

A rare cause of bilateral exophthalmos: about one case with a sphenoid mucocele*

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Abstract

Background: Mucocele is a benign cystic pseudo-tumor that develops within the sinus cavities. The most frequent locations are frontal and frontoethmoidal. The sphenoidal forms are rare. We report a case of sphenoidal mucocele revealed by bilateral exophthalmos.

Methods and results:

We report the case of a 14 year old male patient, without any particular pathological history, referred by his ophthalmologist for bilateral nasal obstruction, bilateral exophthalmos and visual acuity decrease evolving since one year. The ENT examination found a large, renitrant mass, filling the entire right nasal cavity. This mass pushed the nasal septum to the left. The CT scan showed a large sphenoidal, hypodense mass evoking a mucocele. The patient underwent endonasal marsupialization by endoscopic guidance.

Conclusions: Sphenoidal mucocele is a rare cause of bilateral exophthalmos. The diagnosis can be misplaced when the signs are overt extra-sinus. Hence the interest of CT in order to eliminate other differential dignostics.

Key words: sphenoidal mucocele, sphenoid sinus, sinus CT, sphenoidotomy, marsupialization

Introduction

Mucocele is a benign, expansive, slowly growing pseudo-cystic formation that develops within the sinus cavities. The most frequent locations are frontal and frontoethmoidal ⁽¹⁾. The sphenoidal forms are rare. The proximity of the orbit and the endocranion make this sphenoidal location so serious. Symptoms are often borrowed and often delayed compared to the appearance of the mucocele. We report a case of sphenoidal mucocele revealed by bilateral exophthalmos.

Clinical case

This is a 14-year-old male patient with no pathological history, referred by his ophthalmologist for bilateral nasal obstruction, bilateral proptosis and reduced visual acuity that has progressed for a year.

The ENT examination found a large, renitent mass, filling the entire right nasal cavity. It pushes back the nasal septum to the left (Figure 1).

Ophthalmologic examination found bilateral exophthalmos with bilateral optic nerve atrophy.

The CT scan showed a large sphenoidal mass, hypodense thinning its walls with mass effect on the orbital cavities, leading to bilateral exophthalmos (Figure 2 and 3).

The treatment had consisted of marsupialization of the mucocele pocket. The approach was endonasal with optical guidance. When the mucocelic pouch was opened, puriform fluid was drained. After marsupialization, no beating possibly related to the carotid artery was observed in the marsupialization cavity. A wicking of the right nasal cavity was performed at the end of the operation and maintained for 2 days. The operative consequen-



Figure 1. Mass filling the right nasal cavity.

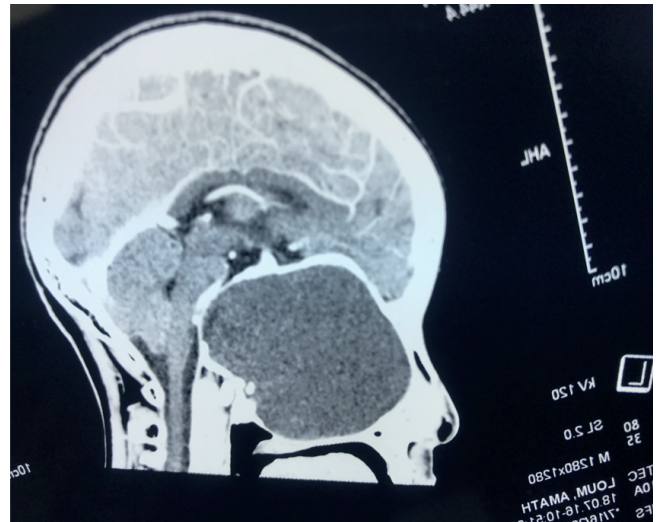


Figure 2. Sphenoid mass extending to the nasal cavity.

ces were simple.

From an ophthalmological point of view, the course was marked by a regression of exophthalmos. The direct photomotor reflex was present and eye tone was normal. However, we found, on the right side, total optic nerve atrophy and on the left side a cup / disc ratio of 9/10 with loss of the foveolar reflex. The follow-up CT scan taken 4 months later showed no recurrence (Figure 4).

Discussion

The diagnosis of sphenoid sinus mucocele can be misplaced when there are overt extrinsic signs. The exophthalmos and the decline in visual acuity, revealing in our case, lead directly to an ophthalmological pathology. It was CT that allowed the ophthalmologist to focus on the diagnosis and to be able to refer the patient to the ENT doctor. In ENT practice, sphenoid mucoceles are very rare, representing 1 to 2%; the most frequent locations being the frontal and ethmoido-frontal sinuses ⁽²⁾. In a series of 58 cases of mucocele, Martel-Martín reported 55% ethmoido-frontal localization, 14% pure ethmoidal localization, 24% maxillary localization and 7% sphenoidal localization ⁽¹⁾.

