-hypersignal, while the blood component appeared as a T1- and T2-hypersignal. Color vision and visual field examinations had not been performed. The patient underwent endoscopic endonasal surgery.

Discussion: Sphenoid mucocele is rare. Functional Endoscopic Sinus Surgery (FESS) is currently the treatment of choice for sphenoid sinus mucocele.

Conclusion: This is a benign condition that progresses slowly and responds well to treatment. Endoscopically guided surgery is the standard of care.

Keywords: Mucocele, Sphenoid, Decreased Visual Acuity.

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INTRODUCTION

Naso-sinus mucocoeles are benign, inflammatory pseudocysts that are space-occupying and progress slowly. A mucocoele develops in response to the combination of two phenomena: ostial obstruction and chronic inflammation. It is a rare condition, the incidence of which has risen sharply since the 1990s, alongside the expansion of endoscopic endonasal surgery. They are most commonly found in the anterior fronto-ethmoidal region [1]. Sphenoid-shaped mucocoeles account for only 1 to 2% of all paranasal sinus mucocoeles, and bilateral involvement is rare [2]. The clinical manifestations of sphenoid mucocoeles are primarily related to their expansive extra-sinus growth; orbital extension leads to serious ophthalmological complications (such as optic neuropathy, oculomotor paralysis, and blindness) associated with tumor expansion. Diagnosis is often made using imaging studies and relies primarily on CT scans [3]. The endoscopic approach is the gold standard for the treatment of sphenoid sinus mucocoeles [4].

We report a rare case of bilateral sphenoid mucocoele diagnosed following a decline in visual acuity at the Dalal Jamm National Hospital in Dakar.

OBSERVATION

The patient was a 45-year-old woman presenting with progressive vision loss in the right eye evolving over more than six years accompanied by headaches, but without ocular pain or diplopia. The otolaryngological examination revealed a congested oropharynx with mild enlargement of the palatine tonsils.

The ophthalmological examination at admission revealed:

- in the right eye: a lack of light perception, exotropia, a relative afferent pupillary deficiency, and, on funduscopy examination, papillary pallor and temporal peripapillary atrophy;
- in the left eye: a normal photomotor reflex and, on funduscopy examination, a healthy macula and a healthy optic disc.

The retinography revealed papillary pallor and temporal peripapillary atrophy in the right eye; the left eye appeared normal (Figure 1).

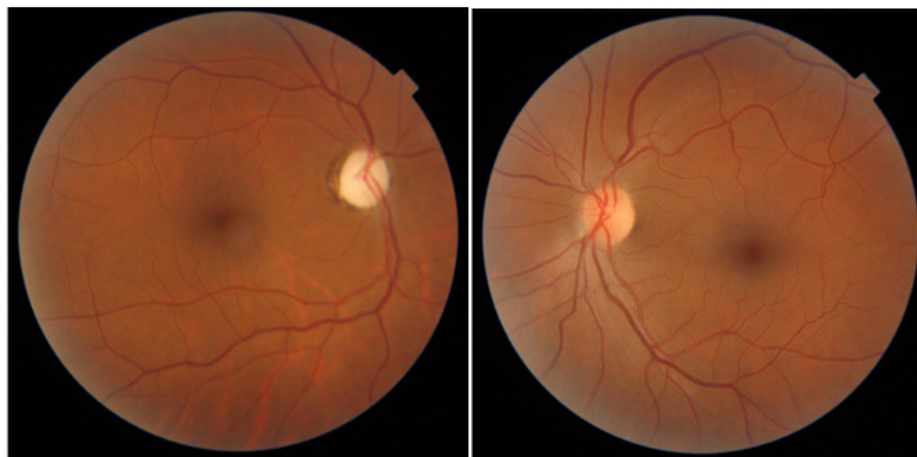


Figure 1: Right and left retinography

The OCT of the nerve fibers and ganglion cells showed:

- in the right eye : severe thinning of the RNFL and GCL, with preservation of the inferonasal sector (Figure 2);

- in the left eye : thickening of the RNFL in the inferonasal region with a sectoral defect in the GCL in the superior region (Figure 3).

