Lesions of the paranasal sinuses often have to be considered in the differential diagnosis of sellar disease. The sphenoid sinus in particular may be a source of diagnostic confusion. Of the various lesions to which this sinus falls prey, one of the rarest is mucocele or pyocele. The following case report illustrates several unusual features of importance to the neurosurgeon in the diagnosis and management of such lesions.

**Case Report**

No4-59-2522. J.W., a 30-year-old white male, began to suffer episodic visual disturbance in 1955. This began as a sudden loss of vision in his left eye which cleared completely within 3 weeks. No further symptoms occurred until 1960 when there was a sudden decrease in vision in both eyes accompanied by headaches. This was described as bifrontal pain of a continuous nature, enhanced and often precipitated by lying down and relieved by standing up. A diagnosis of retrobulbar neuritis was made, and a course of systemic steroids was given. Roentgenograms of the skull at this time were reported as normal as were studies of the cerebrospinal fluid. The visual acuity improved in the right eye to 20/50 within 6 weeks but remained at counting fingers in the left eye. It was noted for the first time that he had bilateral corneal dystrophy, worse on the left side.

In early 1962, he was aware of the complete loss of vision in the nasol field of the left eye, and only motion of the hand was perceived in the rest of the field; vision was unchanged in the right eye. In June 1962, a constant left-sided headache developed in the parietal region; this too was relieved by the erect posture. October 1962 roentgenograms of the skull revealed erosion in the floor of the pituitary fossa. A pneumoencephalogram and bilateral carotid angiograms were performed and a presumptive diagnosis of pituitary tumor was made. On Oct. 25, 1962 a right frontal craniotomy revealed extensive frontal cerebral edema and the sellar region could not be explored. Because of the edema the bone flap was left out. A second exploration at the same site on Nov. 6, 1962 was also unsuccessful for the same reasons.

In November 1962 the patient was referred to the Branch of Ophthalmology of the National Institute of Neurological Diseases and Blindness, for treatment of the corneal dystrophy, and further neurosurgical evaluation was requested from the Branch of Surgical Neurology, where he was seen for the first time on Nov. 20, 1962.

**Examination.** He was a well-looking individual with a healed coronal incision of the scalp and absence of bone in the right frontal area. The remaining findings were entirely within normal limits with the exception of the ocular findings which were compatible with corneal dystrophy and were summarized briefly as follows: dystrophic changes in both corneas, mainly in the left; visual acuity 20/60 and J2 in the right eye and light perception only in the left. Fundi were normal and visual fields were full bilaterally.

All blood studies and urinalysis were within normal limits. Roentgenograms of the skull on Dec. 7, 1962 revealed a mass of soft tissue involving the left nasopharynx and sphenoid sinus with destruction of the floor of the pituitary fossa (Figs. 1 and 2). A large surgical defect was present in the right frontal region. Views of the optic foramina showed bony destruction of the left optic foramen with only the inferior margin remaining intact. Tomograms of the sella turcica and adjacent sphenoid bone showed destruction of the air cells of the ethmoid, sphenoid sinus, and medial portion of the lesser wing of the sphenoid bone on the left. This opaque area of erosion extended into the nasopharynx through the sphenoid bone adjacent to the pterygoid process (Fig. 3).
Fig. 2. Lateral roentgenogram of skull showing the area of sphenoidal opacity and destruction of sellar floor.

The pneumoencephalogram and carotid angiograms done in October 1962 were reviewed and our interpretation excluded any intracranial extension of the lesion. Diagnosis at this stage was “a nasopharyngeal destructive mass involving the sphenoid sinus and extending into the floor of the pituitary fossa; probably a nasopharyngeal carcinoma.”

Operations. On Dec. 13, 1962 the patient underwent nasopharyngoscopy with biopsy under topical analgesia. Fullness in the upper anterolateral aspect of the nasopharynx on the left side was the only abnormality found. Multiple biopsies in this area were negative for tumor.

On Jan. 9, 1963, under local analgesia, the middle turbinate of the left nostril was removed surgically and entrance into the left sphenoid sinus was then obtained with the use of a punch. Polaroid roentgenograms were taken during the procedure to locate the exact position of the biopsy instrument (Fig. 4). As the left sphenoid sinus was entered, about 30 cc. of yellow-green pus escaped. A considerable amount of remaining pus was removed with a suction catheter and saline irrigations of the cavity. The patient tolerated the procedure well.

Histological examination of tissue taken from the wall of the cavity showed only necrotic epithelium of the sphenoid sinus, and cultures of the pus and this tissue grew only nasal commensals without any pathogens.

