

## CASE REPORT



## Fibrous dysplasia of the pediatric anterior maxilla: A case report and review of literature

Sunil Vasudev, Partha Pratim Debnath, S. Deepak, G. R. Maneeshashree

Department of Oral and Maxillofacial Surgery, DAPM RV Dental College, Bengaluru, Karnataka, India

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**Corresponding author**

Dr. Partha Pratim Debnath, Department of Oral and Maxillofacial Surgery, DAPM RV Dental College, Bengaluru, Karnataka, India. Phone:+918094395473. E-mail: debnathpartha29@gmail.com

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**Abstract**

Fibrous dysplasia (FD) is a benign fibro-osseous lesion in which the fibrous connective tissue containing abnormal bone replaces normal bone. FD is linked to GNAS1 guanine nucleotide-binding protein, alpha-stimulating activity peptide 1, and the gene mutation resulting in the abnormal proliferation and differentiation into pre-osteoblast. In this case report, we present a case of 9 years old boy with monostotic FD affecting the left maxilla causing bony expansion and asymmetry of the right side of face.

**Introduction**

Fibrous dysplasia (FD) is a benign fibro-osseous lesion in which the fibrous connective tissue containing abnormal bone replaces normal bone. In 1891, Von Recklinghausen described “generalist fibrous osteitis” pathological conditions that characterized deformities and alterations in the bone. In the year 1938, Liechtenstein and Jaffe recognized these lesions and described as a well-characterized pathology and named it as FD.<sup>[1,2]</sup> FD is linked to GNAS1 guanine nucleotide-binding protein, alpha-stimulating activity peptide 1 and the gene mutation resulting in the abnormal proliferation and differentiation into pre-osteoblast.<sup>[3]</sup> If it is affecting only one bone termed as monostotic form, multiple bone-polyostotic form or may be seen in combination form with hyperfunctioning endocrinopathies and hyperpigmented skin lesions such as “café-au lait” spots in skin known as Mc Cune-Albright syndrome. FD is also found in Mazabraud’s syndrome, an atypical benign disease where soft-tissue myxoma is associated with Mc Cune-Albright syndrome. The distribution of FD is noted as follows 74% monostotic, 13% polyostotic, and 13% craniofacial form.<sup>[4]</sup> Malignant transformation has been reported rarely. Clinically, the lesion presents as asymptomatic and gradual enlargement of the involved bone seen which causes facial asymmetry and

deformity. In this case report, we present a case of 9 years old boy with monostotic FD affecting the left maxilla causing bony expansion and asymmetry of the face.

**Case Report**

A 9-year-old boy reported to the Department of Oral and Maxillofacial Surgery, DAPM RV Dental College and Hospital with the complaint of swelling on the right side of the face since past 2 years [Figure 1]. On clinical history, the swelling was noticed by his parents 2 years ago but the timing of occurrence was not known. On extra oral examination diffused swelling noted measuring about 30 cm on right side of the face extending antero-posteriorly from the lateral wall of the alar base to the lateral canthal line; superio-inferiorly extending from 1 cm below the infra orbital rim to the line joining corner of mouth to tragus. On extraoral palpation, the consistency of the swelling was bony hard, nontender, immovable, and no local rise of temperature noted. On intra-oral examination mouth opening of 38 mm noted with deranged occlusion on the right side. Increased volume with vestibular obliteration noted from the right canine region extending up to the infra zygomatic region with expansion of the buccal and palatal cortical plates. The overlying mucosa was normal in appearance and no tenderness noted

[Figures 2 and 3]. Computed tomography face revealed a massive radiopaque lesion with ground glass appearance extending from the lateral wall of the nose to the right infra-zygomatic region. Axial and coronal sections confirmed the complete obliteration of the maxillary sinus with deviation of

nasal septum toward the contralateral side [Figures 4 and 5]. After the clinical and radiological examination, the provisional diagnosis was made as monostotic FD of the right maxilla. Osteoplasty was done to excise and contour the bony lesion of the right maxilla using vestibular approach under general anesthesia and collected bony specimens were sent



