

Unilateral tibia vara in a toddler caused by focal fibrocartilaginous dysplasia

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ABSTRACT

Focal fibrocartilaginous dysplasia (FFCD) of the tibia is a rare tumor like lesion probably caused by a failure of differentiation in the pes anserinus area. The children usually present with unilateral tibia vara and conspicuous walking features. The radiographic appearance of FFCD is pathognomic. In most cases this benign condition undergoes spontaneous resolution. Curettage or corrective osteotomy is only indicated when the deformity is persistent or progressive. We report the case of a 14 months old toddler diagnosed with FFCD. The characteristic radiographic and MRI features are presented. Further, we present for the first time the sonographic appearance of FFCD.

CASE REPORT

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A 14 months old male toddler was referred to our hospital because of unilateral bowlegs. At the age of 12 months the parents had noted conspicuous limping and frequent stumbling. There was no history of fever or pain. Inspection revealed discrete hyperpigmentation above the tibial tuberosity.

Radiographs of the lower limb depicted a well defined elliptic lucent defect in the medial cortex of the proximal tibia and cortical sclerosis along the lower border (figure 1). The defect was clearly separated from the metaphyseal bone marrow by a segment of intact cortex. Ultrasonography showed a hypoechoic lesion (15 x 10 x 5 mm) visible upon the cortex (figure 2). The cortex was interrupted in just one place about 25 mm distal to the metaphysis as seen in a gaping fracture. The findings in the surrounding soft tissue were unobtrusive. There was no proof of hyperperfusion in color duplex sonography. For further characterization of the

apparent benign lesion an MRI was performed (1,5 T, Signa GE). MRI revealed a lesion hypointense on T1 as well as on T2 weighted images without uptake of contrast media after injection of Gadolinium (figure 3). The signal of the bone marrow was normal for the patient's age.

The findings of the imaging studies were interpreted as a benign focal fibrocartilaginous lesion, biopsy was considered unnecessary, as the features of this lesion are characteristic. On the orthopedic part solely a shoe elevation (3 mm) was attached on the medial foot border for a period of six months. Four years follow-up showed complete restitution of the defective position and resolution of the varus deformity without residual standing or walking disability (figure 4).

DISCUSSION

Bilateral bowlegs in toddlers is well known as physiologic tibia vara. Unilateral tibia vara is an uncommon feature and is

seen in only four conditions: 1) infantile tibia vara (Blount's disease), 2) adolescent tibia vara caused by partial closure of the growth plate after trauma or infection between the ages of six and 13 years, 3) late-onset tibia vara in obese black children between the ages of six and 15 years, 4) tibia vara caused by focal fibrocartilaginous dysplasia (FFCD) (1).

FFCD is a benign condition first described in 1985 by Bell et al. (2), which is probably caused by a failure of differentiation in the pes anserinus area resulting in a growth disturbance of the proximal tibia along with varus deformity. Up to date there have been about 92 published cases of FFCD, the majority occurring in the proximal tibia (n=67), fewer in the femur (n=11), ulna (n=7), humerus (n=3), radius (n=2) or phalanx (n=2).

The radiographic appearance of FFCD is pathognomic. Diagnosis can be confirmed by MRI in ambiguous cases. In most patients the deformity resolves spontaneous, in more severe cases curettage or osteotomy might become inevitable.

FFCD is a rare tumor like lesion probably caused by displaced islets of the growth plate into the cortex of the juxtametaphyseal region. Jouve et al. proposed the term "fibrous periosteal inclusion" as they believed that this entity represents a bony anchor preventing natural sliding of the periosteum during growth (3). Histological these lesions show regional variation in cellularity, amount of fibrous and cartilaginous components (4).

Most important differential diagnosis is unilateral Blount's disease, a developmental disorder with progressive genu varum (5). Further differential diagnosis include dyschondrosteosis, fibrous dysplasia, Ollier's disease, neurofibromatosis or trauma (2). A fibrous tether at the distal aspect of femur may also cause genu valgum or genu varum, depending on the location of the tether. In contrast to FFCD these lesions are purely fibrous and show no evidence of any cartilaginous or osseous elements (6).

The children usual present at the age of 12-24 months because of unilateral tibia vara, slight shortening of the leg and distinctive walking features observed by the parents. If they have reached standing age, hyperextension of the knee may also be present. Patients with upper limb involvement, however, usually present at older ages (7).

