Mucocele of the sphenoid sinus with bilateral internal carotid artery occlusion

Case report

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A young man with a large sphenoid sinus mucocele developed hypopituitarism, headaches, and visual difficulties. Subsequently the lesion caused complete occlusion of both internal carotid arteries in the parasellar region. The headaches and visual difficulty improved after simple transoral drainage of the cyst.

KEY WORDS  
Mucocele  
Sphenoid sinus  
Pituitary  
Parasellar lesion  
Carotid occlusion

Mucocele of the sphenoid sinus is unusual. Nugent, et al., recently reviewed the world literature, finding 81 cases, reporting 63 of these, and adding two of their own.

The lesion is of interest to the neurosurgeon because the signs, symptoms, and radiographs have frequently prompted a craniotomy in search of a suspected intracranial tumor. However, with the correct preoperative diagnosis, the preferred approach is usually via transmaxillary, endonasal, or other anterior routes. This case is presented because of the previously unreported development of bilateral occlusion of the intracranial internal carotid arteries at the site of the mucocele.

Case Report

First Admission. This patient was first admitted to Pennsylvania Hospital in 1966 when he was 15 years old with a chief complaint of severe right frontal headaches of 3 weeks’ duration, worse behind the right eye and associated with twitching of the right eyelid. He denied visual disturbances and symptoms of acute or chronic upper respiratory tract infection. He was then of normal size for his age, but lacked pubic hair. Optic fundi and visual fields were normal as was the remainder of the general and central nervous system examination. Skull films demonstrated a destructive lesion involving the sphenoid sinus and the retropharyngeal space. Planograms confirmed this and showed an indistinct sellar floor, apparently destroyed. The clinoids were intact. Bilateral carotid arteriography showed a right internal carotid artery which narrowed markedly at a point 2 cm above its origin and remained narrow throughout its course. Also, the right carotid siphon was elevated and displaced laterally (Fig. 1). The left internal carotid artery was entirely normal. Pneumoenceph-
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Fig. 1. Lateral left carotid arteriogram in 1966 (left) and comparable drawing (right) demonstrating marked narrowing of the internal carotid artery, particularly in the parasellar area. Arrow indicates obstruction of the left internal carotid artery in the cavernous sinus.

angiography was normal except for absence of air in the chiasmatic cistern. The electroencephalogram (EEG) and echoencephalogram were normal. Nasopharyngeal biopsy under local anesthesia obtained only lymphoid tissue. More extensive nasopharyngeal biopsy under general anesthesia, carried down to bone, obtained more lymphoid tissue. At craniotomy, the optic chiasm was exposed, and a small piece of reddish tissue anterior to the chiasm was biopsied. There was no gross distortion of the right optic nerve or chiasm. The intracranial portion of the right internal carotid artery appeared normal. The histology of the biopsied tissue showed primarily amphophilic cells, although acidophilic and basophilic cells were scattered throughout. In retrospect this was probably normal anterior hypophyseal tissue displaced through the diaphragma sellae by pressure from below. Postoperatively, he developed transient diabetes insipidis, but otherwise did well. An endocrine work-up indicated a definite deficiency in growth hormone and probable TSH and LH deficiencies. He was treated with growth hormone for 1 year and was started on testosterone because of clinical evidence of slow sexual maturation. He received a total of 4600 R in air to the midline of the skull through a 5 × 5 cm portal. Treatment was delivered on a 1 MEV apparatus over 44 days, through parallel opposed portals, and was completed November 23, 1966.

Second Admission. Three years later in 1969 the patient noted periods of blurred and double vision, and again complained of headaches which he described as a sensation of pressure behind his eyes. Corrected vision was 20/30 left and 20/40 right, and there was now a definite right lateral rectus weakness. Both optic discs were pale, the right more than the left. External ocular muscles were intact. There was an enlarged right blind spot and bitemporal hemichromatopsia to a 5 mm red test object. Skull films showed that the huge, soft tissue mass was much larger than in 1966; it now involved the sphenoid sinus and bulged into the retropophysyalgeal space, destroying the floor of the sella turcica (Fig. 2). A right retrograde brachial arteriogram demonstrated complete occlusion of the right internal carotid artery at 2 cm above the bifurcation, with a greatly dilated vertebrobasilar system supplying the entire intracranial circulation including both anterior cerebral arteries (Fig. 3). A left carotid arteriogram demonstrated a severely narrowed left intracranial internal carotid artery, elevated and displaced laterally. It appeared to be stretched over a rounded suprasellar mass. A nasopharyngeal biopsy under local anesthesia was unrewarding.

