The Journal of Laryngology and Otology
May 1981. Vol. **95**. pp. 493–502

Clinical records

The sphenoid sinus mucocoele

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Summary

MUCOCOELES, mucopyocoeles and empyemas of the sphenoid sinus are rare, but potentially dangerous. Their occurrence is accompanied by a multitude of clinical symptoms and signs, which confuse the diagnosis and may lead to its mismanagement. The treatment of this condition is surgical and in its early states essentially simple.

At Groote Schuur Hospital over a 6-year period (1972–1977) 6 patients with mucocoeles, mucopyocoeles or abscesses of the sphenoid sinuses have been seen. Their clinical presentation, radiological findings and surgical treatment are described and discussed. Correctly managed this condition should completely resolve; however, there is a mortality associated with delay or misdiagnosis.

Introduction

The clinical manifestations of sphenoid sinus disease are many and the sphenoid sinus mucocoele may initially present to physician, ophthalmologist, neurologist, or neurosurgeon rather than to the otorhinolaryngologist. Although rare, this condition is of considerable practical importance to the otorhinolaryngologist and to the neurosurgeon.

The sphenoid sinuses at birth are rudimentary. During early childhood pneumatization of the sphenoid body proceeds from before backwards until eventually these sinuses assume a close relationship with important parasellar structures. This development, though, is inconsistent, and the eventual capacity of each sinus varies from 0·1 to 14 ml., with an average volume of 5 ml. They may therefore remain rudimentary or may become grossly enlarged, with direct extension into the greater and lesser sphenoidal wings (Onodi, 1910).

Because of the variable degree of pneumatization, the inaccessibility to direct and easy clinical examination, the associated bizarre symptomatology, and the reluctance to examine this region radiologically and to explore it surgically, sphenoid sinus disease has remained in comparative obscurity.

The sphenoid sinus mucocoele was first described in 1889 by Berg and since then occasional reports have appeared in world medical literature, notably neurosurgical and otorhinolaryngological. Difficulty exists in differentiation between sphenoid sinus mucocoeles, pyocoeles, abscesses and cysts and furthermore, when the posterior ethmoidal sinuses are involved as well, exact definition of the sinus of origin may be impossible. However as the surgical treatment is the same for all of these, differentiation is of little practical importance.

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https://doi.org/10.1017/S0022215100091003 Published online by Cambridge University Press

The aetiology of paranasal sinus mucocoeles is uncertain but most probably one or more of three factors are involved:

obstruction to the sinus ostium by inflamed mucosa;
cystic degeneration of an epithelial mucous gland;
cystic degeneration of an inflammatory polyp.

Pyogenic infection of the retained fluid produces the mucopyocoele.

Sphenoid surgery was introduced by Schäffer in 1885. He first explored this sinus transnasally by blunt instrumentation and the opening was further widened with gouge or punch forceps. Because of the very variable dimensions of the anterior face of the sphenoid body, different surgical approaches to this sinus were then developed.

Initially surgical interest in the sphenoid sinus is unlikely to have been great, but at that time this structure was taking on a new importance since it was becoming recognized as the gateway to the sella turcica and the 'open sesame' to pituitary surgery. Although the first hypophysectomy, performed in 1906 by Sir Victor Horsley, was carried out trans-temporally, this route to the pituitary was beset by so many problems that the sphenoid sinus soon assumed great surgical importance and two approaches to it, developed at that time by Kocher transnasally, and by Hirsch transethmoidally, today remain as standard procedures. By 1917, a multitude of pathways to this sinus and to the pituitary gland beyond it had been explored (Cope, 1917).

The symptoms and signs of sphenoid sinus disease (mucocoele) can be truly bizarre. Proetz in 1948 clarified this to some extent by listing 13 vulnerable neighbouring structures, which may be involved by disease of this sinus.

Clinically the symptoms and signs which together carry such names as orbital apex syndrome, sphenoidal fissure syndrome, orbital inlet syndrome, superior orbital fissure syndrome and anterior cavernous sinus syndrome, are non-specific. The differential diagnosis of sphenoid sinus pathology is large. As this is a clinically difficult structure to examine, reliance on the radiological features is considerable. Indeed, these findings may constitute the only tangible evidence of any of these diagnoses prior to surgery and only astute radiology can differentiate the mucopyocoele from those other neighbouring conditions.

