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CASE REPORT

FIBROUS DYSPLASIA – A CASE REPORT

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Abstract

Fibrous dysplasia is an uncommon benign fibro-osseous lesion of unknown etiology and often affecting single bone. The case presented here has typical "Ground glass" appearance of skull and facial bones, i.e. maxilla, sphenoid, zygoma. Hence the terminology craniofacial fibrous dysplasia would be more appropriate. The histopathological report was consistent of fibrous dysplasia.

Key words- Fibrous dysplasia, Ground glass, Woven bone.

INTRODUCTION

Fibrous dysplasia is considered as hamartomatous fibroosseous lesion not of periodontal ligament origin. Though the exact etiology is unknown,[5] it is a lesion of bone that produces lysis of bone with fibrous proliferation as a replacement in its early stage. As it ages, immature bone is laid down in Chinese-character spicules.[13]

Fibrous dysplasia was first described by Lichtenstein 1938 as a disorder characterized by progressive replacement of normal bone element by fibrous tissue. [2,4,10, 18,21] Fibrous dysplasia affecting the jaws is an uncommon developmental anomaly. It may present as Monostotic (74%), Polystotic (13%) and Craniofacial (13%) varieties. [1,3,4,5,12, 1519,21] According to Scully C et al four

subgroups of fibrous dysplasia have been described: Monostotic (Jaffe-Lichtenstein syndrome), Polyostotic, Polyostotic fibrous dysplasia of Albright's syndrome, and a form confined to the craniofacial complex (craniofacial fibrous dysplasia).[17,20] Although craniofacial bone is the second common site of fibrous dysplasia involvement, it is rarely found in the para nasal sinus.[7] Among fibrous dysplasia of the head and neck, the maxilla and mandible are the most frequent sites to be involved.[8,16,17 20] Although mandibular lesions are truly monostotic, maxillary lesions often involve adjacent bone (such as the zygoma, sphenoid, and occiput) and are not strictly monostotic hence the designation of craniofacial dysplasia is appropriate.[17] Fibrous dysplasia becomes dormant in adolescence and early life which is more common in females.[7] Sometimes it occurs with precocious puberty, endocrine disorder and "café au lait" skin pigmentation termed McCune-Albright syndrome.[3,9,10,16,17,20] Craniofacial category was identified by Davis and Yardley; appear to be confined to the face and jaws involving two or more bone.[4]

Fibrous dysplasia is a sporadic condition that results from a post zygotic mutation in the GNAS 1(guanine nucleotide-binding protein, alpha stimulating activity polypeptide 1) gene. Clinically it may manifest as a localized process involving only one bone, as a condition involving multiple bones, or as multiple bone lesion in conjunction with cutaneous and endocrine abnormalities. The clinical severity of the condition presumably depends on the point in time during fetal and post-natal life that the mutation of the GNAS 1 occurs.[4,9,11,17,19] This mutation activates adenylate cyclase and consequently increases intracellular concentrations of cAMP resulting in abnormal osteoblast differentiation and production of dysplastic bone.[3,16,19]

Radiological signs of fibrous dysplasia consist mainly of radiolucent lytic and cystic lesion, with reduction of cortical thickness and sometimes widening of the diaphysis. The most frequent description used is, "Ground glass", appearance. [10,15-20] Other patterns were reported by Waldron and Giansanti as "Smoky" and "Cloudy" and by Obisesan and coworkers as "Peau d'Orange", "Whorled" or "Diffuse sclerosis".[1]

Case Report

A 11-year old boy presented with complaints of facial asymmetry with slow growing painless, bony hard swelling in the right cheek region since last 1 year. As per patient's father he had noticed the swelling 3-4 year back but as age advanced the swelling was more visible since past one year. Medical history was non contributory.

On extra oral examination single diffuse swelling present on right side of maxilla. (Fig-1)

On intra oral examination the swelling was present anteriorly at the region of right maxillary lateral incisor extending posteriorly up to the right maxillary first molar region and superiorly to the depth of vestibule, measuring 3x4.5 cms in diameter. On palpation consistency was hard and expansion of buccal cortical plate was seen (Fig-2). A working diagnosis of benign bony tumor was made. The differential diagnosis of periapical cyst, dentigerous cyst, ossifying fibroma, aneurysmal bone cyst was considered.

Intra oral periapical radiograph revealed increased density of bone with typical "Ground Glass" appearance in the region of right maxillary lateral incisor up to the right maxillary molar (Fig-3). Occlusal view shows expansion of right buccal cortical plate and typical "Ground glass" appearance of the bone. Panoramic and PNS view radiographs also revealed "Ground Glass" appreance on right maxilla (Fig-4). CT scan report revealed sclerosis with bony expansion with

few cystic changes seen involving right portion of maxillary bone nearly obliterating the right maxillary sinus. The lesion also extends into right alveolar process and right pterygoid process. Expansion of bone and typical "Ground glass" appearance involving body of sphenoid and floor of sella was seen. Obliteration of right sphenoid sinus and involvement of part of greater wings and clivus was evident. Similar involvement of right zygomatic bone is seen. There is no obvious major fissure involvement seen (Fig-5).

