Juvenile Active Ossifying Fibroma

Brandon R. Peterson · Brenda L. Nelson

Received: 7 October 2014 / Accepted: 1 December 2014 / Published online: 7 December 2014
© Springer Science+Business Media New York (outside the USA) 2014

Abstract A case of juvenile active ossifying fibroma affecting a 27 year-old man will be discussed. The characteristic radiologic and histologic features of the entity will be described.

Keywords Juvenile active ossifying fibroma · Active ossifying fibroma · Aggressive psammomatoid ossifying fibroma

History

A 27-year-old African-American male presented to his primary care physician with long-standing nasal congestion and recurrent sinus infections. After initial symptomatic treatment with nasal decongestants and anti-histamines, radiographic studies were obtained to evaluate for chronic sinusitis.

Radiographic Features

Computed tomography images of the sinuses demonstrated a 2.1 cm round bony lesion with ground-glass attenuation within the superior orbital rim of the left frontal sinus with remodeling of the skull base and orbit (Fig. 1).

Disclaimer The opinions and assertions expressed herein are those of the author and are not to be construed as official or representing the views of the Department of the Navy or the Department of Defense.

B. R. Peterson (✉) · B. L. Nelson
Department of Anatomic Pathology, Naval Medical Center San Diego, 34800 Bob Wilson Drive, San Diego, CA 92134-5000, USA
e-mail: Brandon.peterson2@med.navy.mil

Additionally, magnetic resonance imaging (MRI) also shows a smoothly margined lesion centered in the left frontal sinus. A peripheral rim, thought to be residual surrounding inspissated secretions is identified (Fig. 2). Noted on both images, although more easily seen on the MRI, is the nearly complete opacification of the right paranasal sinuses. This was later diagnosed as sinonasal inflammatory polyps.

Diagnosis

A diagnostic biopsy was performed. Histologic examination of the biopsy and subsequent excision specimens demonstrated numerous acellular, concentrically lamellated particles with a central basophilic area surrounded by an eosinophilic rim (Figs. 3, 4). Focally, these ossicles coalesced to form irregular trabeculae. The cellular stroma was composed of uniform stellate fibroblast-like cells. The surrounding normal bone demonstrated active osteoclastic resorption.

Discussion

Juvenile active ossifying fibroma (JAOF, also referred to as “active ossifying fibroma” or “aggressive psammomatoid ossifying fibroma”) is a rare variant of conventional ossifying fibroma (OF) with many important distinguishing clinical and histologic features. As the name implies, the average age of JAOF is younger than conventional OF. Most cases occur in patients <12 years old, though a wide age range has been reported (3 months to 72 years) [1]. There is an equal gender distribution, as opposed to the female predominance seen in conventional OF. As in our

Fig. 1 Computed topography (CT) of the sinuses demonstrating a well-demarcated, radiolucent lesion with central radiopacities within the left frontal sinus

Fig. 2 Magnetic resonance imaging (MRI) shows an expansile lesion of the left frontal sinus. Note the lesions of the right paranasal sinuses, diagnosed as sinonasal inflammatory polyps

case, the most common location is the paranasal sinuses, where it may cause obstructive symptoms. Less common locations include the jaws (maxilla > mandible) and temporal bone [2, 3]. Radiographically, JAOF presents as a well-demarcated, expansile, radiolucent lesion surrounded by an ossified rim. Rare cases demonstrate an invasive growth pattern with involvement of adjacent structures, including intracranial extension [1].

Macroscopically, the lesions are well-circumscribed and have a smooth surface. They are often described as

Fig. 3 A medium power H&E image showing cellular connective tissue with spherical ossicles composed of a basophilic center with an eosinophilic rim

Fig. 4 A high power image showing ossicles that are focally fused

“shelling out” intraoperatively. Microscopically, a network of osteoid is seen within a cellular stroma composed of bland spindled to stellate cells. Scattered mitotic figures may be seen. The most distinctive finding, which is variably present, is the presence of small spherical structures with concentric lamellations, reminiscent of psammoma bodies. These structures are usually acellular, though entrapment of osteoblasts may be seen. The ossicles often have a basophilic center surrounded by a thick eosinophilic rim. Multinucleated giant cells are common, and there are numerous reports of secondary aneurysmal bone cyst formation [1, 4, 5].

