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SURGICAL MANAGEMENT OF FIBROUS DYSPLASIA USING STL MODEL & GUIDING TEMPLATE: CASE REPORT

Akash Tiwari, Srijon Mukherji, Prasanna Kumar, Ankur Rathaur, Ankita Raj, Ayushi Agarwal

Rama Dental College Hospital and Research Centre, Rama University,
Mandhana, Kanpur, Uttar Pradesh, India

ABSTRACT

A skeletal condition known as fibrous dysplasia (FD) is characterized by the replacement of healthy bone with fibrous bone tissue. Patients may experience numbness, discomfort, or swelling on the affected side. Monostotic fibrous dysplasia (MFD) is four times more common than polyostotic fibrous dysplasia. The maxilla is more frequently impacted than the mandible in MFD.

The clinical behaviour and fast course of FD make treatment difficult. Untreated cases have a 0.5% malignant potential. We show a case of FD involving the mandible here. Clinical diagnostics and therapy techniques have been developed utilising a guiding template.

INTRODUCTION

Fibrous dysplasia is a benign bone lesion characterized by replacement of the normal substance of the interior of the bone by fibroosseous connective tissue histologically exhibiting varying degrees of osseous metaplasia. It is a primary developmental abnormality of the bone forming mesenchyme in which fibrous tissue gradually expands and replaces the bone.

In 1938, Lichtenstein described the disease entity of fibrous dysplasia for the first time[8]. Albright et al. described histologically similar lesions linked with female premature puberty and patches of skin hyperpigmentation in 1937, using the term osteitis fibrosa dissemina. The syndrome is known as McCune Albright syndrome[9].

Fibrous dysplasia can affect one or many bones (monostotic or polyostotic).The third type of the condition is McCune Albright syndrome. Furthermore, cherubism is a hereditary familial type of localised fibrous dysplasia identified by Jones. symmetrical in both directions

A cherubic facial look is caused by involvement of the mandible and maxilla, as well as increased fibrotic submandibular lymph nodes and an upward tilt of the eyes. The illness typically manifests itself in childhood and continues through puberty and adolescence. The growth usually stops after adolescence.

Unfortunately, the few patients in whom the lesions continue to progress rapidly before, during, and after puberty, causing deformity and/or functional problems, cannot be ascertained when they are first seen. The clinical findings show asymptomatic enlargement of the involved bone, which produces facial asymmetry, a tilted occlusal plane, loss of teeth, or a grotesque facial deformity. In some patients, fibrous dysplasia involves most of the craniomaxillofacial bone, causing megacranium and a result known as "lion face." Skull involvement occurs in 27% of monostotic patients and in up to 50% of polyostotic patients[7,8]. Lesions involving the frontal, orbital, and sphenoid bones may cause visual disturbances, proptosis, orbital dystopia, or facial asymmetry[8,9]. Neurological complications such as fifth nerve impairment, hearing loss, and seizure disorders have also been reported.Systemic dysplasia can occur simultaneously, leading to syndromes such as craniometaphyseal dysplasia, or Pyle disease, and craniodiaphysial dysplasia.

The lesion may involve any facial bone but is more commonly seen in the maxilla, usually as a solitary lesion. In one series, however, the mandible was more often involved. Less frequently there may be multiple lesions involving both jaws[10].

Case Report

A 20-year-old female presented with painless swelling over the right side of her face for the past 11 years. When the patient first observed swelling on the right side of her face, it had no prior history of prodromal symptoms and was slowly growing and progressing. At the extra-oral clinical examination, in an inferior-superior view, facial asymmetry was observed, presenting a volumetric increase in the right mandible. The right side of the face had a well-defined, hard, bony swelling, with no alteration of skin colour, lymphadenopathy or any other alteration.

At the intraoral examination, there was an increase in volume in the region of end of sulcus from the canine to right mandibular ramus region with erasure of the vestibular sulcus. Of hard consistency to the palpation, with oral mucosa of normal appearance, smooth surface, regular and without ulcerations.

