

# An unusual case of sphenoid sinus mucocoele with severe intracranial extension\*

John Daniilidis<sup>1</sup>, Angelos Nikolaou<sup>1</sup>, Vassilios Kondopoulos<sup>2</sup>

<sup>1</sup> University Department of Otorhinolaryngology, AHEPA University Hospital, Thessaloniki, Greece

<sup>2</sup> University Department of Neurosurgery, AHEPA University Hospital, Thessaloniki, Greece

## SUMMARY

*We present an unusual case of sphenoid sinus mucocoele with severe intracranial extension, which was diagnosed and treated at the AHEPA General Hospital, University of Thessaloniki, Greece. The patient was a 54-year-old female who presented with endocrine, nasal and ophthalmological symptoms, and headache. She was successfully treated by surgery using an intranasal approach. While presenting this case, we briefly review the literature on the subject of sphenoid sinus mucocoeles.*

*Key words: mucocoele, sphenoid sinus, surgery*

## INTRODUCTION

In 1889, Berg first described the existence of sphenoid sinus mucocoeles and there have been about 150 such cases presented in the medical literature since then (cf., Stankiewicz, 1989; Lidov et al., 1990). Only 1% of the paranasal sinus mucocoeles arise within the sphenoid sinuses. Precise diagnosis can be achieved with the use of CT scan and MR imaging, provided that the great variety of the presenting symptoms will lead to the suspicion of mucocoele existence in the sphenoid sinus.

Differential diagnosis should be made from: (1) cystic adenoma of the pituitary gland; (2) a cyst derived from remnants of Rathke's pouch; (3) a neoplasm (benign or malignant) of this area; and (4) a retention cyst arising from an obstruction of the duct of a minor salivary gland. In the latter case the differential diagnosis is purely clinical and not histological, being based mainly on the size of the lesion. Mucocoeles are larger, occupying the whole sphenoid sinus, and extending beyond the boundaries of the sinus, since they result from obstruction of the sinus ostium. Retention cysts are smaller, leaving some residual air within the sinus (Lidov et al., 1990). "Empty sella" with a meningocele in the sphenoid sinus should also be considered in the differential diagnosis, since it produces symptoms similar to those of a mucocoele.

## CASE REPORT

A 54-year-old woman was presented with a 10-month history of hypothyroidism and a 4-month history of nasal block-

age due to polyps, which were removed under local anaesthesia on an out-patient basis at another hospital. Three months before her admission to our hospital, she experienced headaches and during the last 15 months visual complaints were added to the picture. Physical examination upon admission revealed recurrent nasal polyps, and ophthalmologic examination showed decreased visual acuity (more severe in the right eye) as well as a reduction of the visual fields. Plain radiographs demonstrated severe widening of the sella turcica (Figure 1). Diagnosis was finally established by means of CT scan and MR imaging (Figure 2) which revealed a 4.5-cm expansile lesion, occupying both sphenoid sinuses and extending intracranially into the sella turcica, displacing the pituitary gland and the optic chiasm upwards. The post-contrast image of the wall of the cystic lesion showed contrast-enhancement.

The patient was operated upon under general anaesthesia. A transnasal approach was used to reach the site of the mucocoele. The first surgical steps included septoplasty, polypectomy and total intranasal ethmoidectomy. The sphenoid sinus was then opened, under the operating microscope, and was found to be totally occupied by the mucocoele. Part of the epithelial wall was removed and sent for pathological examination. The mucoid fluid was then drained, the remaining epithelial wall of the cele was resected and the operation was completed with marsupialization of the sphenoid sinus. The histology report revealed that the wall of the cystic lesion was lined with a secretory cuboidal epithelium, suggestive of a mucocoele. The patient recovered



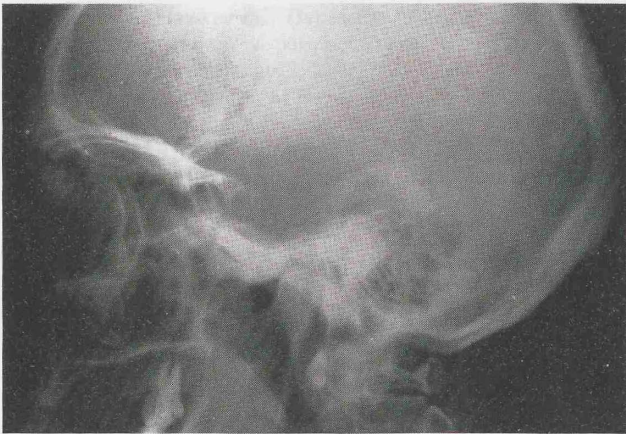


Figure 1. Lateral radiograph of the skull showing extensive widening of the sella turcica.

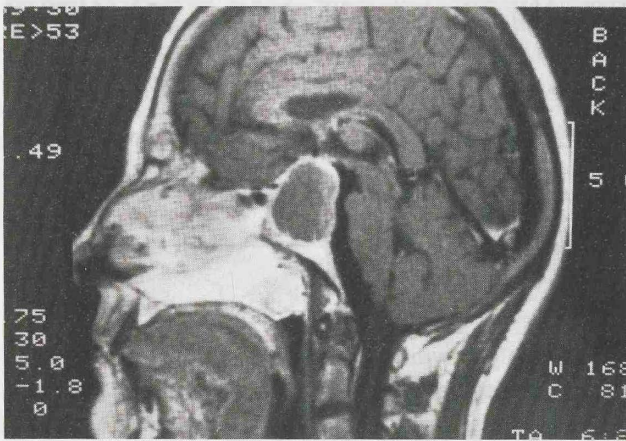


Figure 2. Sagittal scan in T1-weighted MRI showing clearly the intracranial extension of the mucocoele.

uneventfully. Within the first post-operative days there was a rapid recession of both the visual problems and the headaches. A follow-up CT scan, 5 months post-operatively, revealed persistence of the defect in the sella turcica, but absence of sphenoid mucocoele.

