Tibia vara in focal fibrocartilaginous dysplasia

A report of 2 cases

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Submitted 91-12-26. Accepted 92-01-19

Focal fibrocartilaginous dysplasia, which has only recently been recognised, is characterised by severe unilateral tibia vara in early childhood. Radiography typically shows a cortical defect with a surrounding area of sclerosis between the metaphysis and diaphysis of the medial proximal tibia.

The first case descriptions were published by Bell et al. (1985), who called the lesion Focal Fibrinocartilaginous Dysplasia (FFD) because in two of their reported cases the biopsy showed dense hypocellular tissue resembling fibrocartilage in some areas and tendon in others. The etiology is obscure. Hitherto only 12 cases have been reported. The author adds another two cases.

Case 1

A girl, who started to walk at the age of eight months developed a progressive varus deformity in the right knee. There was no history of pre- or postnatal abnormality, or a history of trauma, infection or bone disorders in the family.

At the age of one year her right leg had a severe varus deformity and medial tibial torsion. Radiographs disclosed the characteristic lesion in the right leg (Figure 1). Four months later her right leg had 45° varus deformity with tibial torsion, and radiographs showed the same characteristic appearance. The lesion was explored and found to be a plug resembling cartilage into which the pes anserinus was inserted. Microscopic examination showed hypocellular tissue of fibrocartilaginous appearance with some thick-walled blood vessels in some areas and tendon in others. No treatment was given. At the age of 3 years the same clinical and radiographical features were found with a shortening of the right leg by 1.5 cm.

At the age of 8 years, there was complete resolution, with normal, straight legs. Radiographs showed complete healing with a slight curvature of the tibia. The results of the final clinical and radiographic examinations at 30 years of age were normal.

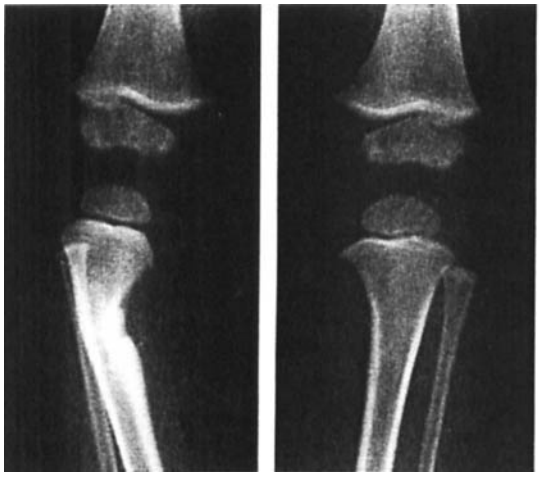
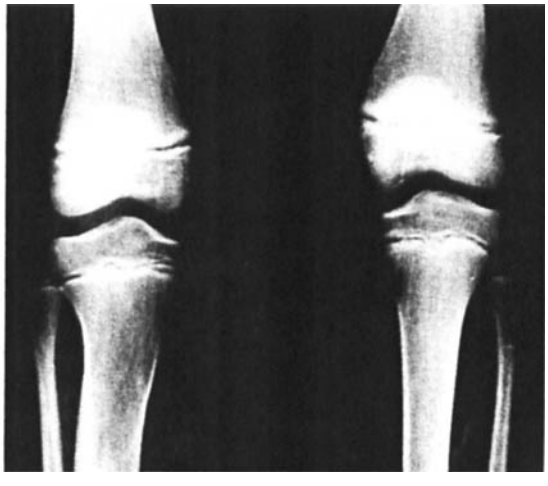
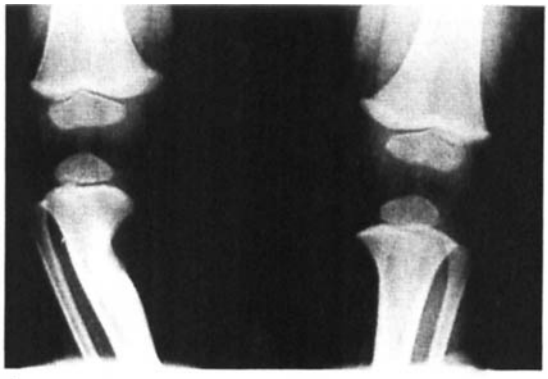


