



# Trabecular Variant of Juvenile Ossifying Fibroma of Maxilla

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## ABSTRACT

Juvenile ossifying fibroma (JOF) is a benign, but potentially aggressive, fibro-osseous lesion of the craniofacial bones which is considered by many to be a unique lesion because of its reported tendency to occur in children and adolescents, it is more complex histological features, and its purported tendency for locally aggressive growth. Because of this lesion's aggressive nature and high recurrence rate, early detection and complete surgical excision are essential. Reported here is a case of a juvenile ossifying fibroma of the maxilla in a 5-year-old female child.

**Keywords:** Juvenile ossifying fibroma, Benign, Fibro-osseous lesions.

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## INTRODUCTION

Juvenile ossifying fibroma (JOF) is a fibro-osseous neoplasm described as an actively growing lesion consisting of a cell-rich fibrous stroma, containing bands of cellular osteoid without osteoblastic lining, together with trabeculae of more typical woven bone. Small foci of giant cells may also be present. The lesion is nonencapsulated but well demarcated from surrounding bone (WHO).<sup>1</sup> Although JOF was described in the past as a variant of ossifying fibroma; more recently, it is segregated into two distinct subtypes are as follows:

1. Trabecular—WHO type
2. Psammomatoid variety, with varied clinical and histological features,<sup>2</sup> the maxilla, paranasal sinus, orbit and frontoethmoid bone are the predilected sites for the involvement of the JOF.

Very few cases of mandibular JOF have been reported.<sup>3</sup> Clinically in most instances, JOF is slowly growing asymptomatic swelling causing facial asymmetry, but tumor can progress to considerable size and behaves, as aggressive lesion. Less commonly, pain and paresthesia are noted. Depending upon anatomical site involvement, nasal obstruction, epistaxis and exophthalmus are observed. Slight male predominance with age ranging from 2 to 15 years has been reported. The appearance of JOF radiographically varies from unilocular to multilocular radiolucency with well-defined borders and occasionally opacification depending upon the stage and time of radiographic examination. The presence of cortical thinning, perforation, tooth displacement and root resorption, are suggestive of feature of aggressive JOF. The advanced imaging modalities, like computed tomography (CT) and magnetic resonance imaging (MRI), have shown to reveal more invasive and destructive features of JOF apart from conventional radiographic features.<sup>4</sup>

Histopathologically, both subtypes are typically nonencapsulated but well demarcated from surrounding bone. The tumor consists of varying neoplastic cellular stroma formed by spindle- or oval-shaped fibroblast cells. The mineralized component in the both patterns is distinct. The trabecular variant shows irregular strands of highly cellular osteoid encasing plump and irregular osteocytes. The plump osteoblasts often line the strands and focal areas of multinucleated giant cell are also observed. In contrast, psammomatoid pattern forms concentric laminated and spherical ossicles that vary in shape and typically have basophilic centers with peripheral eosinophilic osteoid rims.<sup>5</sup> The overall clinical, imaging and histopathological features are required to label the diagnosis of JOF. Because of unpredictable rapid and progressive growth of some JOF, the management and the prognosis of JOF are uncertain. Nonaggressive JOF is treated by curettage and local excision. Aggressive JOF requires the complete surgical excision, en bloc or hemimaxillectomy to prevent recurrences.<sup>6</sup>

## CASE REPORT

A 5-year-old female child with no general history of interest was referred to our department for a unilateral swelling localized in the right maxillary region with an evolution of 2 months (Fig. 1). Her parents had been aware of the swelling for approximately 2 months. The

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lesion had been slowly increasing in size since it was first noticed. Physical examination showed a healthy, normally developed young girl. There was significant facial asymmetry caused by an approximately 5 × 5 cm mass involving the right maxilla. The mass was bony hard and nontender to palpation and not adherent to the overlying skin. No bruits or pulsations were detected. Intraoral examination

showed expansion of the right buccal cortex, with obliteration of maxillary vestibule (Fig. 2). There was no evidence of tooth mobility, abscess formation, dehiscence or malocclusion. Computed tomography scan of the right maxilla showed medial and lateral cortical expansion and areas of central calcification (Fig. 3). Laboratory values were within normal limits. An incisional biopsy was subsequently performed, which showed a cellular connective tissue stroma consisting of fibroblasts arranged in a whorled or storiform pattern, which merged into anastomosing areas of cellular condensation. There were ribbon-like osteoid trabeculae interspersed in the fibrous stroma (Fig. 4). Approximately, 2 weeks later, the lesion was excised with the patient under general anesthesia, the lesion was approached intraorally using vestibular incision. Grossly, the specimen measured approximately 2.5 × 2.5 × 1.5 cm and had a smooth, lobulated outer surface with a pale, firm, homogenous cut surface and was well encapsulated (Fig. 5). Based on the clinical history, radiographic and histologic features of the lesion, a diagnosis of JOF of trabecular type was established.



