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CASE REPORT

Craniofacial Fibrous Dysplasia with Facial Asymmetry, Canted Occlusion and Open Bite: A Case Report with 2 Years Follow-up

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ABSTRACT

Aim: Fibrous dysplasia is a benign fibro-osseous disorder of unknown etiology that may affect the craniofacial region causing significant impairment of facial esthetics and function. This paper reports a case of a 21-year-old male, which was investigated and diagnosed with craniofacial fibrous dysplasia involving an overgrowth over right maxillary and mandibular region associated with facial asymmetry, severe occlusal cant and anterior open bite. Management of the case included surgical excision, cosmetic contouring and orthodontic intervention. The case was reviewed over a period of two years, which demonstrate stable treatment outcome.

Keywords: Fibrous dysplasia, Canted occlusion, Open bite, surgery, Orthodontics.

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INTRODUCTION

Fibrous dysplasia (FD) is a benign dysplastic process of altered osteogenesis in which bone is replaced by a variable amount of structurally weak fibrous and osseous tissue that may occur within a single bone (monostotic) or multiple bones (polyostotic).[1] Fibrous dysplasia constitutes 5 to 7% of all benign bone neoplasm.[2] FD typically arises in the first or second decade as a slowly enlarging painless expansion of the involved bone(s).[3] There is no sex or race predilection.[4] FD can be classified into monostotic, polyostotic, craniofacial and syndromic types.[5] The syndromic types arise with polyostotic FD and include McCune-Albright syndrome (triad of polyostotic FD, café-au-lait spots and multiple endocrinopathies) and Mazabraud syndrome (polyostatic FD and intramuscular myxomas).[6] Fibrous dysplasia is a congenital, non-heritable skeletal disorder. Although the precise etiology of FD is currently unknown,[7] several researches have proposed that the condition results from a mutation in a gene known as GNAS1. The mutation leads the bone cells to deposit fibrous tissue instead of healthy bone.[6]

Approximately, 25% of patients with FD have involvement of the craniofacial bones.[3] The craniofacial form of FD is not restricted to a single bone, but may be confined to a single anatomic site. These lesions affect primarily the maxilla but may also involve mandible, cross sutures into the sphenoid, zygoma, temporal, orbital and frontonasal bones and base of skull. Although multiple contiguous bones are involved, craniofacial FD is usually a unilateral process.[8]

Clinically, a slow growing osseous expansion of the affected bone is the most common presenting symptom giving a nontender facial asymmetry of variable degree.[6] Progressive growth may cause significant functional and esthetic impairment. As a rule of thumb, lesion usually stops growing when skeletal growth ceases, but cases of continual enlargement have been reported.[9] The overlying skin or mucosa appears normal and teeth may show migration or displacement but are rarely loosened. Laboratory assay of serum calcium and phosphate levels of affected patients are normal, but alkaline phosphatase level may be slightly elevated. Conversely, lack of an elevation should not rule out FD.[9]

Radiographic features vary depending upon the stage of the disease. Early onset lesions are radiolucent and later progressively calcify, culminating in a 'ground glass' or mottled mixed radiolucent/radiopaque pattern. Critical to the diagnosis is the fact that fibrous dysplasia fails to manifest any discrete margins in the jaws (although usually well defined elsewhere in the skeleton), rather, the lesional bone subtly blends into the surrounding normal appearing bone,[6] this feature is helpful in distinguishing FD from other

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radiopaque lesions.[10] Expansion occurs, usually fusiform in the mandible and more complex in the maxilla, reflecting the more complex anatomy of the latter; in both cases, the underlying morphology of the bone is largely preserved. Because FD has no dental relationship, it is not confined to the alveolar processes; and may superiorly displace the inferior alveolar canal.[10,11] Radiographic features of FD have been subclassified by some researchers into three different patterns: pagetoid type (56%), sclerotic type (23%) and the radiolucent type (21%). Sclerotic type is most commonly seen in facial bones and bones of the base of the skull, the lytic and pagetoid types usually involve the calvarial bones.[4,12] CT is useful to assess the precise extent of the lesion in the complex facial skeleton, especially when there is orbital involvement or involvement of the neurovascular foramina of the skull base.[13] Scintigraphy is helpful in determining the activity and potential multicentricity of the lesion and is specifically helpful in diagnosing when plain radiographs are equivocal.[4]

The histologic appearance of FD depends on the stage of the lesion.[14] Early FD is characterized by irregular (curvilinear; Chinese characters) trabeculae of woven bone associated with a moderately cellular, haphazard proliferation of bland spindled-ovoid fibroblasts.[15] Lesional bone fuses with adjacent non-lesional bone.[16] Unlike in the long bones, FD of craniofacial bones can undergo maturation to lamellar bone with a decrease in stromal fibroblast cellularity.[15] Other changes that may be seen that are not seen in long bone FD include prominent osteoblastic rimming and cementum-like calcifications.[17]

Treatment of FD involves use of both medications and surgical methods. Medications include drugs like Alendronate, Pamidronate and Calcitonin.[18] Small and well-delineated lesions can be treated with curettage and enucleation or excision. If the lesion is extensive and presents with malocclusion and jaw disproportion, conservative recontouring surgery and repositioning of the affected jaw bone aiming at esthetic correction or functional improvement are recommended. In children, it is wise to delay the surgical procedure until after puberty so that the lesion tends to become static.[19] Radiotherapy in treatment of FD is contraindicated, because of the possibility of radiation-induced sarcomas.[1]

Based on a case report, clinical pathological features and management of a case of craniofacial FD with canted occlusion and open bite is presented.

CASE REPORT

Case History

A 21-year-old male reported to the Department of Oral and Maxillofacial Surgery, Government College of Dentistry, Indore, MP, India, with complaint of swelling on right side of the face since 6 to 7 years. Initially, swelling was small and gradually increased to the present size. There was no history of pain, paresthesia, trauma, epistaxis, loosening of teeth, abscess formation, trismus or diminished vision. There was no other swelling in the body and café-au-lait spots were not seen. General and systemic examination revealed no abnormality. There was no family history with similar findings.

