



CASE REPORT

Juvenile Aggressive Trabecular Ossifying Fibroma of Mandible: A Rare Case Report

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ABSTRACT

Fibroosseous lesions of the jaws, including juvenile ossifying fibroma (JOF), pose diagnostic and therapeutic difficulties due to their clinical, radiological, and histological variability. There are two histological varieties of it, one as psammomatoid type and second as trabecular type; here, we present a trabecular type, which is a rare variety. After the clinical examination, radiological and histological analysis, it was diagnosed as juvenile trabecular ossifying fibroma. Although JOF is an uncommon clinical entity, its aggressive local behavior and high recurrence rate means that it is important to make an early diagnosis, apply the appropriate treatment, and, especially, follow-up the patient over the long term.

Keywords: Fibroosseous lesions of the jaw, Juvenile aggressive trabecular ossifying fibroma, JOF.

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INTRODUCTION

Fibroosseous lesions of the cranial and facial bones are usually benign and tend to grow slowly. Benign fibroosseous lesions have histopathological features similar with fibrous dysplasia, ossifying fibroma, and cemento-ossifying dysplasia.^{1,2}

Ossifying fibroma, a rare tumor entity, is a well-demarcated, benign, fibroosseous tumor with capsule composed of metaplastic bone, fibrous tissue, and varying amounts of osteoid.³⁻⁵ The ossifying fibromas are subdivided into conventional and juvenile clinicopathologic subtypes.³ Conventional ossifying fibromas are usually slow growing and generally seen in the 3rd and 4th decades of life.^{6,7} They are treated with simple curettage and the recurrence is rare.⁸ It affects people of all ages, but

in contrast to the form seen in adults, the juvenile form is clinically more aggressive and tends to be recurrent.³

According to the new edition of the classification of the World Health Organization (WHO),⁹ ossifying fibromas that appear as fast growing mass between 5 and 15 years of age, radiologically well-bordered, and consistent with ossifying fibroma histologically, are referred as juvenile ossifying fibroma (JOF).

Juvenile ossifying fibroma appears at an early age and in 79% of the patients is diagnosed before the age of 15.^{2,3,10} Males and females are equally affected.¹¹ Juvenile ossifying fibroma originates from periodontal ligament and accounts for 2% of oral tumors in children.^{12,13} The JOF is located mainly (85%) in facial bones, in some cases (12%) in calvarium, and very seldom (3%) extracranially.² Ninety percent of the lesions located in the face region involve the sinuses, mainly the maxillary antra.² Mandibular lesions are seen in 10% of the cases.^{2,14} The tumor is well circumscribed by a tiny sclerotic shell of bone. It appears locally aggressive with cortical disruption and involvement of many adjacent anatomical structures. This lesion has predominating soft tissue consistency with variable amounts of internal calcification and/or linear or irregular focal bone.² It usually shows a low density mass due to cystic changes on computed tomography (CT) scans. Following intravenous injection of iodinated contrast, the lesion may show diffuse appearance enhancement.²

Histologically, JOF is characterized by the presence of cellular fibrous stroma, garland-like bony strands, and cement particles.^{2,6,11,13} The JOFs are classified into two distinct clinicopathological entities: The trabecular and the psammomatoid types. Trabecular JOF is distinguished by the presence of trabeculae of fibrillar osteoid and woven bone and psammomatoid JOF is characterized by the presence of small uniform spherical ossicles that resemble psammoma bodies.^{14,15} Psammomatoid JOF is reported more commonly than trabecular JOF.^{14,16} Psammomatoid JOF occurs predominantly in the sinonasal and orbital bones, and trabecular JOF predominantly affects the jaws. Psammomatoid JOF has aggressive behavior and it has a very strong tendency to recur.¹⁵⁻¹⁷

An accurate diagnosis of JOF is made by correlating the clinical, radiological features, and histopathological findings.²

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A case of juvenile trabecular ossifying fibroma (JTOF) of the right mandible in a 34-year-old male is presented here. The rarity of the case is the age of occurrence that is unusual for JTFO, but the lesion was undetected for a long time. The lesion was so aggressive, as it caused expansile destruction of the right mandible. Very few cases of JTOF have been reported in the literature that makes this case a unique one.