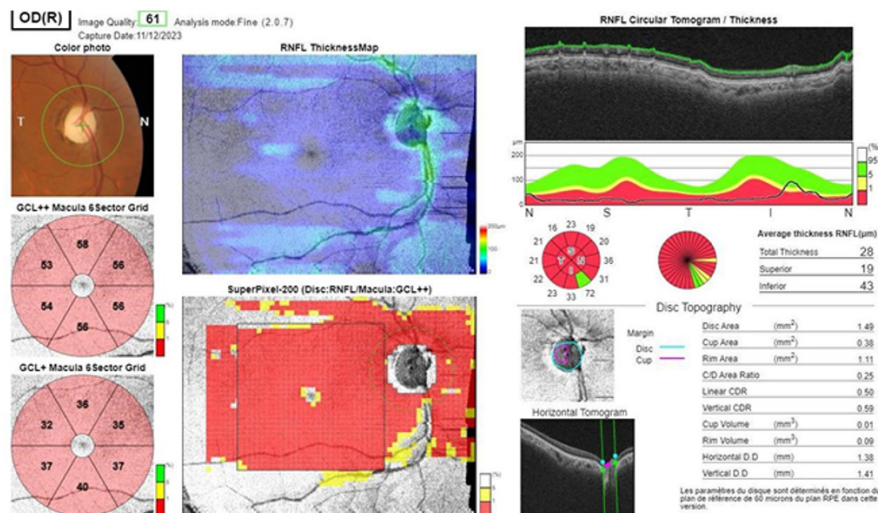


Figure 2: OCT of the optic nerve in the right eye (RNFL and GCL)

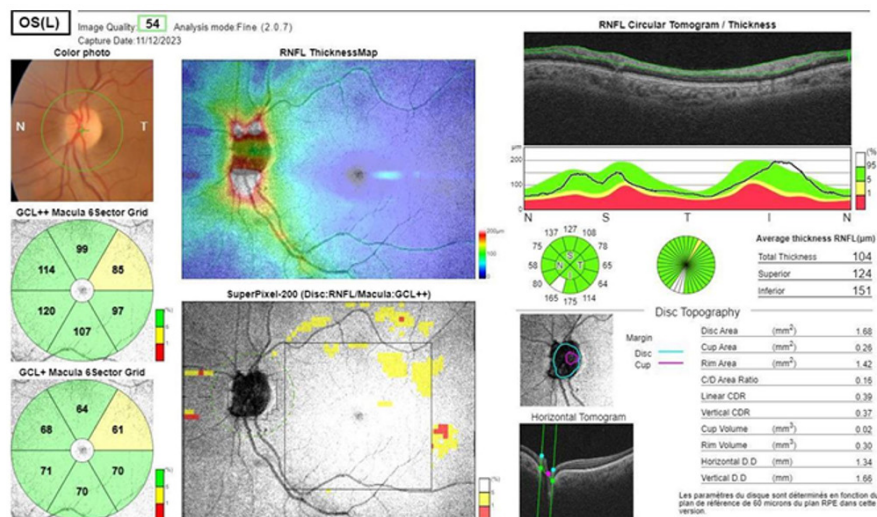


Figure 3: OCT of the optic nerve in the left eye (RNFL and GCL)

The radial OCT showed:

-in the left eye: a normal appearance (Figure 5).

-in the right eye: retinal thinning with disappearance of the interpapillomacular bundle (Figure 4) ;

