Case Report

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CASE REPORT – TRABECULAR TYPE OF JUVENILE OSSIFYING FIBROMA

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ABSTRACT:

Juvenile ossifying fibroma (JOF) is a group of heterogeneous, benign fibro-osseous tumors of the craniofacial skeleton in young people. According to the WHO, it may present as one of two histologic variants: juvenile psammomatoid ossifying fibroma (JPOF) and juvenile trabecular ossifying fibroma (JTOF). We report a case of a 12-year-old girl who was misdiagnosed as fibrous dysplasia with aneurysmal bone cyst (ABC) like changes. The tumor was re-biopsied followed by complete resection and the specimen revealed a trabecular type of Juvenile ossifying fibroma. The clinical, radiographic, histopathologic findings and differential diagnoses of the case are presented.

Key Words: Juvenile ossifying fibroma; juvenile psammomatoid ossifying fibroma; trabecular ossifying fibroma; fibrous dysplasia; aneurysmal bone cyst.

INTRODUCTION

Juvenile Ossifying fibromas are uncommon benign lesion with aggressive local growth. These tumors are believed to originate from the periodontal ligament.[1] Because of their intense osteoblastic activity, these tumours are very aggressive and osteolytic in nature.[2] The juvenile ossifying fibroma(JOF) is a controversial lesion that has been distinguished from the larger group of ossifying fibromas on the basis of the age of the patients, most common sites of involvement and clinical behaviour. On the basis of different histologic and clinical features, El Mofty et al classified JOF into juvenile pasammomatoid ossifying fibroma(JPOF) and juvenile trabecular ossifying fibroma(JTOF).[3]

Among lesions involving the craniofacial skeleton, the JPOF is more prevalent than JTOF by a ratio of approximately 4:1. The JTOFwas first described by Reed and Hagy, in 1965. It most commonly affects maxilla than the mandible. It affects males and females equally. It can lead to facial asymmetry because of expansion of involved bones. Depending upon the site of involvement, symptoms like pain, paresthesia, malocclusion, sinusitis, proptosis, can also occur due to swelling. Root displacement is common and resorption, although rare but can occur. Radiographically, lesion may appear well defined radiolucent, radiopaque or mixed radiolucent-radiopaque, expansile lesion with sclerotic border, may lead to perforation of bone. We report a rare case of JTOF of maxilla in a twelve-year-old

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female patient with its clinical, radiological features, our experience in its management and a brief discussion regarding the case.

CASE DETAIL

A twelve-year-old female patient reported with chief complaint of swelling on left mid-face since 2 years.According to history of present illness; swelling was insidious in onset, initially small in size approximately size of a peanut.Swelling gradually increased to its present size over period of 2 years. Patient went to Guru Gobind Singh Hospital for same, where lesion was biopsied and histopathology was suggestive of fibrous dysplasia (with Aneurysmal Bone Cyst like cyst changes). Patient was referred to our centre for management of same. There was no associated history of trauma. There was no pain, paresthesia associated with the lesion. On extra oral examination, there was facial asymmetry with diffuse swelling on left side of face extending from a line joining medial canthus of eye to ala of nose anteriorly to lateral canthus of eye posteriorly and from a point 1 cm below infraorbital margin superiorly to upper lip inferiorly. There was obliteration of left nasolabial fold. Overlying skin was normal in color and texture. On Palpation, it was afebrile to touch and firm in consistency with no signs of fluctuance. No significant lymphadenopathy was evident. On Intra oral examination, there was dome shaped expansion of buccal and palatal cortex extending from region of 23 mesially to 26 distally and from buccal vestibule to gingival margin of associated teeth superoinferiorly. Overlying mucosa was normal in colour and texture except for 3 sutures present over buccal alveolar mucosa irt 23, 24. 53, 54 was retained and 23, 24 was missing. On Palpation, the swelling was firm in consistency, measuring approximately 4x3 cms in size. There were no signs of fluctuation and swelling was non tender on palpation. Associated teeth were not mobile and non tender. Based of clinical features, provisional diagnosis of benign odontogenic tumor was made with differential diagnosis of Adenomatoid Odontogenic Tumor, fibrous dysplasia, and desmoplastic ameloblastoma. Patient was advised Maxillary Occlusal view, orthopantomogram (OPG) and contrast enhanced computed tomography (CECT) scan of maxilla and mandible.

Maxillary occlusal view revealed well defined radiolucent-radiopaque lesion in left maxillary region with expansion of buccal cortex extending from mesial of 53 to distal of 26. Internally multiple thin trabeculae in criss-cross pattern were evident (FIGURE-3). 23 and 24 were impacted and lying along midpalatal suture and close to left nasal fossa (Figure 3).