Figure 1: Pre-operative profile



Figure 4: CT coronal section



Figure 2: Pre-operative occlusion

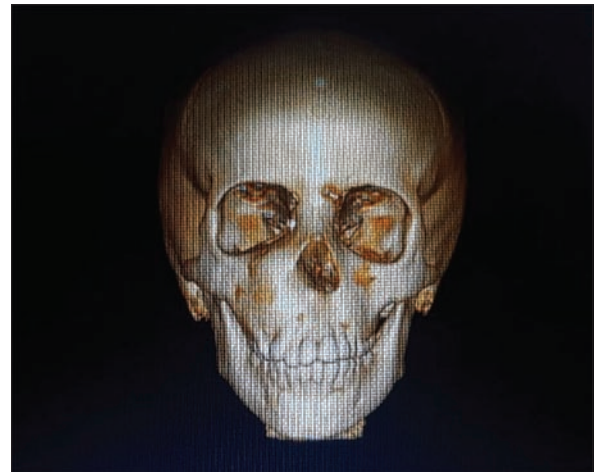


Figure 5: 3D reconstruction



Figure 3: Palatal extension of the lesion

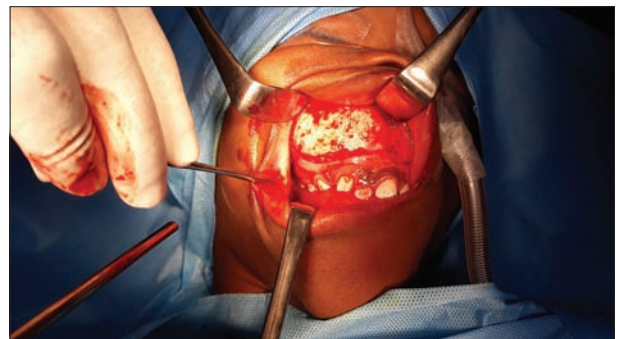


Figure 6: Intraoperative picture

for histopathological examination [Figure 6]. Patient was followed up with clinical and radiological examination for 5 years and no recurrence was noticed. Histopathological examination revealed fibrous stroma made up of fibroblast in a compact stroma of interlacing fibers, irregularly scattered trabeculae of bone seen throughout the lesion giving C-shape or Chinese letter appearance. The clinical, radiological, and histopathological examination confirmed the lesion as FD affecting the left maxilla.

## Discussion

The FD is a benign lesion, characterized by defective bone modeling, with gradual replacement of normal bone by fibrosis with irregularly mineralized osteoids resulting in swelling and expansion of the affected bone. Various theories have been postulated in the literature regarding the etiopathogenesis of FD, including the trauma with a non-specific disturbance in local bone reaction, as a congenital anomaly degenerate activity of mesenchymal bone forming cells.<sup>[5,6]</sup> Binding of ligand to G-protein coupled receptor causes dissociation of alpha subunit from  $G_{\beta}$ ,  $G_{\gamma}$ , and replacement of linked to  $G \alpha$  GDP by GTP leads to activation of adenylyl cyclase by binding to active alpha-subunit, followed by formation of cyclic adenosine monophosphate (c-AMP) from adenosine triphosphate which is secondary messenger that activates intracellular proteins. Overproduction of the c-AMP causes hyperfunction of endocrine organs, increases proliferation of melanocytes, and effects on differentiation of osteoblasts thus leading to FD.<sup>[7]</sup> Monostotic form more commonly seen in children and younger age <10 years with equal gender distribution. Maxillary posterior region is most commonly affected which extend locally to involve maxillary sinus, zygomatic process, floor of the orbit, and skull base. Schlumberger *et al.* described site involved in decreasing order as ribs (28%), femur (23%), tibia (21%), craniofacial form (10–25%), and humerus (4–7%).<sup>[7]</sup> Unilateral swelling may be asymptomatic, painless slow growing in nature and with progressive enlargement causes expansion of the buccal and palatal cortical plate which may lead to obstruction of nose, compression of orbital content and other vital structures in the involved region.<sup>[8]</sup> Radiological features are variable which depends on the stage of the lesion, early stage – unilocular or multilocular radiolucency with well circumscribed border may be seen; intermedial – radiolucent lesion with intermittent irregular, patchy radio-opacities give mottled appearance; matured stage – homogenous radiopacity gives ground glass, pebbled, and orange peel appearance with poorly demarcating margins of lesion blended with surrounding normal bone.<sup>[4]</sup> Routine blood investigation might show increased alkaline phosphatase and urinary hydroxyproline. Recurrence of FD is seen more commonly in growth period but rare when the lesion has occurred in adults.<sup>[2]</sup> The main management for

FD is surgery, which can be divided into conservative and radical resection. If any clinical features of deformity, pain or functional disability suggest the need for intervention and complete surgical resection of the involved area is the treatment of choice. Radiotherapy is avoided as it can encourage transformation into post-irradiation osteosarcoma. Bisphosphonate therapy may be helpful in decreasing pain, function, and reduces the risk of fracture. Post-operative observation should be done through serial radiographs, CT scans, and clinical examination. While planning the management for monostotic FD and craniofacial FD apart from the conventional radiographic evaluation nuclear scintigraphy, molecular or mutational analysis of GNAS1 gene, lab studies to document endocrine disturbances should be considered as it helps in a proper evaluation of and development of treatment goal. According to the literature preferred approach for monostotic FD and craniofacial FD is no treatment with periodic observation<sup>[8]</sup> but in our case considering the facial deformity, deviation of the nasal septum, patient concern about the appearance surgical contouring was performed under general anesthesia.

## Conclusion

Considering the clinical scenario, FD is considered a bony lesion, which may cause functional and unacceptable esthetic appearance for the patient. While making the decision to treat FD, significant attention should be paid to the patient's age, presence or absence of facial asymmetry, and involvement of vital structures leading to significant impairment of function. Since it is a tumor with no exact and relapsing limits, it is important to remove the lesion as much as possible without causing disfigurements to the patient, functional discrepancies. In young-aged monostotic FD cases, regular follow-up or minor bony osteoplasty at affected site is adequate for aesthetic and functional purpose.

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