Synovium as a widespread pathway to the adjacent joint in undifferentiated high-grade pleomorphic sarcoma of the tibia
A case report

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Abstract

Rationale: Undifferentiated high-grade pleomorphic sarcoma (UPS), originated from bone, is a rare tumor, accounting for 2% to 5% of all primary maligment bone neoplasms. Skip lesion can be found in undifferentiated high-grade pleomorphic sarcoma of bone (UPS-B). However, the direct invasion across the articular synovium to bone has not been reported previously.

Patient concerns: We report an unusual case of a 65-year-old man complained of a year history of pain, swelling, and limitation of activity in the left knee joint. At the proximal tibia, there was extensive invasion of articular synovium, which provides a direct anatomic pathway for the tumor invasion to the adjacent bone, including patella and femoral condyle.

Diagnoses: Magnetic resonance imaging was important in defining the marrow involvement and joint invasion, including the thickening articular synovium. Subsequent pathological examination confirmed the diagnosis of UPS.

Interventions: The patient underwent an extensive resection of the knee joint, except for the patellar.

Outcomes: After operation, routine chemotherapy was performed. Unfortunately, half a year later, soft tissue swelling of whole thigh was found. Then this patient came our hospital again. Positron emission tomography imaging showed there was recurrence of UPS with lung metastasis. A week later, this patient died.

Lessons: In contrast to frequent infiltration pathway, the articular synovium as a media for this tumor spread is rare. This study adds a better understanding of this direct invasion way to the medical literature.

Abbreviations: CT = computed tomography, MFH = malignant fibrous histiocytoma, MRI = magnetic resonance imaging, UPS = undifferentiated high-grade pleomorphic sarcoma.

Keywords: articular synovium, diagnosis, undifferentiated high-grade pleomorphic sarcoma

1. Introduction

Undifferentiated high-grade pleomorphic sarcoma (UPS), originated from the bone, is rare and mainly grows in the epiphysis and diaphysis of the long bones,[1] accounting for 2% to 5% of all primary maligment bone neoplasms.[2] UPS of bone (UPS-B) occurs commonly in 50 to 60 years old patients.[3] The incidence rate of UPS-B in men is higher than that in women.[4] The main clinical symptoms are local pain and swelling. Rarely, in patients, a pathological fracture is the primary symptom.[5] Some studies have shown that UPS-B arises secondarily in many pre-existing musculoskeletal disorders,[6] such as bone infarction,[7] chronic osteomyelitis,[8] osteitis deformans, as well as fibrous dysplasia,[9] enchondroma, and giant cell tumor of bone.[10] Radiation therapy[11] can also increase the risk of development of UPS. The knee joint was reported to be the most common site for UPS invasion.[8] Efe et al[2] have reported that it could arise from anterior cruciate ligament reconstruction. Infiltration pattern of this tumor can be different. Direct invasion and skip lesion can be found in UPS-B.[4]

The current paper reports a case of UPS at the proximal tibia with extensive invasion of synovial membrane, which provides a direct anatomic pathway for the tumor invasion to the adjacent bone, including patella and femoral condyle. Reviewing the literature, such spread of tumor has not reported.

Figure 1. X-ray radiographs of the right knee showing the high density signal in the proximal tibia: (A) anteroposterior; (B) lateral.

2. Case presentation

A 65-year-old man worked as a farmer complained of a year history of pain, swelling, and limitation of activity in the left knee joint after a micro-trauma for half a year. No fever appeared in the clinical manifestation of this patient. As the symptom could not be relieved after taking nonsteroidal anti-inflammatory drugs, the patient visited our department of radiology. Physical examination revealed local tenderness and effusion in the left knee joint, but no soft tissue mass could be palpated. The family history of UPS-B was negative.

