

Sphenoid Sinus Mucocele With Intra And Suprasellar Extension - A Diagnostic Dilemma

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Abstract : A case of mucocele of the sphenoid sinus that presented with sellar and parasellar extension is reported. This rare entity should be considered in the differential diagnosis of lesions causing destruction of the sellar floor even if there is lack of historical or radiographic evidence of primary paranasal sinus disease. CT

has greatly aided the demonstration of the mass but a correct differential diagnosis may have to be made in conjunction with a radiologist. Transsphenoidal approach is the procedure of choice, since there exist dreadful complications of craniotomy.

Key Words : Mucocele, Sphenoid sinus, Transsphenoidal surgery

INTRODUCTION

Sphenoid sinus mucocele is rare and often misdiagnosed (3,7). Since its first description by Berg in 1889 (2), less than 150 cases have been reported in the medical literature, notably neurosurgical and otolaryngological (1,3-7,9-13,15-17). This benign, readily treatable lesion is potentially fatal if misdiagnosis prompts an exploratory craniotomy for a presumed intracranial lesion (6,7,11). To make the correct diagnosis preoperatively, a careful radiological examination and interpretation is crucial. The purpose of this paper is to present a case in conjunction with a brief review of the clinical, radiological and surgical highlights of this uncommon disease.

CASE HISTORY

A 48-year-old air force colonel presented with three-week history of double vision. On admission, he was alert and general examination was essentially normal. Neurological examination revealed a partial left sixth nerve palsy. Plain skull x-rays showed destruction of the sellar floor, dorsum sellae, anterior as well as the posterior clinoids and partly the clivus (Fig. 1). CT scans demonstrated a huge mass

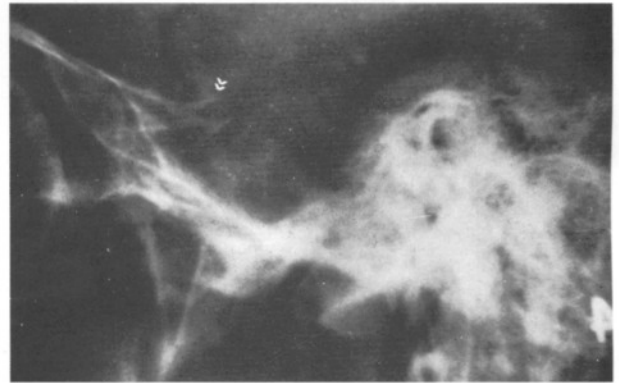


Fig. 1 : Lateral plain skull x-ray demonstrates partial opacification of the sphenoid sinus and destruction of the sellar floor, dorsum sellae, posterior clinoids and the clivus. Note the elevation of the anterior clinoids (white arrowheads).

measuring 3x4 cm filling the sella, sphenoid and posterior ethmoid sinuses which also extended down to the nasopharynx and up to the suprasellar region (Fig. 2 - 3). The contrast enhancement was minimal and inhomogenous. These findings were suggestive of a nasopharynx carcinoma or an invasive pituitary adenoma or less likely a chordoma. An intravenous digital subtraction angiogram revealed lateral displacement of the cavernous segment of both

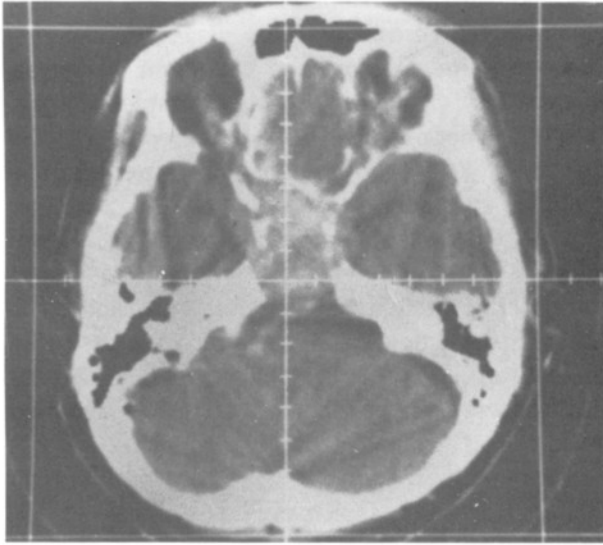


Fig. 2 : Axial CT scan after contrast injection demonstrates an inhomogeneously enhancing slightly hyperdense mass filling the entire sphenoid sinus and extending to anterior posterior and lateral directions.