#### DISCUSSION

We have already referred to the rare incidence of sphenoid sinus mucocoeles. As far as the expansion of these cystic formations is concerned, it is known that with time they erode bone and extend out of the sinus that initially contains them. In experimental studies attempting to interpret the mechanism of bone erosion, fibroblasts have been cultured from excised portions of mucocoele mucosa. When these cultured fibroblasts were stimulated by a mononuclear cell culture, significantly elevated levels of prostaglandin  $E_2$  and collagenase were produced, as compared to normal sinus mucosa fibroblasts. These findings suggest that lining fibroblasts are a major source of bone-resorbing factors (Lund et al., 1988). It should also be considered that trauma accompanied with fractured osseous wall of a paranasal sinus can lead to mucocoele formation.

Sphenoid sinus mucocoeles present with various symptoms, usually when they have reached the "right" size, so as to produce symptoms by exerting pressure to the neighbouring structures. It has been reported that in 71% of the cases there is a history of headache, in 61% there are visual symptoms, in 50% nasal symptoms, and in only 5% endocrine symptoms (Lundgren and Olin, 1961; Stankiewicz, 1989). In our patient, all these symptoms were present and it is interesting to note that the first symptom was hypothyroidism, which is very infrequent according to the literature.

Radiological examination played a primary role in the diagnosis and differential diagnosis of the disease. Plain radiographs show opacity in the sphenoid sinus, and widening of the sella turcica in cases of very large mucocoeles, which was the case with our patient (Figure 1). The CT scan provides precise information on the bony structures, whereas MR imaging with the highest percentage of precision sets the final diagnosis.

At least 5 cases of sphenoid mucocoeles with coexisting neoplasms (benign or malignant) have been reported in the literature (Schaeffer et al., 1985; Alleva et al., 1989; Weaver and Bartley, 1991; Lanzieri et al., 1991). In these cases a myxoma, a mucoepidermoid or other carcinoma initially obstructed the sinus ostium of the sphenoid and consequently a mucocoele developed.

Various surgical approaches have been proposed for the treatment of sphenoid mucocoele. Some of these are the intracranial approach (which has been abandoned), the external sphenoidectomy, the transantral and, finally, the intranasal approach. This latter approach can be performed either transseptally or through the nasal cavity, usually after septoplasty.

During the last 5 years, studies have been published on the intranasal approach using endoscopes (Stankiewicz, 1989). In our case, intranasal ethmoidectomy was elected in order to remove the recurrent polyps and this in combination with septoplasty enabled us to reach the sphenoid sinus with the help of an operating microscope. Regardless of which surgical technique each surgeon uses to approach the sphenoid sinus mucocoele, it is important to mention that the epithelial wall must be removed in total in order to avoid recurrence of the cele. Manipulations in the area of the sphenoid mucocoele must be very careful, because important structures, e.g. internal carotid arteries and optic nerves, are usually found exposed and displaced and can be easily damaged. Intraoperative fluoroscopic control (which was also used in our case) helped to rapidly locate the wall of the sphenoid sinus, and that of the mucocoele as well.

We conclude that despite the fact that sphenoid mucocoele is a rare disease, it should be included in our diagnostic assessment. Early stages of mucocoele development are asymptomatic, but when significant expansion, beyond the osseous walls of the sinus, occurs, a variety of symptoms from organs and systems out of the immediate boundaries of Otolaryngology can be presented. The diagnosis is usually established by means of CT scanning and MR imaging.

## REFERENCES

1. Alleva MD, Werber JL, Kimmelman CP (1989) Pathologic quiz case 2. Arch Otolaryngol Head Neck Surg 115: 878-881.
2. Lanzieri CF, Shah M, Krauss D, Lavertu P (1991) Use of gadolinium-enhanced MR imaging for differentiating mucoceles from neoplasms in the paranasal sinuses. Radiology 178: 425-428.
3. Lidov M, Behin F, Som P (1990) Calcified sphenoid mucocele. Arch Otolaryngol Head Neck Surg 116: 718-720.
4. Lund VJ, Harvey W, Meghji S, Harris M (1988) Prostaglandin synthesis in the pathogenesis of fronto-ethmoidal mucoceles. Acta Otolaryngol (Stockh) 106: 145-151.
5. Lundgren A, Olin T (1961) Mucocele of the sphenoidal sinus. Acta Otolaryngol (Stockh) 53: 61-79.
6. Schaeffer BT, Som PM, Sacher M, Lanzieri CF, Solodnik P, Lawson W, Biller HF (1985) Coexistence of a nasal mucoepidermoid carcinoma and sphenoid mucoceles: CT diagnosis and treatment implications. J Comput Assist Tomogr 9: 803-805.
7. Stankiewicz JA (1989) Sphenoid sinus mucocele. Arch Otolaryngol Head Neck Surg 115: 735-740.
8. Weaver DT, Bartley GB (1991) Malignant neoplasia of the paranasal sinuses associated with mucocele. Ophthalmology 98: 342-346.

Prof. John Daniilidis  
Department of Otorhinolaryngology  
Aristotelean Medical School  
AHEPA University Hospital  
Thessaloniki  
54636 Greece