Case Report

Trabecular Juvenile Ossifying Fibroma With Aneurysmal Bone Cyst: A Rare Presentation

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Abstract: *Juvenile ossifying fibroma is an uncommon, benign, bone-forming neoplasm that is distinguished from other fibro-osseous lesions primarily by its age of onset, clinical presentation, and potential behavior. It mainly occurs in juveniles and has a slight male predilection and more aggressive behavior than a common ossifying fibroma. There are 2 distinct histopathological variants of this lesion: (1) psammomatoid pattern and (2) trabecular pattern. An aneurysmal bone cyst may occur in association with other bone lesions, such as fibrous dysplasia, ossifying fibroma, and giant cell lesion. The clinical management and prognosis of juvenile ossifying fibroma is somewhat uncertain, and this tumor has high rates of recurrence. Such behavior may be related to younger patient age and the concurrent development of aneurysmal bone cyst. The purpose of this paper was to report a case of trabecular juvenile ossifying fibroma in a 9-year-old girl associated with an aneurysmal bone cyst, presenting an aggressive behavior, and causing significant facial asymmetry. (Pediatr Dent 2011;33:388-91) Received February 15, 2010 / Last Revision May 1, 2010 / Accepted May 3, 2010*

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Juvenile ossifying fibroma (**JOF**) is an uncommon benign neoplasm that arises within the craniofacial bones.[1,2] It is a variant of the ossifying fibroma, distinguished from other fibro-osseous lesions primarily by its age of onset, clinical presentation, and potential behavior.[3]

This tumor mainly occurs in juveniles[4-6] and exhibits a more aggressive behavior in comparison with the more common ossifying fibroma seen in adults, with some exceptions.[7] It is usually asymptomatic, reaching a large size and being locally aggressive.[8] Its first clinical presentation is an extraoral mandibular swelling.[9] In the jaws, JOF is considered to develop from undifferentiated cells of the periodontal ligament.[10,11]

These lesions are nonencapsulated neoplasms, but are well delimited by the adjacent bone.[4,8] The radiologic features are variable and depend on the location of the tumor, maturation stage, and the amount of the calcification. Radiographically, they present as circumscribed radiolucent areas, which in some cases contain central radiopacities.[12]

There are 2 histopathological variants of this lesion: (1) a psammomatoid (**PsJOF**) pattern and a trabecular (**TrJOF**) pattern. PsJOF involves the bones of the orbit and paranasal sinuses, whereas TrJOF is predominantly a gnathic lesion with a predilection for the maxilla.[11] There is also an age difference, with the PsJOF occurring in an older and wider age range vs the TrJOF, which occurs in an average range of 8 to 12 years of age[5] Histologically, both variants tend to be unencapsulated and composed of a cellular fibrous connective tissue and have small areas of giant cells. Both differ in their mineralized components; one shows trabeculae of immature woven bone (trabecular type), and the other presents concentric lamellated and spherical ossicles that vary in shape (psammomatoid type).[7]

The clinical management and prognosis of JOF is somewhat uncertain and recurrences range from 38% to 58%.[2-4] Many authors recommend conservative excision or curettage,[4,9,10] and others believe that some lesions may necessitate more aggressive management, such as en-bloc resection.[2,8,13]

Although JOFs are solid tumors, they can develop microscopic or macroscopic cystic lesions.[14] Such cystic spaces have been described as aneurysmal bone cyst-like areas containing blood and giant cells.[15]

The aneurysmal bone cyst is an expansile osteolytic lesion, often multilocular, with blood-filled spaces separated by fibrous septa containing osteoclast-type giant cells and reactive bone.[16] It usually affects the long bones. In the jaws, it is more often found in the mandible, predominantly in the posterior regions, and is more common in the second to third decades of life. In spite of being well recognized, the etiology and pathogenesis have yet to be elucidated.[17]

It is well known that the aneurysmal bone cyst may arise primarily or be associated with other bone lesions,[16] such as ossifying fibroma, chondroblastoma, central giant cell lesions, osteosarcoma, and fibrous dysplasia.[14] In a literature review, the authors found that 22% of aneurysmal bone cysts were associated with another bone lesion.[18]

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Case report

The patient, a 9-year-old Caucasian girl, presented to the Clinic of Oral and Maxillofacial Surgery, Teófilo-Otoni, Minas Gerais, Brazil, with a left facial swelling that had been developing for approximately 6 months. There was nothing noteworthy in her family or past history. The lesion was asymptomatic and had rapidly increased in size since it was first noticed.