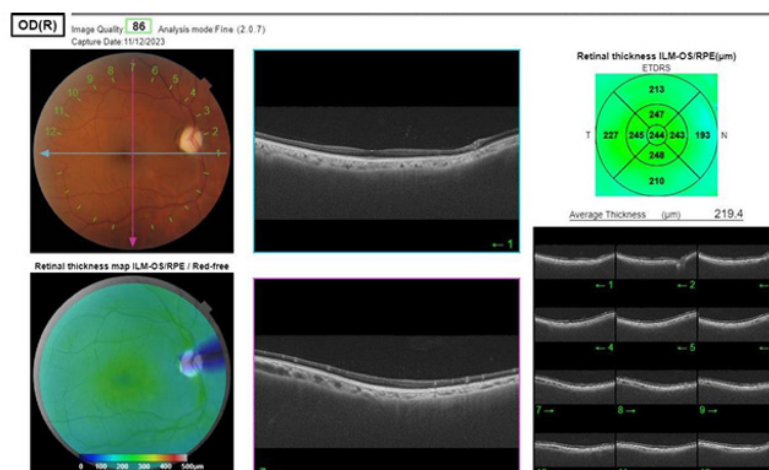


Figure 4: OCT in the right eye

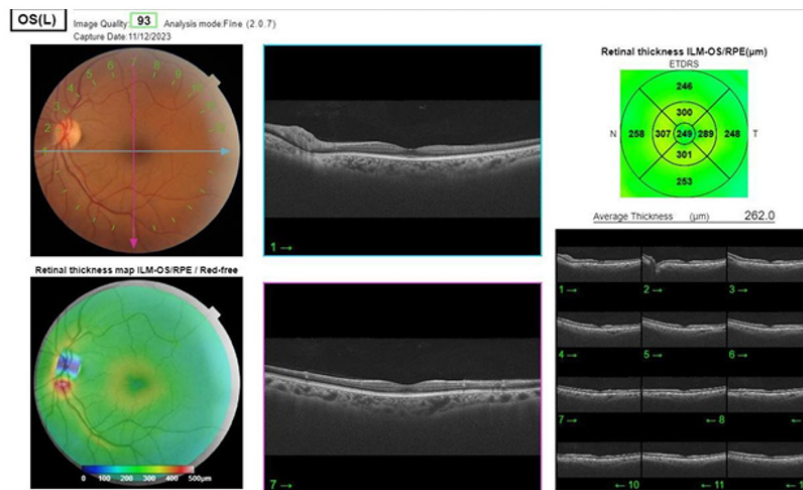


Figure 5: OCT in the left eye

The MRI showed bilateral ballooning of the sphenoid cells with mixed contents fluid and blood in places with the fluid appearing as a T1 hyposignal and T2 hypersignal, and

the blood appearing as a T1 and T2 hypersignal. This was responsible for moderate stenosis of the right orbital fissure with optic nerve atrophy (Figure 6).

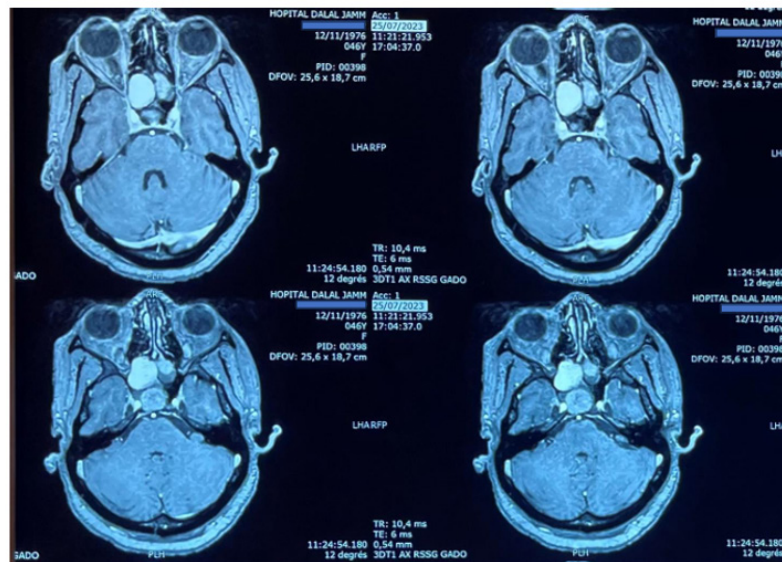


Figure 6: Orbital-cerebral MRI showing a sphenoid sinus mucocoele

The clinical presentation was suggestive of optic nerve compression. The patient underwent treatment for a mucocoele in the otolaryngological department. The contents

of the mucocoele were removed via an endoscopic endonasal approach, which involved marsupialization of the mucocoele sac within the nasal cavity (Figure 7).