Serum alkaline phosphates level was 112.0U/L and serum calcium level was 9.4mg%. Microscopic picture reveals – loosely arranged, fibrous tissue with moderate vascularity containing variable amount of immature woven bone. Trabeculae are thin and irregular in size and are not lined by osteoblast (Fig-6). Focal areas show lamellar bone and few osteocytes entrapped. Few osteoclasts are also seen. Histopathological report suggested of fibrous dysplasia. Patient was referred to department oral surgery for further management.

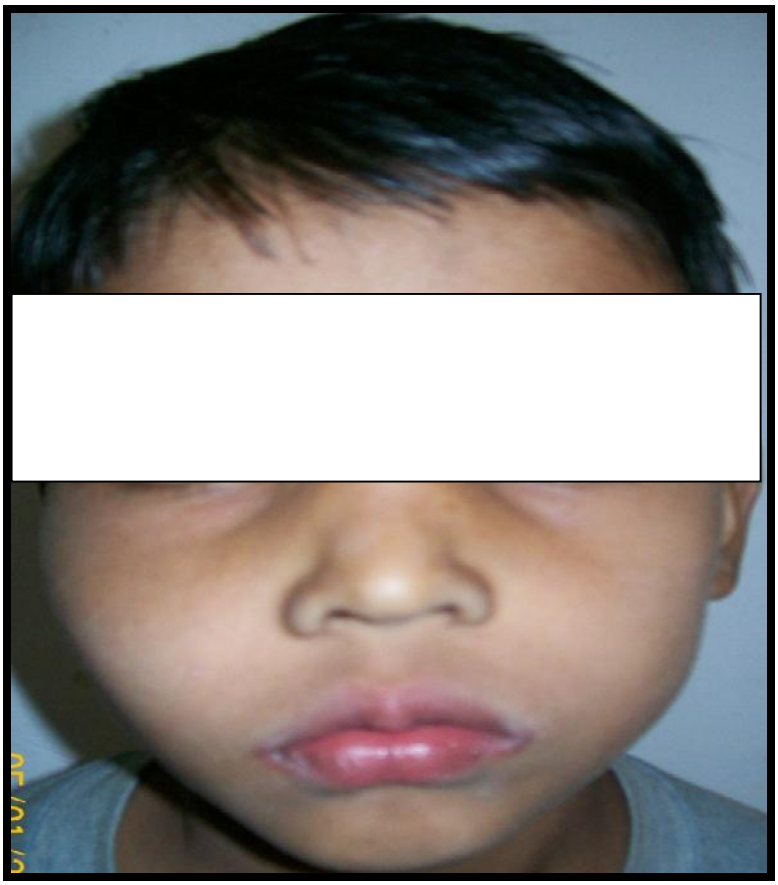


Fig 1: Diffuse swelling on right side of maxilla

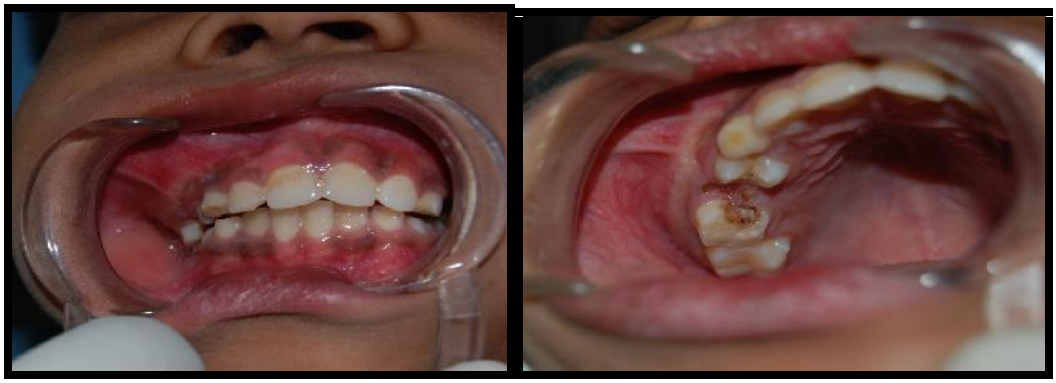


Fig 2: Expansion of buccal cortical plate at vestibular region

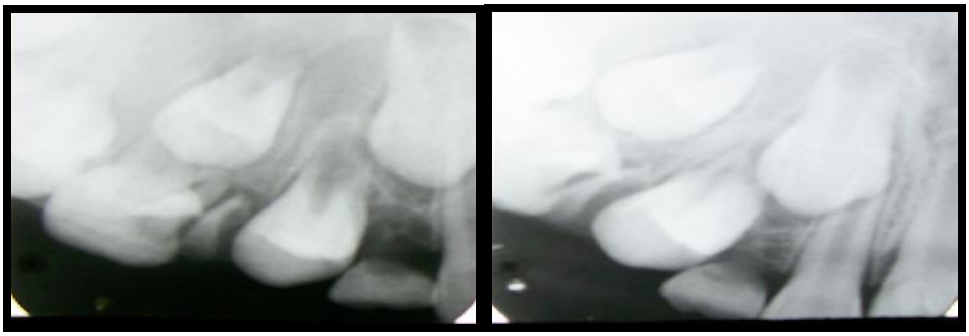


Fig 3: Periapical radiographs showing “Ground glass” appearance

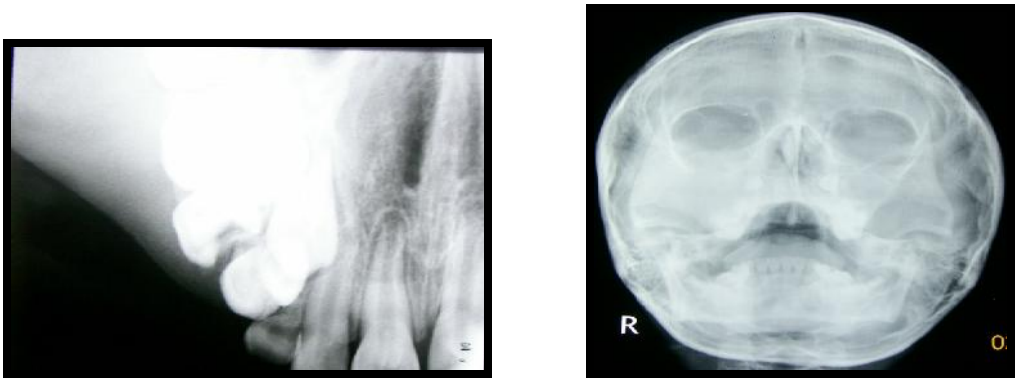


Fig 4: Occlusal & PNS views show “Ground Glass” appreance on right maxilla

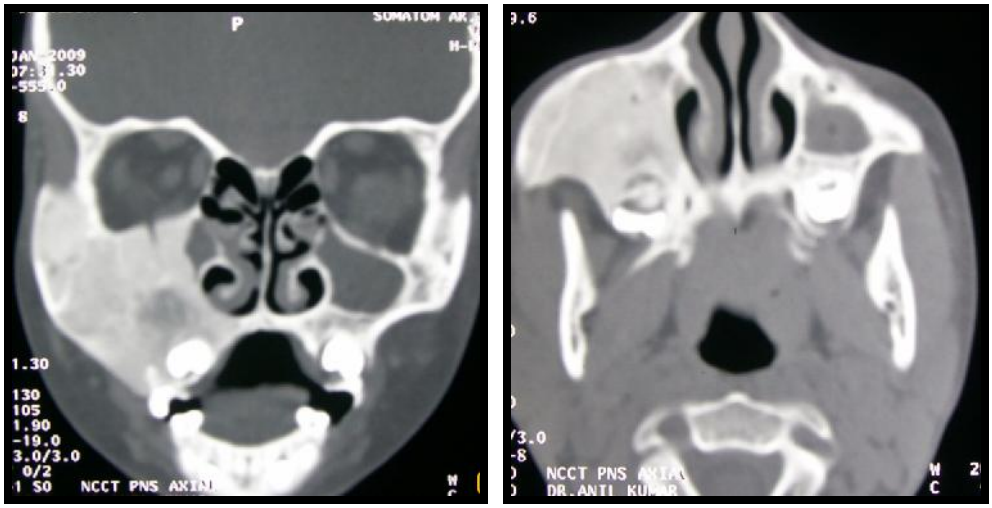
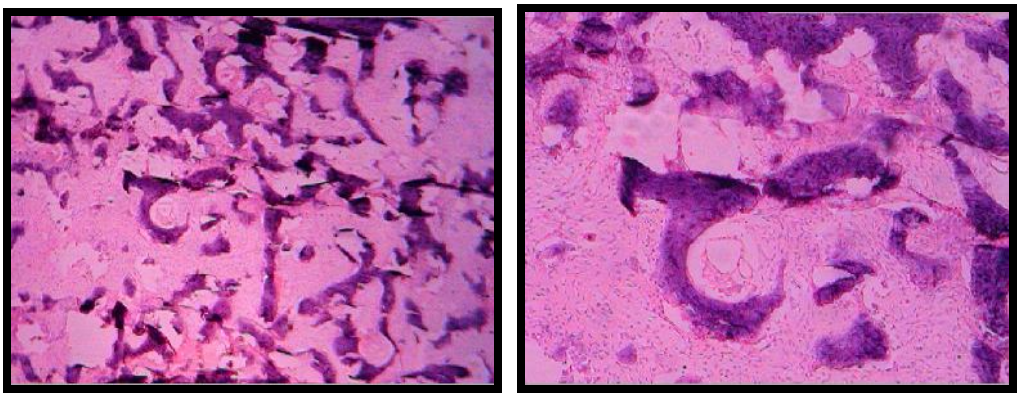


