Case Report

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Fibrous Dysplasia Involving the Right Maxilla: A Case Report and Review of Literature of the Radiographic Feature of Fibrous Dysplasia

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Abstract

Fibrous dysplasia is a common benign skeletal lesion that may involve one bone (monostotic) or multiple bones (polyostotic) and occurs throughout the skeleton with a predilection for the long bones, ribs, and craniofacial bones. The etiology of fibrous dysplasia has been linked to an activating mutation in the gene that encodes the alpha subunit of stimulatory G protein (Gsα) located at 20q13.2-13.3. Most lesions are monostotic, asymptomatic, and identified incidentally and can be treated with clinical observation and patient education. Surgery is indicated for confirmatory biopsy, correction of deformity, prevention of pathologic fracture, and/or eradication of symptomatic lesions. The use of cortical grafts is preferred over cancellous grafts or bone-graft substitutes because of the superior physical qualities of remodeled cortical bone.

Keywords: Fibrous Dysplasia; Lichtenstein-Jaffe's Disease; Maxillofacial; Mccune-Albright's Disease; Osteodystrophia Fibrosa; Osteitis Fibrosa Disseminata; Monostotic Form; Polyostotic Form; Craniofacial Form; Cherubism

Introduction

Maxillofacial fibro-osseous lesions (FOL) consist of lesions that differ, with the exception of fibrous dysplasia, to those found in the rest of the skeleton. FOLs of the face and jaws are cemento-ossifying dysplasia, fibrous dysplasia and cement ossifying fibroma. Radiology is central to their diagnosis because the pathology for all FOLs is similar, although they range widely in behaviour, from dysplasia, hamartoma to benign neoplasia with occasional recurrence. Furthermore, once diagnosed the management of each is different. Maxillofacial FOLs are of particular interest to the radiologist because they emphasize the central role of the radiologist in the diagnostic process. The late Charles Waldron wrote "In absence of good clinical and radiologic information a pathologist can only state that a given biopsy is consistent with a FOL. With adequate clinical and radiologic information most lesions can be assigned with reasonable certainty into one of several categories". Despite the advances in the understanding of these conditions, fibro-osseous lesions continue to present problems in classification, diagnosis, and management due to multiple histological and radiographic similarities. The objective of this article is to review the most current, radiographic, and molecular studies of Benign Fibro-osseous Lesions with the help of a case.

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Case Report

15 year old male patient comes with a chief complaint of painless swelling over his left side of the face (fig. 1) since 2 years. Swelling was insidious in onset which gradually progressed to the present

size. Initially swelling was small to start with. Swelling was not associated with pain or fever. Swelling became visible since 2 years. No associated nasal discharge, mild heaviness present over the left side of the face. No history of change of vision during the course of swelling. On examination swelling was present over the left midface region with diffused margins extending from the ala of the nose up to the zygoma and from intra orbital ridge up to the line joining the angle of the mouth and tragus of ear. Skin over the swelling appears to be normal with no colour change swelling felt bony hard uniformly non-tender non-mobile. No evident lymphadenopathy Intra orally swelling (fig. 2) was present on the left upper jaw extending from 23 to 26 on the buccal aspect. Swelling uniformly extends over the buccal alveolar ridge with vestibular obliteration. The mucosa appears normal on palpation bony hard in consistency. We came to a provisional diagnosis of benign lesion involving the left maxilla which encompasses a wide range of conditions of odontogenic and non-odontogenic origin. Odontogenic keratocysts, ameloblastoma, ameloblastic fibro-odontoma, fibrous dysplasia, ossifying fibroma. Radiographs were taken. Ioparreveals altered trabeculations giving it a ground glass appearance. This is a loss of trabeculations and loss of lamina dura (fig. 4). Opg (fig. 3) reveals full complement of upper and lower teeth with well-defined radio-opacity over the left maxillary sinus and the floor of the sinus is not evident with mild distal tipping of the roots of 26. PNS (fig. 5) reveals complete obliteration of the left side maxillary sinus with a homogeneous well defined radio opacity. There is deviation of the nasal septum towards the right side. CT (fig. 6) reveals obliteration of the maxillary sinus which has a ground glass appearance uniformly with HU of 678 of blending with the surrounding walls of the antrum and there was no post contrast enhancement of the peripheral rim differentiating it from ossifying fibroma. Patient was subject to biochemical investigation of alkaline phosphatase which was 266 IU/L. Skeletal survey was done to rule out any other bone involvement and rule out presence of any café au lait spots. With all this the final diagnosis was given as monostotic