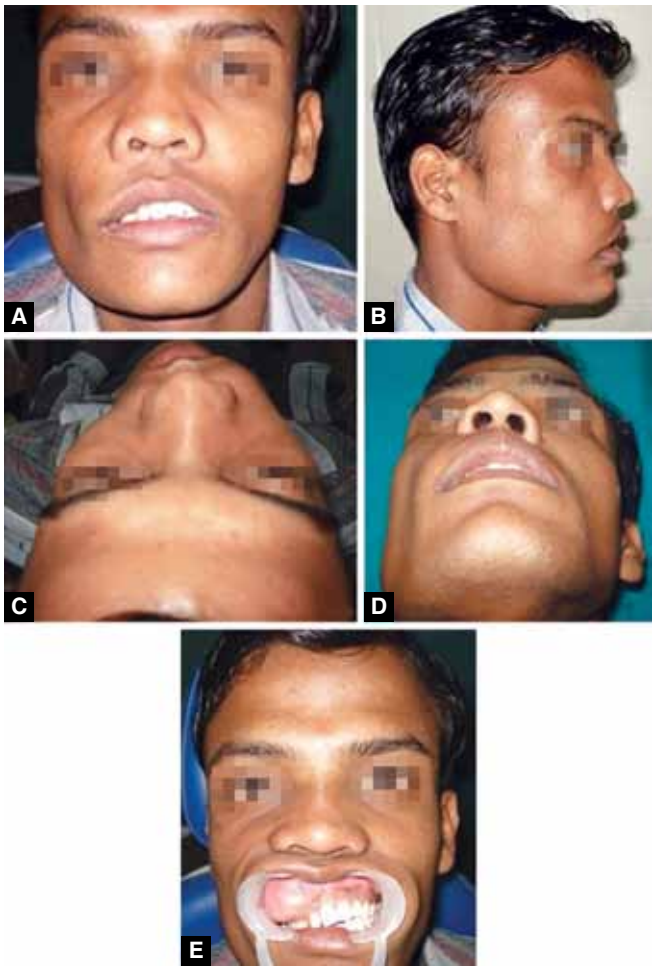
Clinical Evaluation

On extraoral assessment a diffused unilateral swelling was present on the right face extending vertically from side and below orbit to below angle of mouth and horizontally from philtrum below ala tragus line extending posteroinferiorly toward ramus, angle and body of mandible (Figs 1A and B). The lips were incompetent with an 8 mm incisal show at rest associated with canting of teeth and anterior open bite of 1.75 mm (Figs 1A and E). The chin position was slightly deviated to the left. The upper dental midline was deviated 1.8 mm to the left (Fig. 1A). On lateral profile view, patient had slight facial convexity with normal nasolabial angle, incompetent and protruded upper and lower lips, and slightly deep mentolabial fold. There was asymmetric broadening of right mandibular ramus and flattening of gonial angle (Fig. 1B). Extraoral submental view confirmed the right maxillary bulge between the side of nose and upper lip with flattening of the zygomatic prominence, broadening of right side of mandibular ramus, angle and thickening of the right inferior border of mandible (Figs 1C and D). The maximum mouth opening was normal at 45 mm. The speech was normal and there was no evidence of temporomandibular joint dysfunction. Breathing was both nasal and oral.

On palpation the consistency was bony hard, nontender and no rise in temperature was noted. Intraoral examination demonstrated a pronounced swelling, bulging from the upper right gingivobuccal sulcus extending labially from above maxillary right central incisor to maxillary right first molar (Fig. 1E). The right buccal cortical plate was expanded; overlying mucosa appeared normal, firm and nontender. The maxillary arch on occlusal view appeared slightly asymmetric. The occlusion was class I with distally tipped crown of maxillary right canine and mesially tipped crown of maxillary right first premolar and spacing between maxillary right canine and lateral incisor. The upper occlusal plane showed an S-shaped canting toward right creating an anterior open bite (Fig. 1E).

Radiographic Evaluation

The orthopantomogram revealed a ground glass appearance with poor discernible borders, loss of normal trabecular



Figs 1A to E: Pretreatment facial and intraoral photographs: (A) Front view, (B) lateral view, right side, (C) coronal view, (D) submental view and (E) intraoral front view

pattern of bone and significant expansion with thinning of outer cortex of the right mandible extending from the mesial of mandibular right first premolar root tip till the right condyl (involving the part of body, angle, ramus of mandible and coronoid process). Lower border of mandible showed downward displacement and flattening of gonial angle. The periodontal ligament space and lamina dura were diminished of the affected side, although there was no evidence of root resorption on the OPG, the roots of mandibular right second molar appeared diminished in length as compared to the contralateral mandibular left second molar. The maxillary involvement was not clearly seen on the OPG except displacement of roots of maxillary right first premolar and maxillary right canine (Fig. 2).

CT scan revealed dysplastic bone affecting most of the right facial skeleton (Figs 3A to J). Features of expansion with ground glass appearance with widened diploic spaces were noted in the frontal bone, zygoma, nasal bone and right maxilla including palatine process and frontal process of maxilla and right side of mandible. Frontal, ethmoid and sphenoid air cells appeared to be obliterated on the right side (Figs 3A to C). Bone scintigraphy was also carried

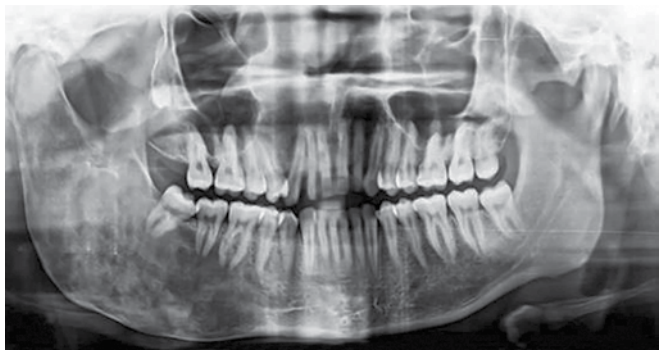


Fig. 2: Pretreatment panoramic radiograph

out which ruled out involvement of any other bones of the body (Figs 4A and B).

