Psammomatoid variant of juvenile ossifying fibroma involving mandible: A rare case report

Saurabh Kumar, Arun Paul, Abhishek Ghosh, Rohan Raut

Abstract

The term juvenile ossifying fibroma (JOF) is used in literature in naming two microscopically distinct fibro-osseous lesions of the craniofacial skeleton. One is characterized by small uniform spherical ossicles resembling psammoma bodies (psammomatoid JOF [PsJOF]). The other is distinguished by trabeculae of fibrillary osteoid and woven bone (trabecular JOF). Psammomatoid ossifying fibromas represent a unique subset of fibro-osseous lesions of the craniofacial region. PsJOF has been distinguished because of its location, clinical behavior, and age of occurrence. They have distinctive histomorphologic features and a tendency toward locally aggressive behavior, including invasion and destruction of adjacent anatomic structures. It is generally seen in the younger age group, and the most common site is paranasal sinuses, orbits, and frontoethmoidal complex. We report a case of JPOF involving mandible which is rarely been described in literature. An insight into the radiographic progression of this rare entity along with the clinical feature and surgical management is discussed.

Keywords: Fibro-osseous lesion, juvenile ossifying fibroma, psammomatoid variant

Introduction

Ossifying fibromas are the well-defined type of a benign fibro-osseous lesion of jaws. It has two types conventional and juvenile ossifying fibroma (JOF). The JOF is a controversial lesion that has been distinguished from the larger group of ossifying fibroma on the basis of the age of the patients, the most common sites of involvement, and clinical behavior. JOF is an uncommon, benign, osteogenic, and nonodontogenic tumor. It is considered a separate entity from ossifying fibroma due to its locally aggressive behavior and propensity to occur at an adolescent age.[1,2] The most distinguishing feature of ossifying fibroma is the well-circumscribed appearance of the lesion radiographically, clinically, and the ease with which it is separated from the normal bone surgically.[3] The juvenile variety is usually present in children and adolescent. The lesion has a more aggressive behavior than does the conventional ossifying fibroma. Microscopically, it has a characteristic feature of irregularly mineralized cellular osteoid strands lined by plump osteoblasts.

Most JOFs arise in the vicinity of the paranasal sinuses.[4] With regard to the incidence of JOF in the jaws, there are conflicting reports of maxillary and mandibular[4,5] predilections. A recent study by El-Mofty identified two histopathological variants, trabecular JOF (TrJOF) and psammomatoid JOF (PsJOF). One clinical feature that helps differentiate TrJOF from PsJOF is the site of involvement, with PsJOF occurring in the paranasal sinuses and TrJOF occurring in the maxilla. This makes our case report unique and rare, which reports a psammomatoid variant in mandibular region.[6]

Case Report

A 20-year-old male patient reported to our unit with a complaint of swelling in the right lower jaw since 1 year. He also complained of loose mobile teeth in the same region. He has been evaluated at various places in his hometown, and previous biopsies reported were inconclusive. Extraoral examination revealed a diffuse swelling present over the right lower third of the face which was hard and nontender on palpation with no localized rise in temperature; there

Department of Dental and Oral Surgery Unit-1, Christian Medical College Hospital, Vellore, Tamil Nadu, India

Correspondence: Dr. Saurabh Kumar,
Department of Dental and Oral Surgery Unit-1, OPD Block,
Christian Medical College Hospital, Ida Scudder Road,
Thottapalayam, Vellore - 632 004, Tamil Nadu, India.
E-mail: dr.s.kumar.bds@gmail.com

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was no associated clinical lymphadenopathy. Intraorally bony expansion of alveolus was present with obliteration of gingivobuccal sulcus [Figure 1]. Orthopantomogram (OPG) done 3 months prior to the presentation [Figure 2] showed a well-defined sharply demarcated border involving the roots of right side lower canine extending up to the first molar. However, OPG at the time of presentation showed increase in radio-opacity as well as size of the lesion [Figure 3]. All the radiographs showed root resorption of the associated roots of the teeth. An incisional bone biopsy was done which showed lamellar bone along with islands of round to spindle-shaped cells displaying bland chromatin and eosinophilic cytoplasm, arranged in sheets. Spherical basophilic structures resembling cementum were evident within these islands. There was no evidence of malignancy. Based on the biopsy report and clinical presentation, a provisional diagnosis of benign fibro-osseous lesion was made. Due to the aggressiveness of the lesion, complete excision of the tumor was done, and the surgical defect was reconstructed with titanium reconstruction plate [Figure 4]. The surgical specimen on histopathology was reported as consistent with PsJOF.

