

Intramedullary nailing for fibrous dysplasia of lower limbs

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Abstract. Fibrous dysplasia (FD) of the bone is rare and self-limiting. However, lesion expansion may occur, causing pain, deformity and pathological fracture, in which case surgery is occasionally required. Indicators of FD have not been previously described, although there are several surgical procedures reported (curettage, curettage and graft, and internal fixation). In this study we discuss whether intramedullary nailing of these lesions could result in more favorable outcomes in correcting deformities, including the prevention of secondary fractures and maintenance of the bone, compared to other internal fixation methods. A total of 39 patients with FD of the bone treated with intramedullary nailing were retrospectively analyzed. The surgical procedures involved curettage, grafting and intramedullary nailing. No infection, thromboembolism or other notable complications occurred. The patients resumed full activities of daily living. At the last follow-up, 33 patients presented no pain and seven patients had occasional mild pain. The clinical score according to the modified criteria of Guille improved from an average of 4.4 points prior to surgery to an average of 8 points following surgery. The neck shaft angle of the femur improved from an average of 90° prior to surgery to 125° following surgery. Intramedullary nailing may be used to correct deformity and prevent pain and refracture in FD of the bone of the lower limbs with large lesions, pathological fracture or deformities. All patients were allowed full athletic recovery following surgery.

Introduction

Fibrous dysplasia (FD) of the bone is a battery of tumor-like lesions in which fibrous tissue proliferation replaces normal bone architecture. FD accounts for only 5% of benign bone tumors (1). The incidence of this disease is 10-30 cases per million and accounts for 5.89% of benign

bone tumors in China. In 1891, FD was first described by Von Recklinghausen (2), but it was Lichtenstein (3) who labeled it polyostotic FD (PFD) in 1938. Lichtenstein and Jaffe (4) initially described the spectrum of the clinical, radiographic and histological symptoms. FD may involve a single skeletal site with an isolated lesion [monostotic FD (MFD)] or it may involve multiple bones [polyostotic FD (PFD)]. In addition, it may occur in association with distinct café-au-lait skin spots and/or a number of hyperfunctioning endocrinopathies labeled as McCune-Albright syndrome (MAS). In addition, FD may occur in association with intramuscular myxomas, which is referred to as Mazabraud's syndrome (5,6). It has been reported that the monostotic form is 8-10 times more common than the polyostotic form (7). Compared with the spine, ribs and skull, FD is more common in the bones of the limbs. Following puberty or epiphyseal closure, the asymptomatic lesions of the majority of patients are restrictive and do not develop or progress. However, development and progression may occasionally be detected in lesions of patients with symptoms of pain, deformity and/or pathological microfracture.

In the present study, we retrospectively examined patient outcome following intramedullary nailing for FD of the lower limbs via long-term clinical follow-up.

Patients and methods

Patients. We retrospectively reviewed patients with FD of the lower limbs treated by intramedullary nailing between 2003 and 2010. A total of 39 patients participated in the study. The study was approved by the ethics committee of Anhui Medical University. Informed consent was obtained from the patients or the patient's family. The mean age of the patients was 31 years (range, 17-55), of which 22 were male and 17 were female. The mean follow-up period was 50 months (range, 4-93 months). Types of tumor included monostotic (33 patients) and polyostotic (7 patients). Symptoms included pain (15 patients), pathological fracture (10 patients), swelling/deformity (13 patients), coxa vara (8 patients) and a limp (1 patient). Two patients had already undergone several treatments. One patient aged 12 years who presented with FD of the right humerus had received conservative treatment. Curettage and grafts of the left tibia were performed at 15 years old and were pathologically confirmed to be FD. Pathological fracture of the left tibia occurred at 17 years. Another patient presented with a lesion and received conservative treatment in

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Table I. Patient characteristics.

