Journal of Cardiovascular Disease Research

ISSN: 0975-3583, 0976-2833 VOL12, ISSUE01, 2021

POLYOSTOTIC FIBROUS DYSPLASIA: A RARE CASE REPORT

Dr.Sejal Samson
Lecturer, Department of Oral Medicine and Radiology
D Y Patil University, School of Dentistry, Navi Mumbai
Email-id -sssejal9@gmail.com
Lecturer, Department of Oral Medicine and Radiology
D Y Patil University, School of Dentistry, Navi Mumbai
Email-id-drhemantbhutani@gmail.com
Dr.Bhavika Vhatkar
Associate professor, Department of Oral Medicine and Radiology
D Y Patil University, School of Dentistry, Navi Mumbai
Email Id- drpolvhatkar@gmail.com
Dr Janhavi Chandure
Post Graduate Student
Department of Oral Medicine and Radiology
D Y Patil University, School of Dentistry, Navi Mumbai
Email id- janvichandure96@gmail.com
Dr. Shrutika Mhatre
Post Graduate Student
Department of Oral Medicine and Radiology
D Y Patil University, School of Dentistry, Navi Mumbai
Email id - shrutamh97@gmail.com
Dr .Anindya debnath
Post Graduate Student
Department of Oral Medicine and Radiology
D Y Patil University, School of Dentistry, Navi Mumbai
Email Id –anindyad78@gmail.com

Abstract:
A fibro-osseous lesion known as ‘fibrous dysplasia’ has an unidentified underlying etiology. Based on the number of affected bones, it is divided into three categories: monostotic (80–85%), polyostotic (20–30%), and Albright's disease. A condition known as "polyostotic"—which is described as somewhat uncommon disorder-occurs when two or more bones are involved. This case report describes a rare instance of polyostotic fibrous dysplasia in a female patient who was 19 years old. The lesion affected her maxilla, zygomatic bone, sphenoid bone, and orbital bones.

Key words: Fibrous dysplasia, polyostotic, CBCT.

INTRODUCTION

A defect in osteoblastic differentiation and maturation results in fibrous dysplasia, a developmental aberration. The impacted bones exhibit sclerosis, thickening, and enlargement.[1]

ISSN: 0975-3583, 0976-2833 VOL12, ISSUE01, 2021

It is distinguished by the abnormal growth of cellular fibrous connective tissue that replaces normal bone, which is mixed with uneven bone trabeculae.[2,3] Clinically, it is classified into 3 types, monostotic (80-85%), polyostotic (20-30%), and Albright's disease with the incidence in monostotic being the highest. As opposed to polyostotic, which is multifocal and affects multiple bones at once, monostotic is focal and limited to a single bone.[4] The sex predilection in monostotic fibrous dysplasia is not significant whereas polyostotic fibrous dysplasia is seen mostly in females.

Clinically, it is a bony hard swelling with irregular margins, seen unilaterally in most cases. The overlying skin appears normal. The lesion shows slow growth and usually, the patient may not recollect when the lesion was initially perceived.[5]

CASE REPORT

A patient who was 19 years old presented to the Oral Medicine and Radiology department with the primary complaint of swelling in the upper right anterior and posterior region of the face for 12–13 years.

History revealed that the swelling appeared when the patient was 6 years old and has been increasing since then. The patient has no pain associated. The swelling has been slow growing and progressive in nature with no history of any treatment performed. No secondary changes were seen since then. The patient has no difficulty chewing or speaking.

The patient shows no relevant medical history, no relevant family history and no deleterious lifestyle habits. She was conscious, well-oriented, and cooperative.

Extraoral Examination shows facial asymmetry with diffuse swelling on the right side of the face extending anteroposteriorly, from the bridge of the nose to the preauricular region. Supero-inferiorly, from the infra-orbital margin to the level of the corner of the mouth with measuring 5 inches x 3 inches x 3 cms approximately. (Figure 1) Swelling is bony-hard in consistency with irregular margins. No change in color of the overlying skin or change in temperature.

