Trabeculae type of juvenile aggressive ossifying fibroma of the maxilla: Report of two cases

Shalu Rai, Mandeep Kaur[1], Sumit Goel[2], Mukul Prabhat

Abstract

Juvenile ossifying fibroma (JOF) is a rare controversial fibroosseous lesion affecting the craniofacial skeleton and occurring commonly in children and young adults. It is highly aggressive and has a high tendency to recur. It is distinguished from the adult variant of ossifying fibroma on the basis of age, site, clinical behavior and microscopic appearance. Because of its high recurrence rate, which is 30–58%, complete excision is essential. Early diagnosis will circumvent the necessity of radical treatment. We report a rare case of trabecular JOF of maxilla where a computed tomography scan was taken to further support the characteristic feature of the lesion.

Keywords: Aggressive, CT scan, juvenile ossifying fibroma, trabecular variant

Introduction

The fibrous osseous lesions of the jaw represent a diverse group of entities, and aree characterized by replacement of normal bony architecture by fibrous connective tissue matrix with varying degrees of osteoid, immature and mature bone.[1]

The second edition of the WHO classification of odontogenic tumor defines juvenile ossifying fibroma (JOF) as a lesion consisting of cell-rich fibrous tissue containing bands of cellular osteoid without osteoblastic rimming with trabeculae of more typical woven bone.[2] The lesions having this morphology have been reported as young ossifying fibroma, juvenile active ossifying fibroma, aggressive ossifying fibroma, trabecular desmo-osteoblastoma and active fibrous dysplasia.[2-4] JOF occurs predominantly but not exclusively in children. It affects males and females equally without any gender predilection. The maxilla is more commonly affected than the mandible.[5,6] Although it can occur anywhere in the skeleton, its highest incidence is in the facial bone.[4-6] It can expand the involved bones, leading to facial asymmetry. Depending on the site, symptoms such as pain, paresthesia, malocclusion, sinusitis, proptosis, etc. can also occur due to the swelling.[6-8] Root displacement is common and resorption, although rare, can occur.[6-8] The lesion can cause expansion as well as perforation.[6] Radiographically, they appear radiolucent, radioopaque or mixed radiolucent–radioopaque with a well-defined sclerotic border.[3-5,7]

Two histopathologic variants of ossifying fibroma of craniofacial skeleton have been described – trabecular juvenile ossifying fibroma (TrJOF) and psammomatoid juvenile ossifying fibroma (PsJOF).[5,6,9]

Department of Oral Medicine and Radiology, Institute of Dental Studies and Technologies, Kadrabad, Modinagar, Uttar Pradesh, [1]Dental College, Jamia Milia Islamia, New Delhi, [2]Subharti Dental College, Meerut, India

Correspondence: Dr. Shalu Rai, Department of Oral Medicine and Radiology, Institute of Dental Studies and Technologies, Kadrabad, Modinagar - 201 201, Uttar Pradesh, India.
E-mail: drshalurai@gmail.com

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Case Reports

Case 1

A 15-year-old female patient reported to our Out Patient Department (OPD) with a painless swelling on the right side of the face since 2 1/2 years. The patient related this swelling to extractions done in that region 2 years back at a private clinic. The swelling had gradually grown to its present size and was asymptomatic. No contributory medical or family history was found.

On general physical examination, the patient was conscious, cooperative, healthy and thin built and well nourished. All vitals were within the normal range. Extraoral examination showed facial asymmetry due to a diffuse swelling on the right side of the face [Figure 1]. The swelling measured approximately 2 cm x 3 cm in size and extended from the ala of nose till just in front of the tragus of the right ear. Superoinferiorly, it extended from the right infraorbital region to 2–3 cm short of the inferior border of the mandible. Skin over the swelling and the surrounding area appeared normal. The swelling was hard in consistency.

Intraoral examination revealed a solitary, well-defined swelling obliterating the vestibule and extending from the right maxillary canine region (13) to the right second molar region (17) posteriorly [Figure 2]. Superoinferior borders could not be evaluated. On palpation, the swelling was hard and tender, causing expansion of the buccal cortical plates.

