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Fibrous Dysplasia: A Case Report and Review of Literature.

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ABSTRACT

Fibrous dysplasia is a pathologic condition of bone with no apparent familial, hereditary or congenital basis. Fibrous dysplasia constitutes 2.5% of all bony neoplasms and 7% of all benign bony tumours. The treatment can be either conservative or complete resection. Here we report a case of fibrous dysplasia in a 22-year-old male patient on the right side of mandible.

Keywords: Fibrous dysplasia, neoplasm, resection.

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INTRODUCTION

Lichtenstein first coined the term in 1938 [1] and in 1942 he and Jaffe separated it from other fibro-osseous lesions [2]. In fact, there were thirty-three synonyms listed in their paper in 1942, and the term fibrous dysplaisa was not 1isted in the index medicus until 1967. Fibrous dysplasia is divided into *monostotic and polyostotic* variety [3]. It may involve multiple bones associated with cutaneous pigmentation and precocious puberty known as Albright's syndrome [4]. Fibrous dysplasia of craniofacial region commonly occurs in maxilla

In this paper, we present a case of fibrous dysplasia in a 22-year-old male patient on the right side of mandible.

Case Report

A 22-year-old male patient reported to our dental OP with a chief complaint of painless swelling in right side of lower third of face since 10 years. Patient gave a history of road traffic accident 10 years back.

Patient was apparently normal 10 years back. History of swelling which was gradual onset, initially small in size and later progressed to present size. No history of associated pain and fever.

On extraoral examination, a single diffuse swelling extending from 1cm away from right parasymphysis, crossing the midline to left parasymphysis region. On palpation, inspectory findings with respect to site, size, shape and extent are confirmed. Hard in consistency and was not tender and not warm. No pulsations palpable.

On intraoral examination, a single diffuse swelling was evident which extended from 32 to 46 region anteroposteriorly and from gingival margin to inferior border of mandible superoinferiorly. The swelling was hard in consistency and non-tender. No secondary changes were evident and not pulsatile.

Based on the history, clinical examination and investigations the case was provisionally diagnosed as fibrous dysplasia. Differential diagnosis for the bony hard swelling considered were ossifying fibroma, pagets disease, focal cementosseous dysplasia and osteoma.

Orthopantamograph (OPG) revealed a mixed radiopaque - radiolucent area in right mandibular quadrant in relation to 42 to 45. It measured approximately 6 X 3cm in size and is roughly oval in shape. The borders were ill-defined borders.

Computed tomography (CT) scan revealed a well-defined expansile lesion with lytic areas and ground glass density noted involving the body of the right hemi mandible extending across the midline to the parasymphyseal region of the left hemi mandible. The lesion measured approximately 5 x 3.6 x 2.7 cm. The lesion is seen involving the roots of the overlying mandibular teeth. CT value of the soft tissue density within the lytic areas measured of about 15-20 HU. No cortical breach. These features were suggestive of fibrous dysplasia.

An incisional biopsy was performed which revealed numerous interconnecting trabeculae of coarse woven immature bone. The immature bone is irregular in shape. The connective tissue is moderately collagenized with high cellularity. Numerous resting and reversal lines are observed in the trabeculae of bone. The irregular trabeculae of bone show osteocytes residing in their lacunae. Moderate vascularity is seen. The features were suggestive of fibrous dysplasia.

Based on the history, clinical examination and investigations the case was finally diagnosed as fibrous dysplasia on mandible.

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Figure 1: Facial asymmetry due to a diffuse swelling on right lower third of face

