Case Reports

MUCOCELES OF THE PARANASAL SINUSES WITH INTRAORBITAL AND INTRACRANIAL INVASION

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The authors review five cases of paranasal sinus mucocele invading the orbit and cranial cavities which were treated in their neurosurgical unit over a 7-year period. The clinical features of the patients, locations of the lesions, CT findings and operative results are analysed. Early investigation by CT scan and prompt surgical treatment are important if irreversible neurological deficits are to be avoided.
Saudi J Ophthalmol 1994;8:131-136

Key words
paranasal sinus diseases – mucocele – orbital diseases

PARANASAL SINUS MUCOCELES are expanding lesions lined by sinus mucosa and containing inspissated mucus.[1] If they are not treated, they slowly spread to surrounding bone and soft tissues and invade the orbit and cranial cavity.[2,3] They are an uncommon pathology, accounting for 0.4% of intracranial space-occupying lesions in our hospital.[4]

A localised paranasal sinus mucocele is usually managed by ear, nose and throat (ENT) surgeons or ophthalmic surgeons. However, the presence of an extension of the mucocele into the cranial cavity or orbit is, in most centres, an indication for neurosurgical intervention.[2,5-8] Invasive mucoceles commonly produce a variety of ophthalmological signs which may aggravate rapidly and require urgent surgical intervention. Greater awareness of the various clinical manifestations of this pathology is essential for the early diagnosis, referral, and surgical treatment necessary to achieve the best possible outcome for patients.

In this article we present the clinical, radiological and operative features of five cases of paranasal sinus mucoceles with intraorbital and intracranial extension that we treated in our neurosurgical unit over the last 7 years. To our knowledge, this is the first report of invasive mucoceles from Saudi Arabia.

Case Reports

Case 1

A 60-year-old Saudi female presented in 1993 with a 1-year history of headache and blurred vision in the left eye. She had previously undergone cataract extraction in the left eye without implantation of an IOL. Examination revealed proptosis of the left eye. Visual acuity was hand movement in both eyes. The right eye had an opaque cornea. The left eye showed mild conjunctival injection and a clear corneal graft. Fundoscopy revealed an atrophied left optic nerve. CT scan (Figure 1a) showed an isodense mucocele involving the sphenoid and ethmoid sinuses which extended into the left orbit, anterior cranial fossa and nasal cavity. The patient underwent transnasal drainage of the mucocele. A wide opening was made in the capsule to allow

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drainage of the mucinous material into the nose. The mucocele contained yellow mucinous fluid, the culture of which was negative. Histology of the excised part of the capsule revealed tissue lined by pseudostratified columnar ciliated epithelium infiltrated by chronic inflammatory cells. The patient made a good postoperative recovery. At the 4-month follow-up appointment, the headache and proptosis had improved although visual acuity was unchanged. CT scan (Figure 1b) showed an empty mucocele cavity.

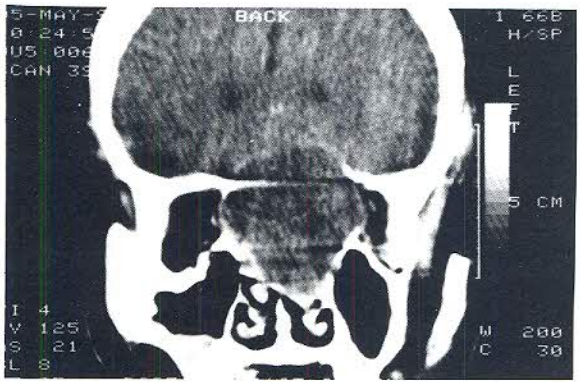

Figure 1a. Case 1. CT scan (with IV contrast) taken preoperatively shows a mucocele involving the sphenoid and ethmoid sinuses and extending into the left orbit, anterior cranial fossa and nasal cavity.

