

Desmoplastic fibroma of the femur with atypical image findings

A case report

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Abstract

Rationale: Desmoplastic fibroma of the bone (DFB) is an extremely rare benign bone tumor, which can occur in any bone at all ages. Herein, we report a case of non-specific imaging findings.

Patient concerns: A 25-year-old female patient was consulted in the Armed Police General Hospital (Beijing, China) due to repeated pain in the right thigh lasting over 1 year. Imaging examination revealed a space-occupying lesion in the right femur.

Diagnosis: Desmoplastic fibroma of the femur

Interventions: Wide surgical resection

Outcomes: At the 1-year follow-up visit, no relapse in the tumor was observed.

Lessons: In this case report, we described and emphasized the particularity of the case examined and the possible non-specificity of the imaging results of some DFB cases.

Abbreviations: DFB = desmoplastic fibroma of bone, LOM = limitation of motion, MRI = magnetic resonance imaging.

Keywords: case report, desmoplastic fibroma, femur

1. Introduction

Desmoplastic fibroma of bone (DFB) is an extremely rare benign bone tumor, which accounts for approximately 0.06% of the primary bone tumors and 0.3% of benign bone tumors worldwide.^[1] DFB can occur in any bone, but is predominant in the jaw and the femur bones.^[2] Typically, DFB is more common in the distal femoral metaphysis when it occurs in the femur.^[3] Moreover, although DFB can occur at any age, the morbidity in young people is much higher than that in the elders. Evidence has revealed that DFB is lytic, occasionally with poorly defined borders on X-ray. On CT images, it can be detected as mildly hyperdense structures with thin sclerotic margins, with or without destruction of the contiguous cortex^[4]; low-signal

intensity is observed on T2-weighted MRI scans. However, some DFB imaging findings are different. Due to the rarity of DFB, it has been difficult to conduct relevant trials, leading to a lack of clinical data. Therefore, here, we report a case of DFB with atypical image findings.

2. Case report

A 25-year-old female patient was consulted in the Armed Police General Hospital (Haidian District, Beijing, China) due to repeated pain in the right thigh for over 1 year that had aggravated for 1 week. The patient reported to have had right thigh pain and swelling 1 year ago, accompanied by limitation of motion (LOM), which was relieved after rest. In the following year and thereafter, the patient felt pain and swelling in the right thigh when catching a cold, which was mitigated by rest and was thus long neglected. Approximately 1 week ago, the patient had this right thigh pain again, but this time it apparently aggravated more than ever before.

2.1. Physical examination

No deformity or hypoesthesia was found in the right thigh and right knee joint. Flushed and swollen skin was noted in the lower segment of the right thigh, whose skin temperature was higher than that of the left one. Meanwhile, distal right femoral tenderness (+) and right knee joint LOM were detected.

2.2. Imaging data

X-ray showed an uneven density in the distal left femur, in which lamellate high-density opacity and bone destruction were observed (Fig. 1). Furthermore, the CT findings indicated proximal femoral thickening with irregular, low-density opacity, and a long-axis parallel that was to the femur. The well-defined

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Patients have signed an informed consent form, understand thoroughfare her cases.

All authors have read the final version of the manuscript and confirm its accuracy as well as that the data have not been copied from or published elsewhere.

