CASE REPORT

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Vision impairment as presenting symptom of a sphenoidal mucocele*

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SUMMARY

We present a case of a 27-year-old woman with a sphenoid mucocele, presenting with bilateral visual disturbance and pituitary gland dysfunction but without nasal or sinus complaints. We would like to emphasize the importance of early diagnosis and prompt surgical treatment.

Key words : mucocele, sphenoid sinus, endoscopic surgery, ophthalmology, hypopituitarism

INTRODUCTION

Mucoceles are cystic benign formations containing mucus, covered by a respiratory mucosa and situated in the paranasal cavities. Capable of expansion, they gradually accumulate mucous material leading to a progressive distension of the sinus bony walls. In the particular context of the sphenoid sinus, the direct compression of the optic nerve associated with a surrounding inflammatory reaction may cause severe visual impairment. Because the primary symptoms of ethmoidal or sphenoidal mucoceles may be ophthalmological such as reduced ocular mobility, exophthalmus, diplopia and loss of visual acuity, patients may first visit ophthalmologists (Casteels et al., 1992; Moriyama et al., 1992; Hao, 1994; Muneer et al., 1997). Complications of sinus diseases such as acute or chronic sinusitis are also known to induce vision disturbances (Slavin et al., 1987; Patt et al., 1991; Maniglia et al., 1995; Pitkaranta et al., 2000; Muller et al., 2001).

Unusual presentations of sphenoidal sinuspathology include hypopituitarism (Valvassori et al., 1973; Benton et al., 1975; Rosselet et al., 1984).

We report a case of massive sphenoid mucocele initially presented with isolated bilateral visual dysfunction. We stress the importance of both, ophthalmological and ENT diagnostic procedures, and point out the necessity of a prompt surgical treatment to avoid permanent visual loss.

CASE REPORT

A 27-year-old caucasian woman presented to the Department of Ophthalmology for a rapidly progressive bilateral loss of vision. There was no history of previous trauma, sinus surgery or chronic rhinopathy and she had no particular ENT complaints. Two months before the onset of symptoms, she had a spontaneous abortion and recent laboratory examinations revealed a thyroid dysfunction: TSH was 0.26 mcU/ml (normal range 0.3-4.67).

The ophthalmological examination of visual acuity revealed only finger counting at the right side and 2/10 at the left side. The fundus and intraocular pressure were normal and there was a normal, not painful ocular motility. The Goldmann test (perimetry) revealed a caecocentral scotoma on the right side. Colorimetry showed bilateral impaired colour vision and visual evoked potentials were suggestive for conductive disorders, for which she immediately received intravenous corticosteroids: Solumedrol® (methylprednisolone) 250 mg per day during 3 days. This treatment resulted only in a partial restoration of her vision on the left side. Complementary imaging and ENT examination were requested.

Magnetic Resonance showed an expansive mass in the sphenoidal region, suggestive of a mucocele. The finding of a high signal intensity area on T1-weighted MR images with, and

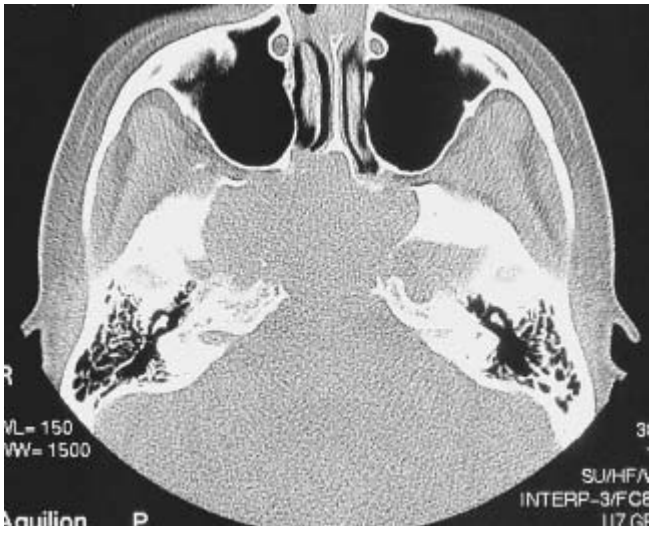

Figure 1. Axial CT scan at the level of the sphenoidal sinus demonstrates a large mucocele.