Patient	Age (years)	Gender	Location	Symptoms	Return to full activity (months)	Guille ^a	Guille ^b	Neck shaft angle ^a	Neck shaft angle ^b
1	35	F	Femur (L)	Pain	4.8				
2	44	M	Bilateral limb	Fracture	14.1				
3	42	F	Femur (R)	Pain	5.1				
4	38	M	Tibia (R)	Pain	4.9				
5	31	F	Femur (R)	Pain	4.2				
6	20	M	Tibia (R)	Fracture	3.8				
7	29	M	Femur (L)	Fracture	5.0				
8	26	M	Femur (L)	Deformity	5.8	3	9	70	119
9	33	M	Femur (R)	Pain	3.9				
10	41	M	Femur (R)	Deformity	4.7				
11	35	F	Tibia (R)	Pain	4.0				
12	17	M	Tibia (R)	Fracture	2.6				
13	18	F	Femur (R) + skull	Deformity	5.2	4	9	85	120
14	45	M	Tibia (R) + L3	Pain	3.6				
15	23	M	Femur (L)	Deformity	4.7				
16	30	M	Femur (L)	Fracture	5.4				
17	51	F	Tibia (L)	Pain	6.4				
18	17	M	Femur (L)	Pain	4.4				
19	18	F	Femur (L)	Deformity	3.5	5	10	104	130
20	55	F	Femur (R) + C6C7	Pain	6.8				
21	18	M	Femur (R) + tibia (R)	Deformity	3.2	3	9	75	120
22	34	M	Femur (L)	Deformity	4.5	6	8	120	140
23	20	F	Tibia (R)	Pain	3.3				
24	18	M	Femur (L)	Deformity	5.5	5	8	90	127
25	20	M	Femur (R)	Fracture	4.7				
26	30	M	Femur (L)	Deformity	5.8				
27	18	F	Femur (L)	Limp	4.2				
28	48	F	Femur (L)	Fracture	6.0				
29	54	F	Femur (L + R)	Pain	5.4				
30	20	M	Femur (L)	Fracture	3.6				
31	52	F	Femur (R)	Deformity	6.5				
32	21	F	Femur (L) + fibula (L)	Pain	3.2				
33	19	F	Femur (R)	Fracture	2.4				
34	47	M	Tibia (L)	Pain	3.1				
35	21	F	Tibia (R)	Pain	2.6				
36	33	F	Femur (L)	Deformity	4.0	4	7	87	120
37	43	F	Femur (R)	Deformity	5.5	5	6	92	125
38	23	M	Tibia (L)	Deformity	4.1				
39	52	M	Tibia (L)	Fracture	5.7				

^aPreoperatively; ^bpostoperatively; L, left; R, right; L3, lumbar 3; C6C7, cervical vertebra 6, cervical vertebra 7.

the neck of the left femur aged 10 years following a two-year limp. The patient underwent curettage due to deformity of the skull at 13 years old, and coxa vara was diagnosed at 18 years. The sites of the lesions were the femur (31 lesions), coxa vara (8 lesions), tibia (14 lesions), fibula (1 lesion), spine (2 lesions), bilateral lower limbs (1 patient), ipsilateral femur and tibia (1 patient) and ipsilateral femur and fibula (1 patient). The lesions located by patient self-observation and clinical manifestations, were detected by radiography, including plain films, computed tomography (CT) and magnetic resonance imaging (MRI). Following surgery, pathology was the ulti-

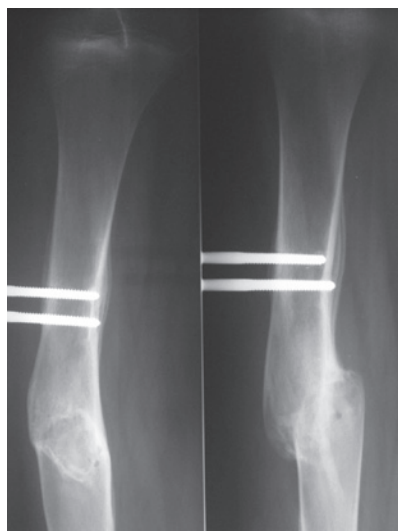
mate diagnostic method. Patients characteristics are shown in Table I.

