Case report

"A pattern based approach to diagnosis of Juvenile Ossifying Fibroma : A Case report and review of literature**"**

Abstract

Juvenile ossifying fibroma is a rare, benign and aggressive osteogenic neoplasm of the jaws commonly occurring in children and young adults. It required a postoperative follow-up over the years to rule out recurrence of the lesion. **Case report:** In this report, we present a case of a 12-year-old girl presented with a swelling in the face at the upper right maxillary region. After clinical, radiological, and histopathological examinations the diagnosis of trabecular juvenile ossifying fibroma was made. The lesion was surgically excised and followed up for two years with no evidence of recurrence.

Key words : juvenile, trabecular , psammamatoid

Introduction

The FOL comprised a diverse, and challenging group of lesions that proposed difficulties in classification and treatment. [(1)] Waldron has defined FOL as, "a group of pathological changes of jaw bones in which normal bone is replaced by fibrous tissue, with or without calcification."[(2)]

Juvenile ossifying fibroma (JOF) is an uncommon benign tumor; it is considered the aggressive counterpart of a central ossifying fibroma. In 2017, the World Health Organization (WHO) defined this lesion as a benign fibro-osseous lesion, with rapid expansive growth, that affects children and adolescents aged 8 to 12 years [(3)] and does not have any significant gender predilection.[(4)] It often diagnosed as 'juvenile ossifying fibroma', 'aggressive ossifying fibroma' or 'active ossifying fibroma' in the literature. [(5)]

This report designates a case of a JOF of a 12 year-old girl without evidence of recurrence.

Case report

A 12-yr-old female patient reported with chief complaint of painless, progressive swelling in the upper right side of face. Initially noticed as a small swelling which grew to the present size within a span of 2 yrs. There was no history of trauma. Past medical and dental history not significant.

Extra-oral examination revealed a diffuse, non-tender, bony hard swelling on the right side of face extending from right ala of nose to mid region of cheek, approx. 3-4 cm in size causing marked facial asymmetry [Figure 1]. Color and temperature of the overlying skin was normal. On intra-oral examination, a single diffuse bony hard, localized swelling extended from 11 to 15 region

with obliteration of buccal vestibule and palate. 13 tooth was missing. There was no significant intraoral finding seen(figure 2).

In radiographic evaluation, Orthopantomograph (OPG) revealed a mixed radiolucent-radiopaque lesion of size approximately 4 × 4 cm extending from 12 to 15 region. The borders of the lesion were non-corticated, well defined and showed some amount of scalloping especially in the supero-lateral aspect [Figure 3]. Cone beam computed tomography scan revealed irregular expansion of buccal and lingual cortical plates with central foci of calcification [Figures 3].

The lesion was excised with conservative approach with the provisional diagnosis of adenomatoid odontogenic tumor. On gross examination, the biopsy specimen was in multiple pieces, Brownish white in color and firm-to-gritty in consistency (figure 4). On microscopic examination, H and E stained section revealed equal amount of calcified material and fibroblastic stroma. The calcified structures consist of irregularly shaped bony trabeculae with an osteoblastic rimming. Focal areas shows clusters of multinucleated giant cells (MNGC) (figure 6). Osteoid tissue lying in cellular fibrous connective tissue stroma with plump fibroblast and spindle-shaped arranged in storiform pattern without much cytoplasm (figure 5A and 5B). Mineralization and haemorrhagic areas seen at a focal places.

Based on the clinical, radiographic and histological features, the final diagnosis given as a "trabecular type of juvenile ossifying fibroma (JOF)". 2 years follow-up showed no signs of recurrence.