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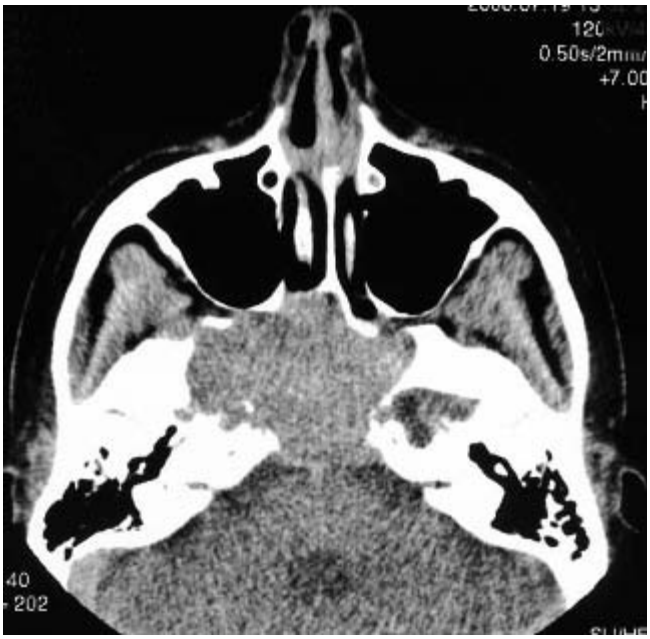

Figure 2. Axial CT scan shows absence of intersinus septum.

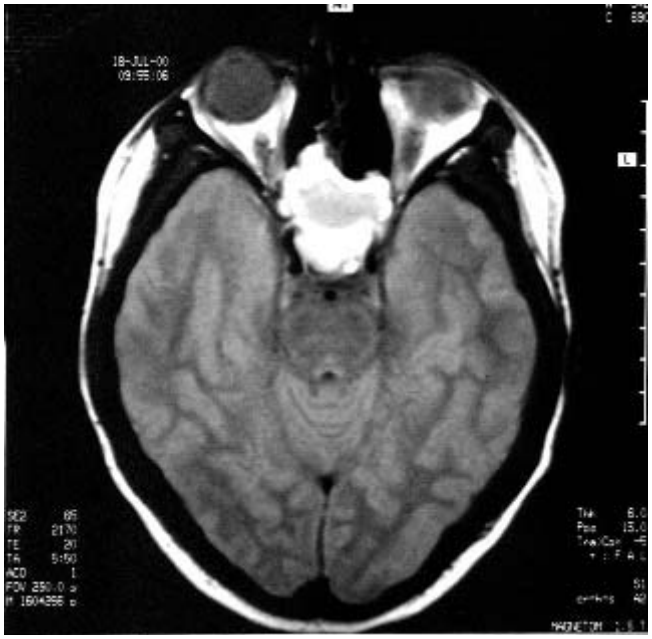

Figure 3. Axial MRI-scan of the head showing a lesion of the sphenoid sinus of mixed intensity.

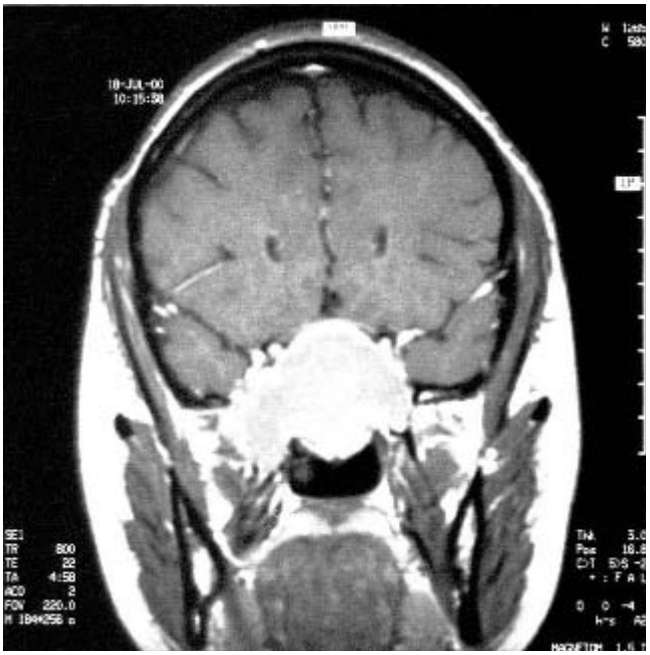

Figure 4. Coronal T1 weighted image with contrast showing expansion of the mucocele on the right side into the fossa pterygopalatina.