OPG revealed diffuse radiolucent lesion with radiopaque foci on left side of maxilla extending mesially from 22 to 25 distally and from alveolar crest of these teeth to floor of nasal fossa superoinferiorly. There was displacement of root of 22 mesially and 25 distally. 53, 54 are retained and 23, 24 are impacted close of floor of nasal fossa. There was no

evidence of root resorption of associated teeth (Figure 4).

Axial section of CECT Maxilla & Mandible revealed lytic expansile lesion arising from left maxilla, measuring 20-29mm with multiple septations within. The teeth are displaced at the periphery of lesion, with intact inner cortex of maxilla. (FIGURE-5)

Based on radiographic findings, differential diagnosis of Ossifying fibroma, Adenomatoid Odontogenic Tumor, and Odontogenic myxoma were considered.

The incisional biopsy was performed under local anaesthesia. Histopathological examination revealed tumour tissue composed of cell rich stroma with highly active proliferative fibroblast, abundant multinucleated giant cells and many bony trabeculae which was lined by osteoblasts. These features were suggestive of Trabecular variant of juvenile Ossifying fibroma.(FIGURE-6). Patient was referred to Department of Oral and Maxillofacial surgery for management. The patient was operated under general anesthesia, as the lesion was less aggressive, complete surgical excision followed by recontouring procedure was performed. And the excised tissues were sent for histopathological examination, which further confirmed the diagnosis.

DISCUSSION

Ossifying fibroma is a rare benign osteogenic neoplasm, which accounts for 2% of oral tumours in children.[4] Juvenile ossifying fibroma is a locally aggressive neoplasm. The juvenile form can be distinguished from ossifying fibroma on the basis of its earlier in onset (childhood or adolescence), locally aggressive growth and osteoid trabeculae on histological examination.[1] According to WHO classification of Fibro-osseous lesions of jaws(2005), juvenile ossifying fibroma is defined as a lesion consisting of cells-rich fibrous tissue containing bands of cellular osteoid without osteoblastic rimming with trabeculae of more typical woven bone. JOF was earlier thought to arise from differentiation of mesenchymal cells of Periodontal Ligament, multipotent precursor cells, forming into fibrous tissue, cementum or osteoid.[5] Lawton *et al* [6] suggested that they perhaps originate from maldevelopment of tissue generating bony septa, between roots of the molar teeth. Pimento FJ et al [7] reported the association of new tumor suppressor gene (HRPT2) mutation, suggesting that the lesion could arise as a result of haploinsufficiency of the particular gene. Based of distinct histological appearance,El-Mofty (2002) classified JOF into two categories, trabecular form (JTOF) and psammomatoid form (JPOF).[3] However, the two categories also have a distinct clinical features with respect to age and site predilection. The average age of occurrence of JTOF is 8½-12 years, whereas that of JPOF is 16-33 years. The clinical feature that helps differentiate JTOF from JPOF is the site of involvement, as JPOF occurs mainly in the paranasal sinuses and JTOF occurs mainly in the maxilla.

The JTOF term was previously used by Reed and Hagy in 1965 under JOF, who reported two cases.According to literature JTOF is more common in 8-12years of age with slight male predilection [3] but in present case, in 9years old female. According to Scully and Regezzi, Initial presentation of the tumour is asymptomatic, but as it progress in size and invade surrounding bone causing the functional alterations and facial deformities [8] whereas in our case, though the swelling was asymptomatic, causing the facial deformity, but invasion to underlying bone and functional alterations were absent. Radiographically, JOF presents as a well-defined expansile, unilocular/multilocular, radiolucent, radiopaque or mixed radioluent-radiopaque lesion with sclerotic border. Involved teeth commonly shows displacement, and rarely resorption, as seen in present case. The well-defined, circumscribed nature of ossifying fibroma which distinguishes it from fibrous dysplasia. Histologically, these lesions are always benign, composed of highly vascular and fibroblast-rich connective tissue, which produces a calcified substance that often cannot be clearly attributed to either cement or bone. Clumps of osteoblasts are also present, which was also seen in present case.