Discussion

FOL literally means replacement of normal bony architecture by fibrous tissues, which may mineralize in various forms like woven, lamellar bone or cementum. They include a broad spectrum of distinct entities like fibrous dysplasia (FD), ossifying fibroma (OF), paget disease, cemento-osseous dysplasia etc. OF are rare fibro-osseous tumors. They are made up of osteogenic calcified matrix, and they are not of odontogenic origin as previously thought. Their epidemiology is poorly known because they have long been confused with cemento-osseous dysplasia. Their location is mainly limited to the craniofacial bones. [6]

Lawton et al, speculated that JOFs originate from the maldevelopment of the tissue generating the bony septa between the roots of molar teeth. [7] Pimenta et al, suggested that haplodifficiency of HRPT2 gene is responsible for occurrence of JOF . [8]

JOFs are benign, aggressive, asymptomatic and osteolytic lesion commonly seen in children and young adults. Trauma suggested as a possible etiologic factor (table no. 1) but not seen in present patient. In total reviews published by HAMNER et al.[5] and SLOOTWEG et al.[9], the mean age of onset was 11.5 and 11.8 years respectively. It may exhibit rapid growth at the involved anatomic site, resulting in considerable facial disfigurement.[10] Clinically, this lesion has in general a more aggressive growth rate than ossifying fibroma. Most cases of maxillary JOF are asymptomatic. The first clinical manifestation is a swelling of locally involved bony architecture. (table no. 1) [5]

Radiologically JOF looks as a uninoculated / multiloculated radiolucent lesion (table no. 1) in early stage of development followed by radiopaque appearance and is surrounded by a uniform radiolucent rimming at later stages.[4]

On gross examination, the specimen appear as whitish yellow, homogeneous mass with variable amount of calcified material.[4] The characteristic histologic features include a benign osteogenic, well-demarcated neoplasm composed of calcified material and a fibroblastic stroma, which may be cellular. The calcified component is usually a combination of bone trabeculae and strongly basophilic cementum-like structures with variable osteoblastic rimming. No malignant changes was observed.[11] Surgical excision is the treatment of choice. The clinical, radiographic and histopathological presentations of our patient were consistent with the features of juvenile ossifying fibroma.

Based on the pattern of mineralization, JOFs are classified into two distinct entities[4]: Trabecular and Psammomatoid types. [10] The trabecular variant is usually affects the jaws with an age range of 2–12 years and distinguished by fibrous trabeculae and osteoid with woven bone formation.[11] The psammomatoid variant is mainly located around paranasal sinuses, orbits with proliferation of benign spindle-shaped fibroblastic cells interspersed with small uniform spherical ossicles resembling psammoma bodies. [12] psammomatoid variant seen in wide age range i.e. 3 months–72 years.[11]

The differential diagnosis of FOL is a challenging task to pathologists because of their similar clinical, radiographic and histopathological behaviour. Fibrous dysplasia (FD) was ruled out, as it typically blends with the normal bone at the margins of the lesion. It comprised calcified structures consist of Chinese letter shaped bony trabeculae without an osteoblastic rimming.[10] The case discussed here presented a well-demarcated lesion from the surrounding bone with highly cellular fibrous stroma and prominent osteoblastic rimming, clinicopathologically indicative of JOF. Absence of cytological atypia and infiltration to adjacent structures in the present case ruled out osteosarcoma.[5] Cemento-ossifying fibroma (COF) considered a histological variant of ossifying fibroma, i.e. giant cells which are evidently found in JOFs are not present in COF.[13] Aspiration yielded negative result, ruling out cystic lesion.

The treatment of JOFs remains controversial. Abuzinad and Alyamani et al; proposed that conservative management with less aggressive approach must be considered.[14] Bohn et al. reported the diagnostic and therapeutic difficulties encountered in their management of JOF with dual histologic features of the trabecular and psammomatoid variants affecting the basal skull in a 15-year-old patient.[12] However, many studies reported a high recurrence rate after conservative with mini-invasive treatment (in 30–56% of cases) and thus, a complete surgical resection remains the preferred line of treatment.[15] Post-operative follow up of patients with JOF is indefinite, especially in the first two years as it is considered to be period of greatest recurrence rate.

Conclusion

JOF is an rare clinical entity. It is locally aggressive & high recurrence rate mean that it is important to make early diagnosis. A careful evaluation of the clinical, radiological, and histological parameter to rule out other FOL.