Fig 1: Diffuse Swelling present over the left mid face region extending from the infra orbital ridge up to the upper border of the lip superio inferiorly

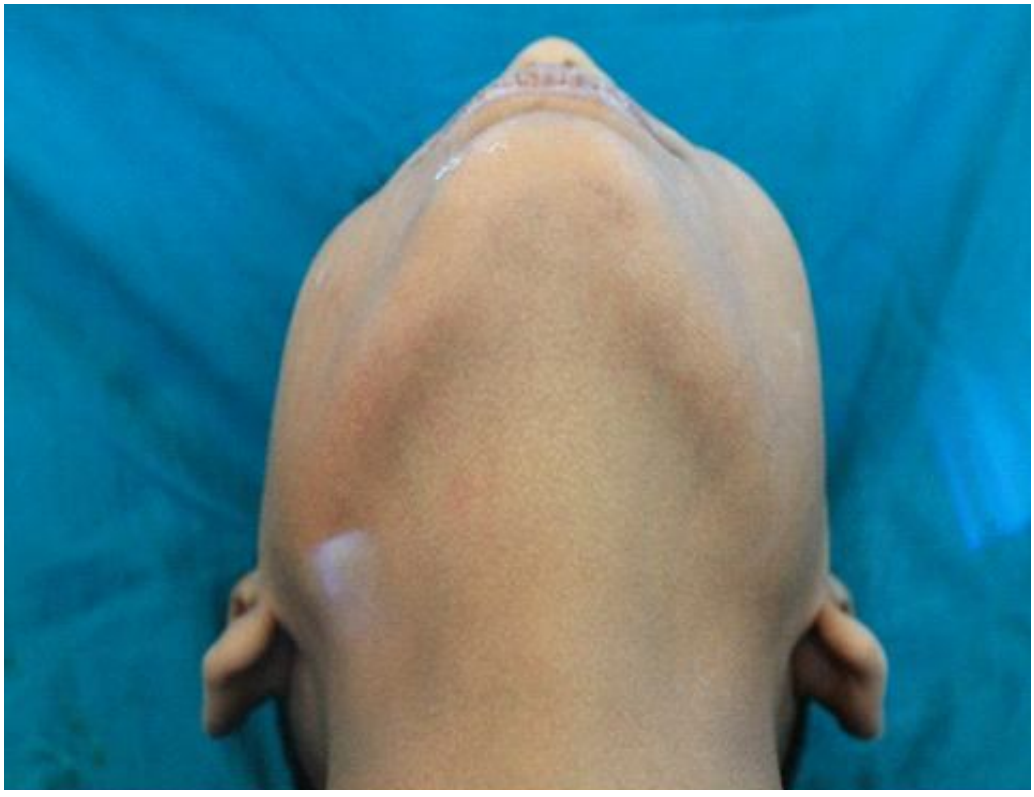


Fig 2: Diffused swelling present along the buccal aspect extending from 23 to 37

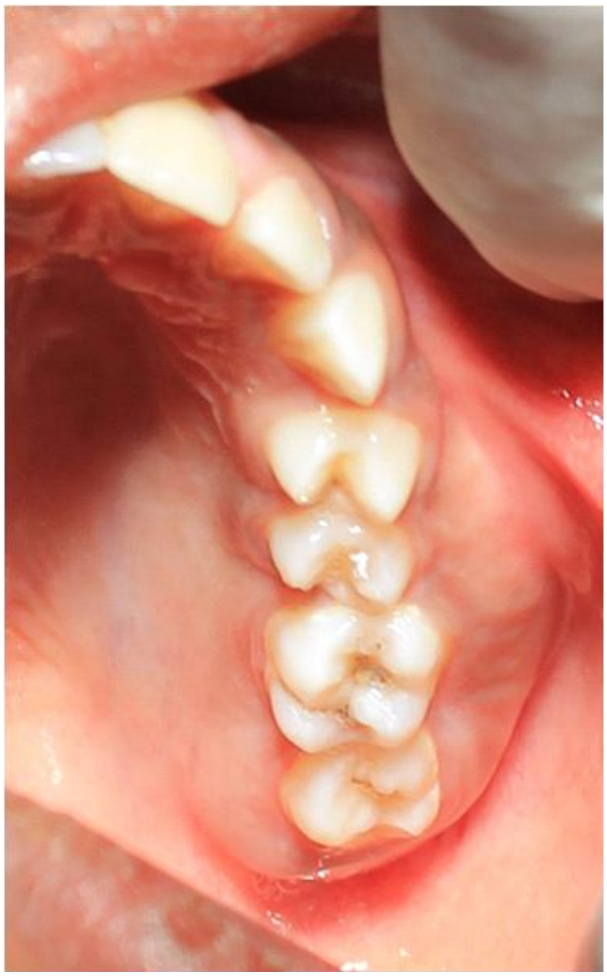


Fig 3: There is an evident homogenous radio opacity with ill defined borders which is extending medialy from the 23 to 28