Clinically the sphenoid sinus mucocoele may produce a variety of symptoms depending on the structures involved around the sinus (see Table I). The most consistent symptom is headache which is usually frontal or retro-orbital, but may be diffuse or localized elsewhere in the head. It tends to be worse towards evening,

TABLE I

CLINICAL FEATURES OF SPHENOID SINUS MUCOCOELE

Neurological	Headache
	Facial pain
	Anosmia
	Visual failure
	Ocular palsy (? intermittent)
Orbital	Exophthalmos
Nasal	Rhinorrhoea
	Polyposis
Endocrine	Hypopituitarism

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may radiate to the face or orbit, and may be present for many years before diagnosis becomes a matter of urgency. In a number of instances this isolated symptom has led to a diagnosis of ophthalmoplegic migraine (Pincus and Daroff, 1964).

The next commonest symptom is visual failure and after that ocular palsy. It is these symptoms of poor- or double-vision and their accompanying clinical signs of failing visual acuity and external ophthalmoplegias that alert the patient and his clinician to the seriousness of the problem. With timely treatment full recovery may occur. Ocular involvement is not, however, a prerequisite to diagnosis.

The third group of significant symptoms and signs is nasal and although these are uncommon in reported series careful examination of the nose would probably reveal a greater incidence of positive clinical signs. In this regard and in the absence of frank rhinosinusitis, the finding of pus on the roof of the nasopharynx is of recognized importance. Other nasal signs include polypi, tumours and anosmia. That present or past evidence of nasal disease may be absent has been stressed repeatedly.

In a review of reported cases up to 1970 Nugent *et al.* found 81 patients with sphenoid sinus mucocoele. From these, including 2 of their own, they were able to effectively analyse 63 case reports. Significantly headache was present in 45 patients, visual disturbances in 41, diplopia in 19 and rhinosinusitis in 13. Nasal abnormality, however, in the form of sinusitis, polypi, nasal mass or frank discharge was present in a total of only 29 patients. Three deaths and two instances of meningitis occurred, all following transcranial surgery. In all, 16 patients underwent craniotomy followed by, on the corrected diagnosis, trans-nasal surgery. Forty-five patients underwent primary nasal surgery to establish diagnosis and drainage.

From experience at the Mayo Clinic over an 18-year period, Wyllie *et al.* (1973) reported sphenoid sinus mucocoeles in 13 of 45 patients with isolated sphenoid sinus lesions. Headache had been present in 10 of these 13 patients from 4 months to 23 years, and blurring of vision or diplopia had been present in 8, in one of whom only was it an early sign. Evidence of infection was present in 3 patients. All 13 patients underwent surgical exploration, 12 transnasally and one via an external ethmoidectomy. One patient, who had a preceding craniotomy, and another, who died of a brain abscess, were the only ones not to recover symptomatically. In all 13 patients radiological evidence of gross abnormalities, seen as soft tissue masses, decalcification, erosion, sinus clouding or fluid level, occurred.

Friedmann and Harrison (1970), from their review of the literature, came to the same broad conclusions as Nugent *et al.* (1970) as regards frequency of symptoms and signs but in addition stressed that anosmia occurs in about 25 per cent of patients. On the basis of their own two cases and a search of the literature, they described the syndrome of fluctuating third nerve palsy associated with severe facial pain and pointed out that in most of these patients, the third nerve palsy was characterized by pupil-sparing.

Radiological Features

The characteristic radiological features of sphenoid sinus mucocoele are produced by a benign expanding lesion which causes thinning, expansion and finally erosion of the sinus walls. Plain skull films usually reveal an opacification

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of the posterior ethmoid and sphenoid sinuses, associated with expansion and thinning of its walls and destruction of the septa (Simon and Tingwald, 1955; Friedmann and Harrison, 1970). Occasionally the sinus appears translucent, particularly if there is extensive bone destruction in the presence of fluid accumulation (Bloom, 1965). The floor of the sella turcica is frequently eroded or even destroyed, particularly anteriorly. Isolated erosion of the sellar floor is probably the feature most commonly noted on lateral skull X-rays (Nugent *et al.*, 1970). The sella may be ballooned, with thinning of the dorsum sellae, suggesting an intra-sellar tumour (Simms *et al.*, 1970). This process of pressure erosion may involve the anterior clinoid processes, optic canals, sphenoid fissure and orbital walls. Very infrequently, this destructive erosion of neighbouring structures may involve the clivus and petrous apex (Phelps and Toland, 1969). Mucosal thickening, fluid levels, and bony sclerosis are features which may be less commonly demonstrated. The multiple curvilinear deformities described by Takahashi *et al.* (1973), are no more than the thinned, bowed and partially destroyed sinus walls and septa in the sphenoethmoidal region.

