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Case report: Management of a case of complex multifocal deformity in monomelic fibrous dysplasia

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ABSTRACT

Fibrous dysplasia occurs due to failure of maturation and differentiation of osteoblasts. The affected bones are subject to progressive deformities and rarely pathologic fractures, requiring surgical management. We report a case of monomelic fibrous dysplasia involving the proximal femur and the shaft of tibia, with a progressive genu valgum, and tibial bowing, who was managed by staged deformity correction and internal fixation.

Keywords: fibrous dysplasia; deformity correction; genu valgum; corrective osteotomy

INTRODUCTION

Fibrous dysplasia is a skeletal developmental anomaly of maturation and differentiation of osteoblasts, with predominant unilateral involvement. The medullary cavity of involved bones fills up with grayish white, gritty fibrous tissue containing newly formed trabeculae of primitive bone. (1) It affects both genders equally, with greatest frequency in the second and third decades of life. (2) It occurs in two forms- monostotic, and polyostotic. In monomelic fibrous dysplasia, usually seen in the lower limb, multiple bones in the corresponding limb segments are involved. The compromised structural integrity of the affected bone, along with the burden of weight bearing, causes progressive deformities and pathologic fractures.

Patients are asymptomatic initially, and the diagnosis may often be incidental. Later, patients present with pain, deformity, altered gait and even pathologic fracture, which may require surgical management.

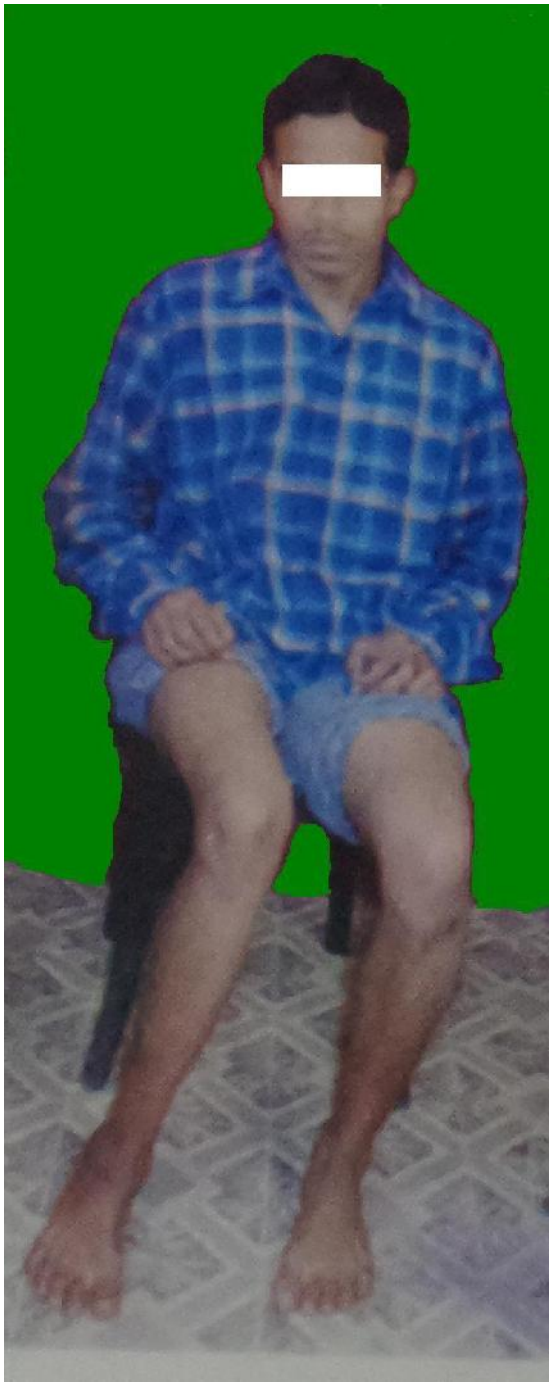
We report our planning and staged correction of a patient with monomelic fibrous dysplasia involving the proximal femur and the shaft of tibia.

