

Case Report

Psammomatoid juvenile ossifying fibroma of mandible in a 6-year-old child

B. S. Manjunatha, Sharad Purohit¹, Sheetal Kiran², V. Naga Mahita³

Department of Dental Anatomy and Oral Biology, Faculty of Dentistry, Al-Hawaiyah, Taif University, Taif, Kingdom of Saudi Arabia, ¹Department of Oral Pathology and Microbiology, Jodhpur Dental College and Hospital, Jodhpur, Rajasthan, ²Department of Pediatric and Preventive Dentistry, Karnavati Dental College and Hospital, Ahmedabad, Gujarat, ³Department of Oral Pathology and Microbiology, Navodaya Dental College and Hospital, Mantralayam Road, Navodaya Nagar, Raichur, Karnataka, India

Abstract

Juvenile ossifying fibroma (JOF) is a rare fibro-osseous lesion of the jaw occurs at the early age of onset frequently under 15 years with a propensity to recur. It appears as a unilobulated/multilobulated lesion at an early stage followed by radiopaque appearance surrounded by radiolucent rimming at advanced stages. The psammomatous type of JOF (PsJOF) principally involves the bones of the orbit and paranasal sinuses. However in some cases, maxilla or rarely the mandible may be pretentious. Here, we report a challenging case of PsJOF in a 6-year-old child with a complaint of swelling imitating a cystic lesion in left back region of the lower jaw for 2 months. The authors propose that a careful examination and correlations of clinical, radiological, and histopathological features are essential to arrive at correct diagnosis and play a vital role in management of such patients.

Key words: Fibro-osseous lesion, juvenile ossifying fibroma, psammoma bodies

INTRODUCTION

OF is a benign neoplasm of bone that has the probable for excessive growth, bone destruction, and recurrence.^[1] Juvenile OF (JOF) is an expansile intraosseous lesion of the jaw that outdoes odontogenic lesions, seldom seen in patients < 15 years of age.^[2] JOF is a broad term used to define two distinct histopathological variants:^[3]

- Trabecular JOF has trabeculae of fibrillar osteoid and woven bone
- PsJOF is characterized by the presence of small uniform spherical ossicles that resemble psammoma bodies.

El-Mofty showed noteworthy demographic differences between the two variants of JOF. A slight male prevalence has been observed in several case reports and clinicopathological studies of PsJOF.^[3-6] Here we report a case of Psammomatoid type of JOF in a 6 year old child with a complaint of swelling in left back region of the lower jaw since two months.

CASE REPORT

A 6-year-old male patient came with a complaint of nontender swelling in the lower right back region for 2 months. The extraoral examination revealed a nontender, diffuse, and hard swelling with slight facial asymmetry located near the right body of the mandible [Figure 1a and b]. The swelling was nontender, diffuse, and hard located near the right body of the mandible. No discharge of pus or drainage from the swelling and paresthesia of the lip or restricted mouth opening were noted. Intraoral examination revealed a painless swelling measuring about 2 cm × 3 cm involving the teeth number 85 and number 46 with obliteration of

Address for correspondence:

Dr. B. S. Manjunatha,
Professor (Associate), Faculty of Dentistry, Taif University, Taif,
Kingdom of Saudi Arabia.
E-mail: drmanju26@hotmail.com

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the buccal vestibule. The associated teeth were noncarious with no lymphadenopathy. Radiographs such as intraoral periapical, panoramic, and lateral views of a skull displayed a large radiolucency with intact inferior border of the mandible on the right side. The lesion was extending from distal to the first molar until the mesial ramus of mandible anteroposteriorly. The panoramic view showed erupted first molar without root completion, and developing second molar tooth bud in posterior part of the mandible [Figure 2]. Routine blood and urine investigations were normal. No other bones of the skeleton were involved, and no other abnormalities were found on general physical examination. The medical history was noncontributory, and there was no family history of similar lesion or other skeletal diseases. Differential diagnosis included odontogenic keratocyst, cemento-ossifying fibroma (OF), and central giant cell granuloma.

Aspiration cytology was accomplished to rule out any arteriovenous malformations, cystic lesions, and the fibro-osseous lesions. Based on the clinical

and radiographic features, a diagnosis of "OF" was made. Aspiration of the lesion exhibited only blood with few surface squamous cells. The lesion was removed by surgical enucleation under local anesthesia. The surgical specimen had multiple tissue fragments [Figure 3]. The histopathological examination showed a fibrocellular connective tissue composed numerous bundles of collagen fibers with proliferating round to stellate fibroblasts. Numerous eosinophilic spherical structures dispersed in the fibrous stroma [Figures 4 and 5] were noted. Spherical structures were basophilic in center and eosinophilic in periphery [Figure 6] termed as psammoma bodies. Histopathology report was suggestive of "psammomatoid type of juvenile OF (PsJOF)." Regular follow-up after 3 months with no evidence of recurrence and lost for follow-up later on.