Subsequently, the radiological imaging, including plain radiography, computed tomography (CT) scan, and magnetic resonance imaging (MRI) was obtained and reviewed. Radiographs showed increased bone density in the proximal tibia (Fig. 1) and localized destruction of lateral condyle of tibia plateau (Fig. 1A) was ambiguous. However, no special changes were seen in the distal femur. Periosteal reactions and expansive growth were not observed. On CT images, the lesions in the left tibial plateau showed uneven osteoblastic bone destruction within which patchy osteolytic lesions were seen with outer cortical destruction (Fig. 2). In the middle of the tibia, the marrow showed to be replaced by soft tissues (Fig. 2B). Slightly high density was showed in the margins of the femoral condyle and capitulum fibulae and the whole patella with the local bone trabecularism disappeared. Soft tissue mass was surrounding the knee joint.

Figure 2. The high-resolution computed tomography imaging showing bone destruction in the margin of lateral condyle of tibia plateau and condyles of femur with the swelling knee joint capsule. (A) and (B): In axial plane images, most of the tibia plateau and the whole patella are affected; (C): the tumor shows an extension to the middle of the tibia and fibulae in the coronally reconstructed image.
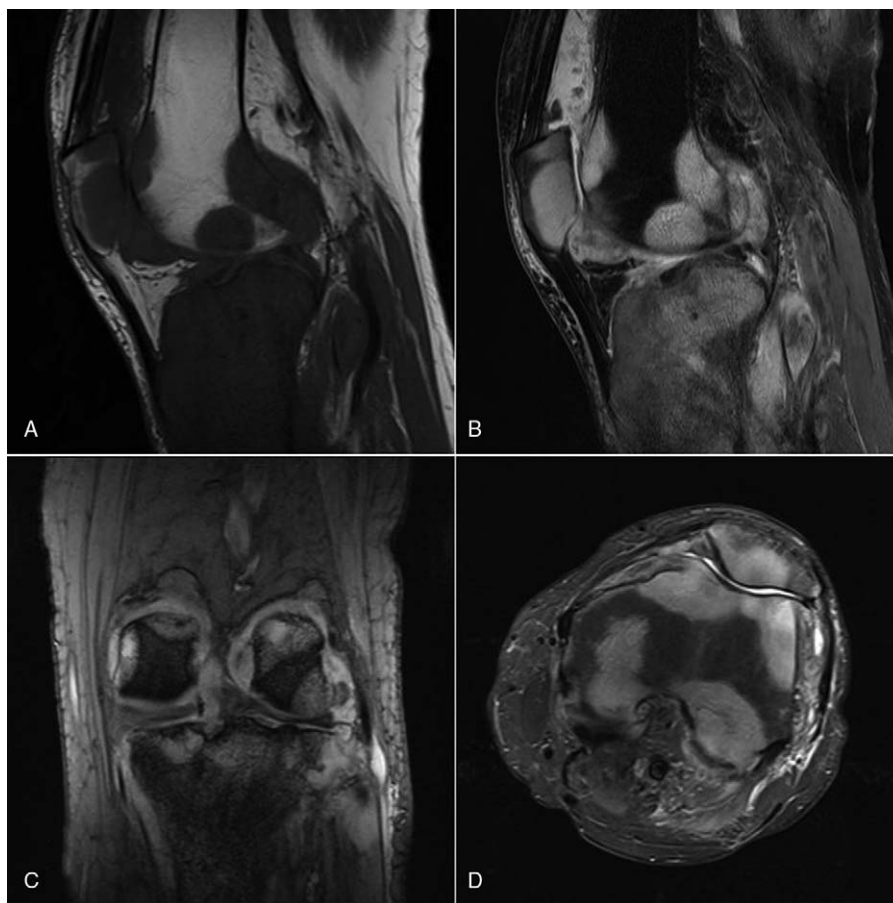
Figure 3. MR images showing the lesion involving the knee joint. (A): The lesion on T1 fast spin-echo-weighted saggital MR image (TR/TE: 512/11 ms) is homogeneous low signal intense to bone marrow; (B): on proton-density (PD) weighted fat saturation saggital MR image (TE/TR = 40/3200 ms), the lesion shows higher signal intense than surrounding muscles. The widespread thickening articular synovium can be observed; (C): T2-weighted multi-echo data image combination sequence demonstrates the local destruction of external condyle of tibia plateau without involvement of meniscus and the articular surface cartilage; (D): axial short T2 turbo inversion recovery (STIR) image (TR/TE: 4500/26, TI: 130) shows many small lesions in the patella and tibia plateau with local cortex erosion accompanied with soft tissue mass.

