

# Amaurosis secondary to sphenoid mucocoele

## Amaurose secundária a mucocoele esfenoidal

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### SUMMARY

**Introduction:** The mucocoele of the sphenoid sinus is a benign rare lesion. Those lesions are probably diagnosed late because they are asymptomatic or cause non-specific symptoms. The clinical characteristics depend on its location and can include fronto-orbital pain, oculomotor nerve palsy, decrease of visual acuity, exophthalmos and olfaction disorders. The findings of the CT and the MRI of nose and paranasal sinuses have increased the diagnostic accuracy. The treatment consists of marsupialization and drainage of the mucocoele via endoscopic sinus. The prognosis for vision depends on the length loss of the visual acuity preoperative.

**Objective:** Report a case of sphenoid mucocoele of big dimensions.

**Case Report:** The authors report a case of sphenoid sinus mucocoele in a male patient of 48 years old, that has suddenly presented amaurosis.

**Final Comments:** The characteristics of the sphenoid mucocoele are reviewed with special attention for the clinical and radiological findings, as well as the surgical treatment.

**Keywords:** mucocoele, sphenoid sinus, visual acuity.

### RESUMO

**Introdução:** A mucocoele do seio esfenoidal é uma lesão rara e benigna. Essas lesões são provavelmente diagnosticadas tardiamente por serem assintomáticas ou causarem sintomas não específicos. As características clínicas dependem de sua localização e podem incluir dor fronto-orbitária, paralisia do nervo oculomotor, diminuição da acuidade visual, exoftalmia e anosmia. Os achados da tomografia computadorizada (TC) e ressonância nuclear magnética (RNM) de nariz e seios paranasais aumentaram a precisão do diagnóstico. O tratamento consiste na marsupialização e drenagem da mucocoele por via endoscópica nasossinusal. O prognóstico em relação à visão depende da duração da perda da acuidade visual pré-operatória.

**Objetivo:** Relatar um caso de mucocoele esfenoidal de grandes dimensões.

**Relato de Caso:** Os autores relatam um caso de mucocoele do seio esfenoidal em um paciente masculino de 48 anos de idade que apresentou amaurose subitamente.

**Comentários Finais:** As características da mucocoele esfenoidal são revistas com especial atenção para os seus achados clínicos e radiológicos, bem como o tratamento cirúrgico.

**Palavras-chave:** mucocoele, seio esfenoidal, acuidade visual.

## INTRODUCTION

The sphenoid mucocoele it's a benign lesion with slow development and late clinical manifestations which may worsen quickly (1). The mucocoeles are relatively infrequent, being more common in fronto-ethmoidal (2). Since the first sphenoid mucocoele identified by ROUGE in 1872, the diagnostic methods have progressed, mainly the Computed Tomography (CT) and more recently the Magnetic Resonance Imaging (MRI), that made the diagnosis possible in an early stage (1). The surgical treatment and the mucocoele prognosis depend on the time of evolution. In this article it's described in a case of mucocoele sphenoid sinus giving special attention to the clinical manifestations, radiological and surgical treatment.

## CASE REPORT

A male patient (S.J.S.), brown, 48 years, was admitted in the Neurological Restoration Hospital service - PE with history of frontal headache for about 20 days and orbital pain on the left. 4 days after the start of the symptoms noticed progressive decrease to bilateral visual impairment being of minor proportion on the right. Referred to bilateral nasal obstruction. Was submitted to CT of the skull, which showed the formation expansive hypodense that has not undergone contrast medium uptake compromising the sphenoid sinus. There was expansion of sphenoid sinus with thinning and remodeling of its bony walls, plus jaw ethmoid mucosal thickening. The lesion superiorly displaced the pituitary parenchyma extending the sella túrcica with superior repulse of the chiasmatic hypothalamic region. There was bulging of the soft tissues of the nasopharynx, obliterating its light and previously extends to posterior portions of the nasal cavities promoting remodeling of some intercellular septa posterior ethmoid. There was compression of optical channels, especially to the left. In view of being a expansive process of the sphenoid sinus of unknown etiology the patient was referred to the Otorhinolaryngology Service of Clinical Hospital of Federal University of Pernambuco (HC-UFPE) to evaluation. To the otorhinolaryngological exam of admission, was observed tumor in the left nasal cavity examination of the remaining normal. The evaluation of ophthalmology showed diffuse hypo pigmentation with iridian atrophy areas, mydriasis mean, glistening vitreous opacities, total pallor of the papilla, total excavation and normal macula in the left eye. In the right eye was observed without changes papilla and macula. The conclusion was optical atrophy and left divergent strabismus. The patient was submitted to endoscopic sinus surgery in December of 2000, observing the intraoperative the expansion of the

anterior wall of sphenoid sinus until the middle third of the nasal passages, most important to the left. A marsupialization was performed by partial resection of the previous wall of the mucocoele through the enlargement of the sphenoid ostium. The patient evolved in the post operative uneventfully, referring improve of the headache quickly and visual acuity on the right, but without any changes of the visual board of the left eye in relation to pre operative. The patient has presented a good clinical outcome over the subsequent revisions and at the end of twelve months there were no endoscopic and radiological signs of recurrence of the disease. The CT of paranasal sinuses after six months of the post operative showed and extensive area in pneumatized sphenoid extending into the nasal passages.

## DISCUSSION

The mucocoele presented in this work was sphenoidal, the rare condition when observing the same frequency among the various sinuses involved. As found in the literature the patient was male and belonged to the most common age of involvement, usually the fourth and fifth decades of life (1). There was no association with previous surgery, and no history of nasal polyposis, however had ethmoid sinus involved in this case, as can happen in about half of the patients with this disease (3,4). The symptoms that emerged in the patient, like nasal obstruction and headache disappeared after the treatment, what makes you think that the indirect compression was the main mechanism of origin of them (5-7). However, the visual loss improved only on the right of which there was irreversible damage to the left. This situation may be because of the indirect compression from vascular origin responsible by the reversible and transient symptoms. The headache which is the earliest and constant sign was one of the symptoms of opening also in this case and distinctively retro-orbital. This exceptional finding, according to the literature, it is in this case a ophthalmological sign, especially the amaurosis, was one of the symptoms that started the clinical picture (1). Although other cranial nerves (III, IV, VI) can be affected, this did not occur in this patient, nor the exophthalmos. Also there were no endocrine signs or intracranial complications as, by the way, are infrequent in these cases (8, 9). As described in the literature, the mucocoele thinned the walls of the sphenoid sinus, filling it fully, but the contrast used in CT did not change its density. Since the CT is the method of choice to diagnosis and the same was defined, was not considered necessary for carrying out the RMI (10). The option of surgical treatment was of endoscopic transnasal access with opening of the sphenoid sinus and its complete marsupialization as it is found as normally described (11). In the same way, there were no important complications because it is a low

morbidity procedure (12). The minimum follow-up of one year was respected and there are no relapses in this period, but should be emphasized that may occur later.

### FINAL COMMENTS

The sphenoidal mucocele is a rare disease and for cursing insidiously have a late diagnosis and which diagnostic method of election is the CT of the paranasal sinus with contrast. It is a disease that must be remembered in the differential diagnosis of the sphenoid lesions by the otorhinolaryngologist. The treatment usually made is the marsupialization and drainage by nasosinusal endoscopy.

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