Skeletal Radiol (2001) 30:530–533

CASE REPORT

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Primary leiomyosarcoma of the patella

Received: 21 November 2000
Revision requested: 8 January 2001
Revision received: 2 April 2001
Accepted: 4 April 2001

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Abstract We report on a case of primary leiomyosarcoma of the patella in a 59-year-old woman. To our knowledge, this is the first report of this rare tumor in a bone where malignant lesions rarely occur.

Keywords Leiomyosarcoma · Patella · Radiographs · MRI

Introduction

Pain and swelling in the knee are common complaints, and most patients have degenerative changes in the knee to account for them. However, few patients have patellar tumors, and rarely are these malignant [1, 2]. Nor do we commonly see primary leiomyosarcoma in bone [3, 4]. As far as we can determine, this is the first report of a case of primary leiomyosarcoma of the patella.

Case report

A 59-year-old woman had complained of pain and swelling in the right knee for about 10 years. The symptoms worsened, and she visited a hospital in June, 1998. Radiographs of her knee joint demonstrated a mixed lytic and sclerotic lesion in the patella. The lesion was ill defined at its margin and showed no cortical changes (not shown). Magnetic resonance imaging (MRI) scans of the patellar lesion showed low intensity on T1-weighted images and heterogeneous low and high intensities on T2-weighted imaging (not shown). A technetium-99m (^{99m}Tc) bone scan showed isolated increased activity in the right patella (not shown). Open biopsy of the patellar lesion was performed; however, histologic examination of the surgical specimen did not permit a clear diagnosis; it suggested that the lesion may be osteomyelitis or a benign fibrohistiocytic tumor with secondary changes. The lesion was subsequently treated with curettage and bone graft with complete resolution of the patient's symptoms.

On July 16, 1999, the patient experienced a sudden onset of pain in the anterior portion of the right knee, following which she was referred to our institution. Clinical examination upon presentation showed swelling and local heat in the right knee, with an elastic hard mass surrounding the scar that traversed the patella. Passive range of motion of the right knee was from 0° to 120°, whereas that of the left knee was from 0° to 140°. The circumference of the right thigh showed muscle atrophy by 1 cm. Clinical hematologic tests including WBC count, serum levels of alkaline phosphatase and C-reactive protein were within normal limits. Radiographs of the right knee showed an osteolytic lesion in the patella.