The extraoral examination showed a swelling and significant left facial asymmetry with disappearance of the nasolabial fold and elevation of the ala of the nose. The overlying skin was normal, and during examination of the neck, no abnormalities were found (Figure 1A).

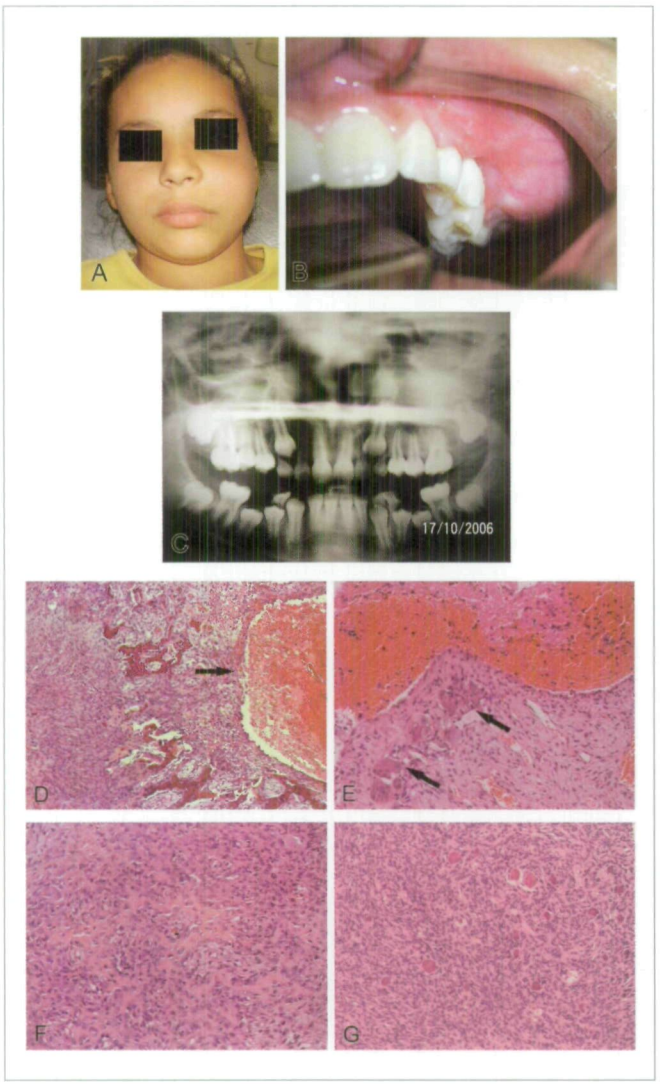

Figure 1. Clinical, radiological, and histopathological aspects upon initial presentation. (A) frontal view of the patient's face showing significant asymmetry. (B) In the intraoral aspect, swelling was observed in the left maxilla covered by normal mucosa. (C) A panoramic radiograph revealing a radiodense lesion with undefined limits and involvement of the maxillary left sinus. (D) Histopathological examination showing the association of the trabecular juvenile ossifying fibroma with the aneurysmal bone cyst (arrow; hematoxylin and eosin, 100X). (E) Aneurysmal bone cyst characterized by blood-filled spaces surrounded by multinucleated giant cells (arrows; hematoxylin and eosin, 400X). (F) Trabecular pattern of the juvenile ossifying fibroma (hematoxylin and eosin, 200X). (G) Psammomatoid pattern of the juvenile ossifying fibroma.

An intraoral examination revealed expansion of the left buccal cortical bone, which was firm and painless to palpation and extended from the maxillary left canine to the maxillary left second molar. There was no evidence of tooth mobility and displacement. The lesion was covered by normal mucosa with no bleeding or purulent discharge (Figure 1B).

A panoramic radiograph showed a radiodense lesion with undefined limits and involvement of the maxillary sinus. There was no evidence of tooth displacement or root resorption (Figure 1C).

An incisional biopsy was performed and the microscopic examination revealed a cellular connective tissue stroma rich in spindle and oval cells, arranged in a whorled or storiform pattern. Osteoid trabeculae and anastomosing immature bone trabeculae, rimmed with osteoblasts and incorporated osteocytes (trabecular pattern), were observed. A focal area with numerous concentric, laminated ossicles, with basophilic centers surrounded by eosinophilic osteoid peripheries and psammoma body-like structures, were also present (psammomatoid pattern). Blood-filled spaces and typical aneurysmal bone cysts were observed. Multinucleated giant cells, usually in association with cystic structures, were also present. Based on the clinical history and the lesion's radiographic and histologic features, a diagnosis of trabecular juvenile ossifying fibroma with central aneurysmal bone cyst was established (Figures 1D-G).

