CASE REPORT

JUVENILE OSSIFYING FIBROMA:
REPORT OF A CASE AND REVIEW OF LITERATURE

ABSTRACT

Juvenile ossifying fibroma (JOF), a rare fibro-osseous lesion of the jaws is an actively growing lesion, well demarcated from the surrounding bone. The two histologic variants viz., WHO type (JOF, WHO) and psammomatous type (JOF, PO) though share clinical features exhibit histological picture characteristic enough to distinguish them from each other. Many authors have suggested that the JOF, PO type occurring in the gnathic regions be clubbed under the diagnosis of cemento-ossifying fibroma (COF) because of the resemblance of the calcified bodies to cementum. In this article we suggest that JOF, PO be called so, until the calcified bodies are proved to be cementum. We report a case of JOF, PO in a 4-year old boy who is probably the youngest patient reported in the literature of JOF, PO.

Key words: juvenile ossifying fibroma, fibro-osseous, psammoma, cementum

INTRODUCTION

Juvenile ossifying fibroma (JOF) is an uncommon fibro-osseous lesion of the jaws that occurs usually in children and shares features with conventional ossifying it fibroma (OF) and fibrous dysplasia (FD)[8]. JOF is a locally aggressive lesion with a tendency to recur. These lesions have been variously called as young ossifying fibroma, juvenile ossifying fibroma (JOF), juvenile aggressive/ active ossifying fibroma and trabecular desmo-osteoblastoma and are unencapsulated but well demarcated from the surrounding bone[6,7]. Here, we report a case of JOF in a 4-year-old boy and briefly review the literature.

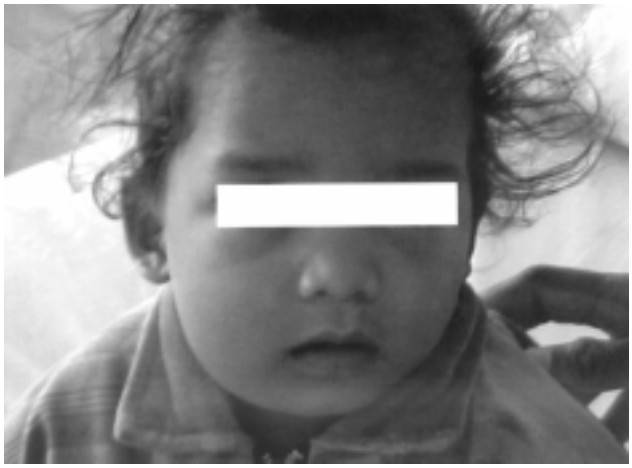

Fig 1: 4 year old boy with swelling of right side of the mandible.

CASE REPORT

In August 1997, a 4-year-old boy presented to our hospital with a swelling of the right posterior mandible of about 10 months duration. History revealed that the swelling had gradually increased in size and that it had caused the patient no pain or discomfort (Fig. 1). On examination, the swelling was bony hard, centered in the molar region with normal appearing skin and mucosa. No cervical or submandibular lymphadenopathy was noted and the jaw movements were within normal limits. A panoramic radiograph revealed a well-circumscribed radiolucency distinctly demarcated from the surrounding normal bone with minute radio-opacities scattered within it. Displacement of the

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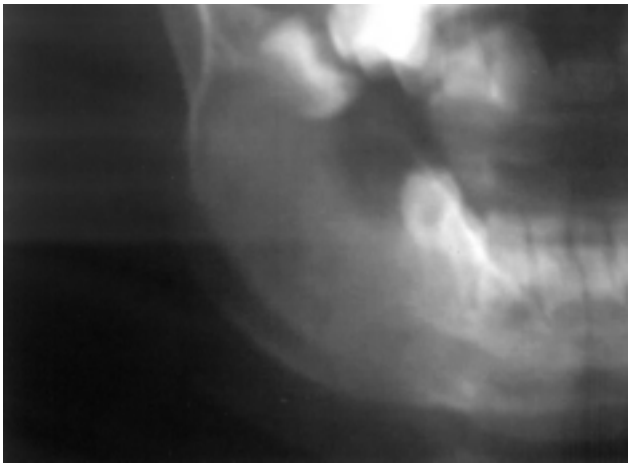

Fig 2: Well circumscribed radiolucency with displacement of deciduous teeth.