Although rare, sphenoidal mucocele is a potentially complicated condition. The seriousness of the sphenoidal involvement lies in its close relationship with the cerebral-meningeal and orbital structures. It is a slowly evolving and expansive pathology often diagnosed at the stages of complications as in our patient: exophthalmos and optic nerve atrophy. Thus, the diagnosis must be made as soon as possible in order to prevent serious ocular complications (optic nerve compression, loss of visual acuity, blindness, mucopyocele) and neuro-meningeal ^(3,4). The etiology of sphenoid sinus mucocele remains unknown, but

three pathogenic mechanisms have been formulated: submucosal edema, obstruction of the secretory duct, and obstruction of the sphenoid sinus ostium. Several contributing factors can be cited: allergic rhinitis, chronic sinusitis, polyposis, etc. ⁽⁵⁾. Another cause to keep in mind is the radiation therapy performed for the treatment of head and neck cancers, especially nasopharyngeal cancer ^(6,7). The cause of sphenoidal mucocele can also be an endonasal surgery that can cause ostial blockage ⁽⁸⁾. In our patient, the cause remains unknown.

The diagnosis requires a tomodensitometric exploration which has a threefold interest: to make the positive diagnosis and extension, to look for anatomical varieties at risk of surgery and to look for complications. The mucocele looks like an expansive, homogeneous formation of regular contours, blowing and sometimes lysing the bone walls. Its density depends on the degree of hydration; spontaneously hypo dense or iso dense, the old forms can appear hyperdense ⁽⁹⁾. MRI is not essential for the diagnosis of mucocele if the CT scan is clear. It is useful for looking for signs of suffering from surrounding elements, especially the eye, meninges, pituitary and optic nerve. The signal from the mucocele is variable depending on the viscosity and the protein content of the intramucocelic retention. After injection of contrast product, there is no modification of the lesion signal with peripheral enhancement ⁽¹⁰⁾.

In addition, CT and MRI are important contributions in the search for differential diagnoses. The sphenoidal mucocele, however, can be confusing with nasosinus and skull base tumors.

Meningioma is a slow-growing benign tumor that is rarer in children. MRI may show a dural tail. The sphenoidal location re-

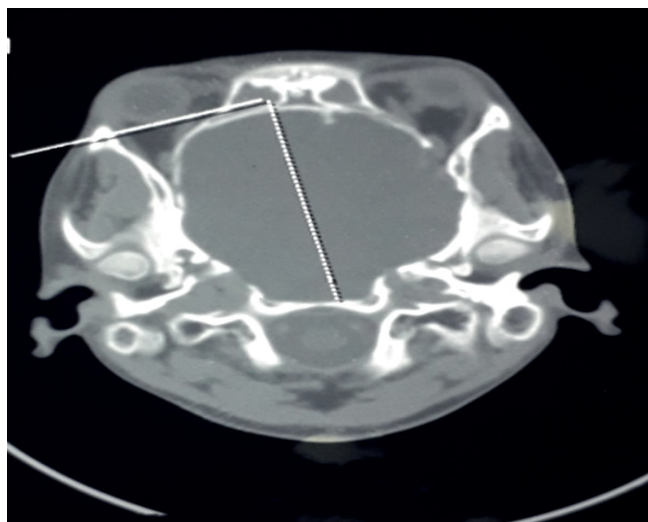


Figure 3. Sphenoid mass pushing back the eyes.

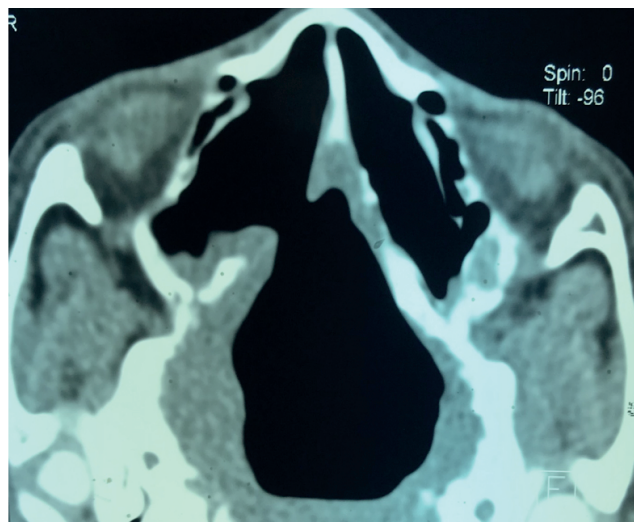


Figure 4. Postoperative control CT.

presents 15 to 20% of the intracranial locations ⁽¹¹⁾. Other tumors such as hypothalamic and chiasmatic astrocytomas, fibrous dysplasia, are also differential diagnoses that should not be overlooked ⁽¹²⁾. Fungal sphenoid sinusitis can take on a tumor-like appearance that can also mimic a mucocele ⁽¹³⁾.

Ophthalmologic recovery was not complete in our patient due to the long duration of optic nerve compression. Indeed, the visual prognosis depends on the degree and duration of the optical compression but also on the speed of the surgical management ⁽¹⁴⁾.

Conclusions

The sphenoid mucocele is a pseudo-cystic tumor with the potential to be aggressive towards the orbital and endocranial structures. Its borrowing symptomatology is a source of diagnostic error. Performing a CT scan is essential for a positive diagnosis. Now, the differential diagnosis of bilateral exophthalmos should include the sphenoid mucocele.

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Authorship contribution

NP and MN wrote the manuscript and all authors discussed the results and contributed to the final manuscript

Conflict of interest

None declared.

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Consent for publication

Informed consent for publication of the clinical details and images was obtained from the patient.

Availability of data and materials

Not applicable.

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