



Focal fibrocartilagenous dysplasia of the tibia : long-term evolution

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Focal fibrocartilagenous dysplasia of the tibia (FFCD) is a rare and benign condition associated with unilateral tibia vara in childhood. The first description was done by Bell in 1985. Since then more than sixty cases have been reported. The aetiology remains unknown.

Five new cases are retrospectively reported. Four of them with tibia vara less than 30° showed a spontaneous correction. One patient had a severe varus deformity (> 30°) leading to physal impairment and underwent a tibial valgus osteotomy.

Biopsy is not necessary as radiographs are typical.

The infantile growth plate is able to correct a tibia vara due to FFCD if less than 30°. In case of spontaneous resolution, a long-term follow-up remains necessary because of a possible progressive leg length discrepancy.

Keywords : tibia vara, childhood, focal fibrocartilagenous dysplasia.

tibia causing a valgus deformity (20), in the femur (1, 5, 7, 13, 15) and in the upper limb (14, 21).

Diagnosis is usually done by plain radiographs (fig 2). Typical radiographic signs are a well-defined, obliquely positioned, lucent defect in the medial tibial metaphyseal cortex, sclerosis along the lateral border of the lesion, absence of a bone margin superomedially and a location distal to the proximal tibial physis (9). MRI can be performed (fig 3) in doubtful cases. MRI also excludes a soft tissue lesion. The typical MRI appearance of FFCD on both T1-weighted and T2-weighted slices discloses a low-signal area (corresponding to the radiolucent area) and an intermediate signal (corresponding to the distal sclerosis) (16).

No biopsy is required as radiographs are typical. Spontaneous resolution can occur without any treatment (6). Corrective osteotomy is rarely required, and only in case of severe deformity or varus progression.

INTRODUCTION

Focal Fibrocartilagenous Dysplasia (FFCD) is a rare and benign condition of unknown aetiology associated with unilateral tibia vara in childhood (fig 1). Since Bell *et al* (3) reported the first case of FFCD in 1985, more than sixty cases have been reported. FFCD has the same prevalence in both sexes and an even distribution between right and left knee (5, 9). The lesion arises at the insertion of the pes anserinus tendon (3). Other localisations of FFCD have been described lateral in the proximal

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Fig. 1. — Clinical appearance of a 24-month-old girl with left tibial FFCD.

Recognition of FFCD is very important because it avoids biopsy or invasive treatment, as many case reports have shown (5, 8-12, 14, 18).

PATIENTS AND METHODS

From 1991 to 2002, five infants (four girls and one boy) with FFCD were referred to our institution by their paediatricians or general practitioners (table I). The mean age at presentation was 24 months (range, 15 to 36 months). All these patients had progressive unilateral tibial bowing (fig 1). There was no side predominance. All infants were healthy and pain free. There was no history of trauma, infection or metabolic bone disease. No relevant family history was found. One was in breech presentation at birth and another remained a long-time in breech position but changed to a cephalic presentation at birth.

Diagnosis was obtained by plain radiography. MRI had been performed in only one case. No biopsy was performed.

No treatment was applied initially except for two patients (case 2 and 4). Case 4 was treated with valgus night splints. Case 2 presented with an uncommon radiographic appearance with physeal impairment (fig 4a). The typical picture of FFCD was present with a cortical defect in the medial tibial metaphysis, surrounded by sclerosis. But the varus deformity was so important (30°) that physeal involvement occurred with metaphyseal fragmentation consistent with secondary Blount's disease. At 27 months of age, a corrective



Fig. 2. — Typical radiographic picture of tibial FFCD : radiolucent area in the medial metaphysis (black arrow) surrounded by sclerosis (white arrows).

valgus osteotomy was performed with hypercorrection in order to protect the medial part of the physis (fig 4b).

Radiographic follow-up was performed with serial clinical examination and weight bearing goniometry.

Mean follow-up was 6.2 years (range, 1.2 to 14.6 years).



Fig. 3. — MRI aspect of tibial FFCD : T1-weighted slice shows a low signal corresponding to cortical lucency in the medial part of the metaphysis (arrows).

RESULTS

The results are shown in table I. Four patients showed spontaneous improvement. Clinical and radiological examination showed progressive regression of the varus deformity after a mean of 26 months (range, 14 to 42 months). The cortical defect disappeared after a mean of 34 months (range, 14 to 65 months). At latest follow up, three children had a leg length discrepancy of 10 mm or more (10, 11 and 13 mm respectively).

Case 2 had been hypercorrected in order to protect the impaired medial physis. At latest follow-up



Fig. 4. — Atypical radiographic appearance of tibial FFCD. Radiolucent area in the medial metaphysis (large black arrow) surrounded by sclerosis (small black arrows). Impairment of the medial physis and fragmentation of the metaphysis (white arrow). 4-b : result 2 months after corrective valgus osteotomy showing healing of the metaphysis.

(4.8 years of age) the radiograph showed complete healing of the physis. The mechanical axis was still in valgus (10 degrees) and leg length discrepancy was 10 mm.

DISCUSSION

FFCD is an uncommon and benign disorder that has been associated with unilateral tibia vara in childhood after walking age. Up to now no patient with bilateral involvement has been reported. The majority of case reports concern Caucasian children, but FFCD has also been described in Asian children (7, 13) and in black infants (8). Age at presentation in the lower extremity is from 3 months to 28 months (5), but in some cases the deformity was first noted at birth (16). In our series one girl was 36 months old at presentation (case 1). In the upper limb, age at presentation varies from 6 months to 4.8 years (21).

