Case 1236

Eurorad

Focal fibrocartilaginous dysplasia of the tibia

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Section: Paediatric radiology
Imaging Technique: MR
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Case Type: Clinical Cases
Authors: E.V. Wood, C. Harris
Patient: 1 years, female

Clinical History:

A case of unilateral tibia vara in an infant.
Imaging Findings:

The patient presented with a 4-month history of increasing varus deformity of the left tibia, noticed by her parents. She was born by planned caesarean section, birth weight 8lbs and had otherwise normal development thus far. Plain films of her lower limbs were taken, revealing a well-defined lesion in the medial aspect of the left proximal tibial metaphysis, with angulation occurring at this site (Fig 1). Initial diagnosis was unclear, but the possibility of congenital pseudarthrosis was raised.
Further imaging was performed in the form of MRI and CT (Fig 2). MRI confirmed a defect at the medial border of the affected tibia, with deformity occurring around this point. There was abnormal marrow signal in the defect; the surrounding soft tissues were normal. CT confirmed these findings.

A biopsy was subsequently undertaken, and histology revealing a prominent layer of fibrous tissue and a thick layer of dense collaginous tissue. This established the diagnosis as Focal fibrocartilaginous dysplasia (FFCD).

The child has been observed over the course of three years and gradual correction of the deformity has been seen, both clinically and on plain films (Fig 3), without any further intervention.

Discussion:

Focal fibrocartilaginous dysplasia is a rare cause of unilateral tibia vara in young children and was first described by Bell in 1985 [1]. It usually presents as a progressive varus deformity of the tibia, often first noticed by the parents. The aetiology is unknown. Bell hypothesises that it is the result of abnormal differentiation of the mesenchymal anlage in the region of the pes anserinus [1].
Characteristic findings on plain film are of a well-defined lucency in the cortex of the medial part of the proximaltibial metaphysis at the insertion of the pes anserinus. Just distal to and at the lateral border of the area of lucency, there is often corresponding cortical sclerosis [4]. Varus angulation of the tibia begins at the level of the defect. These findings alone, if appreciated, are enough to make the diagnosis of FFCD.

Lesions including non-ossifying fibroma, neurofibromatosis, eosinophilic granuloma, chondromyxoid fibroma, and

osteoid osteoma, may also cause elements of the above. However these conditions usually produce soft tissue masses (not seen with FFCD), or do not cause a varus deformity.

If there is doubt over the findings, further imaging such as CT or MRI can be used as an adjunct to diagnosis.

FFCD has a characteristic MRI appearance [3]. Again, there is a demonstrable focal defect correlating with the plain films, without a soft tissue mass. The area of cortical lucency shows as low signal on both T1- and T2-weighted images, consistent with dense fibroconnective tissue. Areas of sclerosis are shown as low signal on T1 and intermediate in T2, consistent with reactive bone and callus.

CT scanning demonstrates an elliptical cortical defect without a discreet soft tissue mass. The soft tissue medial to the defect is indistinguishable from the muscle and tendon [4].

Previously it has been recommended that corrective osteotomy be undertaken in all of these patients [2]. It is now thought that the natural history of FFCD is to resolve spontaneously over the course of a few years [5], occasionally with initial further progression before the deformity corrects. Surgery is therefore not necessary and has potential for complications, such as neurological damage and creation of an iatrogenic valgus deformity [5]. It has been recommended that the progress of these patients be monitored with serial radiographs and clinical assessment.

The diagnosis of FFCD is primarily radiological, and if made correctly avoids the need for biopsy or unnecessary surgical intervention such as corrective osteotomy or biopsy.

