An aggressive psammomatoid ossifying fibroma of the sinonasal tract: Report of a case

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Abstract
Aggressive psammomatoid ossifying fibromas (APOFs) represent a subgroup of related fibro-osseous lesions that appears to be unique to the nasal cavity, paranasal sinuses, and orbit. These rare lesions are characterized by distinctive histomorphologic features and a tendency to affect younger patients. Histologically they are benign, but clinically they are locally aggressive. We report the case of a 15-year-old boy who had a large APOF in the left ethmoid and sphenoid sinuses. The location of this tumor made this case unusual.

Introduction
In recent years, the term fibro-osseous lesion has grown in popularity as an overall designation for a number of rare, histologically benign lesions of the head and neck that are made up of bone, fibrous tissue, and cemetum.1,2 The designation is somewhat generic because there is no universally agreed-upon histopathologic classification for these tumors. The biologic characteristics of fibro-osseous lesions range from indolent to aggressive and from inflammatory to neoplastic. Lesions that involve the midface and paranasal sinuses tend to demonstrate aggressive behavior and rapid growth.

A sub-group of related fibro-osseous lesions that appears to be unique to the nasal cavity, paranasal sinuses, and orbit has been described; these lesions are called aggressive psammomatoid ossifying fibromas (APOFs).2 These particular lesions are characterized by distinctive histomorphologic features (including psammomatoid ossicles), a tendency to affect younger patients, and locally aggressive behavior.2-4 En bloc excision is the treatment of choice.

We report the successful endoscopic removal of a large APOF from the left ethmoid and sphenoid sinuses. The location of this tumor made this case unusual.

Case report
A 15-year-old boy presented to the emergency room with an 8-month history of painless left-eye proptosis, nasal obstruction, frontal headache, and rhinorrhea. He denied any visual disturbances. He had no history of allergic rhinitis or asthma. In fact, his entire medical history was unremarkable.

Physical examination revealed that the proptosis of the left eye was only slight. Anterior rhinoscopy detected a large mass in the left nasal cavity that had originated in the sphenoethmoid area. The mass had displaced the septum and obstructed both nasal cavities. The obstruction of the right nasal cavity had been caused by both the displaced septum and hypertrophy of the inferior turbinate.

Computed tomography (CT) in both axial and coronal planes identified an expansile soft-tissue–density mass that was confined to the left sphenoid and ethmoid sinuses with foci of calcification that had displaced the globe anterolaterally secondary to the extension of the mass through the medial wall of the orbit (figure 1). No intracranial spread was seen.

Figure 1. CT shows the ossifying fibroma in the left sphenoid and ethmoid sinuses.
Inspissated gray-brown and calcific material was removed endoscopically. En bloc histologic examination of the 3 × 5 × 4-cm excised specimen showed the distinct histomorphology of an APOF, including spherical ossicles that contained osteocytes (figure 2). Re-examination 18 months postoperatively detected no evidence of recurrence.

Discussion
The most common clinical manifestation of APOF is proptosis, which occurred in our patient. Other findings include visual disturbances (including progressive blindness in some cases), airway obstruction, headache, and a mass lesion.

The aggressive behavior of APOFs ranges from a bowing or pushing of adjacent bone (like the bowing of the septum in our patient) to direct invasion through bone and extension into adjacent anatomic compartments (again, like the invasion of the orbit in our patient). The invasion can be extensive and involve several sinuses, the nasal cavity, the nasopharynx, the palate, and even the cranial cavity.

The histologic diagnosis of an APOF is difficult. Clinico-pathologic correlation, particularly with the radiographic features, is essential. Radiographic imaging is invaluable for establishing a diagnosis and for determining the extent of the lesion. CT will show a well-demarcated, expansile mass covered by a thick shell of bone density; the content of the mass will appear to be multiloculated and of varying density.

When possible, complete surgical excision of an APOF will relieve symptoms and prevent recurrence. However, depending on the location and extent of the tumor, surgery is not always feasible. In such cases, endoscopic management is an alternative. Craniofacial resection is necessary for lesions that extend into the cranial cavity.

Adjuvant therapy, such as radiotherapy, should not be administered because it can induce malignant changes. Despite the considerable morbidity associated with APOFs, which is a result of their tendency toward local invasive growth and recurrence, there have been no reports of an APOF metastasizing. The prognosis for patients with these lesions is considered good.

References