



(CASE REPORT)



## Juvenile ossifying fibroma (JOF) – A rare case report

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### Abstract

Juvenile ossifying fibroma (JOF) is a rare, benign, bone forming lesion of children and adolescents. When sited in the jaws, it is considered as an aggressive form of ossifying fibroma presenting with a wide range of clinical symptoms and a high incidence of recurrence. Although early detection and management is thought to eradicate it completely. The present case report focuses on the surgical yet conservative management of JOF in a 5 years old boy who reported with a year-long, progressive swelling on the left side of his face. The tumour was excised with minimal surgical intervention into a clearly defined and lobulated single mass using maxillary vestibular incision under general anaesthesia. The postoperative course was uneventful, and no signs of recurrence were found in the next 6 months follow-up period.

**Keywords:** Juvenile ossifying fibroma; Conservative; Benign; Fibro-osseous lesion

### 1. Introduction

Fibro-osseous lesions of the cranio-facial bones like fibrous dysplasia, ossifying fibroma, cemento-ossifying dysplasia etc are benign by nature, slow progressing and present with similar histopathological findings [1]. Ossifying fibroma is a rare, benign, bone forming lesion that has a peculiar age of onset, clinical appearance, aggressiveness [2] and is divided into conventional and juvenile subtypes. Juvenile ossifying fibroma (JOF) was first described by Benjamin et al in 1938 as an “osteoid fibroma with atypical calcification” and in 1952 Johnson et al called it “juvenile active ossifying fibroma” [3]. JOF is also known as juvenile aggressive ossifying fibroma, trabecular osteo-desmoplastoma, and active fibrous dysplasia [4]. Among all oral tumours in children, JOF comprises of 2% with equal predilection in males and females. JOF is found in facial bones (85%), calvarium (12%), the mandibular region (10%), and very rarely, an extracranial location (3%) [5].

90% of the lesions in the facial region involves the maxillary antrum. A little sclerotic shell of bone completely encircles the tumour. With cortical damage and infiltration of several adjacent anatomical structures, it appears to be locally aggressive. Soft tissue consistency predominates, with varying amounts of inner calcification and/or linear or asymmetrical focal bone. On Computed Tomography (CT) scans, it usually appears as a low-density mass due to cystic alterations. The lesion may demonstrate diffuse appearance and enhancement after intravenous infusion of iodinated contrast.

JOF is identified by the presence of cellular fibrous stroma, garland-like bone strands, and cementum particles on histological examination. Clinical, linear radiography, CT scan, and histological findings are adjuncts for appropriate diagnosis of JOF [6]. JOF was further subdivided into two histopathological variants: trabecular (TrJOF) and

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psammomatoid (PsJOF). One clinical feature that helps differentiate between the histopathological variants is the site of involvement; PsJOF presents in the paranasal sinuses, and TrJOF presents in the maxilla [2].

## 2. Case Report

A 5-year-old male reported to the Department of Oral & Maxillofacial Surgery, Institute of Dental Studies & Technologies, Modinagar, UP, India with a chief complaint of swelling over the left side of the face since one year. To begin with the lesion appeared as a pea-nut size swelling one year ago which gradually increased into the present size which is sustained for the last 4 months. Nothing significant was noted in the medical history. Extraoral examination revealed a firm swelling measuring 3.6 x 3.1 x 3 cm in size on the left side of the face, leading to facial asymmetry (Figure 1). Intraoral examination revealed normal mouth opening with a lesion that had almost completely obliterated the buccal sulcus, with mobile deciduous canine and molars of the affected side (Figure 2). Palpation revealed a firm, non-tender mass adhering to the maxilla with a smooth surface. The mucous membrane in the tumour site had no pathological alterations. Cone Beam Computed Tomography (CBCT) of the maxilla revealed well defined, expansile, unilocular, mixed density lesion involving left maxilla involving the maxillary sinus and alveolus. Imaging revealed invasion of soft tissue mass in the superior left maxilla to the left maxillary sinus (Figure 3). On histopathological evaluation, presence of fibrillar osteoid trabeculae and woven bone pieces, highly fibro cellular connective tissue with dense collagen fibre bundles arranged in whorls and swirling pattern led to the diagnosis of JOF and the patient was advised to undergo excision. A maxillary left buccal vestibular incision was made and surgical excision of the tumour mass was performed under general anaesthesia. The tumour was removed in toto as a single mass which was clearly defined and lobulated. The bony defect was filled with medical absorbable gelatin sponge (Figure 4-5). The mobile teeth were extracted at the time of surgery. The patient's postoperative course was uneventful, and he was followed up for six months with no signs of recurrence.