On MR imaging, besides the proximal tibia, the lesions involved the distal femur, capitulum fibulae, and the whole patella. Also, the articular synovium was affected (Fig. 3). The lesions showed predominantly low signal intensity to bone marrow on T1-weighted images (Fig. 3A) and slightly high signal intensity on T2 weighted images with fat suppression (Fig. 3B–D). On T2-weighted images, the lesions showed an inhomogeneous signal. The tumor exhibited a high degree of aggressiveness including infiltration of the surrounding tissues, especially invasion of the joint capsule. There was an uneven thickening of knee joint synovium firmly attaching to the adjacent osseous surfaces of the distal femur and proximal tibia. Infrapatellar fat pad and posterolateral joint capsule are more evident (Fig. 3B and C). There was also an increase in the joint capsule fluid. Effusion was increased in the joint capsule. The soft tissue mass shadow was surrounded the proximal tibia. Additionally, there was soft tissue mass adherence to the capsule wall of knee joint. In the periphery of the cruciate ligaments, the mass could also be detected. However, no erosions were detected in the meniscus and the articular surface cartilage (Fig. 3C). The result of imaging diagnosis of malignant bone tumor in the proximal tibia was provided, invading the surrounding tissues, including knee joint capsule, the distal femur, as well as capitulum fibulae and the whole patella.

During the operation, the cartilages of the bones in the knee joint were exfoliated. Synovial proliferation was extensive and cortex destruction was severe in the lateral condyle of tibia plateau accompany with soft tissue mass. Wide resection of knee joint tumor was performed, including the distal femur, proximal tibia, tumor-infiltrated soft tissue, and synovium. Subsequently, a tumor of domestic custom knee prosthesis was implanted (Fig. 4). During pathologic investigation, the gross specimens showed many soft, translucent, and grayish-white lesions with inhomogeneous distribution in the bone marrow of the distal femur and the proximal tibia. Meanwhile, localized destruction with cortex erosion at lateral condyle of tibia plateau was obvious. Synovial proliferation was extensive. Immunohistochemical staining revealed focal positive reaction for various molecules such as smooth muscle actin (SMA), CD 68, LYS, respectively. The histopathological examination findings of the biopsy specimen of knee bones and synovium confirmed this tumor as a UPS-B (Fig. 5).

After operation, routine chemotherapy was performed. Unfortunately, half a year later, soft tissue swelling of whole thigh was found. Then this patient came to our hospital again. Positron emission tomography imaging showed there was recurrence of UPS with lung metastasis. A week later, this patient died.
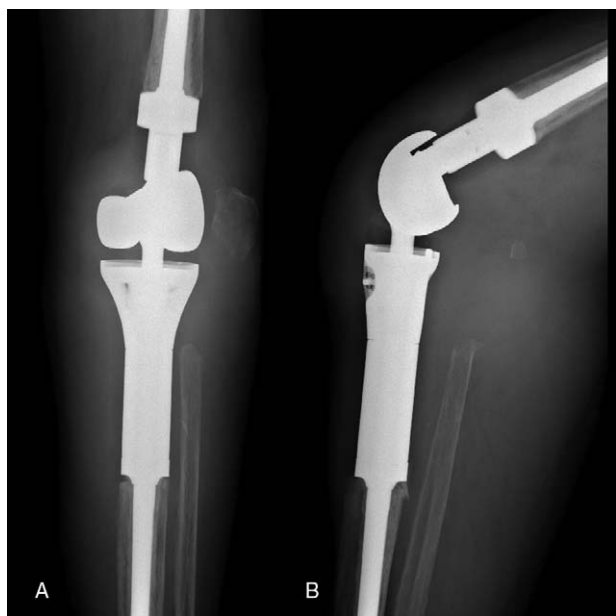
Figure 4. X-ray radiographs of the right knee showing the joint prosthesis in the extremity with the patella keeping by side: (A) anteroposterior; (B) lateral.

