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Fibrous Dysplasia – A Case Report and Review of Literature

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Abstract Fibrous dysplasia (FD) is a fibro-osseous lesion with no apparent familial, hereditary or congenital basis. It is a non-neoplastic developmental hamartomatous disease of the bone, characterised by a blend of fibrous and osseous elements in the region. It constitutes 2.5% of all bony neoplasms and 7% of all benign bony neoplasm. The treatment can be either conservative or complete resection. Here we report a case of fibrous dysplasia in a 64-year-old male patient on the right side of the face.

Keywords: *fibrous dysplasia, neoplasm, fibro-osseous lesion*

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1. Introduction

Lichtenstein first coined the term in 1938 [1] and in 1942 he and Jaffe separated it from other fibro-osseous lesions [2].

Fibrous dysplasia is divided into monostotic and polyostotic variety. [3] Polyostotic variety includes Jaffe syndrome and McCune-Albright syndrome. It may involve multiple bones associated with cutaneous pigmentation and precocious puberty known as Albright's syndrome [4].

In this paper, we present a case of fibrous dysplasia in a 64-year-old male patient on the right side of the face.

2. Case Report

A 64-year-old male patient reported to our dental OP with a chief complaint of painless swelling in right side middle third of face since 30 years.

Patient was apparently normal 30 years back. Patient gave a history of swelling which was gradual in onset, initially small in size and later progressed to attain present size. Patient also gave no relevant history of associated pain and fever. Patient had consulted a private practitioner for the same complaint 30 years back. Patient was wearing a complete denture for completely missing maxillary and mandibular teeth. The complete denture did not fit well as the mass progressed in size.

On extraoral examination, facial asymmetry was evident due to a diffuse swelling on middle third of the face on the right side. Swelling extended superiorly from infraorbital margin, 2cm below ala-tragal line inferiorly, 1cm away from ala of nose anteriorly, 2cm ahead of tragus posteriorly. Nasolabial fold was not obliterated. Swelling measured approximately 3X2 cm in size, roughly oval in shape. Skin over the swelling appeared normal. On palpation, inspectory findings with respect to site, size, shape and extent were confirmed. The swelling was hard in consistency and not tender. Pulsations were not visible or palpable.

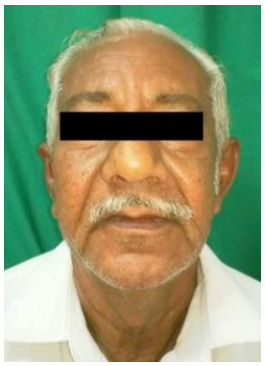

Figure 1. Facial asymmetry due to a mild, diffuse swelling on right middle third of the face

On intraoral examination, a single well-defined swelling was evident on edentulous area on buccal aspect in relation to 15 to 17 region. It extended medially from alveolar ridge and obliterated the buccal vestibule laterally in relation to 15, 16 and 17. The swelling measured approximately 3 X 2 cm in size and was roughly oval in shape. No secondary changes were evident. On palpation, inspectory findings with respect to site, size, shape and extent were confirmed. The swelling was not tender and hard in consistency. No visible or palpable pulsations were evident.

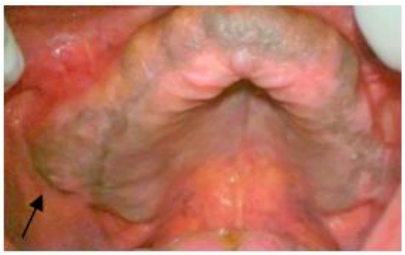

Figure 2. A single well-defined swelling evident on edentulous area on buccal aspect in relation to 15 to 17 region

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 on edentulous region in relation to 14 to 17 extended upto right maxillary sinus. It measured approximately 5 X 4 cm in size and roughly oval in shape. The borders were ill-defined borders and involved of right maxillary antrum.

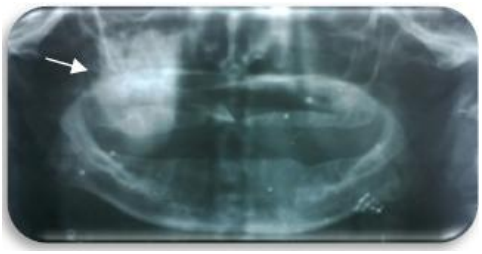

Figure 3. OPG revealed a mixed radiopaque-radiolucent area in right maxillary quadrant

Computed tomography (CT) scan of facial bone in bony window of right maxillary antrum showed a diffuse ground glass opacities of HU 528 IU mixed with areas of soft tissue density of HU 551 IU surrounding cortex appears normal with no evidence of bony erosion. Edentulous jaw with an impacted 13 was evident. CT study shows features suggestive of fibrous dysplasia.