Differential Diagnosis List: Focal fibrocartilaginous dysplasia

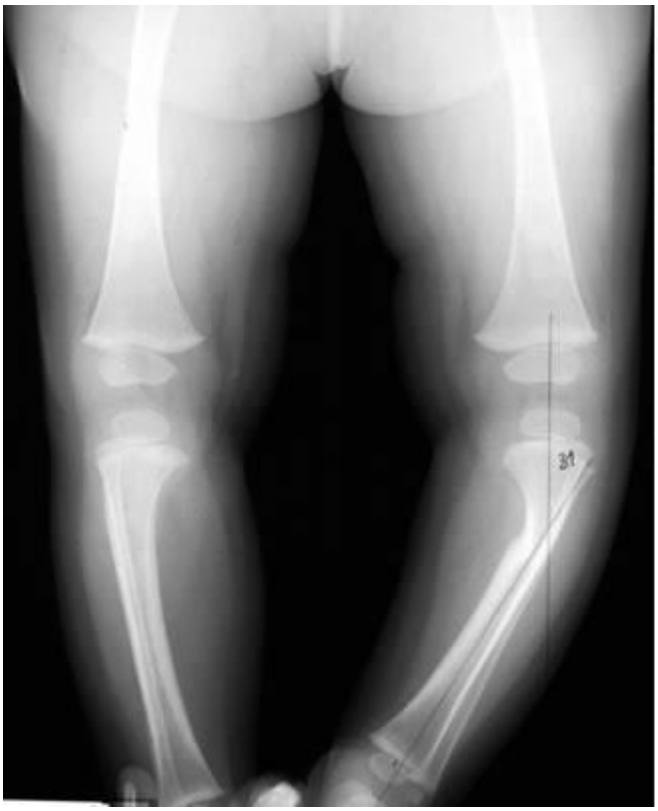
Final Diagnosis: Focal fibrocartilaginous dysplasia

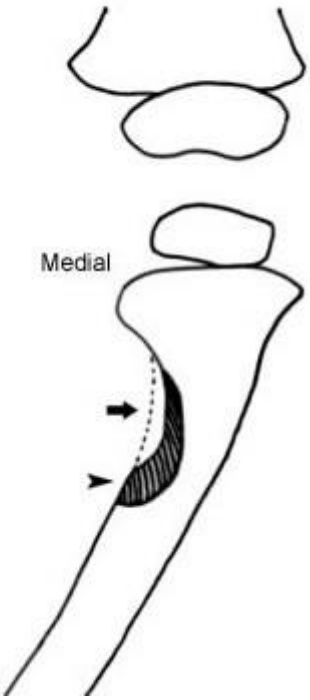
References:

Bell SN, Campbell PE, Cole WG, Menelaus MB. Tibia vara caused by focal fibrocartilaginous dyslpasia. J Bone Joint Surg [Br] 1985;67:780-4. (PMID: 4055881)
Olney BW, Cole WG, Menelaus MB. Three additional cases of focal fibrocartilaginous dysplasia causing tibia vara. J Paediatric Orthop 1990;10:405-7. (PMID: 2355089)
Meyer JS, Davidson RS, Hubbard AM, Conard KA. MRI of focal fibrocartilaginous dysplasia. J Pediatr Orthop 1995;15:304-6. (PMID: 7790484)
Herman TE, Siegel MJ, McAlister WH. Focal fibrocartilaginous dysplasia associated with tibia vara. Radiology 1990;177(3):767-8. (PMID: 2243986)
Bradish CF, Davies SJM, Malone M. Tibia vara due to focal fibrocartilaginous dysplasia: the natural history. J Bone Joint Surg [Br] 1988;70-B:106-8. (PMID: 3339040)

Figure 1

a

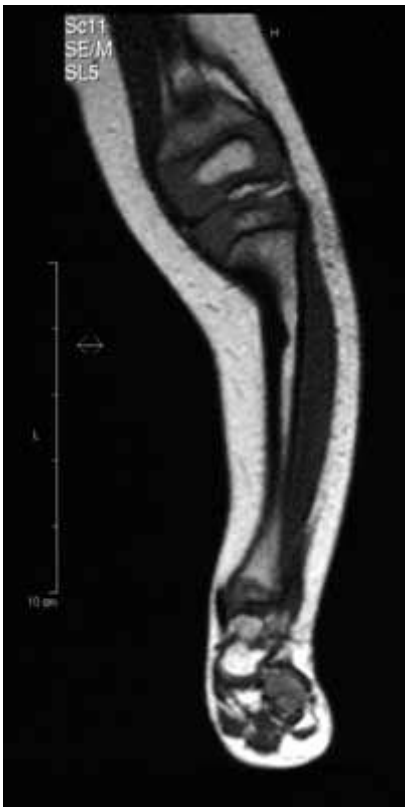




Description: Anteroposterior bilateral view of lower limbs showing a well-defined lesion in the medial aspect of the left tibial metaphysis, with marked varus angulation occurring at this site.
 The diagram demonstrates:
arrow - Cortical lucency,
 arrowhead - Area of sclerosis distal and lateral to lucent area.
 Note that angulation occurs at the level of the lesion, and the absence of a soft tissue lesion. **Origin:**