A provisional diagnosis of monostotic fibrous dysplasia of right maxilla was made, with the differential diagnosis of ossifying fibroma. Hematological findings were unremarkable and serum alkaline phosphatase levels were within normal limits. Radiological investigations included orthopantomogram, maxillary lateral occlusal projection, paranasal (PNS) sinus view and computed tomography (CT) scan. Orthopantomogram revealed a mixed radiolucent–radioopaque lesion in the right maxillary premolar and molar regions giving a ground glass appearance [Figure 3]. PNS view showed an ill-defined radioopacity occupying the floor of the maxillary sinus [Figure 4]. CT scan showed a hyperdense focal expansile area involving the right maxillary bone, premolars and molars, compressing the lateral wall of the right maxillary sinus with thinning of the cortex [Figures 5].

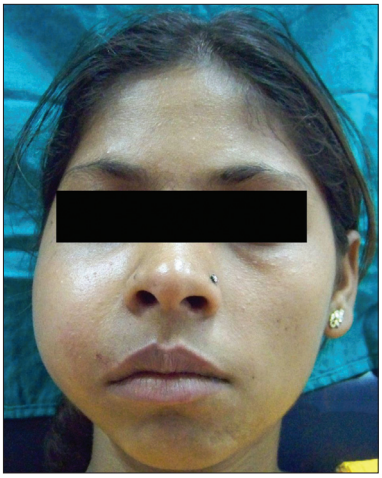

Figure 1: Facial asymmetry due to a diffuse swelling on the right side of the face extending from the ala of nose to posterior in front of the tragus of the right ear

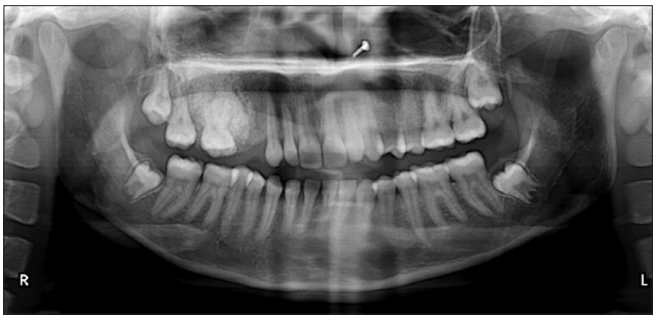

Figure 3: Orthopantomogram showing a mixed radiolucent–radioopaque lesion in the right maxillary premolar and molar region giving a ground glass appearance

The patient was operated under general anesthesia. Because the lesion was slowly growing, a recontouring procedure was performed involving multiple osteotomies. Multiple excised tissues were sent for histopathological examination.

Hematoxylin and Eosin (HandE)-stained sections showed proliferative cellular connective tissue stroma encasing plump and irregular cells [Figure 6a], comprising of mineralized material in the form of trabeculae of woven bone [Figure 6b]. The trabeculae did not show osteoblastic rimming [Figure 6c]. The histologic features were suggestive of trabecular variant of JOF. Post-operative healing was uneventful [Figure 7] and the patient is under regular follow-up considering the high recurrence rates of this neoplasm.

Case 2

A 15-year-old male patient reported to our OPD with the chief complaint of a gradually increasing asymmetry of the face. There was no history of trauma pain, lymphadenopathy,

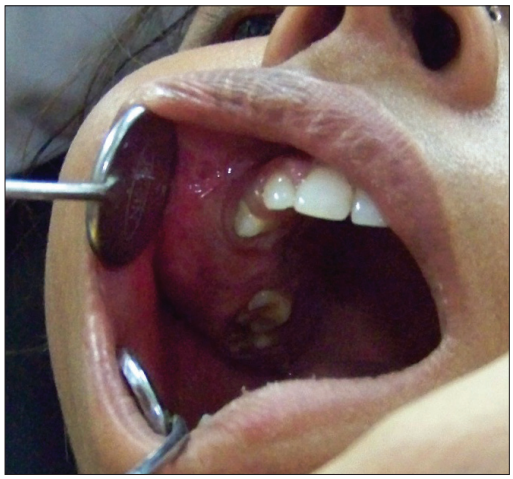

Figure 2: A solitary, well-defined swelling obliterating the vestibule and extending from the right maxillary canine region (13) to the right second molar region (17)

