CASE REPORT

Juvenile Ossifying Fibroma in a Juvenile Diabetic: A Case Report with Surgical Management

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Abstract

Juvenile ossifying fibroma (JOF) is a highly aggressive uncommon fibro-osseous lesion occurring in the facial bones. Two histological variants of JOF, i.e. trabecular and psammomatoid have been reported. Here we present a case of a 20-year-old juvenile diabetic patient with massive growth on her left side of the jaw which was diagnosed as juvenile ossifying fibroma. This huge aggressive mass in the left side of the mandible had lead to complete lingual tilting of molars and premolars. After clinical and radiological examination, a biopsy led to the histopathological diagnosis of mixed variant of juvenile ossifying fibroma. The required surgical treatment (hemimandibulectomy) was undertaken under GA. The aggressive local behavior and high recurrence rate of JOF emphasizes the need for an early diagnosis, and appropriate radical treatment with long-term follow-up. One such case is reviewed and discussed in detail in the following article.

Keywords: Juvenile ossifying fibroma, fibro-osseous lesions, aggressive ossifying fibroma.

INTRODUCTION

Fibro-osseous lesions are thought to be the result of diverse processes in which the normal bone architecture is replaced by fibroblasts and collagen fibers that contain various amounts of mineralized matrix. These lesions include a broad group of several entities like ossifying fibroma, Juvenile ossifying fibroma (JOF), fibrous dysplasia and so on.[1] Most of them have been considered as benign lesions, but JOF has been classified as a different disease because of its local aggressive behavior and its tendency to predominantly occur in children and adolescents.[2-4] Juvenile ossifying fibroma is a benign bone-forming neoplasm, and it is defined as a variant of ossifying fibroma occurring in the craniofacial skeleton of young patients.[2,5] It is known to have two distinct histological subtypes, that is, Juvenile trabecular ossifying fibroma (JTOF) and Juvenile psammomatoid ossifying fibroma (JPOF).[5,6] In this paper, we define specific, precise clinicopathalogical features, different modalities of management and the reasons for recurrence.

CASE HISTORY

A 20-year-old apparently healthy, cooperative female patient visited our department with a history of painless huge swelling on the left side of her face which started as small lump about 2 years back, but had shown a rapid increase in size over the last 1 year. Medical history revealed that she was a juvenile diabetic and was on systemic insulin (Inj. Biphasic), once a day since the last 10 years. There was no history of extraction, pain, paresthesia or discharge of pus.

Extraorally, a huge swelling had led to gross facial asymmetry. The swelling measured approximately 15 × 8 cm extending anteriorly from the central portion of lower lip till the left ear lobe posteriorly, superiorly extending till the