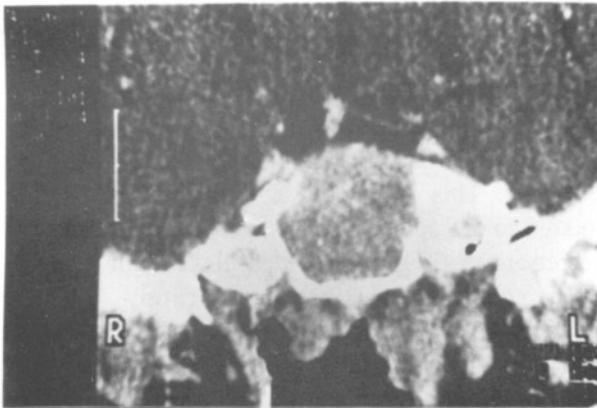


Fig. 3 : Coronal CT scan with contrast delineates the extent of the supra- and infrasellar involvement.

carotid arteries more on the left and elevation of the A1 segment of both anterior cerebral arteries. A pituitary screen revealed normal values for TSH, GH, ACTH, PRL, LH and FSH as well as a.m. cortisol. A computerized perimetry confirmed the normal visual fields as observed on confrontation.

An otolaryngological opinion was then sought in an attempt to verify the nature of the pathology through nasopharynx. Examination revealed a huge nasal polyp filling the posterior choana and the nasopharynx, which was partially excised under local anesthesia. The histopathological examination showed an inflammatory nasal polyp rich in mononuclear cells suggesting an allergic origin.

Two weeks later, the patient was taken to theatre for a standard sublabial transsphenoidal surgery. Upon exploration the mucosa over the floor of the sphenoid sinus was found very thick and after dissection the bony floor was found open on the left. The sinus was filled by a grayish-yellow mucoid mass which was partly cheesy all of which could be sucked out. When the floor of the sella was exposed, it was found to be largely eroded. The extension of the mass into the sella was quite circumscribed and upon removal the pituitary gland was found pushed superio-posteriorly. Subsequently the diaphragma sellae and the normal pulsation of the brain was observed.

Histopathological examination of the mass revealed a tissue rich in acellular mucin and columnar epithelium forming islands within. There appeared scanty necrosis.

The postoperative period was uneventful. The recovery from double vision was prompt. Discharged symptom-free on the 7th postoperative day, the patient is asymptomatic at 4 years and the pituitary function tests have remained normal.

DISCUSSION

The etiology of paranasal mucoceles is controversial (7). They usually occur when drainage of a paranasal sinus is obstructed due to inflammation, fibrosis, trauma, previous surgery or anatomical abnormality (3,11). Mucoceles occur most commonly in the frontal and ethmoid sinuses but are quite infrequent in the sphenoid sinus (9). Nugent et al (11) reviewed the literature in 1970 and were able to find 81 cases of which 63 were available for a thorough review. Interestingly in 29 patients (46%), there was a history of previous otolaryngological disease in the form of sinusitis, intranasal polyps, nasal mass or nasal discharge. An allergic state is a predisposing factor for the formation of a polyp hence obliterating the outflow and therefore should be questioned preoperatively.

The clinical and radiographic manifestations of sphenoid sinus mucoceles are usually related to sinus expansion and extension of the lesion beyond the confines of the sinus (11). This usually follows the path of least resistance, namely anteriorly to involve ethmoid air cells and orbits but occasionally posteriorly into the clivus and superiorly into the

sella turcica (10). Clinically, the symptoms and signs which together bear names as orbital apex syndrome, sphenoidal fissure syndrome, orbital inlet syndrome, superior orbital fissure syndrome and anterior cavernous sinus syndrome are therefore all non-specific (15). The most common complaint in Nugent et al review (11) was headache occurring in 55 out of 63 (87%). The headache is typically frontal or retroorbital, tends to be worse towards evening (15). It may be due to dural stretching if the lesion breaks through the sella turcica (10). However it is the visual symptoms that alert the patient and his clinician to the seriousness of the problem (15). In 41 patients (62%) vision in at least one eye was impaired to some degree. Fifteen were blind in one eye and five others were bilaterally blind. Among the 19 cases presenting with diplopia (30%), 12 patients had third cranial nerve involvement while in seven there was sixth nerve compression (11).

Evaluation of the pituitary function was carried out in four patients in Nugent et al review (11) and in a few cases reported thereafter (1,7,16), but in none including our case was there evidence of pituitary insufficiency.