After analysing the patient's data, a provisional diagnosis of MFD was made, and differential diagnoses of hyperparathyroidism and Albright's syndrome were considered. The patient was referred for further laboratory investigations where her complete blood count and serum calcium, alkaline phosphatase, T3, T4, and thyroid stimulating hormone levels were within normal limits.

Computed tomography revealed an expansile lesion appearing as a hazy opacity and small soft tissue density areas seen in neck, angle and adjacent body of mandible on right side.

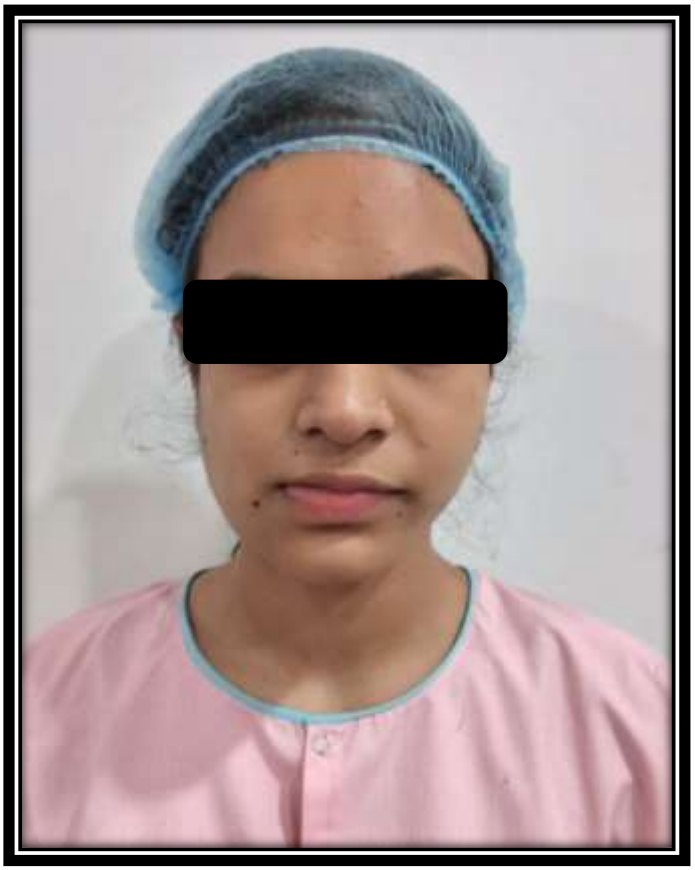
No abnormal periosteal reaction or cortical reaction is seen. No appearance of masseter muscle is seen on right side.

The mandible of left side appears normal.

Rest of the muscular structures appear normal. The submandibular planes are normal.

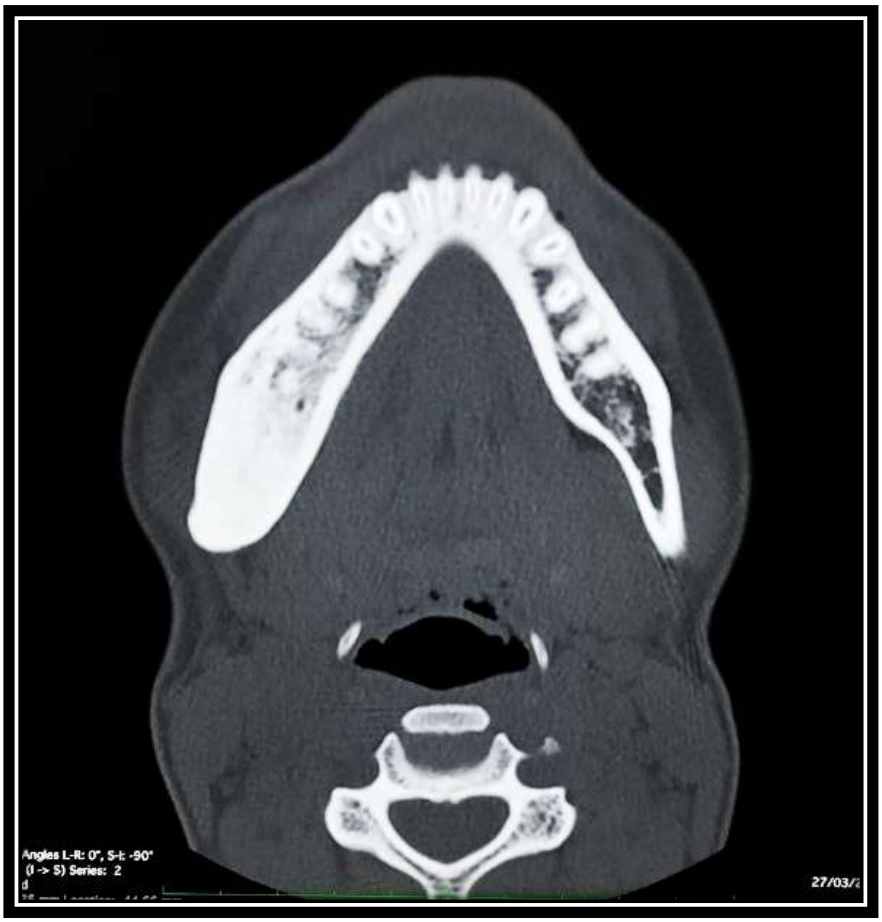
No lymphadenopathy was noted.

