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MAXILLARY FIBROUS DYSPLASIA – A CASE REPORT

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ABSTRACT: Fibrous dysplasia is an idiopathic skeletal disorder in which the trabecular bone is replaced and distorted by poorly organized, structurally unsound fibro-osseous tissue. The lesion is classified into two forms: Monostotic and polyostotic. Characteristic radiographic appearance shows an expanded osseous lesion having poorly defined margins covered by a thin "eggshell" cortex and lacking periosteal new bone formation.

KEYWORDS: Fibrous dysplasia, maxilla, monostotic fibrous dysplasia.

1. INTRODUCTION:

Fibrous dysplasia is defined as a benign fibro – osseous lesion. In this condition, normal bone is replaced by fibrous connective tissue, containing abnormal bone. In the year 1891, Von Recklinghausen coined the term 'generalist fibrous osteitis', which included pathological conditions that characterized deformities and bone alterations 1. The term "fibrous dysplasia" was coined in the year 1938 by Liechtenstein and Jaffe 2, as they mentioned the above-mentioned condition's to be one well defined characteristic disease.

Fibrous dysplasia is classified into monostotic or polyostotic. The monostotic variant involves a single bone, polyostotic involves several bones simultaneously. Monostotic dysplasia is usually characterised by a painless swelling and is most often diagnosed during the second decade of life. It has a female predilection. Polyostotic dysplasia affects a few or up to 75% bones of the entire skeleton 3.

The typical clinical features of this condition are volume increase or slow growth with the bulging of the affected region and facial asymmetry. The soft tissue remains unaffected, pain and parasthesia are also absent.

The radiographic techniques used include conventional radiography, scintigraphy, magnetic resonance imaging and computed tomography.

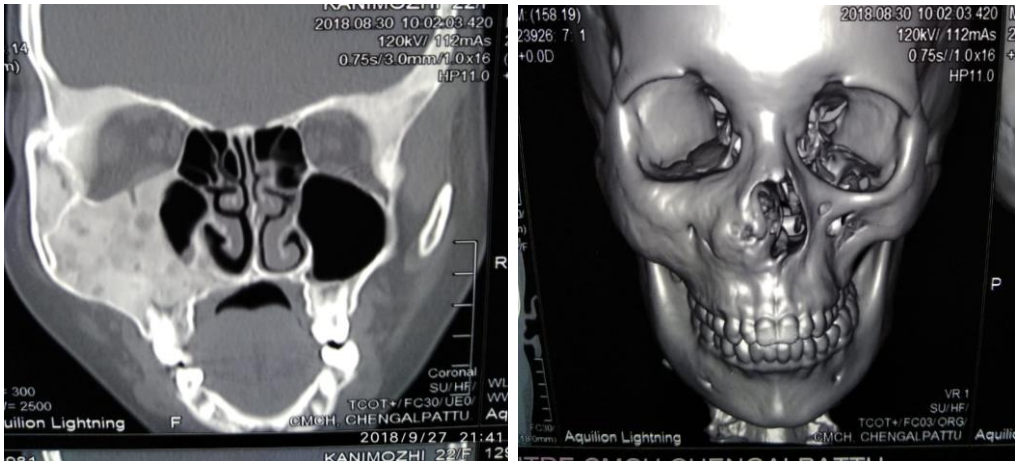
This is a case report of a 22-year-old female patient with fibrous dysplasia in the maxilla.