Conservative local excision of the lesion was performed, and the patient recovered well from the surgery but was lost to follow-up.

Three years after the first management, the patient returned to the same clinic presenting an increased left facial swelling, in the same anatomic site as that of the previous lesion, showing more significant facial asymmetry with displacement in the upward direction of the left orbit. The overlying skin and mucosa were normal. No other abnormalities were present (Figures 2A-C).

A computed tomography scan showed a unilateral, large, multilocular, well-demarcated, radiolucent, homogeneous mass with focal radiopacities, circumscribed by a thin cortical margin. The lesion expanded the anterior wall of the left maxilla and the temporal fossa and displaced the lateral wall of the nasal cavity, pterigoid bone, and floor of the left orbit in the upward direction, but there was no frank perforation of these anatomic structures (Figures 2D-H).

Under general anesthesia, curettage of the entire lesion was performed. The patient recovered well from surgery. Histopathological findings were similar to those of the first lesion, confirming the diagnosis of a recurrence of trabecular juvenile ossifying fibroma associated with an aneurysmal bone cyst. After 6 months of follow-up, no recurrence had been observed.

Discussion

A JOF diagnosis is based on essential characteristics, such as the early age of onset, location of the tumor, radiologic pattern, rapid growth, and tendency to recur.[14] Most of the clinical and histological features of the present case were

consistent with JOF, since our patient was a 9-year-old girl presenting with a large rapidly growing swelling in the maxilla.

Most JOF cases are asymptomatic, making the early diagnosis difficult, and the first clinical manifestation is frequently a swelling of the jaw bones.[2] TrJOF tends to affect the maxilla and is characterized by a progressive and sometimes rapid, aggressive growth.[7,11] The tumor is often a painless swelling and expands the affected bone, leading to facial asymmetry.[5] Depending on the site, symptoms such as pain, paresthesia, malocclusion, sinusitis, proptosis, etc, can also occur due to the swelling.[12,19]

Usually, JOF presents as an expansive lesion, but alternatively it may be diagnosed as a chance finding on a radiograph, demonstrating the importance of this procedure in children and adolescents. A well-defined lesion, partially or completely surrounded with thin, corrugated margins, is often found. The lesions are either radiolucent or show a mixture of radiolucency and radio-density.[5] It was observed that the tumor involved the left maxillary sinus. This agrees with the literature, which states that maxillary tumors tend to extend into and obliterate the maxillary sinus.[1,12]

Radiographic features are nonspecific and, coupled with the absence of clinical findings in premature lesions, early diagnosis is difficult. The differential diagnosis in these cases could be made mainly with fibrous dysplasia and osteosarcoma.[20] Histopathologically, the juvenile ossifying fibroma could be distinguished from ossifying fibroma by the earlier onset, locally aggressive growth, and osteoid trabeculae by histological examination.[21] Fibrous dysplasia can be ruled out, since it typically blends with the normal bone at the lesion's margins and is characterized by a less cellular stroma, and its bony trabeculae do not exhibit osteoblastic rimming.[2,11,14] The histologically benign aspects of this lesion ruled out osteosarcoma. The present case showed a lesion that was well demarcated from the surrounding bone and histologically, presented highly cellular fibrous stroma and prominent osteoblastic rimming, which were suggestive of JOF.

An interesting observation in the present case was that the 2 histological patterns were present within the same lesion, with very few cases reported.[7,15] The predominant component was the trabecular pattern with a focal area of the psammomatoid pattern. Moreover, areas of aneurysmal bone cyst within the lesion were observed.[5,15] Johnson et al.,[14] commented that large aggressive maxillary lesions were commonly associated with aneurysmal bone cyst formation.

Struthers and Shear[22] tested the hypothesis that the aneurysmal bone cyst is a secondary phenomenon, which occurs in a primary bone lesion. They postulated that the initiating process of the aneurysmal bone cyst is the microcyst, which is formed as a result of intercellular edema in a primary lesion with loose, unsupported stroma. Rupture of vessels in the microcysts introduces blood under hemodynamic pressure. With little resistance provided by the stroma, the blood spaces resorb the surrounding bone and lift the periosteum, which produces a thin shell of new bone.

