Case Report

Juvenile Ossifying Fibroma of the Maxilla: A Case Report

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Abstract

Juvenile ossifying fibroma (JOF) is an uncommon, benign, bone-forming neoplasm with aggressive local growth that is distinguished from other fibro-osseous lesions primarily by its age of onset, clinical presentation and potential behaviour. We reported a 10 year old girl presenting with a growth in the left upper buccal gingiva which later was diagnosed as juvenile ossifying fibroma. The nature of the disease and outline of management were discussed.

Keywords: Ossifying fibroma, Juvenile, Maxilla.

Introduction

Juvenile ossifying fibromas are non-odontogenic lesions that clinically imitate odontogenic lesions by symptoms and signs that they produced. This lesion most often occurs in early decades of life, as the name implies. Despite being benign in nature, facial disfigurement is not uncommon due to the rapid growth of the involved anatomic site.

Case Summary

A 10 year old girl presented with one month history of a mass in the upper left buccal gingiva. The lesion was painful, pricking and persistent in nature. It was associated with scanty foul smelly pus discharge from the 26 and 27 tooth. She was treated as having bacterial gingivitis and symptoms resolved with oral amoxicillin and metronidazole. However, the mass kept growing over two month's time. She started to have facial disfigurement. The symptom was associated with painful chewing especially on the left side.

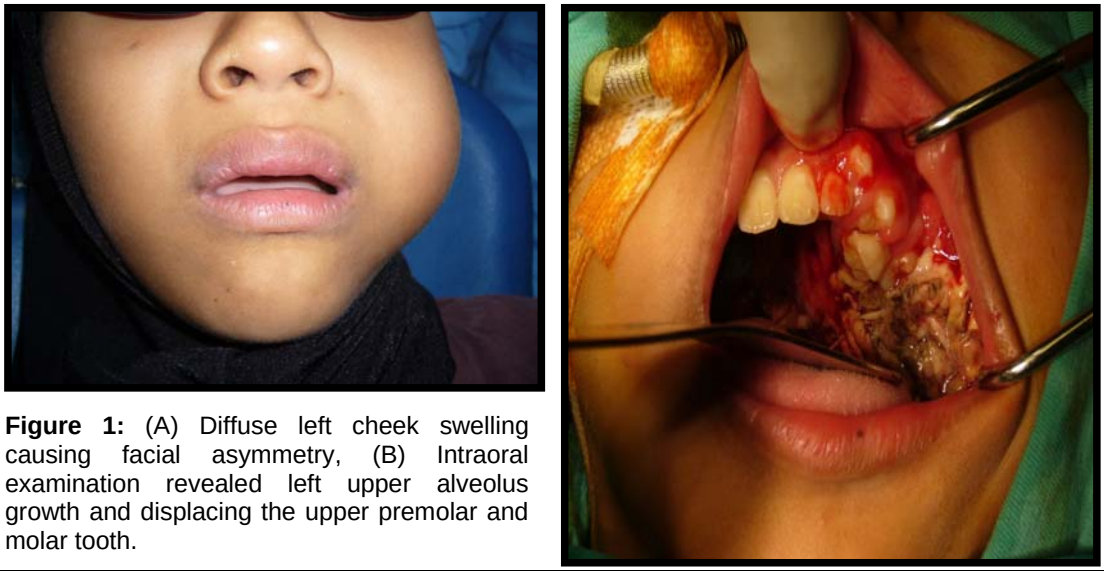

Figure 1: (A) Diffuse left cheek swelling causing facial asymmetry, (B) Intraoral examination revealed left upper alveolus growth and displacing the upper premolar and molar tooth.

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Clinical examination revealed a significant facial asymmetry caused by approximately 6 x 7 cm mass involving the left cheek extending to the left preauricular area. The mass was bony firm, mildly tender, freely mobile and the overlying skin was normal in colour (Figure 1A).

Intraoral examination showed an irregular growth over the left upper alveolus measuring 5 x 5 cm which was extended to the left maxillary tuberosity and displacing the 25 and 26 anterior and superiorly, and 27 and 28 tooth posteriorly. The lesion was mild tender, bony hard in consistency and wrapped by fibrotic tissue. However no pus or bloody discharge noted (Figure 1B). Other structures were normal.

Computed tomographic scan axial view of paranasal sinus showed presence of an expansile soft tissue mass in the left maxilla measuring 3 cm x 3 cm x 5 cm, causing displacement of the teeth superiorly and anteriorly (Figure 2).

Incisional biopsy pointed towards fibromatosis of gingiva. Excisional biopsy of the mass was performed under general anaesthesia via sublabial approach. The mass was well demarcated from its surrounding bone and easily separated from the maxillary sinus wall. Grossly, the specimen within the sinus measured approximately 5 x 8 cm and the intraoral

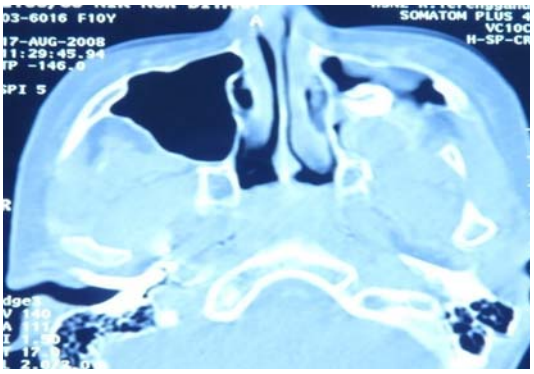

Figure 2: Axial CT scan showed a heterogenous mass in the left maxillary sinus.

specimen around 4 x 6 cm. It has an irregular surface with differing texture from soft, firm and cartilaginous in nature.