3D reconstruction offers a better three-dimensional understanding of the lesion, being more accurate in relation to the volume, density and involvement of adjacent anatomical structures, which facilitates the treatment plan.

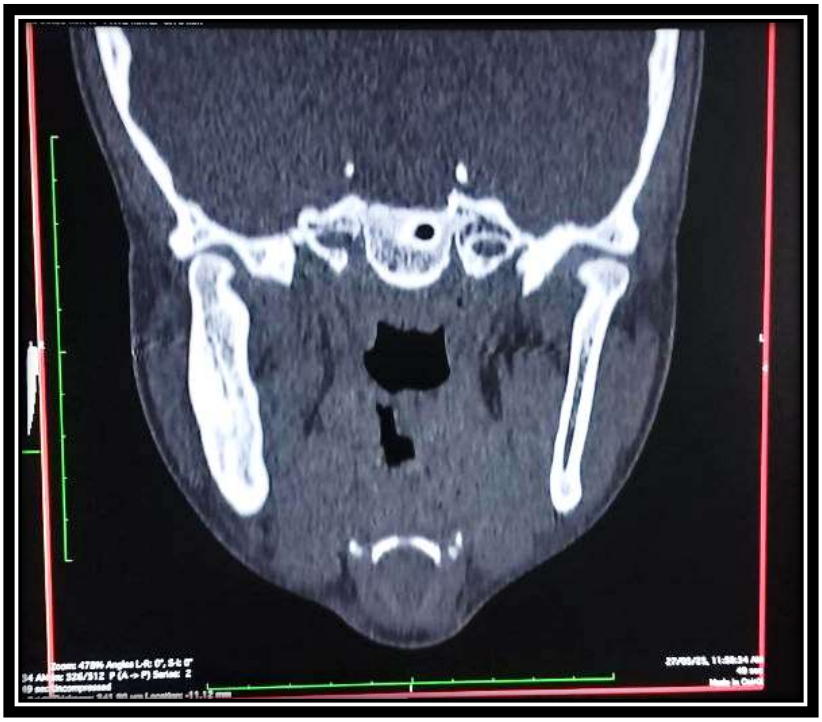


Clinical photograph showing unilateral swelling over the right side of the face.

Basal view showing swelling over right mandibular lower border

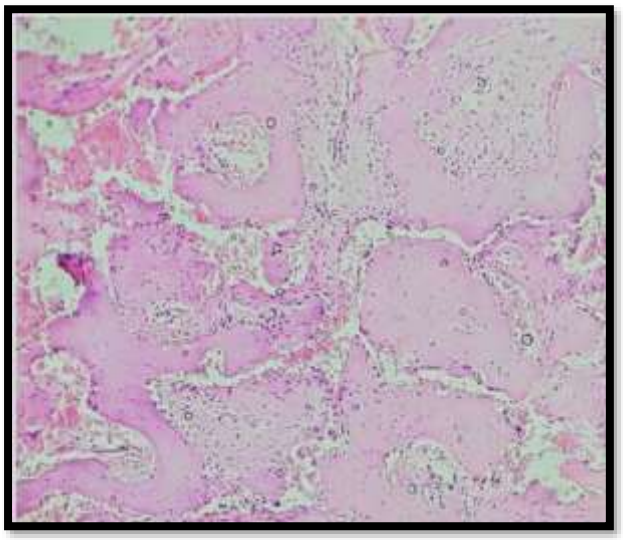


Computed tomography scan demonstrating enlargement of the right mandibular bony expansion