Lateral cephalometric analysis indicated a skeletal class I relationship with increased lower anterior face height. The maxillary incisors were slightly proclined and anterior open bite was evident. The soft tissue analysis showed protruded and incompetent upper and lower lips, normal convexity and normal nasolabial angle. The cervical vertebral maturation staging indicated completion of growth (Figs 5A and B, Table 1).

Posteroanterior cephalogram was assessed using Grummon's analysis, which indicated asymmetry in the middle and lower third of face, right maxillary canting with slight compensatory mandibular canting. Upper dental midline was deviated by about 1.8 mm to the left (Figs 6A and B).

The blood and urine investigations were within the reference range. Based on the clinical and radiographic findings a provisional diagnosis of craniomaxillofacial fibrous dysplasia of right face was made.

Table 1: Lateral cephalometric measurements of the patient

Measurements	*Average value*	*Pretreatment*	*Post-treatment*
Skeletal			
SNA (°)	82 ± 4	84	84
SNB (°)	80 ± 2	82	82
ANB (°)	2 ± 2	2	2
SN-MP (°)	32 ± 5	29	29
UAFH:LAFH (%)	45:55	49:72	49:71
Gonial angle (°)	128 ± 7	116	117
CVMI		VI	VI
Dental			
U1-SN (°)	102 ± 2	118	109
U1-NA (°)	22	34	24
U1-NA (mm)	4	9	5
L1-MP (°)	95 ± 7	91	92
L1-NB (°)	25	24	24
L1-NB (mm)	4	5	5
Soft tissue			
Nasolabial angle (°)	102 ± 8	90	92
Interlabial gap (mm)	2 ± 2	9	1
Upper lip to E line (mm)	–4 ± 2	0	–3
Lower lip to E line (mm)	–2 ± 2	+2.75	–1

Treatment Objectives

The following treatment objectives were defined:

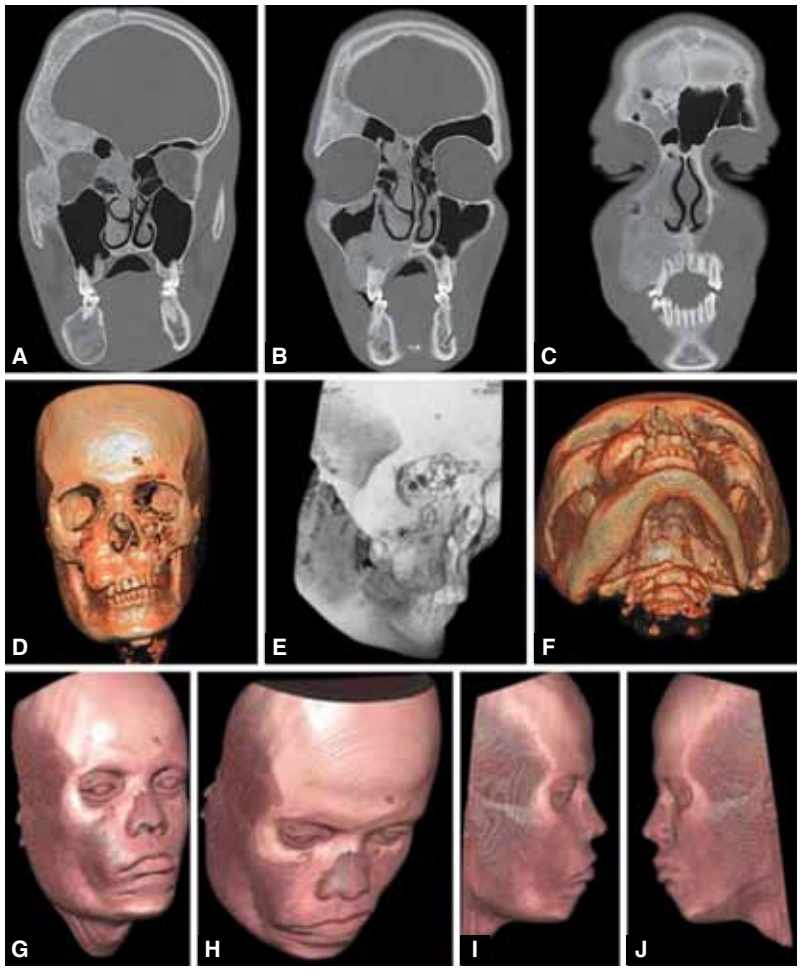
1. To correct facial asymmetry.
2. To align and level all teeth to favorable function and esthetics.
3. To obtain optimum overbite and overjet.
4. To correct upper dental midline shift.
5. Watch out for any growth in the long-term.

