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SPHENOID MUCOCOELE AS A COMPLICATION OF FIBROUS DYSPLASIA OF THE FACIAL BONES

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We report on a 28-year-old man with severe headache. Imaging studies included CT and MR. A large sphenoid mucocoele was demonstrated as well as findings compatible with fibrous dysplasia of the facial bones. Both disorders had been previously unknown. Imaging findings suggested that the sphenoid mucocoele was related to an obstruction caused by the expansile bone of fibrous dysplasia. These findings were confirmed surgically. Fibrous dysplasia with subsequent outflow obstruction is an extremely rare cause of sphenoid mucocoele development.

Key-word: Mucocele.

A mucocoele is a benign encapsulated expansile mass filled with mucous material. It is lined by respiratory mucosa. It enlarges gradually and may result in erosion of the bony walls of the sinus. One reported cause of the development of a mucocoele is an obstruction of the ostium of the sinus. Mucocoeles occurring in the sphenoid sinus make up 2% of paranasal sinus mucocoeles (1).

Fibrous dysplasia is a skeletal developmental abnormality of the mesenchyme caused by a defect in osteoblastic maturation and differentiation. Bone is replaced by fibrous tissue which is variably calcified and may lead to the relatively typical ground glass appearance. Any bone in the body may be affected but sites of predilection in the skull include the frontal, sphenoid, maxillary, and ethmoidal bones (2).

Mucocoeles occurring as a complication of tumors or tumorlike lesions are exceedingly rare with only a few cases reported in the literature (3).

Case report

A 28-year-old man presented to the emergency department with a history of severe occipital headache for two days. He had self medicated with over the counter analgetics but this had been unsuccessful in alleviating the pain. He also reported pain in the neck region irradiating to the right shoulder and hand and tingling in the hand. On further questioning he reported daily headaches for years in addition to a stuffed nose. On physical examination his head was lateroflexed to the right. Paresthesia of the right arm was present but otherwise neurological examination was unremarkable. The results of laboratory tests were normal.

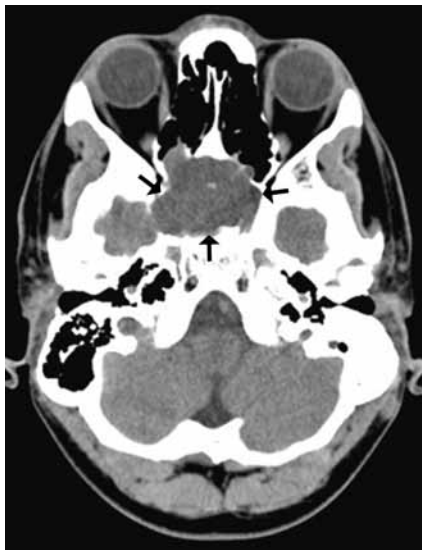

Fig. 1. — Note slightly hypodense mass (arrows) involving the sphenoid sinus area on this transverse non contrast enhanced CT image (soft tissue algorithm).

On the basis of these findings a non-contrast enhanced CT of the skull with multiplanar reconstructions was performed. CT revealed a large slightly hypodense and expansile mass in the sphenoid region (Fig. 1). Erosion of the bony walls of the sphenoid sinus was also present. Findings characteristic of fibrous dysplasia were also depicted in the facial bones and included a ground glass appearance and expansion of the bony structures (Fig. 2, 3). Subsequently an MRI of the skull was performed. On T1-weighted images with intravenous contrast the expansile lesion of the sphenoid sinus appeared hypointense and showed delicate rim enhancement after intravenous contrast administration (Fig. 4). On T2-weighted images the mass appeared hyper - intense (Fig. 5).

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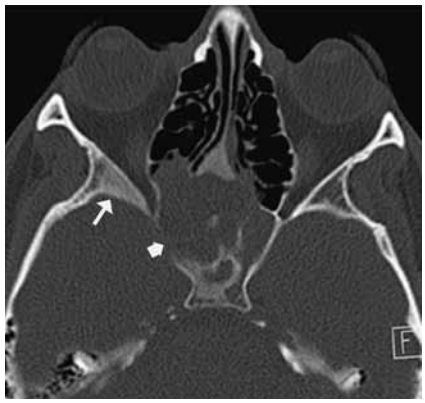

Fig. 2. — Transverse CT image (bone algorithm) shows eroded medial wall of the expanded sphenoid sinus (short arrow). Note ground glass appearance in expanded bone (long arrow) typical of fibrous dysplasia of the skull base.

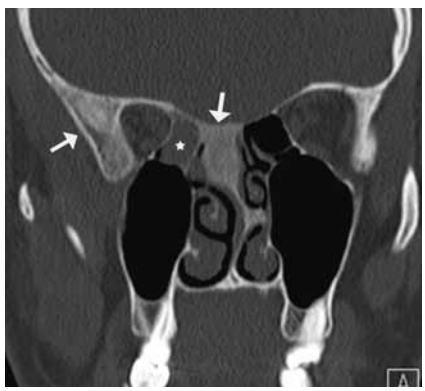

Fig. 3. — Coronally reconstructed multiplanar CT image (bone algorithm). Note the typical ground glass appearance and expansion of the lateral orbital wall and nasal septum (arrows). Anterior part of mucocoele is seen medial to the medial orbital wall (asterisk).

