A Patient with Focal Fibrocartilaginous Dysplasia in the Distal Femur and Review of the Literature

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Focal fibrocartilaginous dysplasia (FFCD) is a rare and benign bone lesion that induces bowing deformity of the long bones in young children. Excessive production of fibrocartilage by abnormal differentiation in the metaphysis or by trauma during delivery or after birth is thought to cause growth disturbance. Radiologically, the lesion is characterized by a lucent defect with marginal sclerosis in the medial metaphysis of the long bone. However, there have been few reports about the initial radiological changes of FFCD before bowing started. We report a patient with FFCD in the left distal femur in whom the radiological changes were serially observed during the course of the disorder. A 2-week-old boy first visited our hospital because of left thigh pain. Plain radiographs did not show any abnormal findings at that time. At 10 weeks, a well-defined lucent defect with bony fragment inside was observed in the distal femoral medial cortex. At 1 year, this bony fragment gradually vanished but varus deformity progressed and reached approximately 40 degrees at the age of 2. After removal of the lesion, osteotomy and immobilization was performed with Ilizarov external fixator comprising rings, rods and wires. Complete bone union was achieved 3 months after operation. It is noteworthy that we could observe the initial radiological changes of FFCD before varus deformity occurred. As far as we know, there have been no descriptions of the bony fragment inside a lucent defect of the lesion. Radiological features may vary in the early phase of FFCD. ———— focal fibrocartilaginous dysplasia; femur; osteotomy; Ilizarov external fixator.


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FFCD in the femur and upper extremities was composed of dense fibrous tissue with areas of cartilaginous differentiation but without an abnormal tendon insertion (Ruchelsman et al. 2004; Smith et al. 2004). The periosteum was involved in a fibrocartilaginous lesion in the most cases and the term ‘fibrous periosteal inclusion’ was recently proposed (Jouve et al. 2007). They suggested that the fibrocartilaginous lesion was a kind of anchor preventing natural sliding of the periosteum during growth.

The natural history of FFCD is benign (Bradish et al. 1988; Choi et al. 2000; Santos et al. 2002; Jouve et al. 2007). It was reported that at least 45% of the cases in the literature healed spontaneously (Choi et al. 2000). Though the lesions initially progressed, spontaneous correction usually began around the age of 24 months because of the rupture of the fibrocartilaginous lesion (Jouve et al. 2007). However, spontaneous correction did not occur as expected in the femur and humerus (Rodríguez et al. 1998; Choi et al. 2000; Eren et al. 2006; Jouve et al. 2007). Femoral and humeral cases with severe deformity or shortening after conservative treatment were reported (Rodríguez et al. 1998; Eren et al. 2006). The potential for spontaneous correction may vary depending on the affected sites (Choi et al. 2000).

Radiologically, the lesion is characterized by a lucent defect with marginal sclerosis in the medial metaphysis of the long bone. We report a patient with FFCD in the left distal femur in whom the radiological changes were serially observed during the course of the disorder.

**Clinical Findings**

A 2-week-old boy was referred to our hospital because of left thigh pain. There were no abnormalities or complications in the gestation period and delivery. On physical examination, swelling, tenderness, and contracture of the knee and hip joints were absent. Laboratory data were within normal range. Plain radiographs did not reveal any abnormal findings (Fig. 1A). At 10 weeks after birth, a sclerotic change with bony fragment was found on the medial side of the distal femoral metaphysis (Fig. 1B). Differential diagnoses at that time were a bone tumor such as osteochondroma, fibrous dysplasia or a trauma.

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**Fig. 1.** Anteroposterior radiograph of the left femur. A, Two weeks after birth. Abnormal change was not apparent. B, Ten weeks. A sclerotic change (arrowheads) with bony island (arrow) became apparent. C, One year. The bony island became smaller (arrow) and varus deformity was apparent. D, Two years. Varus deformity progressed up to 40 degrees.
At the age of 1 year, the bony fragment almost vanished but a unilateral femoral varus deformity with a lucent defect and the marginal bone sclerosis at the medial aspect of the distal femur became apparent (Fig. 1C). Magnetic resonance imaging showed a mass at the defect. This mass had slightly lower intensities on T1-weighted images, higher intensities than those of muscles but lower than those of bone marrow on T2-weighted images (Figs. 2A, B). An extraosseous mass was absent. Computed tomography also showed severe varus deformity with medial cortical defect and marginal sclerosis (Fig. 2C). Bone scan with $^{99m}$Technetium showed a mild uptake in the distal femur (Fig. 2D). Open biopsy was performed and histological findings showed dense fibrous tissues without cartilaginous elements (Fig. 3). The varus deformity progressed subsequently and reached approximately 40 degrees at 2 years (Figs. 1D, 4). Because the varus deformity was progressive and was not acceptable from the biomechanical point of view, an operative correction was decided.