Treatment Plans

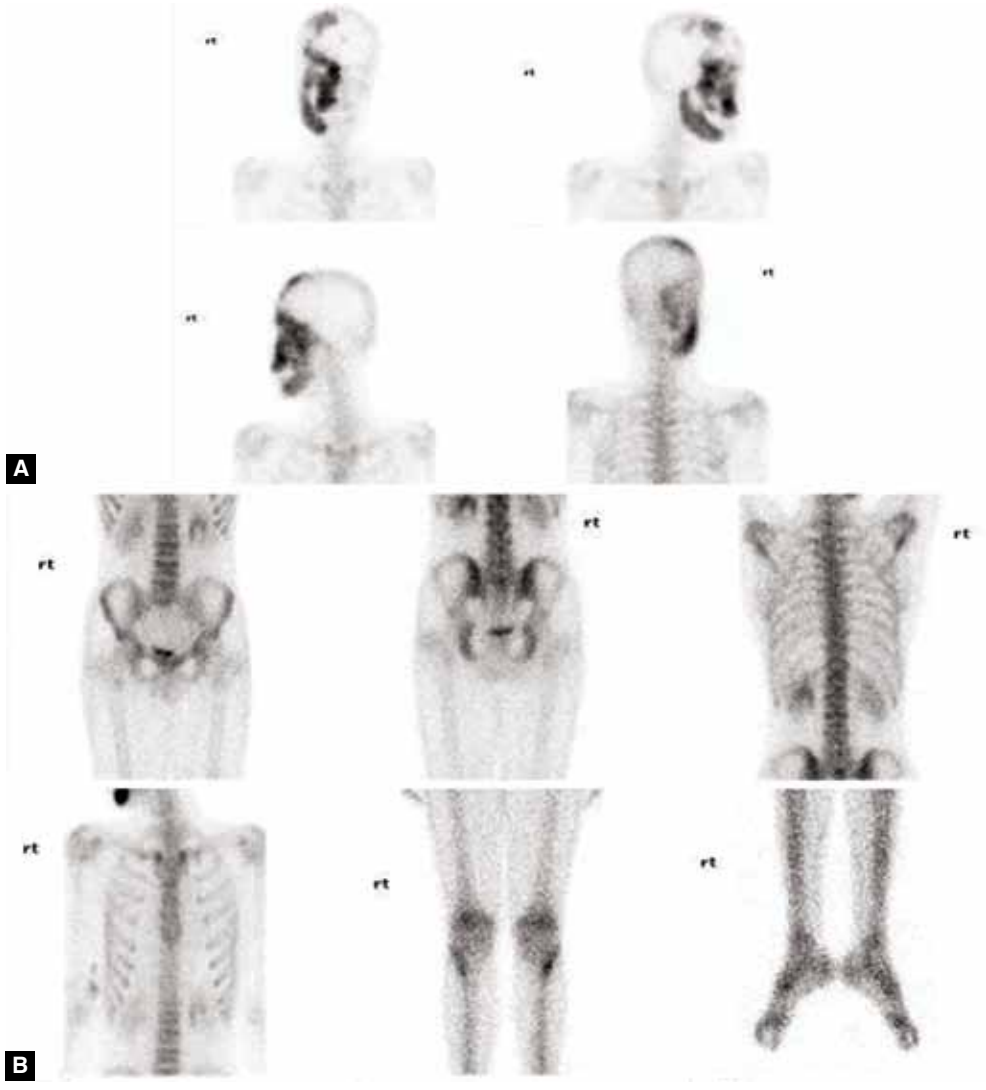
1. Conservative surgical excision of the overgrowth and contouring the affected maxillary and mandibular region.
2. Postsurgical fixed orthodontics for functional and esthetic correction of occlusion.
3. Long-term follow-up.

Treatment Progress

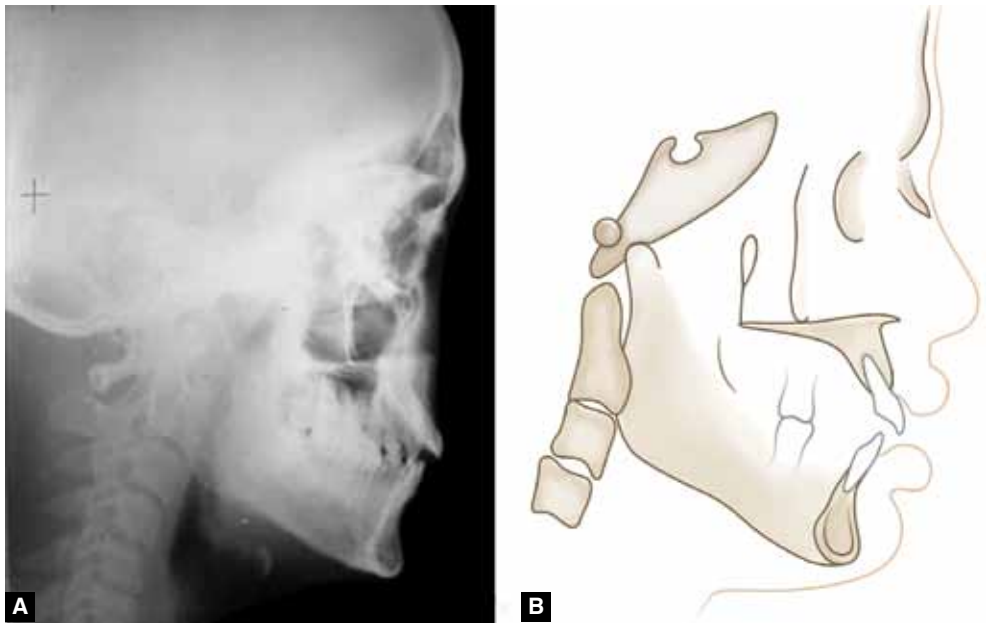
Treatment was begun with surgical excision of the affected bony tissue in the maxilla and mandible with cosmetic contouring under general anesthesia. Sublabial intraoral approach was performed for the maxillary lesion (Fig. 7A) and submandibular extraoral approach for the mandibular lesion (Figs 7B and C). The excised bone segments (Fig. 7D) were sent for histopathology, which confirmed the lesion to be fibrous dysplasia (Figs 8A to D). The postoperative recovery was uneventful and the patient was discharged from the hospital within 48 hours after surgery. After 2 months, fixed orthodontic treatment was initiated with 0.022 × 0.28" MBT appliance. It was observed that post surgery, the anterior open bite had increased to about 4 mm, and mandibular right canine was below the occlusal plane. The patient had also developed secondary tongue thrust. Maxillary canting and asymmetric gingival show and lip incompetence was apparent as before (Figs 9A to C). Simple mechanics of altered bracket positioning and placement of 0.014" NiTi wire was opted. Brackets were bonded slightly incisally on the upper right teeth as compared to the left side to facilitate correction of occlusal cant (Figs 10A to C). Bracket on mandibular right canine was placed gingivally to cause its extrusion. Sequentially, after 0.014" NiTi wire, the upper and lower archwires were changed to stainless steel wires of 0.16", 0.16" × 0.22" and 0.19" × 0.25" respectively. Patient was also asked to do regular tongue exercises. He showed great improvement over a period of 6 to 7 months