DISCUSSION

JOF has distinctive features from the adult variant and is associated with the early age of onset, localization

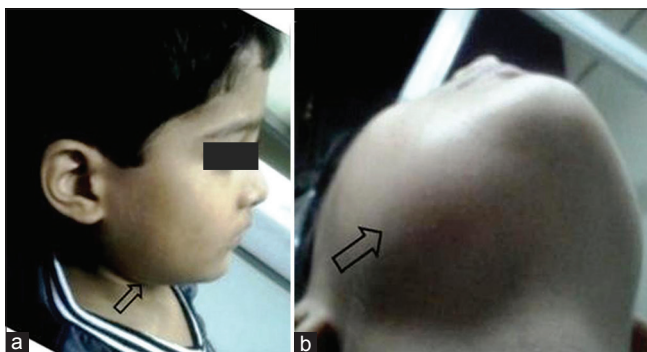


Figure 1: Extraoral photograph of the swelling angle-ramus area of the mandible (a-lateral view and b-lower border of mandible)



Figure 2: Panoramic radiograph showing large radiolucent area on right side of molar area posterior to permanent first molar

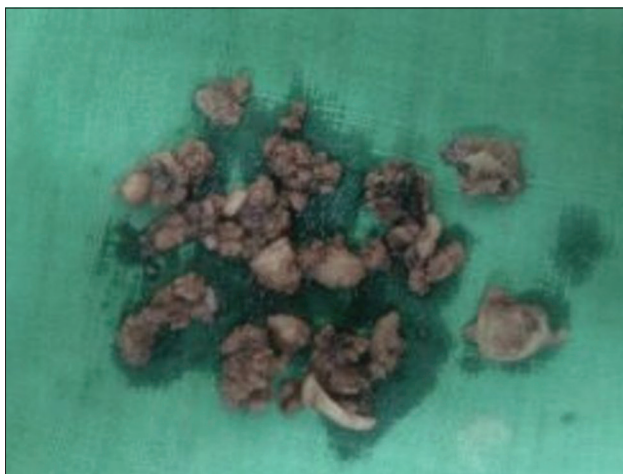


Figure 3: Multiple small bits of tissues fixed in buffered formalin received after surgical enucleation

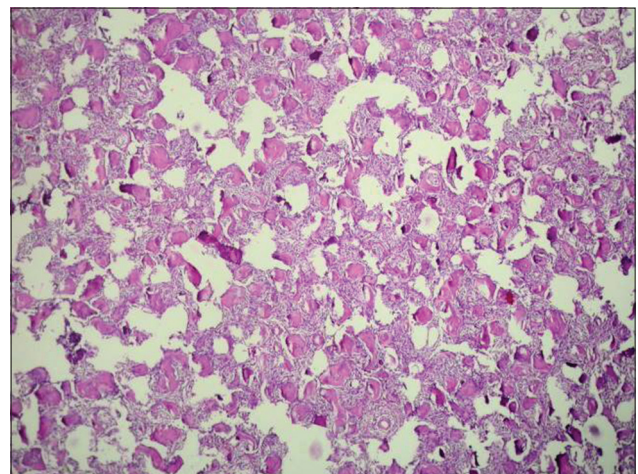


Figure 4: Photomicrograph showing fibrocellular stroma with spherical eosinophilic masses

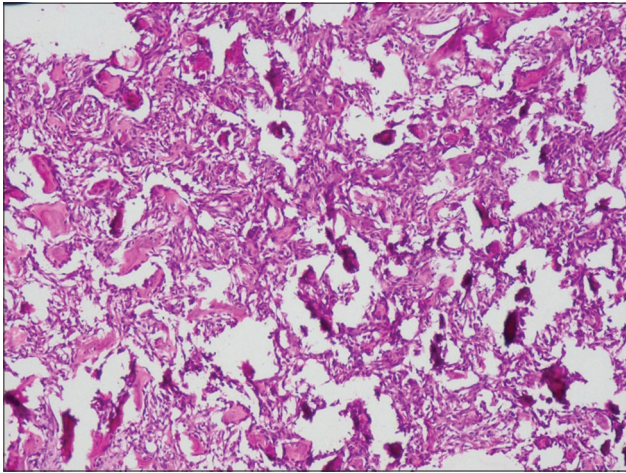


Figure 5: Photomicrograph showing cellular stroma having plump fibroblasts with small psammoma-like bodies

