Mucoceles with intracranial and extracranial extensions

Report of two cases

FERNANDO DIAZ, M.D., RICHARD LATCHOW, M.D., ARNOLD J. DUVALL, III, M.D., CEDRIC A. QUICK, M.D., AND DONALD L. ERICKSON, M.D.

Departments of Neurosurgery, Radiology, and Otolaryngology, University of Minnesota Hospitals, Minneapolis, Minnesota

Two cases of unusual childhood mucocele are presented. These serve to illustrate the great potential these lesions have for slow, insidious development and insinuation of themselves into regions of the head and face where they are difficult to remove surgically. A description of the staged surgical excision is presented to emphasize the importance of radical removal of these benign lesions.

KEY WORDS • mucocele • osteoma • intra-extracranial lesion • total excision • two-stage operation

While mucoceles with intracranial extensions are uncommon in adults, they are even more rare in children. We have recently encountered two cases in children where the lesion had large intracranial and extracranial extensions requiring two-stage resections. In one of our cases the mucocele was associated with a large sinus osteoma, but in the other case there was no evidence of sinus disease and the origin remains unknown. The unusual clinical presentations, diagnostic procedures, and surgical approach will be discussed.

Case Reports

Case 1

This 11-year-old girl was admitted to the Neurosurgery Service at the University of Minnesota Hospitals in December, 1975, because of progressive left proptosis, loss of visual acuity, and facial swelling. At the age of 2 years, this child had surgical correction of a left esotropia, and by 4 years old was found to have decreased visual acuity in that eye. Throughout her life she had recurrent bouts of left facial swelling, and when 8 years old had developed a left sixth nerve palsy. In the 2 months before admission, she had a rather rapid progression of left proptosis and left facial swelling.

Examination. There was facial asymmetry and marked left proptosis. Neurologically, the pupil reflex was sluggish and a left fifth, sixth, and seventh nerve paresis was demonstrated. Visual acuity was limited to finger counting at 12 inches and optic atrophy was present.

Skull x-rays and facial tomograms demonstrated enlargement of the left orbit, deformity and destruction of the great and lesser wings of the left sphenoid, and destruction of the floor of the left orbit and middle cranial fossa (Fig. 1 upper left). A soft-tissue...
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FIG. 1. Case 1. **Upper Left:** Submentovertex view of the skull demonstrates enlargement of the left middle cranial fossa and posterior displacement of the left foramen ovale (arrowhead) relative to the right (arrow). These findings are indicative of focal mass effect and chronic pressure during skull maturation. **Upper Right:** Computerized tomography scan showing the missing left posterior orbital wall (arrows), and the large intraorbital mass (asterisk) producing proptosis. **Lower Left:** Tomography in the anteroposterior projection demonstrates elevation and contour change of the lesser wing of the left sphenoid bone (straight arrow), produced by the long-standing intraorbital mass. The extension into the pterygopalatine fossa has eroded the floor of the orbit (curved arrow), and thinned and elongated the pterygoid plates (arrowheads). **Lower Right:** Lateral projection of left common carotid angiogram illustrates posterior displacement of the cavernous portion of the left internal carotid artery (open arrow) secondary to the retro-orbital component of the mucocele. There is marked inferior displacement of the left internal maxillary artery (black arrows).
FIG. 2. Diagram showing extension of mucocele from the orbital cavity to the pterygoid fossa.

mass was seen ballooning the left pterygoid space and bulging into the back of the maxillary antrum (Fig. 1 lower left). The sinuses were all visualized and appeared normal. The long-standing nature of the mass was indicated by the posterior position of the left foramina ovale and spinosum (Fig. 1 upper left). A computerized tomography (CT) scan demonstrated a huge intraorbital mass with no enhancement (Fig. 1 upper right). Left carotid angiography confirmed the presence of a large avascular intra- and retro-orbital mass. The left internal maxillary artery was displaced inferiorly (Fig. 1 lower right).

Operations. The patient underwent left temporal craniotomy and orbital exploration. A large extradural dumbbell-shaped cystic mass was extending from the pterygoid fossa into the orbit through the superior and inferior orbital fissure (Fig. 2). The intraorbital component measured approximately 4 cm in diameter, and was totally excised with ligation of the more narrow portion at the orbital fissures. The mucoid material from the pterygoid component of this mass was aspirated but no attempt was made at that time to remove that portion of the lesion, which was approximately 6 cm in diameter. Two weeks after craniotomy, the retropterygoid portion was totally removed by a transpalatine retromaxillary approach performed by the otolaryngology staff.

Histological examination of the surgical specimen showed respiratory epithelium without cilia, and glandular elements with mucoid material, compatible with mucocele.

