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Case Report

Tibia vara caused by focal fibrocartilaginous dysplasia: a case report

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Abstract: Focal fibrocartilaginous dysplasia is a relatively rare, benign bone lesion that usually leads to varus deformities of the proximal tibia. We report upon such a case which was surgically excised by piezosurgery. The final examination 18 months later showed normal features both clinically and radiographically. To the best of our knowledge, this is the first case cured with piezosurgery and the first case to report sequestrum by biopsy.

Keywords: Focal fibrocartilaginous dysplasia, tibia vara, piezosurgery

Introduction

Focal fibrocartilaginous dysplasia (FFCD) is a relatively rare, benign bone lesion that usually occurs in children. Cases involving the tibia [1-3], femur [4, 5], ulna [6, 7], humerus [7], radius [8], phalanx [4], and vertebra [9] have been reported. Among these reports, the proximal tibia is believed to be the most frequently involved site. FFCD in the tibia usually leads to varus deformities. It is not completely known what causes this disease and how the deformity occurs [1, 5]. Spontaneous resolution can occur in some patients, while persistent or progressive deformities exist in others. Surgical or conservative treatment is still controversial.

We report a case affecting the proximal tibia with varus deformity, which was surgically excised by piezosurgery. Histopathology demonstrated that the lesion was composed of fibromuscular tissue, cartilage and sequestrum. To the best of our knowledge, this is the first case cured with piezosurgery and the first case to report sequestrum by biopsy.

Case report

A 26-month-old boy with varus deformity of the right leg was evaluated in our department. When he was 14 months old, his parents noticed that his right leg was bowed. The deformity gradually worsened, and he walked with a limp at the age of 20 months. The patient had no history of trauma, infection, or metabolic disease. He did not complain of any pain or other discomfort. Physical examination showed that the affected leg was 2 cm shorter than the normal side. Radiographs demonstrated lucent defects in the medial cortex of the proximal tibia, with sclerosis along the borders of the lesion. His radiological evaluation also revealed a varus deformity with a mechanical medial proximal tibial angle (mMPTA) of 73° (**Figure 1A**). A full-length X-ray was not obtained due to the refusal of his parents. Magnetic resonance imaging (MRI) showed a low-signal area in the medial part of the proximal tibia on both T1- and T2-weighted sequences (**Figure 1B**). The procedure was applied through a medial longitudinal incision along the proximal tibia. The lesion of the tibia was excised by piezosurgery (XD860A, SMTP, Jangsu, China, frequency of 45 Hz with a maximum amplitude of 0.15 mm). Attention was given not to damage the normal cartilage from the epiphysis and the growth plate. Neither guided growth nor an osteotomy procedure was performed to correct the tibial varus deformity. Histopathology demonstrated that the lesion was composed of fibromuscular tissue, cartilage and sequestrum (**Figure 2**). The final examination at 3 years and 8 months

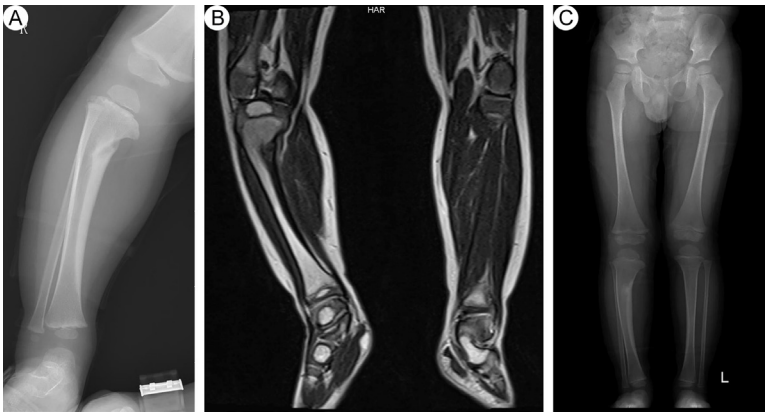

Figure 1. A. Preoperative X-ray showed a lucent defect in the medial cortex and varus deformity of the proximal tibia. B. The T2-weighted MRI sequences showed a low-signal area in the medial part of the proximal tibia. C. The 18-month follow-up X-ray showed good limb alignment.