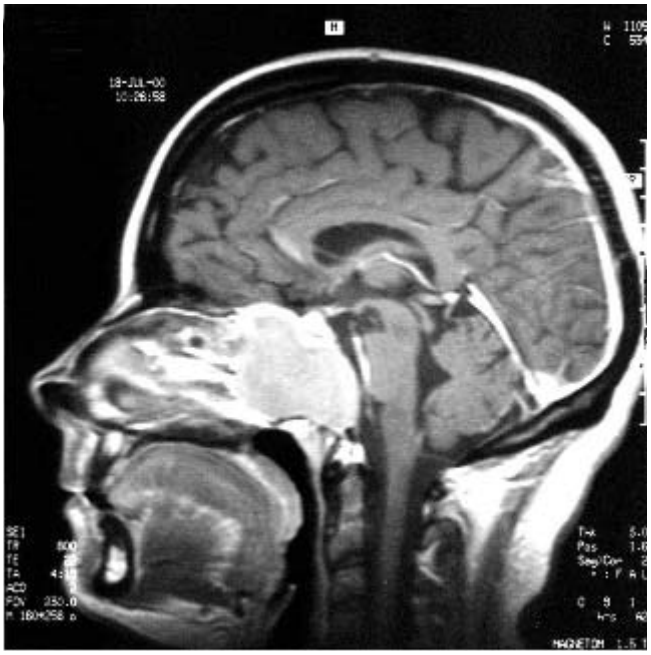

Figure 5. Sagittal image showing a lesion of the sphenoid with projection of the pituitary gland superiorly and backwards.

without, gadolinium was the criterium to differentiate from other expansile masses around the sphenoid sinus. Possible differential diagnoses were lymphangiomas and meningiomas, but they would lead to an enhancement of contrast with gadolinium. A CT-scan of the sinuses clearly displayed the shape and size of the lesion and the involvement of the sphenoidal sinuses, with no intersinus septum (Figures 1 and 2). The mucocele distended and thinned the bony walls like a balloon and compressed the adjacent neurovascular structures (Figures 3 and 4). It displaced both optic nerves laterally and projected the pituitary gland superiorly and backwards (Figure 5). On the right side the mucocele had expanded superolaterally into the fossa pterygopalatina. CT-scans showed a very thin surrounding bony rim displayed both in axial and coronal planes.

Preoperative nasal endoscopic examination revealed a bulging of the anterior sphenoidal sinus wall on the right side without signs of acute infection.

As soon as the patient was seen by the ENT surgeon it was decided to perform an emergency endoscopic sinus surgery under general anaesthesia. After partial conchectomy of the

middle turbinate, a sphenoidotomy and a resection of the sinus anterior wall were realized. With the large opening of the mucocele, large amounts of grayish white, viscous and turbid fluid were released and the presence of *Staphylococcus aureus* was demonstrated on microbiological culture. There was a partial defect of the bony lateral wall on the right side with a clearly exposed pulsating internal carotid artery and optic nerve.

Because of a lack of local bleeding, no postoperative nasal packing nor antibiotics were used. However we administered intravenous corticosteroids for another 3 days: Solucortef® (hydrocortisone) 200 mg/day for two days and 100 mg the last day.

Microscopically, the excised part of the capsule presented a chronic non-specific inflammatory tissue composed of stratified respiratory epithelium made up of ciliated columnar cells with areas of mononuclear cell infiltration and fibrous tissue.