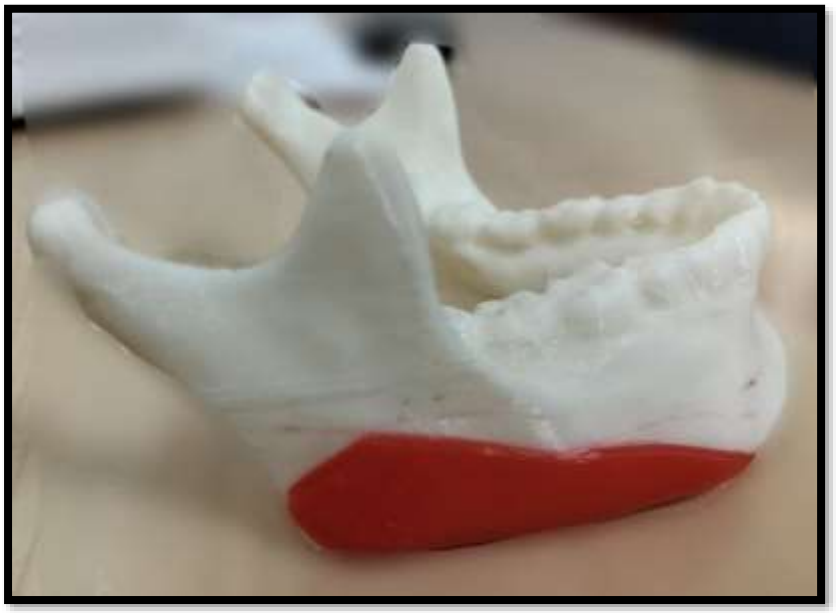
Computed tomography reveals a hazy opacity appearance in the buccal and lingual bone expansion regions and relatively well-defined borders.

An incisional biopsy was advised on the affected side, and histopathological analysis showed the presence of bone trabaculae interspersed with fibrous stroma. Detailed analysis under high magnification revealed the presence of bone trabaculae with entrapped osteocytes and osteoblastic lining, which were suggestive of FD



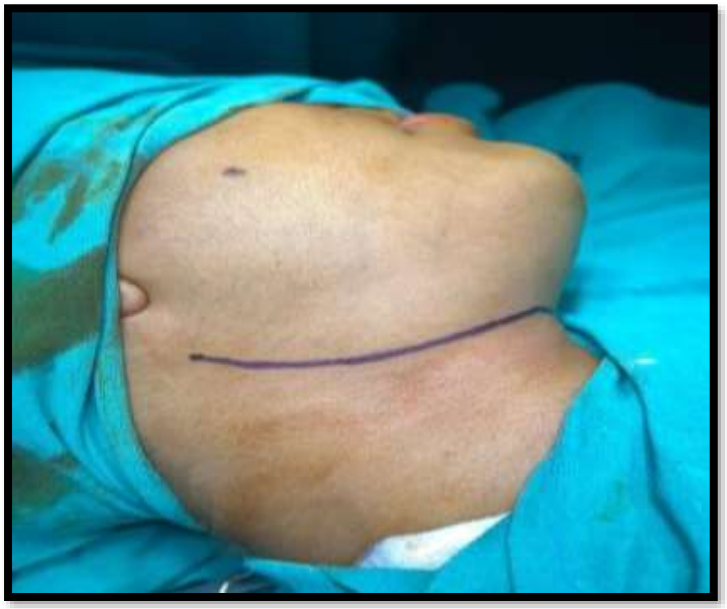
After reviewing all the findings, a guided surgery was decided using, physical 3-D model consisting of the reconstructed and unaffected sites was prepared for a reconstruction protocol.