Fig. 1A: Frontal view

Fig. 1B: Lateral view

Fig. 1C: Shows extension of mass inferiorly

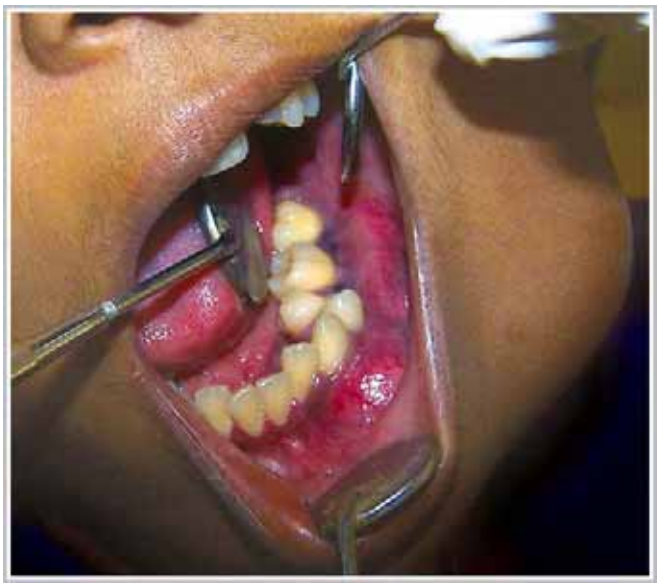

Fig. 2: Intraorally tilting of molars noted

ala tragus line and inferiorly 4 cm below the lower border of mandible (Figs 1A to C). The skin overlying the swelling appeared stretched. No discharging sinus or scars were noted extraorally. On palpation it was nontender, immobile, and hard in consistency with no rise in overlying skin temperature. Intraorally (Fig. 2) there was a diffuse swelling which was extending from lower left central incisor to retromolar area leading to expansion (i.e. from 31 to 37 area with 38 clinically missing) of buccal and lingual cortical plates with obliteration of buccal sulcus. The teeth from 35 to 37 were totally lingually tilted. None of the teeth were mobile. With above history and clinical findings the provisional diagnosis of fibro-osseous lesion was made with a differential diagnosis of ameloblastoma and dentigerous cyst. The patient was advised to undergo orthopantomography, PA skull and lateral skull views and CT (computed tomography) along with RBS and routine hemogram with relevant serum biochemical analysis. Orthopantomograph, PA skull and lateral skull showed a expansive lesion, partially or completely surrounded by thin corrugated margins extending from 31 region till ramus area leading to destruction of the surrounding bony structures (Figs 3A to C). Root resorption was noted with 36, 37. On the same side 38 was impacted and was completely displaced into the ramus. Multiple septae running throughout the lesion with subtle radiopaque flecks were noted. The pressure exerted by this huge mass had led to the compression of the upper left jaw with intrusion of upper left quadrant teeth with displacement of 28. The whole lesion showed

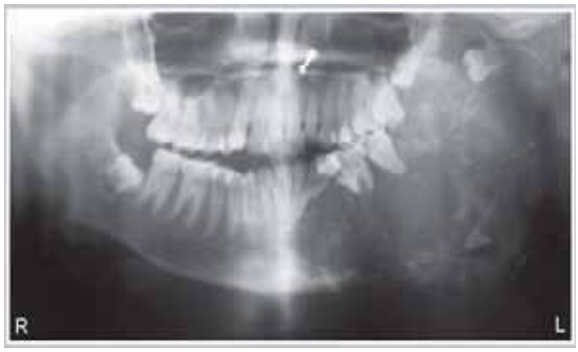

Fig. 3A: OPG shows multiple septae at the centre of lesion with displacement of 38