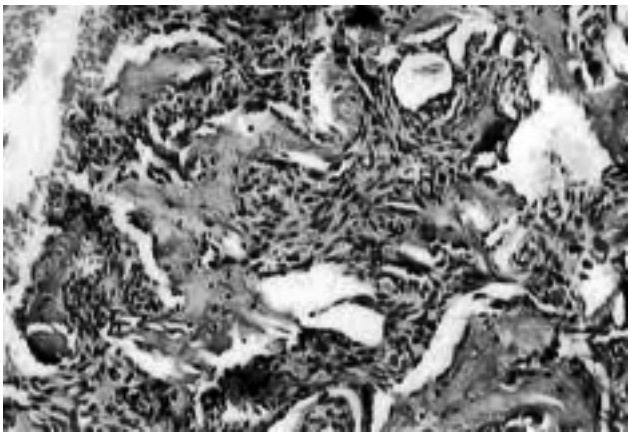

Fig 3: Higly cellular connective tissue stroma with bony trabeculae.

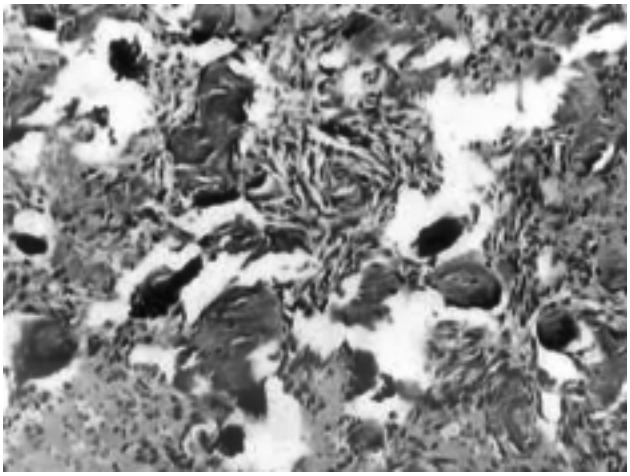

Fig 4: Numerous psammomatous bodies scattered in cellular stroma.

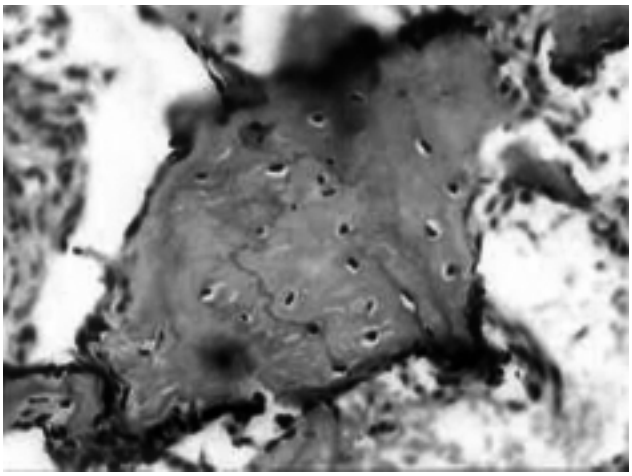

Fig 5: Bony trabeculae with osteoblastic rimming.

deciduous teeth was evident (fig.2). With this clinical and radiographic picture our initial impression was in favour of a fibro-osseous lesion. When the patient was subjected to incisional biopsy, the lesional tissue exhibited a highly fibroblastic cellular stroma in which were scattered numerous trabeculae of bone (Fig.3). A number of almost uniformly sized psammoma-like ossicles were noted strewn in this cellular stroma (Fig.4, 5). Few of the bone trabeculae exhibited osteoblastic rimming (Fig.6). A definitive diagnosis of juvenile ossifying fibroma, psammomatous type (JOF, PO) was given. The patient underwent radical surgical resection and the histopathology of the post-operative specimen was identical with the pre-operative one. The patient recovered uneventfully and is being followed up every six months ever since, with no signs of recurrence.

DISCUSSION

JOF is a relatively rare fibro-osseous lesion of the jaws defined in the WHO histologic classification of odontogenic tumors as an actively growing lesion, well demarcated from surrounding bone that is composed of cell-rich fibrous tissue containing bundles of cellular osteoid and bony trabeculae without osteoblastic rimming. Giant cells may also be present. Contemporarily, two histologic variants of JOF have been reported. JOF which fulfills the criteria described by the WHO is called as JOF,WHO type. Those characterized by the presence of a fibroblastic stroma containing small os-

sicles resembling psammoma bodies (psammoma bodies are gritty, laminated calcifications seen in meningiomas, thyroid adenomas, ovarian tumors, etc and terms like psammoma-like or psammomatous designates resemblance of calcifications to these structures histologically) have been addressed as JOF, psammomatous type (JOF, PO) and fall outside the WHO definition. JOF, PO often exhibits osteoblastic rimming. Various authors have put forward clinical and histologic comparisons between the two types[4,5,8].