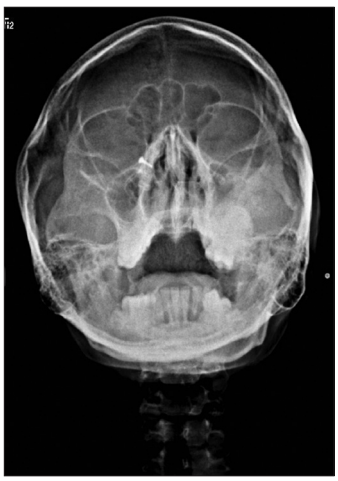

Figure 4: Paranasal view showing an ill-defined radioopacity occupying the floor of the maxillary sinus

paresthesia or dysphagia. He had no significant medical, dental or family history. Clinical examination revealed a moderately large left facial mass over the left molar region [Figure 8]. Intraoral examination revealed gross expansion of the left maxillary alveolar process extending from the 12 region to the 17 region posteriorly, obliterating the buccal vestibule [Figure 9]. On palpation, the swelling was hard and tender causing expansion of the buccal cortical plates. Radiological examination revealed an ill-defined, radioopaque lesion. Maxillary cross-sectional occlusal radiograph revealed expansion of the cortical plates [Figure 10]. HandE-stained sections showed a similar histopathological picture as in Case 1 [Figures 11 and 12], and were again suggestive of trabecular variant of JOF. The lesion was treated by complete surgical excision.

Discussion

JOF is an uncommon benign fibroosseous tumor occurring within facial bones in 85% of the patients. The calvarium is involved in 12% and 4% of the cases as seen extracranially. These lesions are generally more defined, but may displace the teeth and invade the adjacent bone.[10] Minority of the cases, particularly in children (below 15 years), exhibit rapid growth and a tendency to recur and thus have been named JOF. Gender predilection has been a matter of controversy, with some authors claiming no predilection for either sex, whereas Johnson *et al*.[11] found a higher incidence in females[6,11] and El-Mofty reported a male predilection.[9]

JOF has been thought to arise from differentiation of mesenchymal cells of Periodontal Ligament, multipotent precursor cells, forming into fibrous tissue, cementum or osteoid.[5] Lawton *et al*.[12] suggested that they perhaps originate from maldevelopment of tissue generating bony septa, between roots of the molar teeth. Association of new tumor suppressor gene (HRPT2) mutation has been reported by Pimenta *et al.,*[13] suggesting that the lesion could arise as a result

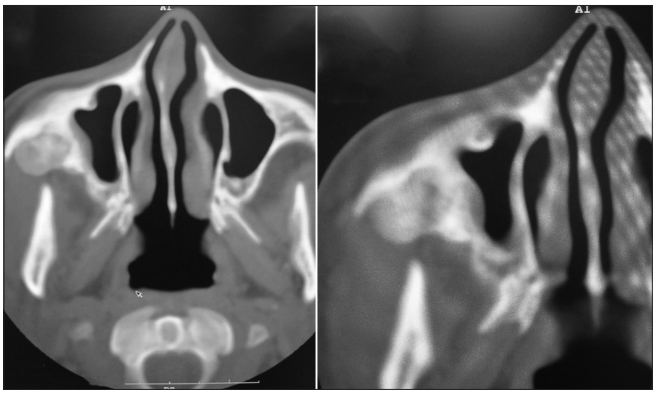

Figure 5: Axial computed tomography scan showing a hyperdense focal expansile area involving the right maxillary bone, premolars and molars compressing the lateral wall of the right maxillary sinus with thinning of the cortex

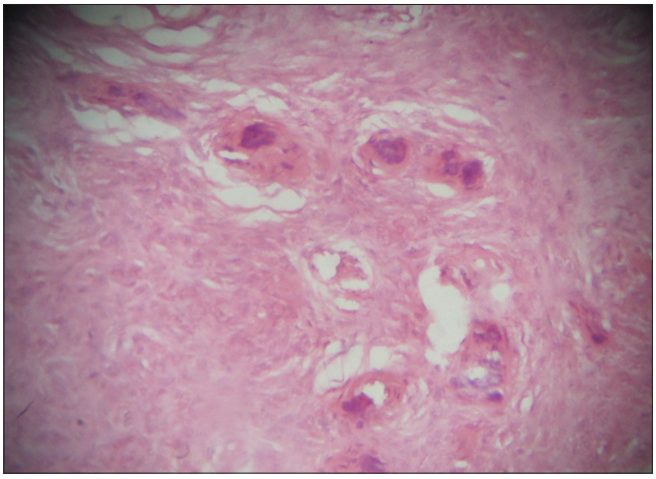

Figure 6b: Hematoxylin and Eosin-stained section (×10) comprising of mineralized material in the form of trabeculae of woven bone