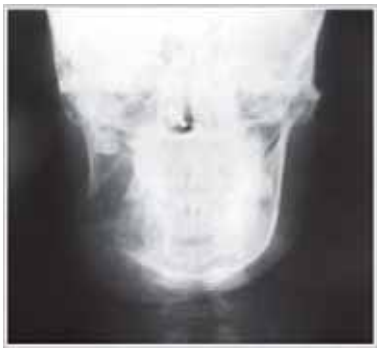

Fig. 3B: PA skull showing extension of lesion

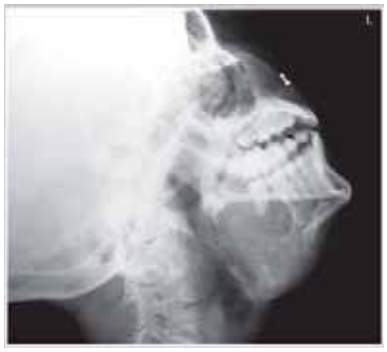

Fig. 3C: Lateral skull showing the extension

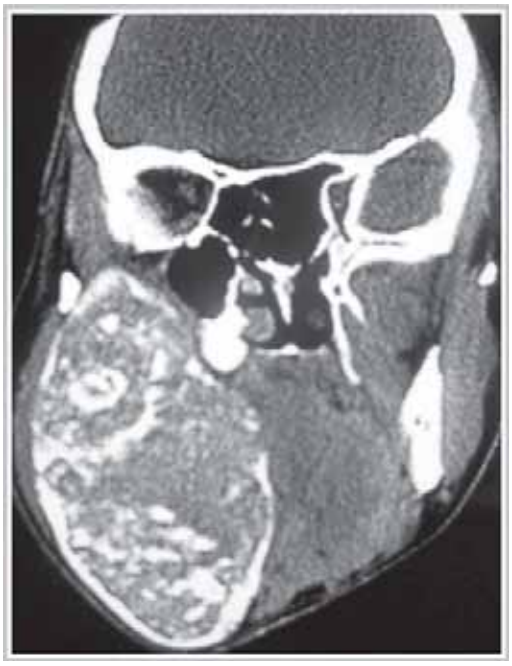

Fig. 4A: Coronal CT showed an exapnsile mass with multiple areas of homogeneous hyperdensity, suggestive of mineralized matrix

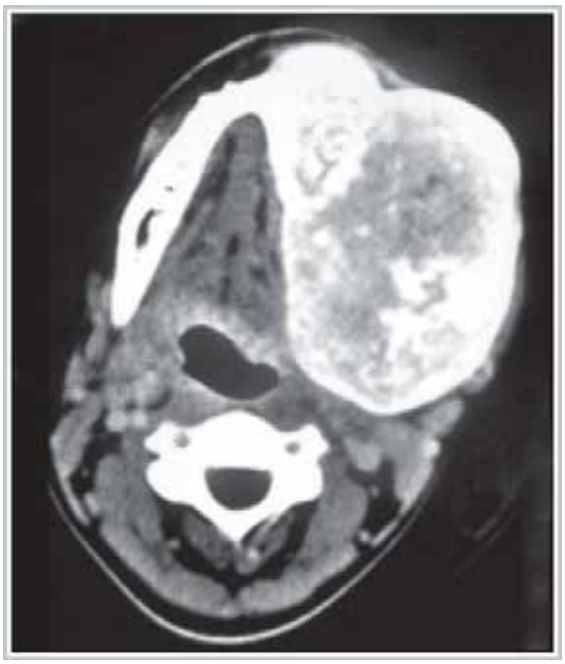

Fig. 4B: Axial CT showed, expansile, osteolytic lesion with gross expansion of both cortical plates

centrifugal growth pattern with downward bowing of the inferior cortex of the mandible.

CT scans, PNS view demonstrated well-defined, expansile, mixed radiolucent and radiodense mass in the left mandible (Figs 4A and B). Coronal CT showed an expansile mass with multiple areas of homogeneous hyperdensity, suggestive of mineralized matrix. Axial CT showed, expansile, osteolytic lesion with gross expansion of both cortical plates. Postprandial blood glucose level checked 4 weeks earlier was 311 mg/dl. The blood glucose level was brought under control by doubling the dosage of systemic insulin for 1 week. An incisional biopsy showed a well-circumscribed, capsulated mass. The capsule is fibrous and is thinned out at places. The subcapsular area comprised of interconnected bony trabeculae encasing plump osteocytes. Most of the trabeculae exhibit osteoblastic rimming. Concentric lamellated and spherical ossicles are seen in one area with basophilic center and peripheral eosinophilic rims. The fibrous tissue is of varying cellularity with a few chronic inflammatory cells. Small clusters of giant cells were also evident. All these findings have led to a final diagnosis of mixed variant of juvenile ossifying fibroma (Figs 5A to C). The patient was then subjected to a hemimandibulectomy under GA where the affected portion