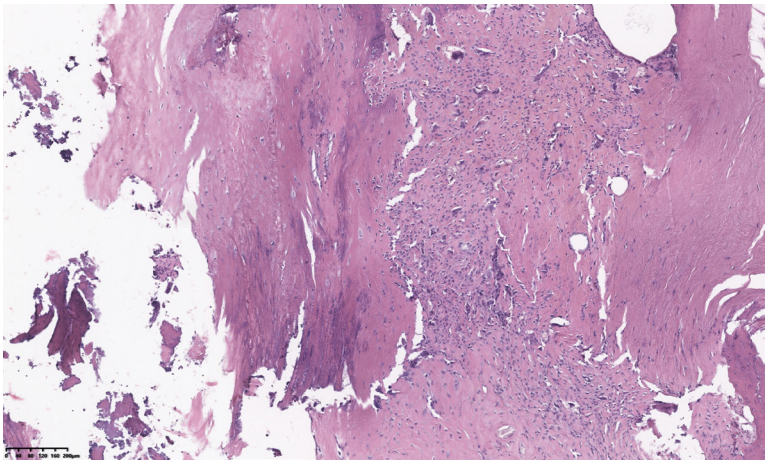

Figure 2. Histopathology demonstrated that the lesion was composed of fibromuscular tissue, cartilage and sequestrum (H&E, 100X).

of age showed normal features both clinically and radiographically (**Figure 1C**).

Discussion

The etiology of FFCD remains unknown. The common pathologic feature is a thick fibrotic band extending from the epiphysis to the metaphysis on one side of the bone. It is supposed that this band behaves as a tether, causing asymmetric growth and angulation. Bell et al. suggested that the mesenchymal anlage of the tibial metaphysis has developed abnormally, which leads to excessive production of fibrocartilage [10].

Overall, 24 reports about FFCD in the tibia with full text are found in English literature, and 71 cases of FFCD in tibia were recorded. There is a small prevalence of males, with a male/female ratio of 1.22 (39/32) and a left/right ratio of 1.15 (38/33). FFCD mainly affects children at the beginning of their walking age, but it has also occurred in an adult patient aged 29 years, with Turner's syndrome [11]. The onset of the deformity occurs at the age of 0 to 36 months in children, while the age at presentation is 2 to 54 months.

Clinical manifestations included unilateral bowing of the leg, tibial torsion, limb length discrepancy and limp. Swelling of the leg [12, 13], a limited range of motion [12], stumbling, and discrete hyperpigmentation [4] have also been reported. Pain, tenderness, and joint contractures are absent. Postovsky reported a case of FFCD of the tibia without clinical abnormalities concomitant with eosinophilic granuloma of the lower jaw [13].

Radiography typically shows a varus deformity and cortical defect with a surrounding sclerosis area of the medial proximal tibia [1], while Mooney reported a 2-year-old female with varus deformity of the distal tibia [14]. In addition, valgus deformity and a lucency of the lateral proximal tibia were also detected [5, 14, 15]. Nakase described a prominent periosteal reaction at the lesion site [12]. One of the five cases reported by Dusabe presented with an uncommon radiographic appearance with physeal impairment [16]. Magnetic resonance imaging (MRI) shows a low-signal area on both T1- and T2-weighted sequences [4, 5]. In the case reported by Ringe, ultrasonography showed a hypoechoic lesion upon the cortex without hyperperfusion in color duplex sonography, and there was a cortex interruption as seen in a gaping fracture [4]. One of the two cases reported by Zayer

underwent scintigraphy, which showed a high uptake of the lesion [1]. A complete body scan with ^{99m}technetium showed a mild reactive lesion in three patients [17].

To clarify or exclude the diagnosis, a biopsy procedure was performed in some cases. The histopathological appearance varies. The first descriptions were published by Bell et al. In two of their cases, the biopsy showed dense hypocellular tissue resembling fibrocartilage in some areas and tendon in others [10]. However, in some later cases, cartilage was absent. In our case, biopsy showed sequestrum in addition to fibromuscular tissue and cartilage. Perhaps different cases were undergoing different phases of the same disease. Biopsy may not always be necessary. The diagnosis can be made when one has typical clinical and radiological features without any other possible causes [3, 10].

In some patients, the deformity resolves after treatment with splints, braces, or shoe elevation [2-4, 17, 18], but spontaneous resolution also occurs after observation alone [3, 10, 19-22].

Most of the surgically treated patients, including fibrous band release, curettage, guided growth, osteotomy, or a combination of different surgical methods, healed clinically and radiologically at the last follow-up [3, 10, 11, 14, 17-20, 22]. However, incomplete correction and overcorrection also occur in some patients after osteotomy or guided growth [20, 22]. Due to the differences in tissue density and elastic properties, ultrasonic osteotomes could play a selective cutting role in hard structure bony tissue rather than in soft tissue [23]. For the fewest adverse effects, we chose the piezosurgery.

In conclusion, the diagnosis of FFCD can be made based on the typical clinical and radiological appearance. When differential diagnosis is difficult, biopsy can be taken into consideration. Lesion resection alone is enough for FFCD in the tibia with secondary deformity. Once surgical resection is decided upon, piezosurgery may be a good choice.

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Disclosure of conflict of interest

None.

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