Course. The patient had complete relief of his headaches for the first 5 days after operation and then began experiencing headaches again. Repeated roentgenograms showed a level of fluid in the left sphenoid sinus and further irrigations were carried out (Fig. 5). The headaches were relieved with these irrigations which were performed twice a week for a few weeks and then for a few occasions, as indicated by repeated roentgenograms of the sinus. The last irrigation was done in March 1968. The patient has remained symptom-free since that time. The bone flap was replaced in August 1968.

Repeated films of the skull with views of the optic foramen and laminography were done on Jan. 17, 1964. These showed reconstitution of bony margins adjacent to the sphenoid sinus without evidence of recurrent sphenoidal disease or other lesions in the nasopharynx (Figs. 6 and 7). The patient is now well and has had no recurrence of headache, or other symptoms.

The visual disturbance caused by the corneal dystrophy is under control and has shown no progression.

Discussion

Mucocele of the sphenoid sinus was first described by Berg in 1889. In earlier reviews of pyocele-mucoceles of the paranasal sinuses, it was made clear that either mucocele or pyocele could produce an identical clinical picture; even the radiological appearance may be similar. Mucoceles of the frontal sinuses, which are the commonest, are caused by obstruction resulting

Fig. 3. Lateral tomogram through sellar region confirming observations made in Fig. 2.

Fig. 4. Lateral Polaroid roentgen-ray view of patient's head taken at the time of drainage of the sphenoidal pyocele.
from fracture, inflammatory conditions such as chronic sinusitis, or tumors, especially osteoma. In the sphenoid sinus the cause of obstruction is usually chronic sinusitis since no case caused by fracture or osteoma has been described as yet.\textsuperscript{20} Retention of the secretion in the sinus results in a dilated cavity filled with a thick dark glairy fluid which on occasion may be thin and rather yellow. If infection supervenes, the contents become pus, which is best described as a pyocele.

The importance of the anatomical relations of the sphenoid sinus in the varied and often obscure clinical syndrome produced by mucocele-pyocele has been stressed in a number of articles pri-
migraine. 

Incidence and History. There is no particular preponderance for age or sex. The history is characteristic episodic with periods of remission. Headaches and visual disturbances form the most common presenting symptoms, and often headaches constitute the only complaint. An important feature of headache related to mucocele-pyoecele of the paranasal sinuses, particularly in the sphenoid sinus, was demonstrated by our patient. This, in the personal experience of one of the authors (J.M.), is the relation of headache to the supine posture. The duration of symptoms may be quite long, in some cases as much as 20 years before the diagnosis is established, and symptom-free periods are common. The etiology of the headaches is uncertain but most probably related to expansion of the sinus. The possibility of associated cerebral edema as a cause of headache comes to mind, but there is no clear evidence for this. There is an almost complete lack of nasal symptoms.

Clinical Signs. In the early stages there may be none at all. Later they are usually related to the eye, when visual disturbances caused by compression of the optic nerves, paresis of the ocular muscle, and exophthalmos may be seen. Purulent nasal discharge is very rare and usually occurs only when a palpable mass is bulging into the nasopharynx. The exophthalmos may be pulsating though usually is not. One case of ophthalmic migraine caused by sphenoidal mucocele has also been reported, in which an aneurysm was excluded by angiography. Nasal signs are usually slight although in about a quarter of the cases disturbance or absence of smell is present. Occasionally, polyposis secondary to chronic sinusitis is seen. An interesting point is that the signs and symptoms are usually unilateral.

Intracranial Involvement. Spontaneous intracranial involvement, other than effects related to the optic nerves and the orbital apex, is quite unusual. Usually intracranial complications follow a craniotomy, and include meningitis, which is often fatal, cerebrospinal-fluid rhinorrhea, and rarely, hydrocephalus. Endocrinological disorders, probably related to pituitary compression, are not common, and only 2 cases have been reported.

Diagnosis. In most cases the diagnosis has been made at operation, as in our case, unless a visible bulging of the mucocele into the nasopharynx is seen. However, the diagnosis of mucocele-pyoecele of the sphenoid sinus would be made more often if careful attention is paid to the radiological features. These may be summarized as follows: opacification of the sinus and expansion in size, rarefaction of the surrounding bone and destruction of the septae. These erosive changes usually are well outlined. Quite often, however, these changes are unilateral or asymmetrical. The effects of expansion are also seen in the sphenoidal fissures and optic canals producing widening and irregularity. Rarely, erosion of the clinoids or upward and lateral displacement of an anterior clinoid or lateral displacement of the lamina papyracea is seen. This lamina may be partially eroded and in some projections may suggest a separate calcified structure, e.g. the wall of an aneurysm. Radiological evaluation must include an adequate demonstration of all the structures mentioned above and laminography also is a useful adjunct. Cerebral angiography and pneumoencephalography usually do not add anything of diagnostic significance except to exclude any intracranial extension of the lesion.

