Fibrous Dysplasia of Mandible : A Case Report.

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Abstract

Fibrous dysplasia is a developmental benign bone lesion characterised by the replacement of normal bone by excessive proliferation of cellular fibrous connective tissue intermixed with irregular bony trabeculae. Fibrous dysplasia of the jaw are believed to be benign, self-limiting, nonencapsulated mainly occurring in young subjects. Although, jaw lesions of fibrous dysplasias are usually monostotic, they may occasionally be part of a polyostotic process. Monostotic fibrous dysplasia of the maxilla is the most common site of involvement in the facial bones and accounts for approximately 70% of those with facial involvement. We report, a case with a slow growing swelling in relation to right and left side of face.

Keywords: Fibrous Dysplasia, Monostotic, Polyostotic, Craniofacial, Maxilla.

Introduction

Fibrous dysplasia(FD) is a nonspecific hamartomatous skeletal developmental anomaly, characterized by slow progressive replacement of a localized area of bone by an abnormal proliferation of isomorphic fibrous tissue.[1]

It is a fairly common, well-recognized and locally circumscribed benign disorder that was originally described by Lichtenstein more than 60 years ago. Although virtually all bones can be affected, the cranial and facial bones are most frequently involved.[2]

FD represents 5-7% of benign bone lesions. Being a sporadic benign skeletal disorder, it can affect one bone (monostotic form), or multiple bones (polyostotic form). Polyostotic form may form part of the McCune-Albright syndrome (MAS) or of the Jaffe-Lichtenstein syndrome (JLS).[3]

Gender prevalence of FD is equal. The monostotic form is more common and affects 20-30 years of age while the polyostotic form has its onset mainly in children younger than 10 years of age. Signs and symptoms of FD include bone pain, pathological fractures and bone deformities.[4]

Any cranial or facial bone can be affected by FD and associated clinical features will depend upon the bone or bones affected. Signs and symptoms can include facial pain, headache, cranial asymmetry, facial deformity, tooth displacement, and visual or auditory impairment.[5]

Here, we present a case of monostotic FD occurring bilaterally in the mandible of a 30 year old female.

Case Report

A 30 year old female patient reported our department of with a chief complaint of swelling in the lower right and left posterior teeth region since 4 years. Patient gave a history of swelling in the lower left and right posterior tooth region which she was aware of since 4 years. Swelling was initially smaller in size which progressed to the present size. Patient gave no history of pain, trauma,

bleeding or fever associated with the swelling Patient gave no history of similar type of swelling elsewhere in the body or difficulty in speaking or weight loss. Her medical ,dental and personal history were non significant.

On extraoral examination, facial asymmetry was visible on the right side of the face, and on inspection a diffused swelling on the right posterior region of the mandible approximately 4.5 × 3.5 cm in its greatest dimention was seen extending antero-posteriorly 1.5 cm from the commissure to 4.5 cm in front of the angle of the mandible and superoinferiorly 1.0 cm above the lower border of the mandible to 0.5 cm from the commissural line. The surface of the overlying skin was normal with that of the surrounding skin. And on palpation all the inspectory findings were confirmed. There was no local rise in temperature, consistency of swelling was firm, the swelling was non tender, non compressible, non reducible.

On intra-oral examination, a solitary ovoid shaped swelling was seen on the right mandibular posterior tooth region of size 3× 1.5 cm in its greatest dimension with well defined margins , extending anterioposteriorly from mesial aspect of 44 to the distal aspect of 47 and superioinferiorly from depth of vestibule to marginal gingival and lingually from the mesial aspect of 45 to the distal aspect of 47 with slight obliteration of the buccal vestibule. The lingual cortex showed slight expansion The surface of the swelling was smooth with no indentations, ulcerations., and the overlying mucosa was normal with the surrounding mucosa. On palpation all the inspectory findings were confirmed, the swelling was non tender, having a bony hard consistency, non mobile, non-compressible, non-fluctuant.