Microscopic examination showed that tumour is composed of spindle cells with adjacent areas showing extensive necrosis and ulceration of the squamous epithelium with focal area of bacterial colonies and some area of dense acute inflammatory exudates. There is some foci calcification but is minimal. Tumour cells shows interlacing cords of osteoid showing osteoblastic rimming. No evidence of malignancy noted.

After obtaining the histopathological examination of the excised mass, the diagnosis of Juvenile Ossifying Fibroma was established. The patient was discharged from the hospital on the third postoperative day with one week follow-

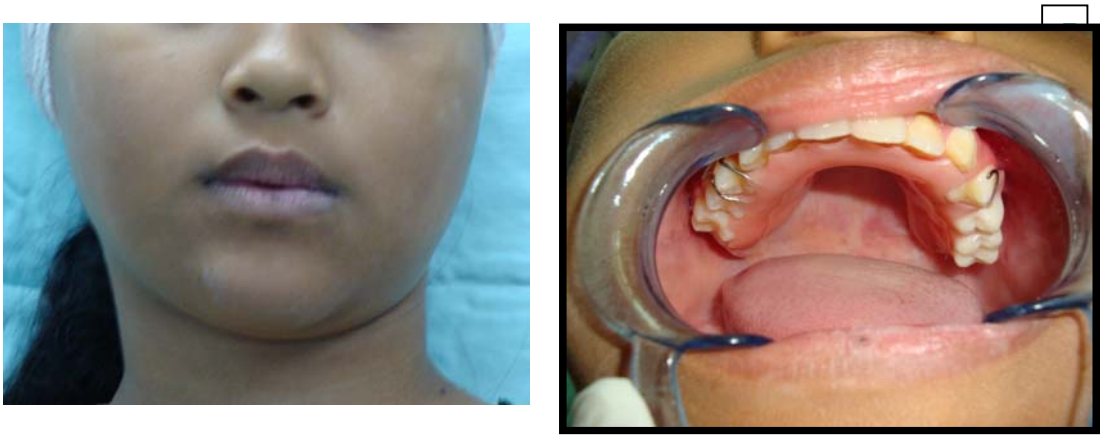

Figure 3A&B: Patient with good aesthetic result, wearing dental prosthesis that restores chewing and swallowing function.

up. White head varnish packing was then removed under general anaesthesia 14 days postoperatively.

The patient was subjected to clinical and radiological follow-up after excision of the lesion, to discard possible relapses or recurrences. On 3rd month's postoperative review, healing process was progressing well with no oroantral fistula noted. She was wearing a removable prosthesis, with satisfactory functional and esthetic results (Figure 3A &B).

Discussion

Juvenile Ossifying fibroma is an uncommon benign osteogenic neoplasm. The lesion in head and neck region is usually seen in the first and second decade of life but it does occur in adults. Wenig BM et al in 1995 in a published a series of patients aged as young as 5 years, also had patients in the fourth and sixth decades[1]. As a term "juvenile" underlines, the tumour largely develops in children, 79% of whom under age of 15 years old[2]. In reviews published by Slootweg et al and Hamner et al, the mean age of onset was 11.5 and 11.8 years, respectively[2,3]. This characteristic was also observed in our case.

Most of JOF occurred within the facial bones. Slootweg et al and Makek described the maxilla as the most frequent site[4,5], with Johnson et al reported that 90% of facial bone fibromas occur in the paranasal sinuses and only 10% in the mandible[6]. Some authors did found that the mandible is the commoner site[3]. Multiple sites of origin have also been reported and these tumours behave aggressively[7].

In general, ossifying fibroma is an asymptomatic lesion until growth causes infection, ulceration, swelling and moderate facial disfigurement. Displacement of the teeth can be an early clinical manifestation. The teeth associated with the lesion may preserve their vitality, but not uncommonly the roots were affected. Root divergence is recorded in 17% of cases while root resorption is seen in 11% to 44%[7].

In this case, the first clinical manifestation is a painful expansion of the maxillary cortex and mucosa, which produced a marked extra-oral facial asymmetry. The painful swelling is most likely due to the lesion being infected and the pain totally resolved with one week course of antibiotics. The teeth implicated in the lesion remained vital and no root resorption was observed, except presence of tooth displacement.

Common clinical features of the lesions arising from paranasal sinuses and orbit include nasal obstruction, exophthalmos, and rarely intracranial manifestation[8]. Mandibular JOF was associated with congenitally missing teeth results from a disturbance in the formation of the tooth socket, which may obliterate the involved developing tooth. Nofke CEE in 1998 noted an absence of the developing permanent first molar at a site of JOF in 4 year-old boy[9], and Johnson et al, 1991 included in their series of mandibular JOFs, a recurrent tumour in a 12 year old girl who also had a missing permanent first molar[6].

The treatment for JOF recommended by most investigators is conservative excision, curettage or more aggressive management in larger lesion. Fu YS, Perzin KH, in 1974 and Damjanov I et al, in 1978 suggested that the aggressive growth and the tendency to recur is age related and is seen more frequently in younger age group[10,11]. In our case, the exact time of onset of the tumour growth could not be defined but the size at presentation and the extremely young age revealed its aggressive behaviour. Han et al in 1991 elaborated a complete surgical removal did not lead to recurrence in the follow up[12]. If complete surgical removal was not practically

possible because of cosmetically unacceptable facial deformity post-operatively, maximum tissue needs to be removed by curettage. However, these cases have a tendency to recur.

Despite the aggressive nature of the lesion and high rate of recurrence, malignant transformation to sarcoma has not been reported[7]. JOF is not radiosensitive and radiotherapy is contraindicated because it can cause malignant transformation[8]. In this case, as the tumour was confined to the maxillary sinus and cortical layer of the upper alveolus, complete surgical removal was done with no evidence of recurrence during the follow-up.

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