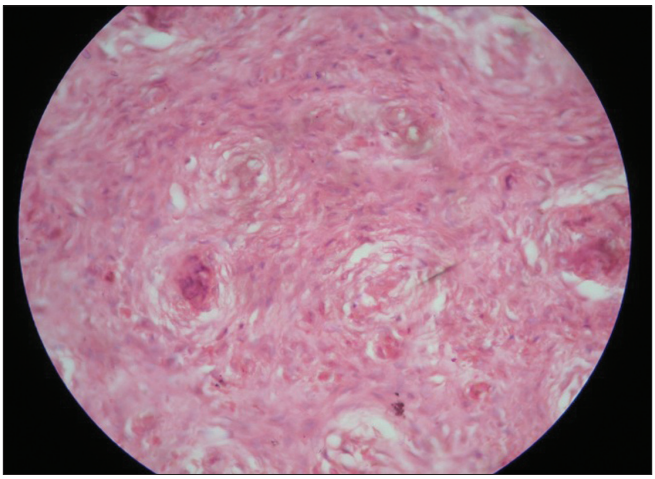

Figure 6a: Hematoxylin and Eosin-stained section (×10) showing proliferative cellular connective tissue stroma encasing plump and irregular cells

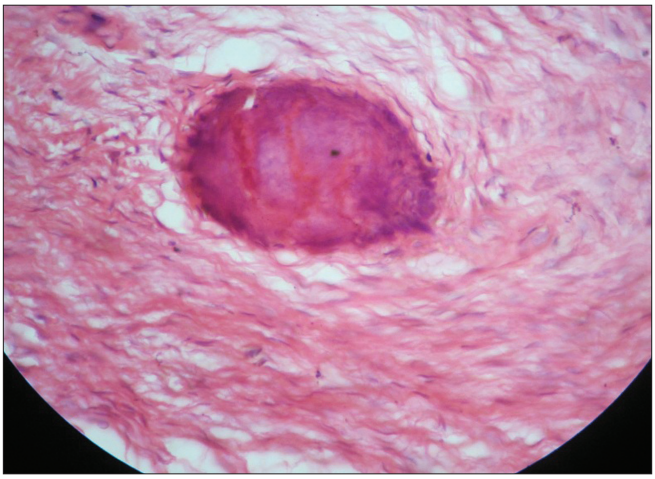

Figure 6c: Hematoxylin and Eosin-stained section (×40) showing trabeculae without osteoblastic rimming

