Annals of Clinical Case Reports

Case Report
Published: 25 Feb, 2025

A Rare Case of Triple - Recurrent Juvenile Trabecular Ossifying Fibroma

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Abstract

Juvenile ossifying fibroma (JOF) is a rare and aggressive benign tumor, most commonly affecting the cranial and facial regions in children and adolescents. It is locally aggressive and has a high recurrence rate if incompletely excised. Based on histology, JOF is divided into juvenile psammomatoid ossifying fibroma (PSOF) and juvenile trabecular ossifying fibroma (JTOF). Complete excision of the tumor is essential to minimize the risk of recurrence. Large lesions result in cosmetic imperfections, which later may require plastic surgery.

We report the case of a 14-year-old male with a progressively enlarging left maxillary swelling over two years, causing facial asymmetry and impaired vision in the left eye. Imaging revealed a large,expansile lesion with calcifications, displacing adjacent structures, suggesting juvenile ossifying fibroma. Surgical excision via Caldwell-Luc approach was performed, and histopathological examination confirmed the diagnosis of trabecular JOF. Post-operative recovery was uneventful. This case highlights the importance of early diagnosis and timely surgical intervention to prevent complications and improve patient outcomes.

Keywords: Trabecular Juvenile Ossifying Fibroma, Recurrence, Facial asymmetry

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Received Date: *08 Feb 2025*
Accepted Date: *24 Feb 2025*
Published Date: *25 Feb 2025*

Citation:
Soni H, Pankajkumar Trivedi D, Pankajkumar Trivedi R, Hasmukhbhai Vaghela M, Bhavesh Desai O, Patel JM. A Rare Case of Triple - Recurrent Juvenile Trabecular Ossifying Fibroma. Ann Clin Case Rep. 2025; 10: 2727.
***ISSN: 2474-1655**.*

Introduction

Fibro-osseous lesions of the bones mainly constitute fibrous dysplasia,ossifying fibroma and cemento-osseous dysplasia. Fibrous dysplasia is a rare bone disorder marked by fibrous tissue proliferation, affecting one or multiple bones [1]. Cemento-osseous dysplasia is a jaw-specific reactive process replacing bone with fibrous tissue [1]. Juvenile Ossifying Fibroma (JOFs) is a rare, benign lesion mainly arising from craniofacial bones. JOF is basically of two types, based on histology: Psammomatoid ossifying fibroma (PSOF) and juvenile trabecular ossifying fibroma (JTOF). It exhibits locally aggressive behaviour and a high likelihood of recurrence [2]. According to a study, JTOF was observed in 55.7% of male children, while PSOF showed no sex predilection, with a mean age of occurrence at 19.5 years [3]. It is crucial to reach at definite diagnosis to ensure proper line of management. Various radiological and histological methods assist in differentiating potential diagnosis.

Case Presentation

A 14 year old male patient presented with recurrent swelling over the left cheek, which progressively increased over a period of 2 years. As the swelling grew significantly, his vision of left eye became impaired. It also led to left nasal obstruction which caused breathing difficulties. Clinical examination revealed prominent facial asymmetry with hard, painless swelling over the left maxillary region of approximately 10cm×8cm, along with upward displacement of the left eye. Intramural examination revealed obliteration of the buccal vestibule on the left side.

There was no evidence of cervical or submandibular lymphadenopathy. He had undergone two previous surgeries for similar swelling in the maxillary region, but of smaller size, one five years ago and another four years ago. The previous surgeries were done via Caldwell-Luc approach, and the complete excision of the lesion was done.

Investigations and findings

For the investigation of this case, hematological tests including complete blood count (CBC), hemoglobin (Hb), hematocrit (Hct), platelet count, erythrocyte sedimentation rate (ESR) and prothrombin time (PT) were performed and found to be within normal ranges. Computed

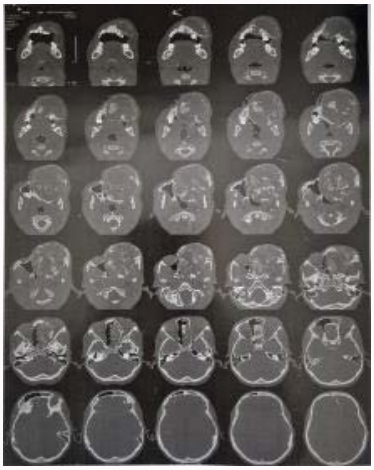

Figure 1: Ossifying fibroma.