Fig 1: Extraoral photograph

Intraoral examination shows diffuse swelling extending from distal of 13 to distal of 17. The color of mucosa over the swelling appears pale pink. Enlargement of the alveolus and obliteration of the muco-buccal in the maxillary right vestibular region. On palpation, the swelling had a bony, hard consistency and was not painful. All teeth were present (except third molars), non-mobile, non-tender. No abnormality was

ISSN: 0975-3583, 0976-2833 VOL12, ISSUE01, 2021

seen in the palatal region. Occlusal caries were seen with respect to 36 and 46 with pulp polyps in both the teeth. (Figure 2)

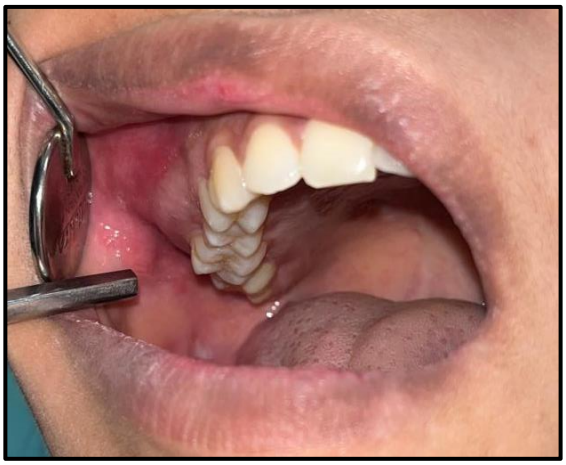

Fig 2: Intraoral photograph

Based on the clinical findings, the provisional diagnosis was a fibro-osseous lesion or a benign tumor or a benign cyst of the maxillary and zygomatic right region of the face.

Differential diagnosis can be fibrous dysplasia, adenomatoid odontogenic tumor, dentigerous cyst

Based on the clinical findings, for radiographic investigations, a cone beam computed tomography (CBCT) must be done along with a bone biopsy.

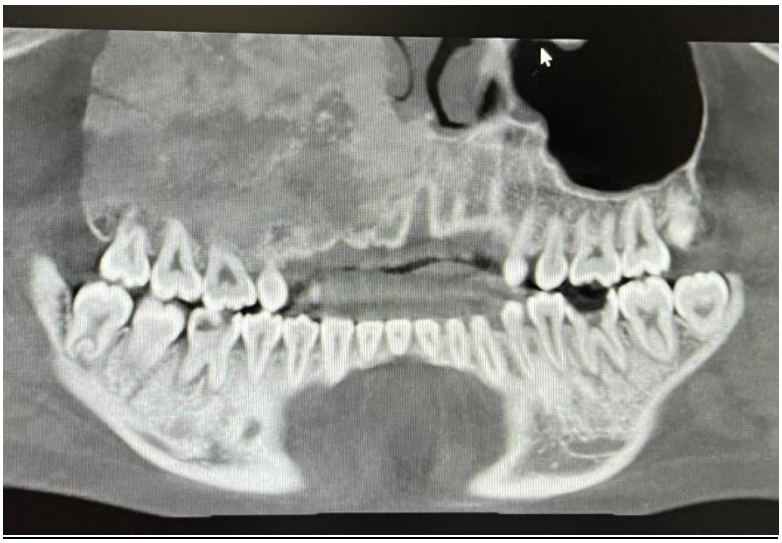

Fig 3: Panoramic Section showing ground glass radio-opacity (characteristic appearance) involving nasal bone, maxillary sinus, and zygomatic bone.

ISSN: 0975-3583, 0976-2833 VOL12, ISSUE01, 2021

The panoramic section shows a heterogenous radiopaque and radiolucent lesion extending from the left canine region to the right side including the entire right maxilla, wall of the nasal floor, and zygomatic bone. It displays effacement of the nasal floor and anterior maxillary sinus wall as well as obliteration of the right maxillary sinus. (Figure 3)

The coronal section shows involvement of the entire right maxilla along with zygomatic and frontal process. It also shows involvement of greater and lesser wing of sphenoid bone with sphenoidal air sinuses. [Figure 4(A), Figure 4(B)]

The axial section shows ground-glass appearance of the maxilla, zygoma, sphenoid and nasal bone. [Figure 4(C)]