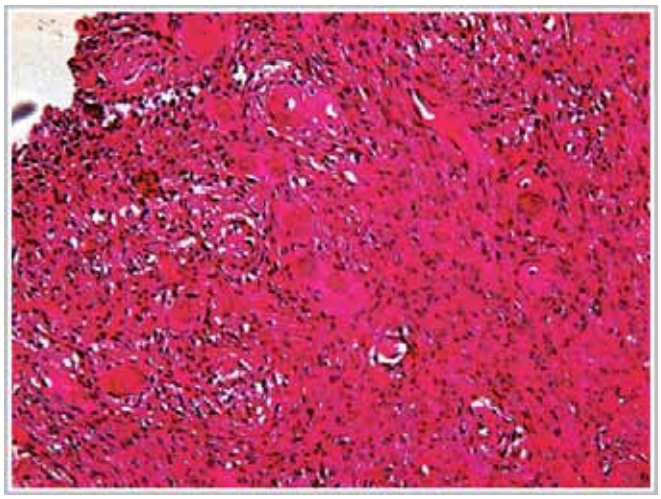

Fig. 5A: Cellular fibrous stroma with psammoma like bodies(H&E, X100)

Fig. 6: Gross specimen after hemimandibulectomy

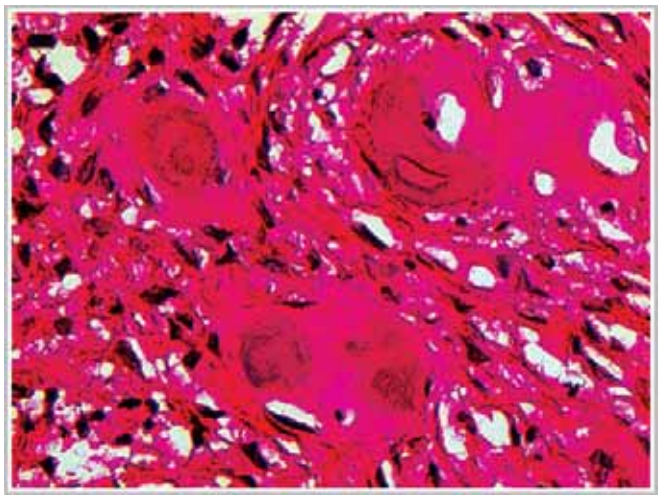

Fig. 5B: Psammoma like ossicles or cementicles (H&E, 400)

Fig. 7: Postoperative photograph

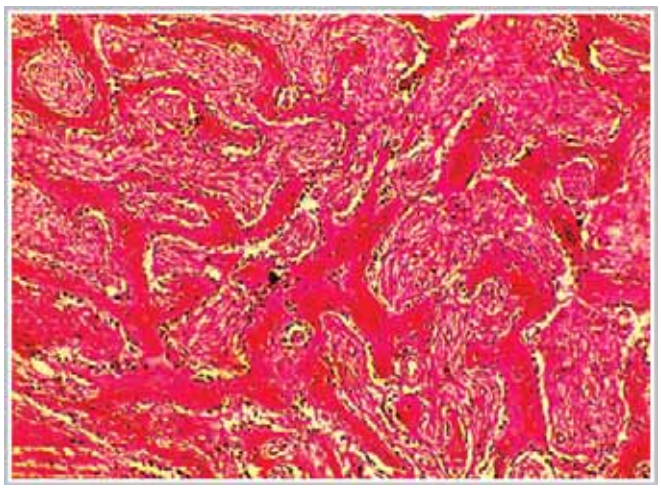

Fig. 5C: Bony trabeculae with osteoblastic rimming interspersed with fibrous stroma (H&E X100)

of the mandible was resected by using modified Aproan incision under general anesthesia. The huge mass resected gross specimen (Fig. 6) sent for histopathological examination confirmed the diagnosis of juvenile ossifying fibroma. Ten months systematic follow-up did not reveal any recurrence (Fig. 7).

DISCUSSION

The main characteristics of juvenile ossifying fibroma are the early age of onset, localization of the tumor, typical radiological pattern (Centrifugal growth with radiopaque flecks) and a tendency to recurrence. As the term "juvenile" underlines, the tumor largely develops in children, 79% of whom are under 15 years old.[7] In reviews published by

Hamner et al[8] and Slootweg et al.[9] The mean age of onset was 11.5 and 11.8 years, respectively. Our present case reported belonged to a higher age group. Authors differ in their reports on the location of the lesion. Slootweg et al.[9] and Makek[10] described the maxilla as the most frequent site, with Johnson et al[11] even reporting that 90% of facial bone fibromas occur in the paranasal sinuses and only 10% in the mandible. On the other hand, Hamner,[8] Leimola et al,[12] Brannon and Fowler[13] and Sánchez Cuéllar et al[14] reported a mandibular predominance. The case described here involved the mandible. According to most authors, there is no predilection for either sex, although Johnson et al[11] found a higher incidence in females.