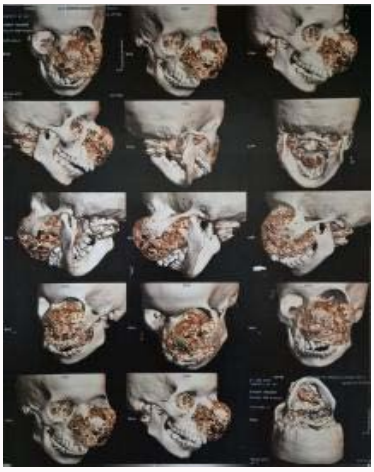

Figure 2: Ossifying fibroma.

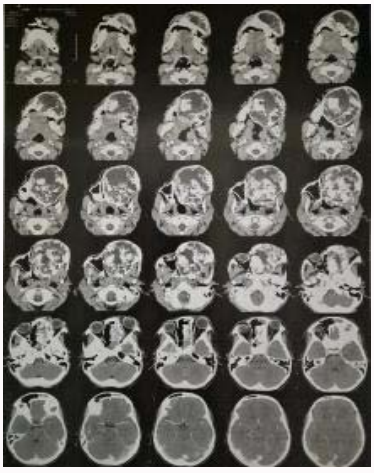

Figure 3: Ossifying fibroma.

tomography (CT) scans, and histopathological tests were employed.

A plain and contrast-enhanced CT scan of the face was performed. The radiographic examination revealed an approximately 90 × 88 × 78 mm, well-defined, oval, heterogeneously enhancing, expansile lesion of mixed density with patchy and confluent areas of calcification,

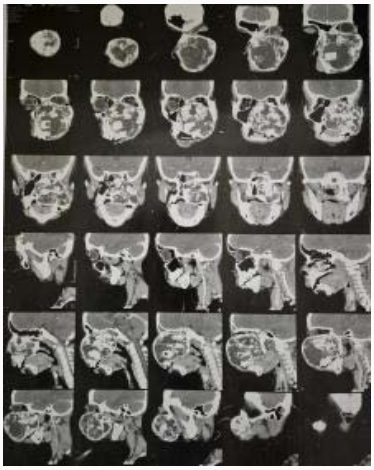

Figure 4: Ossifying fibroma.

located in the region of the left maxillary and left ethmoid sinuses. The lesion caused superior displacement of the inferior wall of the left orbit, lateral displacement and compression of the right nasal cavity, and superolateral displacement of the nasal septum towards the right. It also resulted in obstruction of the left frontal and left sphenoid sinuses. The adjacent bones showed cortical thickening and expansion, along with areas of cortical destruction. These radiological features suggest a neoplastic etiology, likely ossifying fibroma (Figures 1-4).

Diagnostic challenges and intervention

It is important to rule out other fibro-osseous lesions like fibrous dysplasia or cemento ossifying fibroma, using various modalities. The radiological and histological features serve as key differentiators in distinguishing between various possible diagnoses. The ossifying fibroma appears well-defined, frequently mixed radiolucent - radiopaque lesion with a thin, sclerotic border. Fibrous Dysplasia has a ground glass appearance with poorly defined borders, affecting one or more bones. Cemento-ossifying fibroma shows features similar to ossifying fibroma but may have a more radiolucent center and peripheral sclerosis. On histological examination, ossifying fibroma shows mature bone and fibrous tissue with a well-organized trabecular pattern. Fibrous dysplasia shows woven bone in a fibrous stroma with irregular trabeculae. Cemento-ossifying fibroma shows the presence of cementoid material, mature bone and fibrous tissue, often more circumscribed.

