Case Report

Monostotic Fibrous Dysplasia of Maxilla: A Case Report

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Abstract

Fibrous dysplasia (FD) is a frequent benign skeletal disease that can affect one bone (monostotic) or several bones (polyostotic). It can affect any part of the skeleton, although it is more prevalent in the long, ribcage, and craniofacial bones. Monostotic FD makes up 70–80% of all FDs, and 10–25% of cases happen in the craniofacial area. These instances are more prevalent in the 20–30 age range. There is a wide range of clinical manifestations associated with FD, from small monostotic lesions affecting a single bone to severe polyostotic illness affecting the whole skeleton. We report a case of 20-year-old male patient with FD involving right maxilla. For a conclusive diagnosis, the various imaging modalities, the clinical diagnostic strategy, and the techniques for histological analysis have been elaborated.

Keywords: GNAS1 gene, Monostotic fibrous dysplasia, Skeletal disease.

Introduction

Fibrous dysplasia (FD) is a disorder caused by sporadic mutation of the α-subunit of the G stimulatory protein (*mutations of the GNAS1 gene, position at 20q13.32*), in which bone is replaced and distorted by poorly organized, structurally unsound, fibrous tissue.[1] Normal bone is replaced by a newly created aberrant bone that contains fibrous connective tissue. In the beginning, Lichtenstein, and Jaffe in 1938 and, respectively, described FD in 1942. FD is a kind of hamartoma in which improperly calcified and immature bone replaces regular medullary bone.[2] It is an uncommon disease and accounts for 7% of benign tumors. It mainly affects children and young adults.[3]

Bone discomfort and recurrent fractures are the most prevalent symptoms observed in children and adolescents, followed by bunioned deformity and neurological compression, particularly in cases of involvement of the skull and face bones. The long bines, ribs, pelvis, and craniofacial bones are among the skeletal locations that are frequently affected.[4] Computed tomography (CT) imaging may distinguish between three types of FD patterns: a ground-glass pattern, a homogeneously packed pattern, and a cystic pattern. Surgery is the mainstay of treatment; however, the specifics vary based on the lesion's size, location, and symptoms. The use of bisphosphonates to lower osteoclastic activity is being considered as a surgical substitute.[4]

Here, we report a case of FD involving maxilla in a male patient who was diagnosed with the help of various imaging modalities and confirmed the diagnosis with an incisional biopsy.

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Case Report

A 20-year-old male patient (Figure 1) was reported to the Department of Oral and Maxillofacial Surgery, Index Institute of Dental sciences, Indore, Madhya Pradesh with the chief complaint of painless swelling of the left side of his upper face causing disfigurement. The swelling was hard and non-tender to palpation. The onset of the swelling was gradual, initially noticed as a pea size and had since been increasing in size in the past 5 years. No relevant familial history and the swelling extended superiorly from inferior border of orbital rim to the cheek region inferiorly, anteriorly—from the level of ala of nose to 3 cm away from the tragus of ear posteriorly and the skin over the swelling is same as surrounding skin with no visible pulsations. A well-defined swelling was seen over the right labial and buccal vestibule i.r.t 12, 13, 14, 15, 16, 17, anteroposteriorly extending from mesial aspect of 12 to distal aspect of 17, superior inferiorly from 2 mm above the gingival margin to zygomatic arch intraorally. The history and palpatory data were all considered, and a tentative diagnosis of fibro-osseous lesion affecting the right maxilla was made.