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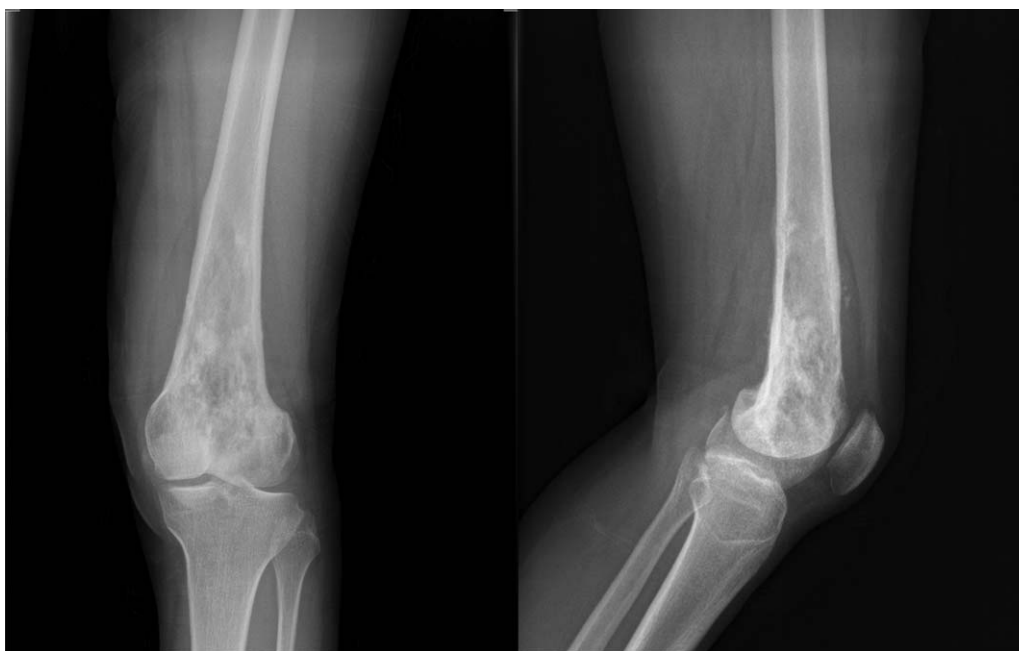


Figure 1. X-ray revealed uneven density in the distal left femur, in which lamellate high-density opacity and bone destruction were observed.

lesion had an even soft tissue density as well as the hardening strap (Fig. 2).

MRI findings revealed massive and lamellate uneven long T1 and short T2 signals in and around the distal left femur, which were ill-defined with the surrounding muscular part, and local femoral cortex signal was non-continuous. A lesion with an approximate size of 6.5 cm × 13.7 cm was identified that had been compressing the quadriceps femoris; a continuous signal in the tendon was obtained (Fig. 3).

2.3. Surgical data

A wide surgical resection of the tumor was performed. The solid mass was found to originate from the distal femur. On gross examination, the encapsulated tumor measured 7 × 17 × 5.5 cm. Next, the tumor was excised with the involved knee joint and femur periosteum, distal up to approximately 9 cm from the knee joint. Reconstruction of the bony defect was implemented using an allogeneic bone graft. Further, a biopsy was performed following the tumor resection, which resulted in a diagnosis of DFB (distal left femur). The immunohistochemical results revealed Actin (+), BCL2 (focal +), CD68 (-), CK (-), Desmin (-), Ki-67 (+5%), S-100 (-), and Vimentin (+) (Fig. 4).

At the 1-year follow-up visit, after the patient had returned to work, the MRI revealed no signs of tumor recurrence. The knee function of the patient was basically normal. However, in this case, due to the short follow-up, the prognosis was uncertain, and the patient was advised to return every 2 years for a further follow-up (Fig. 5).

3. Discussion

3.1. Features of DFB

First reported by Jaffe in 1958, DFB has the global annual morbidity of 2–4/1,000,000^[5] and an etiology that remains obscure and needs to be further studied. From a broad point of view, genetic predisposition, estrogen stimulation, and trauma

stimulation, such as fracture and surgery, are partly associated with the genesis of DFB.^[6] X-ray, CT, and magnetic resonance imaging (MRI) are generally used to determine the tumor range and severity, while pathological examination remains the gold standard in its diagnosis.^[4]

3.1.1. Clinical features. DFB is generally characterized by a high recurrence rate and local invasion. Nevertheless, it does not manifest typical clinical symptoms, and pain; dysfunction or pathological fracture is the most common cause of hospital visit. Its clinical manifestations are usually pain and swelling in the pathological region, as well as symptoms induced by mass compression of the surrounding tissue. Hence, the clinical symptoms in patients are usually closely related to the tumor growth site and the compression of the surrounding tissue.^[7–9]

3.1.2. Imaging findings. DFB has diversified imaging findings but is mainly manifested as oppressive or expansile bone absorption and destruction. Zlotecki^[10] divided DF into a marginal and central type. The marginal type is most frequently manifested as local bone cortex shifting, compression, and absorption, with an adjacent soft-tissue mass.^[11] The central type frequently manifests as local osteolytic swelling, with or without periosteal reaction, but more patients have no periosteal reaction. Meanwhile, osteosclerosis and soft-tissue mass can also be observed in some cases.^[12] The imaging findings in our patient were non-specific, which confirms the diverse imaging results in DFB detection.

