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An Innocuous Variant of Juvenile Ossifying Fibroma of The Maxilla: A Case Report

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Abstract

Juvenile ossifying fibroma (JOF) is a benign bone forming neoplasm arising from undifferentiated cells of the periodontal ligament tissue and is distinguished from other fibro osseous lesions by the age of onset, clinical presentation and aggressiveness.2 This lesion has more predominance to occur in younger individuals than older ones and mostly involves facial bones. It is usually asymptomatic achieving a large size leading to facial asymmetry and expansion of cortical bones. The less common symptoms include pain and paresthesia. 4 JOF has two histological subtypes: psammomatoid (PJOF) and the trabecular (TJOF).

The following article describes a relatively less aggressive, passive form of a Juvenile Ossifying Fibroma arising in a 12 year old female in her left posterior maxilla, and how a timely diagnosis and immediate treatment followed by early prosthetic rehabilitation managed to avoid a more debilitating surgery and further complications later.

Introduction

Benign fibro osseous lesions of the head and neck are a diverse group of lesions which includes fibrous dysplasia, cemento-osseous dysplasia and ossifying fibroma. These lesions are a rare clinical entity and are characterized by the replacement of normal bone by fibrous tissue containing a newly formed mineralized product. **[**1**]**

Ossifying fibroma is a rare benign bone forming neoplasm arising from undifferentiated cells of the periodontal ligament tissue and is distinguished from other fibro osseous lesions by the age of onset, clinical presentation and aggressiveness.**[**2**]** They are characterized by cell rich fibrosis with clear boundaries and presence of variable amounts of calcified tissue similar to bone, cementum or both. Ossifying fibromas are classified as the Conventional and Juvenile ossifying fibroma (JOF). JOF occurs usually in children/ adolescents.**[**3**]** The JOF was first described in the 2nd edition of WHO classification of odontogenic tumors occurring in children below the age of 15 years.**[**4**]** They have been designated as "active"/ "aggressive" reflecting their aggressive nature. JOF exhibits rapid growth with high rate of recurrences which reflects their origin in younger population and the site of origin.**[**3**]**

The JOF was first described by Benjamin (1938) as "Ossifying fibroma with atypical calcification" and the term "Juvenile aggressive ossifying fibroma" was coined by Johnson (1952). They constitute about 2% of the oral tumors occurring in children.**[**5**]** It is usually asymptomatic achieving a large size

leading to facial asymmetry and expansion of cortical bones. The less common symptoms include pain and paresthesia. **[**4**]**

The WHO considers 2 variants based on the histological features: Trabecular JOF (TrJOF) and Psammomatoid JOF (PsJOF).**[**6**]** The clinical features that help us to distinguish between the two variants is the site of origin. TrJOF presents in the maxilla or the gnathic bones whereas PsJOF involves the paranasal sinuses, periorbital and craniofacial skeleton. Owing to the aggressive nature and the high recurrence rates of this lesion, early diagnosis and definitive treatment are essential.

Case Report

A 12-year-old female patient reported to the Department of Pediatric and Preventive Dentistry, College of Dental Sciences, Davanagere, Karnataka with a chief complaint of a progressive swelling in the maxillary posterior region since a year. The swelling had an insidious onset with no history of trauma, pus discharge or paresthesia. However, there was history of pain, which was intermittent, moderate in intensity and aggravated even on touch. Extra oral examination revealed a visible facial asymmetry involving the left maxilla, There was no pus discharge noted. Mouth opening was normal. On palpation, the swelling was hard in consistency, tender, non-mobile and non-adherent to the skin. No pulsations or raise in temperature were noted. The left submandibular lymph node was palpable, tender and mobile.

On intra oral examination, gingival swelling was noted in the maxillary left vestibule in the posterior region involving primary and permanent molars and extending posteriorly to involve the maxillary tuberosity. No abscess or dehiscence was noted. On palpation the swelling was tender, firm in consistency.

On radiographic examination, the panoramic image showed a well circumscribed radio opacity with irregular central opacity in the left maxillary posterior region involving 64, 65,26 and 27. The tooth bud of permanent left first premolar was displaced and root resorption was noted with the mesial root of 26.

Maxillary occlusal radiograph revealed expansion on buccal cortical plate in the region on 64, 65, 26 and 27 with a well circumscribed mixed radiolucent and radio-opaque lesion.

