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INTERNATIONAL JOURNAL OF ADVANCED RESEARCH (IJAR)

Article DOI: 10.21474/IJAR01/19964
DOI URL: http://dx.doi.org/10.21474/IJAR01/19964

RESEARCH ARTICLE

A JUVENILE OSSIFYING FIBROMA IN AN 8-YEAR-OLD BOY: COMPREHENSIVE SURGICAL MANAGEMENT, RECONSTRUCTION, AND LITERATURE REVIEW

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Manuscript Info

Manuscript History
Received: 19 September 2024
Final Accepted: 27 October 2024
Published: November 2024

Key words:-
Juvenile Ossifying Fibroma, Pediatric Tumor, Fibro-Osseous Lesion, Reconstruction

Abstract

Juvenile Osseous Fibroma (JOF) is a rare, benign, yet locally aggressive fibro-osseous lesionprimarily affecting craniofacial bones in children and young adults, with a high recurrencerisk. This report describes an 8-year-old boy with a progressively enlarging, painless facialswelling. Radiological and histopathological examinations identified a trabecular JOF in theleft mandibular ramus, extending to the mandibular angle. Surgical intervention included tumor excision via a combined facial and oral approach,followed by reconstruction using a custom-made patient-specific plate to restore facialsymmetry, support feeding, and accommodate growth.MRI and CT scans are crucial for characterizing jaw tumors before biopsy, aiding in surgicalplanning. Histopathological analysis confirmed the trabecular variant of JOF. This casehighlights the diagnostic and therapeutic challenges of JOF, underscoring the need for amultidisciplinary approach. Although aggressive surgical management, such asmandibulectomy and reconstruction, is unusual for benign tumors, it is essential fortrabecular JOF to prevent recurrence.

Introduction:-

Ossifying fibroma (OF) is a benign but rare fibro-osseous neoplasm involving the jaws and craniofacial skeleton. It predominantly affects females in their third or fourth decade of life, with well-defined clinical and radiological features that typically allow straightforward diagnosis. The prognosis after conservative enucleation is generally favorable, with low recurrence rates. However, more aggressive variants, including Juvenile Trabecular Ossifying Fibroma (JTOF) and Juvenile Psammomatoid Ossifying Fibroma (JPOF), exhibit distinct clinical and radiological characteristics (1). These subtypes primarily affect pediatric patients, where conservative excision is challenging due to the presence of developing dental structures, the unencapsulated nature of the lesions, and their potential for rapid growth, causing expansion of the affected bone and cortical thinning. The management of JTOF and JPOF poses significant challenges, particularly due to the high risk of recurrence following conservative excision in the pediatric population (2). Recurrence or residual disease may necessitate radical bone excision, which complicates treatment given the reconstructive challenges in children. Here, we present a case of JTOF in a pediatric patient, treated with radical mandibulectomy and patient-specific reconstruction.

Case Report:

An 8-year-old boy presented to our department with a progressive left facial swelling that had

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developed over six months (Figure 1A). Physical examination revealed facial asymmetry, with enlargement of the left cheek, buccal widening of the jaw, and deviation of the lower incisor midline and chin toward the right side, along with ipsilateral centimeter-sized cervical lymph nodes. The tumor was painless, with no neurological deficits or dental abnormalities.

CT and MRI scans revealed an intraosseous lesion in the left mandibular ramus and angle, measuring 41 x 37 x 57 mm. The mandible exhibited expansion, a ground-glass; appearance, irregular tissue density, and annular calcifications (Figure 1B–F). Post-contrast imaging showed tissular enhancementon both modalities

