Fibrous dysplasia is a condition in which normal medullary bone is gradually replaced by an abnormal fibrous connective tissue proliferation and there is arrest of bone development in the woven stage with failure to mature to lamellar bone. The resultant fibrosseous tissue is poorly formed and structurally inadequate and weaker than the original bone. Various treatment modalities including medicinal and surgical treatment have been proposed for this lesion, in this case series we share our experience with paring down for such lesion and found it effective and efficient method of treating fibrous dysplasia. Facial symmetry, esthetics, and function were achieved in all the cases.

Keywords: Bone recontouring procedure, Fibrous dysplasia, Paring down, Surgical treatment

INTRODUCTION

Fibrous dysplasia is a non-neoplastic, developmental and hamartomatous disease of bone. The term “fibrous dysplasia” was coined by Lichtenstein as a designation for multiple fibro-osseous lesions. The term “monostotic and polyostotic” have been applied to those forms of the disease in which respectively one or more bones are involved. It is a condition in which normal bone is gradually replaced by an abnormal fibrous connective tissue proliferation. This disease produces solitary or multifocal lesions in which there is arrest of bone development in the woven stage with failure to mature to lamellar bone. The resultant fibrosseous tissue is poorly formed and structurally inadequate and weaker than the original bone. It usually clinically presents as the localized bulging of the either of or both jaws depending on its type with overlying mucosa and skin almost invariably intact over the lesion. Despite of facial disfigurement and disocclusion it also causes neurological and opthalmic complications depending upon its size and location.

Surgical treatment of fibrous dysplasia of jaws includes radical curetage, which is indicated in osteolytic type lesions, however, in more mature or solid type lesions surgical shaving or recontouring is indicated. This report describes four different intraosseous lesions of the maxilla referred to the authors between 1996 and 2013 all of which were treated by paring down technique and 1-year follow-up.

CASE REPORTS

Case 1
A 18 year female patient with complain of painless swelling on right maxilla, on examination swelling was extending from 13 to 18 region (Figure 1a). Computed tomography (CT) reveals gross osseous thickening, sclerosis and new bone formation along the anterior wall of right maxilla extending to the zygomatic crest posterolaterally with encroachment of maxillary antrum (Figure 1b). Paring down procedure was carried out and no reoccurrence was found on 1-year follow-up (Figure 1c).

Case 2
A 16-year-old male patient with complain of painless swelling of right maxilla which was intraorally extending from 11 to 17 region obliterating the labial sulcus (Figure 2a). CT scan reveals gross osseous thickening, sclerosis, and new bone formation along the anterior wall of right maxilla (Figure 2b). Surgical recontouring by paring-
down procedure was done. 1-year follow-up showed no reoccurrence (Figure 2c).

**Case 3**
A 20-year-old female patient with complain of swelling of right maxilla since 6 months, intraorally swelling was extending from 14 to 17 region obliterating the labial sulcus (Figure 3a). CT reveals mixed radiolucent with radio-opaque mass on the right side maxilla with expansion of the labial cortex (Figure 3b). Intra-oral biopsy was performed and reported as fibrous dysplasia (Figure 3c). Surgical recontouring by paring-down procedure done.

**Case 4**
A 24-year-old female patient with complain of painless swelling on left maxilla (Figure 4a). On examination, there was painless swelling intraorally extending from 2 to 27 region, obliterating the labial sulcus. Posterior nasal spine view skull shows radio-opaque mass in the left maxilla with the involvement of maxillary antrum, the lesion was blending with the adjacent tissue (Figure 4b). Surgical recontouring by paring-down procedure was done and the patient was followed up after 1-year without any complications (Figure 4c).

**DISCUSSION**
Fibrous dysplasia of maxilla is a serious form of the disease since it has marked predilection for occurrence in children and almost impossible to eradicate without radical or mutilating surgery. Since these are the lesions not well circumscribed, they extend locally to involve the maxillary sinus, zygomatic process, and floor of the orbit even to the extent of the base of the skull. Severe malocclusion and bulging of the canine fossa or extreme prominence of the zygomatic process producing a marked facial deformity are a typical squeal of cervicofacial type of fibrous dysplasia. The etiology of fibrous dysplasia
is unknown and a variety of possible factors has been suggested and none has found general acceptance. Early investigators suggested that it was caused by aberrant activity in the bone forming mesenchymal tissue. Today it is understood that all types of fibrous dysplasia resulted from a defect in bone maturation that begins in the embryo. At certain time in the histodifferentiation phase of embryo, a genetic mutation or deletion occurs in the gene that encodes for intra-cytoplasmic transducer protein required for bone maturation. Consequently, all the daughter cells of the original aberrant cell will lack these signal transducers and, therefore, a certain population of cells in the individual will be able to produce only fibrous dysplastic bone rather than mature bone. Depending on the time of mutation, the type will be decided. There is clinical evidence, which indicates that local infection and trauma may eventuate in this disease under certain conditions. Some even insist that fibrous dysplasia is a peculiar reparative reaction on the part of the bone to any one variety of injuries. Definitive diagnosis depends on the patient history, blood findings, clinical and radiological examination, and the biopsy.

Fibrous dysplasia of jaws occurs with an apparently equal predilection for males and females. It is more common in children and young adults than in older persons. Fibrous dysplasia is a lesion of slow growing and self-limiting, but growth spurts are not unusual. The lesion may regress after becoming stationary during the fourth decade of life and malignant transformation is rare (<1%). The first clinical sign of the disease is the painless swelling or bulging of the jaws. The bulging usually involves the labial or buccal plate seldom the lingual or palatal aspect. There appears some mal-alignment, tipping or displacement of the teeth due to the progressive expansile nature of the lesion. The mucosa is almost invariably intact over the lesion.

