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CASE REPORT

SYNCHRONOUS JUVENILE OSSIFYING FIBROMA IN MAXILLA AND MANDIBLE-A CASE REPORT

B Shrestha [1]*, S Subedi [1], S Pandey [1]

[1] Chitwan Medical College and Hospital, College of Dental Sciences, Bharatpur, Chitwan, Nepal.

***Correspondence to :** Dr Bijayata Shrestha, Department of Oral Pathology, College of Dental Science,College of Medical College, Bharatpur, Nepal.
Email: bijayata_shr@yahoo.com*

ABSTRACT

Juvenile ossifying fibroma (JOF), one of the fibroosseous lesions is an uncommon clinical entity and involving both jaw is even rarer. Here, we present the case of a 15-year-old female patient with synchronous occurrence of JOF in the right sided maxilla and in the periapical area of mandibular right premolar molar region. After the clinical, radiographical and histopathological examination, the surgical treatment was carried out and the patient was kept under long term follow up. Owning to its aggressive local behaviour and high recurrence, early diagnosis, appropriate treatment and long term regular follow up are of prime importance.

Key Words: *Aggressive; Fibroosseous lesions; Juvenile ossifying fibroma; Recurrence; Synchronous.*

INTRODUCTION

Fibro-osseous lesion (FOL) is characterized by the replacement of bone by a benign connective tissue matrix which displays a varying degree of mineralization in form of woven bone or cementum like round acellular intensely basophilic structures1. Conventional ossifying fibroma (COF), juvenile psammomatoid ossifying fibroma (JPOF), juvenile trabecular ossifying fibroma (JTOF), fibrous dysplasia (FD) and cemento-osseous dysplasia (COD) all belong to the fibro-osseous group of lesions. Some are neoplastic, while others are developmental or reactive in nature2. Juvenile ossifying fibroma (JOF) is a variant of the aggressive subclass of ossifying fibroma. This is uncommon bone-forming neoplasm and is distinguished from other fibro-osseous lesions primarily by its age of onset, clinical presentation, aggressive behavior, and the high tendency to recur3. Onset occurs usually in children below 15 years old. Clinically, it presents as a large asymptomatic swelling of aggressive appearance due to the bone destruction it produces. The lesion is not encapsulated, although it is well demarcated from the surrounding bone. In the jaw, JOF is considered to develop from undifferentiated cells of the periodontal ligament, usually in the premolar and molar region4. Most JOF arise near the paranasal sinuses, but there are conflicting reports regarding the incidence of JOF in the maxilla, and mandible5. The histologic features of JOF include a highly cellular connective tissue stroma often with variations and irregular woven bone trabeculae surrounded by osteoblasts2.

Occurrence of multiple synchronous JOFs is rare in the jaws. The aims of the present report are to present an additional case of synchronous presentation of JOF in the maxilla and mandible and to emphasize the importance of early diagnosis, the appropriate treatment and especially, follow up over the long term owing to its aggressive behavior and high reccurence rate.

CASE REPORT

A 15 years old female presented with a rapidly growing painless swelling of right sided maxilla since 2 months. On clinical examination, she had a diffuse hard swelling of the right side of the maxilla with facial asymmetry. The overlying skin was normal in color but was stretched and the corresponding nasolabial fold was obliterated. Intraorally, bony hard expansion of the alveolar process of maxilla measuring approximately 4 x 3 cm was noted. The swelling extended from the distal aspect of right center incisor to the mesial aspect of right 1st molar region (Figure 1). There was no accompanying cervical lymphadenopathy. A complete blood count, urinalysis, routine blood chemistry, was all found to be within normal limits. Serum calcium, phosphate, and parathyroid hormone (PTH) levels were also found to be within normal limits.

The orthopantomogram (OPG) revealed a large well defined radiolucent lesion with sclerotic border extending from the mesial aspect of maxillary right central incisor to the mesial aspect of second molar. The roots of maxillary right premolars were found to be displaced with no evidence of any root resorption. OPG also revealed two small radiopaque mass surrounded by radiolucent halo located in between two premolars and between second premolar and first molar in right mandibular region (Figure 2).