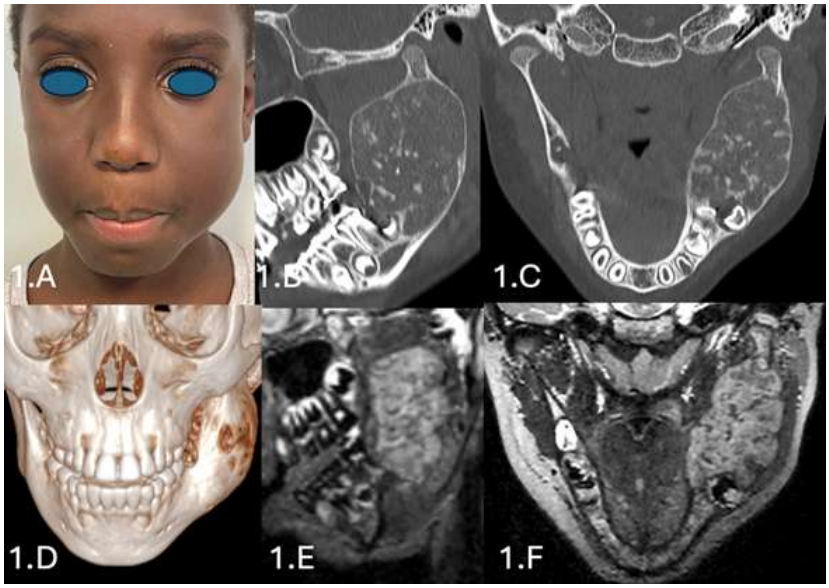

Figure 1 A:- Clinical presentation of facial asymmetry with left cheek enlargement; B, C & D:CT scans showing an expansile tumor in the left mandible, characterized by a ‘’ground-glass’’appearance, irregular tissue density, and annular calcifications; E & F: T2-weighted MRIshowing a well-defined tumor in the left mandibular ramus and angle with hypersignal.

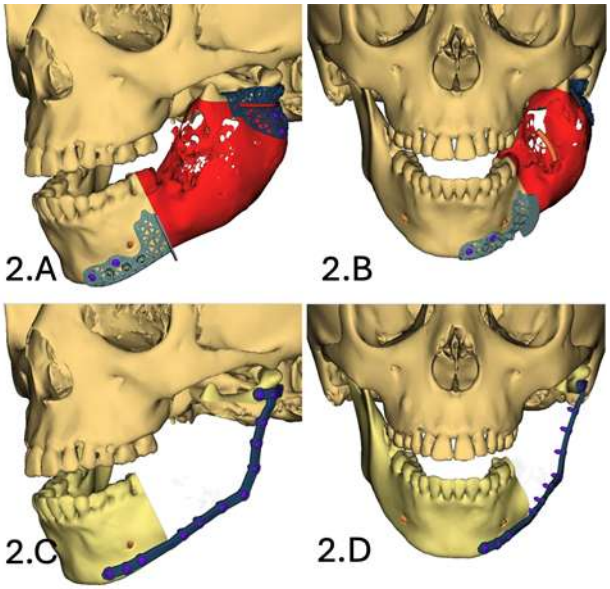

Figure 2 A & B:- Custom-made pre-drilling and cutting guides; C & D: Custom-made patient-specific reconstruction plate.

Due to the inconclusive radiological findings, a biopsy was performed undergeneral anesthesia. Histopathological analysis confirmed the diagnosis of juvenile trabecular ossifyingfibroma, characterized by a proliferation of regular spindle-shaped cells, occasionally arranged in astoriform pattern, with no significant atypia or abnormal mitotic activity. Osteoclasts and a few osteoid deposits, not bordered by osteoblasts, were also present. Additionally, ossified trabeculae, some with a distinct osteoblastic margin, were observed (Figure 2).

To excise the tumor, the patient underwent endoscopy-assisted enucleation via an intraoralapproach under general anesthesia. A buccal mucosal incision was made, followed by bone burringto remove the lesion. Endoscopic assistance facilitated thorough inspection of the enucleation cavity,ensuring adequate control of the posterior and upper margins. However, a postoperative CT scan onday 1 (POD 1) revealed residual tumor at the anterior margin near the first and second molars.

A departmental medical consultation recommended left mandibulectomy for complete tumor removal. Custom-made pre-drilling and cutting guides, along with a reconstruction plate, were designed in collaboration with Materialise (Brussels, Belgium) based on the POD 1 CT scan (Figure 2).

Virtual surgical planning enabled precise removal of the mandible from the first molar to the condylar neck while correcting facial asymmetry using mandibular mirroring.

During a third procedure, under general anesthesia, a pretragal and intraoral approach was used.

Dissection was performed from the left temporomandibular joint to the symphysis, deep to the periosteum. The left mandibular ramus and angle were freed on both the lingual and buccal sides, requiring sectioning of the pterygoid muscles and inferior alveolar nerve. The guides were positioned, and upper and lower osteotomies were performed using a Piezotome device, followed by tumor excision (Figure 3A-B). Intraoperative inspection indicated the lower osteotomy did not reach the lingual side of the first molar, necessitating an additional lingual osteotomy. The specimen was sent for histopathological examination (Figure 3C).