3. Discussion

Undifferentiated pleomorphic sarcoma (UPS) was previously termed "malignant fibrous histiocytoma" (MFH) which was first to be named by Feldman and Norman.[12] There is a controversy about the origin of MFH. A considerable number of authors thought MFH was originated from a neoplasm composed of fibroblast-like and histiocyte-like elements in varying proportions. According to the WHO classification of soft tissue and bone tumor, UPS has replaced MFH and is defined as a sarcoma with cellular pleomorphism and no identifiable line of differentiation.[13,14] The histological appearance of UPS-B can be various. As a result, it is difficult to differentiate it from other malignant bone tumors. The diagnosis of UPS mainly depends on the histopathology and immunohistochemisty.[7]
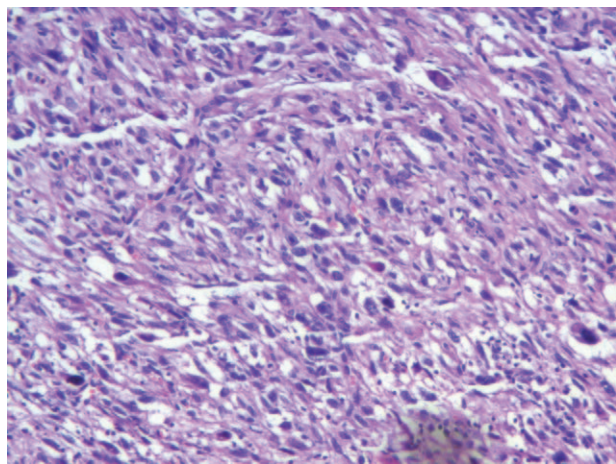
Figure 5. Photomicrograph of the tumor. Microscopic specimen of the lesion shows an admixture of fibroblastic or myofibroblastic spindle cells with a storiform or cartwheel pattern with pronounced inflammatory cell infiltration (Hematoxylin-eosin, H & E, original magnification ×400).

UPS, a high-grade sarcoma, can occur in various bone of the body.[14] The long tubular bones are the most frequently affected sites, such as femur, humerus, tibia, and ulna. It also arises in the short tubular bones, flat bone, and irregular bone, such as sternum, jaw bone, skull, scapula, and spine. It was reported that UPS-B was more around the knee than that in other sites.[8] The characteristic of UPS-B is one bone with the single lesion. Also, skip lesion can be found in UPS-B.[4] The tumor can be transferred to the different positions in a bone by capillary vessel or to the adjacent bone crossing the joint. Our study differs from previous ones in infiltration pattern.

In this case, the lesion in the proximal tibia of the patient, with infiltration of the capsule wall of knee joint, extended to the surrounding bones including the patella and the condyles of femur. No transarticular invasion was found. In the other words, the pattern of metastasis belongs to direct invasion other than crossover. The articular synovium as a media for the tumor diffusion has not been reported much in the literature. A malignant tumor was apparently not observed in a large diarthrodial joint, only few cases were published of sacroiliac and facet joints transarticular involvement.[15,16] This case demonstrated that the articular cartilage, which acted as a barrier to prevent UPS from invading the adjacent tissues, was not involved. All the lesions can be seen on MR images and can be confirmed as UPS-B by the gross specimen.

Imaging characteristics in UPS-B usually are various. Conventional radiograph features of UPS-B show the lesions, accompanying with soft mass, are osteolytic and aggressive growth with cortical destruction. Periosteal reactions, calcification, and ossification may also occur. Reactive sclerosis in the margins of the sites is also the predominantly feature of UPS-B. In spite of no specific criteria, MR images play an important role in describing the lesion borders and dedicating the relationship with the surrounding tissues preoperatively.[8] MR imaging was also of vital importance in defining the marrow involvement and joint invasion.[17] It was reported pre- and post-contrast showed no significant differences for the diagnosis of UPS.[4] No further contrast examination was done in the present study. Link et al[4] suggested the whole skeleton MR scanning in different planes to prevent metastasis in a skipping pattern. As shown by the present case, the lesion appears local osteolytic with cortical destruction in the lateral condyle of tibia plateau. Nevertheless, more lesions demonstrated normal density or osteosclerosis. MR images appeared much more aggressive extent for the tibia lesions than that in x-ray radiographs. Furthermore, the lesion in this case infiltrated the knee joint synovium and ligaments by which the adjacent skeletal were affected. To our knowledge, knee joint synovium in UPS-B, as an infiltration pathway to the surrounding tissues, has not been reported in the previous literature. Such above features are helpful for clinical doctors to give the pretherapy evaluation on this tumor.

