Monostotic Fibrous Dysplasia of Maxilla

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ABSTRACT

Fibrous dysplasia (FD) is a common benign skeletal lesion that may involve one bone (monostotic) or multiple bones (polyostotic) and occurs throughout the skeleton with a predilection for the long bones, ribs and craniofacial bones. Monostotic FD accounts for 70 to 80% of FDs and 10 to 25% of the cases occur in craniofacial region, and these are common in 20 to 30 years age group. Here, we report a case of monostotic FD of maxilla which was treated surgically by surgical recontouring.

Keywords: Fibrous dysplasia, Monostotic, Maxilla, Surgical recontouring.

How to cite this article: Uppala D, Majumdar S, Rao AK, Biswas S. Monostotic Fibrous Dysplasia of Maxilla. Oral Maxillofac Pathol J 2015;6(2):628-631.

INTRODUCTION

Fibrous dysplasias (FDs) are nonhereditary benign fibro-osseous lesions in which fibrous connective tissue stroma and immature bone replace normal medullary bone as a result of abnormal differentiation of osteoblasts originally described by Lichtenstein in 1938 and by Lichtenstein and Jaffe in 1942. Fibrous dysplasia may involve a single bone which is called as monostotic FD or multiple bones which is called as polyostotic FD. The etiology of FD has been linked to an activating mutation in the gene that occurs postzygotically in somatic cells which encodes the α subunit of stimulatory G protein (Gs alpha) located at 20q13.2-13.3. The cells derived from the mutated cells manifest the dysplastic features.

CASE REPORT

A 22-year-old male patient was seen in the Department of Oral Pathology, GITAM Dental College and Hospital, Visakhapatnam, with the chief complaint of painless swelling of left side of his upper face causing disfigurement (Fig. 1). The swelling was hard and nontender to palpation. The onset of the swelling was gradual, initially noticed as a pea size and had since been increasing in size since 10 years. No relevant familial history and the swelling was extending superiorly from the level of ala tragal line to the level of corner of the mouth inferi-orly, anteriorly—from the level of ala of nose to 3 cm away from the tragus of ear posteriorly and the skin over the swelling is same as surrounding skin with no visible pulsations.

A well-defined swelling was seen over the left labial and buccal vestibule in 23, 24, 25, 26, 27. Anteroposteriorly extending from mesial aspect of 23 to distal aspect of 27, superioinferiorly from 2 mm above the gingival margin to zygomatic arch intraorally (Fig. 2).

X-ray posteroanterior (PA) view of skull revealed a well defined radiopacity seen extending from mesial aspect of 23 to distal aspect of 27 (Fig. 3). X-rays of long bones were taken and no abnormality was detected.

The coronal computed tomography (CT) image revealed expansile lesion with patchy areas of ground glass appearance and hypodense areas scattered throughout the left side of maxilla in the middle one-third of the face. The periphery is ill-defined and blended with the surrounding normal bone. Expansion is in the left maxillary sinus and its boundaries are displaced, occupying most of the sinus space. The teeth are not affected (Figs 4A to C).

Provisional diagnosis of central jaw lesion was established and an incisional biopsy was performed.

Fig. 1: Swelling of left side of his upper face causing disfigurement
Microscopic examination revealed cellular fibrous tissue with spindle-shaped cells and immature, isolated trabeculae of woven bone without rimming of osteoblasts. The bone trabeculae are not connected with curvilinear shapes, irregular with no definite pattern which likened to be C-shaped/Chinese script writing. Demonstrated monotonous pattern throughout the lesion (Figs 5A and B).

Final diagnosis of FD was confirmed after histopathological examination. Surgical recontouring of maxilla was done under general anesthesia (Figs 6A and B). Patient is under follow-up for every 3 months.

DISCUSSION

Monostotic FD accounts for 70 to 80% of all FDs, and 10 to 25% of the cases occur in craniofacial region which is common in 20 to 30 years age group. Lesion sizes grow up to the puberty and growth ceases thereafter. Polystotic FD accounts for 20 to 25% of FDs and 40 to 60% of the cases involves craniofacial region. More than one of the bones in the skeletal systems and craniofacial region are affected. The lesions grow during the childhood period and sometimes grow even after the puberty. Three percent of polystotic FD cases associated with various endocrine disorders, such as cutaneous hyperpigmentation (café-au-lait spots), precocious puberty, hyperthyroidism, cushing disease, hyperprolactinemia, and acromegaly called as McCune-Albright syndrome (MAS). Polystotic FD associated with soft tissue mass (intramuscular myxoma) occurring in 1% of population is called as Mazabraud's syndrome.1,13

In FD of the jaws, maxilla is more commonly involved than the mandible and males are commonly involved than females. Signs and symptoms include facial pain, headache, asymmetry of cranial bones, deformity of face, displacement of teeth, and visual or auditory impairment depending on the type of facial or cranial bone involved.

Café-au-lait skin hyperpigmentation is seen in MAS at birth. It has characteristic irregular borders analogous unlike the smooth borders of the hyperpigmentation in neurofibromatosis.2 In our case, there are no Café-au-lait skin hyperpigmentation, and only maxillary bone is involved.

