Sphenoid Sinus Mucoceles

G. Robert Nugent, M.D., Philip Sprinkle, M.D., and Byron M. Bloor, M.D.
Division of Neurological Surgery, and Division of Otolaryngological Surgery,
West Virginia University Medical Center, Morgantown, West Virginia

The neurosurgeon is familiar with the frontal sinus mucocele as a common cause of unilateral exophthalmos and disorders of eye movement, but the less common sphenoid sinus mucocele may have escaped his experience. These lesions are potentially more serious, and are often misdiagnosed and operated on as pituitary tumors. A mucocele of the sphenoid sinus was first described in 1889 by Berg. Since then there have been sporadic reports, primarily in the European literature, the best being that of Lundgren and Olin. It is the purpose of this paper to review the world literature, add two additional cases, and alert the neurosurgeon to a lesion easily mishandled.

Data on 63 cases have been analyzed as the basis of this report. Nineteen other cases have been mentioned but are not included in the analyses because the clinical information was too sparse to be useful. Three cases were not translated. Nine cases are used in this analysis but are not identified in the text.

If the cyst contents are purulent, pyocele might be the more proper term. Frequently, however, the sphenoid sinus mucocele contains a creamy substance which has the appearance of a purulent exudate but is sterile. One case called a pyocele and two others called abscesses all failed to grow pathogens when cultured. Because it is difficult to determine what is a true pyocele and what is a mucocele, we have included the "pyoceles" in this series. We have also included as mucoceles the lesions termed sphenoid "cysts." These lesions frequently involve the posterior ethmoidal cells and have been called spheno-ethmoidal mucoceles, but since the symptoms are related primarily to the sphenoid area, and the posterior ethmoids are only involved secondarily, we prefer the term "sphenoid sinus mucoceles."

The etiology of a mucocele is conjectural. It would be convenient to consider them as simple retention cysts of the sinus resulting from inflammatory blockage of the draining ostium. However, they do occur when the ostium is patent. Nor does a blocked ostium always result in mucoceles. Others have considered them as originating from cystic dilatation of the mucus glands of the epithelial lining of the sinus, or from the cystic degeneration of contained polyps. Schüller has raised the possibility of the development of a hypophyseal cyst from cell rests in the inner or under half of the sella turcica, but evidence for this is lacking. The location of the lesion and the finding of cholesterol crystals in the cyst fluid in 10 cases raises the question of whether these cysts may represent an unusual form of craniopharyngioma. However, cholesterol crystals may be a non-specific finding, and the clinical and pathologic features are not those of a craniopharyngioma.

Whatever the cause, there is no exit for the cyst contents, and the accumulating secretions thin and expand the cavity, ultimately causing compression of surrounding structures. In the case of the sphenoid sinus, the adjacent non-bony structures include the first six cranial nerves, the carotid arteries, the cavernous sinus, and the pituitary gland. It is the involvement of the cranial nerves which brings the patient to the physician. In many individuals, there is only a thin papyraceous partition of bone separating the sinus from the optic nerve, cavernous sinus, tuberculum sella, and medial wall of the superior orbital fissure and orbit. Examination of a disarticulated skull (Fig. 1) shows how easily expansion of the sphenoid sinus could encroach upon these structures.

Case Reports

Case 1. This 33-year-old man was admitted on March 20, 1961, because of right ptosis, severe headache, and limitation of inward gaze with the right eye. He had had in-

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termittent frontal headaches and periorbital pain for 11 years, associated with nasal stuffiness. During a severe headache 8 years before admission he suddenly became blind in the left eye. Operation on the maxillary sinuses improved the headache. A year later the diagnosis of glaucoma was made. A year before admission he began having intermittent visual disturbances in the right eye.

Examination. There was optic atrophy and no light reflex in the blind left eye. Visual acuity in the right eye was 20/70. There was extensive glaucomatous excavation of the left disc, and slight right ptosis. Both eyes were prominent but this was greater on the left. There was no adduction of the left eye. He had a normal sense of smell. Skull x-ray films showed an enlarged sella turcica with erosion of the floor. The posterior clinoids were preserved but the left anterior clinoid was blunted. There appeared to be a pharyngeal mass (Fig. 2). It was felt that the patient had a pituitary tumor, and on April 3 a left carotid arteriogram showed narrowing of the carotid siphon and slight elevation of the first portion of the anterior cerebral artery. The next day his headache was worse, and vision in his right eye suddenly diminished to light perception only.

He was unable to adduct the eye. The diagnosis of pituitary apoplexy was made.