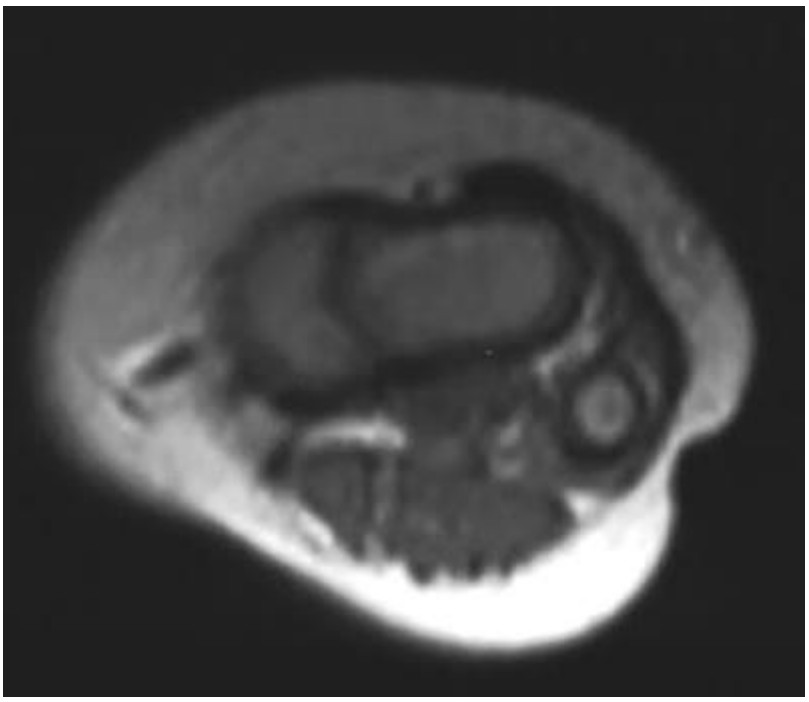
Figure 2

a



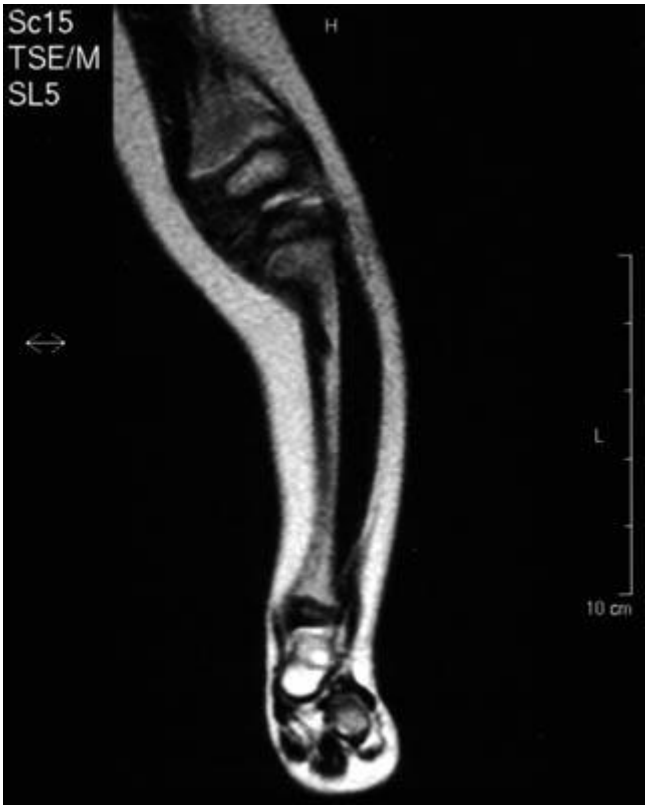
Description: T1 Coronal: MRI confirms the defect at the medial border of the tibial metaphysis. There is abnormal marrow signal in the defect but the surrounding soft tissues are normal. Cortical defect is shown as low signal on both T1 and T2 images, in keeping with dense fibroconnective tissue. Sclerotic areas show as intermediate signal on T2 and low on T1. **Origin:**

