Fibrous Dysplasia of Maxilla - A Case Report and Review of Literature

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Abstract

Fibrous dysplasia (FD) is a non-neoplastic, non-inflamatory condition of the bone that occurs because osteoblastic differentiation and maturation within the bone is defective. In this condition, normal bone is replaced by fibrous tissue. It can affect one or many bones in the body. It has a predilection for long bones, ribs, and craniofacial bones. This article presents a case of fibrous dysplasia of the maxilla in a 21 year old.

Key Words: Fibro osseous, Fibrous Dysplasia, Osteoblasts, Chinese letter.

Introduction

Waldron, in the year 1985 described Fibrous Dysplasia (FD) as "a benign lesion, characterised by the presence of fibrous connective tissue with a whorled pattern and containing trabeculae of immature lamellar bone". A defect in the osteoblastic and osteoclastic activity, due to mutation in the GNAS gene, is thought to be the cause of this lesion. Incidence rate has been reported as 1:4000-1:10,000.[1] It accounts for 2-5% of all bone tumors and 7% of all benign tumors. It seems to affect females more than males.[2] Fibrous Dysplasia can affect one bone (monostotic) or multiple bones (polyostotic). Long bones like femur, ribs and craniofacial region are most commonly affected.[3] Monostotic FD is more prevalent than Polyostotic FD.[4] FD commonly develops in the childhood and continues through puberty and adolescence.[5] This article reports a case of FD observed in a young male.

Case History

A 21 year old male patient presented to the general dentist with a complaint of pain and swelling in upper front teeth for about 2 weeks. On examination, he was found to have an eggshell thin buccal plate in relation to teeth upper left central incisor (11), upper right central incisor (21), upper right lateral incisor (22), upper right canine (23) and upper right first premolar (24). No bone could be felt palatally, and a tentative diagnosis of periapical infection was made. Following intra oral periapical radiograph (IOPA), root canal therapy was commenced for 11, 21, and 22. Even with frequent medication, the infection did not subside. The patient was then recommended to get an Orthopantomogram (OPG) [Fig.1] and Cone Beam Computed Tomography (CBCT). Radiographic examination revealed a mixed radiopaque radiolucent, osteolytic lesion in the maxillary left anterior region. The lesion was approximately 34.01 x 23.55 x 25.24mm in size, round to oval in shape extending from 11, 21, 22, 23, 24 till 25. Both the buccal and palatal cortical plates were involved, with the loss of palatal plate and thinning of buccal plate. Dense radiolucent foci were also seen within the lesion. Root resorption with 22, 23 and 24 and loss of lamina dura with 21, 22, 23, 24, 25 was evident. Radiographic findings suggested an odontogenic cyst/tumor with respect to maxillary anterior region. After an incisional biopsy, a clinico-radiological correlation of traumatic bone cyst was made and surgical removal of the lesion was carried out under general anesthesia.[Fig.2].

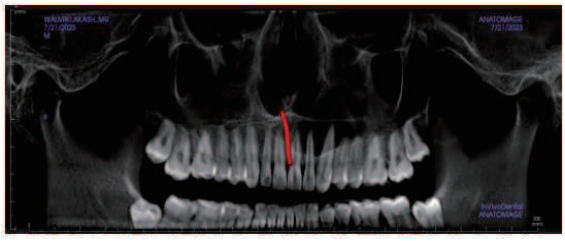

Figure 1: Orthopantomogram showing a well defined mixed radiopaque-radiolucent area with respect to left anterior maxilla.