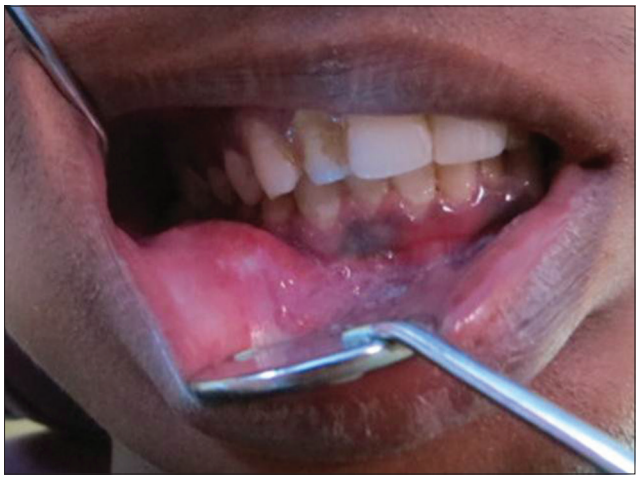

Figure 1: Intraoral examination showed bony hard swelling with vestibular obliteration

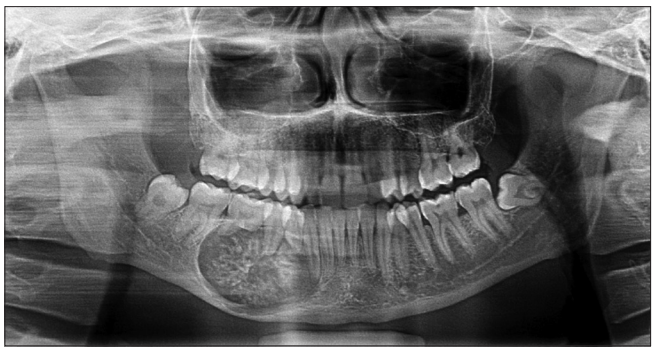

Figure 3: Orthopantomogram at the time of presentation showed increase in the size of the lesion involving the canine and the second molar, highlighting the aggressiveness of the lesion

Discussion

Juvenile aggressive ossifying fibroma (JAOF) is a relatively rare fibro-osseous lesion of the jaw characterized by the early age of onset, the location of the tumor, radiological appearance, and high recurrent potentials. JAOF may present as one of two histologic variants: Juvenile psammomatoid ossifying fibroma (JPOF) and juvenile trabecular ossifying fibroma.[7] The psammomatoid type of JOF is reported more commonly than the trabecular variety and is more aggressive with a strong tendency to recur. JPOF occurs in young patients with peaks occurring in those who are above 15 years of age and is usually characterized by rapid growth and a high tendency for recurrence.[8,9] The age of the patients ranges from 3 months to 72 years, with mean age of occurrence 17.7 years.[4,10,11] It is most commonly seen in the first and second decade of life. It shows slight male predominance with the male:female ratio of 1.2:1. The age and gender of the present case are in confinement of the data from literature. Most cases affect sinonasal area and jaws (90%), out of which 10% cases involve mandible. In the jaw, painless swelling with expansion of cortical plates is present. Mandibular ramus is more common site than the body of the mandible.[11] The present case shows one such rare presentation involving the body of the mandible. The clinical and radiographic presentations of our patient were consistent with the features of JOF with respect to the age, rapidly progressive nature, and the well-delineated radiographic picture.

