MUCOCELE OF THE SPHENOID SINUS*†‡ A BENIGN IMITATOR

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IN recent years, considerable interest has developed in the phenomenon of sphenoid sinus mucocele (Lundgren and Olin, 1961). Important factors are the critical location of this sinus and the various vital structures which may be involved by an essentially benign pathological process originating within its boundaries. We have recently treated a patient in whom the pre-operative diagnosis was that of a malignant parasellar intracranial process, and who has shown good response to simple drainage of the surgically diagnosed mucocele.

Case Report

A 52-year-old housewife entered the hospital on March 8, 1965, because of headaches and poor vision of 6 months' duration. The patient had apparently been in good health until 6 to 12 months before admission, when severe right-sided headaches and recurrent attacks of vertigo were noted. At the same time, she noted progressive diminution in the vision of both eyes, especially the right. After some attempts at self-medication, the progression of the visual loss caused her to seek medical advice.

Her past medical history included a congenital hypoplasia of the left arm and a hysterectomy 15 years previously. She had had frequent attacks of otitis media on the left. Familial and social histories were non-contributory.

Examination.—She appeared as an obese woman in no distress. Temperature, pulse, respiration, and blood pressure were within normal limits.

There were bilateral large central scotomata with marked loss of visual acuity (Fig. 1, opposite). Best corrected visual acuity was 20/200 in the left eye and 2/200 in the right. Severe pallor of the right optic disc and moderate pallor of the left were noted.

The left ear showed a profound sensorineural hearing loss. The external auditory canal ended in a blind pouch about 1 cm. deep. Intranasal examination revealed oedematous nasal mucosa, a thickened and markedly deviated nasal septum, and a small amount of thick yellow discharge. Indirect nasopharyngoscopy was negative.

On neurological examination, the only additional abnormality noted was the spastic, hypoplastic left arm. Detailed examination of the first, fifth, seventh, ninth, tenth, eleventh, and twelfth cranial nerves gave normal results.

X-ray studies and laminograms of the skull revealed destruction of the floor of the sella turcica, and a suggestion of a mass in the sphenoid sinus with displacement of its anterior wall. Clouding of the ethmoid air cells and thinning of the nasal wall of the left orbit were noted. A left carotid arteriogram showed some questionable displacement laterally of the internal carotid artery in the

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FIG. 1.—Visual fields at time of admission (Ajmark perimeter above; tangent screen below), showing bilateral central scotomata and marked visual impairment, especially in the right eye.

cavernous sinus, but this was felt to be within normal limits. There was no evidence of intracranial aneurysm. At this point, the provisional diagnosis was that of an extensive neoplastic process, probably originating in the sphenoid sinus or pituitary gland, and eroding bone.

Laboratory studies which were normal or negative included haemogram, urine analysis, blood urea nitrogen, fasting blood sugar, protein-bound iodine, overnight urinary concentration (208 milliosmols per litre), morning and afternoon plasma cortisols, urinary 17-keto and 17-hydroxy-steroids, and urinary follicular-stimulating hormone. Electro-encephalography and electrocardiography gave normal results.

Operation.—On March 15, 1965, an intranasal transthemoidal sphenoidotomy was performed. Under general anaesthesia, a submucous resection of the nasal septum and resection of the inferior turbinate were performed to allow adequate visualization. (A transthemoidal approach was chosen although a submucous resection had been performed, in order to avoid any possible malignant seeding of the septal space). Using the Zeiss operating microscope with a 300-mm. objective lens, the anterior and posterior ethmoid cells were removed and the anterior wall of the sphenoid was identified. The osseous sinus wall was thin, and the underlying mucosa was unusually friable. The sinus was filled with an odourless, greenish-brown, viscous fluid. Probing the sinus showed that the interseptum was absent and the roof of the sinus in the region of the optic chiasm and sella turcica was soft and apparently devoid of bone (Fig. 2, overleaf). Multiple biopsies were taken. The sphenoidotomy was enlarged, and the sinus packed. Minimal bleeding was encountered.

FIG. 2.—Intra-operative roentgenograms showing probe in sphenoid sinus in region of anterior (a) and posterior (b) clinoid processes. Erosion of bony separation between sphenoid sinus and sella turcica is demonstrated.

Biopsy Specimens.—These showed respiratory epithelium with some chronic inflammatory change. Cultures of the fluid for bacteria and fungi were negative.

Result.—Postoperatively, there was marked reduction in the severity of headaches and subsidence of the vertigo. Subjective improvement in vision was marked. On Snellen chart testing 5 days postoperatively, the visual acuity of the right eye remained unchanged but the left had improved to 20/40. The left central scotoma was noted to have become somewhat smaller.

Discussion

A mucocele is a retention cyst which develops because of the obstruction of the ducts of mucus-secreting glands, or, as in this location, of the ostium of a sinus. Chronic infection or osteoma may be among the factors causing the symptom of obstructed drainage (van Alyea, 1960).