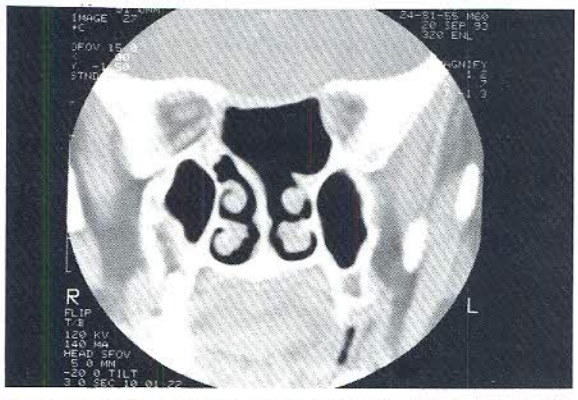

Figure 1b. Case 1. CT scan (with IV contrast) taken 4 months after surgery shows an empty mucocele cavity in the ethmoid and sphenoid sinuses.

Case 2

A 70-year-old Saudi female presented in 1992 with a 2-year history of proptosis of the left eye with displacement of the eyeball laterally. She was known to have bilateral visual failure caused by glaucoma of 10 years' duration. On admission she was found to have no light perception in the left eye and counting fingers in the right eye. CT scan (Figure 2a) showed an isodense mucocele involving the left frontal and ethmoidal sinuses and extending into the left orbit, nasal cavity and anterior cranial fossa. The patient underwent a craniotomy and radical excision of the mucocele and its capsule. The mucocele contained yellow-green fluid, the culture of which was negative. Histology of the capsule revealed fibrous tissue infiltrated by chronic inflammatory cells and pus cells. A CT scan taken 10 days after the surgery (Figure 2b) revealed a resolving postoperative hematoma and air bubbles in the cavity of the excised mucocele. At a follow-up consultation 2 months postoperatively, the patient's proptosis and eye displacement had improved but her visual acuity was unchanged. The patient has not attended the clinic since that consultation.

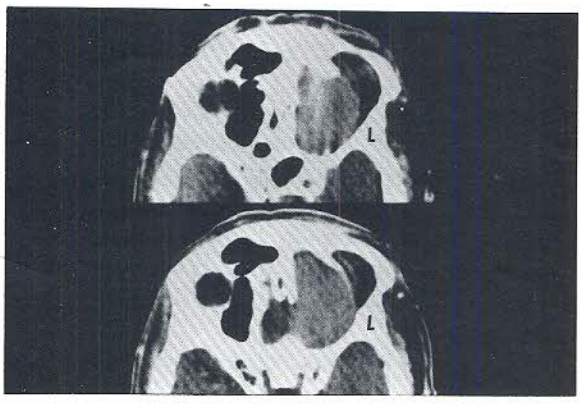

Figure 2a. Case 2. CT scan (without contrast) taken preoperatively shows a mucocele involving the ethmoid sinus and extending into the left orbit and nasal cavity.

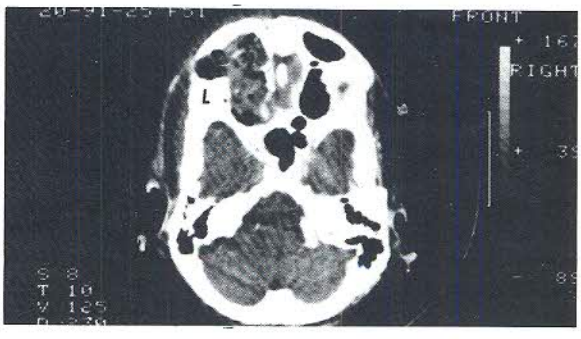

Figure 2b. Case 2. CT scan (without contrast) taken 10 days after surgery shows evidence of a postoperative hematoma and air bubbles in the cavity of the mucocele (note that the left/right scanner notation is the reverse of the notation used in Figure 2a).