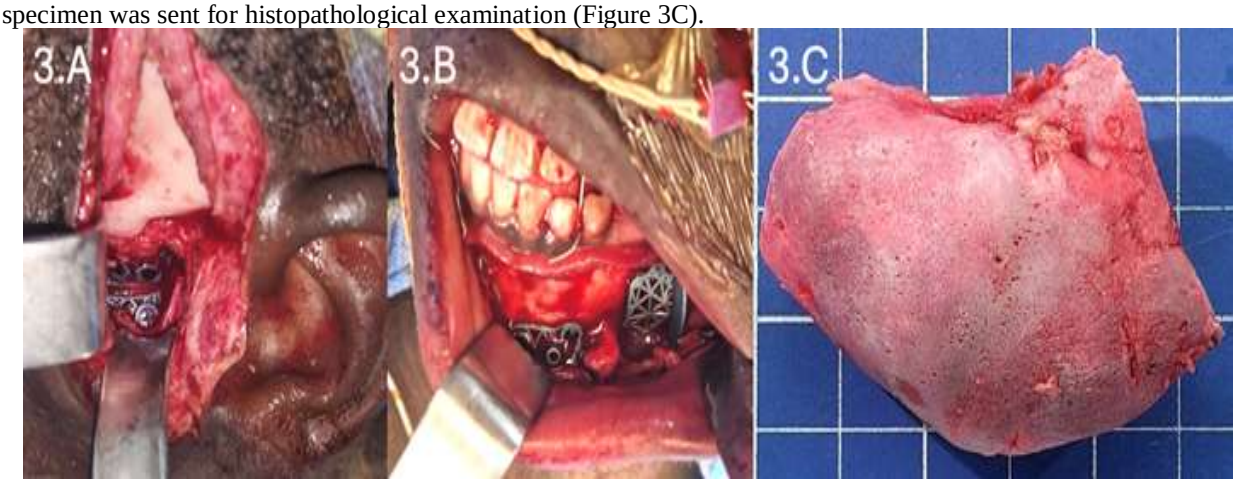

Figure 3 A & B:- Custom-made pre-drilling and cutting guides for the upper and lower osteotomies; C: Excised tumor specimen.

Reconstruction was completed via the intraoral approach, with the patient-specific plate fixed using three 5.5 mm screws at the condyle and three at the mandibular corpus. Passive mandibular mobilization confirmed satisfactory occlusion and alignment of the upper and lower incisor midline.

The incision was closed with a Redon drain. Postoperatively, the patient had mild pain, swelling, and initial difficulty centering his occlusion. A POD 1 CT scan confirmed complete resection with no residual tumor. Antibiotics were continued until POD 7, and a soft food diet was prescribed until POD 45. By one month, the patient showed improved facial symmetry, no pain, and restored incisor midline alignment.

Microscopic examination of formalin-fixed and paraffin-embedded specimens revealed the same histological observations in the initial surgical biopsy and in the two surgical specimens. The tumor was made of anastomosing

trabeculae of woven bone or osteoid, with a conspicuous rim of osteoblasts. In some areas, progressive calcification of the osteoid trabeculae was visible (Figure 4).

