

Genu Valgum Secondary to Focal Fibrocartilaginous Dysplasia of the Distal Femur

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Abstract: We report a case of unilateral genu valgum secondary to focal fibrocartilaginous dysplasia (FFCD) isolated in the posterolateral cortex of the distal femur. This case is the first incidence of a discrete fibrous band occurring in conjunction with a FFCD lesion in the distal posterolateral femur treated with excision of the tether and the overlying periosteum with curettage of the cortical focal fibrocartilaginous defect. Treatment was considered successful with gradual resolution of the 30° valgus deformity over 24 months, and we avoided the necessity of corrective osteotomy and its associated risks. To our knowledge, resolution of genu valgum secondary to FFCD in the distal posterolateral femur after curettage has not been previously described in the literature.

Key Words: children, curettage, deformity, femur, focal fibrocartilaginous dysplasia, genu valgum

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Since the initial report of focal fibrocartilaginous dysplasia (FFCD) as an etiological factor in the onset of unilateral tibia vara during infancy by Bell et al³ in 1985, there has been a total of 48 cases of FFCD in the lower extremity (43 proximal tibia, 5 distal femur) causing unilateral angular deformities in young children reported in the literature.^{1,3–6,8–12,14,17–21,23–25} In addition, there are two reports in the literature presenting three patients with bowing deformities in the upper extremity due to FFCD in the proximal humerus (1 case)¹⁶ and distal ulna^{5,16} (2 cases). We present a case of a child with unilateral genu valgum deformity secondary to FFCD in the posterolateral distal femur occurring in conjunction with a fibrous band. Our patient, treated with resection of the fibrous band and curettage of the cortical FFCD lesion, did not require a corrective

osteotomy. The deformity gradually resolved and the mechanical axis was restored over 2 years of follow-up.

CASE REPORT

A healthy, 20-month-old female, the product of a normal, full-term, vaginal delivery without complications, presented to our institution with progressive genu valgum deformity of the left lower extremity, which had been noticed by the patient's mother at the age of 10 months when the patient began to ambulate. The patient's history for trauma, infection, and metabolic bone disease was negative. Her family history was unremarkable for orthopaedic disorders.

Initial radiographic examination at an outside institution when the patient was 13 months of age demonstrated a distal femoral valgus deformity of 10° and a mechanical lateral distal femoral angle (mLDFA)²² of 77° (Fig. 1). At 24 months of age, radiographs obtained at our institution demonstrated a 25° valgus deformity of the left distal femur and a 1.0-cm limb-length discrepancy secondary to femoral inequality. The mLDFA at that time was 63°. Radiographs also revealed a well-defined, cortically-based sclerotic lesion with cortical thickening in the posterolateral cortex of the distal femur at the apex of the angular deformity (Fig. 2). MRI of the distal femur showed increased signal intensity localized within the area of cortical thickening and a normal distal femoral physis, without evidence of premature fusion, cartilaginous bar formation, or a discrete fibrous band (Fig. 3).

Over a 14-month period of time, the valgus deformity of the left femur increased from 10° to 30° and the mLDFA progressed from 77° to 58°. There was an associated 15° of anterior bowing or an anterolateral bowing of approximately 34° (Fig. 4). Surgical intervention was therefore planned when the patient was 27 months of age. A lateral approach to the distal femur allowed exposure of the lateral distal physis. A fibrous band extending from the physis posterolaterally into a bony cavitory lesion in the metaphysis was identified. The band penetrated the cortex at the distal one third of the femur. The fibrous band and overlying periosteum were excised followed by curettage of the bony lesion. Histologic examination of the bony lesion confirmed the diagnosis, revealing dense collagenized fibrous tissue with focal areas of fibrocartilaginous

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FIGURE 1. Anteroposterior radiograph at presentation of 13-month-old female with genu valgum of the left lower extremity.

changes (Fig. 5). These findings confirmed the diagnosis of focal fibrocartilaginous dysplasia.^{3-5,10,13,19,21,25}

Six months after excision and curettage of the lesion, the valgus deformity had decreased from 30° to 18°. At 1 year postoperatively, the valgus deformity measured 9° and the mL DFA was 80°. The bowing deformity was resolved at this time (Fig. 6). Radiographs obtained at 2 years postoperatively demonstrated restoration of the mechanical axis of the femur and a mL DFA of 88° (Fig. 7). There is a residual 1.0-cm limb-length inequality originating in the femur.