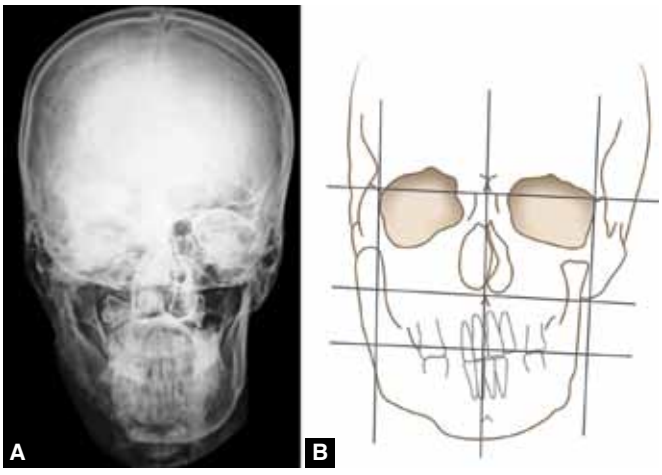
Figs 3A to J: Pretreatment CT scan



Figs 4A and B: Pretreatment bone scintigraphy



Figs 5A and B: Pretreatment lateral cephalometry: (A) Radiograph and (B) Tracing



Figs 6A and B: Pretreatment frontal cephalometry, (A) Radiograph and (B) tracing

(Figs 11A to C and 12A to E). A lingual bonded retainer was placed on the upper anterior teeth from maxillary right canine to maxillary left canine, which was later removed after 8 months on patient's insistence.

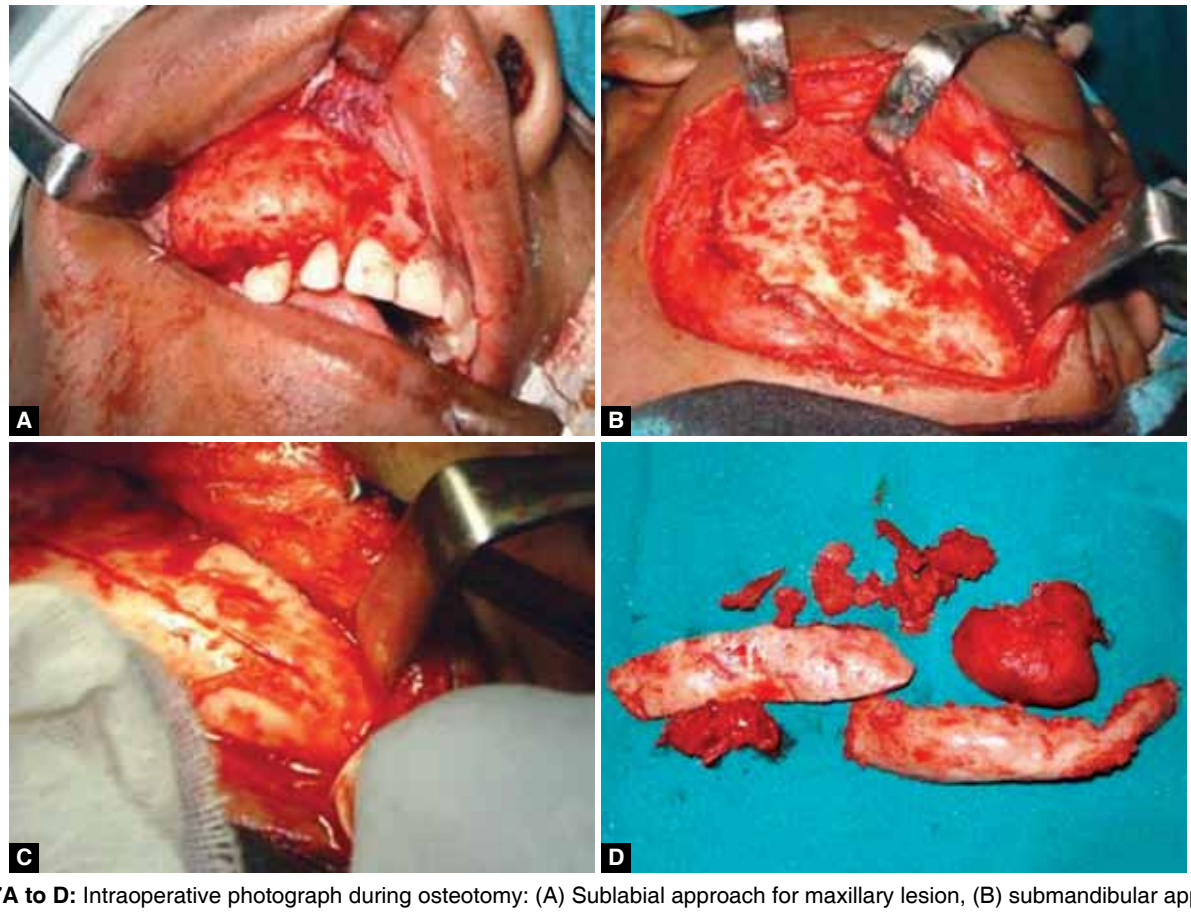
RESULTS

As a result of surgical and orthodontic treatment, the facio-dental asymmetry and occlusal canting was addressed. Favorable overbite, overjet and soft tissue relationship was achieved. Cephalometric evaluation confirmed improvement in dental and soft tissue parameters. The upper incisor angulation was improved and upper and lower lips were harmonized (Figs 13A and B, Table 1). Posteroanterior cephalometric radiograph revealed that the upper midline shift and occlusal canting was corrected (Figs 14A and B). The patient's satisfaction with both facial and occlusal results was very high and allowed him to resume a social life that had previously been severely compromised. Periodic follow-up every 6 months over a period of 2 years was carried out, which confirmed stable treatment outcome with no evidence of active growth (Figs 15A to C and 16A to E).

