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FIBROUS DYSPLASIA OF THE MANDIBLE: a case report

DR.C.AYYADURAI	III YEAR PG, DEPT OF ORALPATHOLOGY AND MICROBIOLOGY, VINAYAKA MISSIONS SANKARACHARIYAR DENTAL COLLEGE, SALEM, TAMILNADU. 636308.
DR.B.SEKAR	M.D.S, PROFESSOR AND H.O.D, DEPT OF ORALPATHOLOGY AND MICROBIOLOGY, VINAYAKA MISSIONS SANKARACHARIYAR DENTAL COLLEGE, SALEM, TAMILNADU. 636308.
DR.MAYA RAMESH	M.D.S, READER, DEPT OF ORALPATHOLOGY AND MICROBIOLOGY, VINAYAKA MISSIONS SANKARACHARIYAR DENTAL COLLEGE, SALEM, TAMILNADU.636308.
DR.SUBHASHINI	III YEAR PG, DEPT OF ORALPATHOLOGY AND MICROBIOLOGY, VINAYAKA MISSIONS SANKARACHARIYAR DENTAL COLLEGE, SALEM, TAMILNADU. 636308.
DR.B.S.SARAVANAN	M.D.S, ORAL AND MAXILLOFACIAL SURGEON, VIMS HOSPITAL, SEERAGAPADI, SALEM, TAMILNADU.636308.

ABSTRACT

Introduction: Fibrous dysplasia is a developmental tumor like condition, characterized by the replacement of normal bone marrow by proliferation of cellular fibrous connective tissue with irregular bony trabeculae. This condition occurs due to mutations in the GNAS gene.

Case presentation: A 17-year-old female patient was referred to our dental unit with a complaint of severe pain and swelling in left side of the face for past 6 years. History of present illness revealed patient was apparently normal 6 years back, then she developed pain and swelling and it attains current size.

Discussion: Monostotic fibrous dysplasia is more common in the maxilla than in the mandible. It has slight female predilection and manifests in the first 3 decades of life.Computed tomography accurately shows the extent of bone involvement and sarcomatous transformation. Biphosphonates, opioids, NSAID are the current and potential therapies for treatment of fibrous dysplasia pain. Skeletal deformities sometimes need a surgical treatment.

Conclusion: There is no complete cure for fibrous dysplasia, only symptomatic treatment is needed according to the situation. But periodical CT scan, clinical assessment and regular health checkup is mandatory to identify the disease progression.

KEYWORDS Craniofacial abnormalities, Fibrous dysplasia, mandible.

Introduction:

In 1891, Von Recklinghausen is a person who coined the name "osteitis fibrosa generalisata" for a patient with skeletal deformities.[1] But the name "Fibrous dysplasia" was first introduced by Lichtenstein in 1938.[2] It is a developmental tumor like condition, characterized by the replacement of normal bone marrow by proliferation of cellular fibrous connective tissue with irregular bony trabeculae. This condition occurs due to mutations in the GNAS gene.[3] It can occur any bone of the skeleton and accounts for 2% to 3% of all bony tumors.[4]

Fibrous dysplasia is mainly of two major types: 1.monostotic, involving a single bone. 2.polyostotic, involving multiple bones and having multiple lesions. The monostotic fibrous dysplasia is the common, comprising 70% of cases, usually occurs at puberty. It commonly involved in femur, tibia , ribs and only 10% of the cases occurs in craniofacial bones.[5] In jaws, the maxilla is more affected than the mandible. Mandibular lesions are truly monostotic but maxillary lesions can extends to adjacent bones. [3] Here we report a case of 17 years old female patient with a histopathological diagnosis of fibrous dysplasia of the mandible.

Case presentation:

A 17-year-old female patient was referred to our dental unit with a complaint of severe pain and swelling in the left side of face for past 6 years. History of present illness revealed that patient was apparently normal 6 years back, then she developed pain and swelling and it attains current size. After medication swelling decreased in size for some days and again attained the same size. General examination revealed that patient is conscious, co-operative, well oriented to time & place, well built, nourished and posture are normal.

Extra oral examination revealed swelling in the left side of face. (FIGURE 1) On inspection, a diffuse swelling on the left lower side of face measuring about 3x4 cm extending superio-inferiorly along the line from corner of mouth to 1cm below to the lower border of mandible and antero-posteriorly 1cm away from symphysis region to 1cm infront of angle of mandible. Colour of the swelling was same that of adjacent skin. On palpation, all inspectory findings in relation to size, shape, and extension were confirmed on palpation. The swelling was hard in consistency, not fluctuant, not compressible. The skin over the swelling was pinchable, No discharge present. The lower border of the mandible was intact. On lymph node examination, Single left and right submandibular lymph nodes were palpable, non tender, soft in consistency, measuring about 1x1 cm in diameter.