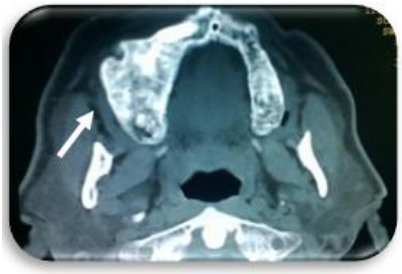

Figure 4. CT scan of facial bone in bony window of right maxillary antrum showed a diffuse ground glass opacities

An incisional biopsy was performed which microscopically revealed a fibrous connective tissue containing many slender irregular trabecular pattern of woven bone focally rimmed by osteoblasts, suggestive of fibrous dysplasia.

Based on the history, clinical examination and investigations the case was diagnosed as fibrous dysplasia or right maxilla.

The mere presence of FD of the craniofacial region is not in itself an indication for treatment. Thus for this patient, treatment was deferred, however he is under regular follow up.

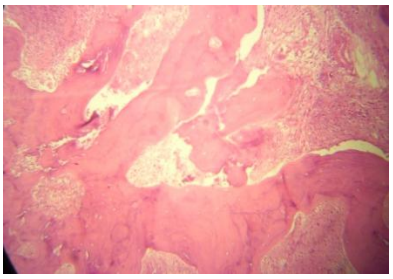

Figure 5. Histopathologic section revealed a fibrous connective tissue containing many slender irregular trabecular pattern of woven bone

3. 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. They usually stabilize when the patient reaches skeletal maturity. However, similar to our case presented here there have also been reports of persistence at later periods underlining their variable clinical behaviour. There is no gender predilection. Common sites of involvement are femur, tibia, fibula, ribs and facial bones. It involves the maxilla almost twice as often as the mandible, frequenting the posterior region and is usually unilateral in nature.[5] The mandible, ethmoid and sphenoidal regions follow in the order. In the present case, right side maxilla was involved.

Various theories have been proposed regarding the etiology of FD, including the trauma with a nonspecific disturbance 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. The most acceptable theory is the abnormal activity of mesenchymal cells.

The pathogenesis of FD 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 the maturation fails, it leaves behind a mass of immature isolated trabeculae enmeshed in dysplastic fibrous tissue that are turning over constantly but never completing the remodeling process. [8] The combination of insufficient mineralization and a lack of stress alignment can lead to loss of mechanical strength. This can lead to deformity, pain 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].

The radiographic features of FD 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 radiographic picture is more radiolucent and well defined in the early stages and becomes mottled and more radio opaque as the disease progresses.

The radiolucent region is composed histologically of a solid fibro-osseous mass of tissue, which may exhibit a cystic component filled with fluid. The lesion is bounded

by a distinct rim of reactive bone that is defined sharply on its inner border and gradually fades into normal cancellous bone. The lesions originate in the medullary canal but consistently replace both cancellous and cortical bone. Thus the sharp distinction between the cortex and the medullary canal is lost. The diameter of the bone is increased by growth of the lesion and is bounded by a shell of reactive bone. The surface of periosteum is smooth and without any periosteal reaction [11].

The main treatment for FD is surgery, which can be divided into conservative and radical resection. Any features of deformity, pain or functional disability suggest the need for intervention. Complete surgical resection of the involved area is the treatment of choice with extensive reconstruction.

Radiotherapy is avoided as it can induce malignant changes in FD. Bisphosphonate therapy may help to improve decrease pain, function and decreases risk of fracture.

The disease nearly always burns itself out around the puberty. In the present case, the patient noticed the swelling only in the third decade of life and has not regressed in size.

The patient reported in this case was 64 years old with a lesion that was reported to be slowly progressive, which is unlike the natural history of the disease, where the growth corresponds to the period of skeletal growth.

The mere presence of FD of the craniofacial region is not in itself an indication for treatment. Small solitary lesions may remain asymptomatic for long periods. Thus for this present case report, treatment was deferred and is under regular follow up.

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