CASE REPORT

A 34-year-old male patient came with a chief complaint of painless swelling in his lower right jaw since last 15 years. Patient gave history of getting hit on the jaw with a cricket ball at school 20 years back. There was pain and swelling following the trauma, but it subsided within a few days without any treatment. However, after 5 years, he noticed a small swelling about the size of a coin in the same region. He neglected it, as it did not cause any discomfort. After a few years, he noticed that the swelling gradually increased in size for about 10 years and then there was a rapid increase in the size of the swelling during the last 5 years, and has reached the present dimension. There has been no history of bleeding, discharge, and parasthesia associated with the swelling. On general examination, the patient was conscious, cooperative, and well-oriented to time, place, and person. He was healthy, well-built, and well nourished. All vital signs were within the normal limits. On extraoral examination, there was a gross facial asymmetry due to a diffuse swelling on the right side of the lower face (Figs 1 to 3).

The swelling measured approximately about 10×6 cm in size anterioposteriorly, extending from right lower



Fig. 2: Right lateral facial profile



Fig. 3: Expansion of the right lower face



Fig. 1: Front facial profile

para symphyseal region till 2.5 cm posterior to the right ear. Superoinferiorly, it extended from right alatrugal line till 1 cm inferior to the right lower mandibular border. Skin over the swelling was tensed, shiny but without any change in color, scar, or pigmentation. On palpation, the entire inspeactory findings were confirmed. The swelling was bony hard, afebrile, and nontender. A single right submandibular lymph node was palpable, tender, and mobile. Intraoral examination revealed obliteration of the right lower buccal vestibule due to the swelling, extending from 43 till the posterior aspect of 48, which



Fig. 4: Intraoral view of lower right mandibular region

on palpation was bony hard in consistency (Figs 1 to 3). The swelling expanded the buccal cortical plate with maximum expansion in relation to 47. The surface of the swelling was of normal color with no sign of discharge. Teeth in this region were normal without any pathology (Fig. 4).

Based on history and clinical examination, a provisional diagnosis of benign odontogenic tumor was given. A differential diagnosis of a fibroosseous lesion was considered.

Orthopantomogram (OPG) (Fig. 5) revealed an expansile mixed lesion on the right side of mandible. The trabecular pattern was altered from the symphyseal region extending posteriorly to the ramus expanding it. In the angle region, the expansion was globular ascending up to the ramus expanding its posterior border involving the condyle obliterating the sigmoid notch and the anterior border of ramus. The internal structure revealed mixed radiolucent radiopaque areas in the anterior aspect and posteriorly a multilocular pattern. Neither there was displacement of teeth in the region nor were the roots resorbed.

Intraoral periapical (IOPA) radiograph from 41 to 48 (Figs 6 to 8) revealed a mixed radiolucent radiopaque lesion in the periapical region with loss of lamina dura in relation to their roots. The radiopacities were patchy with diffuse radiolucent areas in between. Right mandibular lateral occlusal radiograph (Fig. 9) shows a marked expansion of buccal cortical plate from 45 to posteriorly beyond 48. Here, a homogenous radiopaque pattern with radiolucent locules in the anterior region was appreciated.

A posteroanterior (PA) mandible view (Fig. 10) showed an expansile multilocular lesion in the right mandible

