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Ophthalmoplegia and Ptosis as Onset Symptoms of an Isolated Primary Mucocele of the Sphenoid Sinus

Stefano Di Girolamo, MD, Pasquale Cannizzaro, MD,†*
Pasqualina Picciotti, MD,‡ and Cristiano Nardi, MD§

Primary and isolated mucoceles of the sphenoid sinus are very rare,[1-6] but probably the incidence of this disease is underestimated due to the lack of specific symptoms.[7] Symptoms are dependent on the involvement of neuro-ophthalmologic structures. Therefore many clinical signs can be present: retro-orbital pain, paresis of oculomotor muscles (with various levels of ophthalmoplegia and ptosis), sensorial deficit of first branch of fifth cranial nerve with facial and conjunctival hyperesthesia or hypoesthesia, amaurosis (if it involves the second cranial nerve), and exophtalmus (rare).[2]

This article reports the case of a patient who had left diplopia as the only symptoms that masked an incomplete ophthalmoplegia of his left eye as the presentation of a left sphenoid sinus mucocele. This case shows the minimal early symptoms of this sphenoidal mucocele and discuss the diagnostic procedure and the surgical approach.

Report of a Case

A 79-year-old white man was reffered from the neurology department with a 15-day history of ptosis of his left eyelid and diplopia of the left eye. He did not report fever or pain. Clinical examination revealed an isolated and incomplete left-sided oculomotor nerve palsy, and visual fields were normal. No other cranial nerve deficits or neurologic signs were present. A fiberoptic examination of the upper respiratory airways did not reveal any disease or anatomic abnormality. A computed tomography (CT) scan showed a soft tissue mass filling the left sphenoid sinus (Fig 1). Magnetic resonance imaging (MRI) of the skull showed the left sphenoid sinus filled with hypointense material with contrast enhancement (Fig 2). A diagnosis of isolated sphenoid sinus mucocele was made. The patient underwent antibiotic therapy with ceftriazone and steroid therapy for 15 days without any clinical improvement.

A transnasal sphenoidectomy was performed under microscopic guidance.

Histologic specimen showed a chronic mucosal inflammation. Eight days later, the patient was discharged in good general condition with complete resolution of oculomotor nerve palsy. At 2-year follow-up with nasal endoscopic examination and CT scan, the patient showed complete resolution of the disease (Fig 3).

Discussion

Mucocele is usually the consequence of recurrent sinusitis that leads to closure of the sinusal ostium and to the formation of a retention cyst.[8] The walls of a mucocele are made up with sinusal mucosa and contains a viscous liquid with crystals of cholesterol, red cells, neutrophilic granulocytes, and epithelial desquamated cells.[1]

Males and females are equally affected, and there is no age predominance.[9] In the present case, the onset symptoms were only ptosis of the left eyelid and diplopia of the left eye. As far as we know, a similar clinical onset has not been described. In fact, other symptoms generally could be associated with a primary and isolated mucocele of the sphenoid sinus: periorbital or retrorbital unilateral headache (78%) or frontal or temporal headache (33%), sensorial deficit of first branch of the fifth cranial nerve (21%) with facial and conjunctival hyperesthesia or hypoesthesia, exophthalmous (31%), visual defects (56%) generally unilateral, due to superior and anterior growth of a mucocele with compression of the second cranial nerve near orbitary apex, rhinorrhea (13%) and epistaxis (7.5%), dizziness (7%), and hearing loss (5%). If the mucocele grows posteriorly and laterally, it can compress the eighth cranial nerve. Also, anosmia (7.5%), pituitary deficit (5%), and endocranial complications (meningitis) can occur.[1,4,5,7,9] Ophthalmoplegia is present variously associated with the previous signs in 32% of cases due to complete or incomplete

Received from the Institute of Otorhinolaryngology, "Università Cattolica del Sacro Cuore," Rome, Italy.

*ENT Associate Professor.

†ENT Resident.

‡ENT Resident.

§ENT Resident.

Address correspondence and reprint requests to Dr Di Girolamo: Istituto di Clinica Otorinolaringoiatrica, Università Cattolica del Sacro Cuore, Largo Agostino Gemelli No. 8, 00168 Rome, Italy; e-mail: sdigirolamo@virgilio.it

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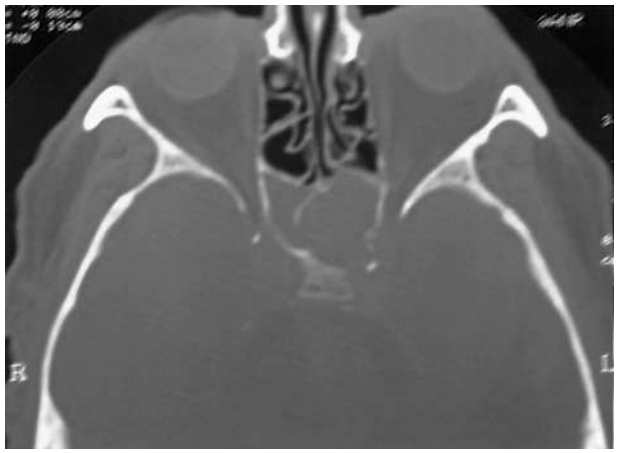

FIGURE 1. Preoperative axial CT scans showing the mucocele filling and expanding of the left sphenoid sinus.