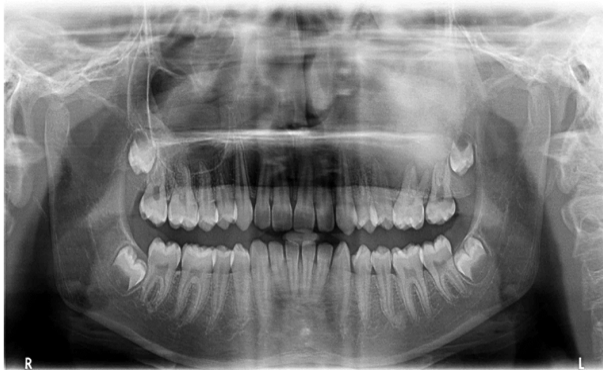


Fig 4: IOPAR revealing typical frosted glass appearance Laminadura is not seen Obliteration of PDL space Floor of the sinus is not seen

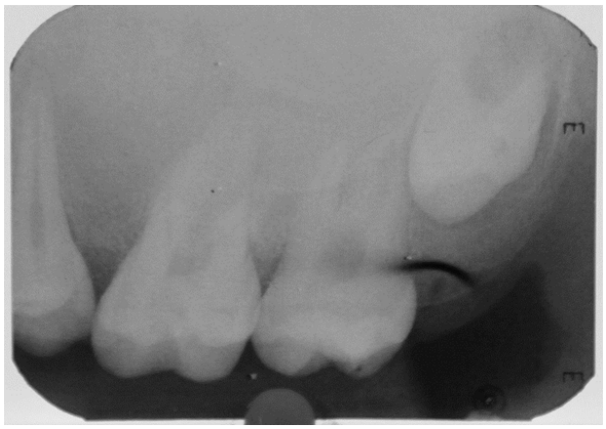


Fig 5: PNS reveals complete obliteration of the left maxillary antrum

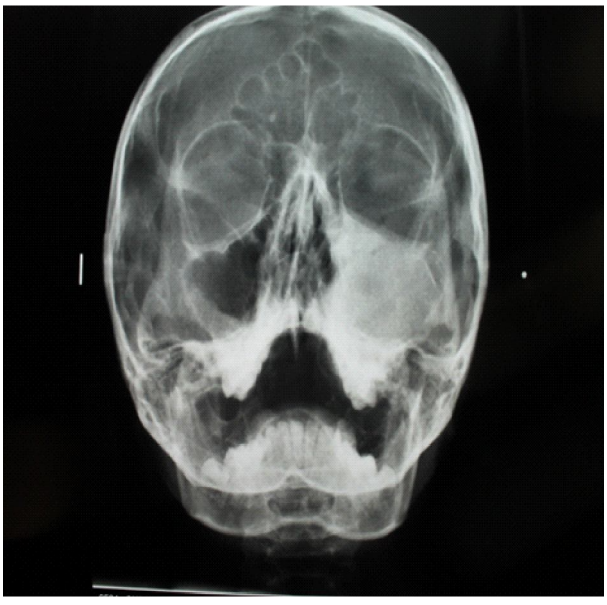
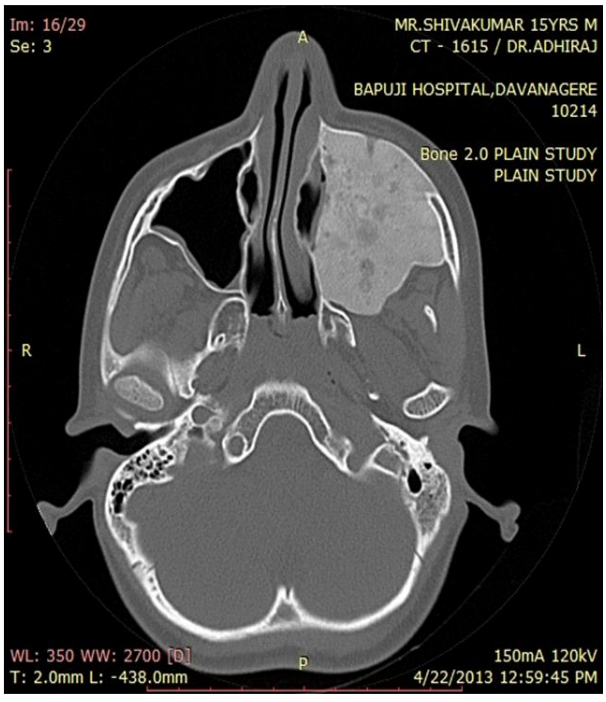