FFCD is a benign lesion and nearly 45% of the tibial FFCD undergo spontaneous resolution. Corrective osteotomy is indicated when the deformity is persistent or progressive (8). In some cases curettage proved to be an alternative procedure to conservative management or more radical surgery (9).

No biopsy is required as the radiographic appearance of FFCD is pathognomic. FFCD of the tibia appears as a well-defined elliptic lucent defect in the medial metadiaphyseal cortex along with sclerosis on the lateral border of the lesion in combination with unilateral tibia vara. It's the radiologist's responsibility to identify this benign lesion in order to avoid further invasive diagnostic procedures or even unnecessary surgery. MRI is needed only in ambiguous cases. We report

for the first time the sonographic appearance of FFCD. To our knowledge this is the first description of the sonographic features of FFCD. Ultrasound is an important imaging modality used in children especially because of the availability and the lack of radiation exposition. It's an appropriate examination especially in the follow-up process of FFCD once the lesion has been characterized radiographically.

TEACHING POINT

Focal fibrocartilaginous dysplasia (FFCD) of the tibia is a rare tumor like lesion with pathognomic radiographic appearance. MRI is needed only in ambiguous cases and ultrasound can be used for follow-up as nearly 45% of the tibial FFCD undergo spontaneous resolution.

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FIGURES



Figure 1: 14 months old boy with unilateral tibia vara caused by focal fibrocartilaginous dysplasia. Radiographs (a.p. and lateral projection) revealed a well defined lucent defect in the medial cortex of the proximal tibia without aggressive features to indicate malignancy. Cortical thickening was visible below the defect which gave the impression of slight cortical compression. Further, medial bending in the metaphysis was existent, as can be seen in a fracture.

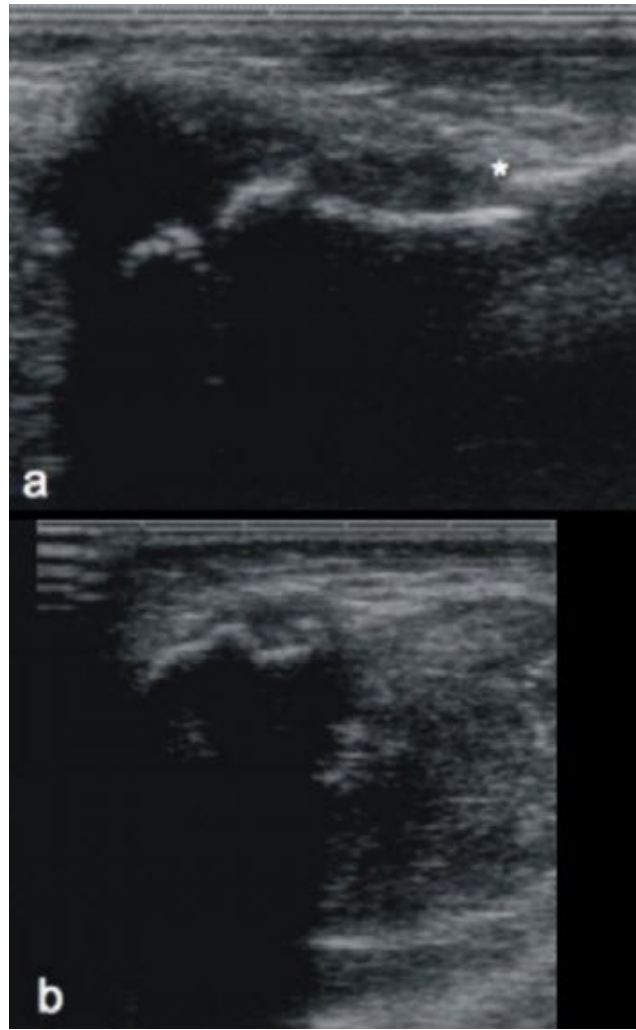


Figure 2 (top): 14 months old boy with unilateral tibia vara caused by focal fibrocartilaginous dysplasia. Ultrasound (a-longitudinal; b-transversal cut; linear transducer 7,5 MHz) depicted a cortical interruption with a slight step (*). There was no sign of hyperperfusion in color duplex sonography (not shown). The findings in the surrounding soft tissue were unobtrusive.