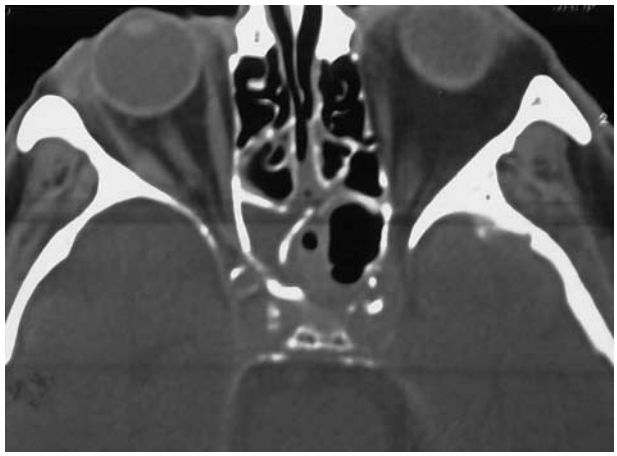

FIGURE 3. Postoperative axial CT scan showing complete removal of the mucocele from the sphenoidal cavity.

paralysis of third, fourth, and sixth cranial nerves with sudden dyplopia with drooping of the upper eyelid[1,5,9] (Table 1).

The onset of ophthalmoplegia is due to the mucocele's growth, after bone resorption of the anterolateral walls of the sphenoid sinus within the superior orbital fissure. Superior orbital fissure remains between the inferior surface of the lesser wing of the sphenoid and the medial margin of the greater wing and contains the third, fourth, and sixth cranial nerves, ophthalmic vein, and artery (Fig 4).

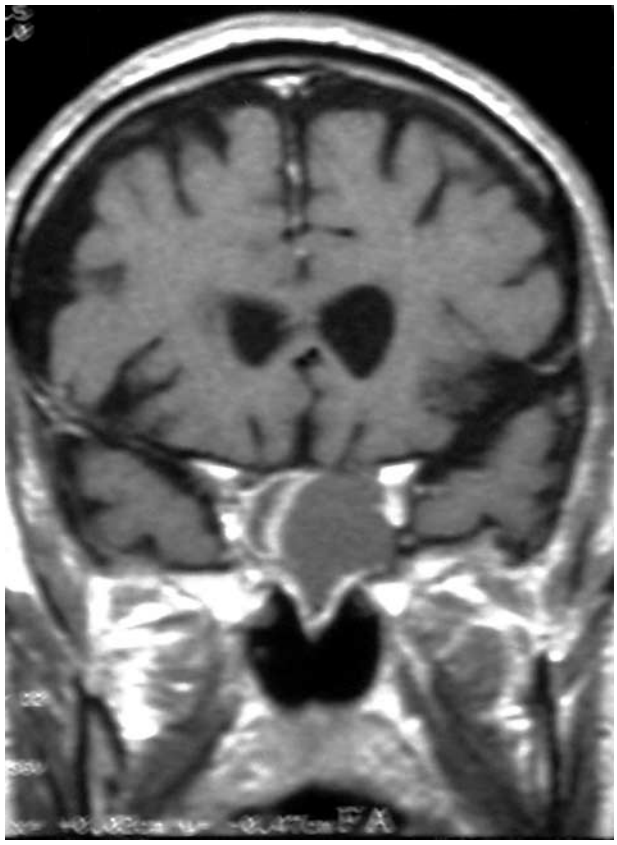

FIGURE 2. Preoperative coronal MRI showing left sphenoid sinus filled by hypointense material with contrast enhancement.

Criteria for diagnosis are based on all of these clinical symptoms, nasal endoscopic evaluation, and CT. Nasal endoscopic evaluation might reveal anatomic modification of the rhinopharynx such as reduction in the sphenoethmoidal recess. Definitive diagnosis is based on imaging. Plain radiology (radiographs of the skull and tomography) may show destruction of the intersinusal septum, of the anterior clinoid processes, or of the back wall of the orbital cavity or erosion of the sella floor and of the internal wall of the sphenoid fissure. CT, with its multiplanar display capability, can show isodense soft tissue mass and determine the intracranial or intraorbital extension of the lesion. To obtain a preoperative diagnosis of the mass, MRI may be useful. Mucocele T1-weighted sequences show hypointensity of the mass, whereas T2-weighted sequences show hyperintensity. Moreover, MRI can analyze the proximity of the mucocele to the optic nerve with use of oblique sagittal planes.[10,11]