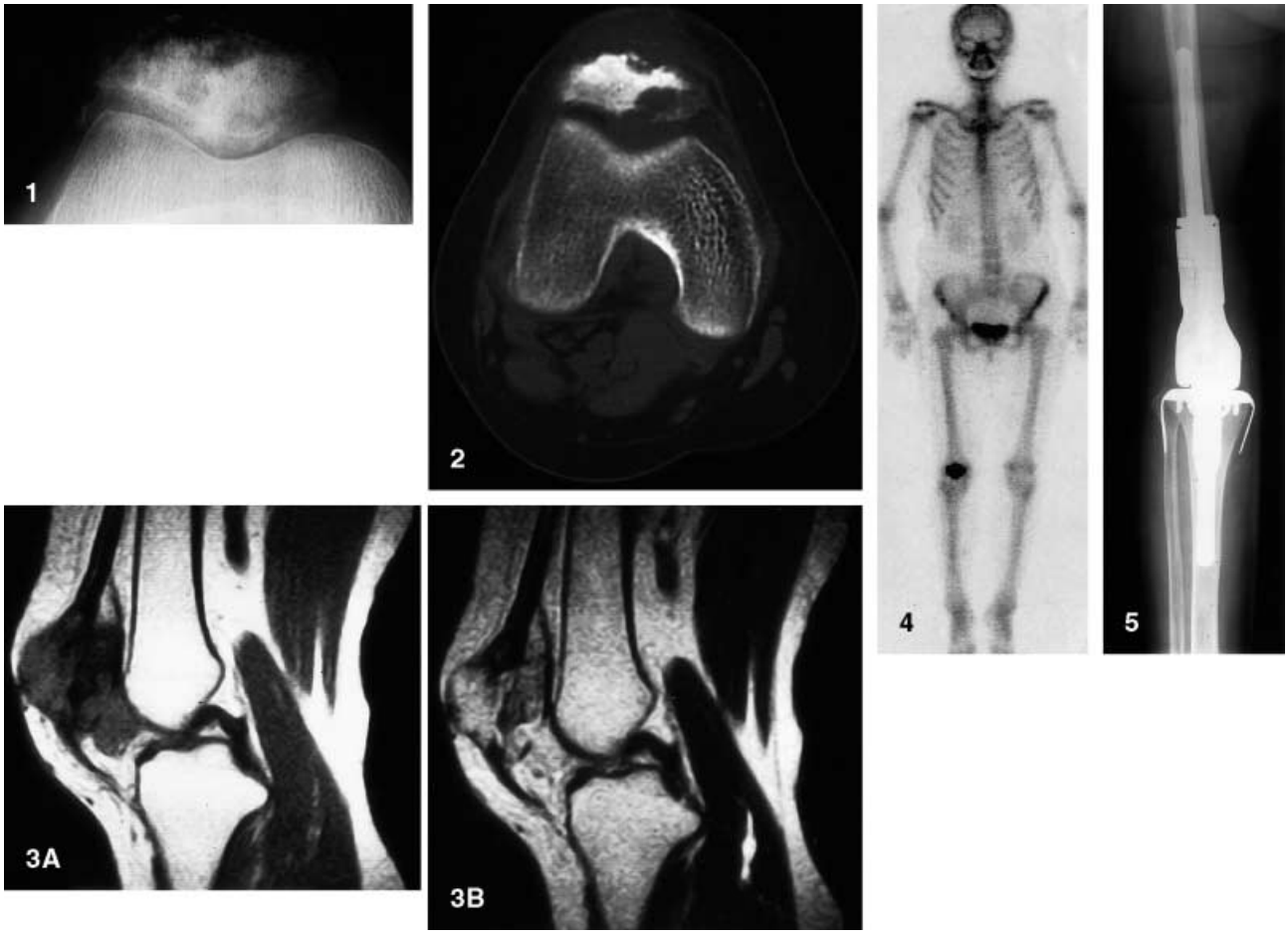

Fig. 1 Axial view of the patella shows a mixed osteolytic and sclerotic lesion. The margin of the lesion is ill defined and associated with cortical disruption

Fig. 2 CT scan through the patella shows admixtures of sclerotic and osteolytic change with cortical disruption

Fig. 3 **A** T1-weighted sagittal MR image shows a low signal intensity lesion with extraosseous intra-articular extension into the patellofemoral aspect of the knee joint. **B** T2-weighted sagittal MR image shows high signal intensity within the patella with the extraosseous infiltration

Fig. 4 A ^{99m}Tc bone scan shows focal increased uptake in the right patella

Fig. 5 A wide resection of the proximal tibia and the distal femur with total patellectomy was performed and a Howmedica modified resection system was inserted

The margins of the lesion were ill defined associated with cortical disruption. Some sclerotic changes in the lesion were also observed; however, periosteal reaction or extraosseous extension of the lesion was not evident (Fig. 1). Computed tomography (CT) scans also showed the heterogeneous osteolytic and sclerotic lesions in the patella (Fig. 2). MRI showed a lesion of low T1- and high T2-weighted signal intensity within the patella with extraosseous infiltration to the subcutis and joint cavity (Fig. 3). The lesion showed focal increased activity on ^{99m}Tc bone scanning (Fig. 4), with corresponding increased activity in the right patella on gallium-67 (^{67}Ga) imaging. No other skeletal sites of increased ^{99m}Tc or ^{67}Ga activity were seen.

Open biopsy was subsequently performed at our hospital. Histologic examination suggested the diagnosis of sarcoma, although this was not conclusive.

The patient was treated with an extra-articular wide resection with total patellectomy and reconstruction using a Howmedica modified resection system (Fig. 5). We resected the patella with a portion of the quadriceps muscles, the joint capsule, distal femur (12 cm proximal to the joint line), and proximal tibia (1 cm distal to the joint line). The distal portion of the femur and proximal portion of the tibia were replaced by a Howmedica modified resection system. After the resection we advanced the quadriceps muscles and sutured with a patellar ligament. The sartorius muscle and gracilis muscle were cut and transferred to augment the quadriceps-patellar ligament junction. Pre- and postoperative chemotherapy were not given. The final pathologic diagnosis of the resected tumor was primary leiomyosarcoma of the patella (Fig. 6).