Operation. A left frontal craniotomy was performed. Upon exposing the left optic nerve, a tense midline, dome-shaped mass was seen in the region of the jugum sphenoidale, elevating the dura and causing destruction of a large area of bone in the midportion of the posterior frontal fossa. No fluid was obtained on aspiration of the mass. Incision of the mass released a brown mucoid substance. Aspiration disclosed a glistening walled chamber 6 × 4 cm in the region of the sphenoid sinus. The diagnosis of sphenoid sinus mucocele was made and the linear dural incision closed with interrupted silk.

The cyst contents were described pathologically as eosinophilic mucous-like material containing filamentous structures, red blood cells, and neutrophils. Routine, fungus, and acid-fast cultures of the cyst fluid all failed to grow organisms.

Postoperative course. The next day the patient’s headache was gone, and he could count fingers with his right eye. The visual field on the right was full to gross confronta-

![Fig. 1. Photograph of interior of sphenoid sinus of normal disarticulated skull. The arrow identifies the optic nerve coursing through the upper lateral sinus covered by only a thin partition of bone. Below the nerve the lateral wall of the sinus forms the medial wall of the superior orbital fissure.](image1)

![Fig. 2. Case 1. Lateral view of skull showing erosion of floor of sella turcica, thinning of dorsum sella and posterior pharyngeal mass.](image2)
tion. A complete postoperative endocrine evaluation showed no evidence of pituitary insufficiency. One week after surgery vision in the right eye was 20/50. The extraocular movements and peripheral fields were normal. X-ray examination 3 weeks after operation revealed no evidence of chronic frontal or maxillary sinus disease. The ethmoids were considered normal. There were prominent nasal turbinates on the right.

Approximately 7 weeks after surgery, through an intranasal approach, an opening 1 cm in diameter was made in the anterior wall of the sphenoid sinus. He has been well ever since.

Case 2. This 54-year-old man was admitted because of right frontal headache for 1 year and double vision on right lateral gaze. He also complained of nasal drainage and conjunctivitis, and loss of libido. He had been operated on 4 years before admission for right frontal sinusitis.

Examination. There was decreased pinprick sensation in the distribution of the right supraorbital nerve, and evidence of a right sixth nerve paralysis. The 17 keto and hydrocorticoids, protein-bound iodide and I\textsuperscript{131} thyroid uptake tests were all normal. Cerebrospinal fluid cell count and protein were normal. Sinus roentgenograms showed pansinusitis, and plain skull films disclosed loss of the floor of the sella turcica and a density in the underlying sphenoid sinus (Fig. 3). Tomograms of the skull, however, disclosed that the erosion of floor of sella turcica was from below rather than from above (Fig. 4). This led to the diagnosis of sphenoid sinus mucocele. Brain scan, electroencephalogram, right retrobrachial arteriogram, and pneumoencephalogram were all normal.

Operation. One month later the sphenoid sinus was marsupialized through a Ferris-Smith incision and anterior ethmoidectomy.

Postoperative course. The patient did well, with complete resolution of the eye findings except for diplopia on downward gaze. Skull films 10 months after operation demonstrated reconstitution of the floor of the sella turcica and an unexplained dense opacification of the sphenoid sinus (Fig. 5).

Discussion

Clinical Features. Some of the cases reviewed were from the older literature and consist of brief case reports without radiographic examination. In other instances the clinical information was so sparse that no attempt could be made to analyze the accumulated clinical data statistically. There was an almost equal distribution between men and women and no age preponderance. The spread in age was from 13 to 75 years with an average age of 42.

Headache. The most common complaints were headaches and visual disturbances. The headache was usually severe and localized to the frontal region (18 cases) or to the region of an eye (27 cases). Frequently there was radiation to the temporal or occipital region. The headache may occur as a result of the upward stretching of the dura in the region of the planum sphenoidale and floor of the frontal fossa. It has been stated that pain originating in the sphenoid sinus may radiate to the vertex but only three patients complained of vertex pain. In only two cases was it specifically stated that headache was not a feature of the illness. The headache was described as intermittent in only 15
cases, but in many others the chronic but fluctuating severe headache was an outstanding feature of the clinical course. In some cases, the headache and other symptoms were relieved coincident with spontaneous drainage from the sinuses. In only four cases was there nausea and vomiting with the headache.

Visual Symptoms. In 41 patients vision in at least one eye was impaired to some degree. Fifteen were blind in one eye (three of these had light perception), and five others were bilaterally blind. Two others could only count fingers with one eye, and in another two cases the vision was reduced to 20/200 in each eye. In 19 cases there was optic atrophy; in three of these it was bilateral. Four patients, including our first case, were blind in one eye but completely recovered their vision following operation.\(^7,14,26\) One patient with 2/200 vision in the right eye and 20/200 vision in the left showed a "gross improvement" in vision the day after surgery,\(^9\) and another patient blind in the left eye had improvement in visual acuity to 8/10 the day after surgery.\(^31\)

In 19 cases there was diplopia. The available information indicated that in 12 patients this appeared to be due to third cranial nerve compression and in seven to sixth nerve involvement.

