Case Report

Aggressive Juvenile Ossifying Fibroma of the Anterior Mandible

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Abstract:
Juvenile ossifying fibroma (JOF) is an uncommon fibro-osseous lesion containing different amounts of calcified tissue scattered in a cellular fibroblastic stroma. It is highly aggressive and has a strong tendency to recur. It has been recognized as a separate histopathological entity among the fibro-osseous group of lesions. The clinical behavior of Juvenile ossifying fibroma is variable and difficult to predict. This article reports a case of Aggressive JOF of the anterior mandible in a 10 year old girl.

Key words: ossifying fibroma; fibro-osseous lesion; juvenile ossifying fibroma; psammomatoid; trabecular; active fibrous dysphasia.

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Introduction:
The fibro osseous lesions of the jaws represent a diverse group of entities that are characterized by replacement of normal bone by a fibrous connective tissue matrix, with in which varying amounts of osteoid, immature and mature bone and in some instances, cementum-like material are deposited¹. Fibro osseous lesions of the jaws include developmental (hamartomatous) lesions, reactive or dysplastic processes and neoplasms.
One such rare or atypical fibro osseous lesion is Juvenile ossifying Fibroma (JOF). The term JOF describes two distinct histopathological variants: Psammomatoid and Trabecular varieties². Psammomatoid JOF is an extra-gnathic lesion occurring
predominantly in the sinonasal and orbital bones. Trabecular JOF is a gnathic lesion affecting the jaws with a predilection for maxilla\(^2\),\(^3\),\(^4\). Cases affecting the mandible have been reported; the ramus-molar area being the most common site\(^3\). This presentation highlights the third documentation of a JOF occurring in the mandibular anterior region; thus making it a rare entity.

Case report:
A 10-year-old girl presented to the department of oral and maxillofacial surgery with a rapidly enlarging mass of 20 days duration over the mandibular anterior region[Figure-1]. On physical examination, an ovoid, firm, intra oral swelling, measuring about 3x3 cm; in relation to 31, 32, 41 and 42, slightly extending into the floor of the mouth with minimal tenderness on palpation and no significant regional lymphadenopathy. There was Lateral displacement with grade II mobility of permanent mandibular incisors and deciduous mandibular canines[Figure-2a,b].

Panoramic and intra-oral periapical radiographs revealed a radiolucent mass in the symphyseal region displacing 31, 32, 41 and 42 laterally and perforating both the cortical plates of the mandible [Figure-3a, b].

CT scan revealed an expansile osteolytic mass, encompassing symphysis of the mandible causing splaying of the incisors; with an intact lower mandibular border. Few bony spicules were seen within the lesion [Figure-4 a,b].

Routine blood examination and biochemical investigations were within the normal limits. An incisional biopsy was performed, fixed in 10% formalin and sent for histopathological examination.

Correlating the age, duration, clinical behavior, radiological pattern and microscopic appearance [Figure-5a,b,c], the lesion was diagnosed as “Aggressive JOF(Trabecular variant).” Under general anaesthesia, an en bloc resection was carried out through a trans-oral approach followed by chemical...
cauterization using carbolic acid.[Figure-6 a,b,c]. The postoperative recovery was uneventful without a significant problem.[Figure-6c]. The patient was reviewed regularly and on a follow up of one year, showed no signs of recurrence.[Figure-8&9].

Discussion:

The most characteristic feature of JOF, as the name suggests, is its higher incidence in children and young adults.\textsuperscript{5,6} However, it can also occur in the older age-groups\textsuperscript{4}. Johnson et al\textsuperscript{5} have reported JOFs occurring at any age between 3 months and 72 years. Among the many classification systems for this lesion, the classification by Slootweg et al\textsuperscript{7} is noteworthy. They have classified JOF into two distinct groups, the JOF-WHO type and JOF-PO (psammoma-like ossicles) type, based primarily on the difference in the age of occurrence: the mean age of occurrence of JOF-WHO is 11.8 years and that of JOF-PO is 22.6 years.\textsuperscript{2} The most recent classification is by El-Mofty\textsuperscript{4} who identified two categories, trabecular JOF (TrJOF) and psammomatoid JOF (PsJOF), based on histologic criteria. However, the two categories also have a distinct predilection for specific age-groups: the average age of occurrence of TrJOF is 8½-12 years, whereas that of PsJOF is 16-33 years.\textsuperscript{4} Although JOF can occur anywhere in the skeleton, its highest incidence is in the facial bones, most commonly the maxilla.\textsuperscript{3,4,5,8} One clinical feature that helps differentiate TrJOF from PsJOF is the site of involvement, with PsJOF occurring mainly in the paranasal sinuses and TrJOF occurring mainly in the maxilla.\textsuperscript{4} Mandibular and extracranial involvement is rare.\textsuperscript{5} Gender predilection has been a matter of controversy, with some authors claiming no predilection for either sex, whereas Johnson et al. found a higher incidence in females\textsuperscript{3,5} and El-Mofty reported a male predilection.\textsuperscript{4}
The histogenesis of this lesion is poorly understood. Johnson et al. believe that mandibular lesions arise from the myxoid dental papilla of the developing tooth. Virtanen et al. consider JOF as a neoplasm that develops from the undifferentiated cells of the periodontal ligament. JOF usually manifests as an asymptomatic bony-hard swelling, the duration and extent of which may vary depending on the site and aggressiveness of the lesion; however, it does not demonstrate the chronic, long-standing evolution of some of the other fibro-osseous lesions. It can expand the involved bones, causing facial asymmetry. Depending on the site, symptoms such as pain, paresthesia, malocclusion, sinusitis, proptosis, etc., can also occur due to the swelling.