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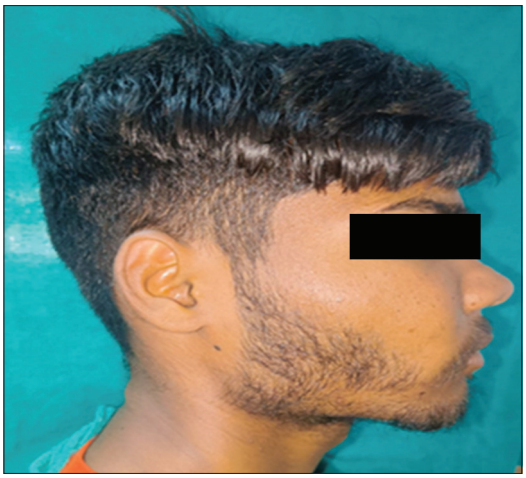

Figure 1: Left side of maxilla

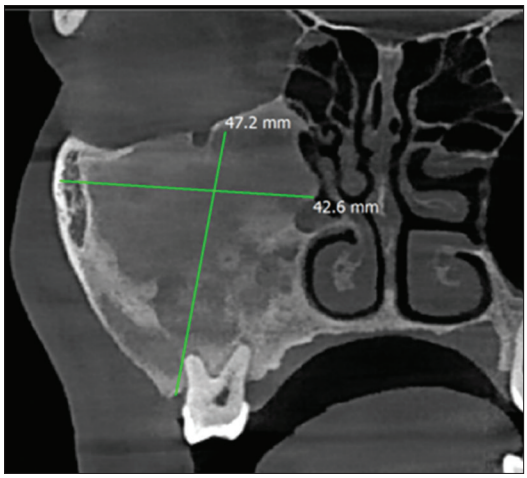

Figure 2: Axial section of cone-beam computed tomography

Every parameter on the complete hemogram was found to be within normal ranges. In addition, serological studies involving serum alkaline phosphatase, serum phosphorus, and serum calcium were within typical ranges.

The cone-beam CT images revealed a prominent homogenous hyperdense area involving right side of maxilla (Figure 2). The hyperdense area is giving a ground glass appearance with complete loss of normal trabecular pattern. The hyperdense area showed Bucco-palatal expansion causing cortical thinning of the involved bone. Sagittal cuts show that the lesion extended from the alveolar crest of the right posterior teeth (from the lateral incisor to the third molar and pterygoid plates) to the right orbital floor in the superior-inferior direction. Axial cuts revealed that the lesion obliterated the right maxillary sinus completely and caused expansion of the anterior, posterior, and lateral walls of the right maxillary sinus while maintaining the maxillary sinus outline. Moreover, the coronal cuts showed that the lesion extended from the lateral wall of the right nasal cavity to the lateral wall of the right maxillary sinus and zygomatic arch with expansion in both lateral and superior walls of the maxillary sinus.

Final diagnosis of FD was confirmed after histopathological examination. Surgical recontouring of maxilla was done by Weber Ferguson approach (Figure 3) under general anesthesia. Closure done in layers by Vicryl 4.0 suture (Figure 4). Patient is under follow-up for every 3 months.

