CASE REPORT

Trabecular Juvenile Ossifying Fibroma: A Rare Case Presentation

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ABSTRACT

Aim: To emphasise the peculiarities of Trabecular Juvenile Ossifying Fibroma (TrJOF) and also signify the anatomic variations along with detailing its radiographic and histopathological aspects.

Summary: Juvenile ossifying fibroma (JOF) is a rare fibro-osseous neoplasm that arises within the craniofacial bones in individuals under 15 years of age. Trabecular juvenile ossifying fibroma is a rare tumour entity with fibrous tissue and varying amounts of osteoid. This has male preponderance and is rare in the mandible. It is locally aggressive and spreads quickly. The following case presentation therefore highlights its unusual aspects emphasizing the clinical, radiographic and histopathological aspects along with management of the same.

Keywords: Fibro osseous lesion, Juvenile ossifying fibroma, Trabecular

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 deposited.[1] Menzel was the first to describe a so called fibro-osseous lesion in 1872, and the term "ossifying" was first popularized in the British literature, but the lesion gained its current nosology from Montgomery in 1927; subsequently with the introduction of the term "fibrous dysplasia" by Lichtenstein in 1938, it was suggested that the lesions of the jaws previously designated as "fibrous osteoma" or "ossifying fibroma" be called by the term fibro osseous lesion.[2] Ossifying fibroma of the craniofacial skeleton is separated into four clinicopathologic entities: ossifying fibroma of odontogenic origin (cemento-ossifying fibroma [COF]), trabecular juvenile ossifying fibroma (TrJOF), psammomatoid juvenile ossifying fibroma (PsJOF), and extragnathic adult ossifying fibroma.[3] According to the new edition of the classification of the World Health Organization, ossifying fibromas which appear as fast growing mass between 5 and 15 years of age, radiologically well bordered, and consistent with ossifying fibroma histologically, are referred as juvenile (aggressive) ossifying fibroma.[4] TrJOF, also known as "trabecular desmo-osteoblastoma", is defined as a lesion affecting the jaws of children composed of a cell-rich fibrous stroma containing bundles of cellular osteoid and bone trabeculae without osteoblastic rimming and aggregates of giant cells.[3] TrJOF is a gnathic lesion affecting the jaws with a predilection for maxilla. The most characteristic feature as the name suggests, is its higher incidence in children and young adults. However, it can also occur in the older age-groups.[1] 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. The recurrence rate for this lesion is about 30% - 60% as stated in literature.[5,6] The following case presentation highlights the case of 7 year old female patient with different clinical manifestations.

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CASE REPORT

A 7 year old female patient visited the Department of Oral and Maxillofacial Pathology, CSMMU, Lucknow, India with chief complaint of swelling in lower right side of the face since one and a half month. Her medical and family history was non-contributory. On examination, a solitary well defined mass having firm consistency and causing facial asymmetry on right side of face due to expansion of buccal cortical plate

could be appreciated in the soft tissue causing obliteration of right buccal vestibule in canine molar region (Fig. 1 and 2). Overlying mucosa was stretched. There was minimal tenderness on palpation and no significant regional lymphadenopathy. The mouth opening was normal and hard tissue examination revealed that patient was in mixed dentition stage. There was no decay and drifting of teeth, but grade 1 mobility was present in relation to deciduous lateral incisor, deciduous canine and deciduous first molar in the same region. Orthopantomograph revealed a well defined radiolucency extending from permanent central incisor to deciduous first molar, also involving the tooth germs of permanent lateral incisor, permanent canine, first premolar and measured roughly 2 cm in greatest dimensions (Fig. 3).

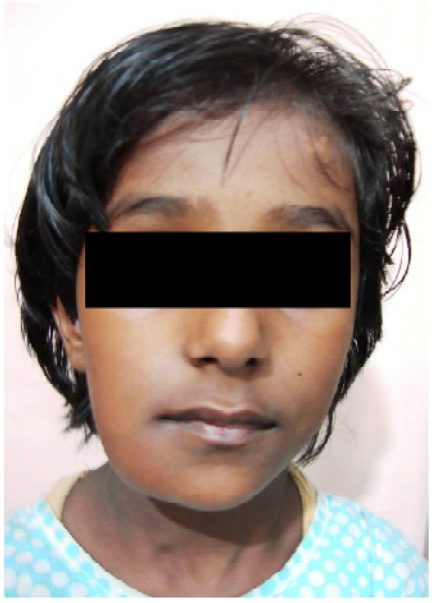

Figure 1: Clinical (extra oral) photograph showing asymmetry over lower right side of face.

