CASE REPORT

Fibrous dysplasia of maxilla: An unusual case report
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Abstract
Bone is a dense connective tissue which is being affected by many diseases and that is independent of any age limits. Fibro-osseous lesions consist of the normal bone is being substituted by benign fibrous tissue comprising varying amount of mineralization which characteristically affect jaws and the craniofacial bone pathology.

Keywords: Craniofacial bones, fibrous dysplasia, fibro-osseous lesions

Introduction
Fibrous dysplasia is a skeletal nonneoplastic developmental lesion of the forming mesenchyme that manifests as an abnormality during differentiation and maturation of odontoblast.[1] It is usually asymptomatic and is associated with change in morphology, which shows the normal bone, is being substituted by fibrous tissue and non-functional trabecula-like osseous structures characteristically. The lesion is of following four types: Monostotic or polyostotic, with or without associated endocrine disturbances, craniofacial form, cherubism. According to reed, when there is a stoppage of the development of bone, with ossification of woven (fibrous) bone resulting from metaplasia of a nonspecific fibro-osseous type it results into fibrous dysplasia.[2] It has been described by various names in the literature as intraosseous calcifying fibroma, osteomatos cyst, osteitis fibrosa cystica, and ossifying fibroma.[3]

In 1891, Von Recklinghausen described fibrous dysplasia of bone. Lichtenstein and Jaffe coined the term fibrous dysplasia in 1938.[4]

The etiology is due to mutation in the Gs a gene in somatic cells 6, 7 which is located on chromosome 20q 13.2-13.3.

All the cells that derive from the mutated cells manifest the dysplastic features. The clinical presentation varies depending on where in the cell mass the mutation is located and the size of the cell mass during embryogenesis when the mutation occurs.

It is a benign bone disorder of an indefinite etiology, indefinite pathogenesis, and varied histopathology.[5]

In approximately 30% of cases, bones of cranium and face are affected with an average age of 25±8 years without any sex preference and manifestation usually occurs before the 30 years of life span.[6,7]

Fibrous dysplasia is of the following four major types: Monostotic, indicates involvement of a single bone; polyostotic, indicates involvement of multiple bones; and is of two types Jaffe and McCune-Albright syndrome. Jaffe type involves a variable number of bones along with pigmented lesions and McCune-Albright, which is a severe type of fibrous dysplasia as it involves nearly all bones along with endocrine abnormalities. The monostotic is the most common type of fibrous dysplasia, affecting younger age group of 20-30 years, comprising 70% of cases involving femur, tibia or ribs, and 25% occurring in the bones of the skull unilaterally. Alkaline phosphatase value remains normal in monostotic but an elevation of about 30% is observed in patients suffering with polyostotic fibrous dysplasia and the third form is craniofacial which most commonly occurs in 10-25% of patients of monostotic form and in 50% of the polyostotic form of fibrous dysplasia. Cherubism is an infrequent type of fibrous dysplasia.

Case Report
A 17-year-old female patient reported to the outpatient department of our department with a chief complaint of...
pain and swelling in left upper gum region since 2 months [Figure 1].

There was no family history with similar findings, but parents gave a history of consanguineous marriage. The past medical history includes right-hand deformity since birth and short-sightedness (2-3 months).

The general physical examination revealed a moderately built patient with normal vital signs.

Extraorally facial asymmetry present on left side with a diffuse swelling in middle the one-third part of the face extending superiorly 1 cm below the infraorbital margin to 1 cm above the inferior border of the mandible and anteroposteriorly 1 cm away from the ala of the nose to 2 cm away from the tragus of the ear without any discoloration of skin on comparing to other side of the face. On palpation tenderness was present with slight raise in temperature; submandibular lymph nodes were palpable which was solitary, mobile, roughly oval in shape, and hard in consistency.

Intraorally swelling was present extending buccally from distal aspect of canine to the third molar region. On palpation, swelling was non-tender, bony hard in consistency. Expansion of buccal cortical plates. Overlying mucosa appeared normal, firm and was non-tender [Figure 2].