CASE REPORT

A 22 year old gentleman, salesman by profession, presented to us with complaints of deformity over his right lower limb since 12 years, with altered gait, thigh pain and pain over his right leg and ankle since 4 months. He noticed the deformity over his leg and knee at the age of 10 years, and it had been progressing gradually since then. He had progressive, dull aching type of pain without any radiation, which aggravated with activity and ambulation. His walking distance had reduced to less than 100 meters due to the pain over a course of 4 months since its onset, thus leading to a restriction to his activities of daily living and continuing with his profession. He gave no history of any constitutional symptoms or similar complaints elsewhere in his body. Family and personal history were not significant.

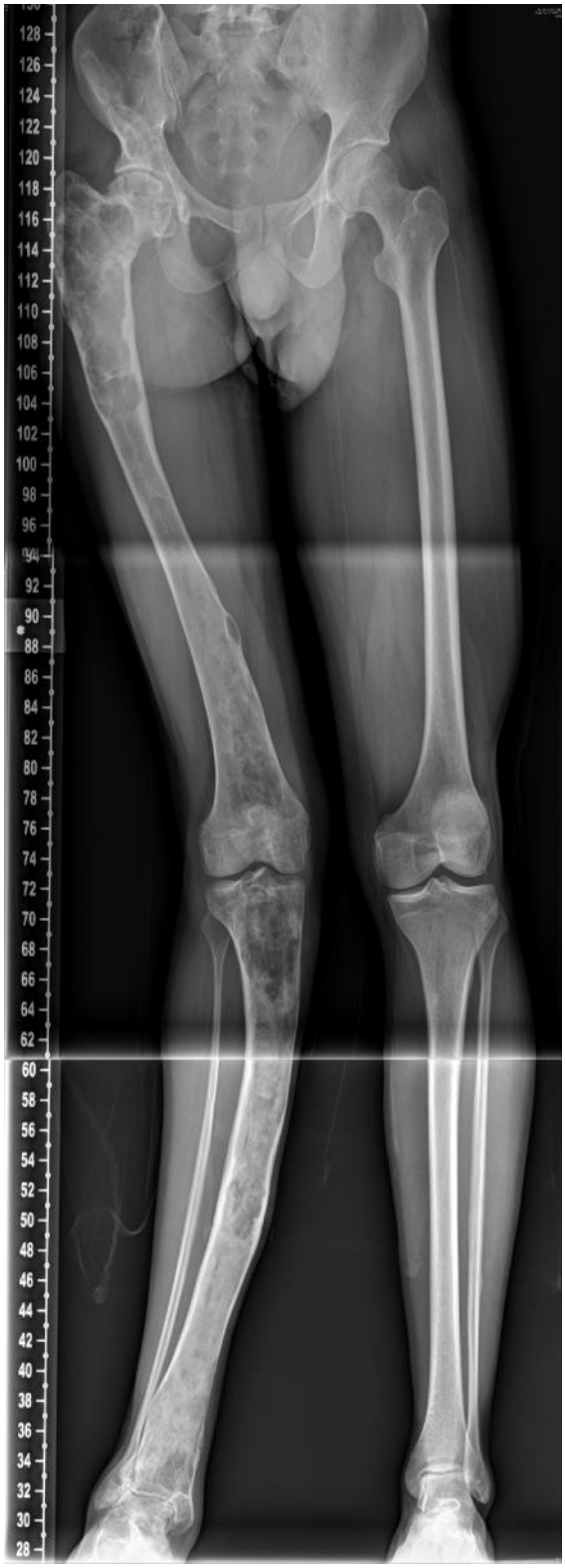
Gait was unaided, stable and antalgic, but the walking distance was severely reduced to less than 100 meters. On inspection, quadriceps wasting was noted. Multiple hyperpigmented patches of skin over his right leg were noted over the anterior aspect. The affected limb was found to have significant valgus deformity (Fig-1) in the knee and the tibial segment. Palpation revealed irregularity over the tibial shaft. Range of movement of the knee and hip revealed increased adduction and restricted external rotation. There was no true limb length discrepancy.