The DICOM (digital imaging and communication in medicine) data acquired from the CT scans of the patient was converted to an STL 3D model The 3-D models were designed based to high resolution CT scans. Assessment of comparative functionality (stability of junction, mobility, mastication ability) and cosmetics



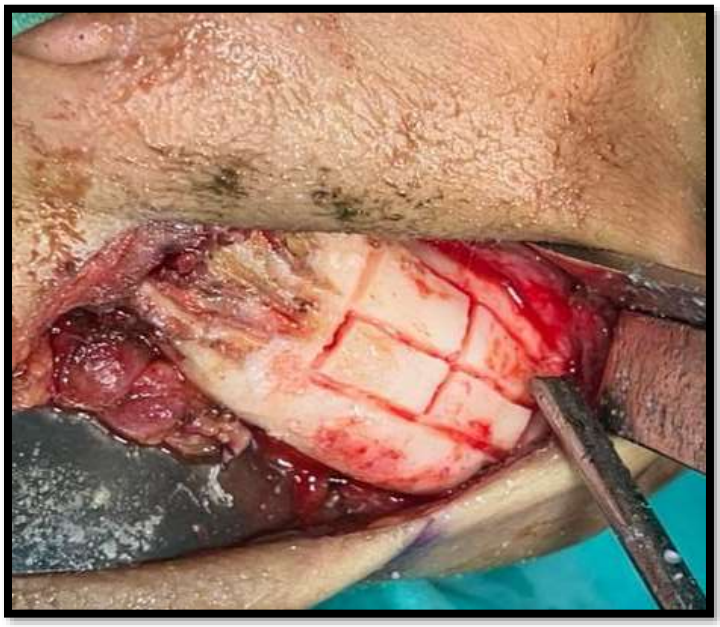
A mirror replication of unaffected site was replicated on right side, wax up was done and a guiding template was prepared using heat cure.

A guiding template showing accurate adaptation on mandibular surface.

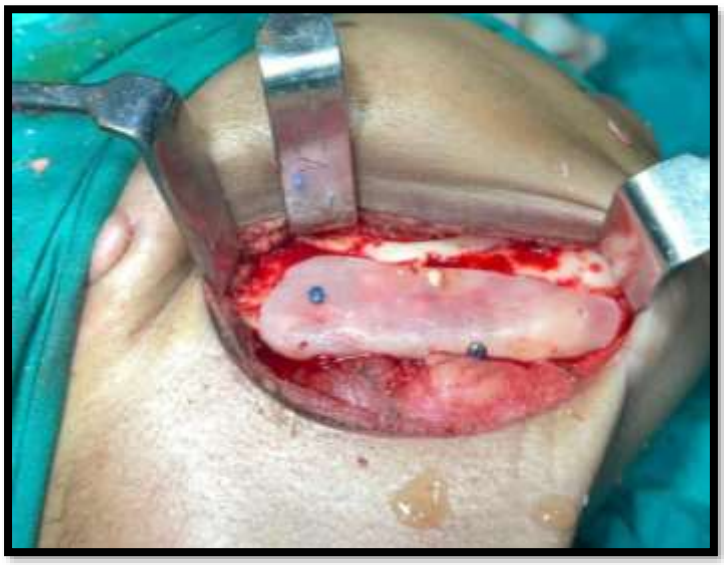


Submandibular incison was placed

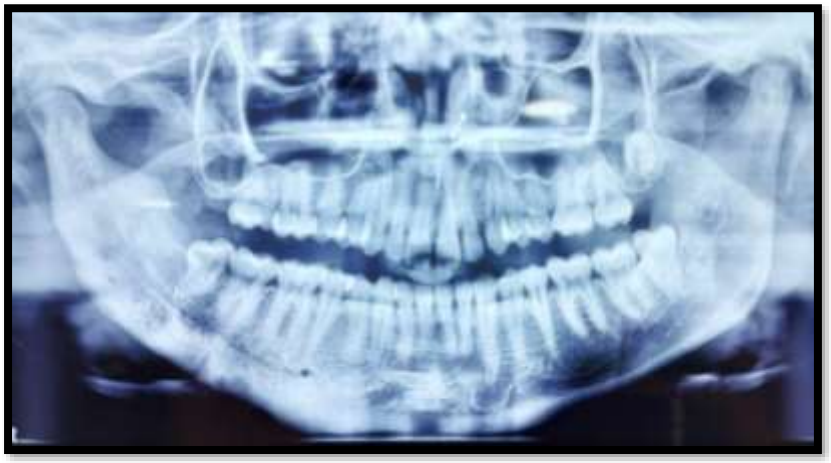
Bony mass was measured before removal



Osteotomy was performed



The guiding template was placed intraoperatively to assess the quantity of bone resection required. Following the removal of that template.