An incisional biopsy was performed under General Anesthesia in the Department of Oral and Maxillofacial surgery. Based on the clinical, radiographic and histological features, a diagnosis of Juvenile Ossifying Fibroma of the Trabecular type was established.

Approximately after 14 days, the lesion was surgically resected under General Anesthesia along with the permanent tooth buds of 24,25,26,27 and 28.

Microscopic examination of the excised specimen revealed a cellular connective tissue stroma with fibroblasts, thereby confirming the diagnosis.

The patient was discharged on the 5th post operative day and a removable feeding plate was fabricated. After 2 months, a partial denture was fabricated. The patient was followed up regularly, and her 1-year follow-up revealed no evidence of any recurrence or infection. Healing was satisfactory and uneventful. Complete aesthetic and functional rehabilitation has been achieved.

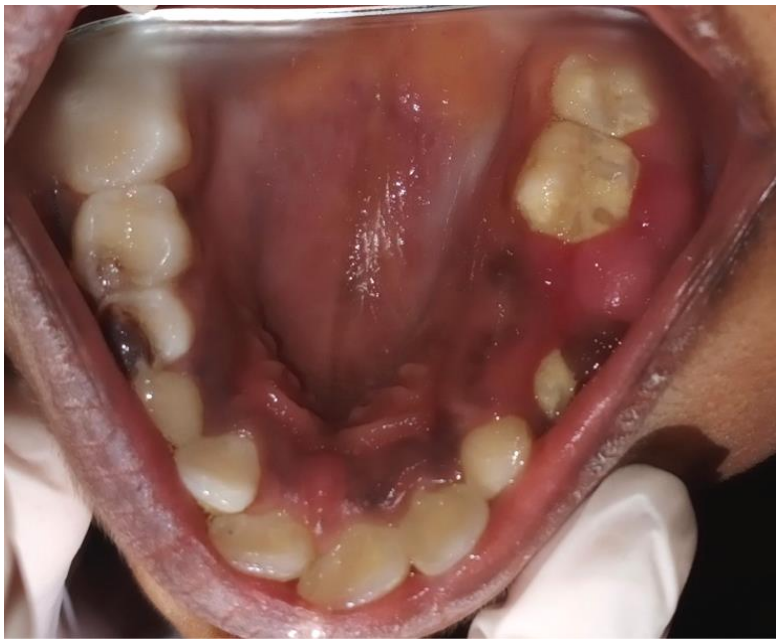

Figure 1 - PRE-OPERATIVE : A mildly expansile lesion over the left posterior maxillary alveolus

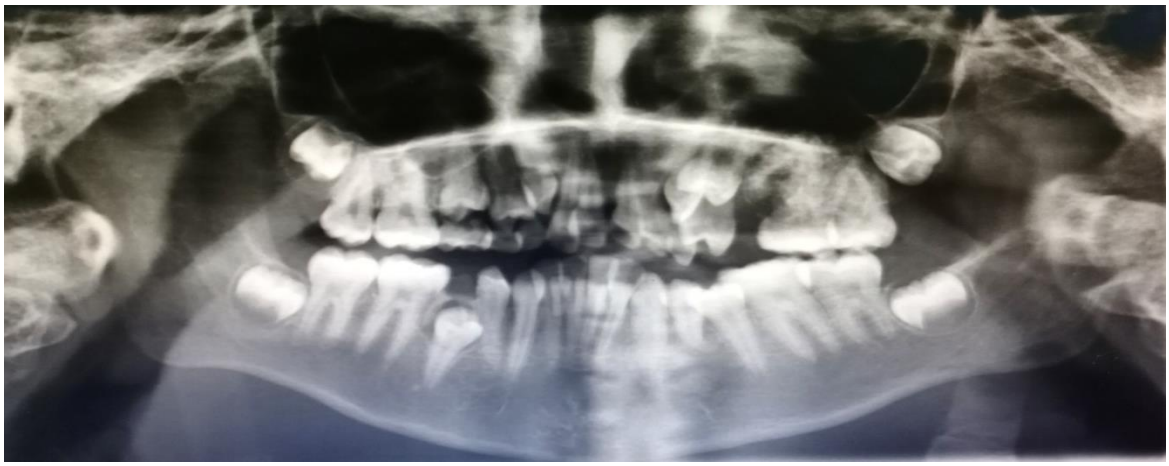

Figure 2A - Orthopantamograph showing a mixed radiolucency and radio-opacity in left posterior maxilla