Fig-1. Pre operatively, the patient had a significant valgus deformity over the right knee as seen in the image.



Evaluation was done with plain radiographs of the pelvis, femur and tibial segments, along with hip to ankle stitch view (Fig-2). They revealed two sites of lesions: the proximal femur, and the tibial diaphysis. The lesions had a characteristic ground-glass appearance, and thinning of affected cortices.

Fig-2. Pre-operative hip to ankle stitch view radiograph of the patient.



He was planned for staged surgical management, the aim of corrective osteotomy was to obtain a well aligned lower limb to restore anatomy and biomechanics. In view of the complex multifocal nature of deformity, correction was planned in two stages. Correction of the femoral deformity required restoration of the neck-shaft angle and offset. This was achieved by subtrochanteric osteotomy. Furthermore medializing the entry point while using a trochanteric entry/ intra medullary nail with a valgus offset (Zimmer® Natural Nail® system) with careful canal preparation prevented a loss of correction with internal fixation.

The distal osteotomy in the femur site was performed at a point, where the bowing prevented further advancement of the nail, with a lateral opening wedge correction and careful preparation of the intra medullary segment. Correction was achieved with a near center position of the nail in the distal femur. Stability was further augmented with interlocking bolts and a neck screw.

Radiographs at 6 months revealed no progress in disease or loss of correction with all osteotomy sites progressing to union. His buttock and thigh pain had significantly reduced, permitting him some return to occupation. He was counselled regarding a second stage correction to correct the valgus deformity at the knee and tibia. Pre-operative planning helped decide the location of the osteotomy to restore mechanical and anatomical axes, with near normalization of the medial proximal tibial angle and the lateral distal tibial angle. Internal fixation with an interlocking intra medullary tibial nail was performed to stabilize the tibial corrective osteotomy. Post operatively, he was advised to continue his physiotherapy exercises which included knee mobilization and quadriceps strengthening, and he recovered full range of motion (Fig-3a,b). He was ambulated with the aid of bilateral axillary crutches with partial weight bearing from the 3rd post-operative day.

Fig-3 a, b. Patient in supine position following deformity correction surgery, demonstrating full range of hip and knee flexion.