DISCUSSION

Focal fibrocartilaginous dysplasia (FFCD) is a rare, benign lesion that may lead to unilateral lower extremity angular deformities in the pediatric population and typically presents when the child begins to ambulate.¹⁵ FFCD has been reported as a unilateral cortical defect between the metaphysis and diaphysis. Unlike infantile tibia vara (Blount's disease), FFCD lesions do not involve the physis. This allows these two clinical entities to be differentiated with radiographic studies. Based on the distribution of cases reported in the literature, there seems to be no discernible predilection for either sex or side of extremity affected. The differential diagnosis based upon radiographic findings includes other cortical lesions such as chondromyxoid fibroma, osteoid osteoma, and eosinophilic granuloma.²³ However, these are not commonly associated with angular deformity.

Since Bell et al³ first described unilateral tibia vara secondary to FFCD in the proximal medial tibia, there have been a growing number of cases reported in the literature of FFCD in the proximal tibia causing varus deformities^{1,3-6,8-12,14,17-21,23,25} and one case of FFCD in the proximal lateral tibial metaphysis with associated tibia valga.²⁴ In 1997, Albinana and colleagues¹ were the first to report unilateral FFCD in the distal femur, followed by Macnicol¹⁷ and most recently by Choi et al.⁵ In four of the five cases of femoral FFCD (Table 1), the lesions were located in the posteromedial aspect on the distal femur and associated with an average varus



FIGURE 2. Leg-length radiographs taken at 24 months of age demonstrated progression to a 25° valgus deformity. A well-defined, cortically-based sclerotic lesion with cortical thickening in the posterolateral cortex of the left distal femur is seen at the apex of the angular deformity.

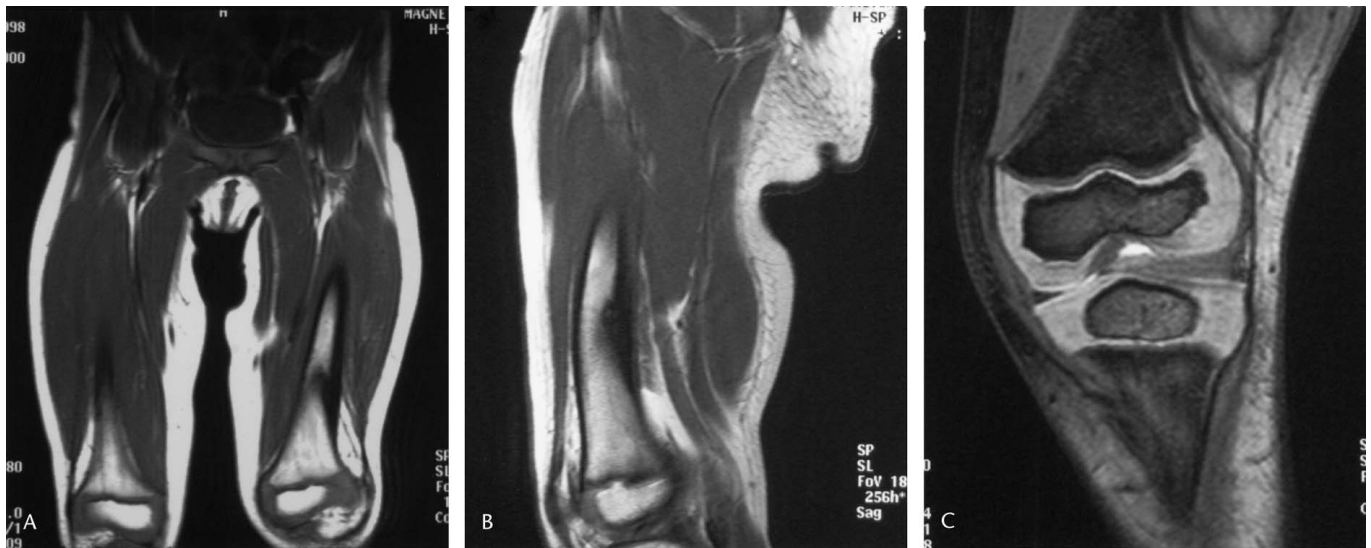


FIGURE 3. Preoperative MRI (A-C) of the distal femur showed increased signal intensity localized within the area of cortical thickening and a normal distal femoral physis, without evidence of premature fusion or cartilaginous bar formation.