In 12 cases there was a central scotoma (two of these were bilateral), and in 14 a peripheral field defect. There were no cases of bitemporal hemianopsia, but two patients blind in one eye had a temporal hemianopsia in the other eye.

Exophthalmos was present in 20 patients. In four cases it was bilateral and in one it was pulsating.\(^49\)
Associated Symptoms. In 29 cases there was a history of previous otolaryngological disease in the form of sinusitis, intranasal polyps, nasal mass, or nasal discharge. Thirteen cases had sinusitis, 6 intranasal polyps, 18 a nasal mass, and 15 a nasal discharge. Seven of the 13 cases of sinusitis were longstanding, and six had previous otolaryngological surgical procedures on either the sinuses or nasal passages. This rather high incidence of nasal symptoms was contrary to another recent report in which it was stated that "there is an almost complete lack of nasal symptoms."24 Nine patients had anosmia.

Evaluation of pituitary function was carried out in four patients, and in none was there evidence of pituitary insufficiency.21,65,68 Two cases, however, described loss of libido, one of which recovered after surgery; one woman had amenorrhea, fatigue, and drowsiness.

Radiographic Aspects. The diagnosis in most instances rests on the radiographic features. Incomplete radiographic study or superficial examination of the films, however, may lead to the erroneous diagnosis of pituitary tumor and establish a trend of thought leading to an ill-advised intracranial operation. Simon and Tingwald46 have summarized the radiographic findings and suggest that the anatomical, clinical, and radiographic features justify the term "orbital inlet syndrome." They point out, and we agree, that the radiographic examination should include satisfactory demonstration of the superior orbital fissures, optic foramina, lateral walls of the ethmoid sinuses, floor of the sella turcica, walls of the sphenoid sinuses, lesser wings of the sphenoids, and walls of the orbits. Anteroposterior and lateral tomography is of greatest importance in examining these areas and should avert misdiagnosis.29,60,66

The most salient radiographic finding was involvement of the sella turcica. This structure was involved in 38 cases, and in 29 instances the floor of the sella was eroded. When this erosion involved the major portion of the floor it was at times difficult to determine whether the mass arose within the sella or below it. In 11 cases there was erosion of an optic canal and in nine cases the medial wall of the orbit was involved. In seven cases the tuberculum sella or planum sphenoidale was destroyed. In only one instance was there x-ray evidence of suprasellar extension of the mass in the plain films.6

Pneumoencephalography or ventriculography was performed in 14 cases. Nine of these studies were normal, three showed changes about the chiasmatic cistern,10,21,76 and two indicated a suprasellar mass.6,53

Arteriography was normal in 10 of 20 studies. In 10 instances the carotid siphon was uncoiled, elevated, or displaced laterally.

Only one patient had a brain scan, and this was normal.

Pathological Study of the Cyst and Its Contents. In 28 cases there was evidence of an inflammatory reaction either on the basis of the histological examination of the cyst wall, the finding of purulent exudate in the cyst, or the culture of organisms from the cyst contents. Of 18 cultures obtained, 12 were sterile. Twice staphylococcus aureus was isolated,10,40 and twice both staphylococcus and streptococcus were cultured.25,52 In one case the culture grew an enterococcus42 and in another a fungus with spores.45

It is of interest that in 10 cases, cholesterol crystals were noted in the cyst fluid. The clinical and pathological evidence indicates that these were not craniopharyngioma cysts and indicates that the cholesterol crystals were probably part of a non-specific reaction to chronically encysted fluid.

The color of the cyst contents was variably described as yellow, greenish-yellow, greenish-brown, dirty green, brown, yellowish-gray, whitish-yellow, ochre, chocolate, milk-chocolate, chocolate-milk, blackish, and straw-colored. The consistency also varied: gummy, rubbery, stringy, creamy, mucoid, viscous, greasy, tenacious, honey-like, glary, and watery.

The normal sphenoid sinus is lined with ciliated columnar epithelium. In two cysts the wall contained cuboidal cells,5,8 in four others columnar epithelium was described,18,24,46,61 in two of these cases the epithelium was ciliated. Five showed respiratory epithelium,21,45,48,55,68 and in five others mucous glands were noted.2,3,39,61 Squamous metaplasia was present in three cases.

The variable pathological features of these
cysts may be related to the presence or absence of infection or spontaneous drainage, and to the length of the development process.