Disclaimer regarding Consent and Ethical Approval:

As per university standard guideline, participant consent and ethical approval have been collected and preserved by the authors

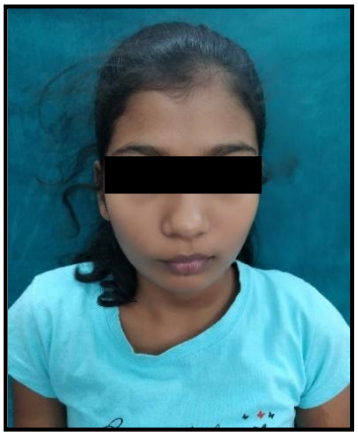


Figure 1:Extraoral view showing a diffuse swelling on right side of maxilla resulting in a slight facial asymmetry

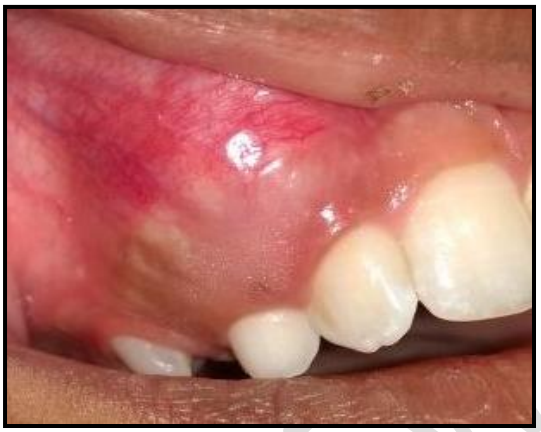


Figure 2: Intraoral view showing swelling at right buccal extending from 12 to 14 region, with displacement of 12

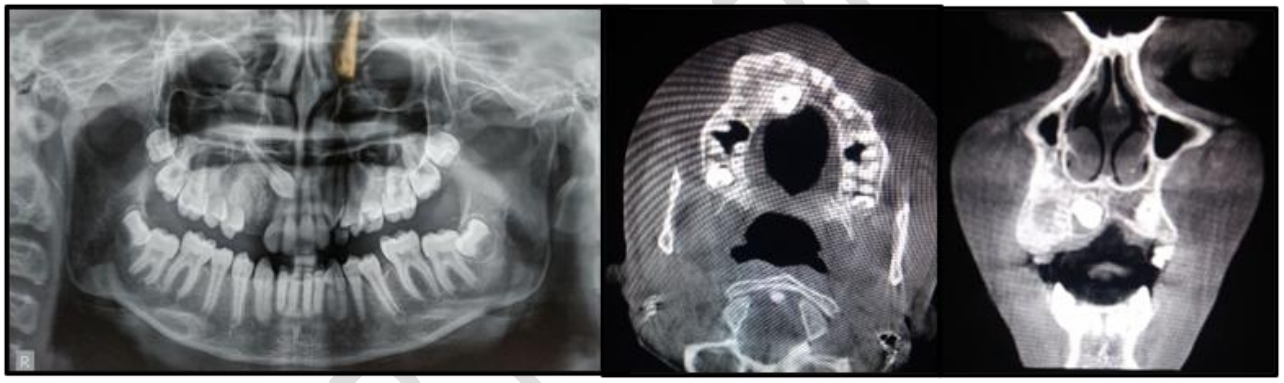


Figure 3 : OPG and CBCT showing a well defined multilocular radiolucency surrounding 13 , extending from the midline to 14 region causing cortical expansion as well as root resorption of 53

