Case Report

Fibrous dysplasia a report of two cases with emphasis on radiographic features

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ABSTRACT

Fibrous dysplasia is also called fibrocystic disease, osteitis fibrosa localisata, focal osteitis fibrosa, fibro osteodystrophy, and Jaffe-Lichtenstein. It is an asymptomatic regional alteration of bone in which the normal architecture of bone is replaced by fibrous tissue and nonfunctional trabeculae-like osseous structures. Fibrous dysplasia may be monostotic or polyostotic, with or without associated endocrine disturbances. Here, we are discussing two cases with monostotic fibrous dysplasia of maxilla with emphasis on various radiographic features that the lesion shows during its different stages.

Key words: Fibrous dysplasia, maxilla, radiographic features, swelling

INTRODUCTION

Fibrous dysplasia was first described by Lichtenstein in 1938 as a disorder characterized by progressive replacement of normal bone elements by fibrous tissue.[1-3] It is although benign, has the potential to cause significant cosmetic and functional disturbance, particularly in the craniofacial skeleton. Incidence ranges from 1:4000–1:10,000 and represents approximately 2.5% of all bone lesions and about 7% of all benign bone tumors.[4-6] Fibrous dysplasia is a sporadic benign skeletal disorder that can affect single bone (monostotic form) or multiple bones (polyostotic form) and if combined with precocious puberty, endocrine disorders, and café au lait skin pigmentation termed McCune - Albright syndrome. Fibrous dysplasia occurs with equal gender prevalence with monostotic form more commonly found in young age group, i.e. 20–30 years, polyostotic form mainly occurs in children younger than 10 years of age. The ratio of monostotic to polyostotic is 7:3.[7-9] Most cases of craniofacial fibrous dysplasia cannot be truly categorized as monostotic because of the involvement of multiple adjacent bones of the craniofacial skeleton. They are also not truly polyostotic because bones outside the craniofacial complex are spared. These conditions have female predilection and mostly seen in the first 3 decades of life and usually stabilize until skeletal maturity.[6]

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CASE REPORTS

Case 1

A 30-year-old female patient reported with a chief complaint of swelling on the right side of the face since 3 years. Earlier swelling was smaller in size and gradually progressed to present size. History of present illness revealed no incidence of trauma, pain epistaxis, loosening of teeth, tismus, diminished vision, no other swelling in the body, and gave no history of systemic diseases and drug allergies. The patient seeks doctor's advice due to facial disfigurement. Extraoral examination revealed that a diffused swelling was present on the right side of the face involving right maxilla, zygoma, temporal region, and

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mandibular region. The skin over the swelling was normal. On palpation, all relevant inspectory findings confirmed that swelling was bony-hard, smooth and nontender on palplation, and skin was not fixed to the underlying bone, but stretched and smooth. Introrally on inspection, an expansion of the buccal cortical plate was noticed extending from first premolar region to molar area on the right side of maxilla [Figure 1]. On palpation, all relevant inspectory findings were confirmed. Swelling was nontender, smooth, and bony-hard in consistency. Based on clinical examination, a provisional diagnosis of fibroseous lesion of the maxilla was made. Radiographic examination which includes orthopantomogram (OPG), para nasal sinus, and posteroanterior view revealed diffuse irregular, thickened, trabecular pattern resembling orange peel appearance in the region of maxilla [Figure 2]. Fibrous dysplasia of right maxilla was considered as final diagnosis. As patient has no sign and symptoms except swelling on the right side of the face, a surgical recontouring was done. The patient was under follow-up for 18 months and no reoccurrence was noticed.