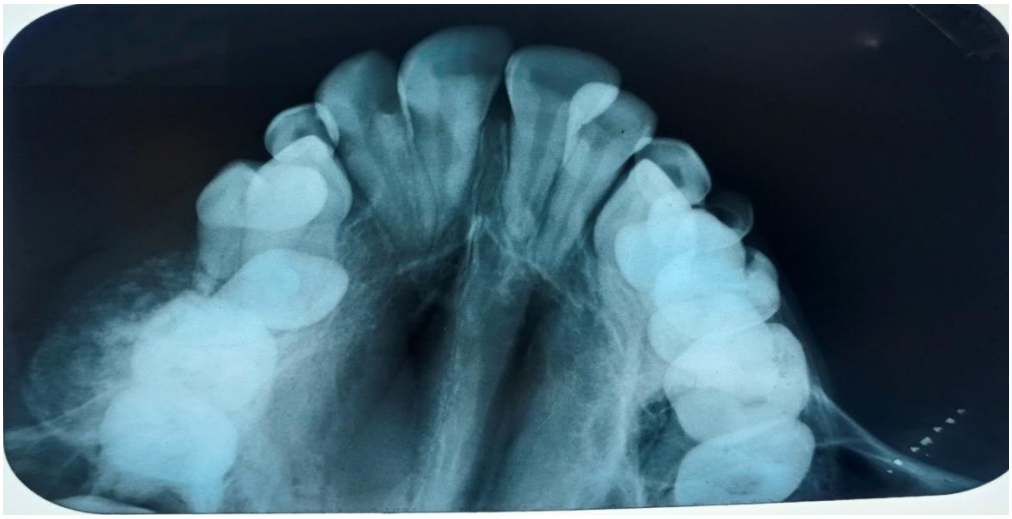

Figure 2B - A Maxillary Occlusal Radiograph showing the same findings

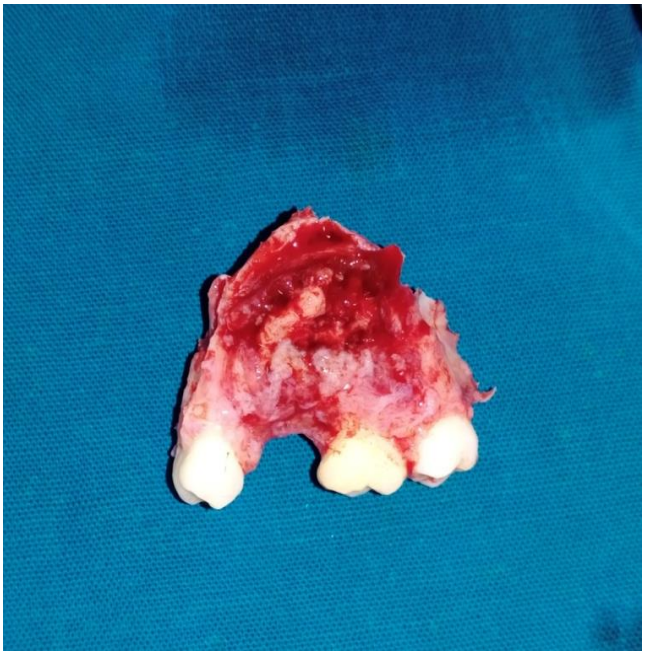

Figure 3 - The resected bone segment

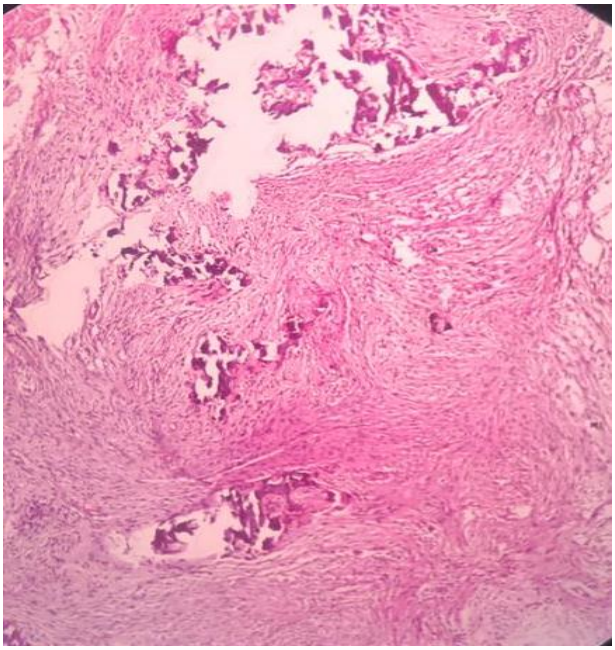

Figure 4 – Histopathological findings

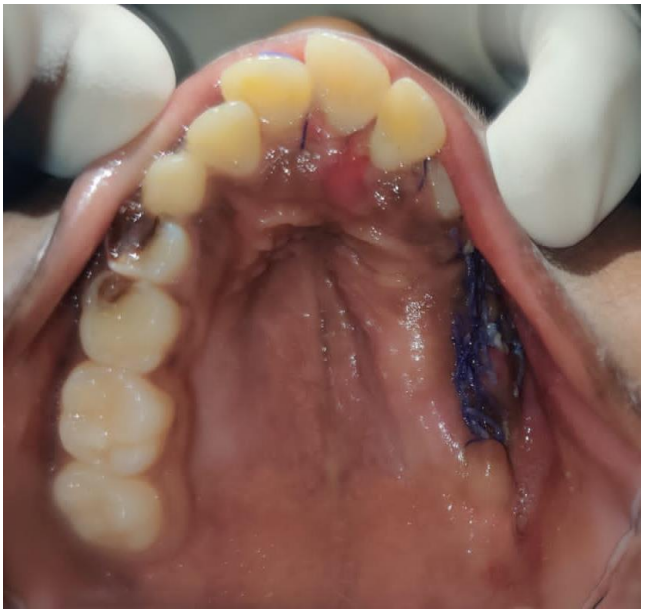