A differential diagnosis of fibrous dysplasia, benign odontogenic tumor, odontogenic cyst, central giant cell granuloma (CGCG), AV malformation was considered. [5,10] Fibrous dysplasia can be ruled out radiographically as its margins blends with adjacent normal bone, whereas as it our case, it was well defined sclerotic margins. Also histologically, fibrous dysplasia has less cellular stroma with large amount of lamellar bone in typical chinese letter appearance, whereas in JTOF is characterised by trabeculae of fibrillar osteoid and woven bone. Odontogenic cyst can be ruled out based on its typical radiographic appearance of well-defined radiolucent lesion with corticated margins. Benign odontogenic tumors can be ruled out based on histogical examination. CGCG is most common in mandibular anteriors, radiographically appears well-defined unilocular/multilocular radiolucent lesion, whereas present case was evident in left maxillary anterior region, and radiographically well-defined mixed radiolucent-radiopaque lesion. Arteriovenous(AV) malformations exhibits rapid growth and usually shows thrills and bruits on palpation, which was not evident in our case.

As far as the management of these lesions is concerned, it is the locally aggressive nature with high recurrence rate of JOF necessitate for complete resection of the lesion with free surgical margins and recontouring. The “aggressive” biologic behavior of the “juvenile” variants appears to be a consequence of increased likelihood to encounter recurrences in sinonasal-based tumors initially treated conservatively. Such aggressive pattern is seen mainly in young patients because high levels of periodontal ligament activity (e.g., formation and degradation) are more commonly seen in children, and the constant irritation associated with both

primary tooth exfoliation and permanent tooth eruption can contribute for the increased prevalence of reactive lesions in younger patients. Although the classic ossifying fibromas occurs two to four times more frequently in females than in males but there is no significant sex predilection is seen in JOF. The lesion may cause a separation of the adjacent teeth, these findings are also evident in the radiograph in this case. [9]

The clinical management and prognosis of JTOF is uncertain. Surgical treatment depending upon the site and size of lesion could be simple curettage, curettage with peripheral ostectomy to block or segmental resection followed by surgical recontouring and bone grafting. Because of the aggressive nature and high recurrence rate (30-50%), JOF should be treated like a benign aggressive neoplasm, so surgical resection is preferred line of treatment.

CONCLUSION

Juvenile trabecular ossifying fibroma of peripheral variety is a rare clinical entity with unpredictable recurrence at varied stages makes the management difficult. The localized aggressive behavior and with high recurrence rate, need for an early diagnosis and prompt treatment followed by long-term follow up of the patient is essential. The possibility of recurrence can be minimized by complete elimination of etiological factors with complete surgical excision.

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FIGURES:

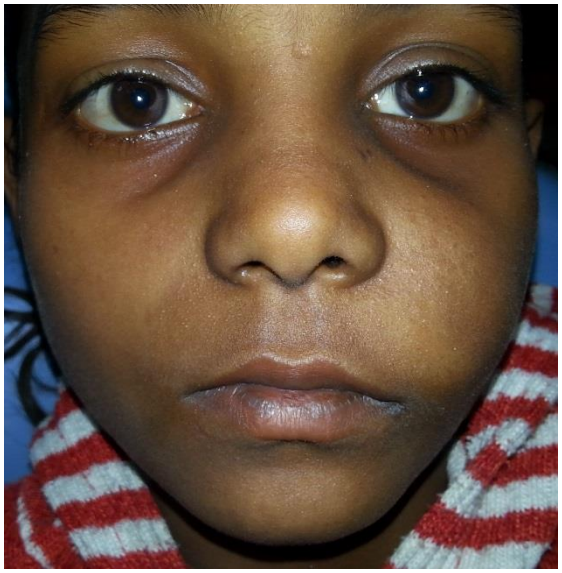


FIGURE-1: Extraoral Picture of Patient Showing Diffuse Swelling over Left Maxillary Region.

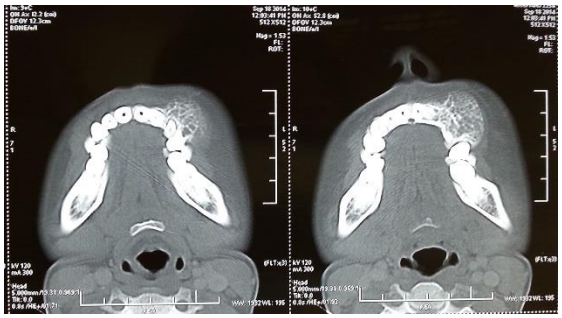


FIGURE-2: Intraoral Picture showing dome shaped Swellingin left maxilla extending from 22 to 26 with retained 53 and 54.

