

Fibrous Dysplasia of Maxillary Bone: A Case Report

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Abstract

Fibrous dysplasia is a developmental tumor like condition that is characterized by replacement of normal bone by excessive proliferation of cellular fibrous connective tissue intermixed with irregular bony trabeculae. It causes bone pain, deformities & pathologic fracture. It may involve one or several bones and consists of one or more foci of fibro osseous tissue within the matrix of the affected bone. Here a case of fibrous dysplasia of maxillary bone has reported.

Key words: Fibrous dysplasia, Maxilla, Monostotic form, Ground glass appearance.

INTRODUCTION

Fibrous dysplasia (FD) is an uncommon developmental bone disease of benign origin. It leads to expansion & replacement of medullary bone by disorganized fibroosseous tissue. It comprises 2.5% of all osseous & 7% of all benign bone tumors. In general, they affect 1 in 4,000 to 10,000 individuals. Male to female ratio is equal. Most frequently found in metaphysodiaphyseal region of long bones i.e. in rib (28%), followed by femur (23%), tibia, craniofacial bone (10-25%). Sarcomatous transformation is rare.

Fibrous dysplasia is a developmental tumor like sporadic condition that results from a post zygotic mutation in GNAS1 gene. GNAS1 gene codes for G protein which stimulates cAMP production in affected tissue which result in;

- (1) Endocrinal disturbances leading to precocious puberty, hyperthyroidism, growth hormone and cortisol overproduction
- (2) Increased proliferation of melanocyte leading to café-au-laitspots
- (3) Aberrant activity during osteoblasts differentiation, which results in normal medullary bone to be replaced by fibrous tissue and appears radiolucent on the radiograph.

CASE REPORT

A 36 years old female patient reported to our dept. with complaint of swelling in the left side of the face since 4 yrs. Initially swelling was small & gradually increased to present size. On extra oral examination diffused swelling of about 3×5 cm was present on left side of the face extending superio-inferiorly 0.5 cm below the cantho tragus line; to line joining corner of mouth to tragus & anteroposteriorly 1.2 cm away from the corner of the mouth to 1 cm in front of left tragus. On palpation the consistency was bony hard, non tender & no local rise of temperature. Intra orally swelling was present extending buccally from first premolar to

about 8mm beyond the tuberosity. Generalized gingival inflammation and bleeding on probing was present. Labiopalatally dimension at premolar region was less than 1 cm and increased gradually to tuberosity region about 2.0 cm. Buccal and palatal cortical plates were expanded. Overlying mucosa appeared normal, firm and was nontender.

Radiological Investigations:

Radiological investigation includes CTscan, Maxillary Occlusal view and Orthopantomogram (OPG). Maxillary Occlusal view and OPG shows gross radio-opacity in the maxillary bone from first premolar region to tuberosity region. Which gives ground glass appearance.



Figure-1:
Extraoral photograph showing solitary swelling present in showing the left middle third of the face.



Figure 2:
Intraoral photograph expansion in left maxilla



Figure 3:
OPG shows diffuse radiopacity in views shows diffuse left maxilla



Figure-4: Occlusal radiopacity with buccopalatal extension from 1st premolar to maxillary tuberosity.

DISCUSSION

Fibrous dysplasia is a bone disorder of unknown origin characterized by slow progressive replacement of medullary bone by abnormal proliferative isomorphic fibrous tissue which appears radiolucent on radiographs with classic ground glass appearance. Fibrous dysplasia has 4 different disease patterns Monostotic (70%), Polyostotic (30%), Craniofacial form and Cherubism (rare). The range of skeletal involvement varies from an asymptomatic monostotic lesion to polyostotic involvement resulting in progressive functional deficit & aesthetic problems. The clinical severity depends on time when the mutation of GNAS-1 occurs.

Polyostotic Form: It is seen if mutation occurs during 6th week of intrauterine life. Multiple bones may get involved. This form commonly involves the skull & facial bones, pelvis, spine & shoulder girdle. Initial symptoms are bone pain and spontaneous fracture of the involved bone. Femur shows shepherd's crook deformity. Polyostotic form is again sub-classified into Jaffe's type & Albright syndrome. Both type consists of variable bony involvement with café-au-lait spot. Albright syndrome has additional feature of endocrine disturbances of varying type. Polyostotic fibrous dysplasia with soft tissue called mazabrauds syndrome.

Monostotic Form: If mutation occurs during postnatal life the progeny of that mutated cells are essentially confined to one site resulting in fibrous dysplasia affecting a single bone. About 70% cases are of monostotic form and they involve mainly ribs, femur, tibia & craniofacial bones. Any bone may be affected, but monostotic forms are never reported as becoming polyostotic. The lesion is asymptomatic and usually discovered incidentally. It causes enlargement & distortion of bone.

Craniofacial Form: 50%-100% of patient with polyostotic disease & 10% patient with

monostotic disease have craniofacial involvement. Maxilla is more commonly involved than mandible. When maxilla is affected it may involve zygomatic & sphenoid bone. Involvement of frontal, sphenoid, nasoethmoid & maxillary bone may lead to nasal obstruction, sinus obstruction & sinusitis.

Hypertelorism, cranial asymmetry, facial deformity, visual impairment, exophthalmos and blindness may occur due to involvement of orbital and parietal bone. Malignant changes with fibrous dysplasia include Osteosarcoma, Fibrosarcoma, Chondrosarcoma, Malignant fibrous histiocytoma & Ewing's sarcoma. Association of Ameloblastoma, Cystic degeneration, Angiosarcoma, Frontal sinus mucocele have also reported. Treatment is primarily surgery. 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 get stabilized after puberty.

CONCLUSION

Fibrous dysplasia may manifest as monostotic or polyostotic form. Diagnosis of polyostotic form is easier due to extra-skeletal involvement. Monostotic form is common in the jaw. Fibrous dysplasia is a tumor like developmental disorder with minimal chances of malignancies. Aesthetic correction is done by surgeries.

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