Abnormal alignment of the left lower extremity and irregular gait

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2-year-old male with abnormal alignment of the left lower extremity and irregular gait.

Focal fibrocartilaginous dysplasia

Focal fibrocartilaginous dysplasia (FFCD) is a rare benign entity first described by Bell in 1985 [1]. The most common site of FFCD is the proximal tibia followed by distal femur, forearm, humerus, phalanx, and vertebra transverse process [2, 3]. FFCD is most commonly noticed when children start crawling and ambulating [4]. Usual presentation is unilateral tibia vara, limb shortening, and tibial torsion [4].

Etiology of FFCD is not completely known. It is believed to be a failure of differentiation of a mesenchymal anlage, resulting in excessive fibrocartilage formation and interfering with growth of the medial tibial metadiaphysis and natural slipping of the periosteum for growth [1, 3]. In proximal tibia, this lesion typically occurs in the region of pes anserine tendons insertion [5]. Prior insult to the medial tibia has been postulated as an etiology [4, 5].

The characteristic radiographic findings are tibia vara associated with an elliptical lucency within the medial tibial metadiaphyseal cortex with surrounding sclerotic bone and absence of cortical margin at the superomedial aspect of the lesion [6]. The cortical thickening is more pronounced along the lateral margin of the lesion [7]. On CT, the corresponding lucency contains the insertion of the pes anserine tendons.

MRI demonstrates characteristic low-signal T1 and T2 intraosseous soft tissue component corresponding to the cortical lucency, surrounded by hypointense sclerotic bone [2]. The signal intensity of the soft tissue component is hypointense to the hyaline cartilage, similar to fibrocartilage. Post-contrast images as in our case typically do not display significant enhancement [7]. FFCD is typically a radiographic diagnosis and CT and MRI should be reserved for problem solving and/or operative planning. Recognition of this entity on radiographs could have prevented our case from undergoing further imaging.

Correct diagnosis of FFCD is crucial to avoid unnecessary invasive procedures. Differential diagnosis for tibia vara in children includes infantile Blount's disease, rickets, and asymmetric closure of the epiphysis secondary to infection or trauma [3]. Blount's disease typically involves the epiphysis and metaphysis and can present bilaterally as opposed to FFCD, which is typically unilateral without direct involvement of the growth plate [4]. Rickets usually involves more than one bone and is accompanied by classic metaphyseal flaring and growth plate widening and can present at the age of 2 in severe cases [8]. Osteoid osteoma in the medial tibia can potentially cause tibia vara. Osteoid osteoma has a lucent nidus, which can be centrally mineralized, and is usually surrounded by reactive sclerotic bone and is seen in a wide age range [9]. On certain imaging slices, FFCD can mimic the appearance of nonmineralized osteoid osteoma nidus, while superomedial cortical bone defect in FFCD can differentiate the diagnosis.

Due to the classic imaging findings, biopsy is generally not recommended. For cases that have undergone biopsy, pathology demonstrated dense hypocellular fibrous tissue interspersed with foci resembling fibrocartilage [1]. The presence of fibrocartilage is not essential for diagnosis [6].

Approximately 45% of the cases of tibial FFCD have spontaneously resolved over the span of approximately 57 months, which may be an underestimation of cases that could potentially spontaneously regress if intervention had not been performed [6]. Conservative management has been recommended if the Levin–Drennan angle, measured by the intersection

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of a line through the transverse plane of the proximal tibial metaphysis with a line perpendicular to the long axis of the tibial diaphysis, is $< 20\text{--}30^\circ$ [5]. Attention to the Levine–Drennan angle can allow for early diagnosis of a bowing deformity as well as assess progression. If conservative treatment fails, surgical options include osteotomy, curettage, and guided growth with plating, with the intent to correct the tibia vara deformity and allow a more rapid recovery of conventional gait [10]. In our case, the Levine–Drennan angle initially measured 21° . Follow-up radiographs at 5 months demonstrated the angle to be 26° and at this time, curettage was offered. The parents elected to continue surveillance and a 3-month radiograph demonstrated stability. Continued surveillance with 5-month interval follow-up radiograph will be performed.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflicts of interest.

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