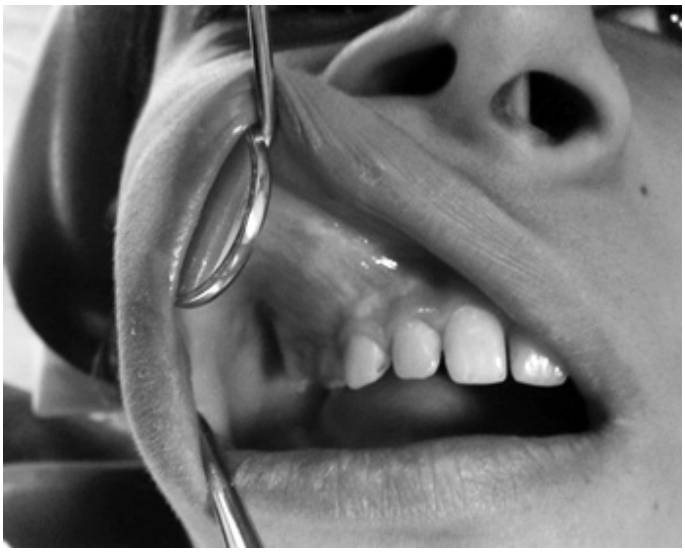

Fig 1: Intraoral photograph showing bony swelling over the alveolar process of the right sided maxilla

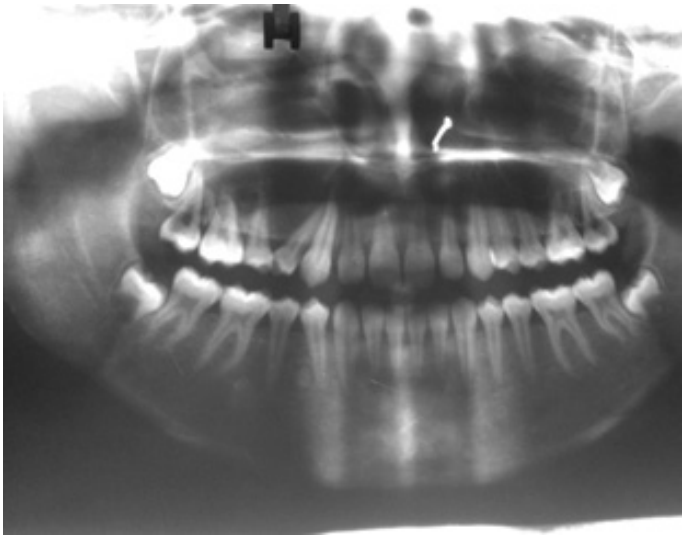

Fig 2: OPG showing a large well defined radiolucent lesion with sclerotic border in incisor to second molar region of maxillary right quadrant and two small radiopaque mass surrounded by radiolucent halo located in premolar molar region of mandibular right quadrant.

Computed tomogram (CT) scan showed an expansile lucent lesion with areas of calcification arising from alveolar process of right maxilla and measuring 3.8 x 3.2 cm. The lesion was bulging into the floor of right maxillary sinus and causing medial deviation of medial wall. Anteriorly the lesion was extending to the level of incisor and posteriorly to the level of premolar tooth (Figure 3).