Case 3

A 36-year-old Saudi male presented in 1990 with a history of pain and swelling in the left eyelid of 3 months' duration. Ten years previously, the patient had had a road traffic accident causing craniofacial fractures and damage to the left optic nerve. On admission the patient had visual acuity of 20/50 in left eye and 20/20 in right eye. There was proptosis of the left eye with downward displacement of the left globe. CT scan (Figure 3a) showed an isodense mucocele involving the left frontal and ethmoidal sinuses and extending into the left orbit, anterior cranial fossa and nasal cavity. The patient underwent a left frontal craniotomy and radical excision of the mucocele and its capsule. The mucocele contained turbid fluid, the culture of which was negative. Histology of the capsule revealed thick hyalinised fibrous tissue with a thick layer of chronic inflammatory cells. The patient made a good postoperative recovery. At a follow-up visit 1 year after surgery there was marked improvement in the patient's left eye proptosis, eyelid swelling and eyeball displacement. Visual acuity, however, was unchanged. A CT scan taken at this visit (Figure 3b) showed that the mucocele had disappeared completely and had not recurred.

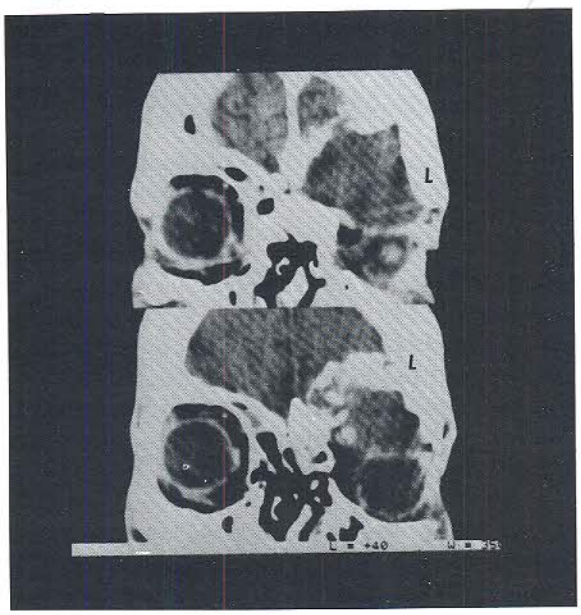

Figure 3a. Case 3. CT scan (without contrast) taken preoperatively shows a mucocele involving the left frontal and ethmoid sinuses and extending into the orbit and anterior cranial fossa.

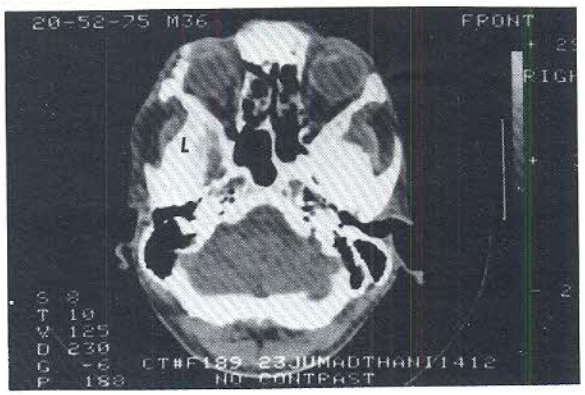

Figure 3b. Case 3. CT scan (without contrast) taken 1 year after surgery shows no evidence of recurrence of the mucocele.

Case 4

A 69-year-old male Saudi had cataract extraction with implantation of an intraocular lens in the left eye in 1987. He did well and vision in his left eye improved. In 1990, he presented with a squint of 6 months' duration which was investigated by the orthoptic department and confirmed to be due to an abducent nerve palsy. CT scan (Figure 4a) showed an isodense mucocele invading the sphenoid and ethmoid sinuses and the middle cranial fossa. The patient underwent a transnasal transsphenoidal evacuation of the mucocele which contained thick black gelatinous material, the culture of which was negative. The patient made a good postoperative recovery. A CT scan (Figure 4b) taken 2 weeks post-operatively showed minimal residual mucinous material in the mucocele capsule. At a follow-up visit one year later, the patient's squint had failed to improve.