Apart from lateral and submento-vertical views of the skull, special views are required to demonstrate the optic canals, petrous apices and orbital fissures. Tomography almost invariably clinches the diagnosis (Figs. 1, 2 and 3).

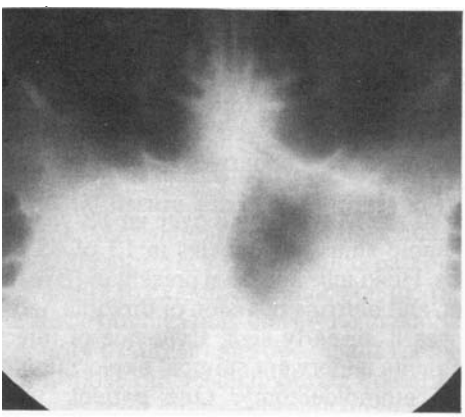


FIG. 1

Base of skull polytomogram showing gross sinus pneumatization, and opacification of the right sphenoid sinus with perisinus sclerosis (patient 2).

Computerized tomography (Fig. 4) has greatly aided the demonstration of masses arising from the sphenoid sinus and extending upwards to involve the sella and neighbouring structures. The use of thin transverse sections in conjunction with computer reconstruction of sagittal and coronal views, has greatly improved the diagnostic accuracy. Orthogonal reconstruction is essential in this region as thin flat bones lying parallel to the X-ray beams cannot be evaluated accurately by the transverse sections only (Marvilla *et al.*, 1978).

Carotid angiography

The need for this will probably fall away except where the intra-sphenoid extension of a carotid aneurysm is suspected, but with a sphenoid sinus mucocoele, the carotid siphon may be displaced laterally and upwards by an

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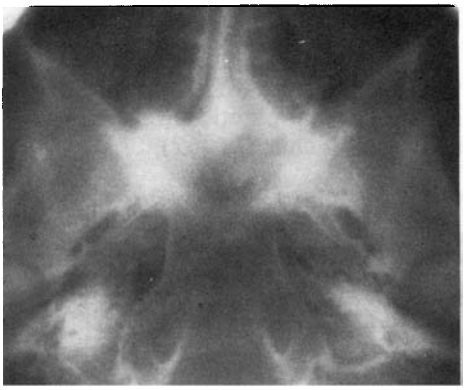


FIG. 2

Base of skull polytomography showing diffuse perisphenoid sinus sclerosis, absence of an intersinus septum and opacification of both sinuses (patient 3).

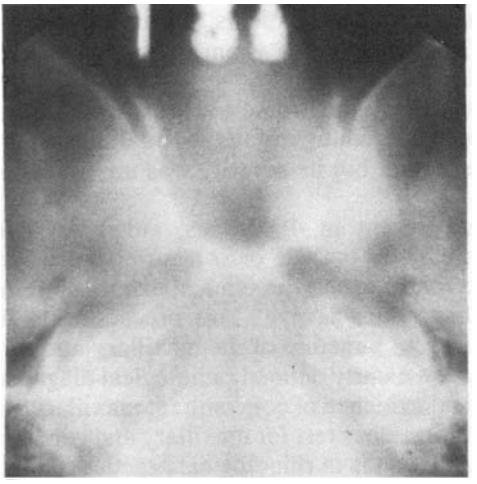


FIG. 3

Base of skull polytomography (see Fig. 4) showing widely expanded sphenoid sinuses with perisinus sclerosis (patient 6).



FIG. 4

CT scan showing a space-occupying mass of the sphenoid sinuses with extension into the posterior ethmoid sinuses, nasal cavity and right orbit (patient 6).

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avascular mass in the sphenoid sinus. This upward displacement of the intra-cavernous portion of the internal carotid artery indicates the presence of an extra-cranial lesion (Bull and Schunk, 1962).

Case Presentation

During a 6-year period (1972–1977) 6 patients with mucocoeles, mucopyocoeles or abscesses of the sphenoid sinuses have been treated at Groote Schuur Hospital. All six were male and their ages ranged from 22 years to 51 years (Table II).

