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## RESEARCH ARTICLE

# LIVING WITH FIBROUS DYSPLASIA- A CONUNDRUM: CASE REPORT

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**ABSTRACT**

Fibrous dysplasia is a fibro-osseous lesion of the jaw bone. It occurs in maxilla and mandible with a more predilection to maxilla. It starts in childhood and progress slowly and mostly ceases after the onset of puberty. However it can affect any bones in the skeletal frame work, including the craniofacial system. Though exact etiology unknown, gene mutation is likely to be the cause .The bone formation is accelerated due to this mutation but the new bone layed down is poorly calcified and immatured. Aesthetic compromise or functional disturbance is less likely, however these cannot be ruled out. This case is unique in its presentation with little or no much esthetic compromise, so we tried to bring out the significance of an intervention in such caes.



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## INTRODUCTION

Fibrous dysplasia is a benign intramedullary fibro-osseous lesion of the bone affecting maxilla and mandible (Mathew, 2005; Shanmugam1, 2017). It was initially explained by Lichtenstein in 1938 and Jaffe in 1942 (Mansi Agarwal, 2014). It is askeletal disorder which starts in childhood characterized by slow progressive enlargementthat can affect one bone (monostotic form), or multiple bones (polyostotic form) which forms a part of the McCune Albright syndrome (MAS) or of the Jaffe Lichtenstein syndrome (Mansi Agarwal, 2014) The new bone formed is poorly calcified but well vascularized and contains trabeculations (Surajit, 2015).This case is unique in its presentation as the patient suffers it from childhood. The changes which were brought about by the lesion was seen in the examination though the patient was unwilling for an intervention. These cases are rare, so we thought such a case of many years of history should be presented to all the medical fraternity. This case is an eye opener for those who violate the ethics of any treatment regime.

**Case Report:** A 22years old male, engineer by profession came to the dept. of Oral Medicine and Radiology, Coorg institute of dental sciences, Karnataka, India, with c/o of swelling on the left side of the face, sine 10years. The swelling was small in size to start with but gradually progressed to the present size over the years. Patient informed consent was taken.There was no history of trauma, fever, or similar swelling elsewhere in body, with anon contributory past medical, dental, family or personnel history but his oral hygiene measures were proper. He had no visual disturbances, no history of headache, earache, and nasal obstruction or discharge. He had no adverse habits. On general examination he was moderately built and nourished with no signs of clubbing, cyanosis, odema. On extraoral examination there was an asymmetry of face with a diffused bony hard swelling in the left malar or maxillary sinus region at the level of line joining the corner of lip and tragus of ear below horizontally and from infraorbital rim superiorly to a line joing from the alae of nose 5 cm posteriorly. The swelling is dome shaped 5x5 cm size limited to the malar region with no change in colour of overlying skin and visible pulsations no lymphadenopathy (Fig 1). Intraoral examination revealed a single bony hard swelling in the region of 23 to 26, dome shaped, size 5x5 cm. The swelling extends from attached gingiva to vestibular sulcus vertically, and from 23 to 26 horizontally in the maxillary buccal shelf area. The mucosa overlying is not stretched and there are no ulceration or discharge. Rest of the oral cavity appeared normal including the periodontal status of the teeth (Fig 2).

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The relevant radiographs taken are intraoral periapical radiograph and maxillary occlusal.