FIGURE 4. Intraoperative anteroposterior (A) and lateral (B) arthrogram of the left leg.

deformity of 32° as measured by the tibiofemoral angle and a leg length discrepancy of 12 mm. All cases were treated surgically (1 medial hemicircumferential periosteal release, 2 corrective valgus osteotomies, 1 valgus osteotomy with concomitant application of an Ilizarov apparatus). Choi et al⁵ reported the only other known case of FFCD isolated in the posterolateral aspect of the distal femoral cortex described as a valgus deformity of 22°, 42° of anterior bowing, and a 3-cm shortened

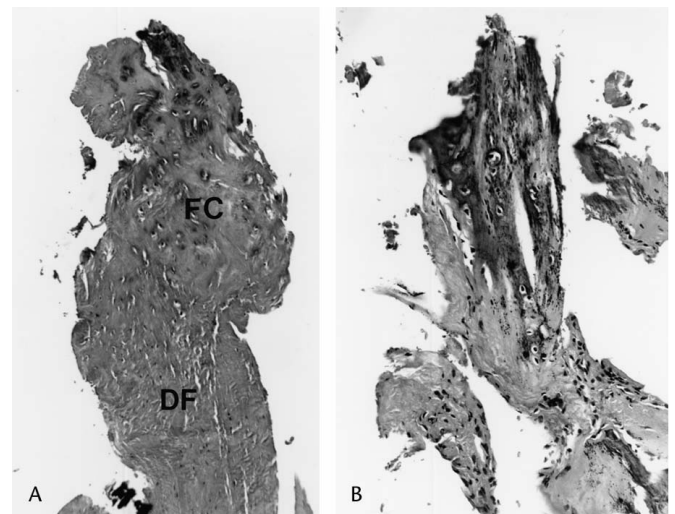


FIGURE 5. (A) Photomicrograph showing dense fibrous tissue (DF, lower section) with transition to fibrocartilage (FC, upper section) (H&E, original magnification $\times 70$). (B) Photomicrograph demonstrating tissue containing fibrocartilage undergoing mineralization of the matrix (H&E, original magnification $\times 100$).

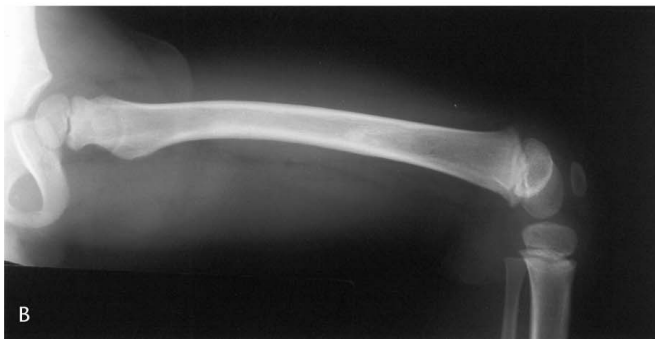


FIGURE 6. (A) Anteroposterior and (B) lateral radiographs of the left leg taken at 1 year postoperatively demonstrating a resolving angular deformity of the femur.

femur, as well as a laterally dislocated patella. Surgical treatment consisted of a corrective varus osteotomy and gradual correction with an Ilizarov apparatus.

Given the rarity of this condition, the natural history of this lesion has not been clearly established. Therefore, the probability that FFCF lesions in the lower extremity will resolve without treatment remains difficult to predict. While 45% of tibial FFCF lesions reported in the literature spontaneously resolved with nonoperative treatment over time, 5 of 6 cases (83%) of femoral lesions progressed and the sixth per-

sisted with clinical observation. It has been proposed that FFCF lesions located in the proximal medial tibia may be self-limiting^{3,4} and resolve with ossification beginning at 24 months of age.¹¹ The limited number of cases of distal femoral



FIGURE 7. (A) Anteroposterior and (B) lateral radiographs taken at 2 years postoperatively showing that restoration of the mechanical axis of the femur has occurred with proximal migration and gradual correction of the femoral deformity.

TABLE 1. Details of the Six Patients With FFCD Isolated in the Distal Femur

Author	Sex; Age at Presentation	Location	Initial Deformity	LLD	Deformity
Albinana et al ¹	F; 16 mo	L femur, PM	Varus 45°	15 mm	Progressed
Macnicol ¹⁷	M; 24 mo	L femur, PM	Varus 25°	10 mm	Progressed to 35°
	M; 14 mo	L femur; PM	Varus +	+	Progressed
Choi et al ⁵	M; 17 mo	R femur, PM	Varus 26°; AB 35°	10 mm	Progressed to 42°
	F; 15 mo	L femur, PL	Valgus 22°; AB 42°	30 mm	Persisted
Authors	F; 13 mo*	L femur, PL	Valgus 10°; mLDFA 77°	10 mm	Progressed to 30°; mLDFA 58°; AB 15°; ALB 34°