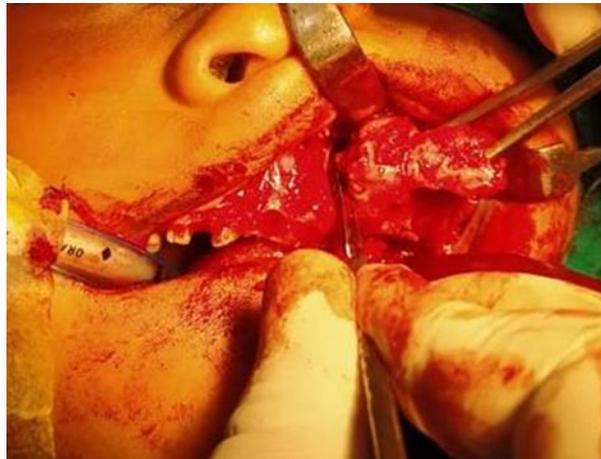
**Figure 1** Clinical picture of the patient depicting swelling on the left maxillary region



**Figure 2** Intraoral picture showing complete obliteration of the buccal sulcus



**Figure 3** Radiographic picture revealed well defined, expansile, unilocular, mixed density lesion involving left maxilla and maxillary sinus



**Figure 4** Surgical excision of the swelling



**Figure 5** Excised specimen

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### 3. Discussion

Ossifying fibromas are uncommon benign tumours with well-defined borders that grow in a concentric, expansile pattern. JOF has an unclear cause. Traumatic, developmental, and odontogenic aetiology are suspected, and the periodontal ligament may play a role in its pathogenesis. HPRT2, a tumour suppressor gene, has also been identified as a possible contributor in JOF [3].

When it originates in the maxilla, JOF affects young children under the age of 15, and its growth is aggressive and asymptomatic. JOF occurs most frequently in the maxillary sinuses (approximately 90% of cases), whereas mandibular lesions are approximately 10% of facial JOF cases [2].

Radiographically, JOF is divided into three stages. Stage I is characterized by a well-defined radiolucency with no inner calcification. The radiolucent area in Stage II (the mixed stage) is characterized by specks of radiopacities. The mature stage (i.e., stage III) is a totally radiopaque structure [3]. In more aggressive cases, there may be erosion and invasion of the surrounding bone. The transition from the radiolucent to the radiopaque stage usually takes at least six years. Hence, most lesions diagnosed are radiolucent or have heterogeneous density. In very rare situations, fast maturation and total radiopacity can occur in less than a year [5]. This case was in Stage II.

Microscopically, the presence of premature woven bone trabeculae, cementum-like tissue, or both in a fibrous stroma characterizes JOF. PsJOF is defined by the multiplication of benign spindle-shaped fibroblastic cells with embedding mineralized components. This can manifest as round to ovoid bone deposits with an osteoid margin. Although multinucleated osteoclast-like large cells are occasionally found with typical mitotic figures, atypia is not a common occurrence. A fibroblastic spindle cell stroma containing osteoid matrix, surrounded by osteoblasts and anastomosing trabeculae of premature woven bone, typically intermixed with dispersed groups of multinucleated giant cells; mitoses can be detectable, however cystic degeneration is infrequent [7].

JOF is equally seen in both the gender, and most cases of JOF, like ours, are asymptomatic. The odds of recurrence following resection range from 30% to 56 %. This could be due to the infiltrative character of the tumour borders following a partial excision, rather than any inherent biological features of the tumour [8]. Despite the aggressive character and increased risk of recurrence, no malignant transformation has been recorded. Secondary alterations associated with JOF include aneurysmal bone cysts. The treatment of choice, as in our situation, is a complete excision of the tumour. JOF can be locally invasive, producing severe morbidity and mortality due to cerebral expansion, even if it is benign. Because JOF is radioresistant, irradiation should be avoided as a treatment option [9].

Taking into consideration about child's age, cosmetic, functional, and growth concerns, a conservative surgical approach was chosen in this case. The tumour was lobulated and measured 3.6 x 3.1 x 3 cm in diameter but did not lost its encapsulation. As a single mass, this characteristic of the tumour made surgical excision easier. The post-surgical site was meticulously examined to rule out any involvement of the maxillary sinus's medial (nasal), roof (orbital floor), or posterior wall. There were no complications throughout the 6-month postoperative follow-up.

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### 4. Conclusion

JOF is a rare, benign tumour of the bone with the potential for rapid development, bone destruction, and recurrence. Surgical excision is the preferred treatment, especially when JOF is located in the maxilla, which necessitates surgery yet conservative excision with healthy margins. Because of its aggressive nature and high recurrence rate, early identification, complete surgical excision of these lesions, and long-term follow-up examinations are critical in the clinical care of JOF cases. Unless JOF recurs locally, the prognosis is favorable.

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### Compliance with ethical standards

#### *Disclosure of conflict of interest*

No conflict of interest.

#### *Statement of informed consent*

Informed consent was obtained from all individual participants included in the study.

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