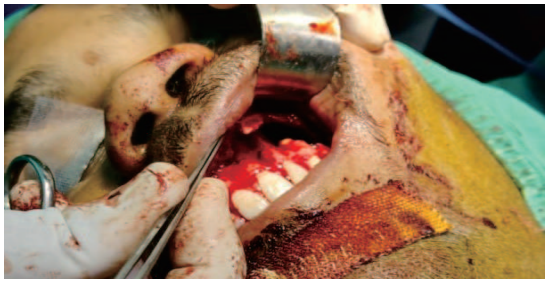

Figure 2: Surgical removal of the lesion

The lesion was curetted and then submitted for histopathological examination. Microscopic examination showed connective tissue that contained trabeculae of woven bone with absence of osteoblastic rimming, resembling a chinese letter pattern. Presence of plump fibroblasts, osteoblasts and dilated capillaries was also noted in the connective tissue stroma.[Fig. 3 & Fig. 4]

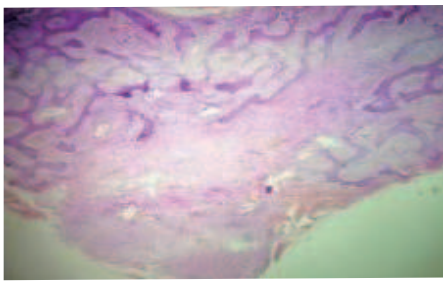

Figure 3: Scanner view - Connective tissue containing trabeculae of woven bone with Chinese letter pattern

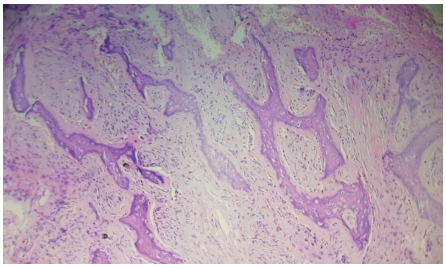

Figure 4: Low power magnification - Woven bone with absence of osteoblastic rimming and dilated capillaries

Based on histopathology a final diagnosis of Fibrous Dysplasia was made.

Discussion

Fibrous dysplasia is an anomaly where normal bone is replaced by fibrous connective tissue, which upon maturation is replaced by trabecular bone of irregular pattern.[6] It can be monostotic, i.e involving a single bone or polyostotic, involving multiple bones. About 70-80% of patients show the monostotic type. The common age of occurrence is second and third decade of life, and the common sites of occurrence are femur, ribs, tibia and cranial bones. Facial and cranial bone involvement in FD occurs in roughly 50% of polyostotic individuals and 10-27% of monostotic patients.[7] Majority of the craniofacial dysplasias are confined to the zygomatico-maxillary complex.[3] Many lesions are unilaterally present.[7]

The most common presenting feature of FD in the craniofacial region is a painless swelling leading to facial deformity, visual changes, nasal congestion and/or auditory disturbances.[2,4] Some other features that can be seen are dysesthesias in the distribution of the trigeminal nerve, epiphora and headaches.[8] Although asymptomatic, complaints of pain and discomfort may arise as a result of encroachment on canals and foramina.[6]

McCune-Albright syndrome (MAS) is associated with about 3% of polyostotic fibrous dysplasias. MAS is characterised by cutaneous hyperpigmentation, i.e. café-au-lait spots, precocious puberty, and fibrous dysplasia. Other endocrine disorders associated with MAS can be hyperthyroidism, hyperprolactinemia, acromegaly and Cushing's disease.[9] Similarly Jaffe-Lichtenstein syndrome may present with fibrous dysplasia and café-au-lait spots.[10]

A sporadic, postzygotic mutation of the GNAS1 gene located on chromosome 20q13 is thought to be the cause.[7]

A recommended diagnostic approach for bony lesions is 3D imaging. A 'ground-glass' appearance is a characteristic radiographic finding of this lesion. Periodontal space thins out and periapical bone becomes abnormal with loss of lamina dura.[2] The radiographic appearance depends on the stage of lesion, with early stages showing radiolucency and maturing stages exhibiting characteristic sclerotic areas.[4] Bone scintigraphy is usually performed, to rule out polyostotic variant.[8] Panda et.al have described 3 distinct patterns, the pagetoid appearance characterised by expansion of bone and scattered bony islands, the sclerotic appearance characterised by homogenous ground glass appearance and the cystic pattern characterised by well-defined lesion with a sclerotic margin.[7]

