

Spindle-cell sarcoma of the popliteal fossa mimicking a benign vascular lesion

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Spindle-cell sarcomas are a group of aggressive malignant soft-tissue tumors with diverse clinical presentations. While some of these tumors may represent de novo malignant transformation of benign entities, others may present with a long indolent course before their invasive nature is apparent. We report the case of a previously healthy 73-year-old female with spindle-cell sarcoma of the left popliteal fossa who initially presented with a painless mass of several months' duration. Magnetic resonance imaging (MRI) suggested a benign vascular lesion/venous angioma, but after the patient's clinical course changed, repeat MRI 18 months later was consistent with an infiltrative soft-tissue malignancy/sarcoma. Excisional biopsy revealed a stage II spindle-cell sarcoma. The patient then underwent radiotherapy and subsequent above-the-knee amputation. Three years following the amputation, the patient is free of local tumor recurrence or metastatic disease.

Introduction

Spindle-cell sarcomas include a wide variety of malignant soft-tissue tumors that are grouped based on their immunophenotype and microscopic structure (1-2). The World Health Organization (WHO) classification divides the spindle-cell sarcoma tumors into the following types: malignant fibrous histiocytoma (the most common type), synovial sarcoma, leiomyosarcoma, malignant peripheral nerve sheath tumors, fibrosarcomas, and unclassified tumors (2-5). These tumors have a variety of clinical presentations and are characterized by an aggressive nature with

metastatic potential (1, 4). On computed tomography (CT) and MRI, they typically appear as heterogeneous masses with avid contrast enhancement and potential local invasion (5-8).

Case report

A 73-year-old female presented to her primary care physician with a history of focal swelling at the posterior aspect of her left knee, of several months' duration. This was initially painless and did not limit mobility at the knee joint. The patient denied any injury to the region of concern and denied any musculoskeletal disorder. Physical examination revealed a palpable mass approximately 2 cm in length in the medial aspect of the popliteal fossa of the left knee. The mass was firm and nontender on palpation, and there was no overlying skin discoloration. Normal arterial pulses were noted proximal to and distal to the lesion, as well as a palpable popliteal artery pulse adjacent to the lesion. There was mild edema of the left calf compared with the right. A normal ankle reflex was noted.

Radiographs of the left knee (not shown) were performed and revealed no significant abnormality, with mild age-related degenerative changes present. Initial MRI examination revealed a subcutaneous soft-tissue lesion posterior to the neurovascular bundle and medial head of the gastrocnemius musculature in the region of the left popliteal fossa and extending along the proximal calf. The lesion was

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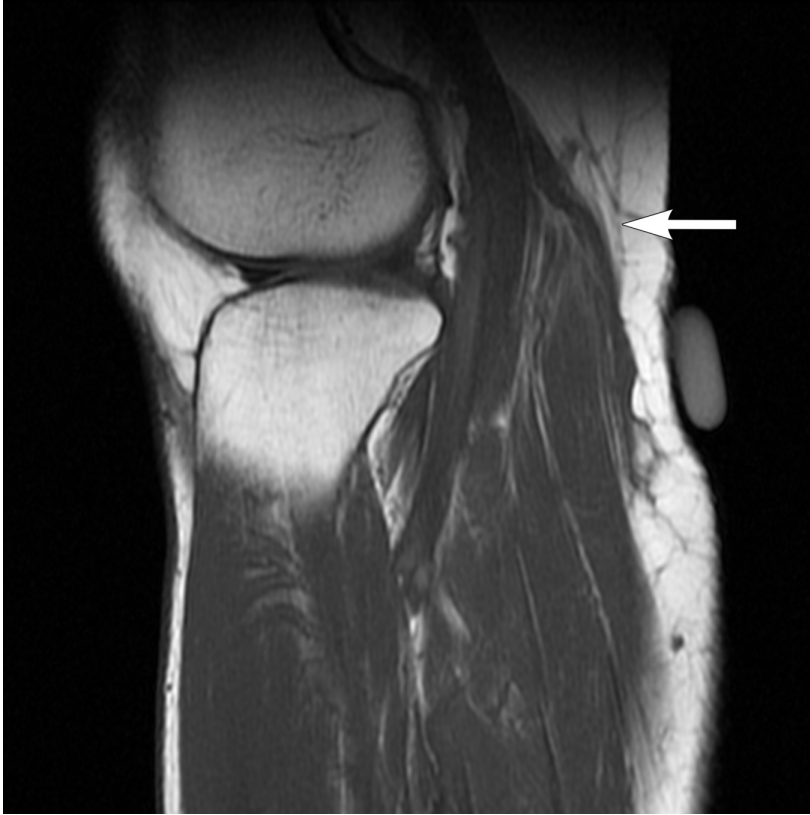