Case 2

A 42-year-old male patient came with a chief complaint of swelling on the left side of the face for 5 years. Earlier swelling was smaller in size and gradually progressed to present size. History of present illness revealed no incidence of trauma, pain epistaxis, loosening of teeth, tismus, diminished vision, no other swelling in the body, and gave no history of systemic diseases and drug allergies. Extraoral examination revealed that a diffused swelling was present on the right side of the face involving right maxilla, zygoma, temporal region, and mandibular region. The skin over the swelling was normal and stretched. On palpation, all relevant inspectory findings confirmed that swelling was bony-hard, smooth, and nontender on palplation. Introrally on inspection, an expansion of the buccal cortical plate was noticed extending from first premolar region to molar area on the right side of maxilla. On palpation, all relevant inspectory findings were confirmed. Swelling was nontender, smooth, and bony-hard in consistency. There was no tenderness during palpation of the swelling. Based on clinical examination, a provisional diagnosis of fibroseous lesion of the maxilla was made. Radiographic examination which includes OPG view revealed a ground glass appearance of the right side of the maxilla. An incisional biopsy was done which revealed highly cellular connective tissue stroma resembling fibroblasts that are spindle-shaped, plump, and benign along with some osteoid-like tissue with osteocytes, but mostly not lined by osteoblasts in various shapes and extravasated red blood cells also seen [Figure 3]. Based on radiographic and histological finding, a diagnosis of fibrous dysplasia of the right maxilla was made. Surgical excision with reconstruction was done and sent for histological examination which revealed fibrous dysplasia. The patient was under follow-up for 1 year and no reoccurrence was noticed.

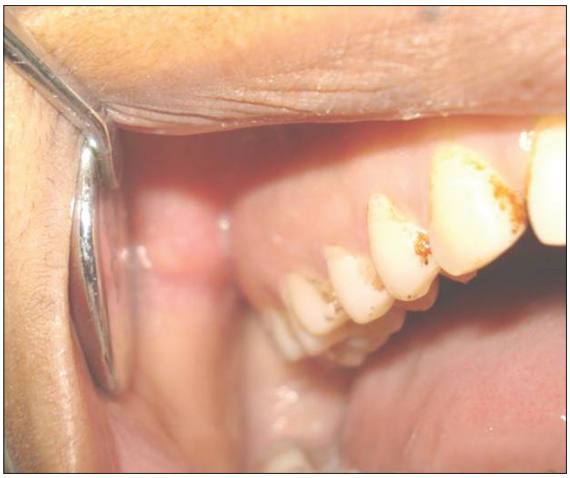

Figure 1: Expansion of buccal cortical plate

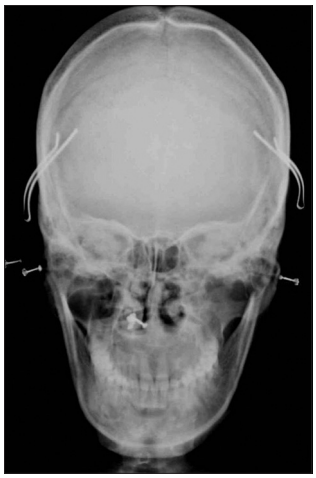

Figure 2: Irregular thickened trabeculae pattern

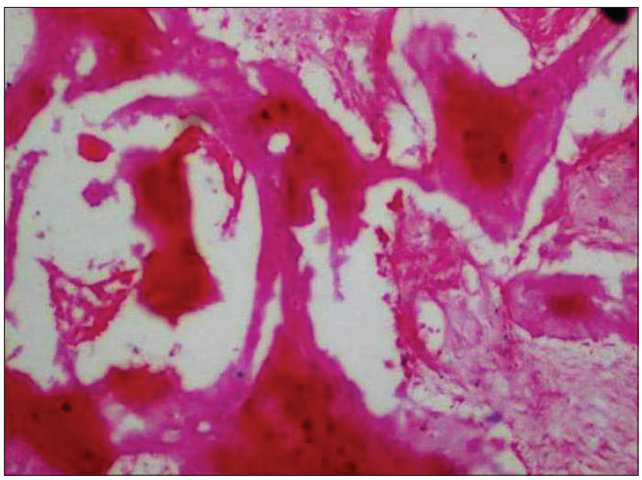

Figure 3: Histological section of fibrous dysplasia

DISCUSSION

Fibrous dysplasia is a hamartomatous disorder of bone metabolism which can cause facial disfigurement of the face as in our cases. Eversole defines fibrous dysplasia of craniofacial bones as "a benign, nonneoplastic intramedullary cellular proliferation of fibroblasts, with formation of irregular trabeculae of bone or ovoid calcifications that shows indistinct, nonencapsulated borders."[10]