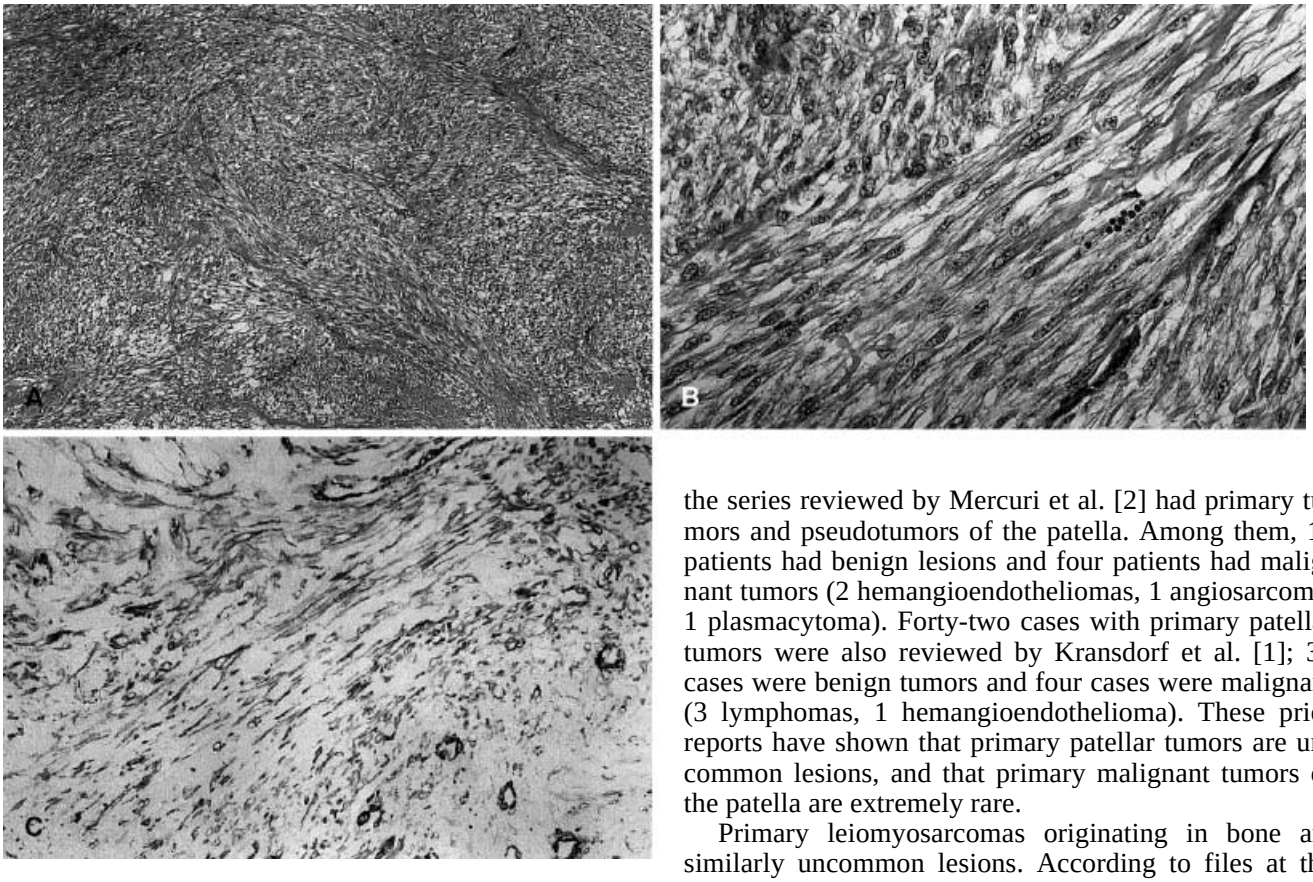

Fig. 6 **A** Low-power photomicrograph depicting fascicles of spindle cells (hematoxylin-eosin stain, ×160). **B** High-power photomicrograph depicting centrally spindle cells with blunt ended nuclei (hematoxylin-eosin stain, ×970). **C** The spindle cells show immunoreactivity for muscle-specific actin (HHF35) (immunoperoxidase, DAB, ×400)

To exclude the possibility that the tumor was metastatic leiomyosarcoma from the gastrointestinal tract or uterus, CT scans through the chest and abdomen were performed; however, no lesions were found in these locations.