Author	Treatment	Age at Operation	Age at Latest Follow-Up	Intraoperative Findings; Pathology	Outcome at Latest Follow-Up
Albinana et al ¹	Medial hermicircumferential periosteal release	NR	33 mo	Abnormal anlagen periosteum; DF tissue with focal areas of FC differentiation	Resolving at latest follow-up
Macnicol ¹⁷	Valgus osteotomy	NR	NR	Dense medial femoral cortex, with fibrotic tissue folded within the lesion and attached to the periosteum; Dense collagenous matrix, with FC tissue merging with fibrous tissue	No limp, no LLD
Choi et al ⁵	Valgus osteotomy	NR	48 mo	NR; NR	No limp, no LLD
	Valgus osteotomy, Ilizarov lengthening	33 mo	66 mo	NR; DF and FC elements	Healed, straight leg
	Varus osteotomy, Ilizarov lengthening	25 mo	84 mo	NR; DF tissue invaginating the cortex, admixed with small areas of cartilaginous differentiation	Healed, straight leg
Authors	Curettage and excision	27 mo	51 mo	Fibrous band extending from the physis posterolaterally, penetrating the cortical lesion at the distal one-third of the femur; DF tissue with transition to focal areas of FC changes	Resolved

L, left; R, right; PM, posteromedial; PL, posterolateral; AB, anterior bow; ALB, anterolateral bow; mLDFA, mechanical lateral distal femoral angle; +, present but magnitude not reported; NR, not reported; DF, dense fibrous; FC, fibrocartilaginous; LLD, limb-length discrepancy.

*Patient presented to an outside institution at 13 months of age.

FFCD lesions in the literature limits our current understanding of the evolution of the secondary angular deformities and limb-length discrepancies and has delayed establishment of an accepted treatment protocol (ie, observation versus curettage versus corrective osteotomy).

Radiographs of our patient's femur revealed characteristic findings consistent with the diagnosis of FFCD, including well-defined sclerosis and cortical thickening at the site of the deformity.^{8,20} Occasionally, a prominent periosteal reaction

has been described.^{8,20} This was absent in our case. In addition, MRI of the area of interest showed findings consistent with those reported in a MRI study of FFCD isolated in the proximal tibia by Meyer et al,¹⁹ who recommend MRI evaluation of the lesion with atypical clinical presentation.

In contrast to the present case, Choi et al⁵ (case #2) used corrective osteotomy with an Ilizarov application for a patient who presented at 15 months with FFCD in the posterolateral cortex of the distal femur. Their patient had a comparable val-

gus deformity (22°) and a limb-length discrepancy (3 cm) to our patient. Intraoperatively, our patient was found to have a discrete fibrous band.² The focal cortical lesion was curetted, and the associated fibrous band was excised with its overlying periosteum.^{11,21,25} Nakase and colleagues²⁰ were the first to have reported curettage of FFCD as effective treatment of this lesion, followed by Khanna et al.¹² Our results confirm that curettage of this lesion occurring in the distal femur is an acceptable treatment alternative that promotes early healing and allows for full correction of persistent or progressive angular deformity. There was no evidence of recurrence at 24 months of follow-up.

Histopathologic findings of FFCD lesions in the femur and tibia have ranged from purely fibrous^{5,17,19,21} in nature to lesions including a fibrocartilaginous component^{1,3-6,8-12,14,16-18,20,21,25} and islands of hyaline cartilage.⁵ Given the broad spectrum of histologic features of FFCD, Kim and colleagues¹³ noted that a definitive distinction between FFCD and fibrous tether^{2,7} remains elusive and suggested that these may encompass a single clinical entity causing unilateral angular deformity in long bones of children. The histopathologic spectrum becomes blurred further and the diagnosis more challenging because fibrocartilaginous components are not essential for the diagnosis of FFCD. As a result, in 1999, Kim et al.¹³ suggested calling this lesion “subperiosteal fibrocartilaginous pseudotumor of long bones.”

Intraoperatively, we did observe a discrete fibrous band extending from the physis, which appeared similar to the entity described in four children by Beaty and Barrett.² This band became continuous with the adjacent cortical lesion, which was characterized as fibroconnective tissue with focal areas of fibrocartilaginous change, findings consistent with those reported in other cases of FFCD.^{3-5,10,13,19,21,25} In contrast, the other authors^{1,5,17} presenting patients with femoral FFCD lesions have reported that obvious fibrous bands were not seen adjacent to the FFCD lesions. Our case suggests that these two entities may occur simultaneously at the site of angular deformity.

We surmise that both the fibrous band in continuity with the FFCD lesion contributed to the patient's valgus deformity in the distal femur. While the fibrous band may act as a valgus producing tether about the knee,² the focus of fibrocartilaginous dysplasia penetrating the cortex potentially disrupts growth at the distal medial femur. It is not possible to speculate if either entity affected a dominant influence on the degree of valgus created.

In conclusion, recognition of focal fibrocartilaginous dysplasia as a cause of femoral deformity is important. Treatment with excision and curettage may be the definitive treatment and avoid the need for corrective osteotomy.

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