Surgical access to the sphenoid sinus may be made through a craniotomy access, which is used only when the mucocele extends to the sella because it is highly invasive, or through a transnasal approach, assisted with a microscope, after the identification of the sphenoethmoidal recess.[12] A careful opening of

Table 1. CLINICAL MANIFESTATIONS OF SPHENOID MUCOCELE IN 133 PATIENTS REPORTED IN THE WORLD LITERATURE

Headache	89%
Decreased visual acuity	57%
Oculomotor palsies	56%
Exophthalmus	25%
Endocrine disorders	3.8%

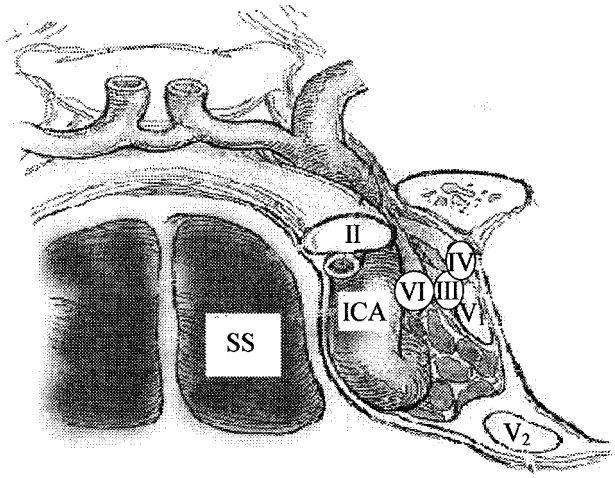

FIGURE 4. Anatomic coronal drawing of the sphenoid region showing the course of the vascular, neurologic and ophthalmologic structures: II, III, IV, V_1, V_2, and VI cranial nerves; internal carotid artery (ICA), and sphenoid sinus (SS).

the anterior wall of the sinus is necessary to avoid the risk of inadvertent injury to adjacent vital structures such as the internal carotid artery or optic nerve. Another method is a sublabial transseptal approach that increases the distance between the operative field and adjacent vital structures, but this can be complicated by septal perforation and purulent sinusitis.[13]

In our experience the transnasal approach allowed easy identification and opening of the sphenoid sinus with a low risk of complications.

The final purpose of this case report was to alert clinicians to consider a mucocele to be a cause of complete or incomplete isolated ophthalmoplegia among other causes, such as pituitary adenoma, craniopharingioma, internal carotid aneurysm, rhinopharyngeal carcinoma, and other neurologic diseases.

References

1. Hamard H, Sale MJ, Labetoulle C, et al: Les mucocèles du sphénoid et leur manifestations optalmologique. J Fr Ophthalmol 8:645, 1985
2. Chen HJ, Kao LY, Lui CC: Mucocele of the sphenoid sinus with apex orbitae syndrome. Surg Neurol 25:101, 1986
3. De Paepe E, Deweder M, Parisse J: Nevrite retrobulbaire dans une mucocele sphenoidale isolee. Acta Otorinolaryngol Belg 43:169, 1989
4. Rothfield RE, de Vries E J, Rueger RG: Isolated sphenoid sinus disease. Head Neck 13:208, 1991
5. Gilain L, Aidan D, Coste A, et al: Functional endoscopy sinus surgery for isolated sphenoid sinus disease. Head Neck 16:433, 1994
6. Sethi DS, Lau DP, Chan C: Sphenoid sinus mucocele presenting with isolated oculomotor nerve palsy. J Laryngol Otol 111:471, 1997
7. Rosier A, Rossazza C, Jan M, et al: Mucocèles du sphénoide. Rev Oto Neuro Ophthalmol 54:335, 1982
8. D'Alatri L, Almadori G, Ottaviani F, et al: Considerazioni sulle sinusiti sfenoidali isolate. Otorinolaryngol 41:93, 1991
9. Meissner R: Beitrag zur Symptomatologie der Keilbeinnhohlenmukozelen. HNO 31:175, 1983
10. Chui MC, Briant TD, Gray T, et al: Computed tomography of sphenoid sinus mucocele. J Otolaryngol 12:263, 1983
11. Dawson RC, Horton JA: MR imaging of mucoceles of the sphenoid sinus. AJNR Am J Neuroradiol 167:803, 1989
12. Hejazi N, Witzmann A, Hassler W: Ocular manifestations of sphenoid mucoceles: Clinical features and neurological management of three cases and review of the literature. Surg Neurol 56:338, 2001
13. Stevens HE, Blair NS: Intranasal sphenoethmoidectomy: 10-Year experience and literature review. J Otolaryngol 17:254, 1988