Surgical technique. The patient was placed in the supine position under an image intensifier. During surgery, a cortical fenestration sufficiently large to be able to visualize the whole lesion was made using an osteotome. The lesion was removed with curettes, and the cavity was enlarged with a burr and irrigated with sterile saline. The removed tissue was sent for pathological examination. For varus of the hips, a lateral closing wedge osteotomy was performed and a guide pin was

A



B



C



Figure 1. (A) Immediate postoperative, (B) 6-month and (C) 1-year follow-up radiographs of a 12-year-old male with a pathological fracture of the left femur that was treated with an external fixator.

inserted into the medullary canal through the distant osteotomized site. This was then reamed, and repeated removal of fibrous tissues of the intramedullary cavity was performed. After reaming, an intramedullary nail was inserted and then



Figure 2. Refracture of the left femur after playing basketball at two years follow-up.

locked with proximal and distal screws. The cavity was then irrigated with sterile saline followed by the placement of a combination of autogenous cancellous bone and/or allograft cancellous chips.

Rehabilitation. Following surgery, all patients were advised against weight bearing but to mobilize joints depending on the location of the lesion. Patients were then provided with crutches to use for walking. Approximately two weeks after surgery, if there was clinically significant restrictive motion, physiotherapy was offered. Patients received a wound check and suture removal two weeks after surgery. Partial weight bearing was allowed approximately 2-3 months later. Time taken to return to normal walking was determined by patients' symptoms and follow-up radiographical imaging of osteotomy sites.

Follow-up. Patients were reexamined using plain film radiography following surgery. Radiography and clinical examinations were repeated every month for the first six months, and once every six months after that. Outcome criteria for the study included: i) pain status at the last follow-up; ii) time taken until return of full activity; iii) repeat/additional surgical interventions. Pain was measured using a patient visual analogue scale. Recovery time was measured between the date of surgery and the date when full activity was resumed. The clinical score was in accordance with the modified criteria of Guille *et al* (8).

Results

Recovery time. None of the patients had infection, thromboembolism or other notable complications. No loosening of screws or refracture was detected in the follow-up period. However, one patient with a lesion in the bilateral lower limbs, who underwent an osteotomy of the femur, had an unrelated pathological fracture on the other femur and was treated with