Preoperatively the diagnosis of a sphenoid mucocoele as a complication of fibrous dysplasia of the facial bones was made. The patient was sheduled for transnasal sphenoidotomy. The anterior and inferior wall of the sphenoid sinus were removed allowing drainage of the mucocoele. Preoperative diagnosis was confirmed surgically. Pathological find-

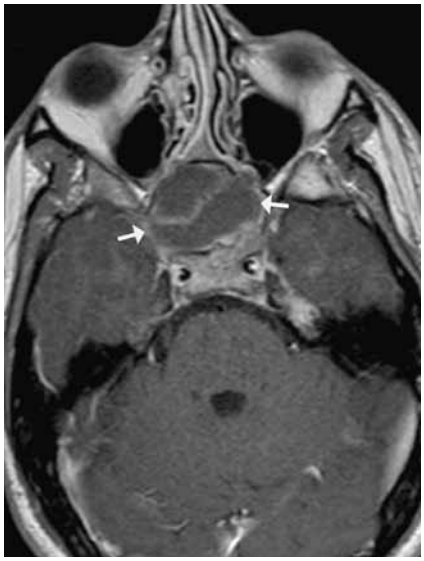

Fig. 4. — Transverse T1-weighted MR image after contrast administration. Note isointense sphenoid sinus mass with rim enhancement (arrows).

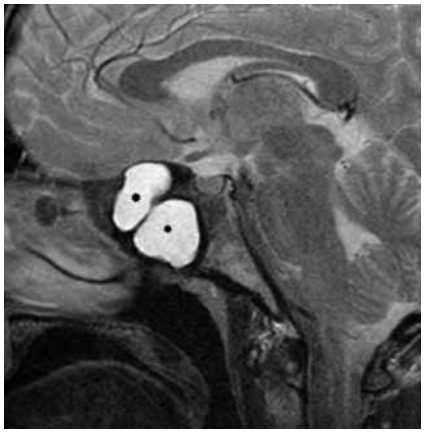

Fig. 5. — Sagittal T2-weighted MR image. Note hyperintense expansile lesion in the sphenoid sinus (asterisk).

ings of the bone fragments confirmed fibrous dysplasia. Postoperative evolution was unremarkable.

Discussion

Only about 2% (1) of all paranasal sinus mucocoeles are located in the sphenoid sinus. Most are simple mucocoeles, and secondary mucocoeles are exceedingly rare. Only a few cases have been reported and causes include carcinoma, osteoma, ossifying fibroma, fibromyxoma, Paget's disease (4), and fibrous dysplasia (3). Transsphenoidal hypophysectomy also may be the cause of a sphenoid mucocoele (5).

Mucocoeles develop chronically, expanding progressively and sometimes leading to erosion of the bony wall of the sinus. Expansion may sometimes be severe with involvement of adjacent intra- and extracranial spaces. In rare cases posterior rupture may even cause brainstem inflammation (6).

The etiology remains somewhat unclear although the initial event is believed to be an obstruction of the draining ostium of the sinus. There are other theories including a cystic dilatation of glandular structures, or a cystic enlargement of embryonic epithelial residual cells.

Clinical symptoms of sphenoid sinus mucocoeles are variable and non-specific. This delays the diagnosis in many cases. Most commonly symptoms include headache, visual disturbances, vertigo, facial pain, nasal discharge, and palsy of the occulomotor and abducens nerves (3). Headache appears to be the most common symptom. Diagnosis is established earlier if there are neurological symptoms.

In our patient headache had been present since many years but was now exacerbated for an unknown reason prompting a visit to the emergency department.

The other clinical findings were seemingly unrelated but contributed to the decision to perform a CT which ultimately showed the mucocoele.

Diagnosis is made with CT and MRI. On CT variable densities may be present in the mucocoele. Precise location and expansile aspect of the lesion are demonstrated. The different densities may be attributable to the protein content and possible surinfection or hemorraghe, but the lesion is usually homogeneous. Typically contrast enhancement is absent except for delicate rim enhancement related to the encapsulated nature of the lesion (3). Such rim enhancement was also present in our patient.

Signal intensities on MRI are likewise variable. The difference is attributable to the variability of cyst content (7). MRI is better able to demonstrate the relationship of the mucocoele to adjacent vessels and nerves.

Differential diagnosis of cystic sphenoidal lesions includes chordoma, pituitary adenoma, craniopharyngeoma (8), dermoid, and arachnoid cyst (3). A mucocoele should also be differentiated from a simple fluid retention which is much more common. A fluid retention typically does not show an expansile aspect.

Fibrous dysplasia can affect virtually any bone in the body and represents a nonhereditary disorder of unknown cause (2). Bone is replaced by fibrous tissue which is variably calcified and may be more or less radiolucent on radiographs and CT scans. Different forms are recognized including a monostotic, polyostotic, craniofacial form, and cherubism. Typical findings of fibrous dysplasia include the slightly expansile nature of the bony structures and the ground glass appearance. These findings are better demonstrated with CT since MRI is less suited to evaluate bony structures.

Treatment of sphenoid mucocoeles is mandatory especially when neurological symptoms are present. It consists of transnasal sphenoidotomy with subsequent drainage of the sinus.

In conclusion mucocoeles of the sphenoid sinus are rare lesions. Mucocoeles secondary to tumor or tumorlike lesions are exceedingly rare. In this article we present a case where a sphenoid mucocoele occurred secondary to fibrous dysplasia of the facial bones, which is an exceedingly rare occurrence. CT and MRI allowed the preoperative diagnosis of the sphenoid mucocoele and the involvement of the skull base by fibrous dysplasia. A secondary cause for sphenoid mucocoeles, although very rare, should always be considered.

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