Figure 1A. 73-year-old female with spindle-cell sarcoma. Initial MRI study. Sagittal T1W T1 fat-suppressed contrast-enhanced image of the left knee shows a tangle of vessels in the subcutaneous soft tissues posterior to the neurovascular bundle and medial head of gastrocnemius musculature. Extension over proximal calf (arrow) with marked enhancement on the postcontrast images (arrow) was erroneously thought to represent a venous angioma. Note intermediate signal intensity of the lesion.

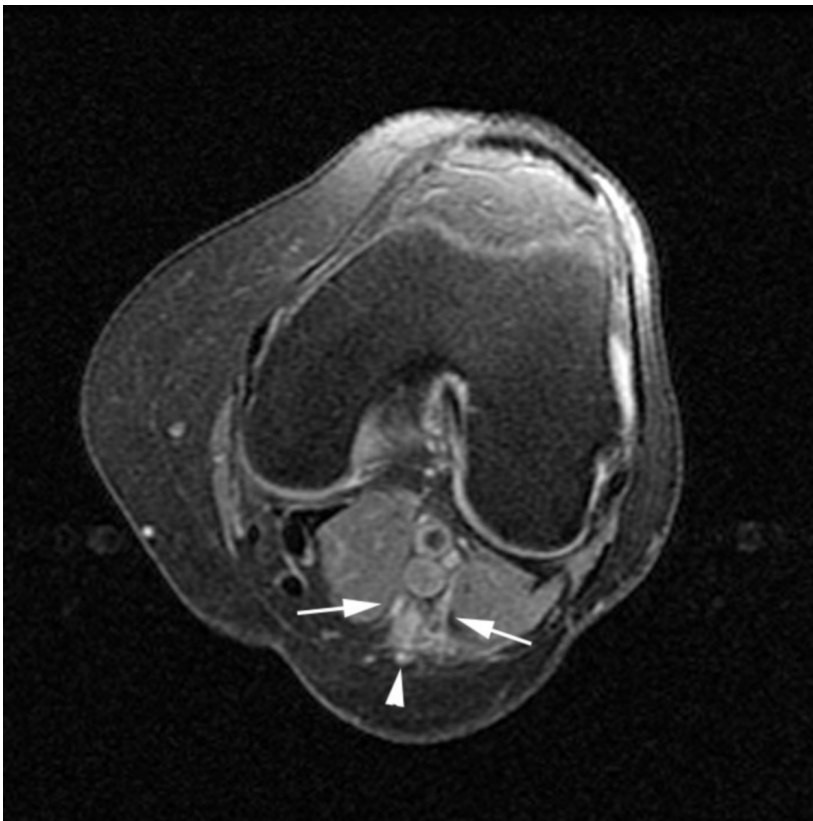


Figure 1B. 73-year-old female with spindle-cell sarcoma. Initial MRI study. Axial PDW fat-suppressed T1 fat-suppressed contrast-enhanced image of the left knee shows a tangle of vessels in the subcutaneous soft tissues posterior to the neurovascular bundle and medial head of the gastrocnemius musculature. Extension over proximal calf with marked enhancement on the postcontrast images (arrows) was erroneously thought to represent a venous angioma. Note intermediate increased signal intensity of the lesion as well as connections with the popliteal vein (arrowhead).

