Desmoplastic Fibroma of Distal Femur: A Case Report

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Desmoplastic fibroma of bone is an extremely rare tumor that was first described by Jaffe in 1958. It histologically resembles the desmoid tumor of soft tissue. It is known as locally aggressive tumor but we experienced definitely benign and resembling simple bone cyst radiographically. We report a case of desmoplastic fibroma of bone and it should be included in the differential diagnosis list of any lytic bone lesion. The radiograph, MR imaging features, radiological and pathological differential diagnosis of the case are described, and literatures are reviewed.

Index words : Desmoplastic fibroma
Distal femur
Female

Introduction

Desmoplastic fibroma(DF) is an extremely rare tumor consisting of thin, wavy fibroblasts set in an abundant matrix of collagen fibers. It histologically resembles the desmoid tumor of soft tissue. This benign tumor is characterized by aggressive local infiltration. Radiographically, the tumors are lucent and expansile lesions. The lesion is difficult to distinguish from other lesions such as a unicameral bone cyst, fibrous dysplasia, chondromyxoid fibroma, non-ossifying fibroma, giant-cell tumor of bone, and fibrosarcoma of bone. We experienced desmoplastic fibroma radiographically resembling simple bone cyst in a 30-year-old female, at the left distal femur. Because of its rarity, we report this case of desmoplastic fibroma.

Case Report

A 30-year-old woman was admitted to our hospital because of left knee discomfort for a month. There was no history of pain. Upon a physical examination, she had no tenderness or limitation of motion. Laboratory test were all within normal limits. The roentgenogram of left knee (Fig. 1A, B) showed well-defined osteolytic lesion with surrounding sclerotic rim at left distal femoral metaphysis. For further evaluation of osteolytic

JKSMRM 13:199-202(2009)
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Received; July 6, 2009, revised; July 16, 2009, accepted; October 14, 2009
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lesion, MR imaging (Intera 1.5T, Philips, Netherland) was performed. T1-weighted sagittal turbo spin echo(TSE) image (Fig. 1C) showed well-defined, lobulated, and homogenous low signal mass at lateral aspect of distal femoral metaphysis with thin markedly low signal border. T2-weighted TSE sagittal, coronal and axial images (Fig. 1D, E, F) showed homogenous high signal intensity with thin low signal border. No cortical disruption or soft tissue extension was visualized. Considering simple X-ray and MR finding, simple bone cyst and chondromyxoid fibroma were suggested radiographically. Because of her intermittent discomfort, curettage and bone-graft procedure was done. The gross specimen photograph (Fig. 1G) showed tear-drop shaped homogenous yellowish content of the mass. The photomicrograph of specimen (Fig. 1H, hematoxylin and eosin ×200.) showed fibroblastic spindle cell proliferation in dense collagenous tissue, consistent with desmoplastic fibroma.

Discussion

Desmoplastic fibroma is a rare bone tumor, accounting for only 0.06% of all bone tumors and 0.3% of the