The primary treatment of JOF is complete surgical excision, due to its high recurrence rate. Excision of large lesions result in noticeable cosmetic imperfections. It often requires plastic surgery. Follow-up at regular intervals is essential to look for recurrence.

Management and outcome

The treatment plan of the patient included surgical excision of the fibroma using the Caldwell-Luc approach under general anesthesia. The lesion was well-defined, with areas of cystic degeneration observed. The excision was performed extensively, removing the swelling up to the zygomatic arch of the infraorbital region laterally, medially to the nasal septum, and posteriorly extending to the anterior pterygopalatine fossa (Figures 5-7).

The excised tissue was preserved in 10% formalin and sent to the Department of Pathology for histopathological analysis. The excised

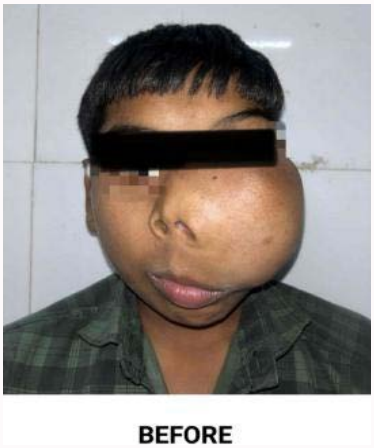

Figure 5: Preoperative images.

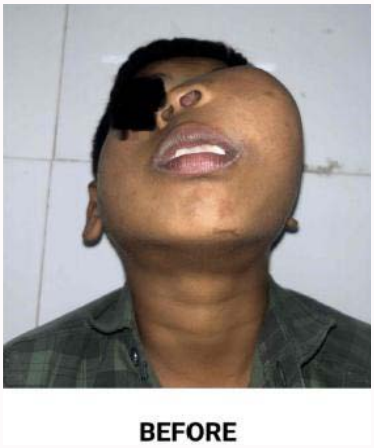

Figure 6: Preoperative images.

Figure 7: Intraoperative image of the patient.

specimen is shown in Figure 8.

In which gross examination revealed multiple brownish soft and hard tissue masses, measuring 14 × 11 × 2.5 cm in aggregate. Microscopic examination showed immature and mature bony trabeculae lined by osteoblastic rimming. The intervening stroma was fibrocollagenous and fibromyxoid, containing spindle, oval, and stellate fibroblast cells. Anastomosing bony trabeculae composed of woven and lamellar bone were observed. Areas of hyalinization and dystrophic calcification were present. In some regions, odontogenic lining was also noted. The intervening stroma exhibited mild to moderate acute inflammatory infiltrates. The imaging findings were

Figure 8: Excised specimen.

consistent with the clinical diagnosis of "maxillary sinus juvenile trabecular ossifying fibroma".

The patient previously developed this condition with similar findings at the ages of 9 and 10, indicating the third recurrence of the disease.

Postoperative recovery was uncomplicated, and the patient was discharged after a 2-week recovery period. Follow-up care was arranged to monitor for any potential complications or recurrence (Figures 9-10).

Discussion

Juvenile ossifying fibroma (JOF) is a rare neoplasm characterized by the replacement of the normal facial bone matrix in children with osteo-fibrous tissue rarely described in literature [7]. An unusual clinical presentation with rapid and destructive growth can be seen when the lesion occurs in younger patients, especially below the age of 15 years [4]. Juvenile Ossifying Fibromas (JOF) are classified into two types: the psammomatoid type, characterized by spherical ossicles, and the less commonly reported trabecular type, which is distinguished by the presence of trabeculae of osteoid and woven bone, as observed in our case [5]. The juvenile psammomatoid variant is characterized by a proliferation of benign spindle-shaped

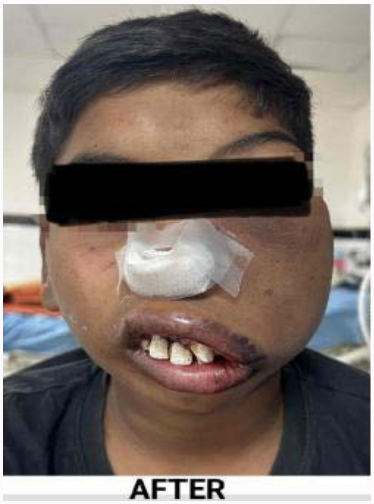

Figure 9: Post-operative images.

