Juvenile Ossifying Fibroma of Maxilla: Report of a case

Manish Juneja * / Mala Kamboj **

Juvenile ossifying fibroma (JOF) is a rare type of fibro osseous lesion affecting the jaws of children under 15 years of age. We report a case of an 8–year–old boy diagnosed with trabecular form of JOF affecting the left maxilla with brief description about its histopathological differential diagnosis.
Keywords: *Fibro-osseous Lesion, Juvenile Ossifying Fibroma, Trabecular type, Maxilla.*
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INTRODUCTION

The term juvenile (aggressive) ossifying fibroma (JOF) was used in the second edition of the World Health Organization classification of odontogenic tumors to describe a lesion which affected the jaws of children under 15 years of age. Histopathologically, it bears a close resemblance to other fibro osseous lesions of the craniofacial region and is characterized by hypercellular fibrous connective tissue containing bands of cellular osteoid, trabeculae of bone, and aggregates of giant cells.[1] Clinically, the tumor is more aggressive and has a much higher rate of recurrence when compared to other fibro osseous lesions.[2] The tumor is generally localized and well demarcated, but is not encapsulated.[1] The aggressive behaviour, and high recurrence rate mandates its early recognition and complete surgical excision. We hereby report a case of an 8-year-old boy diagnosed with trabecular form of JOF affecting the left portion of maxilla.

CASE REPORT:

An 8-year-old boy presented with the chief complaint of painless swelling of the face causing facial deformity since last 8 months. Clinical examination revealed a huge mass on the left side of the face. Oral examination revealed a normal colored, smooth surfaced, round swelling of approximately 10 x 8 cms in size in the left maxillary region causing gross expansion of the buccal plates and palate. On palpation the swelling was hard and non tender with loosening of teeth in the associated region. No nodes were palpable on extra oral examination. There was no relevant family and medical history.

CT scan of the mid face revealed a large expansile, erosive mass in the left maxillary sinus region with multiple foci of calcifications. Anteriorly, the mass was seen infiltrating the overlying cheeks and adjacent gingivo-buccal sulcus. Superiorly, the mass had caused considerable elevation of the orbit with no extensions into it. An incisional biopsy was performed from the gingivo-buccal sulcus region. The histopathological examination revealed features of fibro-osseous lesion with areas of osteoid and woven bone formation and occasional areas showing giant cells. Keeping in mind the aggressive nature of the lesion and younger age of the patient, a segmental resection of the left maxilla was

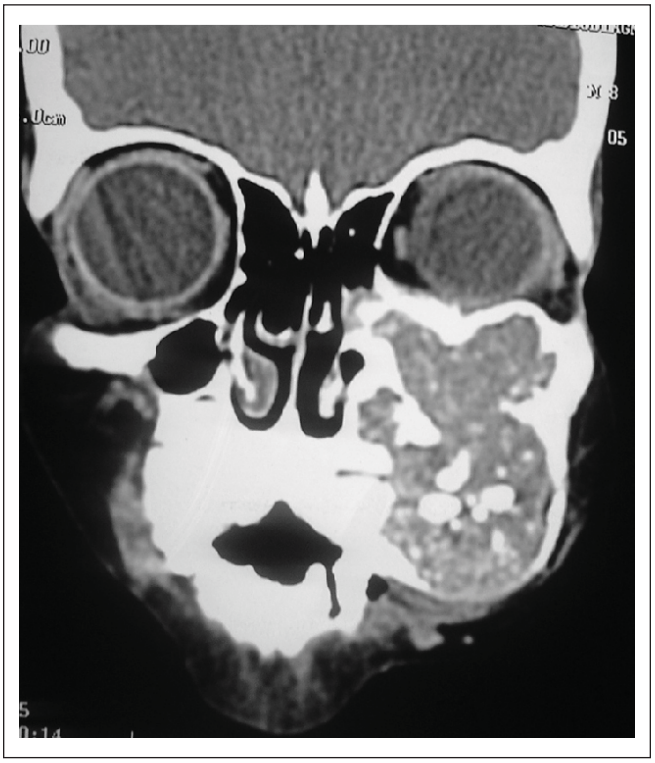

Figure 1. CT scan revealed hypodense areas in the left maxillary sinus region with multiple foci of calcifications.

* Manish Juneja M.D.S., D.N.B., (Oral Pathologist), Senior Lecturer, Department of Oral Pathology and Microbiology, Uttar Pradesh Dental College and Research Centre.
** Mala Kamboj M.D.S. Assistant Professor and HOD. Department of Oral Pathology and Microbiology, CSM Medical University

Send all correspondence to: Dr. Manish Juneja M.D.S., D.N.B., (Oral Pathologist), House No: 22, Road No 6, East Punjabi Bagh, New Delhi-110026, India