Figure 5 - POST OPERATIVE: sutures in situ, satisfactory healing

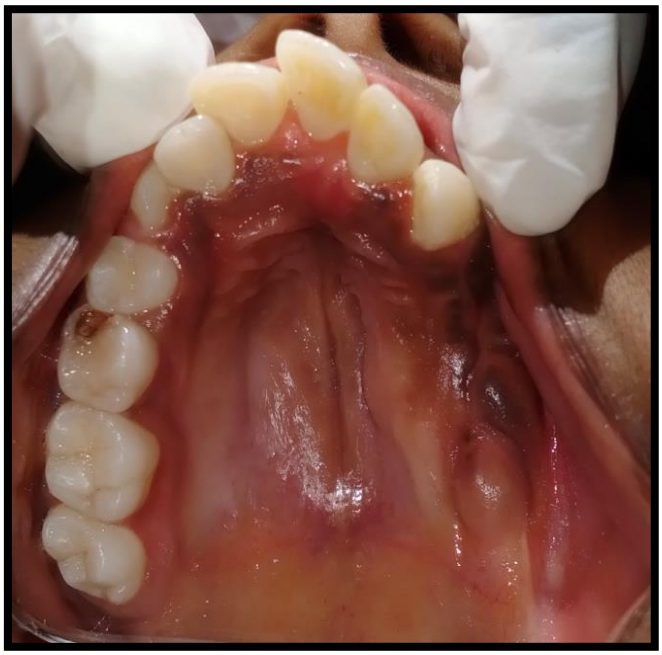

Figure 6 - 7 Months follow up

Figure 7A – Fabrication of a maxillary partial removable prosthesis

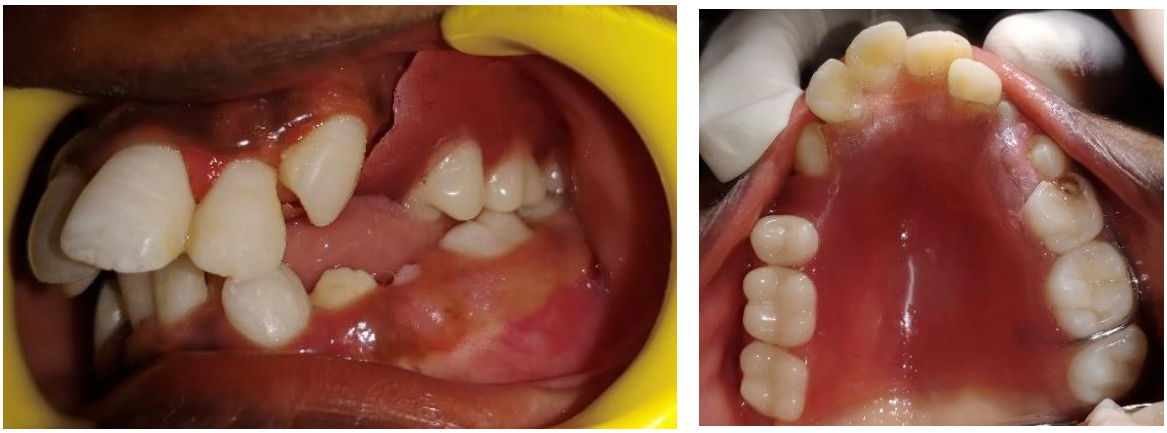

Figure 7B, 7C – Post surgical prosthetic rehabilitation.