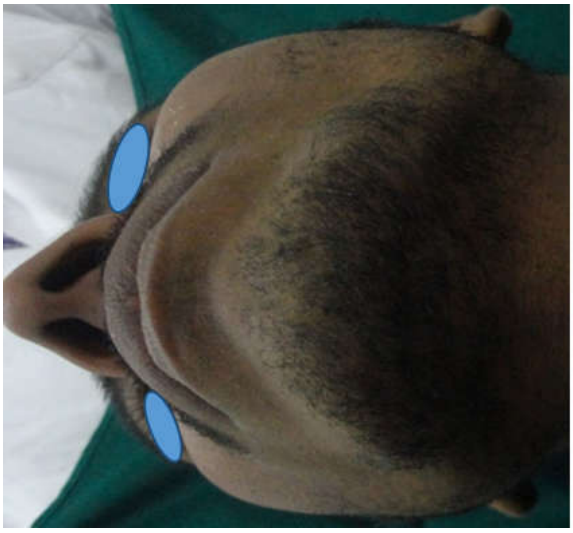

**Photo 1. Extra oral profile picture showing facial asymmetry**

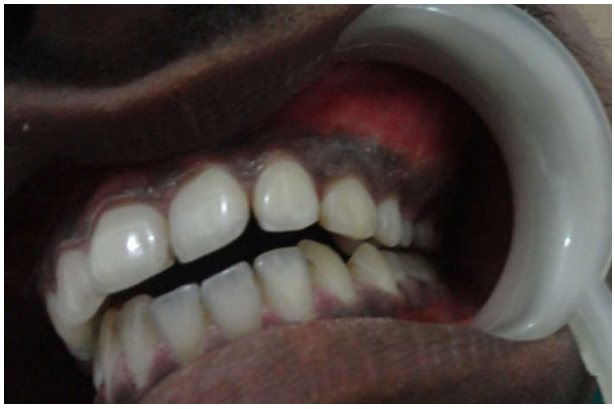

**Photo 2. Intraoral photo showing the lesion**

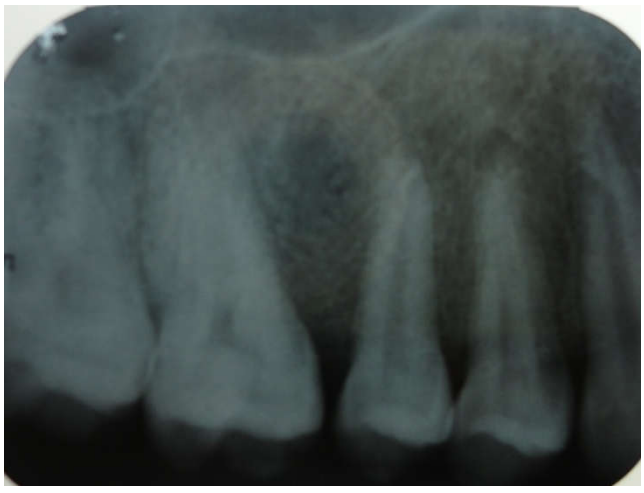

**Photo 3. Intraoral periapical radiograph of the lesion**

Routine blood investigation values were normal. On the basis of history, clinical examination a provisional diagnosis was made as a benign bony lesion - fibrous dysplasia of maxiilaon left side. Radiographic findings- A classical ground glass appearance was able to appreciate in the region in all the radiographs. Based on the history of long duration, asymptomatic nature, clinical features which correlate with the radiographic findings, a provisional diagnosis of fibrous dysplasia was made.

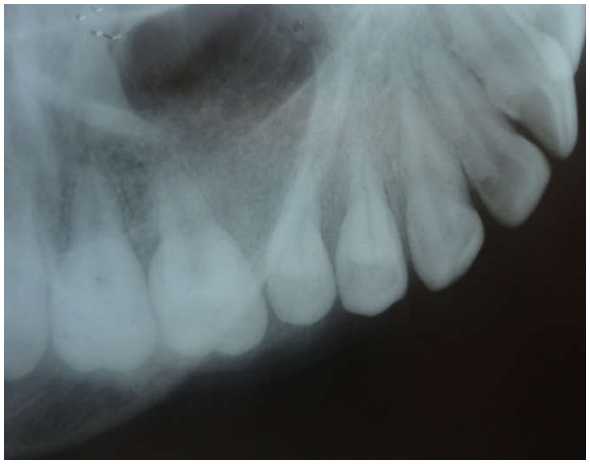

**Photo 4. Maxillary occlusal radiograph showing the lesion**

Due to its asymptomatic nature, patient was not willing for further surgical treatment, so a protocol was followed as regular follow up in every 6 months.

## DISCUSSION

Fibrous dysplasia is a bony lesion of unknown etiology, uncertain pathology. It is a nonmalignant fibro osseous lesion in which the involved bone gradually get converted into expanding cystic and fibrous tissue, which is less calcified but vascularity is not compromised. It may involve in single (monostotic), or multiple bones (polyostotic) but occurs in throughout the skeleton with more predilection for the long bones, ribs, and craniofacial bones (Surajit, 2015; Mathew, 2005)

### Aetiology pathophysiology

It is classified by W.H.O. as developmental in origin.[3]The exact etiology although unknown, different hypothesis have been postulated. The most widely accepted theory is that fibrous dysplasia results from an abnormality in the development of bone forming mesenchyme. So Fibrous dysplasia is a developmental aberration caused due to mutation in the genes that encodes the subunit of stimulatory G protein (Gs) located at 20q13.2. This activates the bone forming cells called mesenchymal cells to differentiate in an uncontrolled manner producing immature mesenchymal tissue. These fail to mature resulting in poorly calcified bone tissue. The mutation takes place after conception in the early stages of foetal development, so this is a somatic rather than germ line mutation. The exact cause for mutation is unknown All cells derived from the mutated ones show the dysplastic features. The clinical presentation varies depending on where in the cell mass the mutation is located and the size of the cell mass during embryogenesis when the mutation occurred (Mathew, 2005; Surajit, 2015). In 2012, Koutlas et al. explained the evaluation to the expression of TWSG1 (Twisted Gastrulation) as an example of a BMP-binding protein because of its known role in regulating BMP activity during mandibular morphogenesis and post natal bone remodeling (Mansi Agarwal, 2014). The syndromes associated with this are McCune-Albright syndrome, Mazabraud's syndrome. Jaffe

Lichtenstein syndrome (Surajit Bhattacharya, 2015). Fibrous dysplasia starts in childhood and generally slows or ceases after puberty, so mostly it's seen in first 3 decades of life. Fibrous dysplasia can occur in any type of bones, endochontral or membranous. In addition to this another type is also noted called as craniofacial type where in involved bones are confined to craniofacial skelton where posterior aspect of maxilla is commonly involved Clinically this presents as a asymptomatic, nontender, bony hard, diffuse, dome shaped swelling, which is gradual in onset with intact overlying mucosa. Since condition is asymptomatic most of the patient seek treatment when there is gross facial asymmetry or any functional disturbances. Recommended treatment options can be divided into 4 categories:

- Observation,
- Medical therapy,
- Surgicalremodeling,
- Radiacl excision and reconstruction (Shanmugam, 2017).

**Conclusion**

The fibrous dysplasia affecting the maxilla is a distinctive entity that can be treated by bone recon touring or shaving. The intervention are done probably after the growth cessation so that chance to recur is minimal.

A follow up protocol is followed considering the probable flare up and continuous growth of the lesion. We are reporting this case due to its rarity and peculiarity of growth.

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