Ph. +919235604142
+919838104065

E mail: kakajuneja@yahoo.com

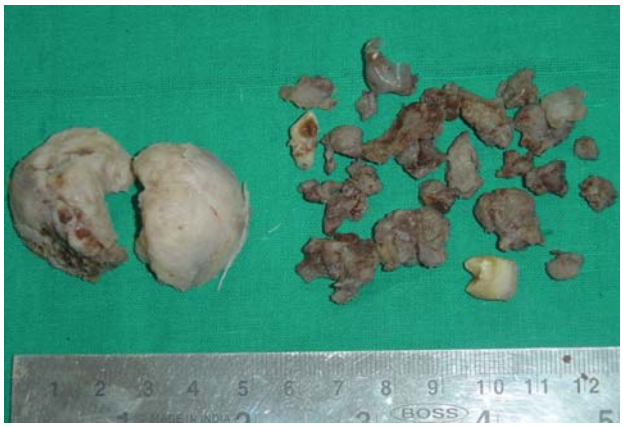

Figure 2. Gross specimen received for histopathological examination

done under general anesthesia and the excised mass was sent for routine histopathological examination.

The excised mass was received in the form of a large tissue with multiple small sized tissue fragments along with a molar tooth. The tissues were fixed and processed routinely.

Histopathologically, the tumor mass was unencapsulated showing infiltration into the surrounding bone. A highly cellular dense connective tissue stroma admixed with areas of osteoid, woven bone and a very few areas resembling cementoid were the predominant findings. A few aggregates of giant cells closely related to trabeculae of woven bone were also visible with multiple foci of myxoid degeneration (Figure 3 and Figure 4). The immature cellular osteoid formed anastomosing trabeculae which were clearly distinguished from the background cellular stroma (Figure 5). Based on the above findings a final diagnosis of juvenile ossifying fibroma-trabecular variety was given.

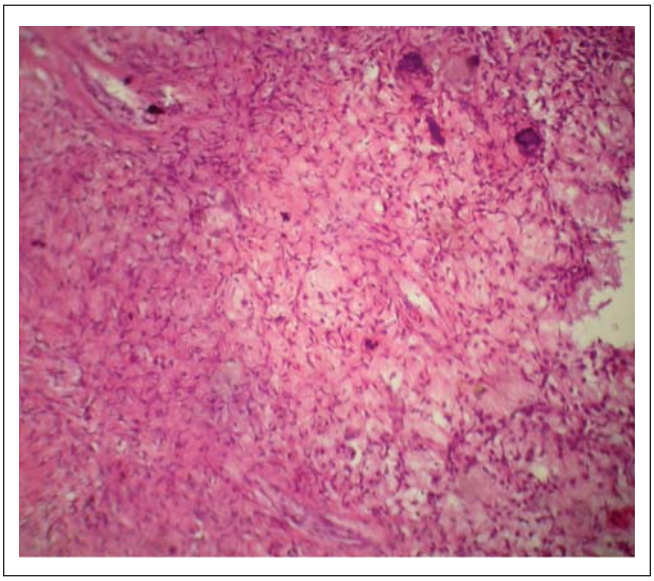

Figure 4. Foci of myxoid areas with few areas of calcification seen. (H &E, 10X)

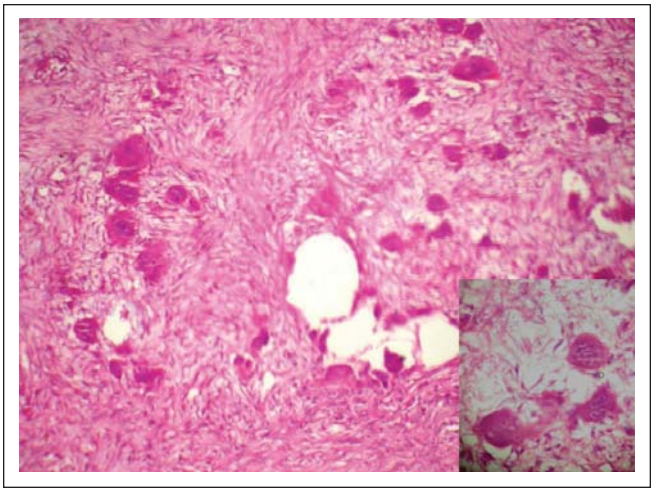

Figure 3. Giant cells seen in numerous clusters. Inset shows multinucleated giant cells. (H &E, 10X, inset shows high power magnification at 40X)

DISCUSSION

Juvenile ossifying fibroma is a rare fibro osseous lesion characterized by aggressive growth pattern and a higher rate of recurrence. Two histopathological variants of JOF have been identified, psammomatoid and trabecular types. Common to both is the age of occurrence, which is much less when compared with other fibro osseous lesions. The trabecular type is seen more in the mean age range of 8.5 years–12 years whereas psammomatoid variant occurs at a slightly older mean age range of 16 years–33 years.[3] Similar age characteristic was reflected in our case with the patient being 8 years of age.

The trabecular form of JOF is more commonly seen in the maxilla as has been reported by Makek (1987)[2] and Slootweg (1994)[4] which was also observed in our case.

