Monostotic Fibrous dysplasia of mandible in an adult – A case report

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ABSTRACT

Fibrous dysplasia (FD) is a developmental anomaly of bone characterized by hamartomatous proliferation of fibrous tissue within the medullary bone, with secondary bony metaplasia, producing immature, newly formed and weakly calcified bone, without maturation of the osteoblast.¹ May involve one or several bones and consists of one or more foci of fibro osseous tissue within the matrix of the affected bone. Patient presents with bone pain, deformities & pathologic fractures according to the site of involvement. FD usually manifests before the 3rd decade of life and is uncommon beyond that age. Fibrous dysplasia of mandible is rare and we report a case of monostotic fibrous dysplasia of mandibular bone in a 48 year old female.

KEYWORDS: Mandible, Fibrous dysplasia, Monostotic form

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. There are three forms of FD- Monostotic, polyostotic and craniofacial.[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 in cases with craniofacial involvement.[2,3] This case report describes its occurrence in the mandible of a 48 yrs old female patient, both site and age being uncommon events.

CASE REPORT

A 48 yrs old female presented to the dental OPD of ESIC Medical College & PGIMSR, with the complaints of swelling left lower jaw for the past 6 months, which progressively increased in size and attained the present size. She also complained of pain and discomfort during mastication. On examination, a well defined hard solitary swelling measuring about 3cms in the labial vestibule in relation to 32,33,31,41,42 teeth region. Obliteration of labial vestibule with lingual extension of the swelling also noted. (Figure:1)

CT facial bones was done which revealed the presence of an expansile lobulated lytic lesion at the symphysis menti, left side body of mandible, measuring 4X2.6 cms in size with thinning of lingual, buccal cortex with ground glass opacities and central calcification. Lesion indenting floor of mouth. Fat plane obliteration between lesion with orbicularis oris muscle and platysma also present. (Figure 2)
A provisional diagnosis of benign odontogenic tumour was made and the tumour was excised and sent for histopathological examination. Gross appearance of the excised specimen was multiple bony fragments measuring 5X3X2 cms aggregate. Microscopic examination revealed the replacement of normal bone by a lesion consisting of characteristic irregular bony trabeculae (Chinese letter pattern) in a cellular fibrous stroma. A diagnosis of fibrous dysplasia was made. (Figure: 3)

**DISCUSSION**

Fibrous dysplasia is a non neoplastic hamartomatous fibrous osseous lesion of bone, which can be monostotic or polyostotic. The latter may form a part of McCune Albright syndrome or Jaffe Lichtenstein syndrome. Fibrous dysplasia represents about 2, 5% of all bone tumors and over 7% of all benign tumours. Gender prevalence of FD is equal and ratio of poly-ostotic to mono-ostotic FD is 3:7.[3,4] The craniofacial bones are affected in about 10% of cases of poly-ostotic fibrous dysplasia (PFD). When only the cranial and facial bones are involved the term craniofacial fibrous dysplasia (CFD) is used. The average age of the patients with FD is 25, 8 years (from 5 to 67) without sex preference(46, 7% male) and usually manifests before the 3rd decade of life.[4,5] Maxilla is more commonly involved than mandible.[5] When maxilla is affected it may involve zygomatic & sphenoid bone.
Involvement of frontal, sphenoid, naso ethmoid & maxillary bone may lead to nasal obstruction, sinus obstruction & sinusitis.[6] Hypertelorism, cranial asymmetry, facial deformity, visual impairment, exophthalmos and blindness may occur due to involvement of orbital and parietal bone. Pain in the lower jaw, being the main complaint of the patients with mandibular involvement, is considered to be caused by compression of the mandible nerve of the enlarged fibrous tissue. The radiological features of FD are diverse and dependent upon the proportion of mineralized bone to fibrous tissue in the lesion. The most frequent radiological description is of ground glass, though other patterns are also reported. Spontaneous resolution of FD does not occur. [7] Differential diagnosis of fibrous dysplasia of mandible is ossifying fibroma.

FD lesions that are not symptomatic, do not progress and do not cause deformities or functional impairment should simply be monitored. Surgery is indicated for confirmatory biopsy, correction of a deformity, prevention of pathological fracture and/or eradication of symptomatic lesions. [8] Malignant changes with fibrous dysplasia include Osteosarcoma, Fibrosarcoma, Chondrosarcoma, Malignant fibrous histiocytoma & Ewings sarcoma. Association of Amelobastoma, Cystic degeneration, Angiosarcoma, Frontal sinus mucocele have also reported. Treatment is primarily surgical.

When the only tooth bearing area is involved conservative treatment is bone shaving. Use of Calcitonin & Pamidronate is also reported for its treatment. Biopsy can be taken to rule out the lesion. Fibrous dysplasia usually gets stabilized after puberty when it presents in prepubertal patients. Radiation should not be employed as there is a risk of sarcomatous transformation of 0.4% that increases to over 40% following radiotherapy. [9]

CONCLUSION

Isolated cases of fibrous displasia in maxillo- mandibular region are rare and can be difficult to differentiate from other benign and malignant bone disorders. Our case is a monostotic fibrous dysplasia involving the mandible in a 48 yr old female patient which was managed by surgery. Each patient may present with variable symptoms and clinical findings, thus the care of these patients must be customized to their needs and sites of involvement. For obtaining the definite diagnosis, treatment and further management of fibrous dysplasia it is mandatory to do imaging studies, histological examination. Aesthetic correction is done by cosmetic maxilla facial surgeries.

REFERENCES


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