Most cases of JOF are asymptomatic with a tendency of aggressive growth, as shown by Hall et al[15] and Zupi et al,[16] and also tendency to recurrence, ranging from the 30% rate observed by Johnson et al[11] to the 58% rate reported by Makek.[11] The tumor expands the affected bone, leading to facial asymmetry.[17,18-23,24-28] Pain and parasthesia are rarely manifested. Root resorption and displacement of involved teeth are observed.[17,23] In the mandible, the angle and ramus are more commonly the site of involvement.[23-25,29]

The cells of the periodontal ligament are able to generate a great variety of benign neoplasms. The lesions generally appear in dentate zones of the upper and lower jaws. In some cases the neoplasms derive from ectopic periodontal ligament cell remains, and can manifest in unusual locations. When present in such ectopic locations, these lesions can become expansive or aggressive, with the need for surgical management.[30]

Type I DM is a immune mediated chronic disease characterized by cellular-mediated autoimmune destruction of the insulin producing beta cell in the pancreatic islets.[31] The genetic differences in cranial fibro-osseous lesions are not well-established, but some genetic studies have been done. Nonrandom chromosomal[32] break points at Xq26 and 2q33, resulting in (X; 2) translocations were identified in some cases of JOPF. However, no evidence of genetic correlations between these two have been recorded.

The treatment[33] of JOF consists of a simple excision. Because they are well-differentiated lesions, they are not radiosensitive and radiotherapy is contraindicated because it can induce malignant change. The prognosis of JOF is uncertain. Although many tumors demonstrate slow but progressive growth, some JOF demonstrate rapid enlargement. The more aggressive neoplasm tends to arise in infants and young children. For smaller lesions, complete local excision or thorough curettage appears adequate. For some rapidly growing lesion, wider resection may be required. Local recurrence is likely if the tumor is not completely removed, although it can also be caused by dysplastic processes in the bone metabolism. Nevertheless, it is important to perform a clinical and radiological follow-up for as many years as possible, because of the possibility of recurrence in this type of new bone formation. Noffke[34] carried out a radiological follow-up of a mandibular JOF and found, at eight years of its enucleation, an aneurysmal bone cyst had developed with reduction in the bone content. Radiographically, demarcation of the tumor from the surrounding bone is well-defined by a radiopaque border, and this characteristic is important in the differential diagnosis between JOF and fibrous dysplasia, because the latter presents a radiographic image with a diffuse border. The radiolucency of the lesion varies, depending on the maturation stage and amount of calcification.[35] In the follow-up orthopantomography performed at one and two years, a favorable clinical evolution of the patient was observed, given that the permanent molars had erupted and no radiolucent angles where the JOF and radicular cyst had been enucleated. Of 112 cases reviewed by Johnson et al,[36] 30% recurred one or more times. Recurrence may be attributed to difficulty in proper resection caused by the location of the lesion and the infiltrative nature of tumor borders.[36] Trabecular JOF is characterized by a progressive and sometimes rapid aggressive growth. A minimum 5-year follow-up of these patients is essential.

The purpose of this article was to present a clinical case of JOF and to analyze its clinical, radiological and histological characteristics, in order to assist us to distinguish this clinical entity from other fibrous lesions and carry out the appropriate treatment.

CONCLUSION

Although juvenile ossifying fibroma is an uncommon clinical entity, its aggressive local behavior and high recurrence rate emphasizes the need for an early diagnosis, complete wide surgical excision and a minimum follow-up period for five years.

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