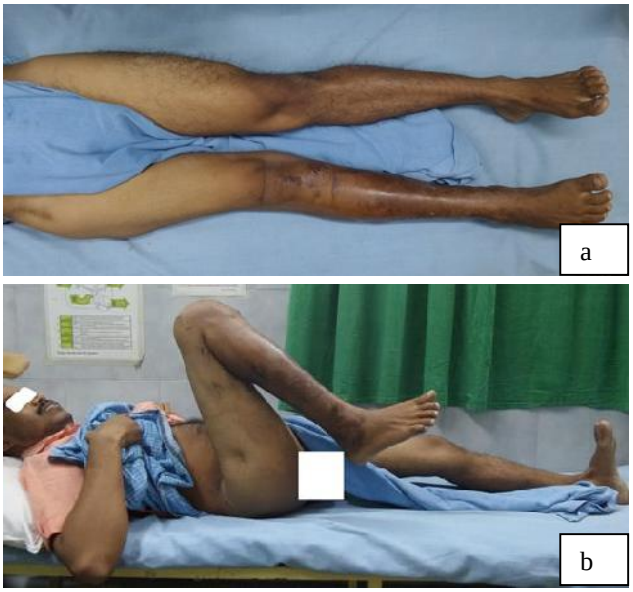
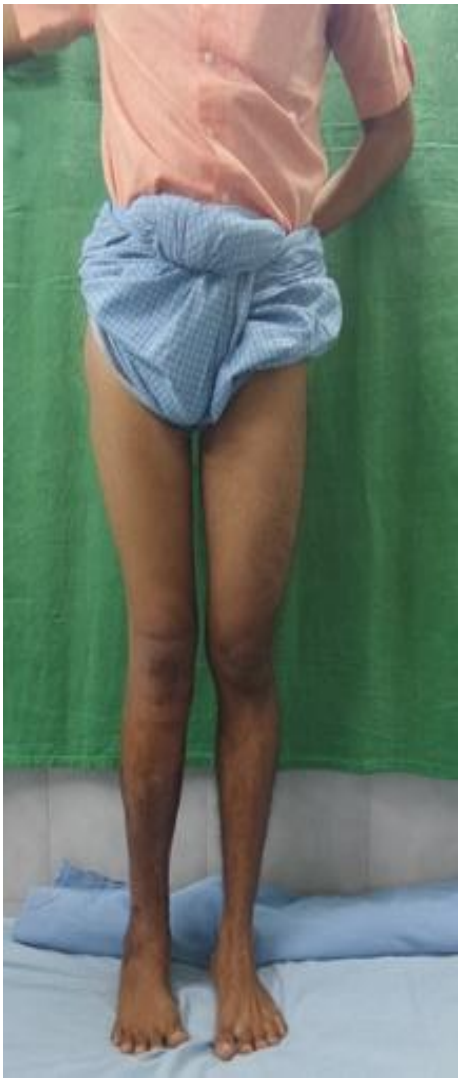
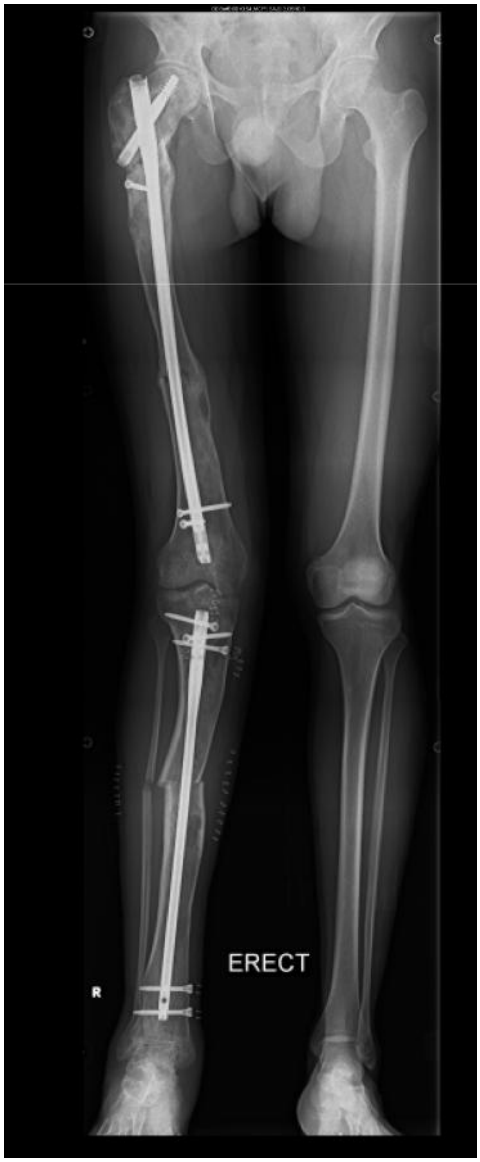


Fig-3(c). Patient standing following deformity correction



At follow-up, the patient was found to be relieved of the pain, and was ambulant and independent for his personal care. All wounds had healed satisfactorily. Fig 3(c) demonstrates the patient in a standing position following the deformity correction surgeries. At follow-up, a hip-to-ankle stitch view radiograph was obtained along with routine imaging of the femoral and tibial segments, to assess the correction of the deformity. (Fig-4)

Fig-4. Hip to ankle stitch view radiograph, post-operatively, following both stages of deformity correction.



The relevant angles measured from the pre-operative and post-operative radiographs has been summarized in table-1. References for normal value ranges, are based on the seminal works of Paley et al (3) and Chao et al.(4)

Table-1. Relevant angles measured from pre and post operative radiographs. Prefixes “a” and “m” have been used to denote that the angle is measured between the joint line, and either the anatomic axis or the mechanical axis of the limb segment respectively. (NSA = femoral neck-shaft angle on the AP view; mLDFA = mechanical lateral distal femoral angle; mMPTA = mechanical medial proximal tibial angle; mLDTA = mechanical lateral distal tibial angle; mTFA = mechanical tibio-femoral angle; aTFA = anatomic tibio femoral angle; MAD = mechanical axis deviation)