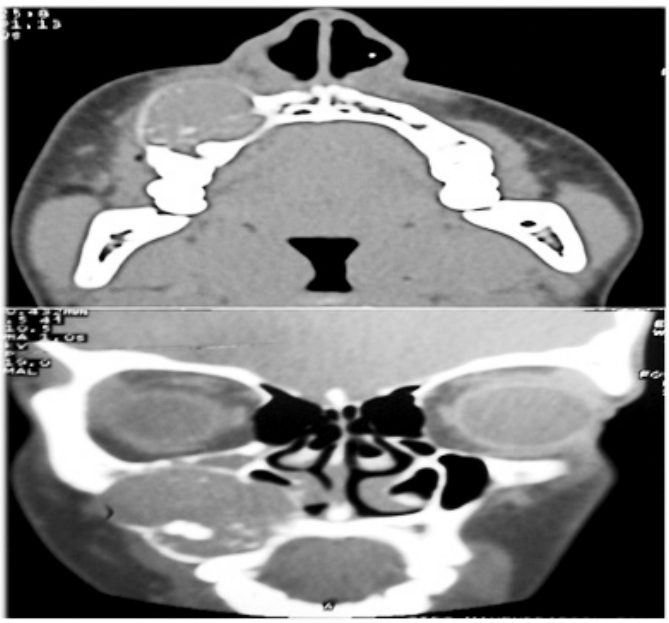

Fig 3. Axial and coronal CT scan showing expansile lucent lesion with areas of calcification arising from alveolar process of right maxilla

Incisional biopsies were taken from the lesions located in both jaws and submitted for histopatological examination. Histopathologically, the lesions showed numerous small bony ossicles within hypercellular stroma consisting of plump actively proliferating fibroblast like cells and loosely arrange fine collagen fibres (Figure 4). The appearances were consistent with the diagnosis of aggressive or juvenile ossifying fibroma. The histopathologic diagnosis of JOF was made for both the lesions.

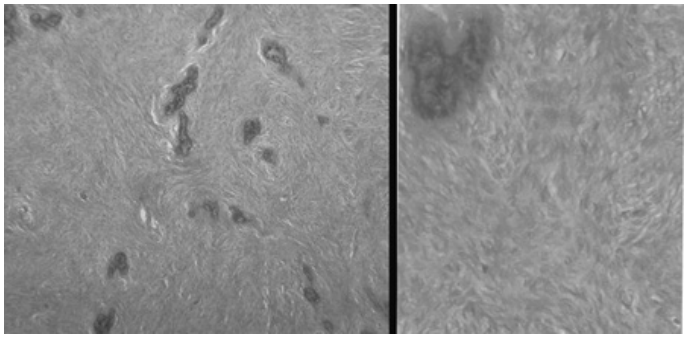

Figure 4. Photomicrograph (Magnification10x and 40x) showing numerous small bony ossicles within hypercellular stroma surrounded by plump actively proliferating fibroblast like cells and loosely arrange fine collagen fibres

The surgical treatment was carried out. Under general anesthesia, a buccal mucoperiosteal flap exposing the buccal cortical plate from the upper left lateral incisor to the distal aspect of right second molar was reflected. Another buccal mucoperiosteal flap exposing the buccal cortical plate from the lower right lateral incisor to mesial aspect of lower right second molar was also reflected. The lesions in maxilla and mandible were excised (Figure 5). Excised mass was again send for further histopathological evaluation (Figure 6) and the diagnosis was

reconfirmed. The patient was then kept under long term follow up.

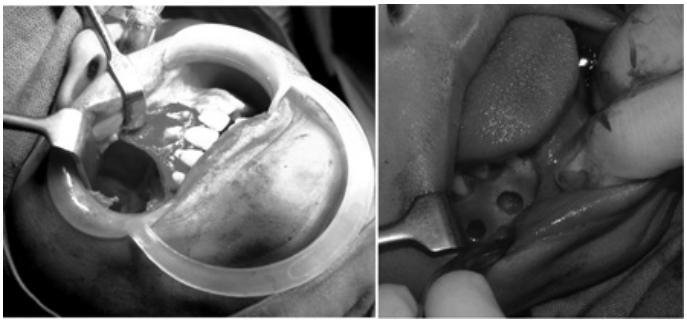

Fig 5: Photograph showing the maxilla and mandible of the patient after the surgical removal of the mass

Fig 6: Gross specimen showing a well circumscribed mass shelled out in one piece.

DISCUSSION

The benign fibro-osseous lesions (BFOL) represent a clinically diverse group of disorders of bone that share similar histopathologic features. Distinguishing specific benign fibro-osseous lesion from one another may still pose significant problems. Diagnosis requires careful clinical, radiologic, and histopathologic correlation6.