**Presenting Diagnosis.** This may be a deceptive lesion. In 15 instances an intracranial lesion was suspected, usually a pituitary tumor (10 cases), but other diagnoses included chordoma, meningioma, and aneurysm. In 13 cases the diagnosis was correctly made preoperatively, but in seven other cases a neoplasm of the sinus was considered.

**Treatment and Results.** In 45 cases the primary approach was an otolaryngological one. Various approaches were used but all were aimed at establishing adequate nasopharyngeal drainage of the cyst by creating a substantial opening in its anterior wall. In almost all of the cases so approached, the procedure was curative although lasting and preexisting blindness was unchanged. In 16 cases, neurosurgeons assumed primary responsibility, and craniotomy was undertaken in an attempt to treat a presumed intracranial lesion. In three of these 16 cases, death resulted from infectious complications of the operation and in two others meningitis ensued but responded to therapy. Not infrequently the craniotomy was followed by a more definitive and curative endonasal approach by the otolaryngologist.

The headache, double vision, exophthalmos, and nasal symptoms usually responded well to adequate drainage of the cyst, but fixed blindness was ordinarily permanent.

**Differential Diagnosis.** Of greatest importance is the exclusion of a pituitary tumor to prevent the misdirection of therapy. Most difficulty is encountered if the physician accepts the initial radiographic findings without special radiographic examination (including tomography), or if he ignores the clinical features, which are usually different from those of a pituitary tumor. The course of the mucocele is apt to be chronic but remittent with the sudden appearance of visual loss, diplopia, and headache while the typical bitemporal hemianopsia and pituitary insufficiency of the pituitary chromophobe adenoma are not present. The sphenoid mucocele is less likely to cause ballooning of the sella turcica or show evidence of suprasellar extension. Tomography, as in our second case, may reveal that the erosion of the floor of the sella is from below rather than from above. The absence of intracranial calcification on tomograms would also rule out craniofaryngioma.

A malignant tumor in the region of the sphenoid sinus pursues a more progressive course, and the roentgenograms show evidence of invasive destruction of bone rather than the pressure erosion seen with the sphenoid mucocele.

An intracranial chordoma grows more posteriorly and is usually calcified. There has been some confusion with sphenoid ridge meningiomas but although the visual symptoms may be similar, the sphenoid ridge meningioma does not ordinarily invade the sphenoid sinus and region of the sella turcica. Other lesions to be considered are: aneurysm, orbital dermoid and epidermoid tumors, orbital meningocele or encephalocele, chronic sphenoiditis, optic nerve glioma, and neurofibromatosis with unilateral involvement of the orbit, sphenoid ridge, and optic foramen.

Also to be considered is the ill-defined entity of ophthalmoplegic migraine which may cause unilateral headache and eye pain followed by third cranial nerve paresis. This is usually seen in younger patients with a shorter history, and nausea and vomiting is more prominent. As has been pointed out by others, however, the diagnosis of sphenoid sinus mucocele should be considered with every case of so-called “ophthalmoplegic migraine.”

It is of historical interest that earlier in this century exploration of the sphenoid sinus, with or without evidence of disease in this area, was considered the treatment of choice for retrobulbar neuritis and other visual disorders. The fact that most patients soon recovered their vision following such surgery was offered as justification for this therapy. Around 1940 it became apparent that most patients with this condition improved spontaneously and the weight of opinion turned against this operative approach. Three of the cases reported here point out, however, that the sphenoid sinus mucocele is one lesion of the sphenoid sinus that may indeed cause symptoms similar to those of retrobulbar neuritis.
Summary

Mucoceles of the sphenoid sinus and posterior ethmoidal cells are really otolaryngological diseases, but the lesions are frequently misdiagnosed as pituitary tumors and, because of the sudden and alarming loss of vision, intracranial surgery may be performed by an unwary neurosurgeon.

The world literature has been reviewed and two cases added, bringing the total cases reported to 81; 63 of these are used as the basis of this report. The clinical features form a rather consistent pattern that includes: severe, and often intermittent, frontal and orbital pain with visual loss in one eye, diplopia, exophthalmos, blindness, and preexisting sinusitis with nasal discharge.

The radiological examination frequently reveals destruction of the floor of the sella turcica, changes in the clinoids (often unilateral), erosion of an optic canal, and destruction of the jugum sphenoidale. The sphenoidal fissure and medial wall of the orbit may also be involved. Tomograms are most helpful in demonstrating these changes.

Those patients first seen by an otolaryngologist are usually properly treated by a transnasal approach. The only complications (three deaths and two cases of meningitis) occurred in the patients who had neurosurgical craniotomies.

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