Table I

Case	Sex	Age at presentation (months)	Side	Initial Varus deformity (°) and LLD (mm)	Treatment	Age at latest follow-up (years)	Follow-up (years)	Final Varus/Valgus deformity (°) and LLD (mm)
1	F	36 m	L	-VR : 15°	observation	17,6	14,6	-VL : 2° -LLD : 11
2	F	24 m	L	-VR : 30° -LLD : 5	valgus osteotomy	4,8	2,8	-VL : 10° -LLD : 10
3	M	29 m	R	-VR : 19° -LLD : 2	observation	12	9,6	-VR : 0° -LLD : 8*
4	F	15 m	L	-VR : 22°	night splints	4,2	2,9	-VR : 2° -LLD : 13
5	F	17 m	R	-VR : 22° -LLD : 4	observation	2,6	1,2	-VL : 2° -LLD : 3

(F = female, M = male, L = left, R = right, VR = varus, VL = valgus, LLD = leg length discrepancy, mm = millimeter, *affected side is longer than normal side).



Fig. 5. — Long-term evolution of a left tibial FFCD from the age of 3 to 17.5 years. Initial radiographic presentation at 36 months. Progressive healing (radiographs at 4.8 years, 6.1 years and 8.4 years). Final result at 17.5 years of age with persistence of a 11-mm leg length discrepancy due to tibial discrepancy.

Clinical features of tibial FFCDD are painless unilateral tibia vara, medial tibial torsion usually accompanied by leg length discrepancy with limping or normal walking. Leg length inequality can be very significant up to 30 mm (3, 17, 22). In the upper limb 7.7 cm of limb length discrepancy has been reported (14). In most cases the deformity has increased both clinically and radiologically after diagnosis, probably because of weight bearing and force distribution imbalance (8). No fracture has been reported in association with FFCDD in the lower limb but in the upper limb one patient presented with a non displaced fracture of the humerus through the maximal bowing area after a fall (14).

The pathogenesis of FFCDD remains unknown. Bell *et al* (3) theorized that a failure of differentiation of the mesenchymal anlage in the area of the pes anserinus and the persistence of a focus of fibrocartilage could hamper the growth on the medial aspect of the proximal tibia. This is not applicable in cases of FFCDD without abnormal tendon insertion as in the upper limb or femur (14, 15). Some authors thought that a focal area of fibrocartilage seems to act like a tether (15). Jouve *et al* (11) hypothesized that FFCDD is a pathology of the pes anserinus insertion which interferes with its physiological migration during growth by creating a pseudo-epiphysiodesis. Langenskiöld suggested that trauma during delivery could be a predisposing factor by causing necrosis of the medial part of the physis (22).

A full 99m Technetium body scan performed by Albiñana *et al* in three cases revealed a mild reactive lesion in the proximal tibia (1) whereas Jouve *et al* did not find any abnormality in one case (11).

In most of the reported cases of FFCDD a biopsy was performed either during corrective osteotomy or by needle biopsy for diagnostic reasons. There were various histopathological findings: dense fibrous and/or hyaline cartilaginous tissue or a combination of both (5, 17, 22). Paucicellular areas of the lesion were composed of dense fibrous and cellular areas of fibrocartilage (7, 13, 18). A biopsy is no longer recommended. Kim *et al* (13) found that histopathology of individual cases was very different, suggesting that the condition is evolutionary. They proposed to rename FFCDD "subperiosteal

fibrocartilaginous pseudotumor of long bone". Beaty and Barret reported four cases of unilateral deformity of the distal femur caused by a focal fibrous tether of which a biopsy showed the presence of fibrous elements only (2).

Differential diagnosis of FFCDD of the tibia includes several conditions such as Blount's disease, benign tumours (chondroma, chondromyxoid fibroma, eosinophilic granuloma, non-ossifying fibroma, lipoma, osteoid osteoma, fibrous dysplasia), fracture malunion, neurofibromatosis, Ollier's disease, osteomyelitis, rachitic tibia and trauma.

In 1985, Bell *et al* (3) described in their series a spontaneous resolution of FFCDD. Conservative treatment has proved that tibia vara caused by FFCDD can spontaneously resolve in 45% of cases (5). Observing the healing process of FFCDD, Kariya *et al* (12) demonstrated that an infantile active growth plate of the proximal tibia is able to correct a varus deformity of up to 30°. In cases treated with osteotomy, peroneal nerve palsy (1, 5) and overcorrection to valgus deformity have been reported as a consequence of osteotomy (5). All authors recognize that surgical treatment can be avoided if there is no increasing deformity or excessive angular deformity at presentation. Radiographs and physical examination are recommended to evaluate the healing process. In all reported cases there has been no recurrence of the deformity either after corrective osteotomy or spontaneous resolution.

In our series we observed spontaneous resolution in four cases with tibia vara of less than 30°. Although a complete healing of the lesion was obtained, a leg length discrepancy persisted. According to our results we continue to abide by a conservative attitude but we recommend a long-term follow-up to skeletal maturity in order to assess a potential leg length discrepancy. In case of severe tibia vara (> 30°), a growth plate involvement can occur and renders surgery necessary.

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