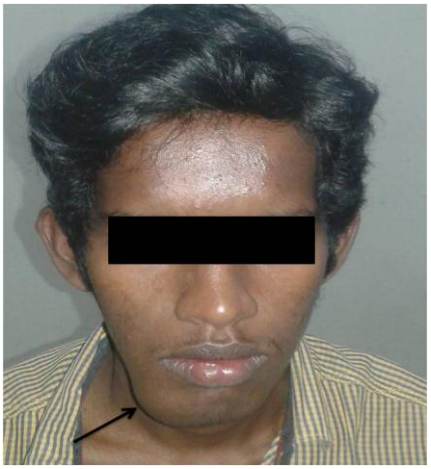

Figure 2: Buccal cortical plate expansion in relation to 42, 43, 44, 45 and 46

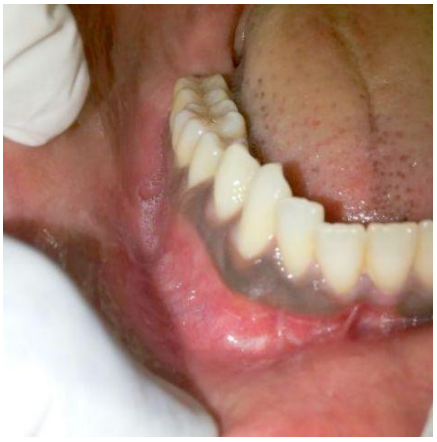

Figure 3: OPG reveals a diffuse mixed lesion in relation to 42, 43

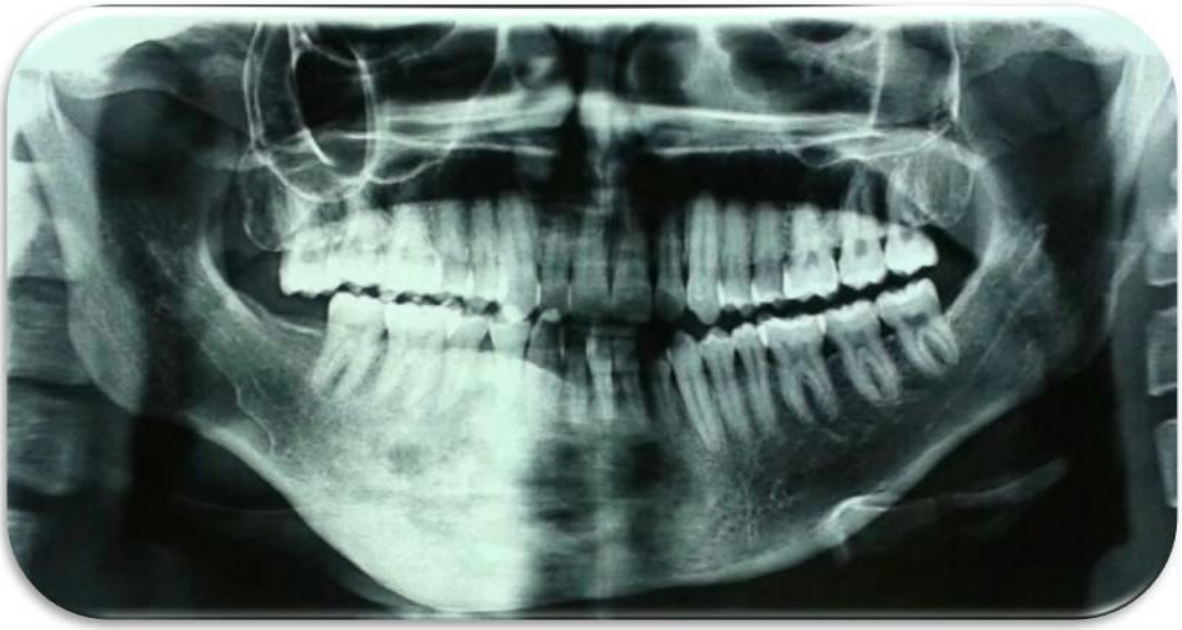

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Figure 4: CT scan revealed well-defined lesion with ground glass density

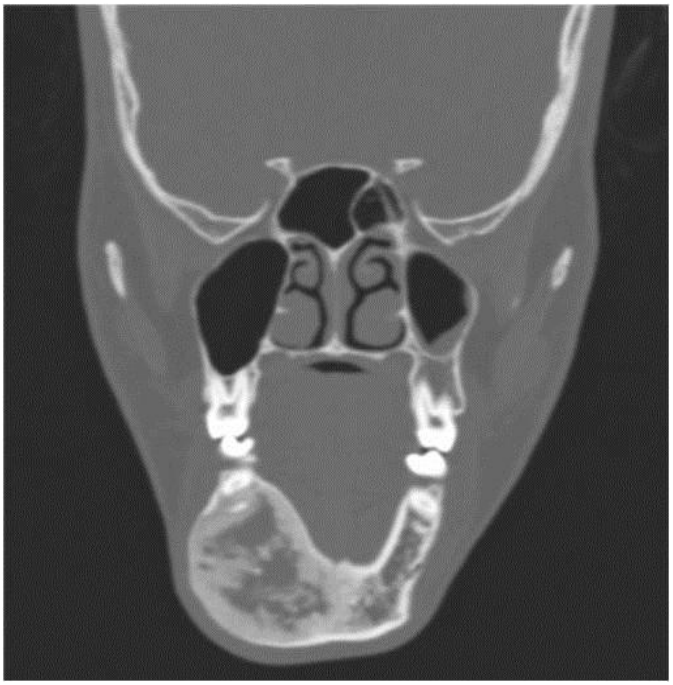
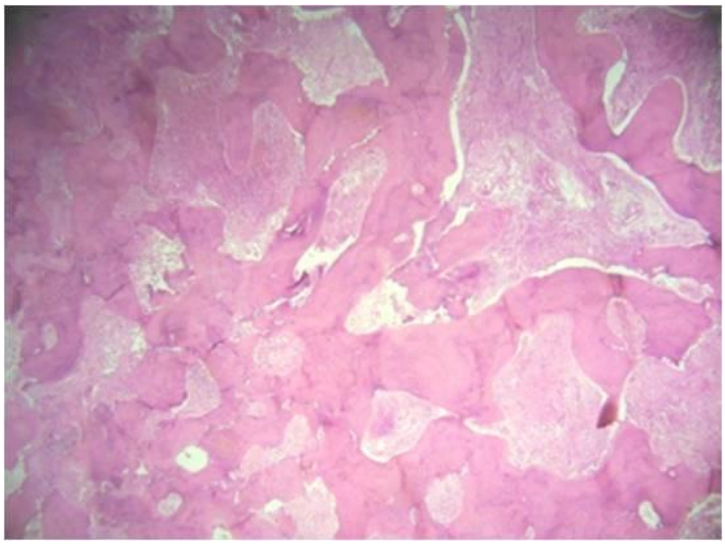