	Normal value	**Right side, Pre op**	**Right side, Post op**	**Left side**
NSA	125° -131°	116°	130°	130°
mLDFA	87.5° +/- 2.5°	80°	83°	87°
mMPTA	87.2° +/- 2.5°	103°	92°	88°
mLDTA	88.6° +/- 3.8°	89°	89°	87°
mTFA	1.3° +/- 2° varus	22° valgus	8° val-gus	1.5° varus
aTFA	6° valgus	38° valgus	15° valgus	5° valgus
MAD	8 +/- 7 mm, Medial	83 mm Lateral	34 mm Lateral	8 mm Medial

DISCUSSION

Fibrous dysplasia is a form of developmental anomaly of the skeleton, wherein there is failure of the mesenchymal osteoblasts to mature and differentiate.(1) (5) As a result, there is abnormal deposition and remodeling of fibro-osseous tissue within the medullary cavity of long bones. Fibrous dysplasia affects both genders, often presents in the second and third decades of life (2), and has no hereditary predisposition. It exists in the monostotic and polyostotic forms(6), and usually affects the metaphyseal and diaphyseal regions of long bones.(5) It accounts for 7% of benign bone lesions.(7) In the monomelic presentation, multiple bones on a single extremity are involved.

Monostotic fibrous dysplasia is often seen in the lower limb unilaterally, most commonly in the femur (5), followed by the tibia to a lesser extent. Less commonly, unilateral fibular involvement may be noted. Skip lesions can also be noted.

The higher concentration of abnormal bone in a weight bearing limb compromises the structural strength of the extremity. This can lead to two kinds of manifestations-deformities, and pathologic fractures. Lesions in the tibia can also lead to anterior bowing.

Clinically, the patients are usually asymptomatic until the condition is discovered as a coincidental finding during examination for another reason.(8) Occasionally, they may present following a pathologic fracture.(3) Majority of patients with a monostotic variant are symptomatic much later compared to the polyostotic form. The lesions usually become quiescent at puberty but the resultant deformity can progress. This probably occurs due to the increased body weight burden with age on the affected limb. Malalignment causes a disturbance in the normal transmission of the forces across the knee, and gradually progresses to degenerative arthropathy. (9)

McCune Albright syndrome comprises polyostotic fibrous dysplasia, endocrine abnormalities, and cutaneous pigmentation abnormalities.(10) Mazabraud syndrome comprises polyostotic fibrous dysplasia, along with intra-muscular myxomas.(11) Associated abnormalities, such as precocious puberty, thyroid disease and abnormal skin pigmentation can also be seen. Malignant transformation has been reported occasionally, and may occur with or without prior radiotherapy, though the incidence of malignant change is low when strict criteria are used.(12)

Radiographically, the lesions are predominantly intra-medullary, and display a radiolucent “ground glass” appearance. These lesions may be eccentric or central, and usually are well circumscribed. They may have a sclerotic border.(13) The “shepherd’s crook” appearance is also a classical finding.

Evaluation of patients with fibrous dysplasia should include a thorough evaluation of endocrine profile and phosphate metabolism. Occasionally, a biopsy may be required for confirmation of diagnosis. Pathologically, cysts of varied sizes, filled with serous fluid or blood are seen. Histopathology shows irregular woven bony spicules within a fibrous stroma. Small areas of cystic changes or cartilaginous metaplasia may be noted.

Treatment is required only in symptomatic patients.(8) Long term outcomes are usually satisfactory regardless of treatment in non-progressive cases. (7) Surgical management is indicated for significant deformity, severe pain, or in the instance of a pathologic fracture. Moreover, the surgical goal in correction of the deformity is to optimize limb alignment and joint orientation, and to restore biomechanics, which will prevent eccentric loading of long bones, which leads to progression of deformity. Correction of deformity also prevents abnormal loading across articular surfaces, which predispose to early degeneration.(14) (3) Internal fixation prevents loss of correction, and helps early mobilization.

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