Juvenile active ossifying fibroma belongs to a morphologic category of benign fibro-osseous lesions in the head and neck which also includes conventional ossifying

fibroma, cemento-osseous dysplasia, and fibrous dysplasia. Due to the morphologic overlap between these entities, clinical and radiographic information is essential to guide the pathologist's interpretation. Compared with JAOF, conventional ossifying fibroma is more common in the jaws and typically occurs in adults. JAOF is also distinguished by a more aggressive clinical behavior. In addition, conventional OF typically lacks the psammomatous calcifications seen in JAOF. Cemento-osseous dysplasia, while morphologically similar to JAOF, rarely presents before age 20, and by definition occurs in the jaws in close association with the roots of teeth. Classic radiographic features of fibrous dysplasia include a "ground glass" appearance with poorly defined borders, in contrast with the well-demarcated features of JAOF. When multiple bones are involved (polyostotic), fibrous dysplasia is favored. Histologically, fibrous dysplasia shows woven bone without osteoblastic rimming which fuses with adjacent uninvolved bone. This continuity of normal and pathologic bone is not seen in JAOF. The psammomatous calcifications of JAOF may mimic the true psammoma bodies of extracranial meningioma, though extracranial meningioma should have plump epithelial cells with a whorled architecture. In difficult cases, immunohistochemical stains for EMA and CK7, which are strongly positive in extracranial meningioma, should clarify the diagnosis.

The treatment of choice for JAOF is surgical excision. There is an excellent prognosis following complete excision, though this may be difficult when an infiltrative growth pattern is present. Incompletely excised tumors

have a recurrence rate of 30–58 %, which may necessitate further surgery with reconstruction [6]. Rare cases with intracranial extension have been associated with secondary bacterial infections with resulting mortality [1]. Despite an aggressive clinical course, true malignant transformation has not been described.

References

1. Johnson LC, Yousefi M, Vinh TN, Heffner DK, Hyams VJ, Hartman KS. Juvenile active ossifying fibroma: its nature, dynamic and origin. *Acta Otolaryngol Suppl.* 1991;488:1–40.
2. Tolentino ES, Centurion BS, Jjioe KC, Casaroto AR, Tobouti PL, Frederique U Jr, Lara VS, Damante JF, Sant'ana E, Goncales ES. Psammomatoid juvenile ossifying fibroma: an analysis of 2 cases affecting the mandible with review of the literature. *Oral Surg Oral Med Oral Pathol Oral Radiol.* 2013;113(6):e40–5.
3. Hasselblatt M, Jundt G, Greiner C, Rama B, Schmal F, Iglesias-Rozas JR, van de Nes JA, Paulus W. Juvenile psammomatoid ossifying fibroma of the neurocranium. Report of four cases. *J Neurosurg.* 2005;102(6):1151–4.
4. El-Mofty S. Psammomatoid and trabecular juvenile ossifying fibroma of the craniofacial skeleton: two distinct clinicopathologic entities. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod.* 2002;93:296–304.
5. Urs AB, Augusting J, Arora S, Kumar P. Rare pediatric presentation of aneurysmal bone cyst with trabecular juvenile ossifying fibroma and ossifying fibroma. *Int J Pediatr Otorhinolaryngol.* 2013;77(4):576–80.
6. Slootweg PJ, Panders AK, Koopmans R, Nikkels PG. Juvenile ossifying fibroma. An analysis of 33 cases with emphasis on histological aspects. *J Oral Pathol Med.* 1994;23:385–8.