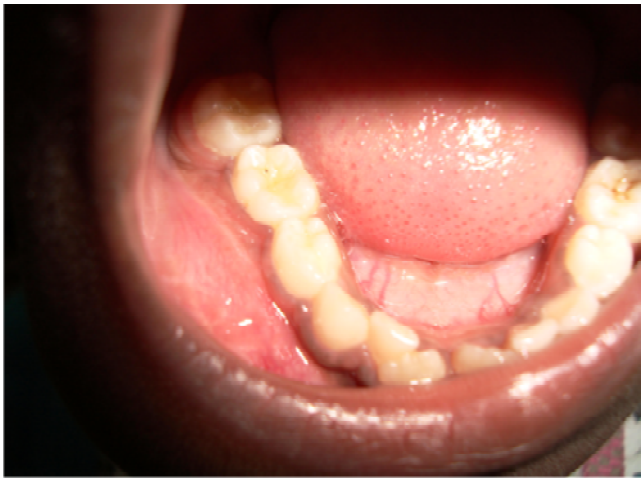

Figure 2: Clinical (intraoral) photograph showing obliteration of right mandibular buccal vestibule due to buccal cortical plate expansion.

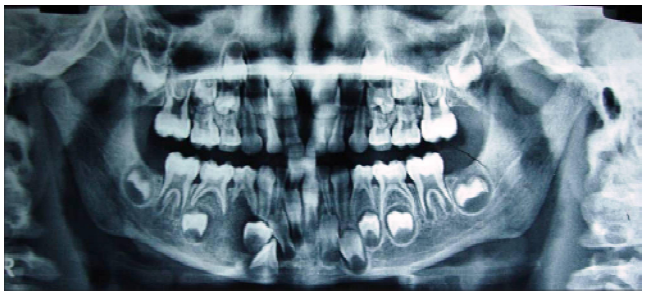

Figure 3: Orthopantomograph showing oval radiolucency extending from permanent central incisor to deciduous first molar on right side of mandible.

Routine blood examination did not demarcate any abnormalities. The biochemical investigations as regard to serum alkaline phosphatase was 346.1IU/l which was higher than the normal but serum calcium was detected to be 9.7 mg/dl which was under normal limits. An incisional biopsy was performed, fixed in 10% neutral buffered formalin and histopathological examination was done. The Hematoxylin and Eosin (H & E) stained sections revealed the presence of trabeculae of fibrillar osteoid and woven bone fragments embedded in the storiform stroma that was cell rich, with spindle or polyhedral cells that produced little collagen (Fig. 4 and 5). These osteoid elements resembled paintbrush strokes and were devoid of osteoblastic rimming at places (Fig. 6). Local aggregates of osteoclastic giant cells were invariably present in the stroma. Mitotic activity of the stromal cells was evident at few places (Fig. 4). Correlating all the above parameters a diagnosis of Juvenile ossifying fibroma was made. Excisional biopsy was planned. Although, an enbloc resection followed by chemical cauterization using carbolic acid is the preferred line of treatment. But, since the patient was in mixed dentition stage; therefore the preservation of tooth germs is of prime importance, and hence conservative curettage was performed as suggested in the literature.[1] The

