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Fibrous Dysplasia - A Rare Case Presentation

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ABSTRACT

A developmental benign medullary fibro-osseous process known as fibrous dysplasia (FD) is typified by the failure to create mature lamellar bone and a stoppage as woven bone, which may be multifocal. Fibrous dysplasia is a rare condition that can affect people of any age in both adults and children. Typically, young adults and children receive the initial diagnosis. Although the actual frequency is unknown, it is thought to account for about 5% of benign bone lesions. This exercise looks at how fibrous dysplasia should be evaluated and emphasizes the need of a multidisciplinary approach in providing care for people with this illness.

Keywords: fibrous dysplasia, craniofacial fibrous dysplasia, maxilla

INTRODUCTION

The characteristic of fibrous dysplasia, which is usually non-neoplastic, is intramedullary fibro-osseous growth resulting from abnormal osteogenesis. (1)

The illness was first identified as "osteitis fibrosa generalisata" in 1891 by von Recklinghausen in a patient who had skeletal abnormalities as a result of fibrotic bone alterations. Lichtenstein coined the term "fibrous dysplasia" in 1938.(2)

There are two types of FD: the monostotic form, which affects one bone in around 70% of cases, and the polyostotic form, which affects at least two bones in about 30% of cases.(3)

The term "craniofacial fibrous dysplasia" (CFD) refers to FD that affects the cranial and facial bones. Typically occurring unilaterally, FD is about twice as common in the maxilla as it is in the jaw bone. (4)

Gs-alpha gene somatic mutations on chromosome 20 can cause McCune-Albright syndrome (MAS), FD, and endocrine malignancies.(5)

In most cases of FD, it can be associated with endocrine abnormalities. calcium, parathyroid hormone, 25-hydroxyvitamin D, and 1,25-dihydroxyvitamin D levels are normal, although serum alkaline phosphatase (ALP) is occasionally increased. Individuals with widespread polyostotic FD may experience osteomalacia, hyperphosphaturia, and hypophosphatemia.(6)

Visible deformity, bone pain, functional impairment (restricted mouth opening, malocclusion, diplopia, etc.), or nerve injury can all be signs of craniofacial fibrous dysplasia (CFD). When the orbit is compromised, the optic nerve is especially vulnerable, and damage to the optic nerve increases the chance of permanent loss of vision acuity and/or visual field defect.(7)

CASE REPORT

A 32-year-old female housewife from Maharashtra presented to our out-patient

department with a complaint of swelling over the left side of her maxillary region.
The questionnaires revealed that the expansion of the swelling was slow, gradual increasing since the past 10-12 years.
The medical, dental, family, and personal histories of the patient were non-contributory.
On general physical examination the patient was conscious, cooperative, moderately nourished and built and showed no signs of anemia, icterus and clubbing. All vital signs were within normal limits.
On examination, the patient had painless unilateral swelling seen in relation to the upper left maxillary region. Swelling was seen extending from the lower eyelid to upper lip. The swelling was obliterating the nasal ala of the left side. Overlying skin was intact and no rise in temperature associated with the swelling was seen. There was no dental finding associated with the swelling.
The patient was referred for further laboratory investigations where her complete blood count, thyroid function test, liver function test, renal function test was within normal limits. Patients' serum calcium level was 8.30mg/dl, serum phosphorus levels were 3.49 mg/dl and parathormone levels were 99.20 pg./ml.

Radiology:
3 tesla MRI neck with contrast showed a mass involving left maxillary sinus, maxillary alveolar process, sphenoid sinus and greater wing of sphenoid likely to be giant cell tumor of the bone.

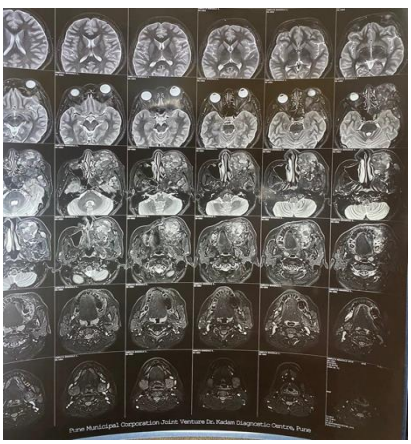


Image 1

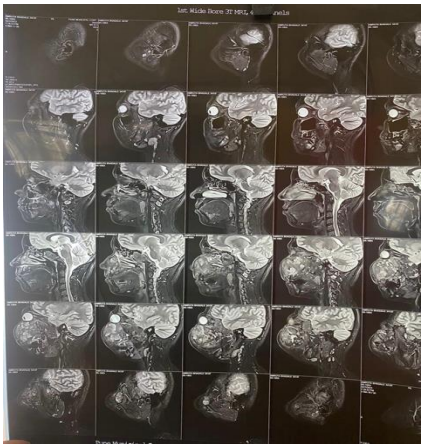


Image 2

CECT Scan of brain showed a large bony lesion involving the left maxillofacial region extending into left temporal and mandibular bone showing bony expansion, some cortical destruction with ground glass matrix and mild soft tissue component. In contrast, study showed minimal post contrast enhancement.
Whole body scan showed evidence of a solitary osteoblastic lesion involving the left maxillary region.
A differential diagnosis of fibrous dysplasia was made.