**Fig. 1:** Depicting preoperative photographs of the child with visible facial asymmetry due to swelling on the right side of maxilla



**Fig. 2:** Intraoral photograph showing obliteration of maxillary buccal vestibule on right side

**DISCUSSION**

Juvenile ossifying fibroma may present clinically as either a gradual or rapid, painless expansion of the affected bone or region. The main characteristics are: a patient under 15 years of age, the location of the tumor, the radiologic pattern, and tendency to recur. Juvenile ossifying fibroma is often seen in a very young child. In reviews published by Hamner et al and Slootweg et al, the mean age of onset was 11.5 and 11.8 years old respectively.<sup>7</sup> Clinically, this lesion has in general a more aggressive growth rate than ossifying fibroma. Most cases of maxillary JOF are asymptomatic, as was the present case. Nasal obstruction, exophthalmos and, rarely, intracranial extension can be associated with those lesions arising within the paranasal sinuses and orbit.<sup>8,9</sup> Radiographic features are nonspecific and depending on the location of the tumor, maturation stage and stage of ossification, they are uni or multilocular well-defined lesions which may be radiolucent, mixed or radiopaque.<sup>9</sup> Aggressive lesions may show cortical thinning and perforation.<sup>10</sup> The most frequent location of JOF is in the paranasal sinuses, accounting for about 90% of cases, whereas mandibular lesions account for approximately 10% of the facial JOF cases. In general, JOF has a more aggressive growth pattern than the adult variant of ossifying fibromas. They are usually asymptomatic, exhibit rapid growth of the involved site and the first presentation will be a clinically obvious swelling. All the above features were seen in our case. Rapidly, expanding central jaw lesions of children and adolescents, when coupled with the above radiographic features, are



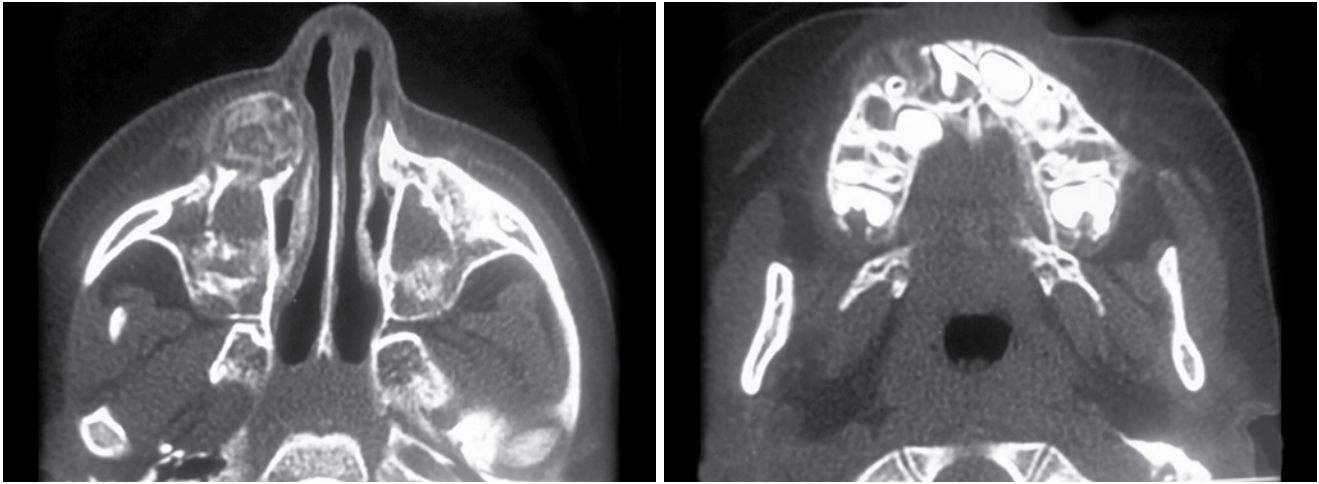


Fig. 3: Symmetric mediolateral expansion of right anterior maxilla with mixed radiopacities