Discussion

JOF may present clinically as either a gradual or rapid, painless expansion of the affected bone or region. JOF is often seen in a very young child. **[**1**]** 85% of JOF lesions originate in the facial bones (maxilla being more common than mandible), amongst which 90% arise within paranasal sinuses. 12% occur in the calvarial bones and 4% are extra-cranial. If the orbital bones or paranasal sinuses are involved, symptoms can vary from proptosis, exophthalmos, epistaxis, nasal congestion and sinusitis.**[**8**]** According to Hamner et al. and Slootweg et al. the mean age of onset was 11.5 and 11.8 years old, respectively.**[**1**]**

The etiology of JAOF is poorly understood. It is thought to arise from mesenchymal cell differentiation of periodontal ligaments—which is a precursor to cementum, fibrous tissue and osteoid.**[**8**]** Others believe that it arises from the primitive mesenchymal cells or remnant cells after incomplete migration of the medial part of the nasal anlage. Mutation of the tumor suppressor gene, HRPT 2 (Pimenta et al.), has also been identified in cases of JOF.**[**9**]** The absence of the tooth germ has also been reported

by Espinosa, et al Noffke and Rinaggio et al based on the theory that JOF would appear like an aberration during odontogenesis.[10] JOF may also be due to maldevelopment in tissues between roots of molar teeth which help in generation of bony septa.[11] Recent studies suggest that non-random chromosome breaks in Xq26 and 2q33 result in a translocation (X;2) may be associated with the origin of this neoplasm. [6]

JOF presents with aggressive pattern which is seen mainly in young patients due to high levels of periodontal ligament activity (e.g., formation and degradation) are more commonly seen in children, contributing to the constant irritation associated with both primary tooth exfoliation and permanent tooth eruption can contribute for the increased prevalence of reactive lesions in younger patients. [12]

All JOF lesions can be categorized into the psammomatoid (Psammomatoid Juvenile Ossifying Fibroma or PJOF) and the trabecular (TJOF) types that were first described by Gogl and Reed, respectively. While both subtypes exhibit similar clinical behavior, PJOF features fibromatous stroma with psammomatous calcifications, while TJOF is characterized by bony trabeculae of varying degrees of mineralization among immature osteoids. TJOF also appears to manifest radiographically with mixed radiolucencies, while PJOF displays a ground-glass phenotype. [4]

Trabecular JOF is an unencapsulated tumour with a hypercellular stroma composed of spindle cells, with little collagen production and with long slender strands of osteoid. The immature bone trabeculae show no maturation and are usually devoid of osteoblastic rimming. The lesions are sharply demarcated from their surroundings, either by a fibrous capsule or by a rim of the pre-existing bone. [13]

Radiographic features are non-specific and depending on the location of the tumor, maturation stage and stage of ossification, Trabecular JOF is almost always well demarcated and often has an even pattern of calcification. The expansive tumor usually appears as a well circumscribed unilocular or multilocular radiolucency with a variable degree of radiopacity. The lesion shows no blending into the surrounding bone or invasion of the soft tissue and is usually described with a well-defined radio-dense border. Aggressive lesions may show cortical thinning and perforation.[1] Root resorption and displacement of related teeth with cortical thinning and perforation can be seen.[14]

Very rarely a ground-glass appearance can be noted.9 Opacification of the sinus is usually seen in cases involving the sinuses.8 On Magnetic Resonance Imaging (MRI), this lesion has a heterogeneous high-intensity signal on T1-weighted sequences and a low intensity signal on T2-weighted sequences. MRI allows defining the extent of the lesion but not clearly its bony component.[3]