Histopathology:
Histopathology on gross examination showed: soft tissue blackish in color, soft firm in consistency with irregular surface and borders.
Histopathologic features: H&E studied section showed highly cellular fibro cellular stroma. The bony trabecular appeared to be in Chinese letter pattern. Bony trabeculae showed central pattern of calcification additionally there was lack of osteoblastic rimming. Foci of giant cells and few extravasated RBCs were noted.
And the overall findings were suggestive of Craniofacial Fibrous Dysplasia.

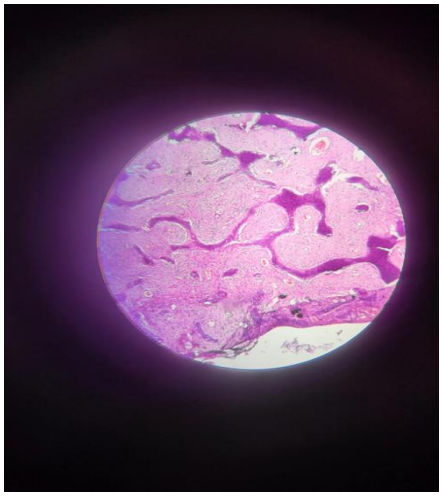
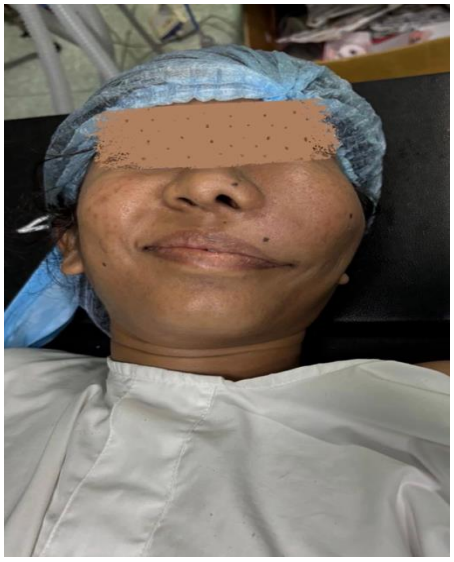


Image 3

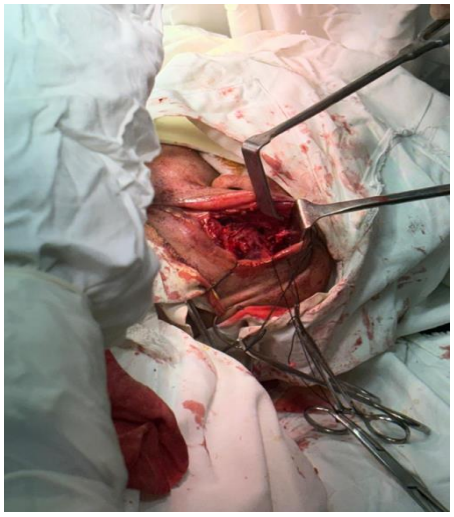
DISCUSSION

Most cases of FD occur gradually, and early oral and maxillofacial symptoms aren't obvious and are easily ignored by patients. As the disease progresses, symptoms such as pain and loosening of teeth may occur.(7) Facial asymmetry runs in families too. It usually occurs in women, usually grows slowly and stabilizes during adolescence, reoccurring in 37% of patients and remaining asymptomatic until age 10. Possible treatments include partial surgery, bone reshaping, surgical reshaping, and bone reduction/reduction to achieve aesthetic results. (8) FD can also affect tooth alignment and cause tooth and root displacement and root resorption, resulting in malocclusion, cavities, or teeth grinding. This is because the alveolar bone is rapidly expanding, which will affect the position of the teeth in the wrong direction.(7)

In this case, patients had complaints of volume increase in the region, hard consistency on palpation, functional impairment, facial asymmetry. Conservative management has been done as a standard of care using an intraoral approach to remove the affected bone. Excision of lesion has been carried out over the left maxilla under general anesthesia. This procedure includes gingival mucoperiosteal flap raised along the left buccal sulcus. Incision was taken from midline to 1st molar teeth. The diseased area was carefully cut in the buccal aspect. Extra neo-bone-fibrous tissue was cut and dissected out. Care was exercised to prevent damage to vital structures such as nerves. Tumor was excised in a piece meal. Removed as much tumor tissue as possible from the inferior incision site.



Pre-operative image



Vestibular approach

Shavings of the bone

Declaration by Authors

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Conflict of Interest: The authors declare no conflict of interest.

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