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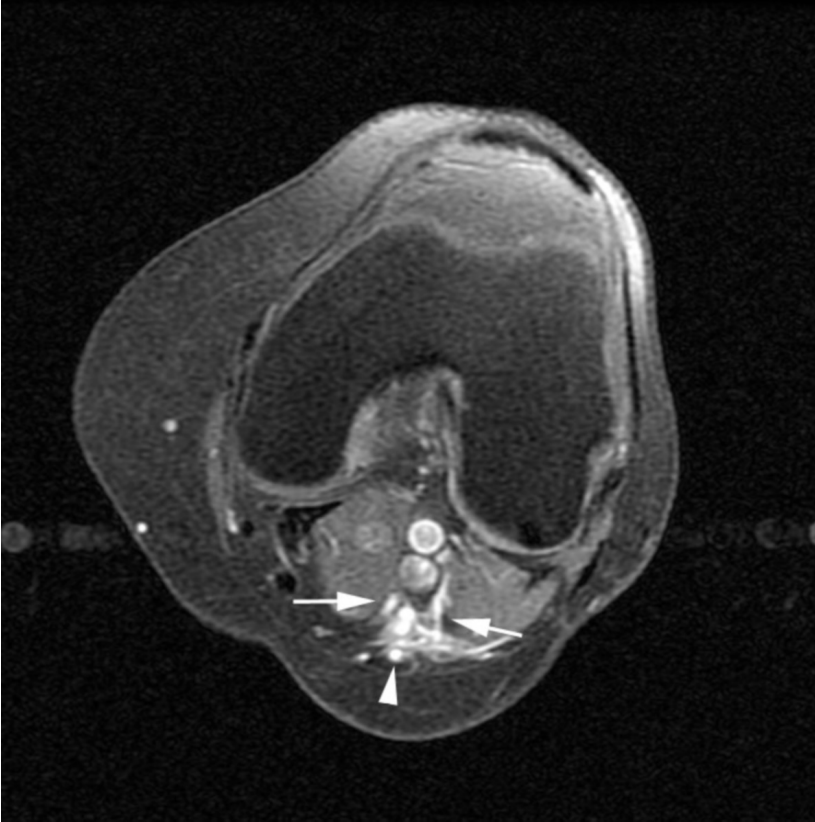


Figure 1C. 73-year-old female with spindle-cell sarcoma. Initial MRI study. Axial T1 fat-suppressed contrast-enhanced image of the left knee shows a tangle of vessels in the subcutaneous soft tissues posterior to the neurovascular bundle and medial head of gastrocnemius musculature. Extension over proximal calf with marked enhancement on the postcontrast images (arrows) was erroneously thought to represent a venous angioma. Note intermediate signal intensity of the lesion as well as connections with the popliteal vein (arrowhead).

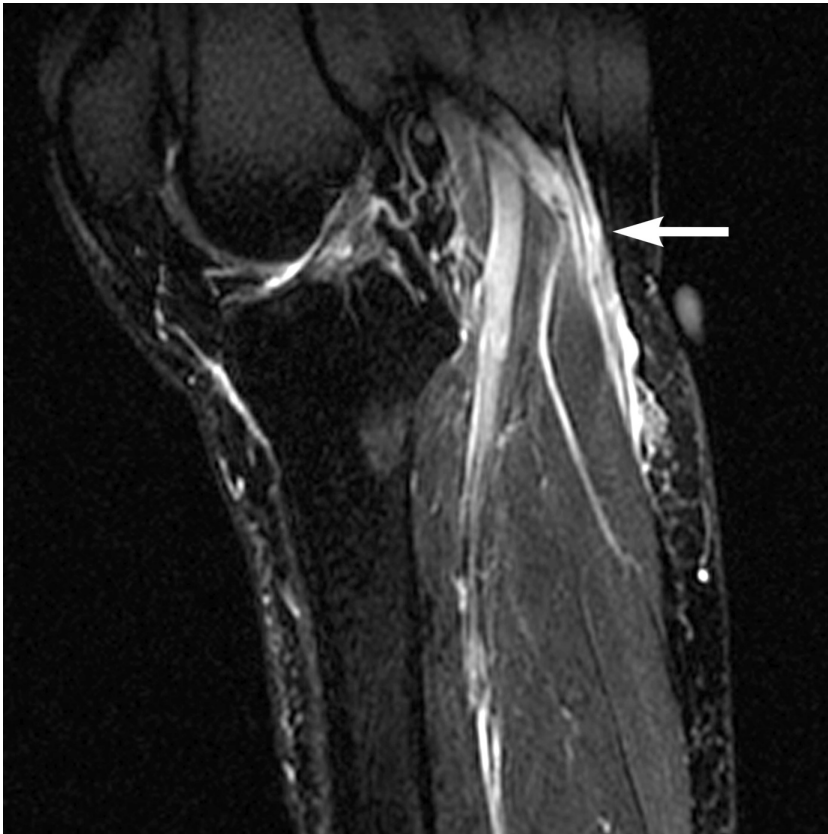


Figure 1D. 73-year-old female with spindle-cell sarcoma. Initial MRI study. Sagittal T1 fat-suppressed contrast-enhanced image of the left knee shows a tangle of vessels in the subcutaneous soft tissues posterior to the neurovascular bundle and medial head of gastrocnemius musculature. Extension over proximal calf (arrow) with marked enhancement on the postcontrast images (arrow) was erroneously thought to represent a venous angioma.