Figure 10: Post-operative images.

fibroblastic cells with embedded mineralized structures, which may present with round to ovoid collections of bone with an osteoid rim [6]. Microscopically, the trabeculae variant is composed of a fibroblastic spindle cell stroma, which contains osteoid matrix surrounded by osteoblasts and anastomosing trabeculae of mostly immature woven bone, often intermixed with scattered clusters of multinucleated giant cells; but cystic degeneration is rare which was seen in our case [7]. The JOF also has the high tendency to recur, unlike other fibro-osseous lesions, such as cemento-ossifying fibroma. Juvenile Ossifying Fibroma (JOF) initially expands within the bone while remaining encapsulated, making it radiologically well-demarcated. However, as the tumor reaches a larger size, it may lose its encapsulation and infiltrate the surrounding tissue by a few millimeters as we saw in our case. Differential diagnoses to consider includes osteoblastoma, osteosarcoma, and odontogenic tumors. Osteoblastoma typically appears radiologically as a cystic bone lesion with a sclerotic boundary, while osteosarcoma presents with an aggressive bone destruction pattern and abnormal soft tissue mass. Odontogenic tumors are characterized by cystic lesions connected to premolar or molar teeth. JOF, on the other side, is distinguished by its expansive growth, sclerotic borders, local aggressiveness, and cortical destruction, as observed on CT imaging. One of the other prominent differential diagnosis also includes fibrous aplasia. But the rapid growth, monostotic nature and radiographic margins differentiates JOF from fibrous aplasia [8].

Treatment of cemento-ossifying fibroma generally has been by conservative enucleation/curettage or radical surgery [7]. Conservative surgery is generally recommended even if the tumour is large with bowing and erosion of the inferior border of the mandible. Radical treatment or complete excision of the tumour such as an en bloc resection or by cadwell luc approach should only be considered if there are recurrences due to its aggressive nature which was seen in our case.

There are several advantages of conservative excision such as there is minimal morbidity after surgery, good bone formation, sensation is preserved as well as no bone graft required from a second surgical site. In long term follow-up cases where contours formed by these large lesions do not disappear completely with time, surgical intervention such as aesthetic recontouring of the bone may then be taken into consideration [9].

In the present case, the patient was diagnosed with the trabecular variant of juvenile ossifying fibroma (JOF). Given the high potential for misdiagnosis, a thorough evaluation of all laboratory investigations was crucial to ensure diagnostic accuracy. This case was particularly challenging due to the recurrent nature of the lesion, having recurred three times over the past six years, and its aggressive growth pattern. The tumor reached a considerable size, measuring 90 × 88 × 78 mm in diameter.

Considering the lesion's extensive growth and recurrence, complete surgical excision was planned to achieve definitive management. Intraoperatively, meticulous inspection of the post-surgical wound was performed to assess any potential involvement of critical anatomical structures, including the medial aspect (nasal cavity), the superior boundary (floor of the orbit), and the posterior wall of the maxillary sinus. This comprehensive surgical approach aimed to minimize the risk of further recurrence and ensure optimal patient outcomes.

Conclusion

Juvenile trabecular ossifying fibroma is a rare benign tumor with a high recurrence risk. Accurate evaluation of its clinical, radiological, and histological features is crucial to address the diagnostic and therapeutic challenges connected with it. Early diagnosis and comprehensive treatment, including complete surgical excision generally done with Cadwell-luc method and long-term follow-up, are very essential to minimize chances of recurrence, facial disfigurement, preserve functionality, and to reduce the severity of outcomes. Increased awareness among clinicians is necessary to enhance understanding and management of this rare clinical entity.

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