Juvenile Aggressive Ossifying Fibroma of the Maxilla: A Case Report
Harsh V Babaji¹, C Saravana Bharathi², P K Pal³, Gurjap Singh⁴, M Anuradha⁵, Vishwajit Rampratap Chaurasia⁴

Contributors:
¹Senior Lecturer, Department of Oral & Maxillofacial Surgery, College of Dental Sciences, Davangere, Karnataka, India; ²Reader, Department of Oral Medicine & Radiology, Rajah Muthiah Dental College & Hospital, Chidambaram, Tamil Nadu, India; ³Professor, Department of Prosthodontics, Institute of Dental Sciences, Bhubaneswar, Odissa, India; ⁴Reader, Department of Oral & Maxillofacial Surgery, ITS Dental College, Greater Noida, NCR, Delhi, India; ⁵PG student, Department of Conservative Dentistry & Endodontics, KLE Dental College, Belgaum, Karnataka, India.

Correspondence:
Dr. Babaji HV. Department of Oral & Maxillofacial Surgery, College of Dental Sciences, Davangere, Karnataka, India.
Email: drharshavbomfs@gmail.com

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Abstract:
Juvenile ossifying fibroma is an uncommon fibro osseous neoplasm at a younger age. It is a well-defined clinical and histological lesion. This lesion is locally invasive and spreads quickly. Early detection and complete surgical excision of this lesion is essential since it has high recurrence rate.

Key Words: Fibro-osseous lesion, juvenile ossifying fibroma, psammoma

Introduction
Juvenile ossifying fibroma (JOF) is a rare benign fibro-osseous lesion affecting craniofacial skeleton of young patients. It is a well-defined clinical and histopathological entity often confused with malignant lesions.¹ These lesions have high recurrence rate. The concepts and criteria of benign fibro-osseous lesions were presented in 1973 by Waldron and Giasanti after reviewing more than 60 cases.³ It affects maxilla more commonly than mandible with no gender predilection. Microscopically JOF can be classified as trabecular and psammomatoid type. Trabecular variant affects predominantly jaws at mean age of 2-12 years, whereas psammomatoid type affects patients from 3 months to 72 years.

JOF is also called as juvenile active ossifying fibroma, juvenile aggressive ossifying fibroma and the aggressive psammomatoid ossifying fibroma. Most of the benign fibro osseous lesions of jaws are slowly growing and asymptomatic with expansion of the affected bone. These lesions are commonly occurs between 5 and 15 years of age.² The tumor can grow rapidly and can expand in an affected area hence they are also referred as active or aggressive ossifying fibroma. Because of its aggressive nature it can be misdiagnosed as sarcomas.¹

Case Report
A 14-year-old patient reported with the complaint of a gradually enlarging asymmetry of the right side of the face. The lesion had been steadily increasing in size over past 3 years; however in the past 6 months, the lesion had rapidly enlarged, producing difficulty in nasal breathing and pus discharge from the medial corner of the right eye.

Extra oral examination revealed facial asymmetry with a moderately large right facial mass extending from the right maxilla to the infra orbital rim superiorly, medially to the lateral wall of the nose, laterally up to the zygoma region (Figure 1).

Intraoral examination revealed gross expansion of the right maxillary alveolar process extending to the hard palate which was bony hard in consistency (Figure 2).

Orthopantomograph showed a unilobulated radiolucent lesion with well-defined radio-opaque borders (Figure 3). Computed tomography (CT) scan revealed extensive, expansile, mixed lesion with central areas of calcification surrounded by well-defined sclerotic borders involving right maxillary sinus, alveolar bone, nasal cavity and floor of the orbit (Figures 4 and 5). Surgical enucleation of the lesion followed by extraction of involved premolar and molars teeth was done under general anesthesia (Figure 6). Post-operative healing of the lesion was uneventful.

Histologically the lesion was non-encapsulated with cell rich fibrous tissue containing bands of cellular osteoid with few giant cells and numerous psammoma body-like ossicles were dispersed in a fibroblastic stroma (Figure 7). On the basis of clinical, histological, radiographic and CT findings, the lesion was diagnosed as a JOF of maxilla (juvenile psammomatoid ossifying fibroma).

Discussion
It has been thought that fibro osseous lesions are the result of replacement of normal bony architectures by fibrous tissue, which may mineralize in various forms such as woven, lamellae bone or cementum with clinical presentations and microscopic features.¹ JOF usually involves the maxilla, paranasal sinuses, orbital and frontoethemoidal bones.⁵
It has been stated that ossifying fibroma of the maxillary sinus should be considered in the differential diagnosis when young patient presents with ptosis and slowly progressive upward displacement of the globe.²

It has been thought that JOF arise as a result of differentiation of multipotential precursor cells or mesenchymal cells of the periodontal ligament to form cementum, osteoid, or fibrous tissue combination. Histologically lesions present with the mineralized component composed of numerous spherical cementum-like bodies (psammomatoid ossicles) as a present

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**Figure 1:** Diffuse swelling present in the right maxillary region.

**Figure 2:** Intraoral view showing diffused expansion of the right maxillary alveolus.

**Figure 3:** Panoramic radiograph showing extensive radiopaque lesion with well-defined corticated border.

**Figure 4:** Computed tomography coronal view showing lesion expansile lesion with corticated border with a central area of calcification. Lesion extends up to the orbital bone.

**Figure 5:** Computed tomography axial view showing lesion in the right maxillary sinus.

**Figure 6:** Enucleation of maxillary lesion followed by extraction of premolars and molars.
Pathogenesis of JOF is unknown; it appears to be spontaneously occurring neoplasm. Radiographic features are highly variable depending on calcification. In the early stages of development JOF appears as a unilobulated or multilobulated radiolucent lesion followed by varying degrees of radiolucency with defined borders and radio-opacity on further progression. With mature lesion bone may take more woven appearance. Clinical, histological and radiographic features help in proper diagnosis of the condition. CT also aids in the diagnosis. CT findings of JOF may include well-defined borders identified by a thin sclerotic shell. The lesion has a predominantly soft tissue consistency with a variable amount of internal calcification or bone. JOF appears to be more invasive and destructive on CT scans when compared with fibrous dysplasia and adult ossifying fibroma. The lesion enhances after gadolinium contrast injection. MRI offers great specificity when there is neurovascular and ocular involvement.

Management of JOF is uncertain, however, small lesions can be conservatively treated by curettage or enucleation. Large and irregular shaped tumors which infiltrating sinuses can be treated by radical resection.

Children with aggressive JOF, in the maxilla, are treated by through curettage in combination with adjuvant interferon alpha therapy for 1 year. It has been reported that its recurrence rate ranges from 30% to 58%. Since it has very high recurrence rate, hence majority of authors suggest early diagnosis and complete surgical excision with long term follow-up.

**Conclusion**

JOF is a non-metastasizing, but locally aggressive and highly recurrent fibro-osseous lesion of the craniofacial skeleton. The knowledge of its possible locations and characteristic features help to differentiate from other lesions. Early detection and complete surgical excision is essential to aid in better prognosis.

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**Figure 7:** High power microscopic picture showing numerous psammoma like ossicles within the cell rich fibrous tissue.