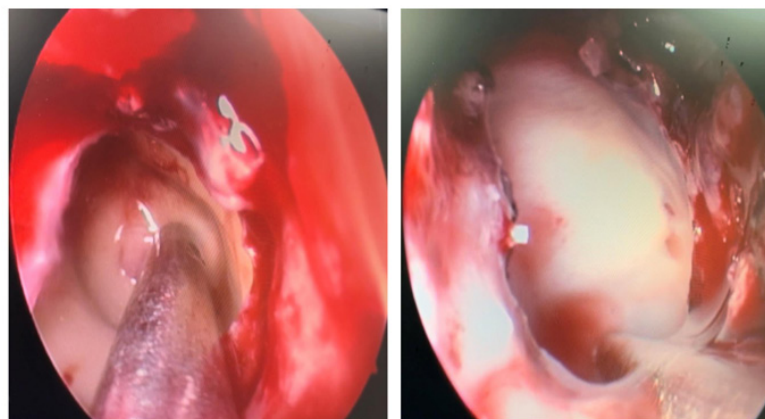


Figure 7: Intraoperative and endoscopic view showing extensive marsupialization of the mucocoele of the sphenoid sinus on the right and left

DISCUSSION

Epidemiology

This is a rare condition, although its incidence appears to have increased significantly in recent years, due to advances in imaging or in tandem with the expansion of endoscopic endonasal surgery [5]. Mucocoeles most commonly occur in adults. The pediatric form is rare and is exclusively located in the ethmoid sinuses. It is often associated with cystic fibrosis, a congenital malformation, or nasosinus polyposis [6]. They affect both sexes equally, with a higher incidence during the third and fourth decades of life [7]. Several factors can cause mucocoeles (sinus surgery, trauma, infection, etc.). The location is often related to the type of initial surgery [1].

Clinical Aspects

The clinical presentation varies, and the mimicking symptoms can lead to diagnostic uncertainty [8]. Ophthalmological signs (decreased visual acuity, exophthalmos, diplopia, etc.) are often the most prominent. The frequency of these signs is related, on the one hand, to the prevalence of mucocoeles in the fronto-ethmoidal complex and, on the other hand, to the fragility of the papyraceous lamina and the orbital roof, which are gradually displaced and lysis by the mucocoele [9]. The ophthalmological examination should look for facial scarring indicative of past accidental or surgical trauma, exophthalmos, or facial swelling, which is a sign of mucocoele protrusion. The decreased visual acuity may be caused by compression of the central retinal vein and the arteries supplying the optic nerve, and sometimes by chiasmatic arachnoiditis extending to the optic nerve [10]. A CT scan may show a hypo- or isodense mass with sharp borders and a cystic appearance of the bony structures. There is no enhancement following the injection of contrast medium [3, 11]. The MRI provides a better understanding of the relationship between the sinuses and the adjacent vital organs [11]. An endoscopically guided sphenoid biopsy is indicated in cases of suspicious swelling [12].

Topography

The most common locations are the fronto-ethmoid and frontal sinuses, followed by the maxillary sinus and, less commonly, the posterior ethmoid and sphenoid sinuses [13]. Sphenoid cysts are rare and account for 1 to 2% of all sinus mucocoeles [2]. The proximity to the meninges and orbits contributes to the severity of this sinus location [14].

Treatment

Treatment involves external and/or endonasal surgery. The choice of technique will depend on the location and extent of the mucocoele. The endonasal endoscopic approach is a safe and effective treatment that ensures adequate drainage with a low recurrence rate [15]. The FESS is currently the treatment of choice for nasosinus mucocoeles [4]. For our patient, the surgical approach was endoscopic endonasal, decided upon in consultation with the otolaryngologist surgeon. A partial middle turbinectomy was performed on each side to provide access to the sphenoid sinus ostium, which was enlarged using Kerrison rongeurs. The mucocoele sac was then ruptured, evacuated, and irrigated. The posterior wall of the sinus was expanded on the left side. Irrigation of the left cavity was performed to check for a communication between the two mucocoele sacs. However, there was no fluid leakage from the right wall. The immediate postoperative period was marked by the onset of vertigo. The patient was prescribed an acetylucine-based antivertigo medication. A lack of light perception in the right eye persisted even after the procedure.

CONCLUSION

Bilateral sphenoid mucocoeles are extremely rare but potentially dangerous. Clinically latent for a long period, their clinical manifestations are associated with expansive extrasinus extension and appear only after the mucocoele has already formed. Orbital extension is responsible for severe and sometimes irreversible ophthalmological complications, such as optic neuropathy. Due to their close anatomical relationship with the orbital apex and neuro-meningeal structures, these sphenoid mucocoeles require prompt surgical management [2].

STATEMENTS

Patient's Consent

We obtained the patient's informed consent.

Conflict of Interests

The authors declare that they have no competing interests.

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