Fig 5: CT scans show sclerosis, bony expansion with few cystic changes



A-X 20 B- X 40

Fig 6: Loosely arranged fibrous tissue containing variable amount of immature woven bone (A-X 20 and B- X40)

DISCUSSION

Fibrous dysplasia is a benign pathologic condition of the bone in which fibrous tissues gradually expand and replace the normal bone. The disease usually begins in childhood and progresses throughout puberty and adolescence, and then becomes dormant in early adult life. About 75% of the cases are found under the age of 30 years. Malignant transformation is rare 0.05% and is usually seen only in polyostotic cases.[7]

Following the ribs and long bones, craniofacial bones are the second most common site of involvement and comprise 25% of the cases. Fibrous dysplasia of the para nasal sinuses is very rare.[7] Most commonly, fibrous dysplasia is asymptomatic until there is encroachment upon adjacent vital structures. Facial asymmetry is the most common sign of fibrous dysplasia in the head and neck, followed by pain, ocular proptosis and neurological changes.[7]

The lesions of fibrous dysplasia are twice as common in maxilla as compared to mandible, and the posterior aspects of the jaw are more frequently affected than the anterior.[6]

The craniofacial form of fibrous dysplasia can be diffuse and may involve multiple bones. When the anatomic spaces and foramina are constricted because of encroachment of the lesions, the patient may experience a variety of symptoms, including headaches, loss of vision, proptosis, diplopia, loss of hearing, anosmia, nasal obstruction, epistaxis, epiphora and symptoms mimicking sinusitis.[4]

The case described here is the patient with the swelling present on right side of maxilla since 3-4 years and more visible since 1 year. On intra oral examination, the swelling was present anteriorly at the region of right maxillary lateral incisor extending posteriorly up to the right maxillary first molar region and expansion of buccal cortical plate was seen. In differential diagnosis, periapical cyst was considered because of carious primary first molar, and dentigerous cyst was considered because of missing maxillary canine. Ossifying fibroma was considered because of swelling extending from right lateral incisor up to first molar region. All the above listed differential diagnosis was eliminated once the ground glass appearance of the bone was noticed on the radiograph. Aneurysmal bone cyst was excluded because it was more common in mandible.

The radiographic appearance revealed increased radiopacity of bone with “Ground glass” appreance in the region of right maxilla. CT scan revealed sclerosis with bony expansion with few cystic changes. Also, it revealed typical “Ground glass” appearance involving multiple bones of the cranium. The trabeculae pattern appeared thin and irregular in size. After the final diagnosis patient was referred to department of Oral Maxillofacial surgery. The surgeons were of the opinion that they would do the surgery, only after completion of patient’s growth spurt. Therefore, patient was advised periodic recall visits at regular intervals to record the bony changes and involvement of other vital structures.

Surgical treatment of fibrous dysplasia consists of either conservative shaving/ contouring or radical excision with immediate reconstruction. The choice of surgical option depends on several factors like site of involvement, rate of growth, aesthetic disturbance, functional disruption, and patient preference, general health of the patient, surgeon’s experience and the availability of a multi-disciplinary team (neurosurgeon, ophthalmologist, orthodontist and endocrinologist). The multi-disciplinary approach becomes even more important in polyostotic fibrous dysplasia.[4]

Oral aluminum acetate can be used to reduce the danger of hyperphosphataemia in severe forms. Steroids have been used with partial success in treating painful lesions. Radiotherapy is contraindicated and has been associated with sarcomatous change.[1]

Surgical treatment for cranio-maxillo-facial lesions is controversial. Excision of all the affected bone is usually fruitless since it is impossible to be sure of the limits of the disease.[1]

Chapurlat et al proposed treatment with the biophosphonate pamidronate without surgical intervention. They noticed that severe bone pain and the number of painful site appeared to be significantly reduced and all biochemical markers of bone remodeling were substantially lowered.[1]

CONCLUSION

Fibrous dysplasia is an uncommon benign neoplasm of bone most commonly occurring in jaws. Primarily, clinician should be able to differentiate from other bony lesions. Surgery should be postponed until the growth spurt is completed. Hence patient should be observed till the surgery is done and regular periodic follow up should be done. Most commonly conservative treatment like shaping and recontouring is preferred. Radical excision and reconstruction is also a choice of surgical management.

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