 DOI: 10.18231/2395-6194.2017.0044

Fibrous Dysplasia – Report of 2 cases

Raghunath Dantu[1,*], Satya Prakash[2], Surekha R. Puranik[3], Praveen Ramdurg[4]

[1,2]Consultant, Dept. of Oral & Maxillofacial Radiology, [3]HOD, [4]Reader, Dept. of Oral Medicine & Radiology, PMNM Dental College & Hospital, Bagalkot

***Corresponding Author:**
Email: raghunath.dantu@gmail.com

Abstract

Fibrous dysplasia (FD) is a benign fibro-osseous bone disease of unknown etiology and uncertain pathogenesis. It is a disorder where normal bone is replaced with fibrous tissue. The lesion frequently affects the craniofacial skeleton. The maxilla is affected twice comparing mandible and occurs more frequently in the posterior area. In this series, two female patients in their third decade presented with asymmetry of the face were diagnosed as Fibrous dysplasia.

Keywords: Monostotic, Ground glass

Introduction

Fibrous Dysplasia according to Charles Waldron is believed to be a non-neoplastic hamartomatous developmental lesion of bone of unknown origin.(1) It should no longer be considered a hamartomatous or developmental disorder of bone, but rather a "benign neoplasm with malignant potential".(2) It is a benign dysplastic process of altered osteogenesis that may occur either in a single bone (Monostotic) or multiple bones (polyostotic). It is an important lesion affecting the maxillofacial region because it can cause severe deformity and asymmetry, and most devastating of all, blindness.

The term Fibrous dysplasia was first suggested by Lichtenstein in 1938 as a designation for multiple (polyostotic) bone lesions of the type described by Albright et al as osteitis fibrosa disseminata. Lichtenstein and Jaffe (1942) later expanded this concept and noted that an isolated (monostotic) form of the disease was considerably more common than the polyostotic form.(3)

The clinical findings are asymptomatic involved bone enlargement which causes facial asymmetry, loss teeth and facial deformity.(4,5) If the craniomaxillofacial bones are affected by FD, due to megacranium, the face of the patient is referred to 'lion face'.(4) The complications of the lesions involving sphenoid, orbital, frontal bones, are proptosis, visual disturbances, facial asymmetry and orbital dystopia.(6,7) The fifth nerve impairment, hearing loss and seizure disorders have been reported as neurological complications.(8)

Case Report 1

A 22 year-old female presented with a chief complaint of swelling on the left side of maxilla. Patient gave history of swelling since childhood which was small in size and has gradually increased in size with no apparent increase since 1 year.

On extra oral examination diffused swelling of about 2 x 2cm was present on left side of the face extending superio-inferiorly 0.5 cm below the ala tragus line; to line joining corner of mouth to angle of mandible and anteroposteriorly 0.5 cm away from the corner of the mouth to 1 cm in front of left tragus. On palpation the consistency was bony hard, nontender & no local rise of temperature.

Intraoral inspection reveals diffuse swelling present on the left side corresponding to 24, 25, 26, 27 measuring 4×2cm extending anteroposteriorly from mesial aspect of 24 to distal aspect of 27 with slight displacement of teeth palatally, superoinferiorly from the upper buccal vestibule to gingival margin of 24, 25, 26, 27 with obliteration of the vestibule (Fig. 1). Surface mucosa appears to be normal. The swelling is bony hard in consistency, nontender on palpation.

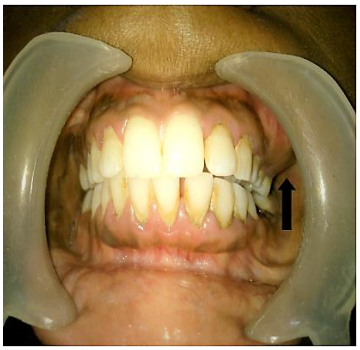
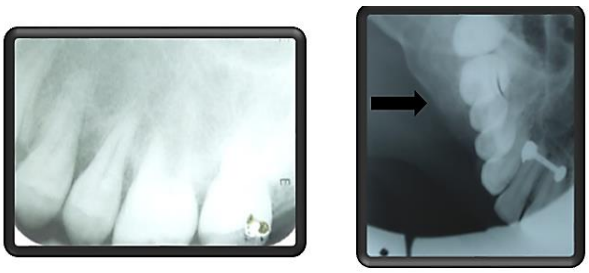


Fig. 1: Intraoral buccal expansion

Radiological investigation included IOPA, Occlusal, OPG and CT scan. Intra oral periapical radiograph revealed ground glass homogenous appearance of trabeculae extending from premolar to the molar area and from alveolar crest region to the sinus area of 24, 25, 26, 27. Surrounding lamina dura and periodontal ligament space appears to be normal (Fig. 2a). Occlusal radiograph revealed expansion of buccal cortex showing homogenous ground glass appearance with few granular areas (Fig. 2b).