Postoperative Course. The patient subsequently made an excellent recovery apart from some episodes of bleeding from the pterygoid space. She has had marked improvement in the proptosis and cranial nerve dysfunction but no changes in her visual acuity.

Case 2

This 15-year-old girl was admitted to the University of Minnesota Hospitals in July, 1976, for evaluation of right frontal headaches of several months’ duration.

Examination. General and neurological examinations were normal, apart from right anosmia. Skull x-ray studies showed the presence of a large right anterior fossa calcification that appeared to arise from the ethmoid sinus (Fig. 3 left). The calcified mass was located over the right olfactory groove area and extended laterally over the sphenoid wing with some displacement of the medial orbital wall. An angiogram demonstrated a subfrontal mass which was larger than the calcified mass, suggesting the presence of an avascular lesion, 2 to 3 cm in thickness, capping what appeared to be a sinus osteoma (Fig. 3 right).

Operations. A right frontal craniotomy revealed the presence of an anterior fossa mucocele surrounding the calcified mass (Fig. 4). The mucocele and intracranial components of the osteoma were excised leaving the inferior portion of the osteoma deep in the ethmoid sinus. One month later, she underwent transethmoidal removal of the remaining portion of the osteoma, by the otolaryngological staff.

Postoperative Course. There were no complications or changes in neurological status related to the operations and the patient subsequently had a benign course.

Discussion

Intracranial and intraorbital mucoceles present as slowly enlarging masses usually arising from one of the paranasal sinuses. A mucocele usually occurs when drainage of a
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Fig. 3. Case 2. *Left:* Anteroposterior projection shows the huge, homogeneously dense osteoma of the right ethmoid air cells, with extension into the right anterior cranial fossa. *Right:* The venous phase of the right carotid angiogram demonstrates a vein (arrows) above the osteoma, separated by a cap of avascular tissue. This cap proved to be a portion of the mucocele.

paranasal sinus is obstructed, due to inflammation, fibrosis, trauma, previous surgery, or anatomical abnormality, or with a mass lesion such as an osteoma. A preceding history of sinusitis occurs in 50% of the cases, trauma in 28%, and allergies in 11%. The first description of an orbital mucocele was in 1785 by Henry Nicolai. In 1888, Berthan revised the concept, and, in 1896, Roulet coined the term "mucocele." The first English report was by L. Turner early in the 20th century.

Frontal sinus mucoceles are the most common form, followed by ethmoidal and maxillary lesions. Sphenoid sinus mucoceles are rare. While mucoceles can occur at any age, they are distinctly rare in infancy and childhood.

Intraorbital mucoceles generally occur as an extension of frontal or ethmoidal sinus involvement. Very few cases of intraorbital mucocele in childhood have been reported, and all of these demonstrated clinical or radiographic evidence of inflammation in the ethmoid sinus.

The origin of the mucocele in Case 1 is obscure. We are postulating the presence of an aberrant sinus without an ostea. The tumor followed the typical course of an intraorbital mucocele with slow progression, remissions, and exacerbations. In Case 2, the origin is obviously related to the presence of the ethmoid osteoma. Proptosis and visual loss typically occur late.

Fig. 4. Diagram demonstrating relationship of osteoma to mucocele.
Tomography is useful in demonstrating the destruction and deformities characteristic of these long-standing benign expansile lesions.\(^4\)\(^8\)\(^9\) Computerized tomography scanning can also contribute by further elucidating the position of the avascular mass. The orbital lesions are particularly well demonstrated by this technique. In both of these patients angiography aided in estimating the size of the mass, in addition to the position of major surrounding vessels.

These two patients present unusual lesions requiring two-stage radical removal. It is unacceptable to leave any significant portion of the secreting lining of these masses without risking recurrence. This is also true of the osteoma, which must be totally excised. These two patients had tumors that had developed in such a fashion as to make removal from one surgical approach technically very difficult. The two-stage approach in these patients has given what we believe to be total removal of these lesions. Certainly in the first patient, there did not appear to be a single approach that would allow safe removal of the intraorbital lesion and adequate exposure of the retropterygoid portion. This transpalatine route has been used to resect other tumors at our institution but has not previously been employed for a mucocele.\(^2\) Likewise, in the second patient, a craniotomy was necessary to remove the intracranial portion but did not provide safe approach to the deep portion of the osteoma within the ethmoid sinus.

**Summary**

Mucoceles may present as complex intra-extracranial mass lesions. Total excision is required to assure permanent resolution. At times, a two-stage approach is warranted if total excision is impossible from one surgical exposure. The two rather unusual cases presented in this paper warranted such treatment.

**References**


*Address reprint requests to: Donald L. Erickson, M.D., Department of Neurosurgery, Box 96 Mayo, University of Minnesota Hospitals, Minneapolis, Minnesota 55455.*