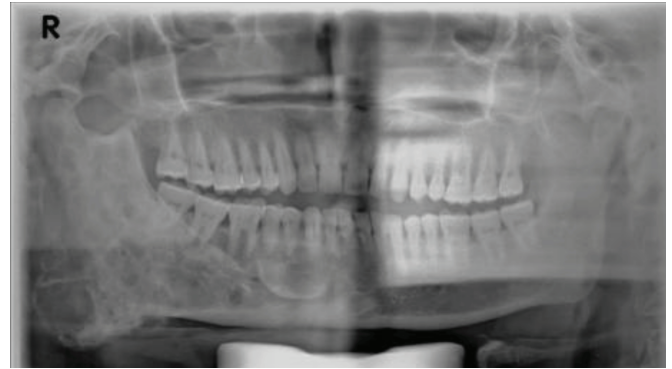


Fig. 5: Orthopantomogram

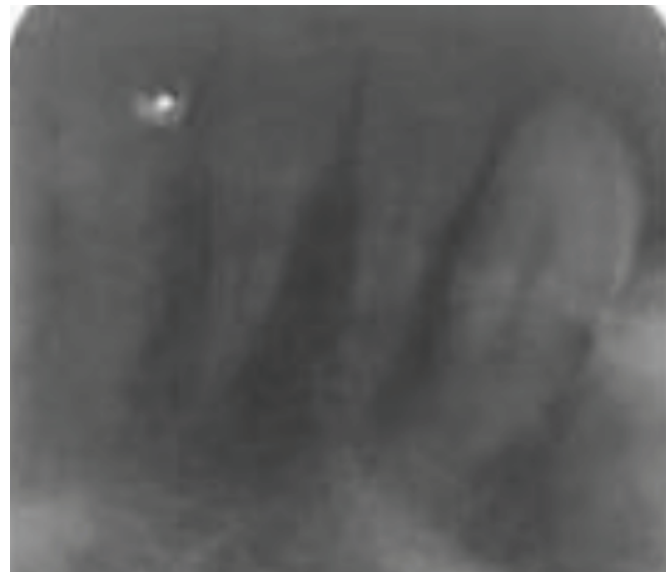


Fig. 6: Intraoral periapical – 41, 42, 43

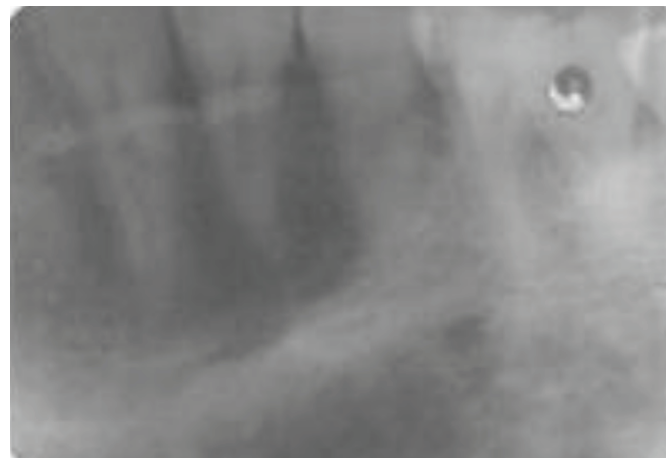


Fig. 7: Intraoral periapical – 44, 45, 46

with a globular expansion of the angle ascending up to the ramus to the condyle. There was irregular expansion of the anterior border of the ramus.

Axial CT scans (Figs 11 to 13) and three-dimensional (3D) reconstruction (Figs 14 to 16) images showed an