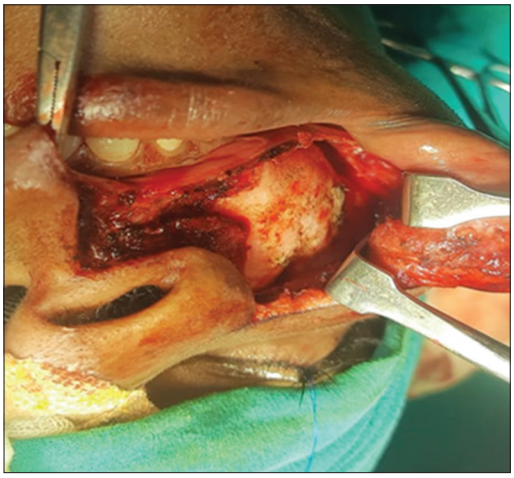

Figure 3: Weber-Ferguson incision

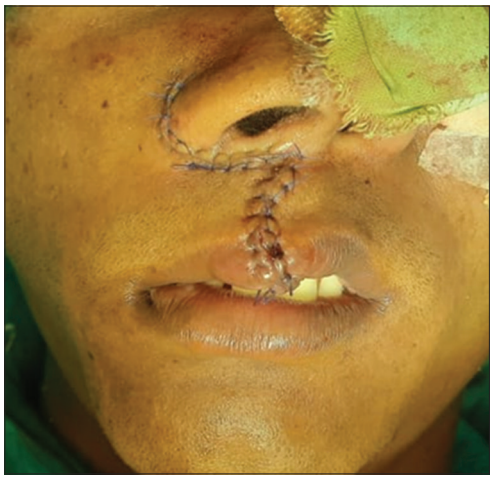

Figure 4: Closure done with Vicryl 4.0

Discussion

FD is a bone disorder in which there is mutation of the GNAS1 gene, position at 20q13.32. The term FD was introduced by Boyce and Collins in 1993. This was first introduced by Lichtenstein in 1938. Lichtenstein and Jaffe in 1942 and originally termed Jaffe-Lichtenstein syndrome. FD may occur in isolation or in association with skin and endocrine features, termed the McCune-Albright syndrome (MAS).[5] The incidence has been estimated at 1 in 5000–10,000. There is no gender predilection.

According to WHO in 1971, classified FD of jaws was in the category of neoplasms and other tumors related to bone in the heading of odontogenic tumors. The WHO in 1992, 2005, and 2017 classified FD's jaws directly under the heading of odontogenic tumors.[6]

There are mainly two types of FD based in the number of bones which are affected with the disease, monostotic and polyostotic. Monostotic form is mainly discovered incidentally and the most common locations in this are ribs followed by skull and femur. However, polyostotic FD is diagnosed during the first few years of life. Most bony lesions become non-silent and clinically significant by age 10 years and no new lesions appear after the age of 15. The most common locations in this type of FD are skull followed by mandible, pelvic bone, and lastly femur.

Monostotic form is most frequently seen (75–80%); clinical features of the monostotic form includes painless swelling, mal-alignment of teeth, tipping, or displacement of teeth due to progressive expansile lesion, tenderness is a late feature and mucosa is almost intact over the lesion.

Polyostotic form has the frequency of 20%; clinical features of the polyostotic form include facial asymmetry diplopia/proptosis/loss of vision, raised intracranial and intraorbital pressure, sinus infection, deafness, oro-nasal obstruction, malocclusion, and cranial nerve involvement.[7,8] Polyostotic form could be associated with syndromes like Jaffe-Lichtenstein and MAS.

According to Lee *et al*. in patients with nonaggressive but active FD, it is better to wait until the lesion stops growing and the operation can be performed when the patient has reached skeletal maturity. Surgical recontouring and debulking may be necessary to achieve acceptable facial proportions. In less than 1% of cases of FD, malignant transformation is seen.[9]

Depending on the patient's age, clinical care of FD might be quite challenging. Surgery, medication, and monitoring are all part of the FD therapy plan. Due to the increased risk of pathologic fracture or deformity associated with FD lesions, appropriate clinical surveillance is necessary.[10] While there is not a single drug that can stop the disease process, medicinal therapy can be used to palliate symptoms, Bisphosphonates, for example, have been shown to increase function, reduce pain, and reduce the chance of fracture. Using an intraoral technique to remove damaged bone, conservative therapy is the standard care management. The employed bone grafts are cortical bone grafts as opposed to cancellous bone grafts or bone graft substitutes because of its superior quality of rebuilt cortical bone.[11,12]

Conclusion

It is uncommon to find a single verified instance of FD affecting the maxilla or mandible. It becomes quite challenging to distinguish between these benign and malignant bone illnesses. To create a complete examination, a good patient history, a radiographic assessment, and a diagnosis of FD are frequently enough. The patient's age, the degree of facial involvement, the existence or lack of facial asymmetry, and the plan for future rehabilitation all influence the therapy strategy. Most of the tissue must be surgically removed without resulting in any deformity, loss of function, or impairment of noble structural function. Any severe deformity, excruciating pain, or pathological fracture might be signs that surgery is necessary. It is important to do appropriate and consistent follow-up to identify any malignant alterations or relapses.

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