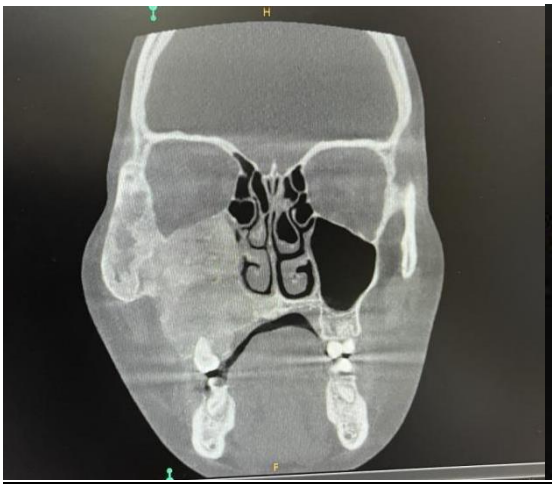

Fig 4 (A): Coronal Section

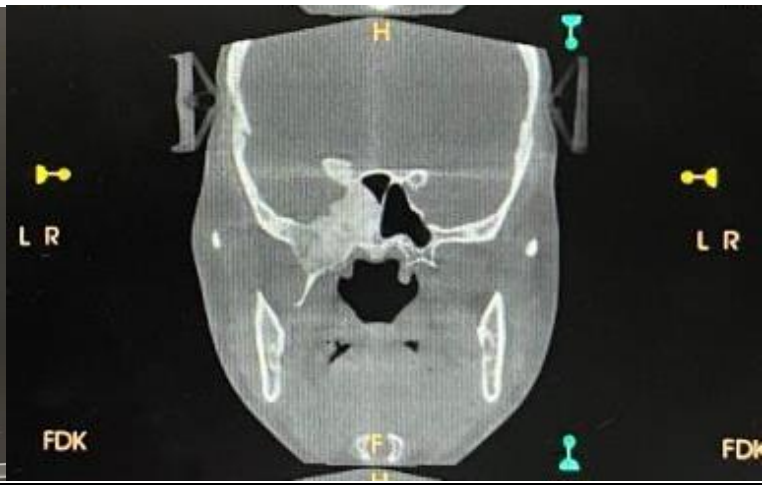

Fig 4 (B): Coronal Section

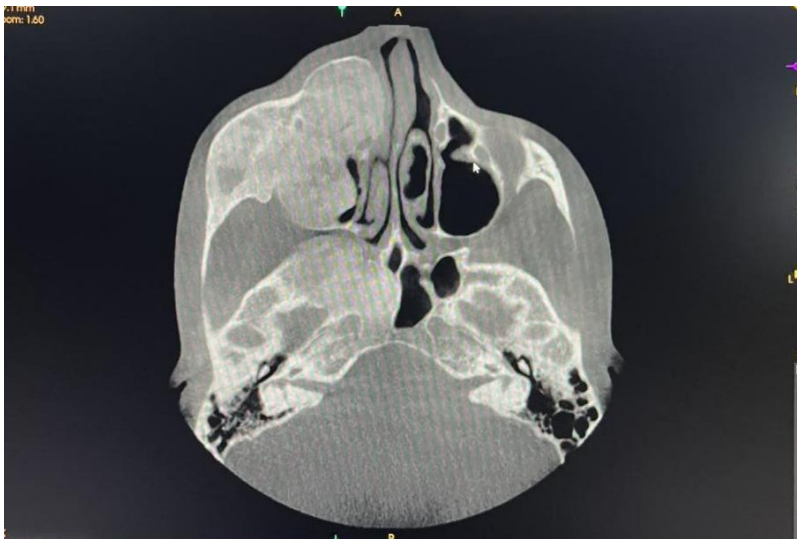

Fig 4(C): Axial Section

ISSN: 0975-3583, 0976-2833 VOL12, ISSUE01, 2021

Histopathological section stained with H and E shows fibro-osseous lesion comprising of osseous tissue and cementum-like tissue interspersed in loose fibro cellular connective tissue stroma. The osseous tissue is in the shape of bony trabaculae comprising of osteocytes and active osteoblastic rimming. Cementum-like dystrophic calcification appears basophilic, irregular in shape and size suggestive of a fibro-osseous lesion.
Hence, the final diagnosis for the case is craniofacial fibrous dysplasia involving the right maxilla, temporal, zygomatic, sphenoidal, and frontal bone.
Re-contouring of the bone is planned as a treatment.

DISCUSSION

Fibrous dysplasia is an uncommon benign disease that is caused due to defective bone modeling. It is believed to be a congenital anomaly of mesenchymal origin.[6] It is a benign lesion caused by bone remodeling, because of constant alterations of the bone. The normal bone is replaced by fibrous immature tissue.
Based on the number of affected bones, it is divided into three categories: monostotic (80–85%), polyostotic (20–30%), and Albright's disease. A condition known as "polyostotic"—which is described as somewhat uncommon disorder, occurs when two or more bones are involved. The prevalence of polyostotic fibrous dysplasia is higher in females. A few up to 75% of the skeleton's bones can be damaged, and almost all individuals with polyostotic fibrous dysplasia have craniofacial involvement, often with a startling preference for one side of the body. [7]

Various authors describe embryological and congenital theories to explain their etiology.[8,9] A post-zygomatic mutation in the GNAS1 gene (protein attached to the guanine nucleotide-stimulating activity of polypeptide 1) causes the rare disease fibrous dysplasia.[8] Fibrous dysplasia causes differentiation, disintegration, and disorganization of the bone.