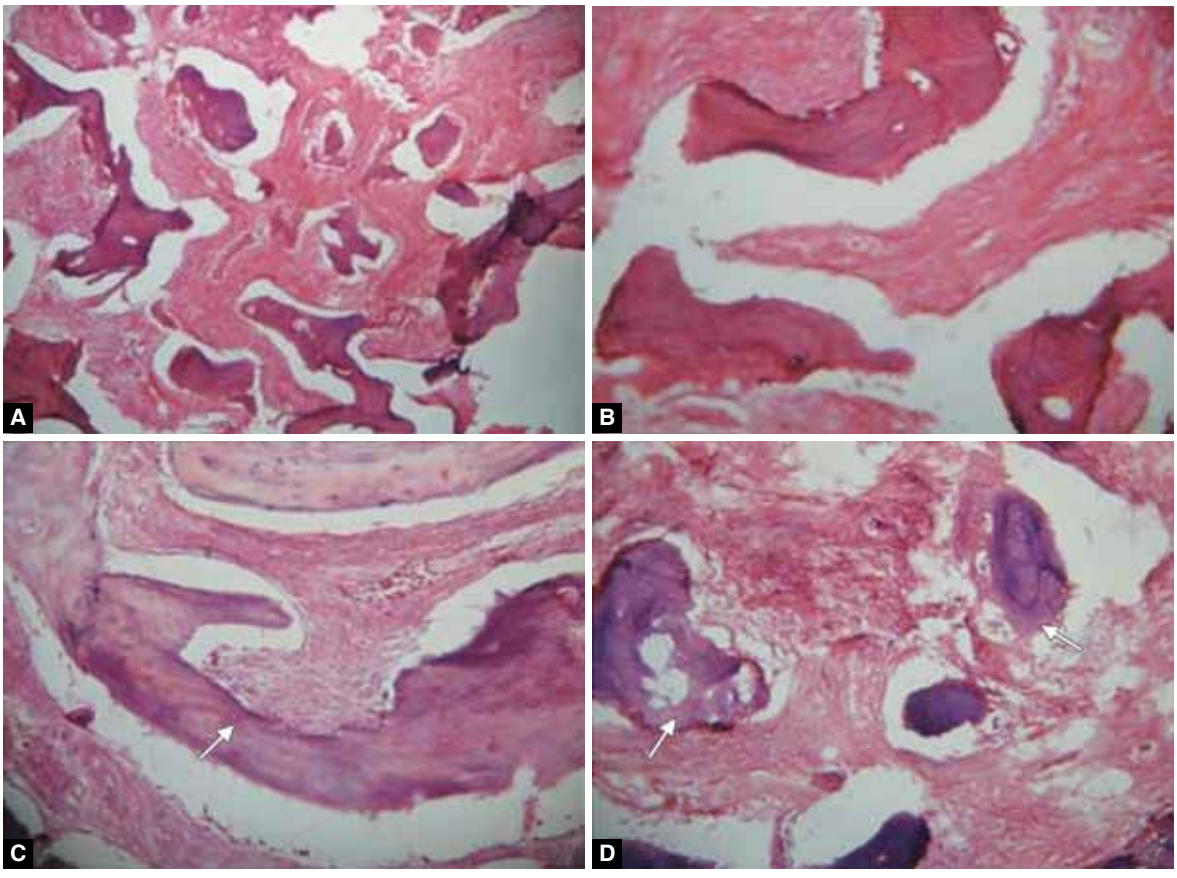
DISCUSSION

Isolated cases of craniomaxillofacial fibrous dysplasia are rare and can be difficult to differentiate from other benign and malignant bone disorders.[20] The differential diagnosis includes chondroma, simple bone cyst, non-ossifying fibroma, osteofibrous dysplasia, Paget's disease of bone, osteoblastoma, chondroblastoma, fibromyxoma of bone, adamantinoma, and low-grade intramedullary osteosarcoma.[2]

The diagnosis in the reported case was suspected by clinical and radiographic findings on the basis of which the treatment was instituted. The 21-year-old patient reported in this article had the lesion slowly enlarging on the right side of the face without causing any discomfort since he was



Figs 7A to D: Intraoperative photograph during osteotomy: (A) Sublabial approach for maxillary lesion, (B) submandibular approach for mandibular lesion, (C) mandibular osteotomy cut and (D) osteotomized bone segments after surgery

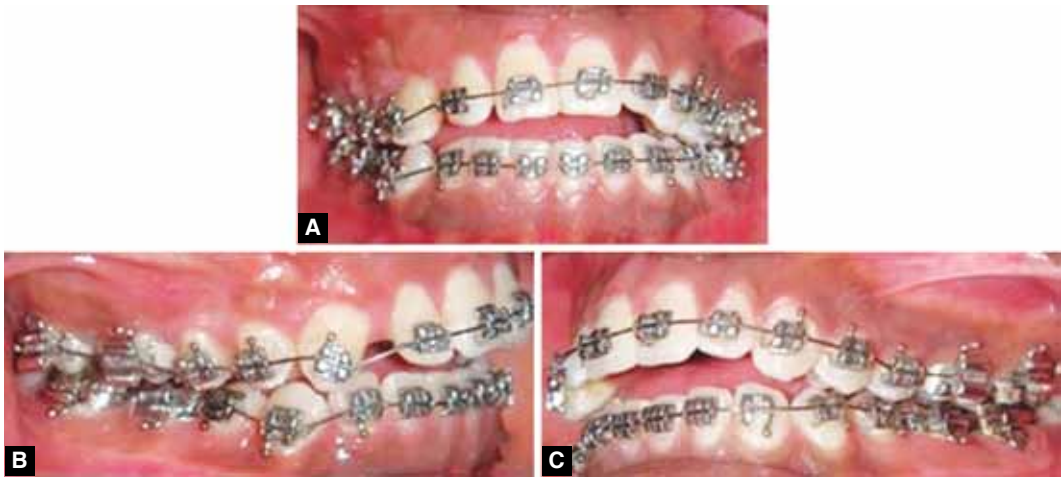


Figs 8A to D: Photomicrographs of the excised tissue: (A) Irregular trabeculae of woven (immature) bone in fibrous stroma (H&E, 100×), (B) irregular trabeculae of immature bone in fibrous stroma (H&E, 400×), (C) bone undergoing maturation to lamellar bone (arrow) (H&E, 400×) and (D) cementum-like calcifications (arrow) (H&E, 400×)

Figs 9A to C: Extraoral photographs, 2 months postsurgery: (A) Extraoral front view, (B) extraoral lateral view right side and (C) extraoral 45° smile view