The radiological appearance of fibrous dysplasia of the jaws is extremely variable. Type I is unilocular or multilocular radiolucency with a rather well-circumscribed border containing a network of fine trabeculae. Type II is similar to the first one except increased trabeculations which renders the lesion more radio-opaque and typically mottled appearance. Type III is quite opaque with many delicate trabeculae giving a “ground glass or Peau d’orange” appearance to the lesion.

Macroscopically the tissue is firm and gritty and easily cuts through with a scalpel. Cut surface is flat and non-compressible and on palpation sensation is similar to that of rubbing fine grade of sand paper. There are considerable microscopic variations in cases of fibrous dysplasia of jaws. The lesion is essentially a fibrous one made up of proliferating fibroblasts in a compact stroma of interlacing collagen fibers. Irregular trabeculae of bone are scattered through the lesion with no definite pattern of arrangement, some of the trabeculae are C-shaped or Chinese character shaped and these trabeculae are of usually coarse woven bone and will mature over period of time and lesional tissue may show lamellar bone. Ossifying fibroma and fibrous dysplasia of the jaws often show similar histological and radiological features making the distinction between the two a diagnostic dilemma. However, they can be differentiated in the precise composition of bone matrix as shown by osteocalcin immunohistochemistry.

The whole process of activity, quiescence, reactivation, and extension of fibrous dysplasia is not understood and therefore the extent of external deformity is unpredictable. Hence, it is clear that surgical management of fibrous dysplasia awaiting the onset of puberty or cessation of growth is not necessarily logical. There is also no evidence that the surgical intervention itself intensifies the process. Samman et al. claim that the indication for surgery for fibrous dysplasia depends upon the degree of deformity and functional disturbance. They further mention that radical approach is unnecessary because of the following reasons:

1. The condition is developmental and benign
2. The lesion merges imperceptibly into the normal bone and the margins cannot be demarcated at surgery
3. Radical excision in the face and the skull is by necessity more destructive than elsewhere in the skeleton.

It is suggested to adopt a conservative approach to the lesion consisting of surgical recontouring, but additionally
osteotomies are performed to correct the dental and skeletal relationships as appropriate. A transoral approach has been advocated in reports of less involved recontouring procedures as in our cases reported.

A very rare case of simultaneous occurrence of facial fibrous dysplasia and ameloblastoma was reported. Surgical shaving was done for the treatment of fibrous dysplasia of maxilla and surgical mandibulectomy and reconstruction of the mandible was done for the mandibular ameloblastoma. No recurrence was reported in both the jaws. Stephen et al. reported a case of polyostotic fibrous dysplasia in which patient showed facial asymmetry and canting of the mandible. Le-fort osteotomy and bilateral sub-condylar osteotomies were performed to achieve the functional as well as esthetic result. Mizuno et al. described a case of facial fibrous dysplasia which involved the left frontal, temporal, malar, and mandibular region. Conservative recontouring was done several times when the patient was 16-23 years of age. The lesion begins in childhood and some authors believe that it does not progress after adolescence. There are others who reported that progression does not stop at puberty. In other words, long-term follow-up is mandatory. In the younger patients, operative procedure should be delayed until the patient reaches the maturity and the growth becomes stabilized. At maturity, the bone can be pared or shaved to produce an acceptable facial appearance.

The treatment of choice for fibro-osseous lesions is almost always surgical. Ma et al. found 31.8% recurrence rate in patients who received conservative bone contouring. Radiotherapy is contraindicated because of the possibility of subsequent development of radiation-induced sarcomas. The bone involved in the young patients is highly vascularized. It is important to achieve complete hemostasis before wound closure or hematoma may occur. The older patients with fibrous dysplasia have sclerotic bone lesions and they are relatively avascular and more liable to post-operative infection. Chronic low-grade osteomyelitis may also occur post-operatively in such patients.

Mercin et al. undertook a study of six children with progressive fibrous dysplasia located in the mandible (3), maxilla (2) and cranial base (1). All the patients received pamidronate infusion 1 mg/kg intravenously for 3 days every 4-6 months. Pain relief was achieved in all the cases, decrease in swelling in three cases and stabilization in other three cases. The local bone density increased and no further spread of the disease occurred. The only side effect was an increase in the body temperature up to 38-40°C. It was concluded that pamidronate appears to be an effective and well-tolerated therapeutic option for the patients with fibrous dysplasia. However, limited non-mutilating surgical intervention is needed in most of the cases.

CONCLUSION

In spite of many research and publications, fibrous dysplasia remains as an entity with no definite etiology, clinical manifestations, radiological appearances as well as treatment modalities. The four cases demonstrate the feasibility of surgical recontouring or paring down procedures as the choice of treatment for facial fibrous dysplasia. The lesion should be evaluated periodically both clinically and radiographically for growth and histologic changes. Imaging follow-up is of utmost importance in patients with fibrous dysplasia. CT remains the “gold standard” imaging modality for diagnosis of fibrous dysplasia and allows early visualization and sarcomatous degeneration. If the lesion is not extensive, transoral, and conservative approach is suggested as carried out in the above cases. Though pamidronate has been advised as the medical management of fibrous dysplasia, the efficiency of the treatment is not definite as further investigations and research are needed in this regard.

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