Extraoral examination of the left side of face revealed a diffused swelling on the left posterior region of the mandible approximately 2.5 × 3.5 in its greatest dimension was seen extending antero-posteriorly 2.0 cm from the commissure to 4 cm in front of the angle of the mandible and superoinferiorly 0.5 cm above the lower border of the mandible to 1cm from the commissural line. The surface of the overlying skin was normal with that of the surrounding skin. And on palpation all the inspectory findings were confirmed. There was no local rise in temperature, consistency of swelling was firm, the swelling was non tender, non compressible, non reducible.

Intra-oral examination of left side revealed , a solitary ovoid shaped swelling in relation to left mandibular posterior tooth region of size 2.5× 1 cm in its greatest dimension with well defined margins , extending anterioposteriorly from mesial aspect of 34 to the distal aspect of 37 and superioinferiorly from depth of vestibule to marginal gingiva and lingually from the mesial aspect of 35 to the distal aspect of 37 with no obliteration of the buccal vestibule. The surface of the swelling was smooth with no indentations, ulcerations., and the overlying mucosa was normal with the surrounding mucosa. On palpation all the inspectory findings were confirmed, the swelling was non tender, having a bony hard consistency, non mobile, non-compressible, non-fluctuant.

On the basis of extra and intra oral examination, a provisional diagnosis of Fibrous Dysplasia was given.

Cementoossifying Fibroma, Calcifying epithelial odontogenic tumor, Osteoma, Dentigerous Cyst were considered in differential diagnosis.

Investigations

Radiographic: Cross sectional Maxillary Occlusal Projection (fig 1), OPG (fig 2).

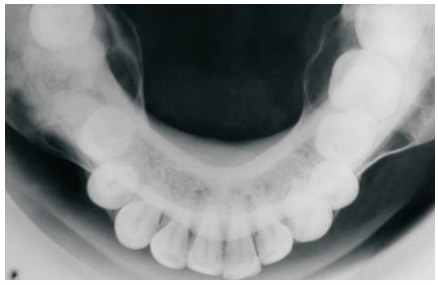

Figure 1: Occlusal radiograph showing expansion and thinning of the buccal and lingual cortex on both sides of the mandible with ground glass appearance.

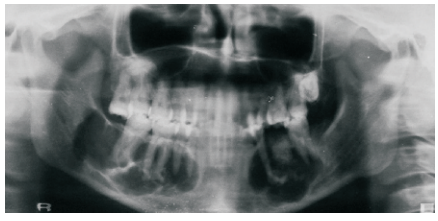

Figure 2: Orthopantomogram showing well-defined multilocular radiolucency with specks of radiopacity in the right and left posterior mandible

Radiographic diagnosis

Fibrous Dysplasia involving the left and right mandible.

Cementoossifying Fibroma, Hyperparathyroidism and Osteogenic Sarcoma were considered in radiographic differential diagnosis.

Histopathological Investigation- Excisional biopsy

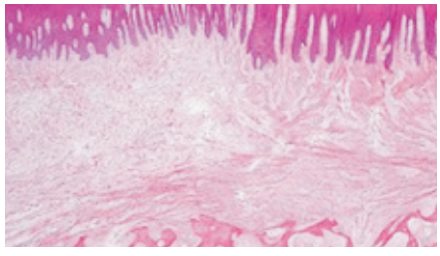

Figure 3: Delicate trabecullae of immature bone arranged in various forms lacking osteoblastic rimming.

Histopathological Findings

Delicate trabeculae of immature bone, with no osteoblastic rimming, enmeshed within a bland fibrous stroma of dysplastic spindle shaped cells without any cellular features of malignancy. Examination of macrosections of intact lesions reveals the margins of the lesion to be separated from surrounding bone by a thin shell of mature lamellar reactive bone. (fig 3)

Final Diagnosis

On the basis of clinical, radiological and histopathological findings, a final diagnosis of fibrous dysplasia was given.

