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.

Preoperatively the diagnosis of a sphenoid mucocoele as a complication of fibrous dysplasia of the facial bones was made. The patient was scheduled for transnasal sphenoidotomy. The anterior and inferior wall of the sphenoid sinus was removed allowing drainage of the mucocoele. Preoperative diagnosis was confirmed surgically. Pathological find-
Disturbances, vertigo, facial pain, sinus in many cases. Most commonly non-specific. This delays the diagnosis. Sinus mucocoeles are variable and epithelial residual cells. A cystic enlargement of embryonic draining ostium of the sinus. There believed to be an obstruction of the inflammation. 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 surfactant in infection or hemorrhage, but the lesion is usually homogeneous. Typically contrast enhancement is absent except for delicate rim enhancement related to the encapsulated nature of the lesion. 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. 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, cranioophygeoma, dermoid, and arachnoid cyst. 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. 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, 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 tumourlike 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.

References