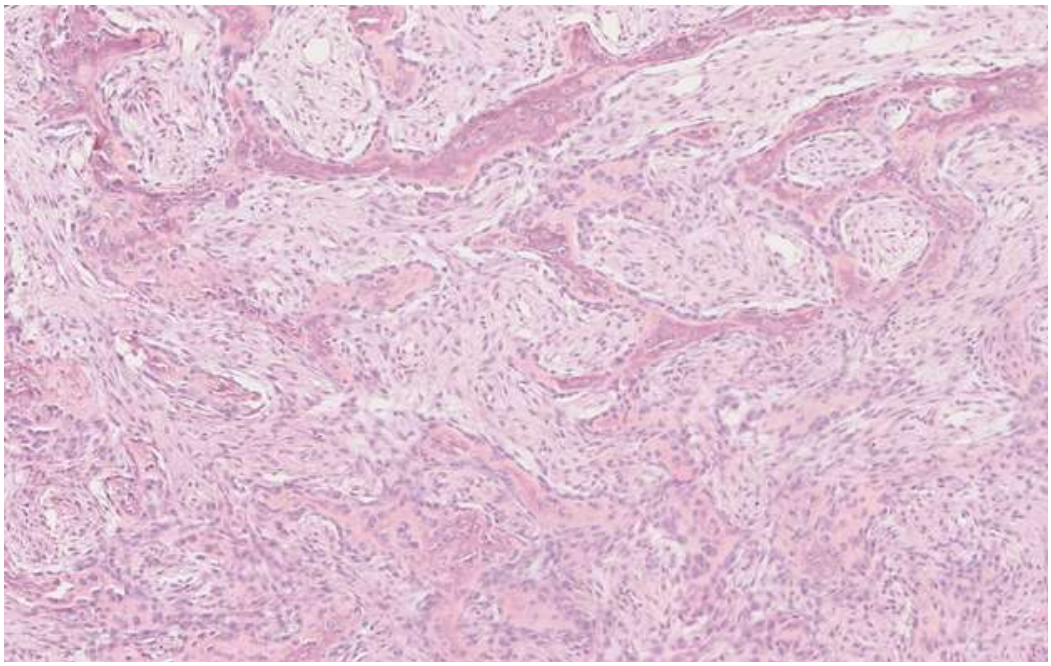

Figure 4:- Microscopic findings (H&E x10). Biphasic lesion made of a bland spindle cell proliferation and trabecular woven bone. Bone trabeculas are lined by osteoblasts. Note the progressive calcification of trabeculas (lower part: uncalcified trabeculas; upper part: calcified trabeculas).

Thetrabeculae were mixed with a moderately dense proliferation of spindle cells arranged in short fascicles or haphazardly. There was no cytological atypia and mitoses were very scarce. The tumor stroma was edematous in some areas, with no significant inflammation. Only in the second surgical specimen, some scar tissue and inflammatory infiltrate were found, due to the first surgery. All those observations were typical for the diagnosis of juvenile trabecular ossifying fibroma (JTOF). Follow-up was scheduled for 3, 6, and 12 months.

Discussion:-

Unlike classical ossifying fibroma which is commoner in the third and fourth decade of life, JOF is a is a rare, benign, yet locally aggressive tumor which primarily affects the craniofacial or the jaws bones of children and young people((3)Two main variants were described in paediatric population, trabecular juvenile ossifying fibroma (tJOF) and psammomatoid juvenile ossifying fibroma (pJOF) which exhibit differencing features in location, local behavior, mean age incidence and histological characteristics (4)

First, tJOF preferentially affects the jaws, with the maxilla being the most common site rather than extragnathic bones. That location tendancy is inversed with pJOF (5–7)Also, there is a slight male predominance and age ranges between 2 to 15 years in tJOF, whereas pJOF can affect adults, with a range from 3 months to 72 years (8)

Progressive intra-bone radiolucent tumor may let to suspect ossifying fibroma. Rapidly growing tumor, especially in children under 15 years-old may suspect its tJOF pJOF variants. However, Histological analysis stays as the cornerstone of the diagnosis in tJOF and pJOF and show a fibro osseous lesion that is characterized by cell rich fibrous tissues, bands of cellular osteoid trabeculae and giant cells (3)Additionally, some rare associations of both tJOF and pJOF variants were reported, one in the skull base and the other in the maxilla.(9).

Although conservative surgery is the treatment in OF, two types of surgery were described in the literature to remove JOF. Conservative surgery includes curettage, enucleation or peripheral osteotomies. High recurrence rates were observed with conservative surgery in case of JOF(2,4)

High recurrence rates is a proper characteristic of tJOF and pJOF compared to OF, which ranges for the tJOF from 28% to 30% (8)

Radical surgery is a more promising approach to reducing the postoperative recurrence. In 2020, Chrcanovic et Gomez, in a systematic review, compared conservatice and radical procedures. Segmental resection achieved zero recurrence in all types of JOF (TrJOF and PsJOF) and in all preferred sites, followed by marginal resection, which also achieved zero recurrence in all types of JOF and in all preferred sites, but was not statistically significant (4). and the conservative recurrence rate was 27% to 31%. Also, in 2023, Adham et al. showed that radical surgery was more effective than simple curettage or enucleation in decreasing the recurrence rate (2), with respective recurrence rates of 10.6% and compared with conservative surgery 19.7%.

After a radical surgery, reconstruction is necessary to maintain feeding, speech and social functions and depends primarily on the size of the defect (9). In children, donor site morbidity, long-term outcomes and recurrences are additional challenges that need to be considered in the choice of the reconstruction.

Reconstruction plates have the advantages of simplicity and one-stage imput possibility. However they are associated with significant disadvantages, such as high rate of exposure of the material, plate fracture, impossibility of dental rehabilitation, and a limited esthetic result (9).