Treatment- The treatment involved conservative surgical removal of the lesion.

Discussion

Fibrous dysplasia (FD) is a benign, non-neoplastic and non-encapsulated fibro-osseous condition involving cranial and extra-cranial bones which show replacement of normal bone by cellular fibrous tissue containing islands of metaplastic bone.[6]

Lichtenstein in 1938 introduced the term Fibrous dysplasia. It is classified by W.H.O. as developmental in origin. Some claim that FD is a dysplastic or hyperplastic process with diffuse margins that blend into adjacent bone. According to some authors, FD is usually caused by mutation in the GNAS1 gene (20q13.2).[7]

FD most frequently occurs in the metaphysial and diaphysial areas of the long bones, the shoulder bones, the bones of the pelvic girdle and those of the jaws and skull. The posterior maxilla is the most common site especially in the area around the first molar. The anterior maxilla is rarely involved. Mandibular lesions are ordinarily found between the mental foramen and angle of the mandible.[8]

In FD, the regular spongiosa is replaced by a fibrous and osseous tissue which is ill suited for expanding and contracting pressure. The cortex becomes narrower due to endosteal bone resorption which can lead to pathological fracture.[9]

Fibrous Dysplasia has been traditionally classified as:[7]

1) Monostotic: Involving only one bone,
2) Polyostotic: Involving more than one bone.

Polyostotic FD is sub-classified as

a) Jaffe's type - Many bones are affected and light brown spots over skin i.e. cafe-au-lait spots.
b) Albright syndrome - Several bones are involved with cafe-au-lait spots or lesions in the affected area and along this endocrine dysfunction, like precocious puberty in females.
c) Craniofacial form - Involving the craniofacial complex only.
d) Cherubism

Monostotic Fibrous Dysplasia

This term is applied to those forms of the disease in which only one bone is affected. It occurs in about 70 - 80 % of the cases of FD. It does not manifest extra-skeletal lesions, but may become polyostotic and affect multiple bones. It is less serious than polyostotic form.

The clinical term "leontiasis ossea" has been applied to cases of Monostotic FD which affects maxilla or facial bones and give the patient a leonine appearance.[10]

Clinical features

It occurs with equal predilection in males and females with a mild predominance for females. It is more common in children and young adults. Mean age of occurrence being 27-34 years.[8] The first clinical sign of the disease is a painless swelling or bulging of the jaw. The swelling usually involves buccal and labial plate and seldom the lingual plate.[9] When it involves the mandible, it sometimes causes a protuberant excrescence of the inferior border. The overlying mucosa is almost invariably intact over the lesion. Tenderness ultimately develops. There may be some malalignment, tipping or displacement of the teeth due to the progressive expansile nature of the lesion.[7]

Radiographic features

The roentgenographic appearance of F.D of the jaw is extremely variable. These are 3 basic patterns which may be seen.[7]

In one type, the lesion is generally a rather small unilocular radiolucency or a somewhat larger multilocular radiolucency, due to marked fibrous proliferation and deposition of osteoid which will subsequently calcify. In second type, the pattern is similar except that increased trabeculation renders the lesion more opaque and typically mottled in appearance.[6] Third type is quite opaque, with many delicate trabeculae giving a ground glass or peau d' orange appearance to the lesion. This type is not well circumscribed but blends into the adjacent normal bone.[9]

In all the three types, cortical bone becomes thinned because of the expansile nature of the growth but this bony plate is seldom perforated.[10]

Histopathological features

The lesion is essentially a fibrous one made up of proliferating fibroblasts in a compact stroma of interlacing collagen fibers.[7]

The trabeculae are thin and located at regular intervals. Irregular trabeculae of bone are scattered through out the lesion, with no definite pattern of arrangement.

