GIANT MUCOCELE OF THE FRONTAL SINUS*

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The naso-mucocele is an encapsulated collection of sterile mucoid material within a nasal sinus, which, as it slowly increases in size, causes bony erosion and displaces neighbouring structures.

The English neurosurgical literature contains little on this subject but relevant papers are by Howarth (1921), Dowman (1923), Alexander (1945), and Linthicum, Rand, and Reeves (1946).

In order of frequency, mucoceles are found in the frontal, ethmoid, and sphenoid sinuses, rarely ever occurring in the maxillary sinuses (Wilkerson, 1941). These are benign lesions which occur equally in both sexes and are usually unilateral but occasionally extend to become bilateral. They occur at any age, but commonly begin in the second decade and slowly progress over many years to become manifest in adult life. There is no established aetiology.

The pathogenesis is one of a cystic dilatation of a mucous gland within the lining mucosa of the sinus (Turner, 1907). A thin-walled sac made up of fibrous tissue with low cuboidal epithelium encapsulates sterile, thick, tenacious, brownish fluid. The continuous production of mucoid material causes the mass to increase in size within the small compartment of the sinus so that the septa gradually disappear and the medial and inferior walls of the sinus compartment are eroded. Although the posterior or inner walls are weaker and thinner, they are more resistive to the pressure and are last to erode; this is most likely due to the intracranial counter pressure. The constancy of the pressure rather than the degree produces the erosion.

Clinical Picture and Diagnosis

Because of their slow, insidious growth mucoceles may not give rise to severe symptoms for a long period. The patients may, as in the case here reported, put off medical consultation and run a course as long as 15 years. The most constant complaint is headache, usually frontal, varying

from mild to severe, a fullness of the head and the cheeks, epiphora is often seen but there is seldom a nasal discharge. Frequently the most striking sign is the displacement of the orbit and the eyeball with diplopia and these patients will first seek attention from the ophthalmologist. The displacement of the eyeball is greatest in the frontal mucoceles and frequently in the advanced cases the displacement may be so extreme that the globe is forced from the socket. The displacement is downward, forward and outward. In those cases in which the posterior plate of the sinus is eroded and in contact with the dura mater and brain, the globe may not only be proptosed but will pulsate.

The radiological findings (Wigh, 1950) include an increase in radiotranslucency in the mucocele area resulting from peripheral bone loss. The areas of destroyed bone will vary in amount and direction, thus giving unusual shapes to the mucocele by allowing it to balloon beyond the confines of the sinus. The supra-orbital plate may be depressed and eroded. The most constant radiographic finding is the absence of scalloping within the sinus. A fluid level may occasionally be seen. The smooth, regular appearance of the bony erosion with marginal sclerosis is diagnostic. Such sclerosis may be sufficiently dense to suggest osteoma or underlying meningioma.

The differential diagnosis should include (1) orbital or nasal meningocele, (2) inflammatory or neoplastic lesions of the sinus, (3) epidermoid or hydatid cyst, and (4) intracranial or orbital meningioma.

Case Report, F.C. (No. 6773), a 58-year-old coloured man, was referred with the diagnosis of right frontal tumour. The illness started 15 years earlier when the patient noticed a mass over his right eye. During the next years, the mass increased in size and it was necessary repeatedly to puncture and aspirate the mass in order to decrease its volume. One year before admission the tumour became painful and grew rapidly over the right eye and nose. The patient started having headaches which were constant and associated with nausea and vomiting.

On examination, the patient showed a large tumour

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FIG. 1.—The patient.

located in the right frontal region with extension over the orbit and occluding the eye completely (Fig. 1). The mass extended also to the opposite side across the bridge of the nose toward the left eye. The nose seemed to be a part of the tumour. The eyeball was pushed downward, outward and forward. The palpebral fissure was occluded and only with extreme traction could the conjunctiva or globe be seen. There was frank conjunctivitis. However, the patient was able to detect light and movement. The globe was fixed but the pupil reacted sluggishly. He was lethargic and appeared chronically ill.