JOF occurs mostly in children and adolescents though infrequently has been reported in patients over the age of 15. It affects both males and females equally, without gender predilection. The propensity to occur in the maxilla more often than in the mandible has been reported[2]. Both the types of JOF behave clinically most often in an aggressive manner with a tendency to recur. They are characterized by progressive growth with sometimes rapid involvement of the affected site. When the maxilla is involved, symptoms like obstruction of the nasal passages and epistaxis are not uncommon[2]. One of the characteristic features of JOF is its distinct radiological appearance with clear-cut demarcation from the surrounding bone distinguishing it from other fibro-osseous lesions of the jaws. The lesion is unencapsulated and given the fact that it is clinically active with its rapid expansion makes it a locally destructive tumor[1].

JOF should be distinguished from fibrous dysplasia (FD) as it shares similar clinical features[1]. But as noted earlier, radiographically JOF can be distinguished by its distinct demarcation of radiolucency from the surrounding bone whereas FD typically blends with normal bone. Histopathologically, FD is characterized by a less cellular stroma and its osteoid or bone do not exhibit osteoblastic rimming and exhibit the typical "Chinese letter" shapes[8]. Unlike JOF, FD does not warrant radical surgical management and hence its distinction is crucial. On the other hand, the stromal cellularity of JOF should be distinguished from osteosarcoma[5]. JOF is only a locally aggressive lesion that does not display any malignant mitotic figures and has never been reported to metastasize.

Data put forth by Slootweg *et al* in 1984 showed that JOF, WHO type predominantly occurred in children with the mean age of 11.8 years. The mean age for the JOF, PO type is 22.6 years. In our case diagnosed as JOF, PO the subject is a 4-yearold boy who probably is the youngest patient reported so far in the literature of JOF,PO. According to the study by Slootweg et al., both JOF, WHO and JOF, PO types did not show any gender predilection[6]. As for the site of occurrence, the data was quite divergent. In JOF, WHO type, occurrence in the maxillary region was more common according to
Slootweg *et al*, while Hamner's study showed a mandibular predilection[6]. JOF, PO was found to be occurring more often in the mandible in contrast to Makek's study, which showed a maxillary predominance[4].

Different authors have reported the recurrence rates of these lesions[4,6]. In Makek's analysis a recurrence rate of about 56% was reported for JOF, PO type and 58% for JOF, WHO type[4] . Johnson LC reported a recurrence rate of 30% for JOF, PO type while Hamner reported a recurrence rate of 50% for the same[6] . The differences in this data could be attributed to the different site distribution in the various studies.

Various authors have suggested that since the JOF, PO type is most often seen in the paranasal sinus area especially in the frontal and the ethmoidal sinuses than in gnathic regions, the formation of cementum by a benign tumor in an extragnathic site is illogical[2,6] and have emphasized that JOF, PO type be considered as a distinct entity and not be clubbed under the umbrella diagnosis of COF. Furtnermore, these authors have suggested that the cases that were diagnosed in the mandible as JOF, PO type were most likely COF with prominent spheroidal cemental masses with similarities to the psammoma-like bodies. Our contention is that when psammoma-like ossicles are seen in gnathic

areas they are not to be grouped under the spectrum of COF unless these calcified bodies are proven to be cementum. Ultrastructural study of one case of JOF, PO by Damjanov *et al* has revealed that these calcifications occurred extracellularly and not along fibrillar matrices unlike the formation of osteoid. These psammoma-like bodies did not exhibit any birefringence under polarized light[1]. Hence, when psammoma-like ossicles are seen in such instances a diagnosis outside the spectrum of COF probably should be considered until these calcifications are proven to be cementum, despite the site of occurrence. This distinction would facilitate the clinician to undertake a radical surgical management of JOF, PO, as this lesion is known for its aggressive biological behaviour and recurrence rate.

CONCLUSION

As suggested by earlier workers, it is rational to distinguish JOF, PO occurring in extragnathic sites from the spectrum of COF as cementum formation in these regions is highly unlikely. Likewise JOF, PO in gnathic regions should be segregated from the spectrum of COF until these calcifications are proven to be cementum.

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