Because the study was a retrospective study, no contrast MR examination was done and intraoperative photos were not acquired. Thus, it must be assumed that the outcome of this case report would be more worth to study.

In conclusion, we present a case of UPS in the knee joint. Imaging diagnosis, including CT and MRI is necessary, and biopsy is very helpful in confirming the diagnosis. In contrast to frequent infiltration pathway, the articular synovium as a media for the tumor diffusion is rare. Radiologists and experts in orthopedics should pay a particular attention to this invasion pattern of UPS-B.

Acknowledgments

References

[1] Peiper M, Zurakowski D, Knoefel WT, et al. Malignant fibrous histiocytoma of the extremities and trunk: an institutional review. Surgery 2004;135:59–66.

[2] Efe T, Heyse TJ, Schofer MD, et al. Malignant fibroushistiocytoma of the distal femur after an arthroscopic anterior cruciate ligament rec onstruction: a case report and a review of the literature. BMC Cancer 2010;10:264.

[3] Jeon DG, Song WS, Kong CB, et al. MFH of bone and osteosarcoma show similar survival and chemosensitivity. Clin Orthop Relat Res 2011;469:584–90.

[4] Link TM, Haeussler MD, Poppek S, et al. Malignant fibrous histiocytoma of bone: conventional X-ray and MR imaging features. Skeletal Radiol 1998;27:552–8.

[5] Capanna R, Bertoni F, Bacchini P, et al. Malignant fibrous histiocytoma of bone. The experience at the Rizzoli Institute: report of 90 cases. Cancer 1984;54:177–87.

[6] Yokoyama R, Tsuneyoshi M, Enjoji M, et al. Prognostic factors of malignant fibrous histiocytoma of bone. A clinical and histopathologic analysis of 34 cases. Cancer 1993;72:1902–8.

[7] Romeo S, Bovee JV, Kroon HM, et al. Malignant fibrous histiocytoma and fibrosarcoma of bone: a re-assessment in the light of currently employed morphological, immunohistochemical and molecular approaches. Virchows Arch 2012;461:561–70.

[8] Koplas MC, Lefkowitz RA, Bauer TW, et al. Imaging findings, prevalence and outcome of de novo and secondary malignant fibrous histiocytoma of bone. Skeletal Radiol 2010;39:791–8.

[9] Hoshi M, Matsumoto S, Manabe J, et al. Malignant change secondary to fibrous dysplasia. Int J Clin Oncol 2006;11:229–35.

[10] Bertoni F, Bacchini P, Staals EL. Malignancy in giant cell tumor of bone. Cancer 2003;97:2520–9.

[11] Shaheen M, Deheshi BM, Riad S, et al. Prognosis of radiation-induced bone sarcoma is similar to primary osteosarcoma. Clin Orthop Relat Res 2006;450:76–81.

[12] Feldman F, Norman D. Intra- and extraosseous malignant histiocytoma (malignant fibrous xanthoma). Radiology 1972;104:497–508.

[13] Kelleher FC, Viterbo A. Histologic and genetic advances in refining the diagnosis of "undifferentiated pleomorphic sarcoma". Cancers 2013;5:218–33.

[14] Fletcher CD. The evolving classification of soft tissue tumours - an update based on the new 2013 WHO classification. Histopathology 2014;64:2–11.

[15] Abdelwahab IF, Miller TT, Hermann G, et al. Transarticular invasion of joints by bone tumors: hypothesis. Skeletal Radiol 1991;20:279–83.

[16] Chhaya S, White LM, Kandel R, et al. Transarticular invasion of bone tumours across the sacroiliac joint. Skeletal Radiol 2005;34:771–7.

[17] Pobirci DD, Bogdan F, Pobirci O, et al. Study of malignant fibrous histiocytoma: clinical, statistic and histopatological interrelation. Rom J Morphol Embryol 2011;52:385–8.