

Fibrous Dysplasia: A Case Report

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ABSTRACT

Fibrous dysplasia (FD) is a rare bony disorder in which normal bone is replaced by abnormal fibro-osseous tissue. In general, FD is found in teenagers, and it usually becomes static after adulthood. FD involves the maxilla almost 2 times more often than the mandible. It frequently appears in the posterior region of the jaw bone and is usually unilateral. Here, we present a case of asymptomatic FD affecting the right maxilla in a 14-year-old female with the clinical, radiographical, and histopathological features, and the treatment procedure involved to recontour the asymmetry of face.

Key words: Fibrous dysplasia, ground glass appearance, maxilla

INTRODUCTION

Fibrous dysplasia (FD) is a benign fibro-osseous pathologic condition characterized by the replacement of bone with fibrous tissue. This disorder was first discovered by Lichtenstein in 1938, who later collaborated with Jaffe in 1942 to first describe the condition in the medical literature.^[1] The replaced bone showed the trabeculae as shorter, thinner, irregularly shaped, and more numerous. It was determined that the abnormal appearance of the bone corresponds to increased weakness and expansion that may further lead to bone deformation and possible fractures. This disorder is caused by a GNAS 1 point mutation on chromosome 20.^[2] These changes inhibit GTPase activity that is normally required to deactivate the G protein, thereby altering signal transduction pathways. The increased amount of cAMP in bone stromal cells leads to unregulated proliferation and differentiation.^[3] It is a sporadic benign skeletal disorder that can affect one bone (monostotic form) or multiple bones (polyostotic form), and the latter may form part of the McCune-Albright syndrome (MAS) or of the Jaffe-Lichtenstein


syndrome (JLS). JLS is characterized by polyostotic FD and café-au-lait pigmented skin lesions, while MAS has additional features of hyperfunctional endocrinopathies manifesting as precocious puberty, hyperthyroidism, or acromegaly.^[4]

The diagnosis of FD is based on physical, radiological, and histopathological examination. There are different treatment approaches, including observation, medical treatment, and surgical treatment.^[5] Treatment modalities differ based on the age and clinical behavior of the neoplasm. Surgical interventions may be difficult as they are more likely to be associated with important anatomical structures. Follow-up plays a major role, if recontouring is done, as the lesions are more likely to re-occur over time.

The present article presents a case of FD of right maxilla in a 14-year-old female patient.

CASE REPORT

A 14-year-old female patient reported to the Department of Oral and Maxillofacial Surgery with the chief complaint of painless bony swelling in the upper right back teeth region for 4 years. The lesion was gradual in onset with intermittent growth patterns and attained to the present size. No known family history was revealed by the patient. There was no history of trauma, paresthesia, and difficulty in chewing food, and it was not associated with any other symptoms like pain. On extraoral examination, gross facial asymmetry was noticed on the right side of the face [Figure 1]. A diffuse swelling

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(3–4 cm approximately) was seen, superoinferiorly starting from 2 cm below the infraorbital margin to the line joining the commissure and the ear lobe, and mediolaterally starting from the right side of nasal septum, causing obliteration of right nasolabial fold, to ramus of the mandible on the right side. On palpation, the lesion was bony hard and non-tender in nature. Intraorally, the swelling extended from the mesial aspect of the upper right first premolar to the distal aspect of the upper right second molar causing obliteration of vestibule and expansion of buccal cortical plate. The overlying mucosa was smooth, and it was bony hard and nontender. No



Figure 1: Extraoral photograph showing facial asymmetry on the right side



Figure 2: Intraoral photograph showing swelling extending from mesial aspect of upper right first premolar to distal aspect of the upper right second molar causing obliteration of vestibule and expansion of buccal cortical plate

surface erosions or ulcerations noticed [Figure 2]. Retained 55 and palatally erupting 15 were observed. Teeth in the vicinity of the lesion were vital.

Orthopantomogram findings revealed homogeneous granular radio-opacity causing obliteration of the right maxillary sinus, blending with adjacent bone. No resorption or displacement of the teeth involved was observed [Figure 3]. The computed tomography of the face sinuses showed a hyperdense, heterogeneous, expansive mass of uneven contour, with aspect of an “opaque glass” involving the maxilla, zygomatic bone, and right maxillary sinus [Figure 4].

A provisional diagnosis of FD of the right maxillary region was made. Differential diagnoses



Figure 3: Orthopantomography reveals homogeneous granular radio-opacity causing obliteration of the right maxillary sinus blending with adjacent bone



Figure 4: Computed tomography (coronal view) shows hyperdense, heterogeneous, expansive mass of uneven contour, with aspect of an “opaque glass” involving the maxilla, zygomatic bone, and right maxillary sinus

of other fibro-osseous lesions such as ossifying fibroma and cemento-osseous dysplasia were considered. Taking into consideration of the age of the patient and to retain, the teeth involved surgical recontouring and long-term follow-up was planned. Routine investigations such as hemogram, serum calcium, and serum alkaline phosphatase (ALP) were performed. All parameters were within normal limits except ALP. ALP was raised to 764 U/L.

The presented case was done under general anesthesia. Markings were placed for lip split incision, which was extending from the labial vestibule intraorally crossing the vermilion border up to the right nasolabial fold and the flap was reflected. Hard bony mass was noted over the right maxillary buttress region [Figure 5]. Corticotomy markings were made with 701 bur over maxillary buttress region. Recontouring of bony mass was done with chisel and mallet. Smoothing of the peripheral bone was done with tungsten carbide bur. The bony bits were sent for histopathological examination [Figure 6], and closure was done [Figure 7].