Characteristically some of these trabeculae are delicate C-shaped or as Chinese character-shaped hook or horse shoe shaped and occasionally may form rings. The trabeculae are usually coarse woven bone but may be lamellar bone. The relationship of osteoblasts and osteoclasts to the trabeculae is similar to that of polyostotic form.[7]

The presence of lamellar bone does not confirm a diagnosis of FD. Other morphological forms of calcification, including small rounded bodies (cementum bodies or globular calcifications) and minute basophilic and laminated calcification may be seen in FD. The histopathology of FD is not pathognomonic and it should be confirmed by correlation of histopathology, radiography and clinical features. Giant cells may also be seen in some cases.[10]

Treatment

The treatment consists of conservative surgical removal of the lesion. It is estimated that between 25% and 50% of patients will show some regrowth after a conservative surgical procedure. Radiation therapy is contraindicated, as it carries the risk of post irradiation bone sarcoma.[10]

Polyostotic Fibrous Dysplasia (Mccune Albright Syndrome)

Weil in 1922, recognized the case of polyostotic FD associated with skin lesions and endocrine disturbance. The condition has been specifically described by Albright, from where the apparent syndrome derives its eponym. 'Polyostotic' have been applied to those lesions in which more than one bone is affected.[7]

Clinical features

It occurs in about 20-30% cases of FD. The disease usually manifests early in life with an evident deformity of long bones, often unilateral in distribution. It has insidious onset. Recurrent bone pain is the most common presenting skeletal symptoms.[11] Because of the severe bone changes, spontaneous fractures are a common complication of the disease. The structural integrity of the bone is weakened and the weight bearing areas become bowed. The curvature of the femoral neck and proximal shaft of the femur markedly increase causing a 'shepherd crook deformity', which is a characteristic sign of the disease. Overgrowth of adjacent soft tissues may be present. Two apparently separate types of polyostotic FD are described as–[7]

Jaffe's type - FD involving a variable number of bones, accompanied by pigmented lesions of the skin or "cafe-au-lait" spots of thin light brown color. It is mild and non-progressive form. This type occurs in about 50% of the cases.[7]

Albright's syndrome - FD even more severe, involving nearly all bones in the skeleton accompanied by pigmented lesions of the skin with endocrine disturbances of varying types. Female patients exhibit precocious puberty, sometimes beginning at the age of 2 or 3 years. Vaginal bleeding is a common manifestation.[7]

Oral manifestations

The oral manifestations of PFD are related to severe disturbance of bony tissue. One third of the polyostotic patients have lesions in the mandible. There may be expansion and deformity of the jaws and the eruption pattern of the teeth is disturbed because of loss of support of the developing teeth. The endocrine disturbance also, may alter the time of eruption of the teeth.[7]

Histological features

Histological features are similar to that of Monostotic FD. The lesion is composed of fibrillar connective tissue within which numerous trabeculae, woven immature bone, irregular in shape, can be seen.[10]

The osteocytes are quite large and collagen fibers of these trabeculae can often be observed extending into fibrous tissue. Bone formation by stellate osteoblasts can be seen but rows of cuboidal osteoblasts remaining on the surface of the trabeculae (osteoblastic rimming) are absent. These trabeculae typically have wide osteoid seams. Osteoclastic activity may also be seen in some areas.[7]

Laboratory findings

No consistent and significant changes in serum calcium or phosphorous can be seen, although, serum alkaline phosphatase level is sometimes elevated. Premature secretion of pituitary follicle stimulating hormone is found. There may be an elevated basal metabolic rate.[8]

Treatment

Mild cases may be treated surgically. Severe forms are treated with X-ray radiation with some success but it is hazardous because of the possibility of development of induced osteosarcomas.[7]

Conclusion

The fibrous dysplasia is significant for the dentists because it may affect the facial, cranial and jaw bones leading to many deformities and dysfunctions. The cells of fibrous dysplasia are committed osteogenic cells with impaired capacity to form normal bone. The mutated protein not only affects osteoblasts but can also affect various hormone receptors leading to endocrinopathies and cafe'-au-lait spots.

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Conflict of Interest: None

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