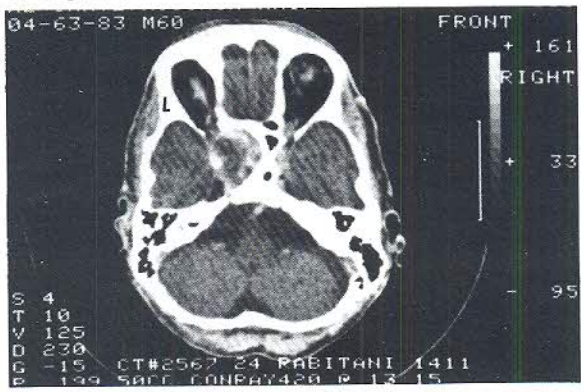

Figure 4a. Case 4. CT scan (with IV contrast) shows a mucocele involving the sphenoid sinus and extending into the left middle fossa.

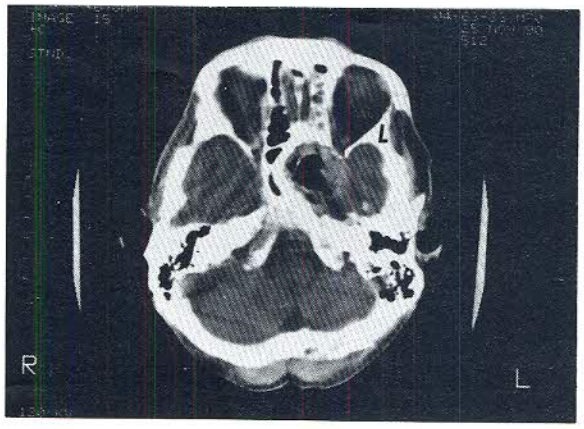

Figure 4b. Case 4. CT scan (with IV contrast) taken 2 weeks postoperatively shows a minimal residual collection of mucinous material in the mucocele cavity (note that the right/left scanner notation is the reverse of the notation used in Figure 4a).

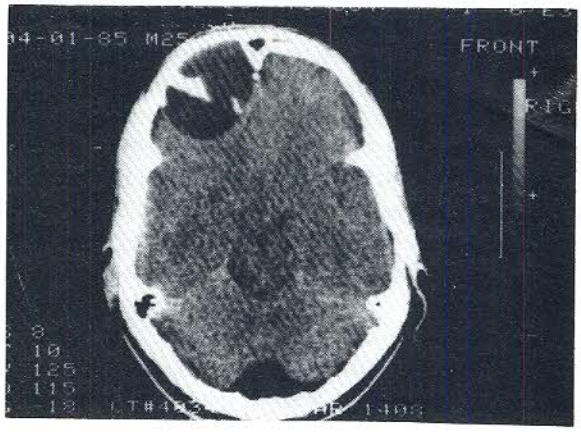

Figure 5a. Case 5. CT scan (without contrast) taken preoperatively shows a hypodense mucocele of the left frontal sinus invading the left frontal lobe.

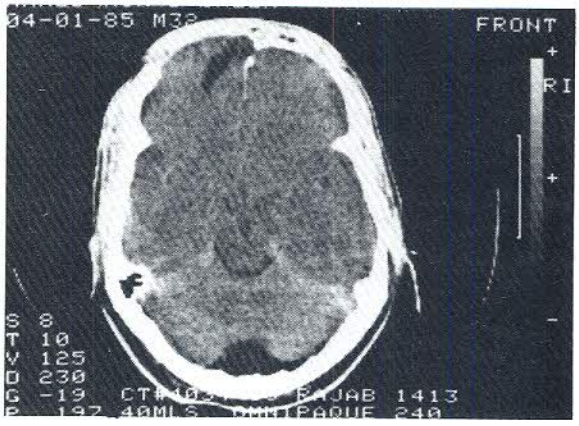

Figure 5b. Case 5. CT scan (with IV contrast) taken 5 years after surgery shows no evidence of residual or recurring mucocele.