In general, JOF has a more aggressive growth pattern than the adult variant of ossifying fibromas. They are usually asymptomatic, exhibit rapid growth of the involved site and the first presentation will be a clinically obvious swelling. All these features were seen in our case.

Radiographically the internal structure can be radiolucent, mixed, or radiopaque, depending on the degree of calcification. Root displacement is common and resorption, though rare, can occur. The lesion can cause expansion as well as perforation. The radiographic features of JOF can resemble that of other lesions, such as fibrous dysplasia and cemento-ossifying fibroma. JOF is not capsulated but is separated from surrounding bone by a radiopaque border, and this finding can help in differentiating it from fibrous dysplasia. A 'ground-glass' appearance on radiographs has been reported. It usually has a concentric or centrifugal growth pattern, which can lead to an erroneous clinical diagnosis of cemento-ossifying fibroma. JOF has also been reported to be associated with other
bony lesions such as aneurysmal bone cyst.\(^4\) Aggressive lesions with marked destruction of adjacent structures may radiographically mimic osteogenic sarcoma.\(^8,9\)

The microscopic features of the lesion are distinctive and include a cell-rich fibrous stroma containing bands of cellular osteoid without osteoblastic lining, osteoid strands, and trabeculae of woven bone.\(^4,7,10\)

PsJOF is slightly more cellular than TrJOF. Due to the resemblance of the psammoma-like ossicles seen in PsJOF to the cementicles in cemento-ossifying fibroma, it has been argued that PsJOF is a type of cemento-ossifying fibroma.\(^2,4\)

However, the marked cellularity of JOF is in sharp contrast to the usually stroma-rich appearance of the latter group of lesions.

The aggressive nature of this entity, along with the reported high rates of recurrence (30-58%),\(^4,8\) suggests that JOF should be treated like a locally aggressive neoplasm, very much like an ameloblastoma. Surgical resection, rather than conservative curettage, is therefore the preferred line of treatment.\(^6,8\)

In the present case we followed modified Troulis et al’s staged protocol for the treatment of the jaw tumours in children.  

**Conclusion:**

Although JOF is an uncommon clinical entity, its aggressive local behaviour and high recurrence rate calls for an early diagnosis, prompt treatment and especially, long term follow up of the patient. The rapid growth rate often exhibited by these lesions can be quite alarming and cause the clinician to suspect the presence of a malignancy.

**FIGURE-1:** A 10-year-old girl presented with a rapidly enlarging mass of 20 days duration over the mandibular anterior region.
Local examination reveals an ovoid, firm, intra oral swelling, measuring about 3x3 cm; in relation to 31, 32, 41 and 42, slightly extending into the floor of the mouth and causing lateral displacement of the above mentioned teeth.

FIGURE-3a, b: OPG and IOPA radiographs revealed a radiolucent mass in the symphyseal region displacing 31, 32, 41 and 42 laterally and perforating both the cortical plates of the mandible.

CT revealed a well-defined radiolucent expansile mass encompassing symphysis of the mandible causing splaying of the incisors; with an intact lower mandibular border. Few bony spicules were seen with in the lesion.
Histopathological examination revealed highly cellular fibrous stroma consisting of spindle to polyhedral shaped fibroblast-like cells with vesiculated nuclei were seen that are arranged in a swirling pattern and a flowing whorled pattern. The highly cellular nature of the fibrous matrix and the woven bone reflected the aggressive behaviour of the tumor. Multi nucleated giant cells were also seen.

An enbloc resection was carried out through a trans oral approach followed by chemical cauterization using carbolic acid.
Figure-7 a

Figure-7b: FIGURE-7a, b: The macroscopic appearance of the lesion and the teeth involved by the mass.

Figure -8a,b:FOLLOW-UP

Pre-operative

Post-operative

FIGURE-8a, b: Follow up of one year, showed no signs of recurrence.(Extra-oral photograph)

Figure -9a,b:FOLLOW-UP

Pre-operative

Post-operative

FIGURE-9a,b: Follow up of one year, showed no signs of recurrence.(Intra-oral photograph)
References