b



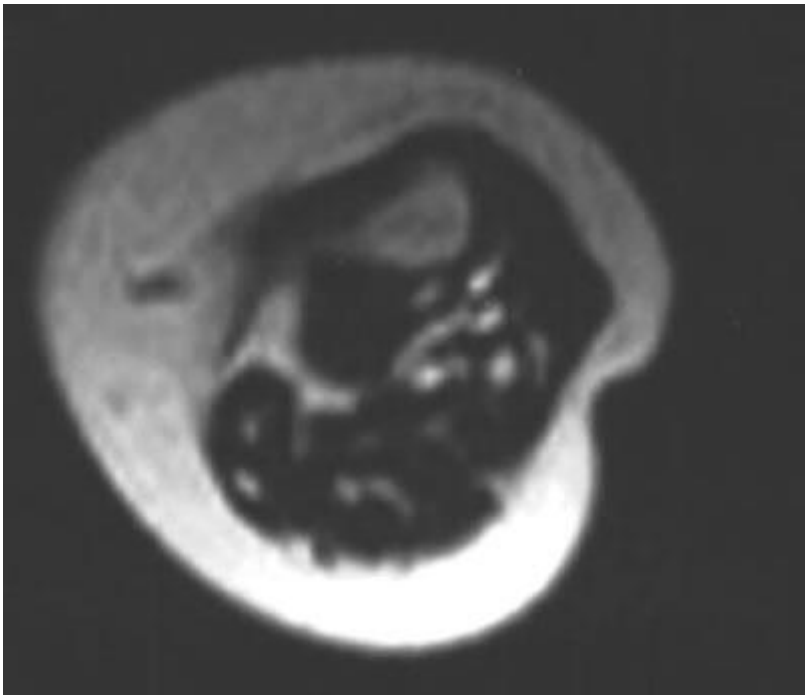
Description: MRI T1 axial **Origin:**

c



Description: MRI T2 Coronal **Origin:**

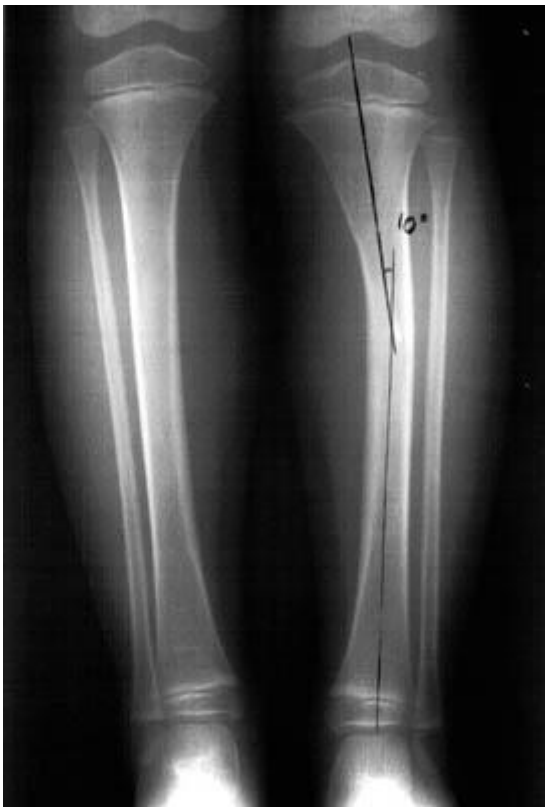
d



Description: MRI T2 axial **Origin:**

Figure 3

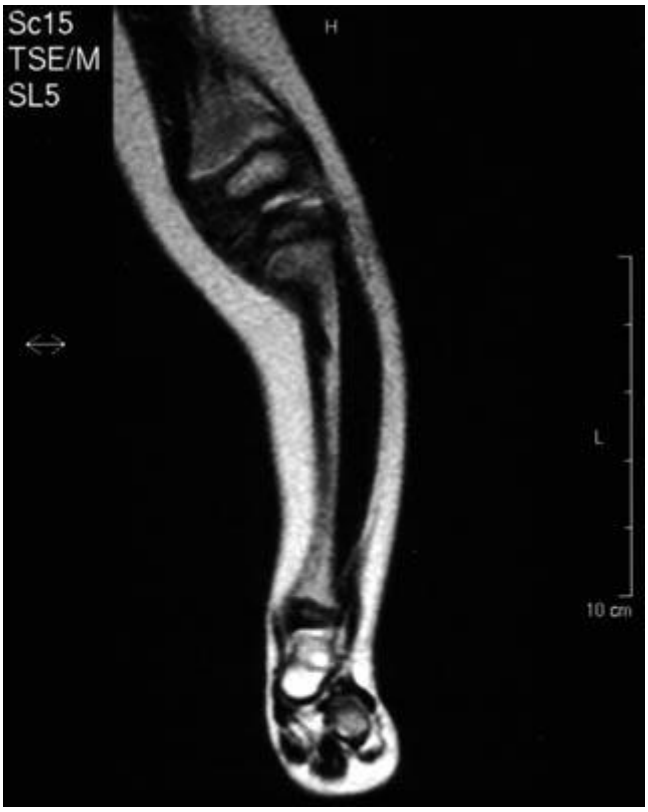
a



Description: These films were taken approximately 3 years following presentation. A large degree of correction has occurred. **Origin:**