Clinical symptoms and signs (Table II)

Headache was present as the earliest symptom in all six patients and was the only clinical abnormality common to all of them. Prior to diagnosis it had been present for a varying duration, from only 3 months to as long as 8 years. It was described as primarily frontal in 2 patients, as primarily retro-orbital in 2 patients, and as temporal in 2 patients, one of whom also complained of headache over the occiput and vertex.

It was commonly fluctuant, worse towards mid-day and evening and was aggravated by stooping, straining or fatigue. In all these patients it had become progressively more severe, but its severity was unrelated to the extent of other clinical signs.

Rhinorrhea was present in 4 patients, either as a frank mucopurulent discharge identifiable by the patient, or as a purulent discharge found within the anterior or posterior nasal spaces on clinical examination of the nose or in the washings on maxillary sinus lavage. This procedure, in these circumstances preferably termed a proof puncture of the maxillary antrum, was performed on those patients without a clearly defined radiological diagnosis and on those with radiological or clinical evidence of concomitant maxillary sinus infection. It is a useful diagnostic confirmatory test for maxillary and generalized sinusitis, and it is a not infrequent experience in rhinological practice to find on proof puncture, despite apparently normal X-rays, pus in the maxillary sinuses. Perhaps an analogy should be made in this respect with chronic sphenoid sinus infection and apparently normal sphenoid sinus X-rays.

Visual and ocular symptoms and signs formed the major group of tangible clinical abnormalities. Failing vision occurred in three of the six patients. It was bilateral in 2 patients and unilateral in one. One patient with bilateral visual deterioration developed complete loss of vision in one eye. One patient with unilateral fluctuating visual deterioration had no evidence of optic atrophy and he made a perfect recovery after operation. In both patients who had evidence of optic atrophy the vision, despite effective surgical treatment of the primary pathology, remained unchanged.

Two patients had incomplete ophthalmoplegias, which recovered promptly after the surgical treatment of the primary pathology. One of these patients had an intermittent sixth nerve paresis. Proptosis occurred in two other patients. It was unilateral in one patient, whose only other problem was bilateral deterioration of vision, and was bilateral in the other patient who also had bilateral optic atrophy. In both patients the proptosis recovered very slowly over a matter of months. This was in sharp contrast to the extra-ocular muscle pareses, which recovered over the first few post-operative weeks.

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TABLE II

SPHENOID SINUS MUCOCOELE (GROOTE SCHUUR HOSPITAL, 1972–1977)

Patient number	Sex age	Headache	Visual failure	Ocular palsy	Anosmia	Nasal symptoms	Proptosis	Initial diagnosis
1	M 51	Bifrontal	+ (Fluctuating)	0	0	+	0	Pituitary tumour
2	M 22	Retro-orbital	0	Partial 3, 4 and 6 bilateral	?	+	Bilateral R>L	Orbital cellulitis
3	M 22	Retro-orbital	0	+ Intermittent 6th nerve palsy	?	+	0	Sphenoid sinus infection
4	M 37	Temporal	+ (Unilateral optic atrophy)	0	?	+	0	Nasopharyngeal carcinoma
5	M 42	Bitemporal	0	0	0	+	0	Cluster headache
6	M 37	Frontal	+ (Bilateral optic atrophy)	0	0	0	Unilateral R	Pituitary tumour

The pattern of these clinical symptoms and signs closely follows that described by Nugent *et al.* (1970) and by Wyllie *et al.* (1973), except for the higher incidence of nasal and paranasal sepsis found in this series.

Radiological findings (Table III)

The diagnosis of sphenoid sinus disease was initially suspected radiologically in 5 of the six patients. In all 6 patients there was radiological opacity of one or both sphenoid sinuses. Indeed the most significant factor in the pre-operative assessment of these patients was the radiological finding on lateral and on submento-vertical skull views of opaque sphenoid sinuses with or without expansion. These findings were further clarified by polytomography, an investigation that was considered necessary in order to identify and localize bone erosion and tumour extension. It was very much on the basis of the radiological appearances that these sphenoid sinus mucocoeles and mucopyocoeles were suspected and that transnasal sphenoid sinus exploration was correctly decided upon.

In only one patient was one sphenoid sinus opaque, but it was this patient who had the most dramatic clinical signs with bilateral 3rd, 4th and 6th cranial nerve paralyses. The radiological picture in this patient was equally dramatic in that he had gross pneumatization of all cranial air spaces and the air space of his very large sphenoid sinuses extended out into the greater sphenoidal wings.