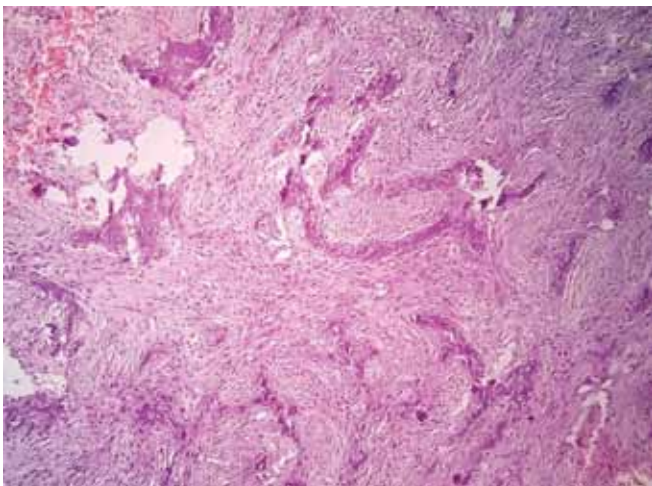


Fig. 4: Photomicrograph showing fibroblastic stroma with irregularly arranged trabeculae interspersed within it

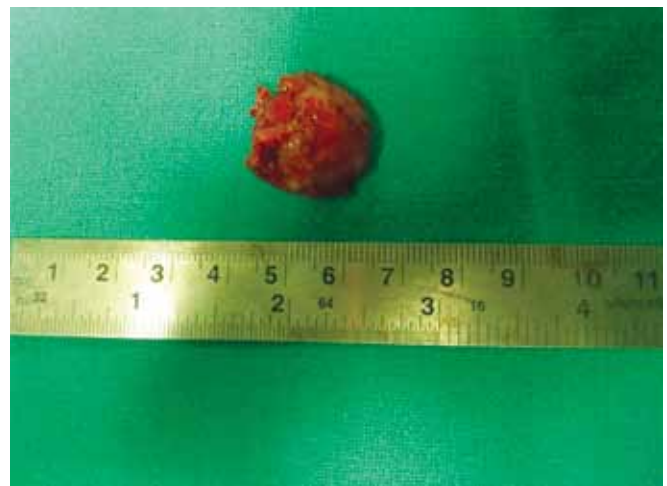


Fig. 5: Intraoperative photograph depicting the smooth, lobulated outer surface of the tumor

worrisome for malignancy. Malignant neoplasms that have a tendency to involve the craniofacial skeleton in this age group include osteosarcoma, chondrosarcoma, Ewing's sarcoma, and the African form of Burkitt's lymphoma. Benign neoplastic or developmental lesions are more commonly encountered in the jaws, and, although they may show differing degrees of aggressiveness, they often present with the radiographic characteristics of JOF. Included among these diseases are osteoblastoma, an intraosseous form of fibromatosis known as desmoplastic fibroma, cemento-ossifying fibroma, fibrous dysplasia, central giant cell granuloma, and cherubism. There is some degree of histological overlap between JOF and cemento-ossifying fibroma. In contrast to cemento-ossifying fibroma, which histologically is characterized by uniformity of pattern, JOF is more likely to show markedly heterogeneous morphology,<sup>7,9,11</sup> a characteristic that may complicate the diagnosis and subsequent management of these tumors. Areas of dense cellularity may alternate with myxomatous regions, and the

distribution of bone trabeculae and ossicles often is uneven. Caution must, therefore, be exercised in the interpretation of incisional biopsies of fibro-osseous lesions. The rapid growth rate often exhibited by these lesions can be quite alarming and cause the clinician to suspect the presence of a malignancy. It is, therefore, important to maintain active communication between surgeon and

pathologist to establish the benign nature of the lesion and prevent overtreatment. Whereas the treatment for JOF recommended by most investigators<sup>9,11-13</sup> is conservative excision or curettage, some lesions may necessitate more aggressive management.<sup>14</sup> Longstanding lesions may show significant cortical destruction and periosteal elevation,<sup>10</sup> which can increase the risk of recurrence. The recurrence rate ranges from 30% observed by Johnson et al to 58% reported by Waldron<sup>8,9</sup> continued follow-up is essential. Despite the aggressive nature of the lesion and high rate of recurrence, malignant transformation to sarcoma has not been reported.

## CONCLUSION

Although JOF an uncommon clinical entity, its aggressive social behavior and high recurrence rate means that is important to make an early diagnosis, apply the appropriate treatment, long-term and follow-up.

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