In the literature, considering bone recontruction, strategies may vary. Costal bone grafts, such as iliac crest, tibia and radius bones, are described (10). In case of large osseous defect, free flaps, such as fibula and scapula free flaps may be used with one-stage reconstructions.(9,11) Others, in extensive tumors, proceed in multi-stage reconstruction. Wong et al. reported a mandible reconstruction with a first reconstruction plate fixated in the same time as the excision and then after histological analysis proving margins free of tumor, proceed a fibula free flap (11)Patient ‘specific implants have a crucial role in patients with secondary deformity of the face and can be designed to mirror the anatomy of the contralateral unaffected side and correct facial asymmetry (11).

In pediatric population, bone reconstruction is challenging as surgeries are performed in growing children. Fibula, iliac crest and scapula free flaps do not have growth potential and secondary deformities can occur at the end of the child growth. In the other hand, chondrocostal graft growth is unpredictable (12). Spontaneous bone regeneration of the mandible is an interesting and specific option in immature patients for managing the bone defect (12). Some factors were reported to help the regeneration such as the preservation of the condyle, of the periosteum, secondary infection.(12)

Donor site morbidity and with secondary free flap use at the end of the growth must be considered.

We choose to maintain the mandible space with a patient-specific implant and get the favorable condition for spontaneous bone regeneration. In case of insufficient bone regeneration, we consider that bone reconstruction using bone graft or free flaps must be harvested after safety marges checking and 1-year follow-up without reccurence.

Conclusion:-

JOF is a rare tumor with a high recurrence rate, unlike OF. Radical surgery offers the advantage of a significant reduction in recurrences. However, its morbidity means that we need to consider reconstructive modalities adapted to the growing paediatric population, whose deleterious effects need to be taken into account. The proposal of a custom-made plate reconstruction avoids these defects and offers the hope of spontaneous mandibular bone regeneration and correction of facial asymmetry, with function preserved.

References:-

1. El-Naggar AK. Editor’s perspective on the 4th edition of the WHO head and neck tumor classification. J Egypt Natl Cancer Inst. juin 2017;29(2):65-6.
2. Adham M, Dewi DJ, Reksodiputro MH, Ranakusuma R. Single stage maxillofacial reconstruction combined radical surgery for managing juvenile ossifying fibroma: A case report. Natl J Maxillofac Surg. août 2023;14(2):334-8.
3. Maria A, Sharma Y, Malik M. Juvenile ossifying fibroma of mandible: a case report. J Maxillofac Oral Surg. déc 2013;12(4):447-50.

4. Chrcanovic BR, Gomez RS. Juvenile ossifying fibroma of the jaws and paranasal sinuses: a systematic review of the cases reported in the literature. Int J Oral Maxillofac Surg. janv 2020;49(1):28-37.
5. Leimola-Virtanen R, Vähätalo K, Syrjänen S. Juvenile active ossifying fibroma of the mandible: a report of 2 cases. J Oral Maxillofac Surg Off J Am Assoc Oral Maxillofac Surg. avr 2001;59(4):439-44.
6. Reaume CE, Schmid RW, Wesley RK. Aggressive ossifying fibroma of the mandible. J Oral Maxillofac Surg Off J Am Assoc Oral Maxillofac Surg. août 1985;43(8):631-5.
7. Rinaggio J, Land M, Cleveland DB. Juvenile ossifying fibroma of the mandible. J Pediatr Surg. avr 2003;38(4):648-50.
8. Gennaro P, Gennari L, Latini L, Cavati G, Vannucchi M, Giovannetti F, et al. Maxillofacial Bone Involvement in Fibro-Osseous Lesions: Emphasizing the Significance of Differential Diagnosis. J Clin Med. 30 mai 2024;13(11).
9. Marx RE. Mandibular reconstruction. J Oral Maxillofac Surg Off J Am Assoc Oral Maxillofac Surg. mai 1993;51(5):466-79.
10. Fauvel F, Pace R, Grimaud F, Marion F, Corre P, Piot B. Costal graft as a support for bone regeneration after mandibular juvenile ossifying fibroma resection: An unusual case report. J Stomatol Oral Maxillofac Surg. oct 2017;118(5):320-5.
11. Wong WW, Martin MC. Reconstruction of Extended Orbitomaxillectomy and Hemimandibulectomy Defects With Fibula Flaps and Patient-Specific Implants. J Craniofac Surg. mars 2016;27(2):380-4.
12. Shahzad F. Pediatric Mandible Reconstruction: Controversies and Considerations. Plast Reconstr Surg Glob Open. déc 2020;8(12):e3285.