FIG. 2.—Preoperative radiograph.

Radiographs of the skull (Fig. 2) revealed advanced osseous destruction of the frontal bone measuring 7×11 cm. The margins of the absorbed bone were smooth, regular, and sclerotic. The zygoma-orbital junction on the right showed an increase in the thickness of the osseous element. The roof of the orbit was displaced downwards. The right anterior frontal bone was elevated and projected forward and anteriorly. The ethmoid sinuses appeared normal.

An electroencephalogram showed slowing in the right frontal area.

On December 6, under general anaesthesia, a coronal incision was made and the scalp was reflected anteriorly (Fig. 3) exposing the mass through the large defect in the frontal bone. When the posterior edge of the cyst was found the cyst was delineated by careful dissection. The capsule adhered firmly to the dura mater being contiguous and vascularized as one membrane. The cyst

FIG. 3.—Artist's conception of the lesion as seen at the operating table.

was punctured and approximately 160 ml. of thick, brownish fluid was withdrawn but a smear of the specimen showed no organisms and cultures were negative. The capsule was dissected from the dura. It extended to the midline and had extended in the extradural space to the coronal sutures and into the temporal area to the level of the zygoma. It extended inferiorly-posteriorly to the sphenoid ridge and to the cribriform plate. The orbital roof was displaced and instead of being convex was concave and depressed into the orbit. The cyst wall was dissected from the plate and by use of rongeurs the supra-orbital plate was completely removed back to the optic foramen. This allowed the orbital contents and globe to rise into the normal position. The frontal ostium was identified and found open; no drain was left. The left frontal sinus was found to be small and displaced to the left. The ethmoid sinus was not entered.

Histological study showed the capsule of the mucocele to be a dense fibrous wall lined by respiratory type

FIG. 4.—Photograph one year after operation.

epithelium with metaplastic changes. Laboratory studies on cyst fluid showed a specific gravity of 1.020. Electrophoresis showed a pattern consistent with serum.

The postoperative course was satisfactory and the patient has been gainfully occupied since dismissal from the hospital. Recent neurological examination shows no abnormalities save a right external squint (Fig. 4). The patient sees only 10/200 in the right eye owing to degeneration of the retina in the macular region; the nerve head is normal. Cranioplasty is to be performed.

Therapy

However mild the symptoms may be, surgical treatment is necessary in order to prevent subsequent osseous, brain, or orbital changes. The treatment of choice is the removal of the mucocele sac and the institution of adequate drainage. This can usually be accomplished in the less extensive cases by the Caldwell-Luc operation or by the Lynch radical frontal surgical exposure. An intranasal operation is occasionally adequate when the ethmoid is only involved but in the case of the frontal mucocele is less satisfactory. In those rare cases of sphenoid mucoceles the approach is endonasal.

It is our belief that if the mucocele has eroded the posterior plate and frontal bone a coronal incision and surgical removal are to be preferred, for the risks of meningeal involvement by a transnasal or radical Lynch procedure are greater. By attacking the mucocele through a coronal incision the operator is working from a clean area and will lessen the chances of a meningitis or a meningeal involvement particularly if there is erosion or a break in the dura mater. If the supra-orbital plate has been depressed with encroachment and reduction in size of the orbit, the coronal transcranial approach is mandatory to carry out a radical orbital decompression allowing the globe and orbital contents to assume their normal relations.

The unusual features of the case presented here rests in (1) its great size—it should be classed as a "giant mucocele"; (2) the extreme displacement of the globe and orbit by the depressed supra-orbital plate; (3) the extensive destruction of the inner table of the skull, the frontal bone, and the anterior upward flaring of the remaining frontal bone edge; (4) the invasion of the extradural space by the dissection of the encapsulated mucocele, much as an extradural haematoma.

The transcranial approach is a logical one though it has not been previously reported. It is indicated (1) whenever the frontal bone is destroyed, as one can then conclude that the cyst wall is in direct vascular continuity with the dura mater and that sharp dissection is necessary; (2) whenever the supra-orbital plate is depressed and an orbital decompression is required.

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