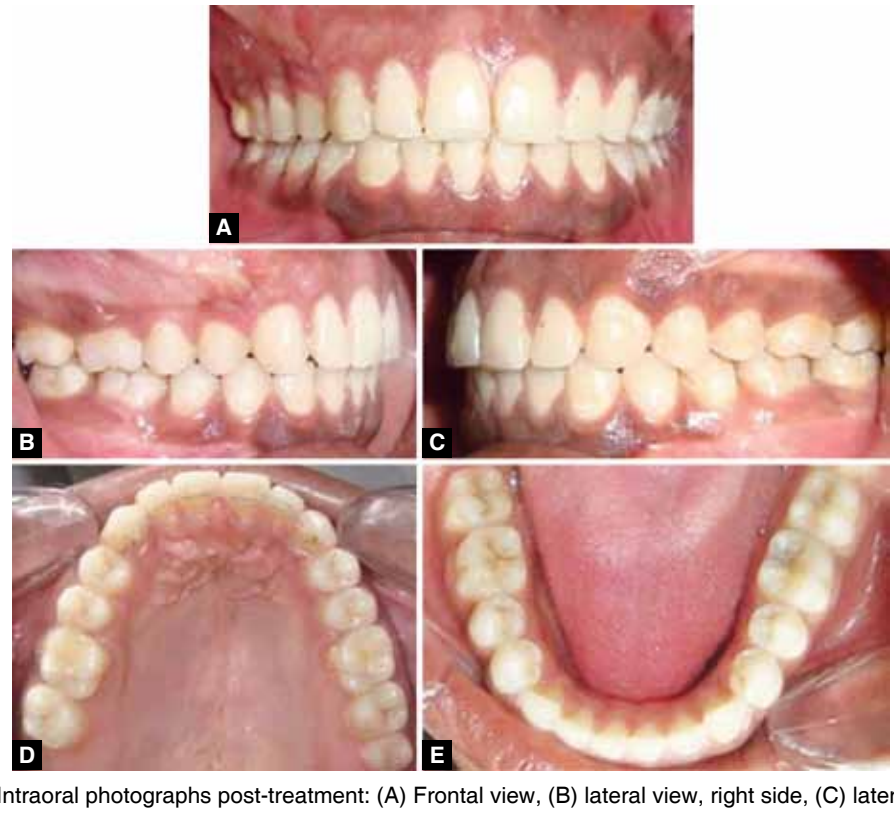
young. The panaromic radiograph and CT scan revealed a 'ground glass' appearance of the majority of the bones of the right facial skeleton. The histopathology of the excised tissue demonstrated classical features of craniofacial fibrous dysplasia. Irregular shaped (Chinese character) trabeculae of immature woven bone were seen embedded within a moderately cellular fibrous stroma (Figs 8A and B). At few places in the section, the bone was seen to undergo maturation to lamellar bone (Fig. 8C). Characteristic cementum-like calcifications were also seen (Fig. 8D). Such unusual findings differentiate craniofacial fibrous dysplasia from fibrous dysplasia of long bones. All these were compatible with diagnosis of craniofacial fibrous dysplasia.

In majority of cases with FD, surgical reduction for cosmetic or functional reason is a treatment of choice. It is usually recommended to delay the surgical procedure until

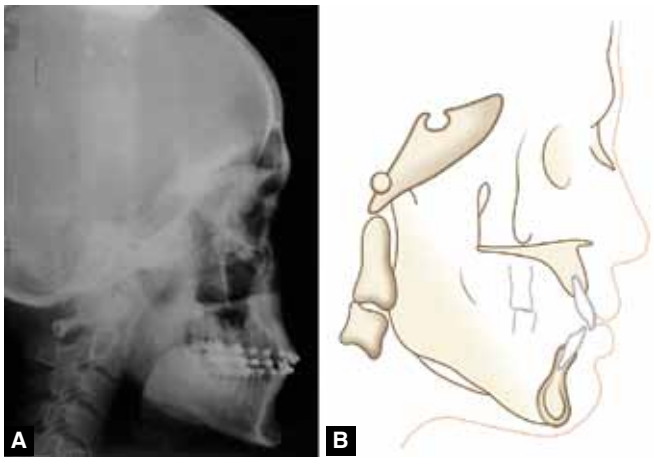


Figs 10A to C: Intraoral photographs showing fixed orthodontic appliance placement after 2 months postsurgery. Anterior open bite has increased postsurgery: (A) Front view, (B) lateral view, right side and (C) lateral view, left side

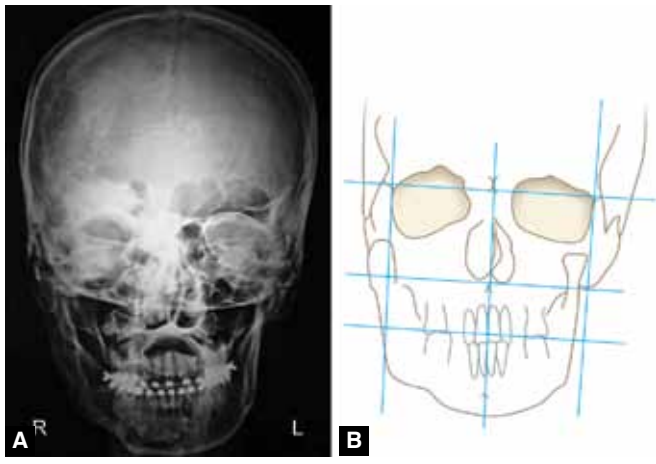
Figs 11A to C: Extraoral photographs post-treatment: (A) Frontal view, (B) lateral view, right side and (C) 45° smile view