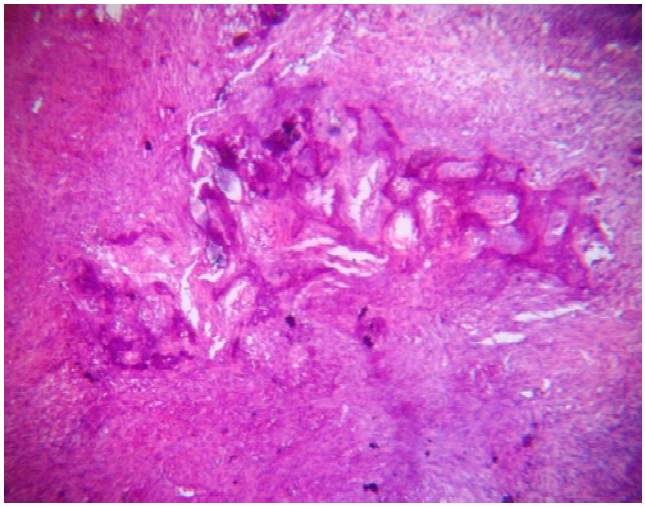

Figure 4: Photomicrograph (H & E stained; 10 X) showing paint brush like trabecular ossifications suspended in cellular stroma.

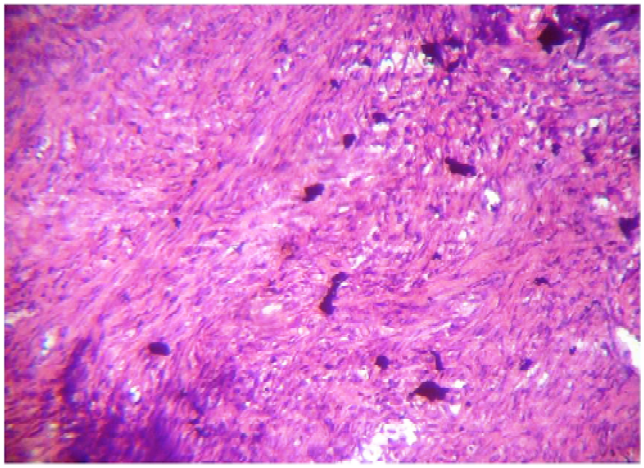

Figure 5: Photomicrograph (H & E stained; 10 X) showing high degree of cellularity in storiform pattern.

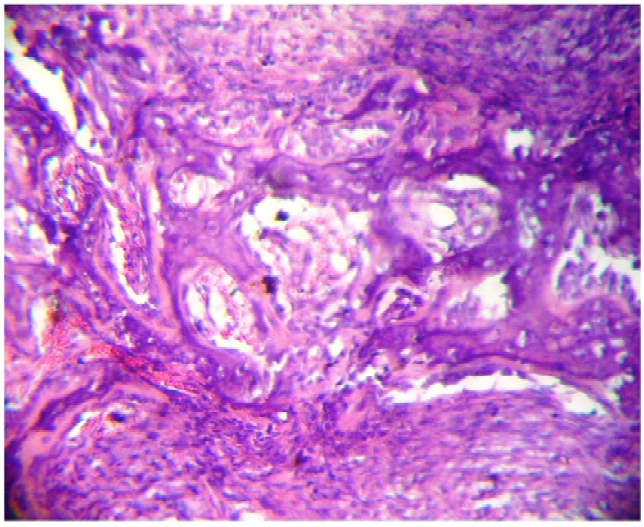

Figure 6: Photomicrohgraph (H & E stained; 40 X) showing trabecular ossifications with/without osteoblastic rimming at places.

postoperative recovery was uneventful. Since the lesion has a strong tendency for recurrence and the conservative treatment was preferred in the present case, the follow-up becomes very important. Hence, patient is currently under regular three months follow up, and remains asymptomatic till she last reported 6 months after the therapy.

DISCUSSION

It is believed that mesodermal jaw tumors arise from cells of odontogenic origin, probably from the periodontal ligament or alternatively from a primitive mesenchymal cell nest or from cells remaining after incomplete migration of the medial part of the nasal anlage.[7] Juvenile ossifying fiboma (JOF) originates from periodontal ligament and ranges 2% of oral tumours in children.[4] Virtanen *et al*.[8] consider JOF as a neoplasm that develops from the undifferentiated cells of the periodontal ligament, whereas Johnson *et al*.[9] believed that mandibular lesions arise from the myxoid dental papilla of the developing tooth, as also appears in our case. In terms of histogenesis, it probably is best described as a dysplastic lesion of membranous bone.[2] Toyosawa *et al*.[10] also suggested that osteogenic differentiation from mesenchymal stem cells is controlled by the transcriptional factor Runx2 and hence in ossifying fibroma, Runx2 staining was observed in the nuclei of spindle cells within fibrous connective tissues as well as cells on the surfaces of mineralized structures.