The etiology of fibrous dysplasia is genetic. It is noninherited condition caused by mutation in the gene GNAS on chromosome 20. This mutation encodes the alpha subunit of the stimulatory G protein – coupled receptor, GSa. These activating mutations occur post zygomatically which leads to the replacement of the arginine residue amino acid with either a cysteine or a histidine amino acid. This mutation selectively inhibits GTPase activity, which results in constitutive stimulation of AMP-protein kinase intracellular signal transduction pathways.[11,12]

This mutated protein form coupled receptor complex which is autonomous in function, cause changes in bone through parathyroid hormone receptor, and changes in skin occur through melanocyte stimulating hormone receptor. Changes can occur even in ovaries through the follicle stimulating hormone receptor and in thyroid and the pituitary gland, through the thyroid and growth hormone receptors, respectively.[11,12]

Clinical features depend upon the number of the affected bones, presence or absence of extraskeletal abnormalities, i.e. the severity of the mutation of the genes. Mostly, it leads to slow progressive enlargement of the affected jaws which is usually painless and typically presents as a unilateral swelling of the face as in our both the cases. Expansion in buccal cortical plate is more common as compared to lingual or palatal aspect similar to the finding reported in our case. In the later stage, it may lead to displacement of teeth leading to malocclusion and disruption in the normal eruption pattern. The mucosa is almost invariably intact over the lesion similar to our cases.[13,14]

Radiographic features

Jaw features can be radiolucent, radiopaque, and mixed. The early lesions are radiolucent with ill-defined borders which blend into the surrounding bone. These defects are often unilocular, but the presence of occasional bony septa may give a multilocular impression. As the lesion matures, a mixed radiolucent and radiopaque lesion appears, depending on the stage of maturity and distribution of fibrous and osseous tissue, the radiolucency and radiopacity of the lesion vary. The new bone forms which appear radiographically a very small radiopacities of poor density and appear granular as they enlarge. The variation is more pronounced in the mandible and more homogenous in the maxilla. The internal density is more opaque in the maxilla and base of the skull as compared to the mandible.

Obisesan *et al.*'s classification (1) Peau'd orange or orange peel; in this type, there are alternating areas of granular density and radiolucency giving a radiographic appearance resembling the peel of an orange. (2) Whorled plaque-like pattern; in this type, the matrix of the well-circumscribed lesion is composed of plaques of the amorphous material of intermediate radiodensity, which on close examination are seen to be arranged in whorled finger print appearance. (3) Diffuse sclerotic type; the lesions of this type are seen as homogenous dense areas, which gradually merge with normal bone. (4) Cyst-like type; in this type, the lesion is radiolucent, unilocular or more often, multilocular with well-defined margins. (5) Pagetoid type; in this type of lesion, the affected area of bone markedly expands and shows alternating areas of opacity and lucency, as seen in Paget's disease. (6) Chalky type; it manifests as a well-circumscribed lesion consisting of an amorphous dense radiopaque material.[15]

Other features include expansion of the involved bone. Eruption pattern of the teeth is disturbed because of the loss of the normal support of developing teeth. Due to its expansile nature, it will cause malalignment, tipping displacement of teeth. When there is an extension into the antrum, teeth may be displaced into the sinus along with the fibro-osseous tissue. Root resorption is not a usual feature of fibrous dysplasia.[15]

Based on histopathology alone, it is impossible to reach the final diagnosis of fibrous dysplasia. Along with histopathological examination, radiographic examination is necessary as we have done in both the cases.

Surgical treatment of fibrous dysplasia consists of shaving/contouring or radical excision with reconstruction. Surgical treatment depends upon the number of bone involved, site of involvement, growth of tumor, functional involvement, and amount of destruction caused by the lesion.

CONCLUSION

FD is a benign disease that has the potential to cause significant cosmetic and functional disturbance. With proper understanding in early diagnosis and proper management by well-qualified team, fibrous dysplasia can be detected and managed properly at early stages without

causing cosmetic disturbances and to prevent further complications. A regular follow up is needed for esthetic and functional purposes.

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Conflicts of interest
There are no conflicts of interest.

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