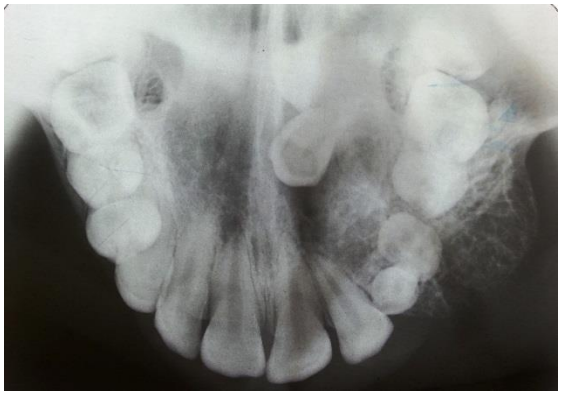


FIGURE-3: Maxillary Occlusal Radiograph Showing Well Defined mixed radiolucent-radiopaque Lesion, With thin Criss-Cross Trabeculae In Left Maxillary region, with impacted 23, 24.

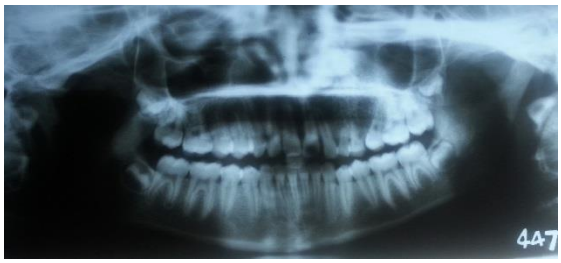


FIGURE-4: Orthopantomograph Showing Well Defined radiolucent lesion with radiopaque foci in Left maxilla extending from 22 to 25 with Displaced AndImpacted 23, 24 and retained 53 and 54.

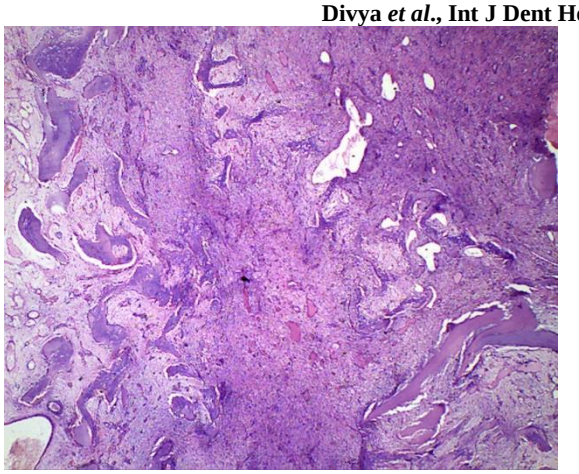
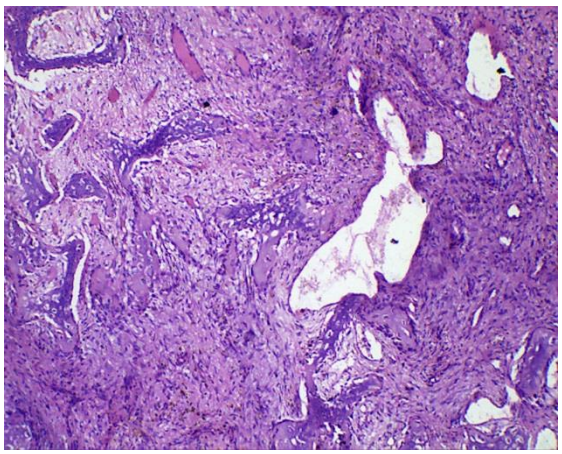


FIGURE-5 : CECT Maxilla & Mandible revealed lytic expansile lesion arising from left maxilla, measuring 20-29mm with multiple septations within.



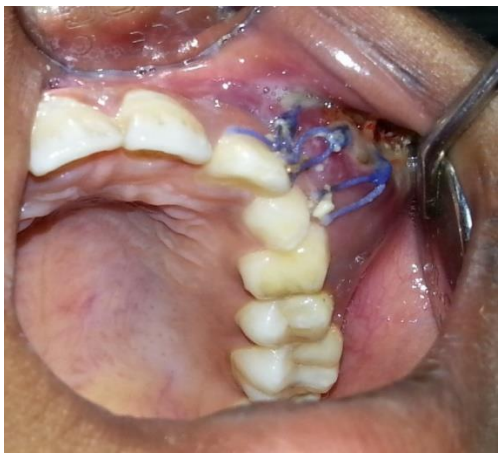


FIGURE-6(A & B): Histopathological examination revealed tumour tissue composed of cell rich stroma with highly active proliferative fibroblast, abundant multinucleated giant cells and many bony trabeculae which was lined by osteoblasts.