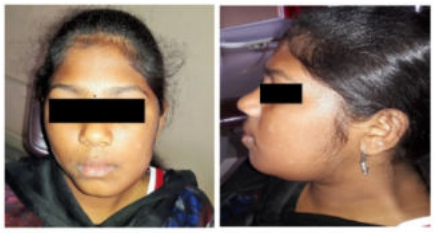

FIGURE 1: Photograph showing front and left side of the face.

On intra oral examination, a diffuse solitary swelling seen in the left buccal vestibule (FIGURE 2) measuring about 3x1cm extending mediolaterally from the gingival margin in 34, 35, 36, 37 to the buccal vestibule in relation to 35, 36, 37 anteroposteriorly from mesial aspect of 34 to distal aspect of 37. Obliteration of buccal vestibule present. Colour of the swelling was same as that of adjacent mucosa. No ulcerations and sinus openings were present. On palpation, all the inspectory findings were confirmed. The swelling was bony hard in consistency, tender,and was not fluctuant.

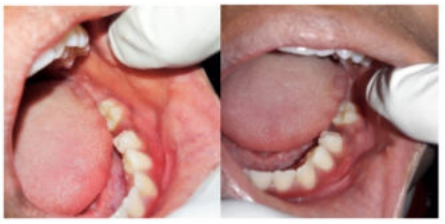

FIGURE 2: Photograph showing intraoral swelling.

A computed tomography (CT) scan (FIGURE 3) showed a solid mass affecting a portion of the mandible on the left side. Microscopic evaluation (FIGURE 4) showed the typical histologic signs of fibrous dysplasia including the presence of irregular bony trabeculae with osteocytes inside the lacunae. Chinese letter pattern and 'c' shaped trabeculae seen in many areas. Connective tissue and blood vessels are seen in between bony trabeculae.

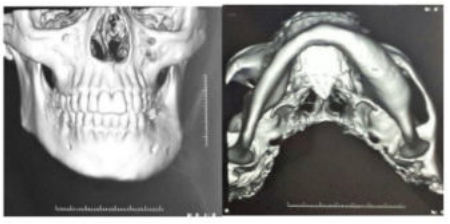

FIGURE 3: CT scan showing swelling in mandible

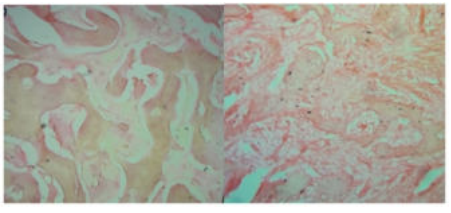

FIGURE 4: Photomigrograph showing typical histologic signs of fibrous dysplasia

Discussion:
Fibrous dysplasia mainly classified as monostotic and polyostotic, in this Polyostotic fibrous dysplasia is further classified as, 1. Fibrous dysplasia involving many bones with pigmented skin lesions (café-au lait spots) 2. Fibrous dysplasia involving nearly all the bones of the skeleton with skin lesions and endocrine disturbances (McCune Albright syndrome). [6] A complete history and physical examination were needed to identify the extent of disease whether it is isolated or extents to multiple bone.[7]

Monostotic fibrous dysplasia is more common in the maxilla than in the mandible.[8] It has slight female predilection and manifests in the first 3 decades of life and it is commonly stabilized during skeletal maturity.[1] Computed tomography accurately shows the extent of bone involvement and sarcomatous transformation.[9] Histopathologic evaluation confirms the diagnosis. Immunohistochemistry serves no role in the diagnosis of fibrous dysplasia.[10] The differential diagnosis may include, odontogenic tumour, giant-cell granuloma , cherubism, ossifying fibroma, osteoma and osteosarcoma. Commonly, monostotic lesions are asymptomatic, localized pain may be the symptom in some patients. Because of estrogen receptors, female patients can get more pain during pregnancy and menstruation.[11] Sometimes facial bone involvement can cause and difficulty in opening of the eyelids, chewing, speaking and obstruction of the nostrils.[8]

Biphosphonates, opioids, NSAID are the current andpotential therapies for treatment of fibrous dysplasia pain.[12] Skeletal deformities sometimes need a surgical treatment like either conservative contouring or radical excision but it should be done after skeletal maturity.[13] Treatment needed mainly depends on lesions effect on regular physiological function, its location, and cosmetics. Malignant transformation may occur rarely (1% average, 4% in McCune-Albright) and the prognosis of fibrous dysplasia is normally very good.[14]

Conclusion: There is no complete cure for fibrous dysplasia, only symptomatic treatment is needed according to the situation. But periodical CT scan, clinical assessment and regular health checkup is mandatory to identify the disease progression. It will helps to early detection of disease progression and for better treatment.

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