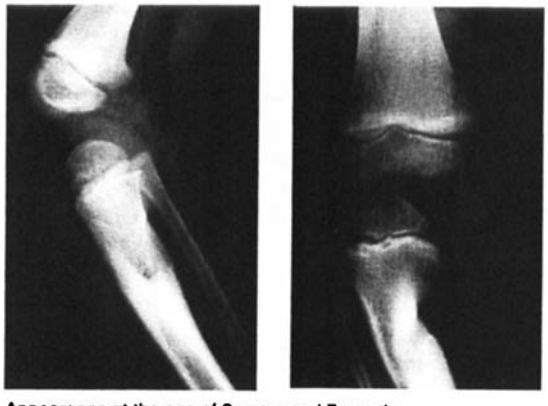
Figure 1. Case 1. At presentation, aged 1 year.



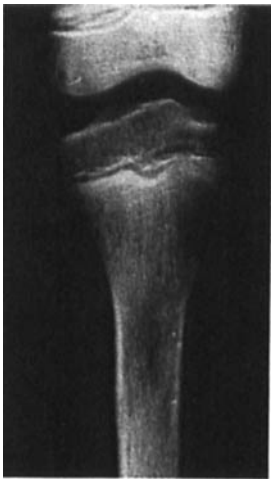
At the age of 8 years.



At presentation, aged one year and three months.



Appearance at the age of 2 years and 7 months.



Normal clinical and radiographic features at final examinaton, 7 years old.

Figure 2. Case 2.

Case 2

A girl, who started to walk at the age of one year developed progressive varus deformity in the right knee with limping. There was no history of pre- or postnatal abnormality, or a history of trauma, infection or bone disorders in the family. On examination at the age of one year and three months her right leg had severe varus and medial tibial torsion. Radiographs disclosed the characteristic lesion in the right leg (Figure 2), and two months later, tomography showed a characteristic cortical defect with varus deformity, but with no signs of a nidus. At the age of two years and seven months 30° varus deformity was present, with medial tibial torsion and limping caused by the shortening of the right leg by 3 cm. Radiographs showed the same characteristic appearance.

A *biopsy* was taken at the age of three years and seven months. The finding was identical to that described by Bell et al. (1985), with dense hypocellular tissue resembling fibrocartilage in some areas and tendon in others. *Scintimetry* showed a high uptake focally within the proximal medial part of the right tibia at the junction of the metaphysis and the diaphysis. *Computed tomography* showed changes in the cortex and increased density of the fatty tissues within the area concerned.

The final examination at seven years of age showed normal features both clinically and radiographically.

Discussion

The 14 cases of FFD reported (Bell et al. 1985, Bradish et al. 1988, Husien and Kale 1989, Olney et al. 1990, this report) show identical clinical, radiographic and histological features that have been recognized only recently. There are 7 boys and 7 girls. The onset of the varus deformity occurred at the age of 11 (3–18) months, while the age at presentation was 18 (5–30) months. Seven cases received no treatment with spontaneous resolution at an average age of 62 (43–89) months, while 7 cases were treated by osteotomy at the average age of 23 (6–30) months, i.e., before spontaneous resolution could take place. The question arises as to whether osteotomy is necessary for such young children. I agree with Bradish et al. (1988) that resolution would have occurred in all patients, and that osteotomy can be avoided.

The etiology of FFD is unkown. Bell et al. (1985) suggest that the mesenchymal anlage of the tibial metaphysis has, for unknown reasons, developed abnormally at the insertion of the pes anserinus. Langenskiöld (personal communication) recognized a similarity between the lesion and his findings in rabbits when localised damage to the growth plates was provoked with X-rays (Langenskiöld and Edgren 1949). He suggested that necrosis of the medial part of the physis, due to trauma at delivery, could be the predisposing factor, with later regeneration and a diaphyseal defect.

Unilateral tibia vara in childhood may be seen in infantile tibia vara (Blount's disease) which is a growth disturbance involving the metaphysis, the physis and the epiphysis of the posteromedial part of the proximal tibia (Zayer 1973), while FFD is a cortical defect outside the growth area with severe varus, always unilateral; it never shows the features of other conditions such as Ollier's disease or neurofibromatosis.

References

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