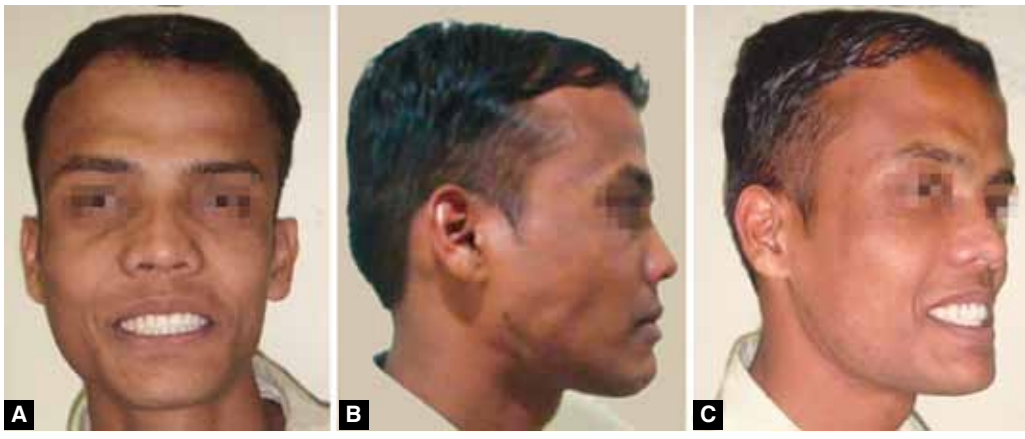
Figs 12A to E: Intraoral photographs post-treatment: (A) Frontal view, (B) lateral view, right side, (C) lateral view, left side, (D) upper occlusal view and (E) lower occlusal view



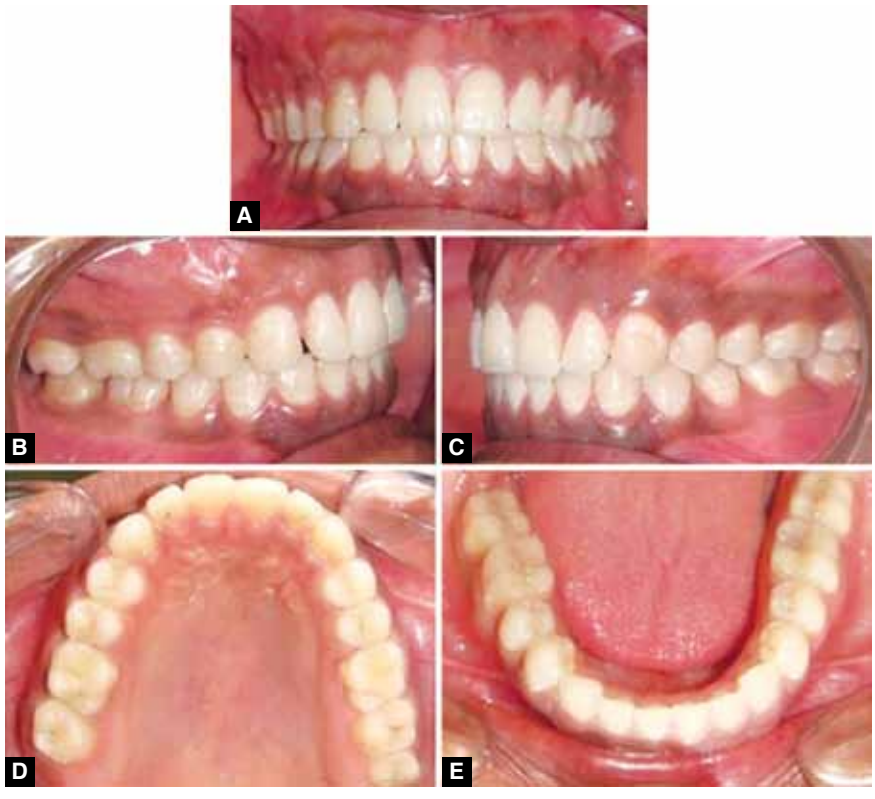
Figs 13A and B: Post-treatment lateral cephalometry: (A) Radiograph and (B) tracing



Figs 14A and B: Post-treatment frontal cephalometry: (A) Radiograph and (B) tracing



Figs 15A to C: Extraoral photographs, 2 years post-treatment: (A) Frontal view, (B) lateral view, right side and (C) 45° smile view



Figs 16A to E: Intraoral photographs, 2 years postsurgery: (A) Frontal view, (B) Lateral view, right side, (C) lateral view, left side, (D) upper occlusal view and (E) lower occlusal view

skeletal growth has completed, so that the lesions tend to be more static.[19] Assessment of the disease activity or stability can be based on subjective evaluation of the patient with regard to the patient's age, CVM stage and alteration in size of the lesion. For the present case, the result after evaluation of the patient indicated static growth of the lesion. Hence, surgical intervention followed by fixed orthodontic treatment was implemented. Surgical procedure included conservative surgical excision and cosmetic contouring of the affected maxillary and mandibular segment only. The surgery was uneventful, however, after healing it was observed that anterior open bite worsened. This may be attributed to be due to the shrinkage of tissue during the healing process. However, orthodontic treatment initiated posthealing with 0.022" MBT appliance using light forces, simplistic mechanics of atypical bracket positioning and NiTi wire placement and three dimensional control of the orthodontic appliance reaped satisfactory results with restoration of function, occlusion and esthetics. The rapidity with which the orthodontic movements were achieved may be ascribed to the pliability of bone postsurgery.

It has to be admitted that treatment selected for the patient might be considered little conservative as not all the affected bone was removed. The treatment plan was based on the belief that growth of the lesion apparently appeared to have ceased, hence, cosmetic correction and later long-term follow-up should be the preferred option, which also meets the chief complaint of the patient. Two-year follow-up, every 6 months demonstrated stability of achieved occlusion and facial esthetics.

Few studies have suggested that approximately 20% of the lesions of FD would continue to grow after treatment, unless radical excision was performed.[21] Cases are also reported to the concomitant development of aneurysmal bone cyst or less commonly, malignant transformation.[14] Such deviant behavior of FD suggest that our understanding of the lesions is still incomplete. The major problem encountered in diagnosing and management of FD lesions is that there is paucity of detailed literature on it and there are only few long-term follow-up case reports on it.[22]

The purpose of this article is to report our experience of a case of FD managed using surgical and orthodontic intervention and carefully followed over 2 years.

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