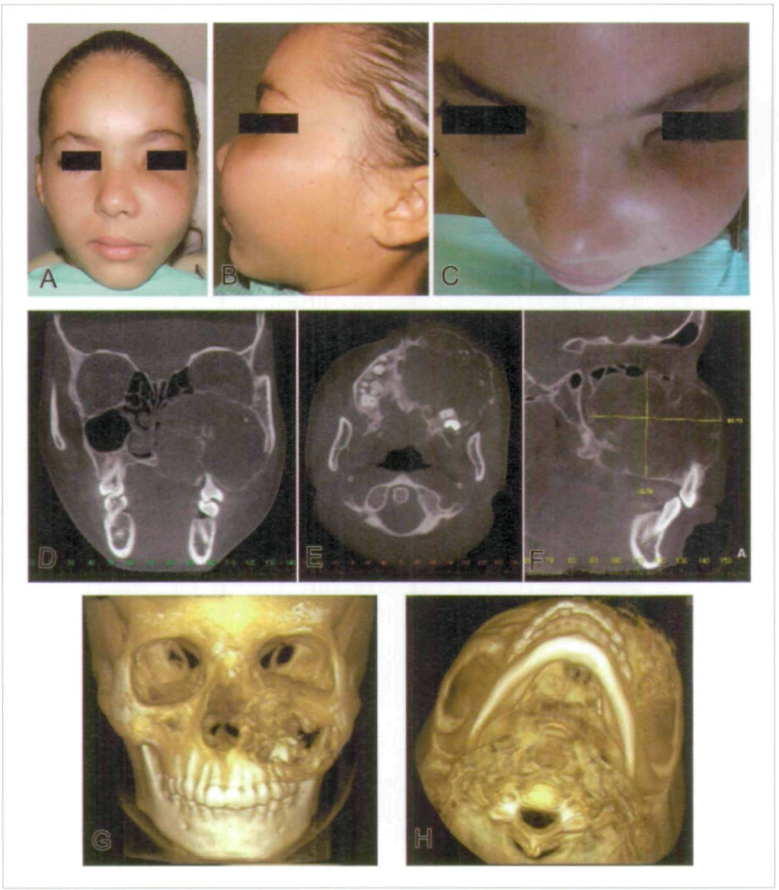

Figure 2. Clinical and radiological aspects 3 years following initial treatment. (A) Frontal view of the patient's face showing significant asymmetry and displacement in the upward direction of the left orbit. (B) Lateral view revealing a large swelling involving the left maxilla. (C) Significant asymmetry is observed in a superior view. (D-F) Computed tomography scan in a coronal, axial, and sagittal view of the lesion demonstrating a large expansile homogeneous mass, well-defined, and radiolucent with focal radiopacities involving the left maxilla, maxillary sinus, nasal cavity, and orbital floor. (G and H) Tridimensional reconstruction of the patient's face showing large involvement of the anatomic structures.

This behavior may be exhibited by both variants of JOF and is more commonly associated with younger patient age and the coexistence of an aneurysmal bone cyst.[14] It is unclear whether this increased aggressive behavior merely represents rapid enlargement of the aneurysmal bone cyst component causing displacement and resorption of adjacent structures or true aggressive behavior of the primary tumor itself.[15]

Development of aneurysmal bone cyst in PsJOF is more commonly reported than in TrJOF.[5] PsJOF and TrJOF seem to behave similarly, and their management is the same.[7]

It is critical to identify patients who have aneurysmal bone cyst associated with JOF, because the treatment plan should be dictated by the associated lesion rather than by the aneurysmal bone cyst, and the recurrence may be related to inadequate treatment of an associated primary lesion rather than the aneurysmal bone cyst per se.[18]

The lesion presented involved the left maxilla, maxillary sinus, nasal cavity, and orbital floor. A radical surgery did not seem to be an appropriate treatment solution. Therefore, local excision and thorough curettage were performed with maximum preservation of the normal skeletal and neurovascular structures.

In most of the medical literature, authors favor conservative surgery rather than radical surgery.[3,4,12] Some authors argue, however, that this lesion requires extensive surgery involving the adjacent bone,[2,8,13] because JOF's high recurrence rate is most likely caused by an incomplete excision resulting from the infiltrative nature of the tumor borders.[1,6,14] The size of the lesion and its relationship with the vital anatomical structures, however, do not allow for radical surgery. Due to JOF's characteristics, continued follow-up is essential.[3] The patient in this case was lost to follow-up, which did not allow early detection and management of the recurrent lesion.

The juvenile forms of ossifying fibroma are uncommon, and the association with aneurysmal bone cyst is a rare condition. Nevertheless, it is very important for surgeons to recognize and manage these lesions appropriately. Early detection and effective treatment with long-term follow-up are essential, considering their aggressive behavior and high recurrence rate.

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