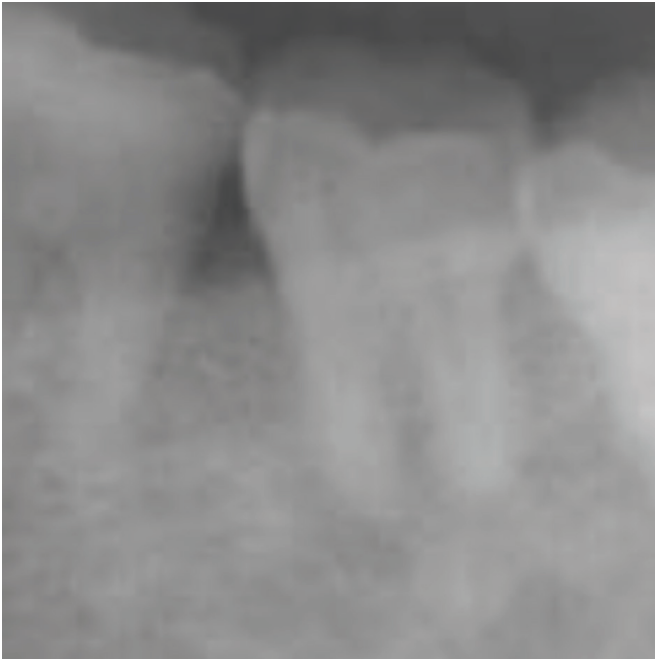


Fig. 8: Intraoral periapical–46, 47, 48



Fig. 9: Right mandibular lateral occlusal radiograph



Fig. 10: Posteroanterior mandible

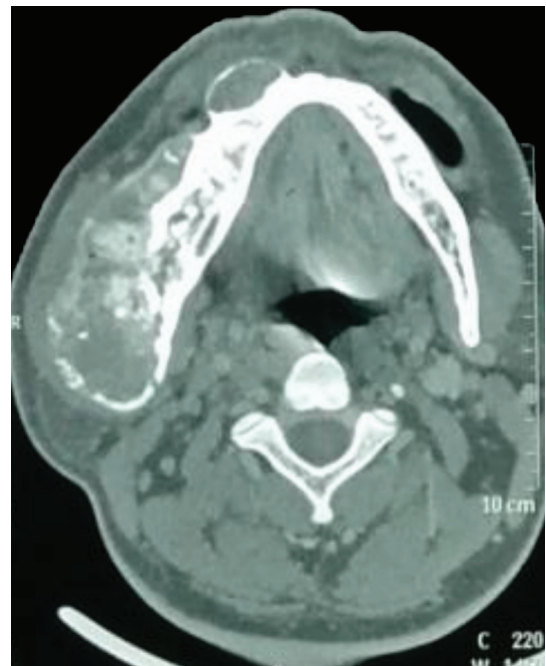


Fig. 11: Computed tomography image

expansile multilocular lesion in the right side of the mandible. The expansion was more in the buccal aspect of the mandible, exaggerated in the angle, involving the ramus, condyle, and coronoid. Small, patchy calcified masses could also be appreciated in the axial sections of CT.

Based on the radiographic pictures, a fibroosseous lesion was considered due to the multilocular radiolucent and patchy calcified areas, with a globular, centrifugal growth pattern in the angle.

A bone biopsy of the swelling revealed an aggressive type of ossifying fibroma in the mandible. Histopathologic report said the H&E section photomicrographs (Figs 17 to 19) show highly cellular connective stroma, which is composed of plump fibroblasts, collagen fibers, and many blood vessels; areas of ossification in the form of bony trabeculae and round masses of lamellar bone are seen interspersed throughout the connective tissue. Some of the trabeculae show osteoblastic rimming indicative of forming bone.