Differential Diagnosis. Tumors are the most common source of error. Hypophysial neoplasia, e.g. chromophobe adenoma, probably comes first with nasopharyngeal carcinoma a close second. Other tumors which have to be excluded are craniopharyngioma and meningioma. Chordomas, cholesteatomas, tumors of the sphenoid and ethmoid sinuses and of the nasopharynx and base of skull, such as juvenile angiofibroma, myeloma, and benign polyps, are quite rare. A rare condition is that of pneumosinus dilatans. This is commonly an asymptomatic finding of an expanded sinus filled with air. It has been described by a number of writers as being capable of producing symptoms which may mimic a mucocele and even produce optic atrophy, presumably by pressure on the optic nerve. Because a pneumosinus dilatans is always radio- translucent, the diagnosis is easily made by roentgenography.

Management. There is little doubt that the mucocele-pyoecele of the sphenoid or adjacent sinuses should be treated by endonasal surgical drainage and never by craniotomy. Since this is also the approach of choice for tumors of the sphenoid bone and adjacent regions (excluding true sellar lesions) the endonasal approach should always be used when a primarily sphenoidal or posterior ethmoidal lesion is suspected. The risk of dangerous complications which may ensue after a craniotomy is well borne out by the published reports of 4 deaths attributable to such an approach.

The actual technique for endonasal drainage is well described in the literature. The simplest procedure is turbinectomy (middle turbinate) and sphenotomy. Other procedures include transeptal sphenotomy, and ethmoidectomy and sphenotomy. Two cases of patients treated by transmaxillary sphenotomy have also been reported.
The results of an adequate endonasal sphenotony are to establish good drainage of the sphenoidal contents. It is not necessary to exterminate the sinus but repeated washouts may be required as described in our patient. Adequate drainage is quickly witnessed by resolution of all the signs and symptoms, headache being the first to go. If vision has been lost for a long period of time, recovery of sight may not occur; even severe visual impairment for as long as a year, however, can be reversed and good sight restored. It is perhaps not too radical to suggest that patients with signs and symptoms as above should not be subjected to nasopharyngeal biopsy as an initial step. A direct sphenotomy with a cannula or trocar would be readily diagnostic, and it may even be possible to cannulate the ostium of the sphenoid sinus to drain it.

Pathology. The pathogenesis has been described as expansion of a cavity by its contained secretions. Secondary suppuration is usually by nasal commensals although the majority of cultures are sterile. A "fungus" has been reported in one case but was not identified.

Our patient illustrated many of the features of the natural history of mucocele-pyocele described above. In addition, the presence of congeal dystrophy made it difficult to evaluate the visual disturbance. The initial diagnosis of a retrobulbar neuritis recalls the older arguments relating sphenoidal disease to this condition. However, this case does not add conclusive evidence one way or the other.

Subsequently a diagnosis of pituitary neoplasm led to two craniotomies; fortunately the extensive cerebral edema prevented probing of the pyocele via the intracranial route. Such an approach can, as shown in the cases reviewed, lead to a fatal meningitis.

Our own preoperative diagnosis of a nasopharyngeal tumor was also wrong, but it led us to the endonasal surgical approach and a correct operative diagnosis of the lesion.

It is of interest to note that in the follow-up roentgenograms of the skull taken about 1 year after the sphenotomy in our patient, there was clear evidence of reconstitution of the bony floor of the sella. This is evidence for absence of any residual collection within the sphenoid sinus. The phenomenon of reconstitution of the floor of the sella has been described by other writers, e.g., following transsphenoidal hypophysectomy.

Summary

A case of supplicative mucocele or pyocele of the sphenoid sinus treated successfully by endonasal drainage is described. The literature is reviewed and the diagnostic difficulties are stressed. Such difficulties often lead to exploratory craniotomy for supposed sellar lesions which may produce a fatal meningitis. For this reason the consensus of opinion is supported that such lesions should always be approached via an endonasal sphenotomy and never by craniotomy. The natural history and radiological features of mucocele-pyocele of the sphenoid sinus are described to facilitate early recognition and correct management.

References

1. BENDESCU, T. Cited by Zejzgabel.