Histopathological examination findings showed the presence of connective tissue stroma with numerous bony trabeculae of varying shapes and sizes. They also showed evidence of osteocytes within them and prominent osteoblastic rimming. The connective tissue is fibro cellular and shows the presence of few endothelial lined blood vessels. Based on the clinical, radiographical and histological features, a final diagnosis of FD of right maxilla was given, and the patient was advised for long-term follow-up. One month follow-up of the patient showed satisfactory healing [Figure 8].

DISCUSSION

FD is a fibro-osseous lesion characterized by developmental hamartoma by blending of fibrous and osseous tissue, with resultant secondary bony metaplasia, without osteoblast maturation producing immature, newly formed, and weakly calcified bone. It is caused due to unknown etiology.^[6] It can be monostotic or polyostotic. The craniofacial bones are affected in 10–25% of cases in monostotic forms and in 50% of cases in polyostotic forms.^[7] The maxilla is more commonly involved than the mandible in Monostotic forms. FD essentially affects children and young adult, such as in the present case, a 14-year-old female. In most cases, the radiographic and clinical findings are sufficient



Figure 5: Reflection of the flap and exposure of bony mass

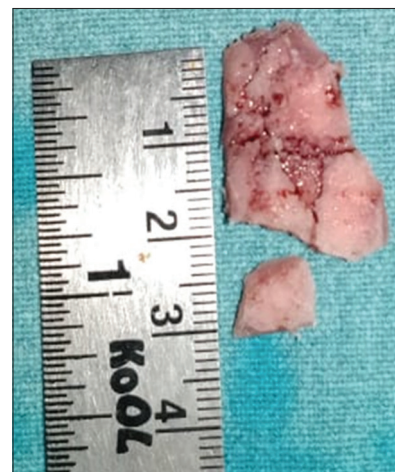


Figure 6: Bony specimen sent for histopathological examination



Figure 7: Closure was done



Figure 8: Extraoral photograph of the patient after 1 month follow-up shows satisfactory healing

to allow the practitioner to diagnose without a biopsy.^[8] The differential diagnosis with similar radiographic appearances, such as ameloblastoma, ameloblastic fibroma, ameloblastic odontoma, ameloblastic fibro-odontoma, central giant cell granuloma, odontogenic cyst, ossifying fibroma, osseous dysplasia, chronic sclerosing osteomyelitis, and osteosarcoma, should be considered.^[9] In the present case, there was no compelling indication to seek a biopsy, any sudden change in the clinical presentation or behavior of the lesion, which might warrant further investigation.

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 are usually shorter, thinner, irregularly shaped, and more numerous than normal trabeculae. This creates a variable radiopaque pattern, it may have a granular appearance (“ground-glass” appearance, and 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.^[8] Prapayastok *et al.* reported a case which was seen a rare radiographic “sun ray” appearance in 19-year-old woman.^[10] In the present case, the computed tomography revealed a “ground glass” appearance of the affected area.

FD is a rare but severe bone disease which may cause fractures in long bones, deformities, and bone pain. Although most lesions appear to stabilize when approaching bone maturity, some cases can reach severe asymmetry, visual impairment, diplopia, pain, paresthesia, proptosis, hearing loss, anosmia, nasal obstruction, epistaxis, and epiphora. The patients generally complain of swelling (94%) and pain (15%).^[11] In the present case, the patient chief complaint was bony swelling over the right cheek region, which caused the facial asymmetry.

The concentration of serum ALP may be an important marker for the detection of the recurrence of the lesion. The patients who had FD, have higher ALP; this may be a reliable marker for estimating tumor progress, and a sudden rise in ALP was correlated with the regrowth of FD by Park *et al.*^[12] This was observed in the present case with raised ALP levels.

Treatment protocols for FD include observation, medical treatment, and surgery. Clinical observation is suggested for FD lesions that have no risk of pathologic fracture or deformity. Medical treatment with bisphosphonates may have benefits, including improvement of function, pain relief, and lower fracture risk in appropriately selected FD patients.^[13] One study reported clinical improvement in children and adults treated with bisphosphonates.^[14] When the lesion involves frontal bone, nasal bones, orbit, ethmoid, zygoma, and upper maxilla, radical surgery is suggested, but this approach is difficult in treatment of recurrences. When the lesion involves frontal bone, nasal bones, orbit, ethmoid, zygoma, and upper maxilla, radical surgery is suggested but this approach is difficult in the treatment of recurrences as radical surgery would possibly increase morbidity by removal of the teeth. Hence, conservative treatment has been the treatment of choice. Shaving and debridement of the lesion are parts of conservative treatment.^[15] Surgery is indicated for confirmatory biopsy, correction of deformity, prevention of pathologic fracture, and/or elimination of symptomatic lesions. Conservative management has been the standard of care, which involves removing the diseased bone through an intraoral approach.

If there is no symptom or evidence of progression during follow-up, surgical treatment is not considered. Recurrence of FD is rare when the lesion has occurred in adults, but it is seen more commonly in the growth period. Because of the conservative surgery and unsuccessful removal of the lesion

cause the increased risk of recurrence. Patients with craniofacial FD have the risk of recurrence ranged from 15% to 20%.^[16] The maxillary area and also other areas of craniomaxillofacial skeleton which includes the structures such as the orbital region, mandibular or the zygomatic bone, and the cranial base, may cause some problems to the surgeon because of their anatomical relationship to important structures.^[12]

CONCLUSION

FD is considered a pathology, which may present functional and esthetic impairment. Proper clinical and radiographic features of the patient are mandatory for confirmative diagnosis. In the moment of decision to treat FD, consideration should be given to the patient's age, presence or absence of facial asymmetry, facial involvement, and future rehabilitation. Recurrences are common for FD; hence, long-term follow-up should be done.

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