Post-operative OPG demonstrating virtually identical amount of bone recontour during surgery.

Discussion

Fibrous dysplasia is a frequently occurring, non-malignant fibro-osseous condition characterized by the replacement of normal bone with an excessive growth of fibrous connective tissue, along with irregular bony trabeculae. The precise cause of fibrous dysplasia remains uncertain, but it is likely associated with genetic predisposition, potentially involving mutations in the GNAS1 gene (guanine nucleotide binding protein), located on chromosome 20q13, or abnormalities in AMPc. These genetic factors may lead to increased melanocyte proliferation, resulting in the development of café au-lait spots and hyperactivity in affected endocrine organs.

Fibrous dysplasia affects both genders equally, with the average age of onset reported as 27 years in 69 patients, as observed by Zimmerman[2]. According to Abdulai et al[3]., fibrous dysplasia is twice as common in the maxilla compared to the mandible and is typically found in the posterior region. In the case at hand, the right posterior maxilla was affected. Polyostotic variants of the condition generally occur in children under the age of 10, while monostotic variants tend to affect individuals in their second and third decades of life. In the present case, the patient was 20 years old.

The diagnosis of FD is difficult in 36.3% of cases because no distinguishing symptoms are present, and 63.6% of patients complain of non-specific symptoms such as discomfort and/or edoema.[10] The maxilla is more commonly affected by MFD than the mandible. In our patient,

however, the mandible was afflicted whereas the maxilla was unaffected. The signs and symptoms of a tumour vary depending on its location. Patients may experience facial deformities, visual changes, nasal congestion, discomfort, and/or auditory impairments. Our patient reported pain and swelling at the affected jaw location. The majority of tumours appear in the premolar region and progress to the third molar region; the anterior region is least impacted. In our case, we discovered similar results.

People's perceptions of some personal attributes, such as attractiveness, worthiness, intelligence, or personality, are significantly influenced by the appearance of the face [5]. In this perspective, some believe that the jaw arrangement is the second most influential facial feature after the eyes in how others perceive our outer qualities [6]. Any disruption of mandibular integrity, whether caused by traumatic events, congenital deformities, or as a result of ablative procedures, can result in considerable impairment and serious aesthetic, psychological, and functional difficulties .

The extra bone can be virtually removed and the defect rebuilt utilising a mirror image of the contralateral side as a guide. In the event of anterior defects, the reconstruction is often accomplished by importing a standard undamaged 3D model of a mandible with the appropriate size and configuration to be precisely inserted in the defect site and adjusted to restore the normal shape[9].

The aim of treatment should be correction of the deformity caused by the tumor for adequate esthetics. Surgical excision of the tumor including bone is a successful treatment modality. However, it leads to considerable functional and esthetic deficits as well as long-term postoperative complications. Other non-surgical treatment modalities such as bisphosphonates have been suggested. They reduce the osteoclastic activity bound to bone surfaces. Its use in adults has shown promising results in controlling FD-induced pain. However, their long-term use should be limited, as they can lead to bone necrosis and are contraindicated in pregnant women. The role of RANK ligand inhibitors (denosumab) in reducing pain and growth should be evaluated Malignant potential is high in patients with polyostotic FD compared to those with MFD[10]

Conclusion

Conservative MFD treatment should be explored, especially if the patient is young. One of the more conservative techniques to treating asymmetry is localised surgical excision of demineralized bone followed by osteoplasty and contour remodelling. These 3D-printed models can be utilised for preoperative planning, surgical practise, and patient education. Close observation and follow-up should be maintained.

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