Fibrous Dysplasia of Mandible

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ABSTRACT

An elderly male presented to the ENT, Outpatient Department at Abbasi Shaheed Hospital (ASH) with a mass on the left side of the face and lower jaw for 8 - 9 years. It was of a tennis ball size. No treatment was sought by him during those years. A diagnosis of benign fibro-osseous lesion of the mandible was made. Later the histopathology proved the same. The first occurrence in a male gender and involvement of the mandible is uncommon.

Key words: Fibrous dysplasia. Benign fibro-osseous lesions of mandible. Benign tumours of the jaw. Male gender. Tumour like condition.

INTRODUCTION

Fibrous dysplasia (FD) is a developmental tumour like condition that is characterized by replacement of normal bone by an excessive proliferation of cellular fibrous connective tissue intermixed with irregular bony trabeculae.¹

It is a genetic, non-inherited disease caused by mutation of the Gs-alpha subunit of protein coupled receptor resulting in up-regulation of cAMP that leads to defects in differentiation of osteoblasts with subsequent production of abnormal bone in an abundant fibrous stroma.² The symptoms of FD vary according to the site it involves. They may be facial pain, headache, craniofacial deformity, tooth loosening and/or displacement, and visual or auditory impairment.^{3,4}

This case report describes its occurrence in the mandible of a male patient, both being uncommon events.

CASE REPORT

An elderly male patient visited the Outpatient Department of ENT at Abbasi Shaheed Hospital with a mass at the left side of the face, especially the lower jaw that was causing deformity, difficulty in mastication and a sense of heaviness of the face. It had been present for the last 8 – 9 years. He sought no treatment for it until now. His general health status was good and he had no known comorbidity. The mass on examination was of a tennis ball size, with normal overlying skin and mucosa. It was hard, fixed and non-tender and seemingly was arising from the left side of the mandible. Fullness of left gingivo-labial and gingivobuccal sulcus were noted. Rest of the ENT examination was un- remarkable. A diagnosis of benign fibro-osseous lesion of the mandible

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was made. He was advised CT scan and orthopantomogram (OPG). The CT scan showed large, expansile, sclerotic mass, arising from the left hemi-mandible, having dense irregular calcifications (Figure 1). OPG showed a radiolucent mass arising from the left mandible.

Subsequently, he was admitted and operated upon for the excision of mass via an external approach. The specimen submitted for histopathology (Figure 2) reported a lesion composed of narrow, curved and

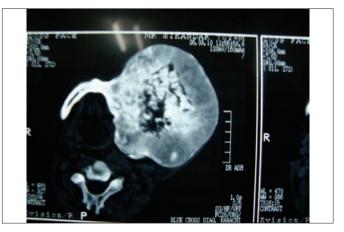


Figure 1: CT scan showing the lesion.



Figure 2: Excised mass.

misshapen bony trabeculae showing hooked configuration lined only focally by osteoblasts with intervening fibroelastic tissue features suggestive of fibrous dysplasia (FD).

DISCUSSION

FD can be monostotic or polyostotic. The latter may form a part of McCune Albright syndrome or Jaffe Lichtenstein syndrome. Gender prevalence of FD is equal and ratio of poly-ostotic to mono-ostotic FD is $3:7.^{3,5}$ The craniofacial bones are affected in about 10% of cases of mono-ostotic fibrous dysplasia (MFD) and 50 - 100% in poly-ostotic fibrous dysplasia (PFD).^{3,4,6} When only the cranial and facial bones are involved the term craniofacial fibrous dysplasia (CFD) is used. The prevalence of PFD ranges from 71 – 91% and of the MFD, from 10 – 29%.^{4,6} FD of the jaws affects the maxilla more frequently than the mandible and females are affected more than males.⁷ In this case both the involved site i.e. mandible and gender i.e. male were uncommon.

The radiological features of FD are diverse and dependent upon the proportion of mineralized bone to fibrous tissue in the lesion.⁸ The most frequent radiological description is of ground glass, though other patterns are also reported.

The existing guidelines for FD treatment aren't universally accepted. Spontaneous resolution of FD does not occur.⁸ FD lesions that are not symptomatic, do not progress and do not cause deformities or functional impairment should simply be monitored.⁴ Surgery is indicated for confirmatory biopsy, correction of a deformity, prevention of pathological fracture and/or eradication of symptomatic lesions.⁹ Radiation should not be employed as there is a risk of sarcomatous transformation of 0.4% that increases to over 40% following radiotherapy. $^{10}\,$

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