Figure 4

a



Description: **Origin:**

Figure 5

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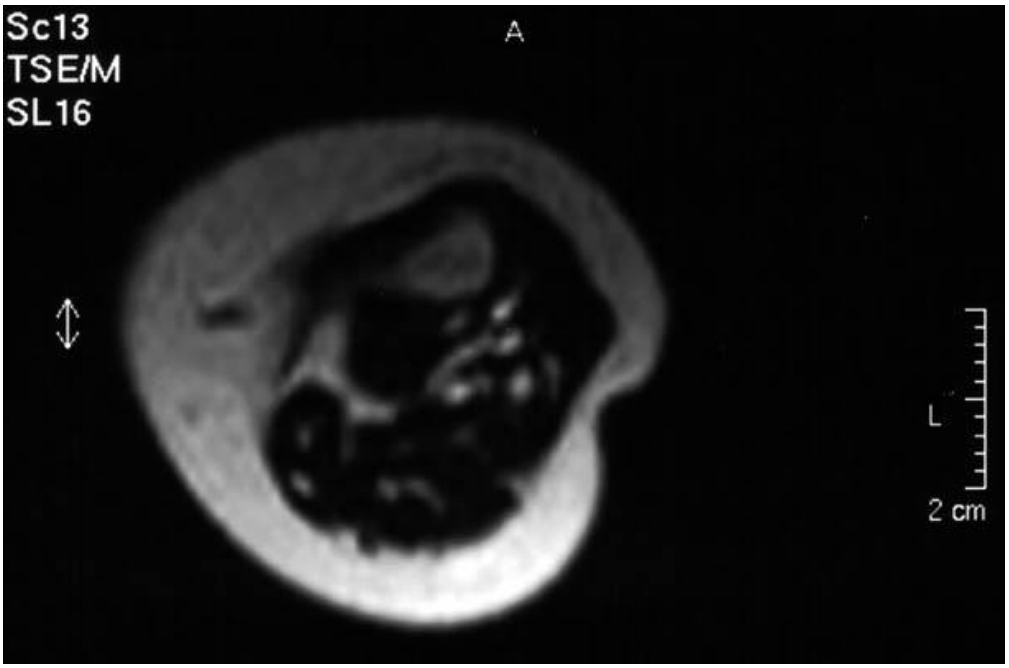

Description: **Origin:**

Figure 6

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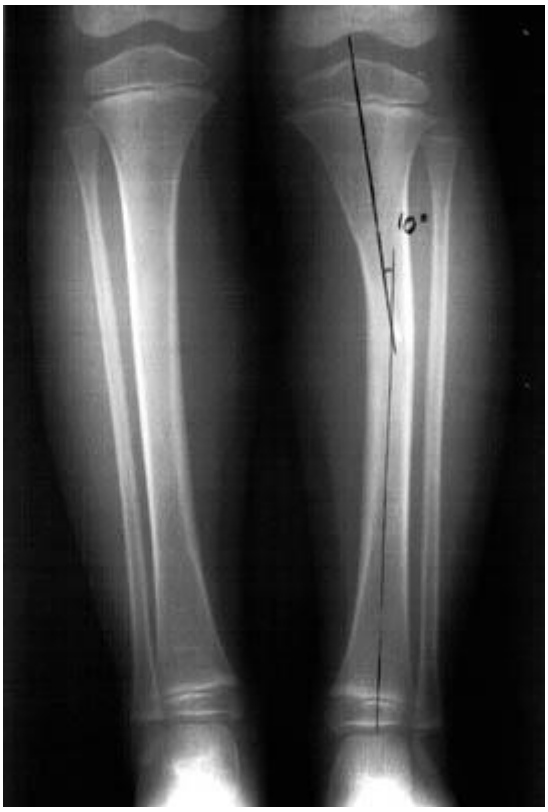

Description: Plain film bilateral lower limbs, taken approximately 3 years following presentation **Origin:**