Twenty months have passed since the operation and there has been no evidence of local recurrence or distant metastasis. Postoperatively, the patient has a passive range of motion of the knee of 0° to 80°. Although she cannot extend the right knee against gravity, she can walk without pain using a knee brace.

Discussion

Primary bone tumors arising in the patella are rare lesions [1, 2, 5, 6, 7, 8]. In a review by Ferguson et al. [5] of eight cases of primary patellar tumors, there were six benign cases and two malignant cases (1 malignant fibrous histiocytoma, 1 osteosarcoma). The 15 patients in the series reviewed by Mercuri et al. [2] had primary tumors and pseudotumors of the patella. Among them, 11 patients had benign lesions and four patients had malignant tumors (2 hemangioendotheliomas, 1 angiosarcoma, 1 plasmacytoma). Forty-two cases with primary patellar tumors were also reviewed by Kransdorf et al. [1]; 38 cases were benign tumors and four cases were malignant (3 lymphomas, 1 hemangioendothelioma). These prior reports have shown that primary patellar tumors are uncommon lesions, and that primary malignant tumors of the patella are extremely rare.

Primary leiomyosarcomas originating in bone are similarly uncommon lesions. According to files at the Basel Bone Tumor Reference Center, the rate of leiomyosarcoma of bone was 0.64% of all malignant neoplasms originating in bone [9]. Since Evans and Sanerkin described the first case in 1965 [4], about 60 cases of primary leiomyosarcoma of bone have been reported in the literature [3, 4, 9, 10, 11, 12, 13, 14, 15]. Antonescu et al. [3] reviewed 33 cases of primary leiomyosarcoma of bone . The mean age of the patients in this series was 44.4 years (range 13–77 years); 16 were male (48%) and 17 female (52%). Presenting complaints in this group of patients included pain (85%), or the presence of a mass and pathologic fracture (15%). The lesions were located predominantly in the long tubular bones, especially in the proximal tibia (12 cases) and the distal femur (7 cases). Other sites of disease in this series included the mandible (6 cases), the pelvic bones (3 cases), the proximal humerus (2 cases), the maxilla (1 case), the vertebrae (1 case) and the clavicle (1 case). Twelve cases of primary leiomyosarcoma of bone were reviewed by Sundaram and coworkers [14], four of which occurred in the femur, three in the tibia and five in the pelvic bones. Other authors have reported cases involving the fibula [12, 13], rib [11] and terminal phalanx of finger [9]. To our knowledge, the present report is the first case of primary leiomyosarcoma of the patella.

In general, it is not easy histologically to differentiate primary leiomyosarcoma originating in bone from other

primary tumors of bone. Differential diagnosis includes metastatic leiomyosarcoma (gastrointestinal tract, uterus) or other sarcoma including fibrosarcoma, malignant fibrous histiocytoma or malignant peripheral nerve sheath tumor [9, 10]. To designate a bone tumor as primary leiomyosarcoma, a metastatic lesion must be excluded. Most soft tissue leiomyosarcomas arise principally in the retroperitoneal space and abdominal cavity, and from the uterus in the pelvis. CT or MRI scans through the abdomen and pelvis are useful to exclude primary tumors in these locations, and CT of the chest would exclude lung metastases. A ^{67}Ga scintigram is also useful to detect systemic lesions.

In the first open biopsy, the tumor was diagnosed as a benign lesion. Although we have been unable to re-evaluate the initial pathologic material, the initial diagnosis is questionable. This presumed misdiagnosis might be due to poor sampling of the tumor in the first biopsy. In the histologic examination of our case, the resection specimen showed proliferation of spindle-shaped cells in interlacing fascicles. The tumor cells had mildly atypical nuclei, some of which were elongated. Mitotic figures including atypical ones were sometimes seen. Kawai et al. [10] reported that immunohistochemical study is very useful for diagnosis. In our case, the tumor cells were positive for myogenic markers (HHF35 and alpha-smooth muscle actin) and weakly positively staining for desmin. These histologic features indicated that the tumor in the present case was in keeping with the diagnosis of osseous leiomyosarcoma. Bone and ^{67}Ga scintigrams confirmed the solitary nature of osseous disease, while CT scans of the chest and abdomen excluded systemic disease or primary malignancy of the uterus or gastrointestinal tract. The patient has remained disease free for 20 months following treatment.

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