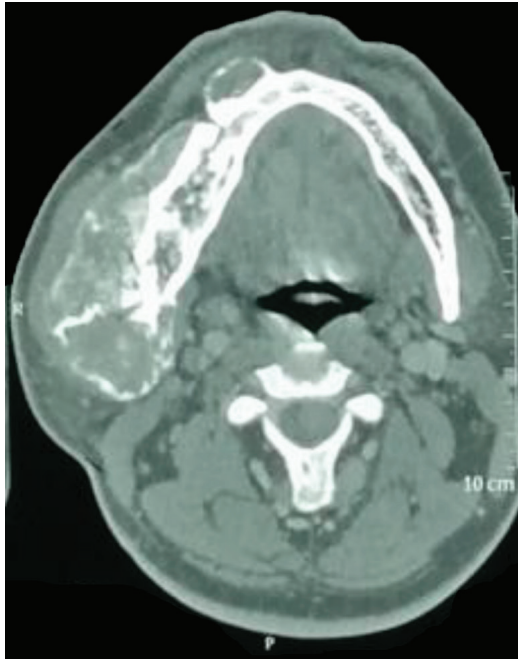


Fig. 12: Computed tomography image axial view

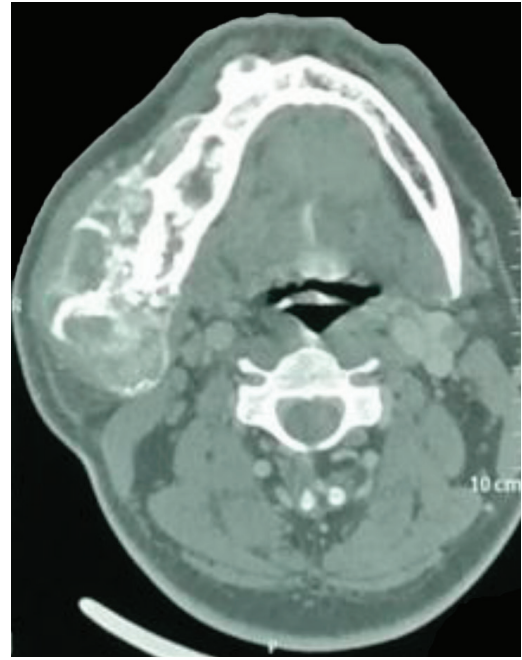


Fig. 13: Computed tomography image axial view

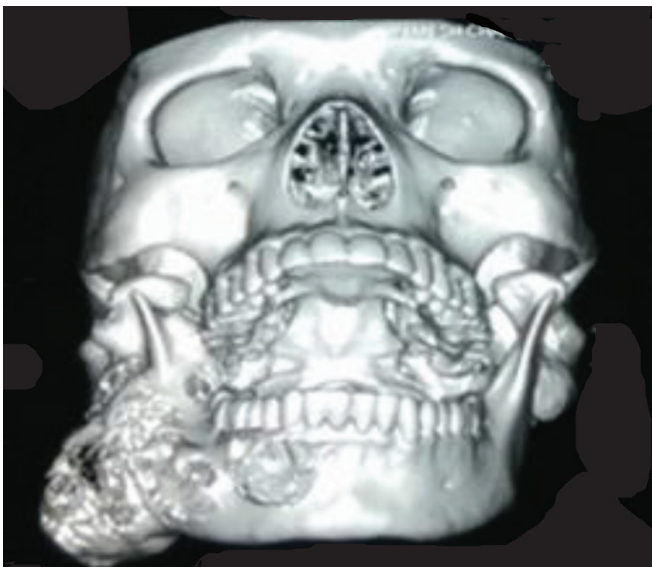


Fig. 14: Three-dimensional reconstruction frontal aspect

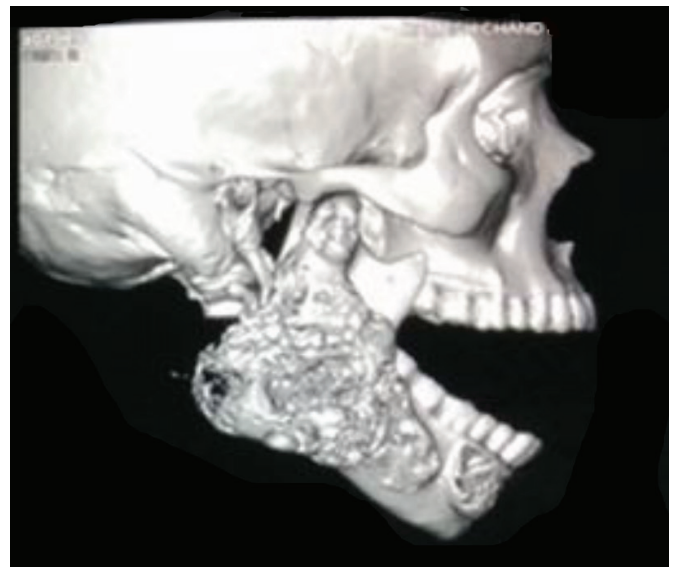


Fig. 15: Three-dimensional reconstruction lateral aspect



Fig. 16: Three-dimensional reconstruction frontosuperior aspect



Fig. 17: Histopathology photomicrogram

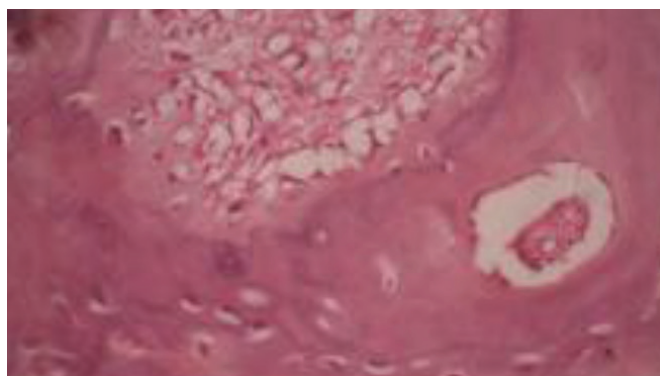


Fig. 18: Histopathology photomicrogram

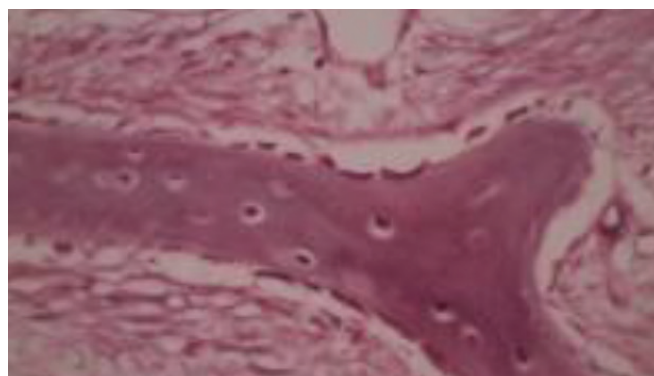


Fig. 19: Histopathology photomicrogram

DISCUSSION

Most benign fibroosseous lesions of jaws are asymptomatic and slowly progressing. Moreover, an unusual clinical presentation with apparent aggressive and destructive growth may be expected when the lesion is encountered in a younger patient.^{18,19} In our patient, the lesion was initiated at a younger age but was neglected by the patient and/or was not detected at that time.