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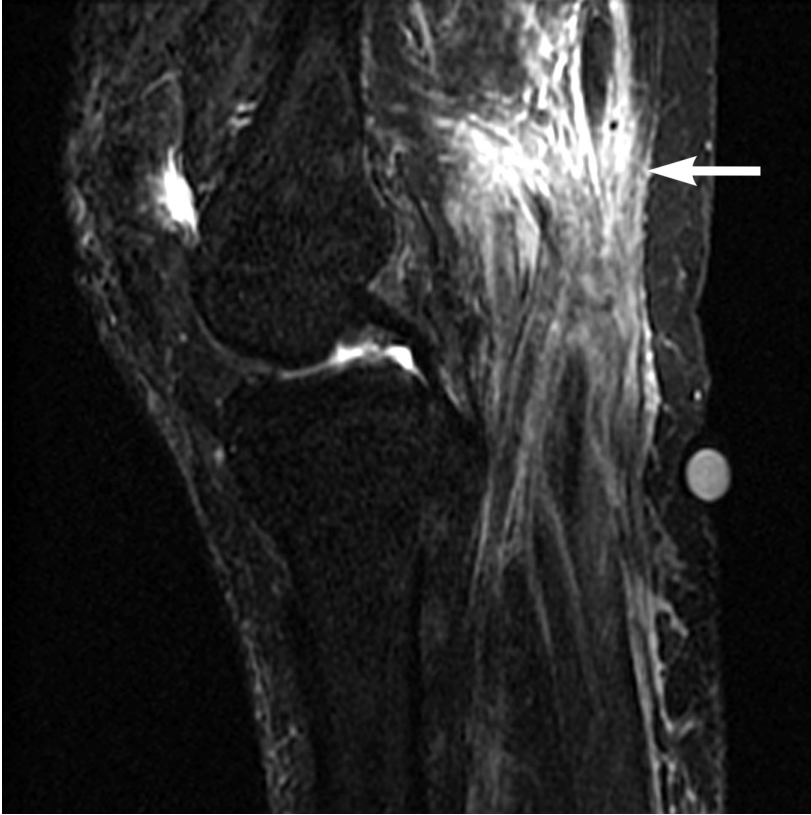


Figure 2A. 73-year-old female with spindle-cell sarcoma. Followup MRI examination 18 months after initial presentation. Sagittal STIR fat-suppressed image of the left knee shows a large ill-defined infiltrative soft-tissue mass centered in the popliteal fossa with invasion of the neurovascular bundle (arrow) consistent with a soft-tissue sarcoma. The lesion shows intermediate-increased signal on the STIR images. Note marked interval enlargement of the lesion since the initial MRI examination (Fig. 1).