Fig 6: CT image revealing ground glass appearance



fibrous dysplasia. Patient was kept under follow up and aesthetic shaving of the excess bone.

Discussion

Fibrous dysplasia (FD) is a bone development anomaly characterized by hamartoma proliferation of fibrous tissue within the medullary bone, with secondary bony metaplasia, producing immature, newly formed and weakly calcified bone, without maturation of the osteoblast which appears radiolucent on radiographs, with the classically described ground-glass appearance [1]. In 1937, McCune and Bruch first suggested that among all of the abnormalities of bone formation, this disorder should have its own place as a distinct clinical entity.

The following year, Lichtenstein introduced the term "fibrous dysplasia" Reed's definition states that fibrous dysplasia is an arrest of bone maturation, woven bone with ossification resulting from metaplasia of a nonspecific fibro osseous type [3]. The etiology of this abnormal growth process is related to a mutation in the gene that encodes the subunit of a stimulatory G protein (Gsα) located on chromosome [20, 1, 2]. As a consequence of this mutation, there is a substitution of the cysteine or the histidine-amino acids of the genomic DNA in the osteoblastic cells-by another amino acid, arginine [3]. Consequently, the osteoblastic cells will elaborate a fibrous tissue in the bone marrow instead of normal bone [8, 9, 10]. It is a benign bone disorder of an unknown etiology, uncertain pathogenesis and diverse histopathology [2]. Fibrous dysplasia represents about 2, 5% of all bone tumours and over 7% of all benign tumours Cranial or facial bones are affected approximately in 30% of the patients [3, 4].

The average age of the patients with FD is from 5 to 67 without sex preference (46, 7% male) and usually manifests before the 3rd decade of life [4, 5]. Fibrous dysplasia is described in terms of three major types: monostotic, involving a single bone; polyostotic, having multiple lesions involving multiple bones; and McCune Albright syndrome, a polyostotic form of fibrous dysplasia that also involves endocrine abnormalities. The monostotic form of fibrous dysplasia is the most common, comprising 70% of cases, most likely to quiesce at puberty. A typical monostotic lesion, usually presented unilateral, will involve the femur, tibia or ribs, with 25% occurring in the bones of the skull. Affection of the craniofacial bone is observed with 10% of the patients suffering from monostotic FD [6, 7].

Twenty-five per cent of fibrous dysplasia involves two or more bones. These lesions may be localized to one region of the body or they may be disseminated, involving virtually every bone. There is a female predilection in polyostotic fibrous dysplasia, and up to 50% may involve bones in the head and neck. These lesions are more likely to continue to progress even after puberty. Deformity is progressive and by mass effect there may be impingement on other structures and functional impairment. These lesions tend to be structurally weak and are therefore prone to pathologic fracture. Alkaline phosphatase may be elevated in up to 30% of patients with polyostotic fibrous dysplasia, and a dramatic rise may herald malignant degeneration. Malignant degeneration occurs in less than 1% of cases of fibrous dysplasia. Malignancies are almost exclusively osteosarcoma. For unknown reasons, monostotic and craniofacial lesions have the greatest potential for malignant degeneration. Pain, rapid growth of a lesion and a dramatic elevation of alkaline phosphatase may herald malignant transformation.