(a) (b)

Fig. 2: IOPA (a) and Occlusal (b) radiograph showing ground glass appearance and buccal expansion

CT scan showed homogenous ground glass appearance involving the left maxillary bone and extending superiorly to floor of orbit and zygomatic arch (Fig. 3).

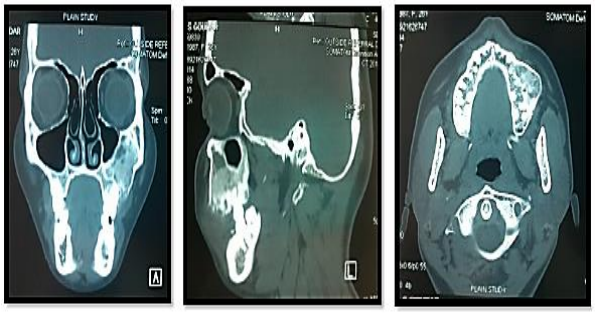


Fig. 3: CT scan reveals ground glass appearance involving the left maxilla

Case Report 2

A 23 years female patient reported with chief complaint of swelling over left mid face region since childhood. History of swelling which was initially smaller in size gradually increased to present size with no apparent changes since one year. Swelling is not associated with pain. Patient concerned of esthetic. No relevant medical and dental history. No history of functional interference because of swelling. On extraoral examination a well-defined swelling seen over the left mid face region measuring about 4×3 cm in dimension, extending superioinferiorely from infraorbital margin to the left corner of mouth, anterioposteriorly 1 cm away from the ala of the nose to the 1 cm in front of tragus. Surface appears stretched with intact skin. On palpation it was bony hard, fixed and non-tender. Intraorally vestibular obliteration seen extending from canine region to the distal to second molar. Buccal expansion was noted (Fig. 4).

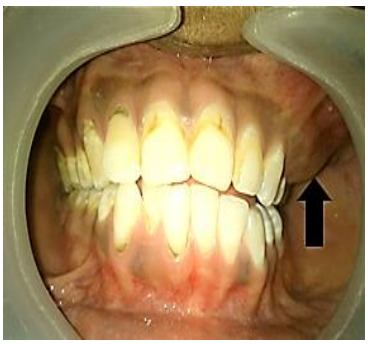
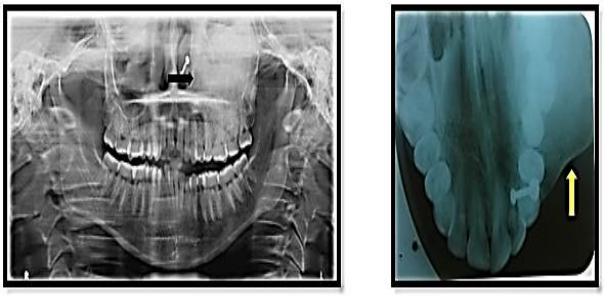


Fig. 4: Intraoral buccal expansion

OPG reveals diffuse radioopacity (ground glass homogenous appearance) in the left side of maxilla extending inferosuperiorely from the apical area of 25, 26, 27 to the floor of orbit involving left maxillary sinus. Maxillary occlusal radiographs shows ground glass radioopaque pattern and a grayish homogeneous appearance with widened maxilla (Fig. 5a, 5b).



(a) (b)

Fig. 5: (a) OPG revealing radio-opacity in Left Maxilla (b) Buccal expansion

CT shows expansile ground glass bony mass involving most of the walls of maxillary sinus, left maxillary alveolus and floor of left orbit (Fig. 6).