At operation, the longitudinal skin incision was made on the medial side of the femur. The fibrous mass was located within the defect, attaching to the surrounding periosteum. The mass was excised and osteotomy at the site of the defect was performed and immobilized with an Ilizarov external fixator. The deformity was corrected with hemicallotasis technique, unilateral lengthening of bone using external fixator (Fig. 5). Complete bone union developed 3 months after the operation and the fixator was removed. Radiographs taken 2 years after the operation showed a normal femorotibial angle and a limb length discrepancy with the affected side being 1 cm longer (Fig. 6).
DISCUSSION

An extensive literature survey reveals that there have been 89 (53 men and 34 women) patients of FFCD (Table 1) (Rodríguez et al. 1998; Ohno et al. 2005; Eren et al. 2006; Bakman and Monu 2007; Jouve et al. 2007). Most patients visited the hospital between 12 and 24 months because this disorder was usually noticed by the parents after the onset of ambulation. Otherwise, the patients with upper limb involvement visited at older ages. All the patients had unilateral limb involvement, 36 on the right and 48 on the left. FFCD was observed in the proximal tibia in 54 patients, distal femur in 17, distal ulna in 11, and proximal humerus in 3. Most patients had a varus deformity, but 2 in the tibial and 5 in the femoral patients had a valgus deformity. In the tibia, an operative treatment was chosen in 27 out of 54, observation (regular medical follow-up) in 25, and unknown in 2. In the femur, an operative treatment was performed in all the reported patients. There were 4 patients of serious complications after osteotomy: peroneal nerve palsy in 2

Fig. 4. The patient before operation. Varus deformity and shortening of the affected limb was apparent.

Fig. 5. Anteroposterior radiograph of the left femur in 1 month after osteotomy. Forty degree correction was completed without any complications. Arrows show the site of osteotomy.

Fig. 6. Two years after operation. The varus deformity was completely corrected. The affected limb was slightly longer.
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and valgus overcorrection in 2 (Jouve et al. 2007). These data revealed that approximately half of the patients with proximal tibial lesions showed spontaneous correction of the deformity. In contrast, surgical treatment was chosen in all the patients of femur probably due to progression of femoral deformity. It was also reported that the proximal humeral lesion led to severe shortening and angular deformity (Eren et al. 2006). It is not known why the tibia has the ability to spontaneously correct the deformity caused by FFCD, whereas the femur and the humerus do not. The natural course of FFCD varies greatly among those with different affected sites (Choi et al. 2000).

The etiology of these lesions remains unknown. This may be caused by the failure of differentiation of the mesenchymal anlage (Bell et al. 1985), or a trauma at delivery or other unknown trauma, and subsequent regeneration (Jouve et al. 2007). FFCD can be considered as an abnormal anchorage of the tendon or pathological fibrous bands in the metaphysis (Bell et al. 1985; Choi et al. 2000; Jouve et al. 2007). As a consequence, a disturbance of the natural sliding of the periosteum occurred during growth, resulting in varus or valgus deformity. A breakage of continuity of the lesion may induce natural correction of the deformity. The lesion shows various histopathological features, ranging from purely dense fibrous-tendon like tissue to benign fibrocartilaginous tissue (Kim et al. 1999). The present case showed pure dense fibrous tissue without cartilaginous components. It is suggested that FFCD may undergo evolutionary changes from its initial cellular phase in cartilaginous form to its late paucicellular phase in a more fibrous form (Choi et al. 2000). The presence of fibrocartilage is not an essential histopathological feature for FFCD (Kim et al. 1999; Choi et al. 2000). Fibrocartilaginous mesenchymoma, fibrous dysplasia or osteofibrous dysplasia was ruled out because of absence of epiphyseal plate-like cartilage, irregular woven bone or osteoblastic rimming (Isefuku et al. 1999; Hatori et al. 2002; Hatori et al. 2006).

The characteristic plain radiographic findings in the tibia are as follows: (a) a well-defined, obliquely positioned lucent defect in the medial tibial metaphyseal cortex, (b) sclerosis along the lateral border of the lesion, (c) absent bone margin superomedially, (d) a location distal to the proximal tibial physis (Rodríguez et al. 1998). There have been few descriptions about the initial radiological changes of FFCD. Only one patient that exhibited a periosteal reaction with an osteolytic lesion in the initial stage leading to the typical appearances was reported (Nakase et al. 1998). Fibrocartilaginous mesenchymoma, fibrous dysplasia or osteofibrous dysplasia was ruled out because of absence of epiphyseal plate-like cartilage, irregular woven bone or osteoblastic rimming (Isefuku et al. 1999; Hatori et al. 2002; Hatori et al. 2006).

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Therapeutic modality for this rare condition was described by Jouve et al (Jouve et al. 2007). They reported as follows: (a) In a patient younger
than 2 years old, 6-month follow-up was recommended to observe the behavior of the deformity; (b) In a patient around 2 years old and the deformity angle was over 20 degrees, curettage was recommended to accelerate the correction and to confirm the diagnosis histologically. However, it is unclear whether a severe deformity like the present case will be corrected by curettage alone, and whether such a severe deformity will be acceptable from biomechanical or cosmetic point of view for a long time. Like other reports of the femur (Rodríguez et al. 1998; Ruchelsman et al. 2004), an osteotomy corrected the varus deformity and achieved complete union in the present case without any complication. Pin and plaster is one of the most common operative procedures to maintain the corrected position. However, Choi et al. reported a patient with valgus overcorrection using this procedure (Choi et al. 2000). There have been few reports about application of Ilizarov external fixator. Ilizarov external fixator is a set of external fixator comprising rings, rods, and wires. This apparatus is different from the conventional one in that it encases the limb as a cylinder, the limb itself being shaped like one. We can adjust the deformity during or even after operation with this apparatus. Choi et al. also corrected deformity using Ilizarov external fixator with satisfactory results. In the present case, the external fixator was useful not only for the maintenance of the corrected position but also for further correction after operation or even lengthening of the affected limb. The osteotomy using Ilizarov apparatus should be taken into consideration in the patients with severe deformity.

References