Most of the maxillary JOF's are asymptomatic in early stages, symptoms appear at a later stage. In the later stage, swelling may be seen leading to facial deformity, as was seen

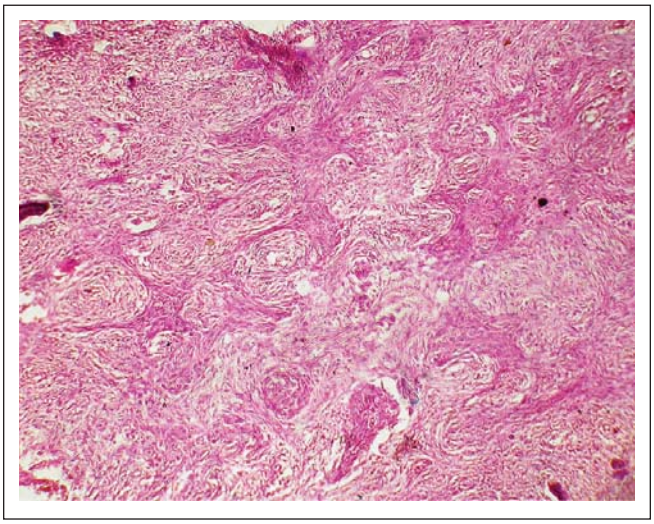

Figure 5. Trabecular pattern formed by cellular osteoid resembling paintbrush strokes (H &E, 10X)

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in our case. Involvement of the orbital bone and paranasal sinuses may cause exopthalmus, bulbar displacement and nasal obstruction.[5] Radiographically, trabecular JOF appears as an expansive, well defined, unilocular or multilocular radiolucency which caused thinning and perforation of the affected bone. The tumor mass is radiolucent with variable calcifications appearing as radio opaque areas, occasionally producing "ground glass appearance."[3] Similar radiographic findings were also found in our case with obliteration of the maxillary sinus.

The absence of the tooth germ has also been reported in cases of juvenile ossifying fibroma. Espinosa *et al*[6], Noffke[7] and Rinaggio *et al* [8] reported absence of inferior second premolar germ, based on the theory that JOF would appear like an aberration during odontogenesis. However no tooth germ was found missing in our case.

Microscopically, these tumors can be confused with other fibro osseous lesions especially cemento-ossifying fibroma, fibrous dysplasia and psammomatoid type of JOF. Cemento-ossifying fibroma is a well demarcated lesion, separated by a zone of fibrous tissue which is similar to JOF. Osteoblastic rimming is present in cemento-ossifying fibroma but according to WHO classification of JOF, it is absent in the latter.[1] However cases presented by Slootweg *et al*[4] and Williams *et al* [9] have shown the presence of osteoblastic rimming. In our case osteoblastic rimming was not seen. Presence of myxoid areas and cellular bands of osteoid resembling a "paint brush" pattern which are generally not seen in cemento-ossifying fibroma also favored a diagnosis of JOF.[3] Earlier age of presentation and aggressive nature of the lesion are not seen in cemento-ossifying fibroma and are features which are more consistent for the diagnosis of JOF.[1,2] Differentiation between fibrous dysplasia and JOF can be made by the presence of immature cellular osteoid in the form of anastomosing trabeculae seen in JOF and the absence of evenly spaced trabeculae of immature woven bone seen in fibrous dysplasia. Presence of myxoid areas was also against the diagnosis of fibrous dysplasia.[5,9] Radiographically, the demarcation of the tumor from the surrounding bone is well-defined by a radio-opaque 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.

Psammomatoid variant of JOF arises exclusively in the extragnathic craniofacial bones, especially around paranasal sinuses and the orbits of young people. Radiographically, this is a well defined osteolytic lesion, often with a sclerotic rim.[10]

It is characterized by presence of ossicles which show a basophilic center and eosinophilic fringe, with surrounding osteoblastic cells becoming incorporated into the ossicles.[3] In our case we did not find any ossicle like structures, further confirming our diagnosis of JOF. Few cementicles seen in our case do not rule out the JOF as paucity of cementicles is accepted for the diagnosis of JOF.[9]

Apart from the above mentioned fibro-osseous lesions JOF should also be distinguished from osteosarcoma of the maxillary bone. The distinction can be made by the presence of atypical cells which are never numerous in JOF. Our case did not show any areas of atypical cells. Although the presence on infiltration into the surrounding structures and myxoid areas were suggestive of osteosarcoma, the absence of atypical cells refuted the same. Absence of epithelial islands or individual epithelial cells also ruled out the possibility of odontogenic fibroma.[1]

Central aneurysmal bone cyst occurring in conjunction with juvenile ossifying fibroma has also been reported by Partridge et al. However, in our case we did not find any such component on thorough microscopic examination.[11]

The clinical management of JOF requires extensive surgery, with wide demolition of the involved bone.[12] Curettage performed as initial treatment for juvenile ossifying fibroma has been reported to have recurrence rates up to 56%[13] and 90%[14] in children younger than 10 years of age. However, the literature does not show any histologic criteria predictable of the biologic behaviour and recurrences rates and that resection results in lowest incidence of recurrence.[3] Partridge *et al* advocate that the treatment thus should be determined by the location, extent and biologic behaviour of the tumor.[11] In our case, segmental resection was done with wide margins and the patient since then has been under constant follow up.

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