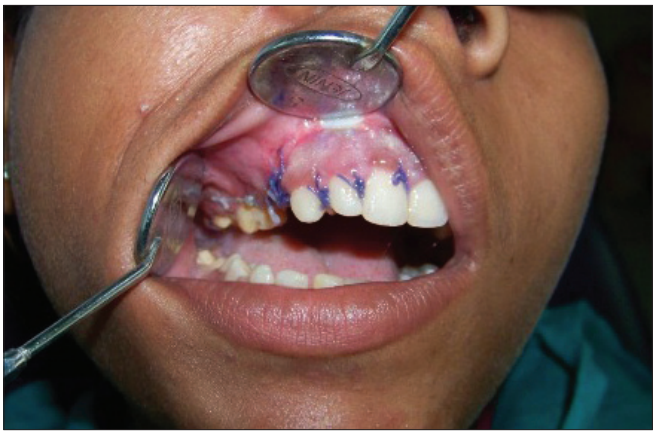

Figure 7: Post-operative picture of the patient

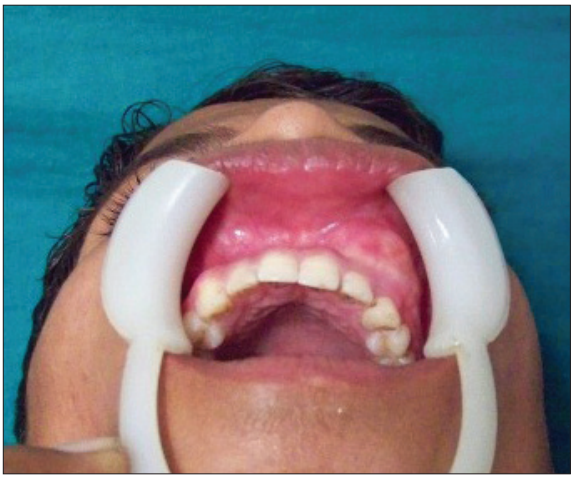

Figure 9: Gross expansion of the left maxillary alveolar process extending from the 12 region to the 17 region posteriorly, obliterating the buccal vestibule

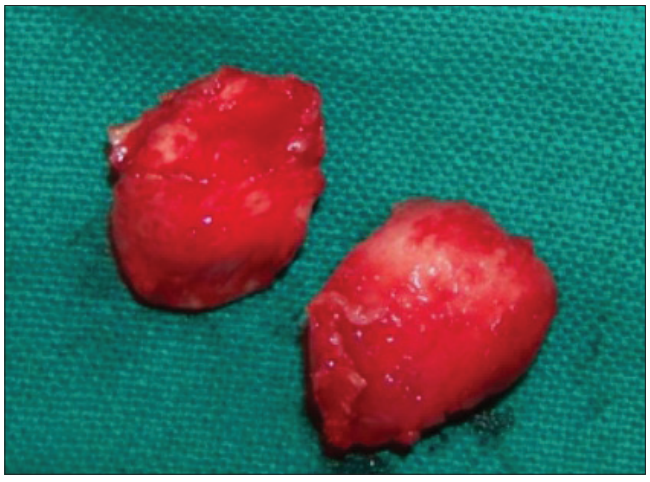

Figure 11: Two bits of excised gross specimen sent for histopathological examination

of heploinsufficiency of the particular gene. Because of its distinct histological features, it has been recognized as a separate histopathological entity among the fibroosseous group of lesions.[2,4,5,9]

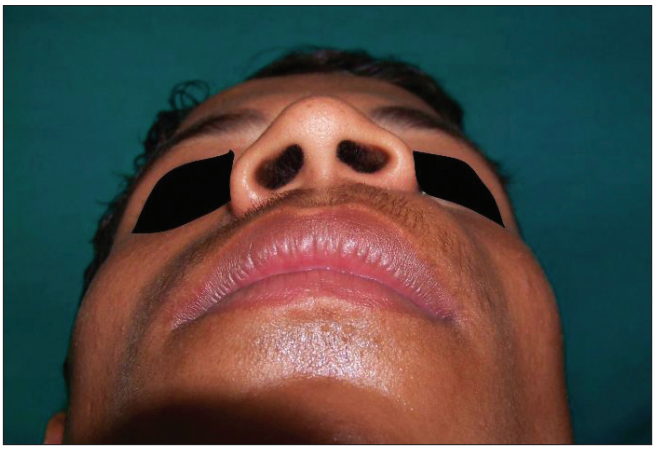

Figure 8: A moderately large left facial mass over the left molar region

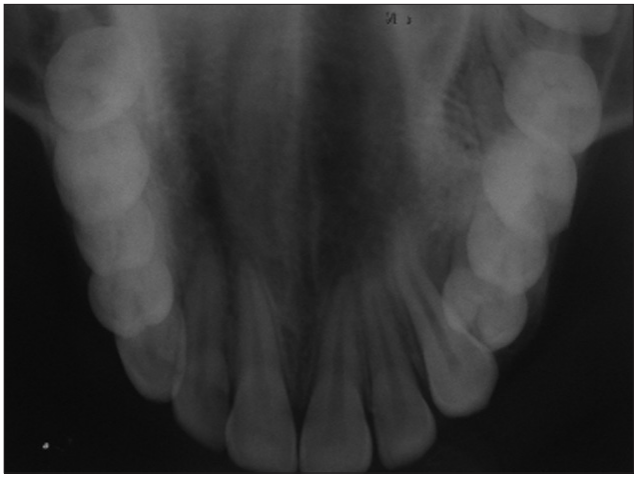

Figure 10: Maxillary cross-sectional occlusal radiograph showing cortical plates expansion