Investigation

Blood and biochemical investigations showed hemoglobin 15%, red blood cell count 5 million/mm³, PCV 45%, TLC 10,000 cells/mm³, platelet count 2.1 lakhs cells/mm³, bleeding time 2 min, clotting time 5 min 30 s, alkaline phosphates 88 IU/l, serum calcium 9.7 mg%, and serum phosphorus 3.1 mg% which were with-in normal range.

Radiological investigations

Radiological investigation included maxillary occlusal view and orthopantomogram (OPG) [Figures 3 and 4]. Maxillary occlusal view show gross radio-opacity in the maxillary bone from distal aspect of canine to the third molar region which revealed presence of radiopaque lesions with abnormal trabeculae suggesting ground glass appearance with buccal extension of the bone and OPG showed presence of radio-opacity along with root resorption in the left maxillary premolar-molar region with break in continuity of maxillary sinus lining [Figures 3 and 4].

Surgical procedure

Incisional biopsy done left upper maxillary premolar - molar region and horizontal mattress sutures were placed [Figure 5].

Histopathological findings

The H and E stained decalciﬁed hard tissue sections showed cellular ﬁbroblastic stroma containing varying shapes of irregular trabecular bone, some of which were merging into stromal tissue. Some of the irregular trabecular bone was in curvilinear pattern [Figure 6].
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Discussion

Fibrous dysplasia is a nonneoplastic developmental lesion showing an abnormality in osteoblastic differentiation and maturation. It has unknown etiology but is usually caused by mutation in the guanine nucleotide-binding protein, α-stimulating activity polypeptide 1 gene (20q 13.2). This gene codes for G protein which stimulates cAMP production in affected tissue resulting in (1) endocrinial disturbances (2) increased production of melanocyte leading to café-au-lait spots (3) increased activity during osteoblastic differentiation as a result of which there is a replacement of normal medullary bone by fibrous tissue and appears to be radiolucent on the radiograph.[1] Fibrous dysplasia is broadly classified into four types monostotic (70%), polyostotic (30%), craniofacial form, and cherubism (rare).[1,8]

Monostotic

These are the most common form of occurring approximately 70-80% of all the types of fibrous dysplasia. The most commonly involved bones are ribs (28%), femur (23%), tibia, craniofacial bones, and humorous with equal predilections for males and females. This lesion is usually asymptomatic, causing enlargement, and distortion of bone and was discovered incidentally.

Polyostotic

As its name suggests, it involves multiple bones, the most common are skull and facial bones, pelvis, spine, and shoulder girdle. It occurs due to mutation during the 6th week of intrauterine life. Its initial symptoms are pain and spontaneous fracture of the involved bone. Femur shows shepherd’s crook deformity.[1] Polyostotic form is further classified as Jaffe’s type and Albright syndrome with café-au-lait spot as a common characteristic and endocrine disturbances as a supplementary feature of Albright syndrome.[1,9-11] Polyostotic fibrous dysplasia when associated with soft tissue called Mazabraud syndrome.[4]

Craniofacial form

This form is usually associated with 50% of patient with polyostotic disease and 10-25% patients with monostotic form of fibrous dysplasia.[9,12] The most common involved bone is maxilla than mandible, and the most common involvement in maxilla is zygomatic and sphenoid bone.[12]

Cherubism

Cherubism is an autosomal dominant disorder. The jaw is broad and protruding. Maxilla and mandible is symmetrically involved. An elevation in serum alkaline phosphatase level is the only distinctive feature in the laboratory diagnosis.

Conclusion

Benign fibro-osseous lesions of the maxillofacial bones represent a diverse group of pathologic conditions that includes developmental lesions, reactive or dysplastic diseases, and neoplasms. Fibrous dysplasia results in aesthetic as well as functional abnormality, especially visual impairment. Although
surgical management is required in fibrous dysplasia, the role of genetic manipulation in the management of the disease has to be explored in the near future.

References