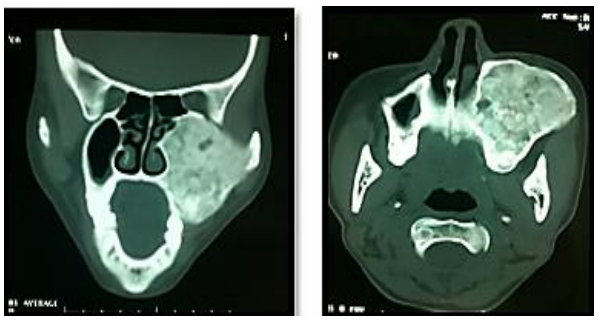


Fig. 6: CT shows expansile ground glass bony mass involving the anterior, mesial and distal walls of maxilla

Based on its distinct clinical and radiologic features, Fibrous dysplasia was given as provisional diagnosis for both cases. Differential diagnosis includes:

1. **Ossifying fibroma:** Ossifying fibroma has a definitive capsule and can be seen most of the times. Ossifying fibroma exhibits well demarcated margins whereas fibrous dysplasia does not. Ossifying fibroma grows in a centrifugal fashion producing a ball like circular lesion. The lesion enlarges equally in all directions, producing expansion of the buccal and lingual cortical plates and most notably the inferior cortex of the mandible. The expanded inferior cortex is exactly parallel to the margin of the tumour mass above. Fibrous dysplasia causes a linear expansion of the cortex thus expanded cortex cannot be in exact parallel relationship to the tumour mass.
2. **Low grade intramedullary Osteosarcoma:** Despite the marked similarities between low-grade central osteosarcoma and fibrous dysplasia, distinction may be made on the basis of lack of a reactive shell, permeative borders, denser mineralization, and more aggressive changes over time in low-grade central osteosarcoma.
3. **Hyperparathyroidism:** These also show ground glass appearance on radiology with elevated serum calcium and reduced serum phosphorous.

Blood & Biochemical investigations were done for both cases which showed alkaline phosphates, Serum Calcium & Serum Phosphorous which were with-in normal range.

Incisional biopsy was performed and decalcified H and E sections shows interconnecting bony trabeculae in a mature fibrous strand. One of the periphery shows thick compact bone, few trabeculae shows prominent basophilic reversal lines which was suggestive of Fibrous dysplasia (Fig. 7). The patients are on periodic checkups for any changes in growth formation and appearance of pain.

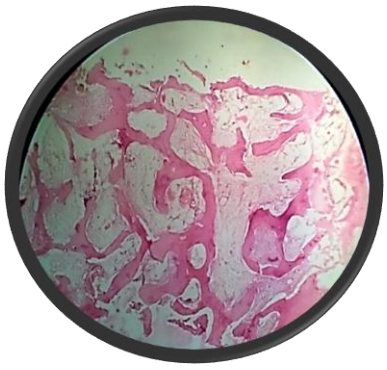


Fig. 7: Interconnecting bony trabeculae in a mature fibrous strand

Since there are no symptoms and evidence of progression surgical treatment was not indicated. Patient was advised to visit us regularly and to observe and inform for any change in growth formation and appearance of pain.

Discussion

FD is commonly benign lesion in which irregularly distributed spicules of bone lie in cellular fibrous stroma.[9,10] 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. Mutations in GNAS associated with different disorders. Gain-of-function mutations have been found in McCune-Albright syndrome (MAS), polyostotic fibrous dysplasia (PFD), monostotic fibrous dysplasia (MFD) and pituitary adenoma (PA).[10] The monostotic form is equally distributed in both genders and ethnic groups and is 6 times more common than polyostotic. Polyostotic is more frequent in females (F/M ratio 3:1). Monostotic and polyostotic are mainly diagnosed in children and young adults. Studies of FD show no sexual predilection except for Mc Cune Albright syndrome which affects females almost exclusively. FD is seen maxilla more than mandible and occurs frequently in the posterior area.[11] Both our cases are seen in posterior maxilla. Keijser et al.[12] reported the case with 13 patients after 20 years of age, the two of cases are polyostotic and the rest of cases are monostotic. In our case, the lesion is also monostotic form of FD.

In most cases, the radiographic and clinical findings are sufficient to allow the practitioner to diagnose without a biopsy.[11] The differential diagnosis with similar radiographic appearance such as ameloblastoma, ameloblastic fibroma, ameloblastic odontoma, ameloblastic fibroodontoma, cental giant cell granuloma, odontogenic cyst, ossifying fibroma, osseous dysplasia, chronic sclerosing osteomyelitis and osteosarcoma should be considered.