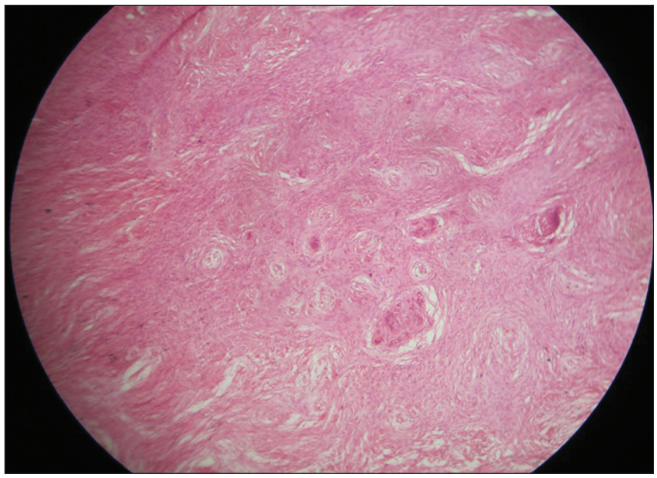

Figure 12: Hematoxylin and Eosin-stained section (×10) showing proliferative cellular connective tissue stroma encasing plump cells comprising of mineralized material in the form of trabeculae of woven bone without osteoblastic rimming

EL Mofty[9] in his study identified two histopathological variants of JOF – TrJOF and PsJOF.

This trabecular variant of JOF was previously used by Reed and Hagy in 1965 under JOF, who reported two cases – one in the maxilla and the other in the mandible.[9] Makek in 1983[14] published the largest series of tumor to date and reviewed 24 cases, 15 from the world literature and nine of his own, from files of the Zurich University Hospital and called the tumor trabecular desmo-osteoblastoma. He showed a slight male predilection (1.3:1), with maxilla being more commonly affected. Slootweg *et al*.[2] reported 10 cases showing a mean age of 11.8 years, again showing a male dominance, with 60% of the cases occurring in the maxilla. Williams *et al*.,[15] Dehner,[16] and Slootweg and Miller[17] reported a few more cases of TrJOF. Both our cases occurred in the maxilla, one in a male patient and the other in a female patient.

TrJOF is characterized by progressive and sometimes rapid aggressive growth. It might expand the facial bone, causing facial asymmetry as seen in the present cases. After reviewing the literature, it was concluded that the patients' age ranged from 2 to 33 years, with an average age range of 8 1/2 to 12 years and with a male dominance.[9] In the maxilla, the tumor may cause nasal obstruction, epistaxis and eye displacement. Pain and paresthesia are rarely manifested. Not all cases are characterized by a fast growth rate. Slow growth has also been reported in a few cases.[4-6,9]

Histopathological examination of the TrJOF shows a well-defined but encapsulated lesion that infiltrates the surrounding bone. It is mainly composed of cell-rich fibroblastic spindle cell stroma. Osteoid matrix may develop encorporating plump eosinophilic osteoblastic cells. Progressive calcification of the osteoid results in anastomosing trabeculae of immature woven bone, as seen in both our cases. Multinucleated giant cells are commonly seen. Collagenization might be seen in older lesions. Cystic degeneration and ABC formation have been described in a few cases.[2,4-6,9]

Presence of proliferating cellular connective tissue with plump cells is suggestive of the aggressive nature of this neoplasm,[2,4,5] as seen with the present two cases.