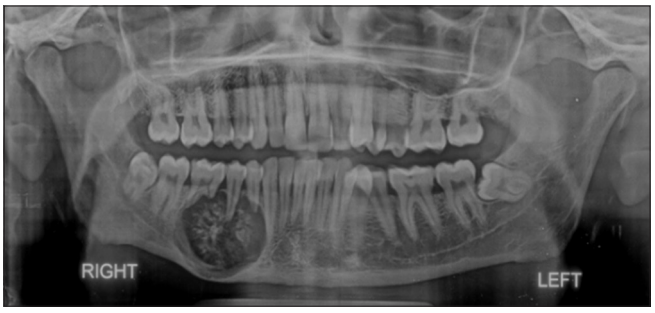

Figure 2: Orthopantomogram done 3 months prior to presentation showing well-circumscribed mixed radio-opaque and radiolucent lesion involving the roots of premolar and first molar

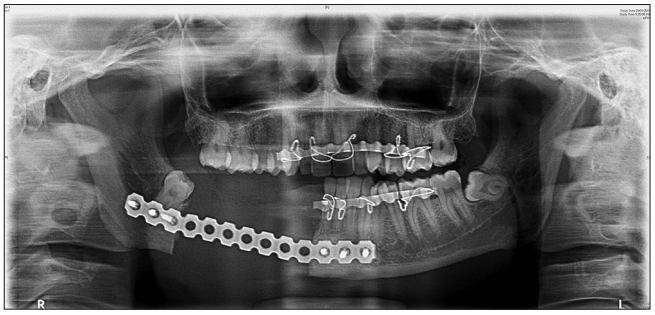

Figure 4: Postoperative orthopantomogram showing resection defect with adequate margin and reconstruction with titanium reconstruction plate

The imaging study of the present case showed a multilocular expansile lesion with a narrow transitional zone with the adjacent normal bone. These lesions usually presents with internal structure that can be radiolucent, mixed or radiopaque depending on the degree of calcification and presence of cystic areas. Root displacement usually is common and resorption, though rare, can occur. The present case demonstrated a mixed radio-opaque and lucent structure with resorption of the roots of the second molar. The lesion can cause expansion as well as perforation. The radiographic features can resemble that of fibrous dysplasia and cemento-ossifying fibroma. JPOF is not capsulated but is separated from the surrounding bone by radiopaque borders, and this finding is helpful in differentiating it from fibrous dysplasia. It usually has a centrifugal growth pattern, which can lead to an erroneous clinical diagnosis of cemento-ossifying fibroma. Other major conditions in the differential diagnosis include aneurismal bone cyst, osteoblastoma, osteosarcoma, and cemento-osseous dysplasia. Burkitt lymphoma should also be considered in the differential diagnosis of JAOF because of the similarity in the age and site of presentation, rapidity of growth, and radiolucent radiographic appearance. The characteristic histologic feature is the presence of numerous small, round ossicles, or "psammomatoid" bodies embedded in a cellular fibrous stroma.[11] The aggressive nature of this entity with high rates of recurrence (30–56%) suggests that JPOF should be treated with surgical resection, rather than conservative curettage. Recurrence may be attributed to difficulty in proper resection caused by the location of the lesion and the infiltrative nature of the tumor borders. Complete resection results in no recurrence for 6 months to 7 years[12] or more.

Because of high recurrence rate, immediate reconstruction is not advised. Secondary reconstruction may be undertaken sooner for slow-growing lesions and be delayed for fast-growing lesions. Although there are no cases of malignant transformation, rapidly growing lesions should raise suspicion and add a reason for delaying reconstruction. The prognosis is good because malignant changes and metastases have not been reported.[11,12] Radiotherapy is contraindicated because of the risk of malignant transformation and potential harmful late effects in children. Thus, appropriate recommended treatment is aggressive surgical approach followed by clinical and radiological follow-up.

Conclusion

JAOF is a rare, clinical entity often misdiagnosed and mismanaged because of its rapidly progressive and osteolytic nature. The diagnosis of psammomatoid variant of JOF, especially when it affects structures other than the paranasal sinuses can be quite challenging as was seen in this case where the mandible was involved. JOAF is treated by surgical excision and may recur if local resection is not complete. Long-term follow-up is necessary owing to its locally aggressive nature and high recurrent potentials. A careful assessment of its clinical, radiographic, and histopathologic features is necessary to overcome the diagnostic and therapeutic challenges associated with this lesion.

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Conflicts of interest

There are no conflicts of interest.

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