Case 5

A 28-year-old Saudi male, who was known to suffer from recurrent attacks of sinusitis, presented in 1988 with 6 months' history of ptosis of the left eyelid with downward displacement of the left eyeball. Visual acuity was 20/70 in the left eye and 20/25 in the right eye. The patient also reported that a mucinous secretion came out of his nostril after pressure on the eye (rhinomucorrhea) and that he had become anosmic. CT scan (Figure 5a) revealed a hypodense mucocele in the left frontal and ethmoidal sinuses invading the orbit and anterior cranial fossa. The patient underwent a craniotomy and radical excision of the mucocele and its capsule. The mucocele contained turbid fluid, the culture of which was negative. Histology of the excised capsule revealed fibrous tissue with a thick layer of chronic inflammatory cells. The patient made a good postoperative recovery. At a 5-year follow-up visit, there was improvement in the ptosis and displacement of the eye as well as in the rhinomucorrhea. Visual acuity in the right eye had improved to 20/25 (bilateral visual acuity was therefore 20/25) but the anosmia persisted. A follow-up CT scan (Figure 5b) showed no evidence that the mucocele had recurred.

Discussion

Though Langenbeck described the basic clinical features of paranasal sinus mucoceles in 1819, it was not until 1886 that Rollet introduced the term "mucocele" to describe these lesions.[1] The first English report of a mucocele was by L. Turner early in the 20th century.[6]

Paranasal sinus mucoceles are caused by obstruction of the sinus ostia which is often secondary to infection, allergy, trauma and surgery. Occasionally, benign neoplasms such as osteomas, fibrous dysplasia and primary or

metastatic malignant tumours obstruct the sinus ostia and cause mucoceles to form.[1] Following obstruction, bone resorption with elevated mucocele levels of PGE2 and collagenase occurs and the mucocele acts as a mass.[9] It is reported that paranasal mucoceles are associated with a history of sinusitis in 35% to 50% of cases, trauma in 11% to 77% and allergy in 11% to 33%.[1,2,6] One of our patients had a history of trauma while another had a history of sinusitis.

Paranasal sinuses mucoceles affect both sexes, usually during the 3rd and 4th decades of life.[1] The mean age of our patients, however, was 53 years. The disease commonly involves the frontal and ethmoidal sinuses. Involvement of the sphenoid sinus is relatively rare with only 100 cases reported in the literature up to 1992.[10] The ethmoid sinus was involved in all of our cases, while the frontal sinus was involved in three patients and the sphenoid sinus in two. The mucocele extended into the orbit and anterior cranial fossa of four patients; in one patient, the lesion extended into the middle cranial fossa.

Various clinical features related to invasive mucoceles have been described. Proptosis, which is reported in 22% to 83% of cases in various series,[1,2,11] was found in three (60%) of our patients. Reduction of visual acuity is a recognised important feature which may necessitate urgent drainage of the mucocele since a correlation can be made between the length of history and improvement in vision. In recent reports,[2,7] 22% to 50% of patients with visual impairment did not regain visual acuity after the mucocele surgery; all of these cases had a history of mucocele of more than 1 year's duration.

Optic atrophy, attributed to the pressure exerted by the mucocele on the optic nerve, is also likely to be associated with poor outcome, as are inflammation and orbital vessel disturbances; the latter two are implicated in poor outcome because of the improvement in vision that sometimes occurs with steroid treatment.[1,7]

Of the four cases with visual impairment reported here, only one, Case 5, showed improvement in visual acuity after surgery. Because records of visual acuity prior to mucocele development were not available in the other three cases with visual impairment, it is not clear whether failure to improve was related to existing ocular pathology (Cases 1 and 2 had a long-standing history of primary ocular disease and Case 3 had optic nerve damage following a craniofacial injury) or to the optic nerve damage caused by the mucocele, particularly as the history of mucocele was relatively long in these patients (3 months to 2 years).