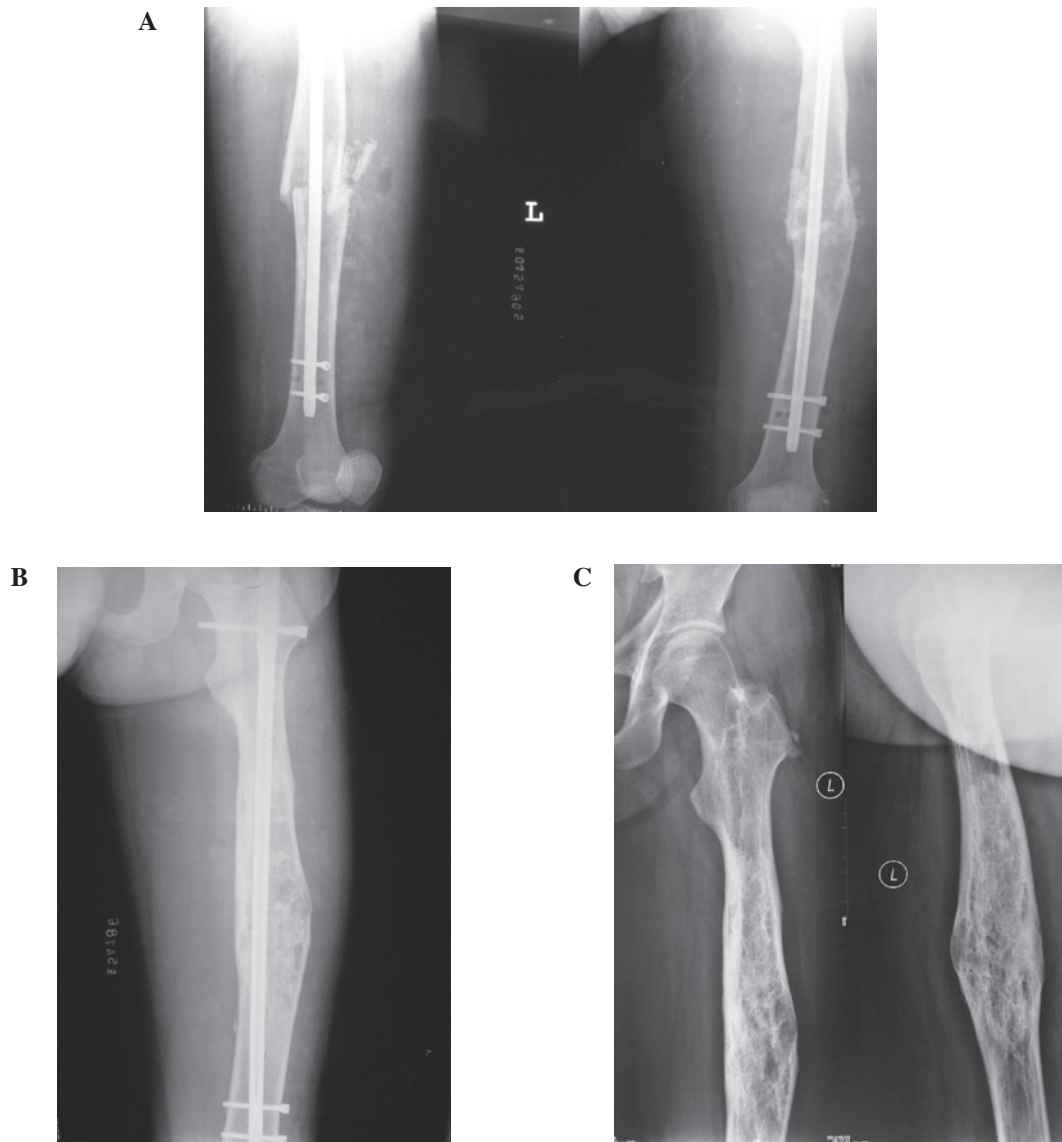


Figure 3. (A) Immediate postoperative, (B) 1-year and (C) 3-year follow-up radiographs of intramedullary nailing, curettage and bone grafting of the left femur.

further intramedullary nailing surgery. Therefore, the time taken to return to full activity was prolonged. One patient aged 12 years presented with a pathological fracture of FD and was treated with an external fixator (Fig. 1A). The fracture resulted in a malunion as demonstrated in the six-month and one-year follow-up radiographs (Fig. 1B and C, respectively). A refracture of the lesion (Fig. 2) was detected and treated with intramedullary nailing at two years follow-up (Fig. 3A). No loosening of screws or refracture was detected in the follow-up period (Fig. 3B). The fixator was removed at three years follow-up (Fig. 3C).

Patient recovery and symptoms. A total of 32 patients (82.1%) presented with no pain and 7 patients (17.9%) had mild and occasional pain at the last follow-up. All patients resumed full activity and function. One patient took longer to return to full activity due to pathological fracture of another limb. The clinical score was in accordance with the modified criteria described by Guille *et al* (8). The clinical score improved from an average of 4.4 (range, 3-6) points prior to surgery to an average of 8 (range,

6-10) points at the last follow-up. The neck shaft angle of the femur was corrected from an average of 90° (range, 70-120°) prior to surgery to an average of 125° (range, 119-140°).