FD is an important lesion affecting the maxillofacial region because it can cause severe deformity and asymmetry, and most devastating of all, blindness. Although according to various authorities, including Waldron [5], the majority of cases "burn out" in early adulthood when skeletal maturity has been reached, according to Eisenberg and Eisenbud [6] there are no studies of FD cases followed up over a long period to substantiate that view. Their contention is supported by later recurrence or reactivation in a small number of FD lesions followed over a long time, such as a report of two White cases [25], and one of a Chinese case [26]. Other cases of FD have either been reactivated or first activated by pregnancy, suggesting that sex hormones could influence at least some of them.

Furthermore, a number of cases continue growing into adulthood or first present clinically in adulthood [27, 28]. Sakamoto and co-workers [27] report six of their 62 Japanese cases of FD presenting in the sixth and seventh decades. Garau and co-authors [28] reported that nine out their 12 cases of gnathic FD presented in Italians over 20 years of age; two in the seventh and eighth decades. It is possible that these reports may merely reflect the ages the lesions were first detected, diagnosed and recorded rather than a later age of commencement of growth. A late detection of a long-standing lesion is entirely possible because many cases of FD are painless [26].

The range of behaviours suggests that the pathogenesis of FD may be complex. Chapurlat and Meunier [29] has proposed recently such a

pathogenesis, which interrelates many of the salient features, elevated cAMP, increased expression of the proto-oncogene, c-fos, abnormally differentiated osteoblasts, formation of abnormal bone, increase in sex steroid receptors, increased interleukin-6 (target of diphosphanate treatment) and osteoclasts. Furthermore, the classical division of FD into monostotic, polyostotic and McCune-Albright forms may reflect the timing of the mutation, and thereby, the initial size of the mass of FD precursor cells [30]. The polyostotic form may arise in foetal life whereas the monostotic form may arise postnatally [29, 30]. This correlates with the evidence that the monostotic form is not a precursor of the polyostotic form [31].

The monostotic form accounts for 80-85% of cases of FD. Three per cent of the polyostotic form have endocrinopathies [23] and are cases of McCune Albright syndrome (precocious puberty and cafe´- au-lait spots). McCune-Albright syndrome will not be considered further, because Fahmy and co-authors [32] have already fully discussed the radiology of precocious puberty and its extensive differential diagnosis. Although the term "monostotic" can be readily applied to cases of FD affecting the mandible alone, this is generally not so for FD affecting the maxilla or face. There FD can affect contiguous bones such as the zygoma and the sphenoid. These cases have been called "craniofacial FD" [23]. Polyostotic and McCune-Albright forms are easily diagnosed on clinical and radiological investigation alone. This is not so with the monostotic form, which has a number of other important lesions in its differential diagnosis requiring bone biopsy. Bone biopsy is generally avoided particularly where the risk of pathological fracture is high [29]. FD of the mandible differs in another important aspect from FD affecting long bones in that there does not appear to have been a report of pathological fracture of a dysplastic mandible [26].

A radiological regime for polyostotic FD used by radiologists is scintigraphy and then plain film radiography of areas of increased radiolabelled uptake or activity. These films may be complimented by computed tomography (CT), which is especially useful for confirming the diagnosis and assessing the extent of the FD in the craniofacial skeleton.

The radiology of FD affecting the face and jaws gives an insight to its behaviour. Eversole's contention that in FD the teeth generally remain un-displaced with resorption, whereas COF may displace them or even resorb their roots [33], is partly supported by a recent systematic review that found that over half of the teeth sited in dysplastic bone were undisplaced.[26] Furthermore, Petrikowski and co-authors34 suggested that "alteration of the lamina dura to the abnormal bone pattern, and narrowing of the periodontal ligament space areprimary distinguishing features" for FD.

These phenomena in FD may reflect "programmed 'field effect' of abnormal osseous development in congenitally predisposed bone matrix" [6]. This may account for the fusiform (spindle-shaped) expansionof FD of the affected bone. In contrast, the displacement of teeth or resorption of their roots in COF represents the almost spherical centrifugal expansion that is associated with a benign tumour growing out from the probable site of origin.