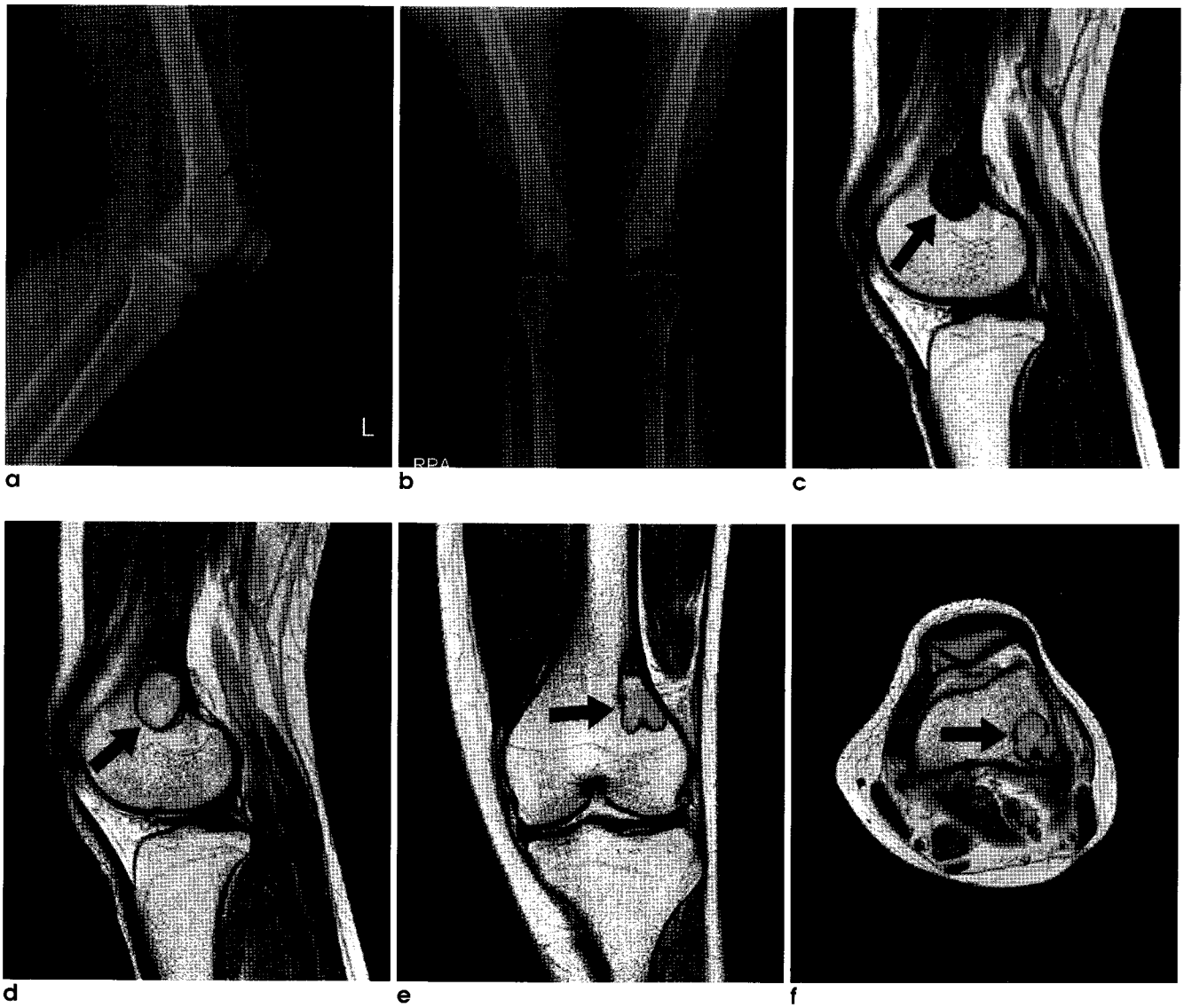

Fig. 1. a, b. The roentgenogram of left knee showed well-defined osteolytic lesion with surrounding sclerotic rim at left distal femoral metaphysis. **c-f.** MR image of left knee. T1-weighted sagittal turbo spin echo (TSE) image (**c**) showed well-defined, lobulated, and homogenous low signal mass at lateral aspect of distal femoral metaphysis with thin markedly low signal border. T2-weighted spin echo sagittal, coronal and axial images (**d-f**) showed homogenous high signal intensity with thin low signal border.

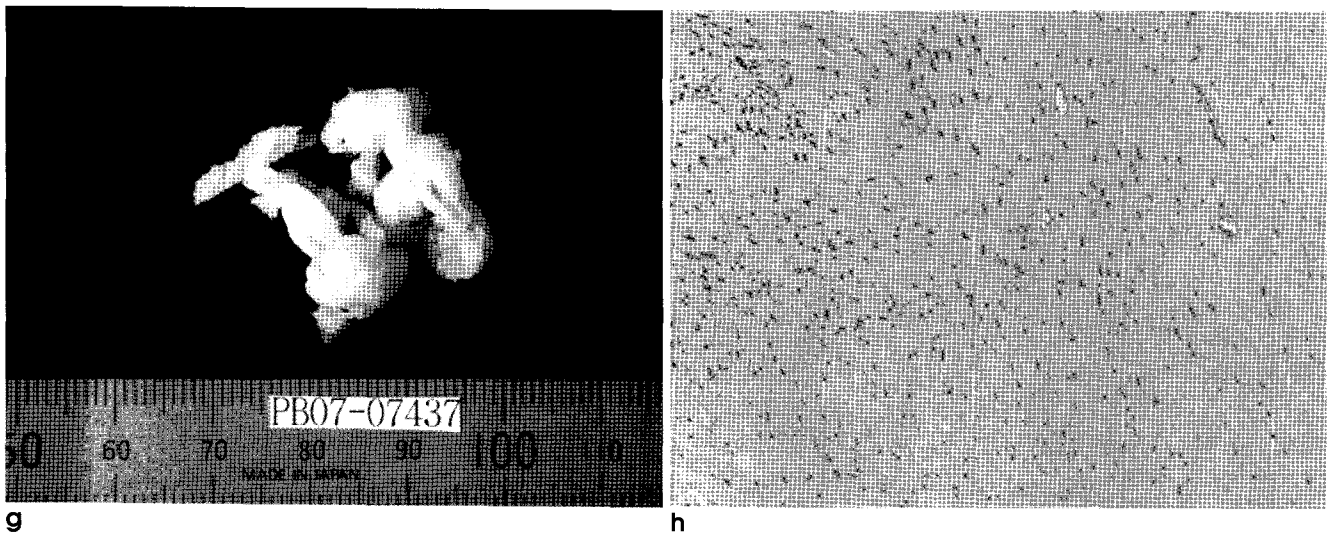

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Fig. 1. g. The gross specimen photograph showed tear-drop shaped homogenous yellowish content of the mass. **h.** Photomicrograph of specimen (e, hematoxylin and eosin ×200) showed fibroblastic spindle cell proliferation in dense collagenous tissue, consistent with desmoplastic fibroma.

benign bone tumors in the Mayo Clinic series (1). It was first described as a distinct entity and given the name desmoplastic fibroma by Jaffe in 1958. He described it as a fibrous tumor that was distinct from other intraosseous fibrous tumors such as non-ossifying fibroma, ossifying fibroma and chondro-myxoid fibroma.