Figure 5: Histopathology revealed numerous interconnecting trabeculae of coarse woven immature bone



DISCUSSION

Fibrous dysplasia is a rare benign bone disorder characterized by the replacement of normal bone and marrow with fibrous tissue intermixed with irregular woven bone.

It begins around 10 years of age and then progresses throughout adolescence with no gender predilection. Common sites of involvement are femur, tibia, fibula, ribs and facial bones. The maxilla is more frequently involved than mandible in the craniofacial region [5]. The mandible, ethmoid and sphenoidal regions follow in the order.

Many theories have been suggested regarding the cause of fibrous dysplasia, including the trauma with a nonspecific distrubance in local bone reaction,[6] a congenital anomaly "Perverted" activity of mesenchymal bone-forming cells [7], and a complex endocrine disturbance with local bone susceptibility. However, the most readily acceptable theory at this time is the abnormal activity of mesenchymal cells. The etiology has been linked with a mutation in the Gsα gene that occurs after fertilization in somatic cells and is

located at chromosome 20q13.2-13.3. The cells that originate from the mutated cells manifest the dysplastic features.

Fibrous dysplasia is postulated to occur as a result of a developmental failure in the remodeling of primitive bone to mature lamellar bone and a failure of the bone to realign in response to mechanical stress. When maturation fails, it leaves a mass of immature isolated trabeculae enmeshed in dysplastic fibrous tissue that are turning over constantly but never completing the remodeling process and [8]. The combination of an insufficient mineralization and lack of stress alignment results in substantial loss of mechanical strength causing pain, deformity and pathologic fractures.

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 [9,10]. Severe disease may be associated with an earlier mutational event that leads to a larger number or a more widespread distribution of mutated cells. The occurrence of these diseases and the characteristic lateralized pattern of skin and bone involvement in the polyostotic forms of fibrous dysplasia suggest this mosaic distribution of abnormal cells. The mutation of Gsα gene was first identified in patients with McCune-Albright syndrome, a disorder that is characterized by polyostotic fibrous dysplasia, skin pigmentation and endocrinopathies. The Gsα gene has also been linked to other endocrine tumors and human diseases.

The radiographic features of fibrous dysplasia vary widely. The normal bone is replaced by tissue that is more radiolucent, with a grayish “ground-glass” pattern that is similar to the density of cancellous bone but is homogeneous, with no visible trabecular pattern. The radiolucent region is composed histologically of a solid fibro-osseous tissue, which may contain a cystic component with a fluid-filled cavity. The characteristic feature of the lesion is that it is bounded by a distinct rim or shell of reactive bone that is defined sharply on its inner border and may fade gradually into normal cancellous bone. The lesions originating within the medullary canal but consistently replace both cortical and cancellous bone which obscures the usual sharp distinction between the cortex and the medullary canal.

Often, the diameter of the bone is increased by growth of the lesion and is bounded by a shell of reactive bone. Cortical thickness variations are caused by slow resorption of the endosteal surface commonly referred to as “endosteal scalloping.” The surface of periosteum is smooth and without any periosteal reaction [11].

The main treatment for CFD is surgery, which can be divided into conservative and radical resection. The mere presence of fibrous dysplasia of the craniofacial region is not in itself an indication for treatment. Small solitary lesions will remain static and asymptomatic for long duration. A progressive or marked deformity, pain or functional disability suggests the need for intervention. Complete surgical resection of the involved area is the treatment of choice, but this result in considerable functional and cosmetic defect demanding extensive reconstruction.

Conservative surgery is the basic surgical promise. [3] Also, the disease nearly always burns itself out around the puberty. Radiotherapy is to be avoided because it can induce malignant changes in the fibrous dysplasia. Bisphosphonate therapy may help to decrease pain, improve function and lower fracture risk in appropriately selected patients of fibrous dysplasia. Surgery is indicated for prevention of pathologic fracture, correction of deformity and/or eradication of symptomatic lesions.

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