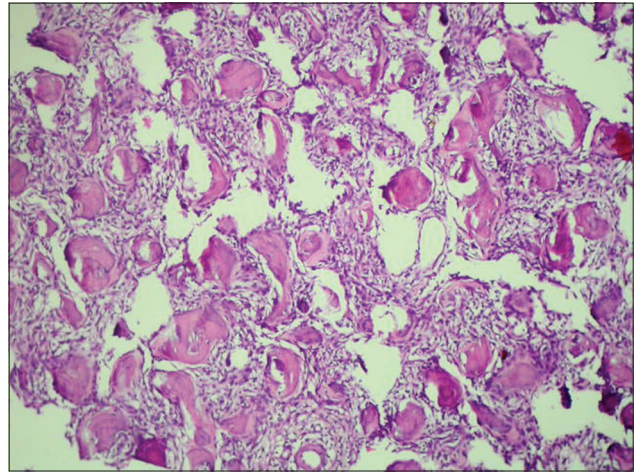


Figure 6: Photomicrograph showing spherical structures having basophilic in the center and eosinophilic in periphery

of the tumor, clinical behavior and microscopic advent, and a tendency to recur.^[7] JOF is variously named as young OF, juvenile aggressive or active OF, and trabecular desmo-osteoblastoma.^[8] Reports by Johnson *et al.*^[9] and Makek^[4] found that 70% of the PsJOF's are recognized in the paranasal sinus, 20% in the maxilla, and solitary about 10% in the mandible. Histogenesis of JOF seems to be of two probable origins and are as under:

- The extreme proliferation of periodontal ligaments and
- A metaplastic process is happening in the connective tissue fibers (nonperiodontal in origin).^[10]

PsJOF was first reported by Benjamins, in 1938, who contributed it by the title osteoid fibroma with atypical ossification of the frontal sinus. It was later termed psammomatoid OF by Gögl, in 1949 of the nose and paranasal sinuses.^[11] Margo *et al.* in 1985 familiarized the term PsJOF.^[5]

The pathogenesis for these jaw lesions is associated with the improper development of basal generative machinery that is indispensable for root formation.^[11] The periodontal derivation is being more common.^[10] Recently, Pimenta described the connotation of new tumor suppressor gene (HRPT2) mutation with OF and recommended that these lesions could ascend as a result of haploinsufficiency of the specific gene.^[12]

In this report, the patient is very young of only 6 year old and the lesion occurred in the mandible with very aggressive clinical findings having bony hard swelling with facial asymmetry. This variant of JOF is mostly a craniofacial lesion and found rarely in the mandible. This case highlights such rare case occurring in the mandibular posterior region, hence,

the documentation and to make this lesion a familiar entity to clinicians.

The pathognomonic feature of this fibro-osseous lesion is the presence of eosinophilic spherical structures discrete in a fibrous stroma. These sole spherical structures are psammoma-like bodies and differ in appearance, but typically have a central basophilic area and outlying eosinophilic fringe. Psammoma-like bodies are the trademark of this neoplasm.^[6,11] The term "psammoma" was resulting from a Greek term "psammos" sense "sand."^[13] It has been supposed to rise as a consequence of differentiation of mesenchymal cells of periodontal ligament, the multipotential predecessor cells, into cementum, osteoid, or fibrous tissue.^[14]

Clinical, radiographic, and histopathological features will be beneficial in arriving at the precise diagnosis.^[11] Until date, the accurate clinical management of JOFs is not clearly agreed. Smaller lesions are cured unadventurously by curettage or enucleation. Surgical methodology is a good decision for large and multilobulated tumors, which penetrate sinuses or fronto-nasal bones. Numerous authors have advocated that complete removal of the lesion at the initial, possible stage is the finest management choice. Late diagnosis, particularly in young patients is related with a speedy increase/growth of the tumor.^[3,15] In the present case, few clinical findings such as intact inferior border, no involvement of adjacent structures, no paresthesia of lip, and age of the patient, the case was treated by curettage without resection.

CONCLUSION

JOF is an uncommon, benign, bone-forming neoplasm with aggressive local growth. PsJOF is predominantly

a craniofacial lesion and occurs rarely in the jaws. The clinical management and prognosis of JOF is uncertain. Smaller lesions can be simply excised with surrounding marginal bone. Larger lesions, however, warrant more aggressive surgical management.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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