Discussion

FD of the bone is a type of benign bone lesion. A single skeletal site with an isolated lesion is involved in the majority of patients, but the disease may be polyostotic. Previous studies have not reported indicators for FD, although there are several methods of treatment, including conservative treatment (medications and braces) and surgical procedures (curettage, curettage and graft, and internal fixation). Kusano *et al* (9) revealed that the majority of lesions of FD usually cease to progress or develop following adolescence, with the exception of McCune-Albright syndrome. Therefore, conservative treatment is recommended for adolescents. The mass of the lesion may not require surgery, unless ongoing pain, lesion expansion or bone percentage predisposes the bone to pathological fracture or a fracture has already occurred (10-15). Pain is associated with lesion

expansion. Generally, curettage of the lesion leaves a cavity which predisposes the bone to destabilization or even pathological fracture. Therefore, an external or internal fixation is performed for stabilization. In PFD patients, particularly those with shepherd's crook deformity, curettage and bone grafting are not advised due to reabsorption of bone graft and lesion expansion following surgery. In this case, internal fixation and correction are recommended for patients with distinct deformity and/or pathological fracture, which result in poor quality of life and complications due to bed rest.

In the case of large lesions (more than two-thirds of the bone) or upon enlargement of the lesion, the bone becomes unstable and predisposed to fracture when a cavity is left following curettage. Muscle atrophy, thromboembolism, ankylosis or other notable complications may occur in the long-term external fixation period. However, it is difficult to provide sufficient stability with plates and screws in the vicinity of the weakened bones, and fracture or refracture may occur due to the stress-shielding effect of the distal part of the plate. Guille *et al* (8) suggested that a refracture may easily occur after the plate and screws are removed in patients with a lesion involving a large area. O'Sullivan and Zacharin (16) reported that intramedullary nailing and bisphosphonate treatment of 10 femurs with McCune-Albright syndrome prevented fractures and resulted in improved walking. Thus, we consider that intramedullary nailing may be used in other anatomical sites, with the exception of proximal femoral lesions of the lower limbs. This approach results in fewer sequelae at the site of the lesion or fracture than when using plates and screws, and provides sufficient stability with fixation in the normal distal bones. Particularly in cases with a lesion covering a large area, the incision has to be enlarged to fit a long metal plate due to the vicinity of weakened bones, and loss of fixation, delayed union or non-union of the fracture may occur more frequently. When a valgus osteotomy is performed, overcorrection of the neck shaft angle is recommended (more than 130°) in anticipation of postoperative loss of valgus in lesions of the proximal femur, including shepherd's crook deformity. However, a satisfactory clinical result can be expected when the neck shaft angle is at least 90° (8). We recommended overcorrection of the neck shaft angle in anticipation of loss of valgus angle after surgery, which is limited by muscle contracture between the greater trochanteric area and the ilium. This approach is prevented in adolescents due to epiphyseal injuries. However, intramedullary nailing is performed on the occurrence of epiphysis fusion or marked changes of the epiphysis.

During surgery, the cortical fenestration should be sufficiently large to allow visualization of the whole lesion with an osteotome. Due to the internal fixation, the bone was stable and the cortical fenestration did not result in pathological fracture during or following surgery. We recommend removal of the lesion using curettes and enlargement of the cavity using a burr. However, normal trabecular bone reduction due to repeated curettes may cause delayed union or non-union of the fracture.

A number of authors have recommended that the defect should be filled with bone grafts or substitutes following curettage. Autogenous bone grafting is recommended due to the advantages of lack of immunological reaction and successful bone induction. However, obtaining sufficient bone for large

cavities may be difficult and sequelae may be generated at the donor site. In addition, certain authors have recommended curettage of benign bone tumors without grafts (17). We recommend curettage and grafting with autogenous bone from the ilium. However, large defects could alternatively be filled with substitutes including cement, hydroxyapatite or tricalcium phosphate, as bone grafts are difficult to obtain.

In conclusion, indicators for FD of the bone have not yet been established due to low incidence and small sample sizes. However, we propose an approach which allows patients to return to full, painless activity quickly in the majority of cases. The approach used in this study is recommended as a reliable and effective surgery for FD of the lower limbs.

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