Ossifying fibroma of the jaw was first described by Montgomery in 1927, as a benign fibro-osseous lesion. Differentiating between fibrous dysplasia and ossifying fibroma may be difficult. Generally, ossifying fibroma is a well circumscribed lesion unlike fibrous dysplasia and it is usually surrounded by a fibrous capsule5. The term 'juvenile ossifying fibroma' was first used by Johnson in 1952, while describing aggressive forms of ossifying fibroma which occurred in the craniofacial bones of children5. JOF is a rare benign neoplasm with two histologic variant, trabecular and psammomatous type2. The main characteristics of JOF are its higher incidence in children and young adults, its aggressiveness and recurrence potential. JOF occurs mainly in the maxilla and the mandibular while its extracranial involvement is rare. Gender predilection has been a matter of controversy, with some authors claiming no predilection for either sex7. To the best of our knowledge, the occurrence of synchronous JOFs of the maxilla and mandible is rare, with only eight previously reported cases of ossifying fibroma8 and probably the first reported case of synchronous JOF. Khanna and Andrade (1992) et al9 reported a patient who had 2 cemento-ossifying fibromas involving the mandible and maxilla, and the maxillary lesion achieved enormous size, similar to the present case. The occurrence of multiple OFs in the jaws has been associated with hormonal abnormalities, such as hypercalcemia associated with hyperparathyroidism. Hyperparathyroidism– jaw tumor syndrome (HPT-JT), an inherited autosomal dominant disorder. This syndrome is characterized by the occurrence of parathyroid adenomas or carcinomas, fibro-osseous lesions of the jaws, some renal disorders, and pancreatic adenocarcinoma8. Normal levels of serum calcium, phosphorus, and PTH in the present patient ruled out HPT-JT.

Radiographically, the lesions can be radiolucent, mixed or radiopaque, depending on the degree of calcification. These features can resemble those of other lesions, such as fibrous dysplasia and cemento-ossifying fibroma. JOF may cause expansion and perforation of cortical bone10. Most of the cases of JOF, affecting the jaws, presented as radiolucent areas with irregular radiopaque areas as seen in maxillary lesion of our case. The mandibular lesion in our case showed radio-opacity with a radiolucent rim7. Oval well defined radiolucency without root resorption in our case is consistent with a benign neoplasm. BFOL are conditions characterized by the replacement of bone with varying amounts of fibrous and mineralized tissues. Microscopic examination of JOF shows hypercellularity, fibrillar osteoid incorporating plump osteoblast and progressive calcification of the osteoid or spherical cementum-like ossicles with psammomatoid calcification whereas OF reveals moderately cellular fibrous stroma with trabeculae of woven bone and occasionally cementum like ossicles11. Unlike the smoothly contoured OF particles, the ossicles in JOF have a thick, irregular collagenous rim. JOF is recognized by its trabeculae of woven bone with coarse lacunae, swollen osteocytes, and a lining of plump osteoblasts. Bands of cellular osteoid are also found. In addition, there may be some difficulty in differentiating JOF from cementoosseous dysplasia and fibrous dysplasia; the latter typically shows c-shaped or Chinese figure like osseous trabeculae and absence of osteoblastic rimming and of cementicles. Moreover, its margins tend to be ill defined radiographically, and blend with the adjacent bone. This finding is not seen in both patterns of JOF7. The histopathological features of this case were consistent with JOF.

The recommended treatment for JOF is conservative excision or curettage and some lesions may necessitate more aggressive management. Longstanding lesions may show significant cortical destruction and periosteal elevation as seen in our case, which can increase the risk of recurrence. The recurrence rate ranges from 30%-58% so continued followup is essential. Keeping in account of the high recurrence rate the case was kept under long term followup after conservative excision. Despite the aggressive nature of the lesion and high rate of recurrence, malignant transformation to sarcoma has not been reported12.

CONCLUSION

The present case represents an unusual presentation of JOF and highlights the clinical, radiographic, and histopathologic findings and emphasizes on early diagnosis, management and long term follow up.

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