The radiological diagnosis in these patients was further facilitated by demonstration of bulging of the sinus walls and erosion of the parasellar bony architecture. Sclerosis of the sinus walls denoted a mucopyocoele in each of the 3 patients in whom it was found. Definite erosion occurred in 4 patients. In one it was accompanied by right-sided proptosis and in another by left optic nerve atrophy.

Carotid angiography has, in our experience, offered little in the way of positive diagnostic information and only served to exclude the presence of a vascular tumour or aneurysm in the sphenoid region.

Computerized tomography was only performed on two of our patients. In one the diagnosis of sphenoid sinus disease was not suspected and this region therefore was not adequately examined. In the other patient, however, the mass in the sinus, expansion of the sinus, destruction of sinus walls and encroachment onto neighbouring structures were clearly demonstrated. From experience with other lesions in this region there is little doubt that computerized tomography with the aid of orthogonal reconstruction should prove the examination of choice in the patient with disease processes arising from the sphenoid sinus.

Surgical management

Surgery was performed under general anaesthesia by the intranasal submucous trans-septal approach. A routine submucoperichondrial resection of a segment of septal cartilage and a subperiosteal resection of part of the vomerine bone was performed. The posterior limit of this resection was then taken to the anterior face of the sphenoid sinus. By avulsing the rostrum of the vomer the sphenoid sinus cavity in some was opened and in others this opening was made with hammer and gouge. Further enlargement of this access to the sinus was made with punch forceps (Sellars *et al.*, 1975).

By this stage, the diagnosis of a mucocoele or sphenoid sinus abscess was

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TABLE III

RADIOLOGICAL FEATURES OF SPHENOID SINUS MUCOCOELE (GROOTE SCHUUR HOSPITAL, 1972–1977)

Patient number	Sphenoid sinus							Sella						Carotid angiography		CT scanning		
	Opacification	Hyperlucency	Bulging walls	Walls thinned	Walls destroyed	Septa destroyed	Other sinuses opacified	Destruction of floor	Ballooning	Sclerosis of floor	Sphenoid sclerosis	Optic canal erosion	Orbital fissure erosion	Lateral displacement of cav. carotid	Upward displacement of siphon	Bone erosion	Soft tissue mass	Contrast enhancement
1	+	0	+	+	+	+	0	+	0	0	0	+(R)	0	0	0	+	Not done	0
2	(R+2½)	+	+	+	0	0	0	0	0	0	+(R)	0	0	Not done			Not done	
3	+	0	0	0	0	+	0	0	0	+	+	0	0	Not done			Not done	
4	+	0	+	+	+	+	+	0	0	+	+	+(R)	+	0	0		Not done	
5	+	0	0	0	0	0	0	0	0	0	0	0	0	Not done		Sphenoid sinus region not examined		
6	+	0	0	+	+	+	+	+	+	0	+	0	0	0	0	+	+	0

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obvious to the naked eye. The contents were simply evacuated and in order to maintain a permanently enlarged ostium between sphenoid sinus and nasal cavity a segment of the nasal mucoperiostium overlying the anterior face of the sphenoid on one side only was removed. A ribbon gauze dressing impregnated with antibiotic ointment was packed into the sphenoid sinus cavity via the nares on that side and a counter-pressure dressing placed into the opposite anterior nasal space. These packs were removed the following day. The post-operative recovery was prompt and uncomplicated in all 6 patients.

Results

Following surgery headache in all patients settled promptly and the onset of recovery of visual defects was noticeable within a week. Except for the two patients with pre-operatively diagnosed optic atrophy the visual symptoms and signs recovered to normal. One patient 8 months later experienced recurrence of headaches and it was evident on examination of the nose that the surgically created ostium had closed. This was re-opened under general anaesthesia with subsequent resolution of the headaches.

Comment

This presentation attempts to highlight the importance of the accurate clinical and pre-operative diagnosis of this surgically treatable condition. It further highlights the considerable reliance that must be placed upon the radiology of this area. It is as 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. When ocular symptoms and signs occur there comes an urgent need to consider this diagnosis since to ignore or to misdiagnose the sphenoid sinus mucopyocoele, rare as it may seem, will lead to distressing consequences which are wholly and so very simply avoidable.

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