Abducent nerve palsy, which is commonly involved with invasive sphenoid sinus mucoceles,[10] was found in our Case 4. This also failed to improve after drainage of the mucocele. Headache, a common symptom with most mucoceles, was documented in only one of our patients. Retro-ocular pain, considered to be characteristic of sphenoid sinus mucoceles,[8] was not encountered in Cases 1 and 4. Only one of our patients had a history of rhinomucorrhoea; this is reported to occur in 7% to 44% of cases[1,2] and is usually accompanied by some attenuation of symptoms. Spontaneous drainage of the lesion presenting as "empty mucocele" has also been reported.[10] Most patients, however, do not complain of any nasal symptoms.[7]

The CT signs specific to the diagnosis of a paranasal mucocele have been well described by Perugini et al.[3] On CT scans without contrast, the mucocele appears as a mass of the same density as cerebral parenchyma occupying the sinus structure, often with exophytic development. It has clear-cut margins on the orbital and intracranial sides without signs of infiltration of adjacent anatomical structures. In addition, there is osteolysis of the sinus structures with regular but thickened margins. After enhancement, the lesion can be seen to have a hyperdense and regular rim on the orbital and intracranial side but there is no variation in the density of the intrinsic tissue. The MRI appearance of paranasal sinuses mucoceles has been described in few cases. It varies from homogenous to non-homogenous, being hypointense or hyperintense on T1, weighted images.[1,12]

Echography, which has been used for the diagnosis of a variety of orbital and periorbital lesions,[13] is likely to prove useful in assessing mucoceles in the sino-orbital area. This technique was not available to us, however.

The differential diagnosis of paranasal sinus mucoceles includes a partially cystic hypophyseal adenoma with sphenoidal involvement, schwanno-

ma, chondroma, ethmoidal chondomyosarcoma, lymphoma of the paranasal sinuses, aspergilloma, and retrobulbar cysts. However, none of these diseases will have simultaneously all the mentioned CT signs specific for mucoceles with the exception of some cases of partially cystic hypophyseal adenomas.[3]

The main objective of surgical treatment of a paranasal sinus mucocele is to prevent a relapse.[1,2,6] The traditional method is to evacuate the lesion and radically remove the sinus mucosa. A variety of surgical approaches have been described that give the maximal exposure of the lesion required for this method; these include frontal, frontotemporal, or bifrontal craniotomy; transfrontal sinusotomy; transnasal, sublabial, transseptal sphenoidotomy; rhinoseptal, transpalatal transseptal, transethmoidal intranasal, or transorbital ethmoidectomy; and two-stage procedures.[2,3,5,6,7,8] Three of our patients (Cases 2, 3, and 5) who had an anterior mucocele invading the anterior cranial cavity and orbit underwent frontal craniotomy and radical excision of the mucocele wall.

Recent advances in the surgical management of mucoceles, however, suggest that extirpation of all mucosal lining of the sinus may not be necessary and that the building of a new, patent communication between the lesion and the nasal cavity is associated with good results.[14,15] We decided to perform simple transnasal drainage in the two elderly patients (Cases 1 and 4) who were in too poor condition to withstand major surgery. Case 1 improved following the surgery but Case 4 did not. There were no postoperative complications in any of the cases. At a mean follow-up of 8 months, only Case 4 had a residual collection of mucus in the mucocele. There have been no relapses, a recognised complication occurring in up to 7% of patients[2] when the capsule is not excised or when a patent communication between the lesion and the nasal cavity is not established.

Conclusion

Invasive mucoceles of the paranasal sinuses are a cause of visual impairment in some patients. Early investigation by CT scan and early surgical treatment is recommended before the development of irreversible neurological deficits. The object of treatment in most patients is the relief of symptoms and prevention of relapse. The traditional method has been to evacuate the lesion and radically remove the sinus mucosa. However, drainage alone is increasingly recognized to be an effective method of treatment.

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