3.1.3. Pathological features. 1. The tumor size (2–6 cm), established by visual inspection, was considerably different. The tumor had an off-white color and a tough texture; it was ill-defined and frequently invasive into the surrounding tissue;

2. The tumor was composed of well-differentiated slender spindle fibroblasts, myofibroblasts, and collagen fibers, which was determined by microscopic examination. These 3 components were unevenly distributed in different tumors or sites of the same tumor. Immunophenotypic analysis showed that DFB

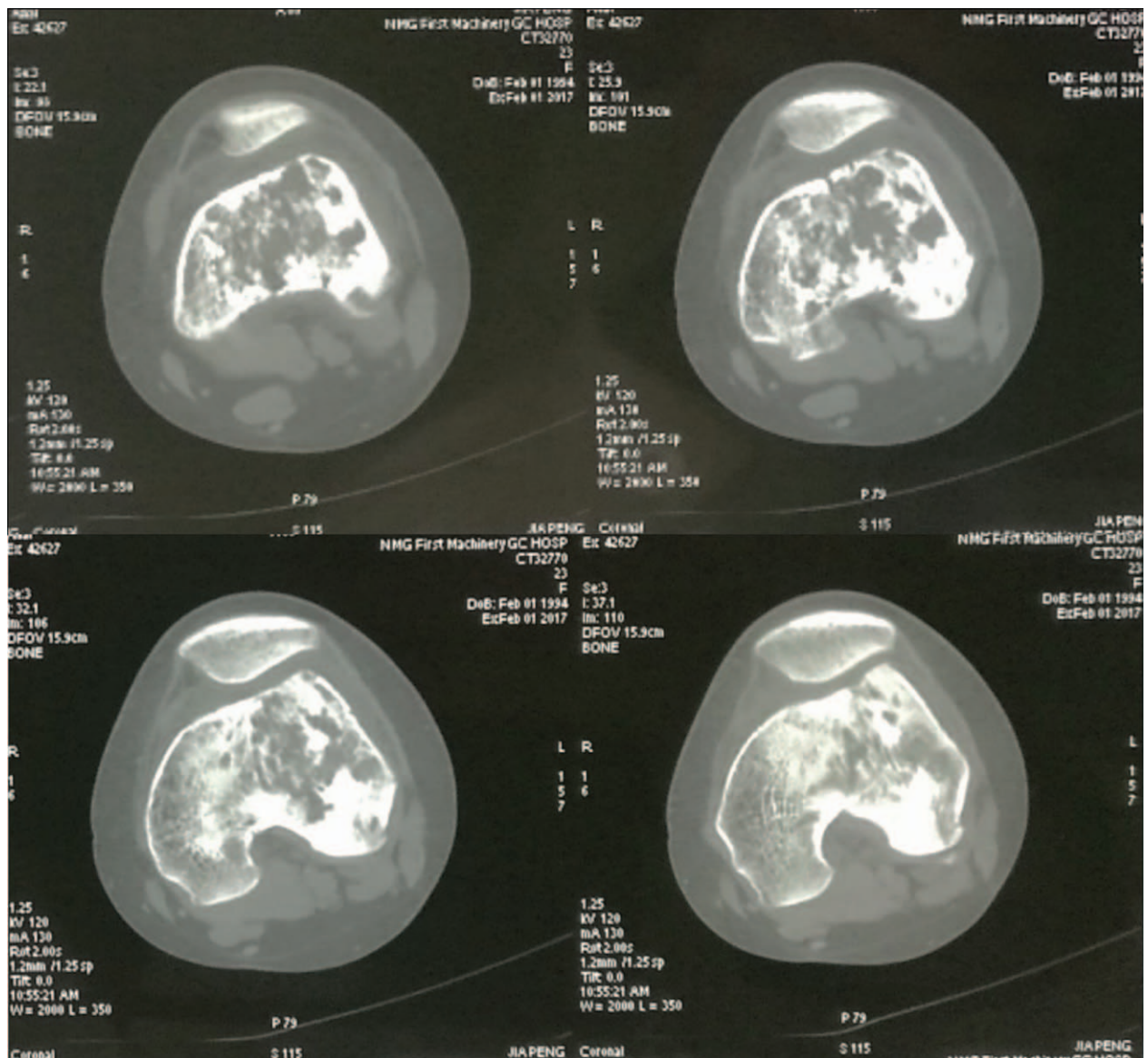


Figure 2. CT showed proximal femoral thickening, in which irregular low-density opacity was observed. Its long-axis was parallel to the femur. The well-defined lesion displayed the even soft tissue density as well as the hardening strap. No obvious periosteal reaction was detected, and a clear surrounding soft tissue space was observed, with no swelling.

shared features identical to those of aggressive fibromatosis of the soft tissue, including positive β -catenin (nuclear-positive) and vimentin expression, whereas the expression of Desmin, S-100, CD34, and MDM2 was negative.^[13,14]

3.2. Treatment and prognosis for DFB

Extensive local surgical resection is recommended in most cases due to the locally invasive behavior of this tumor.^[15-17] Reportedly, the recurrence rate after resection is 17% but can reach as high as 55% after abrasion.^[18] Nevertheless, the extensive surgical resection and high recurrence rate may affect the dysfunction in the affected region. Therefore, multiple factors should be taken into consideration when selecting the resection region. This is particularly important in repeatedly recurrent cases, in which the resection surgery may affect the limb function. However, extensive resection in such patients is still required,

followed by corresponding functional reconstruction, to guarantee their infinite recovery to working and daily life of the patients. Based on the results in previously published reports, the authors suggest that no definite preoperative diagnostic criteria are currently available for DFB.^[19-28] Nonetheless, DFB is associated with low morbidity, and a certain misdiagnosis rate might be expected among pathologists. As a result, clinicians should always be extremely careful in cases of first-onset patients. It is therefore crucial that such patients are to be reminded to receive regular re-examination apart from pathological biopsy for the successful and timely distinction from a malignant bone tumor.

4. Conclusions

In this case report, we described and emphasized the particularity of the case examined and the possible non-specificity of the imaging results of some DFB cases. Therefore, the imaging