Juvenile ossifying fibroma is a relatively rare fibroosseous lesion of the jaws characterized by its early age of occurrence, the location, radiological appearance, and the tendency to recur.²⁵ Juvenile ossifying fibroma affects both males and females equally without any significant gender predilection. However, some researchers report that it is more common among men.²⁰ The patient whose case is reported here is also a 34-year-old male. In contrast, Johnson et al²⁰ stated that mandibular tumors are more frequently common in girls between the age of 5 and 11 years or during the 2nd to 4th decades of life.⁶

A few cases of facial trauma have been suggested as a possible etiologic factor in the JOF development¹⁰ as was found in this patient also.

Juvenile ossifying fibroma is characterized as an expansile, yet with well-defined sclerotic borders, locally aggressive, and destructive at cortex on CT scan. This lesion is observed as a soft tissue mass with internal calcification, linear or irregular bone foci^{2,21-23} as was observed in our case.

Ong and Siar²⁴ presented JOF as a progressively growing lesion that can attain an enormous size with resultant deformity if left untreated. They presented a case of large cemento-ossifying fibroma involving the left mandible in a male patient. The long-lasting history of untreated JOF resulted to spontaneous fracture of mandible. Furthermore, if JOF does not have adequate surgical treatment, it may have a high rate of recurrence.^{4,24} The recurrences are generally seen at an early stage and they are more aggressive than primary lesions.⁴

As per Table 1,¹⁵ for differentiation of ossifying fibroma, juvenile aggressive trabecular ossifying fibroma,

Table 1: A working classification of the three clinicopathological variants of ossifying fibroma¹⁵

	<i>Ossifying fibroma</i>	<i>Juvenile trabecular ossifying fibroma</i>	<i>Juvenile psammomatoid ossifying fibroma</i>
Age (mean)	10–50 years (35)	2–30 years (10)	3 months–72 years (20)
Female:male	2:1	1.2:1	1.3:1
Site	Tooth-bearing areas Mandible 70% Maxilla 30%	Mandible 50% Maxilla 44% Sinonasal 6%	Sinonasal 62% Maxilla 20% Mandible (ramus) 10% Cranium 8%
Radiology	Well circumscribed Unilocular Variable radiopacity	Well circumscribed Speckled calcifications	Well circumscribed Expands and fills paranasal sinuses— “aggressive pattern” tends to be radiopaque
Histopathology	Cellular fibrous tissue, storiform pattern, separate trabeculae of woven bone occasional spherical, psammomatoid calcifications, encapsulated	Densely cellular, immature osteoid and in trabecular pattern, osteoblast rimming and cellular osteoid.	Densely cellular, spherical cementumlike psammomatoid calcifications, may permeate cancellous bone, myoid areas giant cells, hemorrhage
Synonyms	1. Cemento-ossifying fibroma 2. Cementifying fibroma 3. Periodontoma	1. Juvenile aggressive ossifying fibroma 2. Juvenile ossifying fibroma WHO type 3. Trabecular desosteoblastoma	1. Cemento-ossifying fibroma 2. Juvenile ossifying fibroma with psammomatoid ossicles 3. Juvenile ossifying fibroma psammomatous desosteoblastoma

WHO: World Health Organization

Table 2: Differences in clinical and dermographic presentations of psammomatoid juvenile ossifying fibroma and trabecular juvenile ossifying fibroma (adapted and modified Ref.¹⁵)

Type	Site	Age	Mean age	Gender	Histopathology
Psammomatoid juvenile ossifying fibroma	More common in sinonasal and orbital bone	3 months 72 years	16–33 years	M>F More common 1.2:1	Small uniform spherical ossicles resembling psammona bodies
Trabecular juvenile ossifying fibroma	Maxilla>mandible In mandible, body>ramus	2–12 years	8 1/2–12 years	M>F less common	Trabeculae of fibrillar osteoid and woven bone

and juvenile aggressive psammomatoid fibroma, the features of the present case can be diagnosed as juvenile aggressive trabecular ossifying fibroma.

According to the clinical features given by the El Mofty (Table 2),¹⁵ which states that a predominance of maxilla for trabecular pattern does not hold good in our case as it is in the mandible. Further, the table mentions about a predominance for body of mandible as compared with ramus. Age does not coincide with our case, as it was detected late but was initiated early. Histopathologically, findings of the biopsy coincide with both Tables 1 and 2.¹⁵

CONCLUSION

Although JOF is an uncommon clinical entity, its aggressive local behavior and high recurrence rate means that it is important to make an early diagnosis, to apply the appropriate treatment, and follow-up the patient over the long term. It is also important to evaluate the radiographs and correlate with its clinical findings and confirm with histopathology, as there are limited cases of this entity reported so far.

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