Operation. Under general anesthesia, the posterior nasopharynx was exposed transo-
Carotid occlusion due to mucocele

Fig. 2. Skull film in 1969, 3 years after original operation. There is now marked destruction of the entire floor of the sella. A soft tissue mass is seen extending into the sphenoid sinus and retropharyngeal area.

rally, and incised with the intent of biopsying the mass. A huge cyst, extending posteriorly and superiorly, was entered and drained of 50 to 75 cc of brown thick fluid under pressure. The interior of the cyst was palpated and found to be bony. The anterior wall (posterior nasopharynx) was removed, creating a permanent dependent ostium. Section of this tissue showed respiratory epithelium overlying dense fibrous tissue. On the other side of the fibrous margin was a squamous to cuboidal respiratory epithelium. The findings were considered consistent with the diagnosis of mucocele.

Postoperative Course. X-ray films taken immediately postoperatively showed complete pneumatization of the soft tissue mass with air extending to the diaphragma sella (Fig. 4). The pressure headaches, diplopia, and blurring of vision disappeared immediately and have not returned. The right disc pallor and hemichromatopsia have persisted. When last seen 1 year postoperatively, the patient was asymptomatic.

Discussion

Although some doubt exists concerning the etiology of mucoceles, they probably arise either primarily from obstruction and enlargement of a goblet cell gland, or secondarily as a result of obstruction of a sinus ostium, with retention of secretions. Age and sex are not predisposing factors.

Symptoms are remarkably variable and frequently nonspecific. Headache occurred in 71% of the cases of Nugent, et al., and is

Fig. 3. Right retrograde brachial arteriogram in 1969, 3 years after the original carotid arteriograms, now demonstrates filling of the entire cerebral circulation via the basilar and posterior communicating arteries. Arrows indicate sites of occlusion of both intracranial internal carotid arteries in the parasellar area.

Fig. 4. X-ray film shortly after drainage of the sphenoid mucocele shows air filling the mucocele cavity, which extends well beyond the confines of the sphenoid sinus. In the upright position air extends to the presumed level of the diaphragma sella (arrow) but does not enter the intracranial space.
commonly located either in the frontal region or behind the eye. Eye signs include diminution of vision, diplopia, exophthalmos, and scotoma, and occur in two thirds of the cases. Nasal symptoms or sinus disease occurred in 46% of the reported cases. Signs of pituitary insufficiency have been seen rarely and pituitary insufficiency documented in only one case and that postoperatively. Radiographic signs include enlargement and opacification of the sphenoid sinus with thinning of its walls and destructive changes of the sphenoid interseptum, sella turcica, and clinoids. The superior orbital fissure may be enlarged and show irregular margins. There may be lateral displacement of the lamina papyracea. Arteriograms may be normal or may show narrowing or displacement of the carotid siphon.

No cases of complete uni- or bilateral internal carotid artery obstructions have been reported previously. Carotid thrombosis has been reported with extrasellar extension of a pituitary tumor.

Treatment consists of drainage and formation of an adequate ostium. If possible, the lining membrane should be removed, although in very large mucoceles this may be omitted if the optic nerves would be endangered by instrumentation within the cyst. Kruger reported cure by repeated sphenoid sinus irrigation under radiologic guidance (Simon, and Tingwald, 1955). The transnasal route is preferred, and craniotomy should be avoided not only because of the higher morbidity and mortality associated with the procedure itself, but because of potential postoperative infection and cerebrospinal rhinorrhea.

Recovery from most symptoms including headache, external ocular muscle palsies, and nasal symptoms is usually prompt and complete. Visual acuity, if only mildly diminished, will usually return to normal. Unilateral blindness disappears in one half of cases reported, but bilateral blindness is usually permanent.

Summary
We have presented a patient who had a large sphenoid sinus mucocele that produced headaches, decreased vision, and occlusion of both internal carotid arteries in the parasellar region. The symptoms ultimately subsided when the mucocele was drained by the transoral route.

References

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