The vision recovered rapidly and, on day four postoperatively, the visual acuity was 8/10 right and 10/10 left. There were no neurological disturbances. After three months there was a total recovery of the vision and the hormonal disorders of the patient disappeared; there was no longer a thyroid dysfunction. A couple of months after the operation the patient became pregnant and delivered a healthy boy. As demonstrated by nasal endoscopy, the sphenoid sinus keeps widely open. Although there is some decrease of the cavity volume with a further displacement of the tegmen and hypophyse anterocaudally, the internal carotid artery on the right side remains recognisable due to pulsation.

DISCUSSION

The anatomical proximity and the fragility of orbital structures explain the high occurrence of ocular complications in sinusopathy. The orbit is a preferential way of extension for inflammatory or neoplastic sinus diseases, and the optic nerve can frequently be affected. As the optic nerve enters the orbital apex from the intracranial portion, the nerve is encompassed by the narrow bony canal and is in close proximity to the posterior ethmoid and/or sphenoid sinuses. In most cases the bony wall between the nerve and the sinuses is as thin as 40-60 µm (Slavin et al., 1987; Kainz et al., 1992). Intracranially, the optic nerve is covered by the pia mater, arachnoid, and CSF. As the nerve enters the canal, it is surrounded by two layers of a nerve sheath, the outer of which is continuous with the periorbita and the inner of which ensheats the nerve to the eye ball. The optic nerve canal can be identified as a semicircular tube when viewed from the inside of the posterior ethmoid or sphenoid sinus. Since the optic nerve canal contains the nerve and two layers of nerve sheath without any soft tissues such as fat, pressure due to expansion of the mucocele may easily impact the nerve if the thin protecting bony wall is resorbed or if it is dehiscent congenitally.

Mucoceles are dilated mucous-filled sinuses that are lined by a mucous membrane, and result from a chronically obstructed sinus ostium with enlargement of the bony walls caused by mucous secretions filling the sinus cavity (Delfini et al., 1993). They are commonly caused by post-operative ostial obstruction (iatrogenic) or secondary to trauma, but also can present as primary disease. Paranasal sinus mucoceles predominantly occur in the fronto-ethmoidal region (64%), followed by the maxillary sinus (18.6%), the sphenoid sinus (8.4%) and the posterior ethmoid sinus (6.7%) (Rombaux et al., 2000). Primary sphenoid mucoceles can be found in association with chronic ethmoidal sinusitis and/or nasal polyposis and are uncommon in isolation (Daniilidis et al., 1992).

The natural development of sinus mucoceles consists of gradual expansion. Acting as a slow-growning benign neoplasm, this expansion can result in bone remodelling, bulging and erosion, and reaches adjacent structures such as the other sinuses, orbit, clivus, skull base or brain. The cause of bony destruction in large mucoceles has been debated. Altough pressure-induced osteolysis and devascularization of bone play an important role, osteoclastic bone resorption in which prostaglandin E2 from fibroblasts and interleukin-1 from epithelial cells with tumor necrosis-factor-α are thought to be the main responsible agents which could also account for the agressive nature of mucoceles (Lund et al., 1993). In case of a complication, infection may even lead to conditions such as meningitis, subdural or brain abscess (Nugent et al., 1970).

By extension into adjacent structures, the mucocele gives rise to a variety of clinical manifestations. Headache, facial pain, anosmia, ocular displacement, ocular palsy and visual failure are the most common symptoms reported. Visual symptoms in patients with sphenoid sinus mucoceles include diplopia, ocular muscle paresis, exophthalmus and complete visual loss (McCarthy et al., 1972; Casteels et al., 1992; Hao, 1994; Muneer et al., 1997). One should also be aware of unusual presentations including hypopituitarism (Valvassori et al., 1973; Rosselet et al., 1984; Pitkaranta et al., 2000).

Vision impairments were noted in six out of fifteen patients by Benninger et al. (1995). Five of the latter were identified to have either limitation of gaze or progressive visual loss (Benninger et al., 1995). Visual symptoms commonly lead patients to seek an ophthalmological opinion. In a review of 47 patients suffering from ethmoid or sphenoid mucoceles, Moriyama et al. (1992) reported that 70 per cent of patients first sought help from the ophthalmology department.