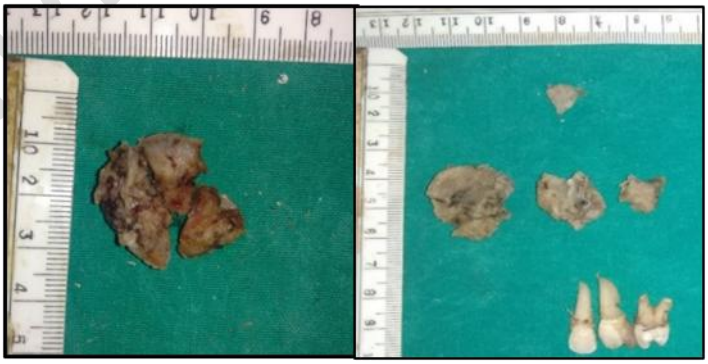
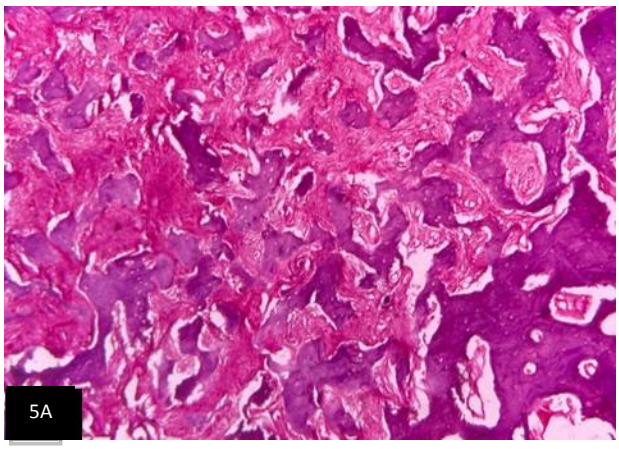


Figure 4 :On macroscopic examination, the biopsy specimen in multiple pieces 2cmx1cm in size, whitish in color, transparent and soft-to-gritty in consistency



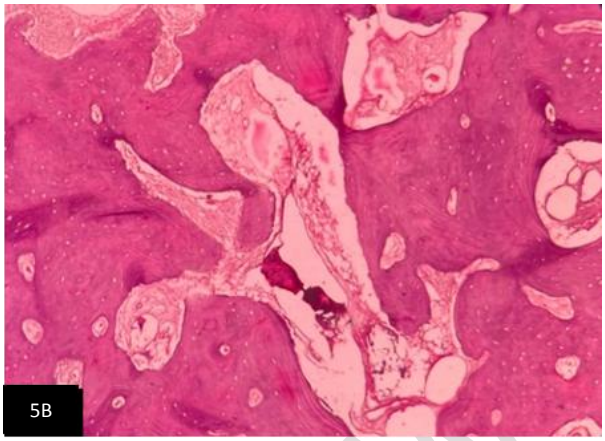


Figure 5A : osteoid lying in a cellular fibrous connective tissue stroma with fibroblast without much cytoplasm (H & E, X4), Figure 5B : calcified structure with osteoblastic rimming (H & E, X10)

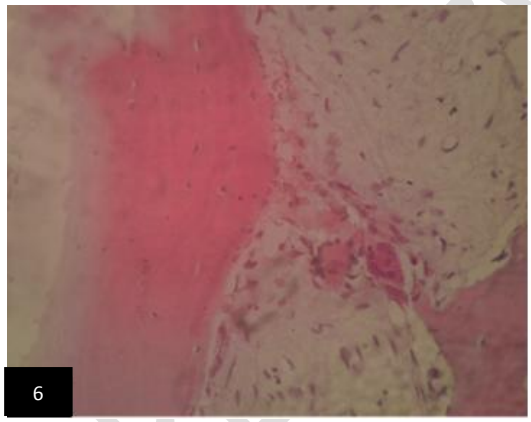


Figure 6 : On microscopic examination, osteoid lining by osteoblast and multinucleated giant cell present. (H & E X40)

Table 1 : comparison of Clinical Presentation of total 17 cases & Review of literature	
Clinical presentation	**Number of cases (Percentage)**
Buccal Expansion	9 (52%)
Mobility of tooth	3 (17.64%)
Tooth Displacement	2 (11.76%)
Previous reactive stimuli at the site of the lesion	1 (5.88%)
Radiology	10 MLRL (58.82%) 07 ULRL (41.2%)
ULRL : unilocular radiolucency, MLRL : multilocular radiolucency	

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