Perhaps the greatest interest in sphenoid sinus mucocele has been engendered by the multiplicity of ophthalmological findings it can produce. A pulsating exophthalmos in association with this condition was first reported more than 30 years ago (O'Shea, 1932). In their review of 55 cases, Lundgren and Olin (1961) reported that twenty showed ophthalmoplegias, 24 severe visual impairment, and twenty exophthalmos. Pincus and Daroff (1964) and Norman and Yanagisawa (1964) reported a case in which symptoms were identical to those of ophthalmoplegic migraine. The severity of headache has been extremely variable, and it may be referred to virtually any part of the cranium (Lundgren and Olin, 1961). Endocrine symptoms and signs may also be the presenting symptom (Meisels, 1926) though our present case showed normal endocrine functions. The diagnostic difficulties have been commented upon by McHenry, Sullivan, Ryan, and Miller (1960) and have led to craniotomy for approach to the mass (MacCarty, Lillie, Daly, Hollenhorst, and Holman, 1957).

As is apparent, the diagnosis of sphenoid sinus mucocele requires careful physical and radiological evaluation, followed by surgical confirmation. While it is not always possible to exclude all possible disorders in the differential diagnosis (Table I, opposite) pre-operatively, this does not mean that a major surgical procedure, *i.e.* craniotomy, must necessarily be carried out.

TABLE I
DIFFERENTIAL DIAGNOSIS OF SPHENOID SINUS MUCOCELE*

1. Ophthalmoplegic migraine	11. Adamantinoma
2. Aneurysm of the internal carotid artery	12. Plasmocytoma of the base of the skull
3. Hypophyseal tumours	13. Congenital anomaly
4. Nasopharyngeal tumours	14. Orbital meningocele
5. Craniopharyngioma	15. Metastatic tumour
6. Tumours of the sphenoid or ethmoid sinuses	16. Xanthomatosis
7. Meningioma of the sphenoid ridge	17. Myeloma
8. Glioma of the optic nerve or chiasm	18. Pneumosinus dilatans
9. Intracranial dermoid or epidermoid tumour	19. Retinoblastoma†
10. Chordoma	20. Neurofibromatosis

* Simon and Tingwald, 1955; Lundgren and Olin, 1961; Norman and Yanagisawa, 1964

† In children

Before surgery is undertaken, an adequate series of x-ray studies should demonstrate all the structures listed in Table II, and the radiological signs listed in Table III should be evaluated. Any indicated laminographic and contrast techniques should be used. In particular, carotid angiography is ordinarily mandatory, as an aneurysm of the circle of Willis may mimic this radiographic appearance.

TABLE II
FOR AN ADEQUATE X-RAY EVALUATION, THE FOLLOWING STRUCTURES SHOULD BE
RADIOGRAPHICALLY DEMONSTRATED*

1. Superior orbital fissures	5. Lateral walls of ethmoid sinuses
2. Optic foramina	6. Floor of sella turcica
3. Walls of orbits	7. Lesser wing of sphenoid
4. Walls of sphenoid sinuses	8. Carotid artery (by angiography)

* Simon and Tingwald, 1955.

TABLE III
RADIOLOGICAL SIGNS OF SPHENOID MUCOCELE*

1. Opacification of sphenoid sinus	5. Displacement of the adjacent internal carotid artery on arteriogram
2. Smooth outlining and rarefaction of the wall of the sinus	6. Enlargement of the superior orbital fissure with irregularity of its medial and inferior margins
3. Destructive changes of the sella turcica and occasionally of the anterior and posterior clinoids	7. Lateral displacement or thinning of the lamina papyracea
4. Destruction of the sphenoid interseptum	

* Simon and Tingwald, 1955; Norman and Yanagisawa, 1964.

After this preoperative evaluation has been carried out, we feel that the inclusion of sphenoid sinus mucocele in the differential diagnosis is an indication for surgical approach by the intranasal route. This is a much safer approach than the sub-frontal transcranial technique, as it eliminates opening the dura to a possibly infected sphenoid sinus (Lundgren and Olin, 1961; Norman and Yanagisawa, 1964). A lesion other than a mucocele may necessitate the opening of the dura for biopsy, but the safety of such an approach is confirmed by its long use in surgery on the pituitary gland (Schlosser, 1906; Kanavel, 1909; Halstead, 1910; Cushing, 1912; Atkins, 1963; James, 1964). The variation of the trans-sphenoid approach used in the present case is similar to that described 55 years ago by Hirsch (1910).

The marked variability in the signs and symptoms of an expanding intra-sphenoid

lesion is readily understood when one realizes the normal variations in the size and shape of the sinus (van Alyea, 1941; Peele, 1957), as well as its intimate relationships with dura, pituitary, optic nerve, optic chiasm, abducens nerve, oculomotor nerve, trochlear nerve, ophthalmic and maxillary branches of the trigeminal nerve, sphenopalatine nerve, nerve of the pterygoid canal, cavernous sinus, and internal carotid artery.

Summary

A case of mucocoele of the sphenoid sinus is presented with visual loss, central scotomata, headaches, and vertigo as presenting complaints. The multiple clinical syndromes which this benign lesion may produce are discussed. Transnasal biopsy may offer a relatively simple diagnostic tool, combined at times with definitive therapy.

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