Histopathologically, JTOF is usually well demarcated, but is unencapsulated and may infiltrate surrounding bone. The stroma shows variable cellularity from loose to more cellular fibroblastic spindle cells. The lesion is characterised by long and slender immature trabeculae of osteoid that fuse together to form anastomosing networks with large eosinophilic osteoblasts. Cellular osteoid is usually present in association with more mature trabeculae of woven bone lined by osteoblasts or osteoclasts and occasional spherical calcifications may be seen. Some cases have been reported with cystic degeneration with focal aggregates of multinucleated giant cells and aneurysmal bone cyst formation. Trabecular JOF may have scattered mitoses, but these are not prominent and are normal.**[**14**]**

JOF has to be distinguished from other fibro-osseous jaw lesions. The main differential diagnosis is fibrous dysplasia. The clinical and radiological demarcation between the lesion and the surrounding tissue preclude considering JOF as a variant of fibrous dysplasia as this latter lesion typically blends imperceptibly into the adjacent bone. JOF is not capsulated but is separated from the surrounding bone by radiopaque borders and lacks ground-glass attenuation. These findings are helpful in differentiating it from fibrous dysplasia. As far as histology is concerned, JOF can be distinguished from fibrous dysplasia by the very prominent osteoblastic rimming of the bony trabeculae and by the demarcation between lesional bone and adjacent jaw bone provided that surrounding tissue is included in the biopsy specimen. Moreover, the continuous growth that is shown by JOF refutes a dysplastic nature and makes it more appropriate to consider JOF a neoplasm. **[**5**]** Aneurysmal bone cyst, osteoblastoma, osteosarcoma, and cemento-osseous dysplasia are other major conditions to be included in the differential diagnosis.6 Burkitt lymphoma should also be considered in the differential diagnosis of Juvenile Aggressive Ossifying Fibroma (JAOF) because of the similarity in the age and site of presentation, rapidity of growth, and radiolucent radiographic appearance. The characteristic gross mobility and displacement of the associated teeth in what is usually termed “dental anarchy” may serve as a dividing line between the two lesions clinically. In addition, patients with Burkitt’s lymphoma may present with varying abdominal symptoms.**[**15**]**

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.**[**1**]** Partridge et al advocate that the treatment thus should be determined by the location, extent and biologic behavior of the tumor.**[**10**]**

The clinical management and prognosis of JOF is somewhat uncertain. Surgical treatment is the mainstay. Lesions have been reported to be removed by radical resection or in a conservative way by

local excision or enucleation with subsequent curettage. As there is no difference in outcome between patients treated in a more limited way and those treated by major surgery, it seems appropriate to recommend a conservative surgical approach when managing JOF.[5]

Surgical resection of JOFs resulted in a virtual absence of recurrence. Recurrence of these lesions has been associated with an incomplete excision due to the infiltrative nature of the tumor borders. It was found that enucleation and curettage had a considerably high recurrence rate, regardless of the anatomical location or the variant type of the lesion. It has been recommended that the tumor mass should be removed down to the level of normal bone with preservation of the adjacent vital structures as much as possible. Although the surgical resection of JOFs resulted in a virtual absence of recurrence, enucleation followed by peripheral osteotomy/ curettage should be the treatment of choice for both JOF variants to avoid the disfigurement usually associated with surgical resection.[13]

JOF is radioresistant, therefore, radiotherapy is contraindicated because it can cause malignant change. Despite the aggressive nature of the lesion and high rate of recurrence, malignant transformation to sarcoma has not been reported. [1,14,16] Immediate reconstruction is not advised in these cases, as the prognosis is uncertain due to the high recurrence rate. Recurrence rate ranges from 30% to 58% as reported by Johnson et al and Makek respectively and occurs after a period ranging from 6 months to 19 years.[16]

The treatment of choice in the presented case was surgical resection and is been followed up for 12 months with no signs of recurrences. Reconstruction of these post-tumour ablative defects poses a big challenge, particularly in children.[8] In this case, buccal fat pad allowed for interim reconstruction of the patient, but it is likely that the patient will require multidisciplinary treatment approach at a later stage.

Conclusion

Juvenile aggressive ossifying fibroma is a rare clinical entity often misdiagnosed and mismanaged due of its rapidly progressive and osteolytic nature. Careful assessment of its clinical, radiographic, and histopathologic features is necessary to overcome the diagnostic and therapeutic challenges associated with this lesion. The present case being a relatively innocuous lesion, allowed for a more conservative approach, due to lack of involvement of other facial bones. This could be attributed to awareness on the patient's end which prompted them to report early on. Timely diagnosis and intervention due to the active collaboration of two departments also played a role. Since high rates of recurrence have been reported in the literature, long term follow-up of the patients is recommended.

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