Plain skull x-rays still are the essentials for a correct preoperative diagnosis. Opacification of the sphenoid sinus in mucocele, has been considered as the constant feature of this entity usually depicted by laminography (13). Plain x-rays delineate well the destruction of the sellar floor, the clinoids and the clivus (13,17). Ballooning of the sella turcica characteristic of pituitary adenoma is unusual with sphenoid sinus mucocele, which rather produce pressure erosion from below (13). Very infrequently the petrous apex may be eroded (15). Lamina papyracea is very thin and offers little resistance to an expanding mucocele (15). Simon and Tingwald (17) summarizing the radiographic findings of sphenoid sinus mucocele, suggested that radiography should include satisfactory demonstration of the superior orbital fissure, optic foramina, lateral walls of the ethmoid sinuses, the floor of the sella turcica, walls of the sphenoid sinus, lesser wings of the sphenoid and the medial walls of the orbits (17). In the review by Nugent et al (11), the floor of the sella was eroded in 29 cases out of 63 cases reviewed (46%). In only two instance there was suprasellar extension of the mucocele as demonstrated by the plain radiographs. An intracranial lesion was suspected in

15 instances. The presumed pathology was a pituitary tumor in ten cases. The other diagnoses included chordoma, meningioma and aneurysm. Not infrequently, with radiographs showing complete destruction of the floor of the sella as occurred in our case and possible opacification of the entire sinus by pituitary adenoma, distinction between adenoma and mucocele may not be possible on radiological grounds (13). It is important for the radiologist to bear in mind this alternative diagnosis as his clinical colleague, for it is he who may first recognize its possible existence. Pure intrasellar mucocele without involvement of the sphenoid sinus has been reported on only one occasion (1) and in such a case it is not possible to differentiate between a mucocele and an adenoma by plain skull x-rays.

CT has greatly aided the demonstration of a mass arising from the sphenoid sinus and extending upwards to involve the sella and the neighbouring structures. Coronal scans and bone window studies are essential for evaluating the cribriform plate, the roof of the ethmoid sinuses, the involvement of the supraorbital ethmoid extension and extension into the superior orbital fissure (9). Mucoceles may appear isodense and occasionally hypo- or hyperdense (3,4,9,12). This difference in attenuation may be related to the age and consistency of the entrapped secretions (6). In general, mucoceles do not enhance with contrast infusion, but acutely infected mucopyoceles may show rim enhancement (4,9,12). It is rather difficult to suggest a characteristic CT pattern for sphenoid sinus mucoceles but the absence of contrast enhancement is understandable in the light of the avascular mucoid contents of the mucocele, therefore the presence of intrinsic enhancement tends to suggest various pathologies either originating from above as pituitary tumors, craniopharyngiomas, meningiomas, optic gliomas, aneurysms, germinomas and metastatic lesions or originating from below as juvenile nasopharyngeal angiofibroma, sphenoid sinus carcinoma, nasopharynx carcinoma and chordoma. Carotid angiography offers little in the way of positive diagnostic information and only serves to exclude the presence of a vascular tumor or an aneurysm in the sphenoid region (15). Magnetic resonance (MR) scans can be as sensitive as CT for demonstrating bony destruction (14), however CT has better spatial resolution and will show the bony destruction in more

detail (1). MR, on the other hand is more sensitive than CT in defining the margins of the lesion and their relationship to normal parasellar anatomy (14). Mucoceles, particularly long-standing lesions, have higher protein content, resulting in a shorter T1- and longer T2 that produce high signal on both T1- and T2-weighted images (8). Unfortunately MR was not available in Ankara at the time of diagnosis.

As of surgical technique, the primary approach was an otolaryngological one in 45 patients among the 63 reviewed (11) which was curative in all excluding cases with preexisting blindness. External ethmoidectomy, transantral-, transseptal- and endonasal-sphenoidotomy were the procedures preferred by the ENT surgeons (10,15). Nevertheless, in 16 cases out of 63 (25%), a craniotomy was undertaken by the neurosurgeons in an attempt to treat a presumed intracranial lesion. In 3 patients out of this 16 (19%), death resulted from infectious complications of the operation, and in two others meningitis ensued but responded to therapy. Such a high morbidity is because of the high probability of the contents of the mucocele getting infected. Among the 18 patients having had cultures in the classical review (11), there were six with positive cultures for staphylococci, streptococci, enterococci and fungus. Though many neurosurgeons with a correct preoperative diagnosis prefer a transsphenoidal approach with which they are more familiar with, Hakuba et al (7) from a neurosurgical department also reported successful results with transpalatal approach. Interestingly, polypectomy alone cured such a lesion reported recently which is rather unexpected (5). In conclusion, we presented the case of a patient harboring an extensive mucocele of the sphenoid sinus. This rare but benign entity is readily treatable if diagnosed correctly preoperatively and approached transsphenoidally. Incomplete radiographic study or superficial examination of the films may lead to the erroneous diagnosis of a pituitary tumor and establish a trend of thought leading to an ill-advised operation.

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