Postzygotic point mutations in GNAS gene results in replacement of arginine at position 201 most commonly.
Figs 5A and B: Demonstrating cellular fibrous tissue with spindle-shaped cells and immature, isolated trabeculae of woven bone without rimming of osteoblasts. The bone trabeculae are not connected with curvilinear shapes, irregular with no definite pattern which likened to be C-shaped/Chinese script writing and monotonous pattern throughout the lesion (H&E staining: 10×)

Figs 6A and B: Demonstrating surgical recontouring of maxilla

with histidine or cysteine and rarely with serine, leucine or glycine because of which the disease is never inherited. As a result of mutation, GTPase activity of the mutated protein is decreased, along with co-activation of adenyl cyclase resulting in over production of 3', 5'-cyclic adenosine monophosphate (cAMP).

McCune-albright syndrome is due to the mutation in all three germ layers. Mutations if occur later in embryogenesis, the result will be isolated FD or endocrinopathies without associated bone disease. Mosaic distribution of abnormal cells is suggested by the characteristic lateralized pattern of skin and bone involvement in the polyostotic forms of fibrous dysplasia.

In normal bone remodeling, a coordinated cycle of sequential osteoclastic bone resorption followed by osteoblastic bone matrix deposition occurs. In FD bone, remodeling is altered by replacement of normal bone by abnormal osteogenic tissue and bone trabeculae.

Yamamoto et al found increased levels of interleukin-6 (IL-6) in two patient with MAS. This increased IL-6 secretion may be responsible for the increased numbers of osteoclasts and the bone resorption of FD. However, serum IL-6 levels were not increased in MAS, suggesting that the increased IL-6 synthesis is restricted to the local bone lesions where a GSα mutation exists.

The radiological features of FD are different and depends on the proportion of mineralized bone to fibrous tissue in the lesion. Initially, FD of craniofacial bones may be unilocular or multilocular, radiolucent with either ill defined or well defined borders. As the lesions mature, the bony defects will have mixed radiolucent/radiopaque appearance, and established FD exhibits ill-defined
borders blending into the normal adjacent bone with mottled radiopaque patterns often described as resembling ground glass, orange peel.9

Conventional X-rays may not so useful as they only reveal the tumor as a patch of haziness because of overlapping anatomical structures. Computed tomography scan is the best method of imaging in fibrous dysplasia.10 Radiographic classifications of fibrous dysplasia includes ground glass—pagetoid, (56%), homogeneous dense-sclerotic (23%) and radiolucent-cystic (21%).11

Leontiasis ossea is the name given to asymmetrical widening of facial bones leading to cosmetic impairment which can be seen with three-dimensional CT images. Magnetic resonance imaging (MRI) shows bone marrow changes and compression of surrounding soft tissues. Increased fibroblastic activity are seen as hypointense images on T1- and T2-weighted sequences, the regions with cystic/necrotic degeneration and increased chondroid matrix are seen as hyperintense images on the T2-weighted sequences.12

Fibrous dysplasia shows low to moderate cellular fibrous stroma surrounding irregular, curvilinear trabeculae of woven bone, commonly referred to Chinese letter characters.14 The dysplastic trabeculae are surrounded by a relatively hypocellular area composed of spindle-shaped cells. Osteoblastic rimming are characteristically absent.5 Chondroid metaplasia can be seen as a secondary change.14

Nonossifying fibroma, osteofibrous dysplasia, aneurysmal bone cyst, adamantinoma, giant cell tumor, and low-grade central osteosarcoma are to be considered in differential diagnosis. If there is a prominent chondroid component low-grade chondrosarcoma can be considered in differential diagnosis. Fibrous dysplasia shows low to moderate cellular fibrous stroma surrounding irregular, curvilinear trabeculae of woven bone, commonly referred to Chinese letter characters.14 The dysplastic trabeculae are surrounded by a relatively hypocellular area composed of spindle-shaped cells. Osteoblastic rimming are characteristically absent.5 Chondroid metaplasia can be seen as a secondary change.14

Nonossifying fibroma, osteofibrous dysplasia, aneurysmal bone cyst, adamantinoma, giant cell tumor, and low-grade central osteosarcoma are to be considered in differential diagnosis. If there is a prominent chondroid component low-grade chondrosarcoma can be considered in differential diagnosis. Osteoblastic rimming around the bone trabeculae differentiates FD from osteofibrous dysplasia and fracture callus and fracture callus should have a history of trauma.14

Parisi MS et al found that patients with FD receiving long-term treatment with IV pamidronate, there is improvement in bone pain and bone mineral density (BMD).

According to Lee et al in patients with nonaggressive but active FD, it is better to wait until the lesion stops growing and the operation can be performed when the patient has reached skeletal maturity. Surgical recontouring and debulking may be necessary to achieve acceptable facial proportions. In less than 1% of cases of FD, malignant transformation is seen.15

REFERENCES
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