FD of the craniofacial complex may differ both radiologically and histologically from its counterparts in the axial skeleton. FD appears frequently in the latter as a circumscribed radiolucency with athin sclerotic periphery, whereas cases of craniofacial FD, certainly those affecting the jaws and adjacent bones, are poorer defined and more radiopaque. A reason for the difference in appearance between maxillofacial FD and FD of the long bones is that the former occurs in skeleton derived from membrane bone [33]. The woven bone, which is well-mineralized, is arranged in a network of broad trabeculae. Furthermore, lamellar bone, generally absent in FD in the axial skeleton, occurs occasionally in FD of the face [20].

Particularly in the monostotic form, FD commonly displays an abnormal opacification, which ranges from the very numerous, small and diffusely distributed opacities "ground glass" and "peaud' orange" to sclerosis, classically described as cotton-wool". Different patterns may not only be present in different parts of the same lesion, but may also depend on whether the film used is "direct exposure" or "fluorescent screen film" [20, 35]. The margins of extra-gnathic FD appear welldefined, whereas they are poorly-defined in the jaws. An objective definition of marginal definition has been described by Slootweg and Muller [36].

A lesion with a zone of transition less than 1mm can be considered to be well-defined. This can be quickly and cheaply appreciated on plain film radiographs. The expansion of FD of the mandible is classically spindle (or fusiform)-shaped when viewed on a true (axial) occlusal film or on a posterior-anterior projection of the mandible. The degree of expansion can be remarkable. Although the shape of the FD affected maxilla appears to be more complex, reflecting the maxilla's complex structure, the overall effect is similar to that seen on the mandible.

The expansion of the external surface of the affected bone assumes a more grotesque, but still recognizable shape, whereas the internal surfaces expand into orbital, nasal and sinus cavities, fissures, fossae and neural and vascular canals. The lesion, if large, often nearly completely obliterates the maxillary sinus. The above pattern is altered if the FD undergoes cystic degeneration with formation of a large aneurysmal bone cyst. Then the affected part of the lesion may lose its anatomical shape and becomes spherical as stated by Ferretti [37].

If FD affects the orbital cavity or more particularlythe optic canal, then blindness can result. Although the onset of blindness is generally gradualand may be intermittent, urgent surgery is frequentlyrequired to recover sight. If specialized surgeons are not available then corticosteroids may help to alleviate optic nerve compression [29].

Radiologists are familiar with an association of aneurysmal bone cysts (ABCs) and FD. Although the ABC is a well-recognized accompaniment to FD of the skull base it is not of FD of the jaws. Examples of the radiology of ABC secondary to FD of the mandible and of the maxilla are illustrated by Dorfman and Czerniak [24, 39] and Lustig and co-authors [31], respectively.

Another concern particularly in long-standing polyostotic FD is sarcomatous transformation, whichcan occur in absence of radiation therapy, 4% for patients with McCune-Albright, and 0.5% those with other FD forms [40]. Nevertheless, head and neck practitioners should be vigilant because the most frequent site for sarcomatous transformation is the craniofacial skeleton [41]. The features on conventional radiography that allow sarcoma to be differentiated from FD are permeative ill-defined borders, destroyed cortical outline and / or speculated periosteal new bone formation and widening of the entire periodontal ligament space [34, 38]. CT has become an essential tool particularly for the investigation of the face and upper jaw, where the anatomy is not only complex, but because of the Proximity of the eyes and optic nerves.

Conclusion

Fibrous dysplasia is a benign skeletal lesion that can involve one or more bones. Its etiology has been linked to an activating mutation of Gsα and the downstream effects of the resultant increase in cAMP. Polyostotic lesions tend to be larger than monostotic lesions and result in more skeletal complications, including pain, deformity, and fractures. Some patients with polyostotic bone involvement also have skin lesions and endocrinopathies (McCune-Albright disease) or multiple myxomas (Mazabraud syndrome). Monostotic lesions frequently are discovered incidentally and require only clinical observation. Confirmatory biopsy is indicated if the radiographic findings are not characteristic of fibrous dysplasia. Bisphosphonates have been shown to offer pain relief and improve skeletal strength in appropriately selected patients with either polyostotic or monostotic fibrous dysplasia. Occasionally, operative treatment is needed to correct deformity or to prevent or stabilize a pathologic or fatigue fracture. Cortical allograft or intra-medullary fixation of the entire long bone provides the best material properties for patients who require operative intervention.

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