The number of cases reported of TrJOF is far less than that of PsJOF. Both the tumor illustrate differences in clinical and dermograpic presentation as shown in Table 1 (adapted and modified).[9]

Radiographically, TrJOF shows unilocular or multilocular radiolucency with a variable degree of calcification manifesting as fine specks and occasionally producing "ground glass appearance." Increase in radiodensity may be observed over a period of time. The tumor may reveal cortical thinning and perforation with root resorption and displacement of involved teeth.[5-7]

CT findings depend on the stage of development and amount of mineralized matrix present. CT attenuation levels have been reported to range from 34 to 513 Hounsfield (HU) depending on the fibrous tissue and bone content.[18] CT findings have been described in only two reports in the radiology literature and in few surgical reports. Areas of low-CT density may be noted due to cystic changes. Following IV injection of iodinated contrast, the lesion may show diffuse enhancement. On CT, the main differential diagnosis would be adult form of ossifying fibroma, fibrous dysplasia and cementoosseous dysplasia.[19]

JOF will show CT changes similar to the conventional form, but, sometimes, may appear more aggressive with cortical destruction. The adult form may show more sclerotic components. Fibrous dysplasia has a typical ground glass appearance, and expands the bone throughout its length and has poorly defined borders that blend with the surrounding normal bone. Cementoosseous dysplasia (apical, focal and florid) is a self-limiting localized lesion present exclusively in the tooth-bearing area of the jaws.[19,20] CT greatly increases the diagnosis and treatment plan by giving an accurate site, extent of the lesion and size of the tumor mass.

On magnetic resonance imaging, ossifying fibroma appears to be heterogenous. They reflect an intermediate signal intensity on T1-weighted imaging and an hypointense signal on T2-weighted imaging with moderate enhancement following IV administration of contrast on T1-weighted imaging.[19]

Fibroosseous lesions present a diagnostic dilemma owing to overlapping clinical and histopathological findings. A differential diagnosis of fibrous dysplasia, malignant bony tumors, aneurysmal bone cyst, central giant cell granuloma, osteogenic sarcoma, osteoblastoma, Calcifying Odontogenic cyst, Adenomatoid Odontogenic Tumor and primordial cysts

Table 1: Differences in clinical and dermograpic presentations of Psammomatoid juvenile ossifying fibroma (PsJOF) and Trabecular juvenile ossifying fibroma (TrJOF) (adapted and modified).[9]

Type	Site	Age	Mean age	Gender		Histopathology
Psammomatoid juvenile ossifying fibroma	More common in sinonasal and orbital bone	3 months 72 years	16–33 years	M>F 1.2:1	More common	Small uniform spherical ossicles resembling psammona bodies
Trabecular juvenile ossifying fibroma	Maxilla>mandible In mandible, body>ramus	2–12 years	8 1/2–12 years	M>F	Less common	Trabeculae of fibrillar osteoid and woven bone

(keratocyst) should be considered.[4,5,20] Fibrous dysplasia can be ruled out as it typically blends with normal bone at the margin of the lesion and has less cellular stroma, and its osteoid does not exhibit osteoblastic rimming and large amount of lamellar bone is found rather than woven bone.[5,18,19] Malignant bony tumors can be excluded by histological examination. Central giant cell granuloma occur more commonly in the anterior mandible, resulting in generally painless expansion of bone and appearing radiographically as unilocular or multilocular radiolucent defects with well-delineated, non-corticated margins. JOF can often be differentiated from osteosarcoma on the basis of radiographic appearance. The radiographic features of osteosarcoma are orthoradial striations, destruction of cortices with an outgrowth of the soft tissue component, generalized widening of the periodontal ligament spaces and destruction of the lamina Dura.[4] Vascular tumors like central hemangioma occur mainly in children and young adults and should also be taken in the differential diagnosis of JOF. AV malformations also exhibit rapid growth, but usually display thrills or bruits on examination.[4,5,19,20]

The clinical management and prognosis of JOF is uncertain. Treatment protocol ranges from simple curettage and curettage with peripheral ostectomy to block resection and segmental resection of mandible. Bone grafting has been used in most severe cases.[3,6,8]

Radiotherapy has been proven ineffective and contraindicated due to the increased incidence of malignant transformation ranging from 0.4% to 0%. Despite the aggressive behavior, no metastasis has been reported.[21]

Because of the aggressive nature of this entity and its high recurrence rate (30–50%), JOF should be treated like a locally aggressive neoplasm, very much like an ameloblastoma. Surgical resection, rather than conservative curettage, is therefore the preferred line of treatment.[3,6,8]

Conclusion

JOF is a rare fibroosseous neoplasm found in the young age group, which is considered locally aggressive than the conventional form and spreads quickly. Therefore, it is important to diagnose the lesion early and correlate all available clinical, radiological, CT scan and histologic data for better management.

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