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.[1] In a review of a number of case series, the mean age range was found to be 8.5 to 12 years[11] as also reported in our case. Males and females are equally affected. Ninety percent of the lesions located in the face region and involve the sinuses, mainly the maxillary antra. Mandibular lesions are seen in only 10% of the cases.[4] The maxilla and the mandible are the dominant sites of incidence. Occurrence in the maxilla is slightly more frequent than in the mandible.[11] However, this trabecular variant involved the mandible. Origin in extragnathic locations is extremely rare.[3] Radiographically the internal structure can be radiolucent, mixed, or radiopaque, depending on the degree of calcification.[1] This lesion has predominating soft tissue consistency with variable amounts of internal calcification and/or linear or irregular focal bone.[4] Since the defect in our case was densely radiolucent, it manifests the early stage of the pathology. Root displacement is common and resorption, though rare, can occur[1] which was also observed with deciduous canine in this presentation. 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.[1]

In TrJOF, delicate "seams" of osteoid rimmed by osteoblasts seem to arise from within the stroma. Plump, eosinophilic osteoblasts often are incorporated within the osteoid. Scattered, irregularly shaped trabeculae of woven bone and occasional calcified spherules also are seen.[12] 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.[1] Maturation to lamellar bone is not observed.[11] The marked cellularity of JOF,[1] with a somewhat storiform pattern,[6] is in sharp contrast to the usually stroma-rich appearance of cemento ossifying fibroma.[1] Small foci of giant cells may be

present. Hemorrhage is not prominent and when present is scattered rather than seen as foci. No obvious islands of epithelium or individual epithelial cells are present.[5] Furthermore, osteblastoma, osteosarcoma and odontogenic tumours should be considered in the differential diagnosis of JOF. While the osteoblastoma is radiologically seen as cystic bone lesion with sclerotic boundary, abnormal soft tissue mass and aggressive bone destruction is seen in the osteosarcoma, and cystic lesion with unilocular or multilocular radiolucency connected to premolar or molar teeth is seen in odontogenic tumours.[4]

Slootweg and Müller[13] suggested that there were no differences between the cases that have limited surgical treatment and those with major surgery in terms of results and recommended conservative surgery, whereas Waldron *et al*.[14] emphasized that local excision and curettage should be a more preferable method and added that local surgical excision can be applied for recurrent tumour treatment. Incomplete resection causes recurrence in aggressive tumours. Therefore, some authors recommended en block resection as an adequate treatment. Curettage together with peripheral osteotomy or sometimes segmental mandibular resection and mandibular reconstruction are suggested in prevalent or recurrent cases. Sarcomatous degeneration is reported to develop in lesions that have recurrence in long term.[4] It is generally agreed that juvenile ossifying fibroma behaves aggressively locally and has a high recurrence rate when not adequately treated, the correct treatment being en bloc resection with free surgical margins.[15] Radiotherapy is contraindicated in the management of ossifying fibroma, and a "wait-and-see" policy is not generally recommended.[2] Marginal resection should be carried out where the lesion is very large with perforation/ extreme thinning of the cortical plates. Total resection or partial mandibulectomy is only advocated where the lower border of the mandible cannot be defined.[16]

However, as regards to both Trabecular and Psammomatoid variants, conservative surgical excision is the treatment of choice. Multiple recurrences have been reported but local excision is still recommended. The extent of surgical resection in both types should be dictated by the age of the patient, anatomic location of the tumor, and the effect of the tumor on surrounding vital structures.[12]

The aggressiveness and rapid growth of Juvenile ossifying fibroma calls for its early diagnosis, keen histopathological assessment, and thorough management alongwith follow up due to its high recurrence rate. This case illustrates eloquently the need of identifying trabecular variant of juvenile ossifying fibroma which had appeared in mandible of a female patient of mixed dentition stage and accounts for its conservative treatment to preserve the tooth germs of permanent teeth in contrast to its aggressive management.

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