The characteristic microscopic findings of FDs are Chinese letter patterns which are irregular, curvilinear trabeculae of woven bone seen in a low to moderately cellular connective tissue stroma of spindle -shaped cells surrounding them.[9,11]

Central ossifying fibroma, central giant cell granuloma aneurysmal bone cyst should be considered as the differential diagnoses for fibrous dysplasia.[9]

A fibrous stroma with varied mineralized products is a characteristic histologic feature of benign fibro-osseous lesions. Cemento-Osseous Dysplasias, ossifying fibromas and fibrous dysplasias are the conditions that classify as being fibro-osseous lesions of the jaws.[12]

Cemento-osseous dysplasia (COD) is a non-neoplastic abnormality of exclusively the tooth-bearing areas that affects the maxilla more commonly than the mandible. COD are further classified into, periapical, florid and focal types. COD is found commonly in middle-aged black women. The lesion is generally asymptomatic, except for the florid variant, and found incidentally on radiography. Radiographically the lesion presents with an outer irregular radiolucent zone and central amorphous radiopacity. Microscopically it contains a fibrous stroma with loose collagen with associated mineralised tissue, that can consist of woven or lamellar bone, osteoid and/or cementum-like material.[12]

Cemento-Ossifying fibroma is another benign bone pathology that affects the craniofacial bones. There are three variants, namely cemento-ossifying fibroma, juvenile trabecular ossifying fibroma and juvenile psammomatoid ossifying fibroma. Cemento-Ossifying fibromas occur in the mandible rather than the maxilla.[12] Smaller tumors may be painless. The juvenile variants show rapid growth and expansion which may cause facial swelling and asymmetry. Radiographically the cemento-ossifying fibromas are well-defined and concentric, with a uniform radiolucent border. A cemento-ossifying fibroma may cause root resorption and displacement of adjacent teeth.[12]

Microscopically a hypercellular fibrous tissue with mineralized content is seen. Osteoblastic rimming is a characteristic feature of cemento-ossifying fibroma. In juvenile trabecular type, cellular osteoid trabeculae with focal mineralization at the centre is a characteristic feature. Psammoatoid bodies are characteristically seen in the juvenile psammomatoid type.[12]

The goal of treatment for fibrous dysplasia is to address any functional disturbance while also improving the aesthetics.[13]

A wide range of presentations of this disease may make it difficult to establish a single modality of treatment. An individualized approach based on patient's functional and aesthetic needs is the way to go about it.[14] Medical therapy in the form of bisphosphonates has been advised in the literature. Bisphosphonates control bone erosion by inhibiting the osteoclastic activity. Pamidronate intravenously has also been suggested as it may help reduce bone pain intensity and bony resorption.[7]

Abnormal osteoprogenitor cells are seen to be increased and is a significant observation in the molecular mechanism of FD. Abnormal formation and orientation of non-collagenous and collagenous proteins were observed. Dysplastic bone showed increased osteonectin levels and a decrease in osteopontin and bone sialoprotein. These changes in the matrix reduce the adherence of the osteoclasts which leads to cell retraction and disorientation. This transforms into a disorganized immature bone matrix. Zones of resorption seen in dysplastic bone are characterized by the presence of multiple osteoclasts resorbing the mineralized matrix which help in the formation of local lytic areas.[15]

Dysplastic cells show an increased proliferation rate and increased intracellular cyclic AMP. Decreased cell differentiation and increased cell proliferation of pre-osteoblasts in dysplastic lesions bring about rapid deposition of an immature, poorly organized woven bone at the histological level.[15]

A recurrence rate of 15% to 20% is noted[14]. The risk of malignant transformation with fibrous dysplasia is 0.4%. Serum alkaline phosphatase levels are important for spotting recurrences.[1] In about 25%-50% of patients, the lesions may regrow after the initial surgery. Although the lesion may stop growing once skeletal maturity is attained, some lesions continue to grow, making follow-up assessments a necessity.[4]

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