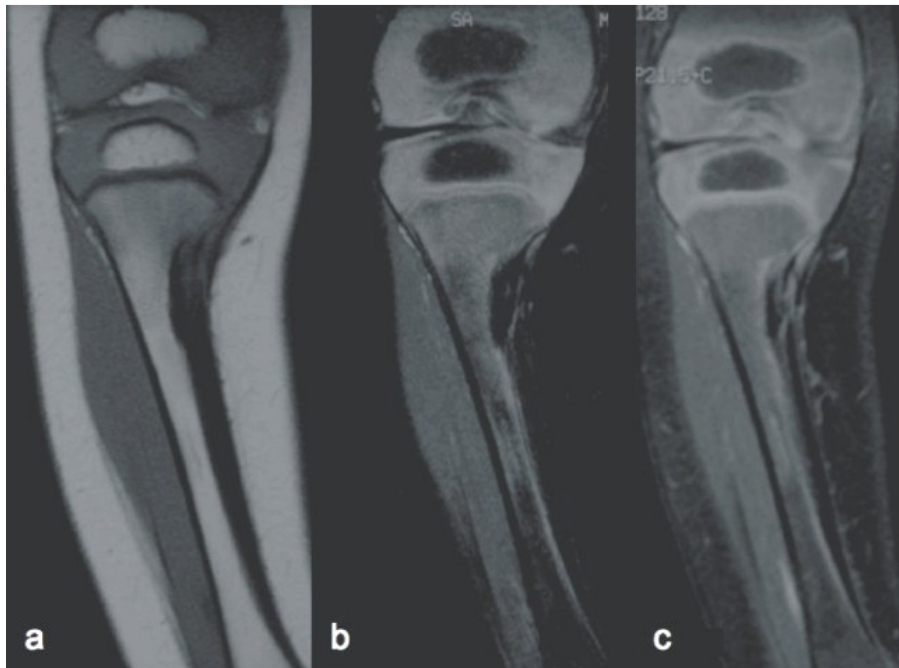


Figure 3 (left): 14 months old boy with unilateral tibia vara caused by focal fibrocartilaginous dysplasia. Coronal T1 weighted images (a) (TE 9, TR 500), T2 weighted images with fat suppression (b) (TE 42,2, TR 2660) and T1 weighted images after contrast injection with fat suppression (c) (TE 20, TR 500). MRI revealed a lesion hypointense on T1 as well as on T2 weighted images without uptake of contrast media after injection of Gadolinium.

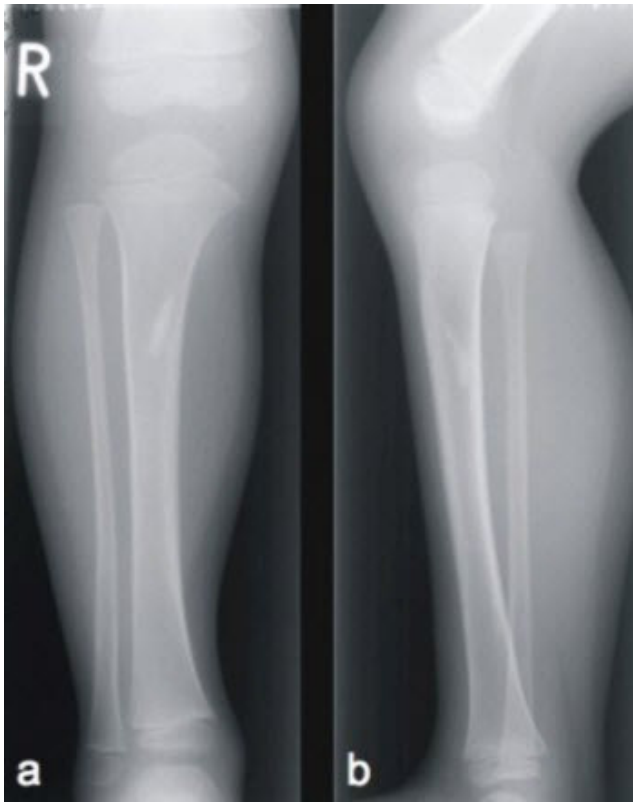


Figure 4: 5 year old boy with unilateral tibia vara caused by focal fibrocartilagenous dysplasia. Radiographs taken four years after initial diagnosis (at 14 months of age) showed nearly complete resolution of the varus deformity with only a residual subcortical area of sclerosis. The lucent defect in the medial cortex has dissolved.

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ABBREVIATIONS

FFCD = focal fibrocartilagenous dysplasia
MRI = Magnetic Resonance Imaging

KEYWORDS

Focal fibrocartilagenous dysplasia, FFCD, tibia vara