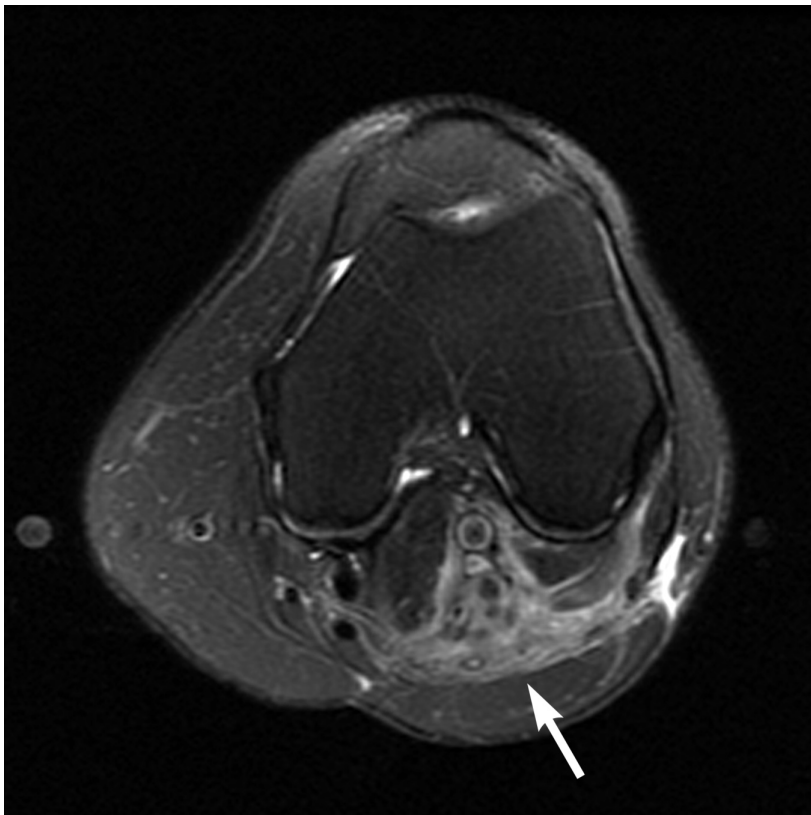


Figure 2B. 73-year-old female with spindle-cell sarcoma. Followup MRI examination 18 months after initial presentation. Axial STIR fat-suppressed image of the left knee shows a large ill-defined infiltrative soft-tissue mass centered in the popliteal fossa with invasion of the neurovascular bundle (arrow) consistent with a soft-tissue sarcoma. The lesion shows intermediate-increased signal on the STIR images. Note marked interval enlargement of the lesion since the initial MRI examination (Fig. 1).

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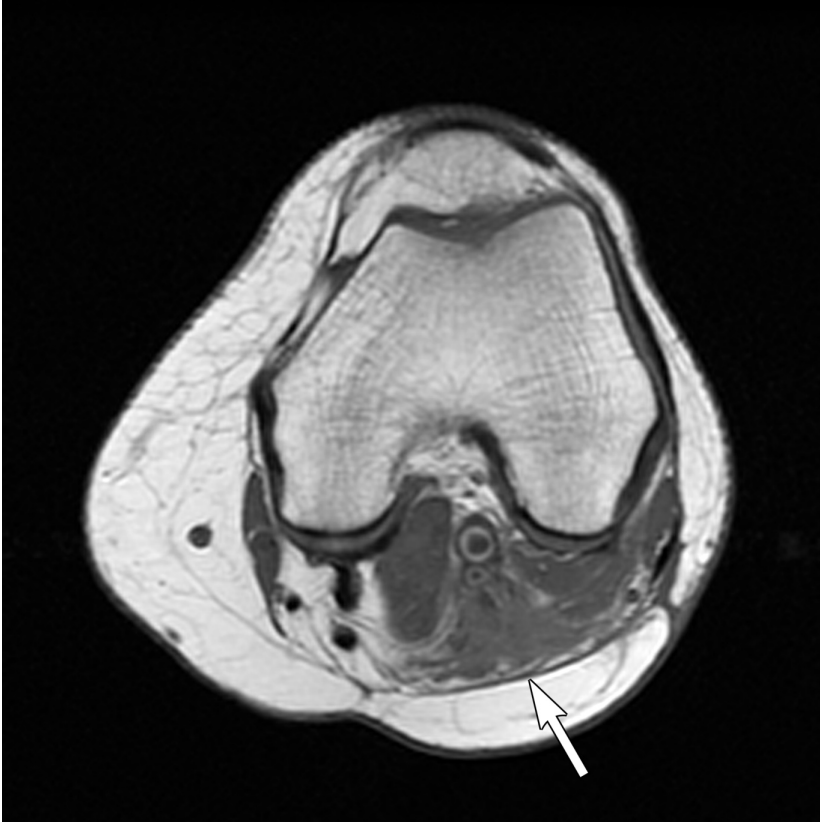


Figure 2C. 73-year-old female with spindle-cell sarcoma. Followup MRI examination 18 months after initial presentation. Axial T1W image of the left knee shows a large ill-defined infiltrative soft-tissue mass centered in the popliteal fossa with invasion of the neurovascular bundle (arrow) consistent with a soft-tissue sarcoma. The lesion shows intermediate signal intensity on the T1W image. Note marked interval enlargement of the lesion since the initial MRI examination (Fig. 1).