Generally, the disease is asymptomatic. The face shows asymmetry in the zygomatic-maxillary region, due to obvious swelling unilaterally. There may be the presence of pathological or physiological fractures in the surrounding area. There may be a loss of nasomaxillary angle due to the expansion of the maxilla leading to a typical feline facial appearance.[10] Obstruction of paranasal sinuses can lead to recurrent infection in the sinus region and cysts. Loss of visual acuity caused by optic nerve compression along with hearing loss, ptosis, exophthalmos, strabismus, and altered eye motions.
Conventional radiography, scintigraphy, magnetic resonance imaging, and computed tomography scans (CT) can all be used to evaluate it, while CT is the exam of choice for evaluating craniofacial lesions.[10,11]
The differential diagnosis includes benign lesions: Non-ossifying fibroma, Solitary unilocular cyst, meningioma, eosinophilic granuloma, cholesteatoma, Paget's disease, osteochondroma, exostoses, ossifying fibroma, aneurysmal bone cyst, giant cell reparative granuloma, cystic fibrous osteitis; and malignancies, such as metastatic osteoblastic lesions and sarcoma.[1]
Bone biopsy specimen showed the presence of trabeculae and interspersed fibrous connective tissue suggestive of fibrous dysplasia.
The best choice of treatment will be restricted to bone remodeling for this particular case. Other intensive surgeries can be done in cases where vision is impaired and where correction of the dentition is required.

ISSN: 0975-3583, 0976-2833 VOL12, ISSUE01, 2021

CONCLUSION

Fibrous dysplasia is of pathological origin. Post-treatment of the disease, regular follow-ups are required to examine the patient for any malignant transformations or relapses.

Fibrous dysplasia causes asymmetry, due to which facial aesthetics are compromised. Hence, bone remodeling must be performed according to age, gender, and patient's facial profile.

References:

1. Sivapathasundharam N. Bone and Joint Diseases. Shafer's Textbook of Oral Pathology. 8th ed. Elsevier; 2016. p. 462-6.
2. Costanzi MA, Velasco e Cruz AA (2007) Envolvimento orbitário difuso por displasia fibrosa na síndrome de mccune albright: Relato de caso. Arq Bras Oftalmol 70: 1021-1023.
3. Neville BW, Damm DD, Allen CM, Bouquot JR (2004) Patologia Oral & Maxilofacial. (2nd edn), Rio de Janeiro: Guanabara Koogan.
4. Simon E, Matee M, Shubi F, Mselle T (1999) Cranio-facial fibrous dysplasia in a 38-year-old African woman: A case history. Oral Dis 5: 247-249.
5. Chen YR, Wong FH, Hsueh C, Lo LJ (2002) Computed tomography characteristics of non-syndromic craniofacial fibrous dysplasia. Chang Gung Med J 25: 1-8.
6. Jan M, Dweik A, Destrieux C, Djebbari Y (1994) Fronto-orbital sphenoidal fibrous dysplasia. Neurosurgery 34: 544-547.
7. Botelho RA, Tornin OS, Yamashiro I, Menezes MC, Furlan S, et al. (2006) Características tomográficas da displasia fibrosa craniofacial: Estudo retrospectivo de 14 casos. Radiol Bras 39: 269-272.
8. Shafer WG, Hine MK, Levy BM (1987) Doenças dos ossos e das articulações. In: Shafer WG, Hine MK, Levy BM, Tratado de Patologia Bucal. Rio de Janeiro: Guanabara Koogan.
9. Akintoye SO, Otis LL, Atkinson JC, Brahim J, Kushner H, et al. (2004) Analyses of variable panoramic radiographic characteristics of maxillo-mandibular fibrous dysplasia in Mccune-albright syndrome. Oral Dis 10: 36-43.
10. Lisle DA, Monsour PA, Maskiell CD (2008) Imaging of craniofacial fibrous dysplasia. J Med Imaging Radiat Oncol 52: 325-332.
11. Mac Donald-Jankowski DS (2004) Fibro-osseous lesions of the face and jaws. Clin Radiol 59: 11-25.