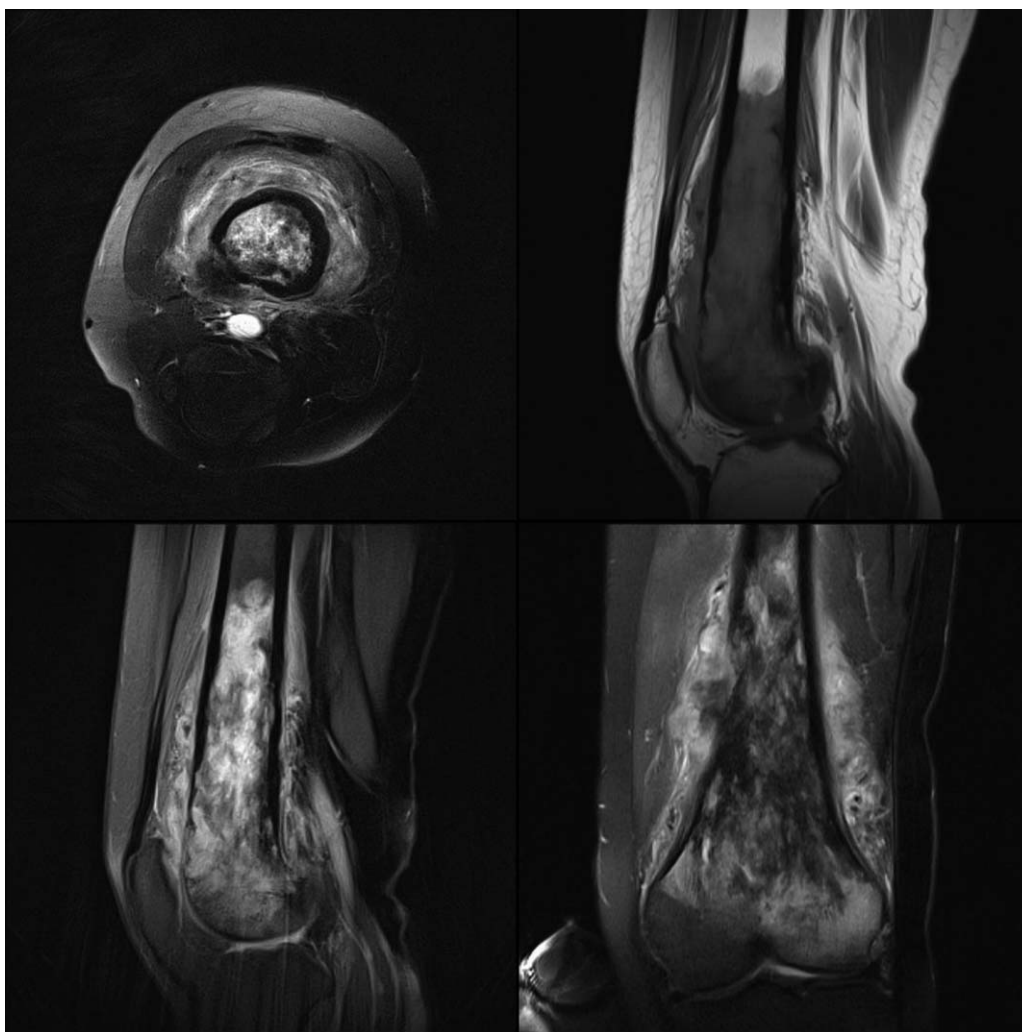


Figure 3. MRI revealed massive and lamellate uneven long T1 and short T2 signals in and around the distal left femur, which were ill-defined with the surrounding muscular part; the local femoral cortex signal was non-continuous. A lesion with an approximate size of 6.5 cm × 13.7 cm was detected, which had compressed the quadriceps femoris; a continuous tendon signal was detected.

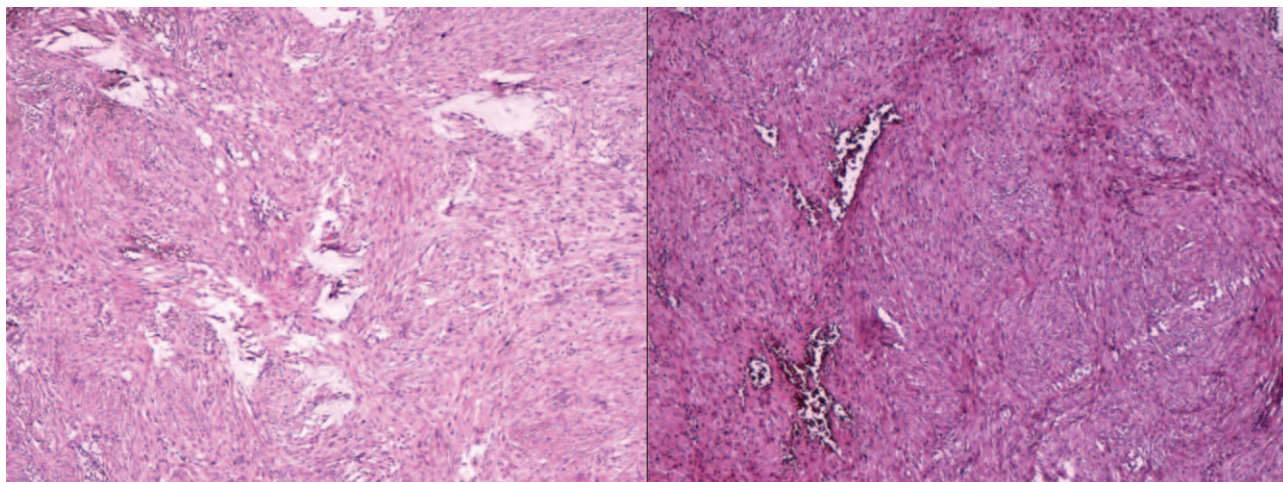


Figure 4. Pathology: (distal left femur) DFB. Immunohistochemical results suggested Actin (+), BCL2 (focal +), CD68 (-), CK (-), Desmin (-), Ki-67 (+5%), S-100 (-), and Vimentin (+).



Figure 5. Post-tumor resection image.

findings of this disease cannot be used as a basis for diagnosis. Clinicians should treat it with caution.

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Author contributions

Writing – original draft: Yongming Xu.

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