The differential diagnosis of isolated bilateral vision impairment includes neuritis optica caused by a viral infection, toxic agents, multiple slerosis, or unknown reasons (idiopathic) and expansile masses. Yumoto et al. (1997) stated that optic neuropathy secondary to paranasal sinus disease occurs much less frequently than due to multiple sclerosis or idiopathic origin. Therefore glucocorticosteroids often are the first treatment of choice.

Visual dysfunctions due to a mucocele have been suggested to be caused by one of two possible mechanisms. The mucocele may compress the optic canal and cause visual disturbance, leading to loss of eyesight in severe cases. In addition to direct

pressure, ischemia or venous congestion around the optic nerve subsequently occurs as suggested by Moriyama et al. (1992). Besides the compression and its local effects, inflammation due to infection of the mucocele can spread to the nerve through zones of bony erosion. This hypothesis was supported by Fujitani et al. (1984) showing that the visual loss may be due to a local inflammatory response which responds to steroid therapy. But even with response to steroids, further diagnosis and immediate surgical drainage are imperative.

The preoperative diagnosis is usually based on CT and magnetic resonance imaging. On CT scan, mucoceles usually fill a sinus structure and bulge against adjacent anatomical structures, but without infiltration. The bony margins of the lesions are well defined. Differential diagnosis may be difficult, if CT shows isodense lesions without enhancement by contrast medium (Yokoyama et al., 1996). Also, the magnetic resonance image of mucoceles varies widely, mucoceles appear to vary from homogeneous to nonhomogeneous, being hyperintense or hypointense on T1-weighted images (Delfini et al., 1993). CT scans are preferable for definitive evaluation, assessment of bony involvement and presurgical planning. MRI scans are helpful in the evaluation of intracranial and orbital extension, and in ruling out a neoplasm or fungus disease (Kennedy et al., 1989). Therefore, CT and MR imaging are complementary for the diagnostic evaluation of an expansive mass around the posterior ethmoid and sphenoid sinus. As possible differential diagnosis one should consider a mucocele, pyocele, sinus malignancy, hypophyseal tumour, craniopharyngioma, meningioma or optic nerve glioma, intracranial chordoma and cholesteatoma as well as neoplastic lesions of the nasopharynx.

Surgical treatment of a sphenoidal mucocele is urgent in cases of vision loss. Because of the easy access and the minimal invasive surgery the endoscopic endonasal approach is the most convenient to treat paranasal sinus mucoceles. Endoscopic sinus surgery has been advocated for the treatment of mucoceles, with lower morbidity and a reduction in potential complications compared to intracranial approaches (Beasley et al., 1995). The mucocele is widely opened and a sufficiently part of the anterior wall is resected for drainage and aeration. With exposure of the dura or peri-orbital wall, the wall of the mucocele acts as a cover and no attempts are made to remove this mucosal lining. Attempts to remove the mucosa may cause injury to the underlying structures such as dura, optical nerve or carotid artery. One should expect a rapid improvement of visual acuity. However, in cases of a complete loss of vision, the prognosis may be much poorer. The degree of improvement of the visual acuity after mucocele marsupialisation depends on the severity of the initial loss before it, the mode of development and the location of the mucocele and the time from onset of the ophthalmological disorder until the surgical marsupialisation (Rombaux et al., 2000). Prompt surgical treatment is necessary in order to avoid permanent visual impairment and other sequelae (Maniglia et al., 1995; Muneer et al., 1997; Yomoto et al., 1997). Especially if vision is seriously impaired, immediate surgery should be performed, preferably within 24 hours after the onset of visual disturbance (Yumoto et al., 1997). Therefore, a good knowledge of this disease by ophthalmologists and otorhinolaryngologists is essential for early diagnosis and prompt surgical treatment.

CONCLUSIONS

Mucoceles as well as other sinus diseases may cause ophthalmological problems or in rare cases pituitary gland dysfunctions, without a specific history of rhinopathy. Their presenting symptoms can mimic central optical disorders and imaging (CT-scan and MRI) remains the best way to establish the diagnosis. Endoscopic sinus surgery is, in these cases, perfectly indicated and this case report clearly emphasizes the efficacy and the safety of this technique.

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