Although almost any bone can be affected, the long bones have accounted for about 50% of the lesions. Of these, occurrence in the femur and tibia predominated. The mandible was the third and the pelvis, the fourth most common site (2).

The symptoms of desmoplastic fibroma were nonspecific and similar to those of other tumors. Some patients had no symptoms and the tumor was an incidental finding after plain radiographs had been obtained after a trauma (3).

Radiographically the lesion is lytic and often has a honeycombed appearance. The tumor increases the diameter of the bone by endosteal resorption and periosteal reaction and is usually contained by a rim of fibrous or osseous periosteum (4).

Radiological appearance of desmoplastic fibroma is nonspecific. Diagnosis is only established by microscopical examination. It is characterised by uniform appearing fibroblastic cells in a stroma containing various amounts of thick, wavy, interlacing collagen fibers (5). Desmoplastic fibroma of bone and medullary fibrosarcoma have similar pathological appearance with spindle-shaped fibroblasts and collagenous matrix. But in well-differentiated fibrosarcoma, fibroblasts are long and spindle-shaped with plump and elongated hyperchromatic nuclei that are almost twice as large as the ones seen in desmoplastic fibroma. Absence of mitotic figures in desmoplastic fibroma of bone is a helpful sign in differential diagnosis also. However, presence of mitotic figures is not a faithful sign for differential diagnosis as mitosis is not a prominent feature in well-differentiated fibrosarcoma of bone (6). Low grade intraosseous osteosarcoma, another tumor that can mimic desmoplastic fibroma, can also be excluded by identification of bone formation. Presence of multinucleated giant cells, foam cells and hemosiderin-laden macrophages in fibrous cortical defects and non-ossifying fibromas distinguishes these lesions from desmoplastic fibroma of bone (7).

CT and MR imaging are useful techniques to delineate the lesion and its relationship to the adjacent structures CT was found to be superior to MR in showing the cortical breakthrough, but MR imaging is better suited than CT to show the extraosseous tumor extent. Although MR studies of the lesion are scarce, it usually has low to intermediate signal on T1-weighted images that has considerable overlap with MR features of other bone tumors. But presence of low to intermediate signal intensity foci on T2-weighted images, which does not correspond to calcifications may help to narrow differential diagnosis (8). The

reason of low to intermediate signal on MR is due to fibroblast and collagen mixture. Transverse T2-weighted images displayed the extraosseous tumor component more clearly than the plain T1-weighted scan, even no extraosseous tumor extent in our case. Clear separation of intraosseous tumor from normal bone marrow along with coronal and/or sagittal images of long bones make MR imaging the most valuable imaging method in surgical planning.

The histologic differential diagnosis includes spindle cell tumors and tumor-like lesions of bone. Low grade fibrosarcoma is the most difficult differential diagnosis. The typical fibrosarcoma is more cellular with a herringbone pattern showing more polymorphism and a higher mitotic activity. However in some cases of low grade fibrosarcoma, mitoses are not manifested and areas with predominant collagen tissue may be found, which makes the distinction extremely difficult. In such cases, only after follow-up imaging makes the final diagnosis established (9). Desmoplastic fibroma of bone can imitate the morphologic pattern of low grade intraosseous osteosarcoma but desmoplastic fibroma lacks osteoid features (10).

In this case, there is no honeycombing change or extraosseous extension. So it makes difficult to differentiate with simple bone cyst or chondromyxoid fibroma.

As a conclusion, desmoplastic fibroma of bone should be included in the differential diagnosis list of any lytic bone lesion especially in young patients.

Because of its rarity, we report a case of desmoplastic fibroma.

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대한자기공명의과학회지 13:199-202(2009)

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결합조직형성 섬유종은 골에 발생하는 종양중 매우 드문종양으로 조직학적으로 연부조직에서 발생하는 유건종과 유사하다. 이 양성 종양은 국소적으로 주위 조직에 침윤성으로 성장한다. 방사선학적으로 고립성 골낭종, 섬유성 골이형성증, 거대세포종, 골육종과 감별이 쉽지 않다. 저자들은 방사선학적으로 골낭종과 비슷한 형태를 보이는 원위 대퇴골에 생긴 결합조직형성 섬유종을 경험하고 드문 골 종양임으로 참고문헌과 함께 보고하는 바이다.

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