The density and trabecular pattern of FD lesions is variable. Early lesions may be more radiolucent than mature lesions and in rare cases may appear to have granular internal septa, giving the internal aspect a multilocular appearance. The abnormal trabeculae usually shorter, thinner, irregularly shaped and more numerous than normal trabeculae. This creates a variable radyopaque pattern, it may have a granular appearance (‘ground-glass’ appearance, resembling the small fragments of a shattered windshield), a pattern resembling the surface of an orange (peau d’orange), a wispy arrangement (cotton wool), or an amorphous, dense pattern. A distinctive characteristic is the organization of the abnormal trabeculae into a swirling pattern similar to a fingerprint.[11]

Different treatment modalities are advised based on the symptoms, extent of the lesion. For most of the monostotic cases choice of treatment is to wait and watch. Many lesions are discovered incidentally on radiographs and are asymptomatic. If the radiographic findings are characteristic of fibrous dysplasia, a biopsy is not indicated. Such lesions ordinarily pose no risk for pathologic fracture or deformity, and only clinical observation is warranted. Follow-up radiographs should

be made every six months to verify that there has been no progression. As a result of the radiolucency of fibrous dysplasia and despite the absence of histologic evidence of abnormal osteoclastic activity, bisphosphonate therapy has been utilized for patients with symptomatic polyostotic disease. Patients were treated with intravenous infusions of pamidronate over three days, with a total dose of 180 mg (60 mg/day), repeated every six months, supplemented with calcium (500 to 1500 mg/day) and vitamin D (800 to 1200 IU/ day). Each infusion was administered over a four-hour period. Surgical procedures may be required for correction of a deformity, prevention of pathologic fracture, and/or eradication of symptomatic lesions. Patient age is important because monostotic lesions remain active only until skeletal maturity, whereas polyostotic lesions may progress during adulthood.[13]

If FD is asymptomatic, it can be noticed incidentally in CT scans and radiographs. If there is no symptom or evidence of progression during follow-up, surgical treatment isn't considered.

In this two cases both clinical and radiographic features are supportive to the diagnosis of fibrous dysplasia. Histopathology is a gold standard which confirmed the diagnosis.

Conclusion

By having good knowledge of clinical and radiologic features of Fibrous dysplasia, it can easily be distinguished from others lesions of the maxilla and mandible.

References

1. Waldron CA; Fibro-osseous lesions of the jaws; Journal of Oral & Maxillofacial surgery; 51; 828, 1993.
2. Faizan Alaw. Benign Fibro-osseous Diseases of the Maxillofacial Bones. A Review and Differential Diagnosis. Am J Clin Pathol 2002;118(Suppl 1):S50-S70.
3. Waldron CA. Fibro-osseous lesions of the jaws. J oral maxillofac surg 43;248-262, 1985.
4. Chen YR, Noordhoff MS (1990) Treatment of cranio-maxillo-facial fibrous dysplasia: how early and how extensive. Plast Reconstr Surg 86: 835-842.
5. Karja J, Rasanen O (1972) Fibrous dysplasia of the jawbones: analysis of five cases. Acta Otolaryngol 74: 130-138.
6. Sassin JF, Rosenberg RN (1968) Neurological complications of fibrous dysplasia of the skull. Arch Neurol 18: 363-369.
7. Chen YR, Fairholm D (1985) Fronto-orbito-sphenoidal fibrous dysplasia. Ann Plast Surg 15: 190-203.
8. Schonder A (1977) Fibrous dysplasia of bone with proptosis. Am J Dis Child 131: 678-679.
9. Siegal GP, Bianco P, Dal Cin P (2013) Fibrous dysplasia. Pathology and genetics of tumours of soft tissue and bones. IARC, Lyon.
10. Dicaprio MR, Enneking WF (2005) Fibrous dysplasia. Pathophysiology, evaluation, and treatment. J Bone Joint Surg (Am) 87: 1848-1864.
11. White SC, Pharoah MJ (2009) Oral Radiology: Principles and Interpretation. (6thedn), Elsevier.
12. Keijser LC, Van Tienen TG, Schreuder HW, Lemmens JA, Pruszczynski M, et al. (2001) Fibrous dysplasia of bone: management and outcome of 20 cases. J Surg Oncol 76: 157-166.
13. Clinical guidelines for the management of craniofacial fibrous dysplasia. Lee JS, Fitz Gibbon EJ, Chen YR. Orphanet Journal of Rare Diseases 2012, 7(Suppl 1):S2.