2. CASE REPORT:

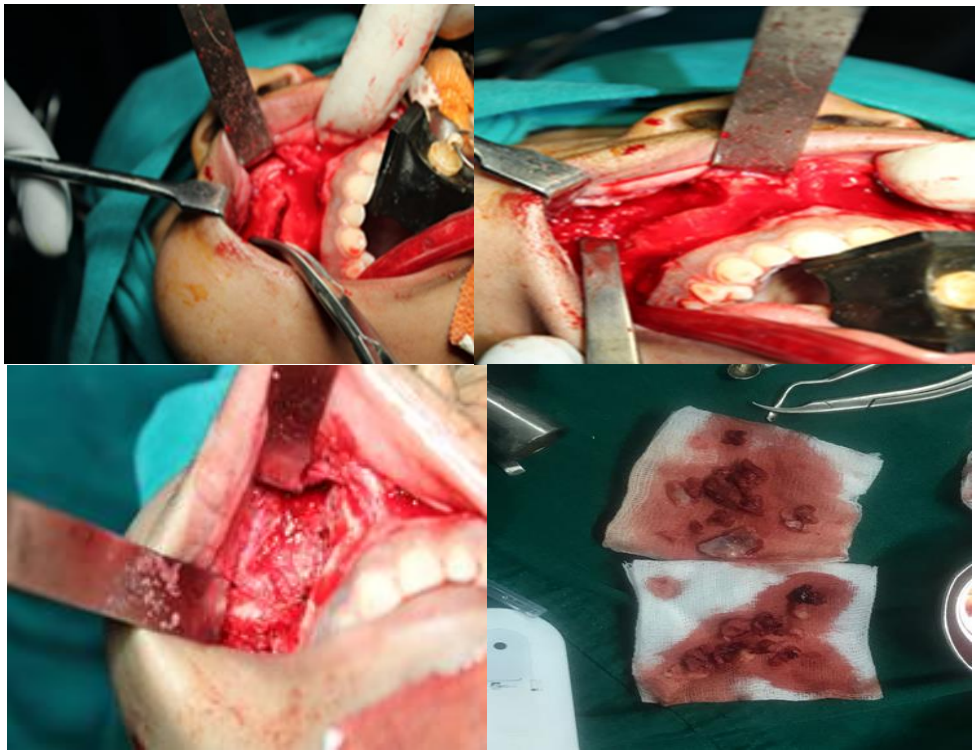
A 22-year-old female patient reported to the Department of Oral and maxillofacial surgery, Sree Balaji Dental College, Chennai. The patient complained of a swelling, extra orally in the right maxillary region, which had gradually increased in size. The patient's primary concern was facial aesthetics. There was no history of pain or parasthesia .

On examination, an extra oral diffuse swelling, measuring 3x4 cm was seen in the right maxillary region, super inferiorly starting 2cm below the infraorbital margin to the line joining the commissure and the ear lobe and mediolaterally starting from the right side of the nasal septum to ramus of the mandible on the right side, which was bony hard and non-tender in nature. Mild facial asymmetry was observed. Intra orally, the swelling extended from 22 to 25 tooth, with expansion of buccal cortical plate and obliteration of the vestibule.

Facial CT revealed a hyperdense image, an expansive mass in the right maxillary region. It revealed am insufflating lesion with dense glass density in the maxillary bone, partially occupying the right maxillary sinus and the ipsilateral nasal cavity and lowering the hard palate with involvement of the dental alveoli.



The patient was operated under general an aesthesia. An incision was made in the upper right vestibule, mucoperiosteal flap was elevated and the lesion was exposed. Partial resection of the lesion, followed by osteoplasty for correction of bone spikes with spherical drill was carried out.



3. DISCUSSION:

Fibrous dysplasia is a benign lesion, which is relatively uncommon. It causes defective bone modelling, wherein the normal bone is gradually replaced with irregularly mineralised osteoid 4. As quoted by Harri SWH et al., this disease is seen among the young population with an incidence rate of 1: 4000 – 1:10,000. It results in facial symmetry 5. Crawford explained this disease in simple terms as bone being replaced by fibrous tissue6.

As stated by Waldron, it occurs due to a mutation of GNAS1 gene7. The signs and symptoms depend upon the location of the lesion and the compressive effect on the neighboring structures. Some of clinical features and signs include - Asymmetry and facial deformity, Pathological fractures, Obstruction of the paranasal sinuses generating recurrent infections, cysts and mucoceles, anosmia, headache, loss of visual acuity by compression of the optic nerve, alteration of ocular movements, ptosis, exophthalmos, strabismus, conductive hearing loss etc 8.

In this case, the patient had the swelling for almost five years but did not have any signs of paresthesia or pain. The swelling had started as a pea sized swelling and had gradually increased in size. But for that, the patient was asymptomatic.

Treatment of this disease is still an issue of debate. The best choice is partial resection and bone remodelling. Aggressive surgeries are carried out in those cases where hearing is impaired or the dentition is affected. Radiotherapy is contra indicated as it can lead to malignant transformation of the lesion. Chemotherapy has been found to be ineffective 9. In this case, we have carried out partial resection and osteoplasty, as the lesion was not found to be aggressive.

Since there is a chance of recurrence, it is always advisable to have regular clinical and radiological follow ups of the patient. We carried detailed and regular checkups for the patient, clinically as well radiologically.

4. CONCLUSION:

Fibrous Dysplasia is considered a pathology, which may present functional and aesthetic impairment. Treatment should be based on the patient's age, presence or absence of facial asymmetry, facial involvement and future rehabilitation. It is important to remove as much tissue as possible without causing mutilations to the patient, functional deficits or lesions of noble structures. Surgical treatment is indicated in case of significant deformity, significant pain or pathological fracture. Follow up is of fundamental importance in order to detect relapses or a possible, malignant change at an early stage.

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