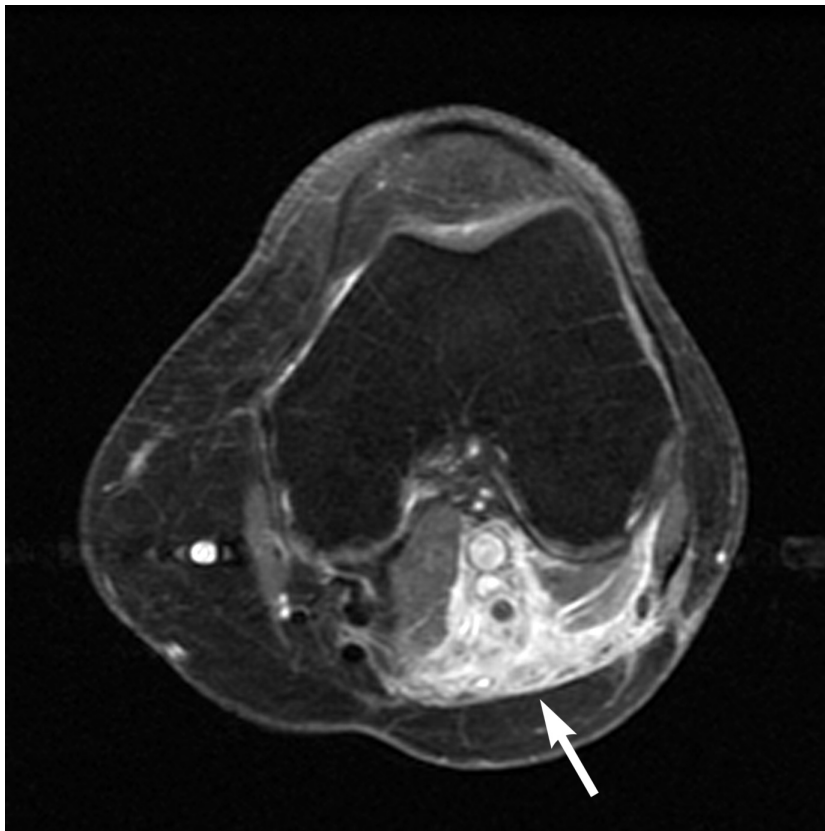


Figure 2D. 73-year-old female with spindle-cell sarcoma. Followup MRI examination 18 months after initial presentation. Axial T1W fat-suppressed with intravenous contrast image of the left knee shows a large ill-defined infiltrative soft-tissue mass centered in the popliteal fossa with invasion of the neurovascular bundle (arrow) consistent with a soft-tissue sarcoma. The lesion shows significant enhancement on the postcontrast image. Note marked interval enlargement of the lesion since the initial MRI examination (Fig. 1).

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composed of a tangle of vessels with connection to the popliteal vein, demonstrating intermediate signal intensity on the T1W and intermediate increased signal intensity on the fluid-sensitive sequences, with marked enhancement on the T1W fat-suppressed sequences following intravenous administration of gadolinium-based contrast (Fig. 1). The lesion measured approximately 6.3 (craniocaudal) x 3.2 (transverse) x 0.7 (anteroposterior) cm. There were no other focal soft-tissue or bony abnormalities. Given the imaging characteristics and clinical presentation, a diagnosis of a probable venous angioma was made by two radiologists, musculoskeletal and vascular-interventional subspecialists in consensus.

One year later, the patient noticed increasing weakness in her left calf and gradual enlargement of the mass. Eighteen months after the initial presentation, she was re-evaluated by her primary care physician. At that time the patient reported pain at the site of the lesion. On clinical examination, the mass was noted to have significantly increased in size since the initial presentation and was slightly tender to palpation.

MRI imaging was then repeated and revealed the presence of a large ill-defined infiltrative soft-tissue mass centered in the popliteal fossa with invasion of the neurovascular bundle. The lesion showed nonspecific intermediate signal intensity on the T1W images and intermediate-to-increased signal on the fluid-sensitive sequences. The previously seen, presumed “venous angioma” was visualized at the posterior aspect of the lesion and appeared ill-defined (Fig. 2). These findings were consistent with an aggressive/malignant, locally invasive, soft-tissue tumor/sarcoma that prompted an excisional biopsy.

On histopathology, the lesion demonstrated strong staining for vimentin and CD68, with low and intermediate staining for cytokeratins, neurofilament, desmin, CD34, S-100, and actin. The neoplastic cells were mitotically active and pleomorphic, with some areas of myxoid stroma (Fig. 3). There was no necrosis. This was felt to represent a poorly differentiated spindle-cell sarcoma, stage II, according to American Joint Committee on Cancer (AJCC) staging criteria (G3T1bN0M0). Surgical margins were positive. The differential diagnosis included a myxofibrosarcoma, undifferentiated liposarcoma, and sarcoma not otherwise specified. Immunohistochemical studies helped to exclude a carcinoma or melanoma.

Given the difficulty in completely excising the lesion at the time of surgery, the patient underwent neoadjuvant radiotherapy and subsequent above-the-knee amputation. A CT scan of the chest, abdomen, and pelvis revealed no evidence for metastatic disease.

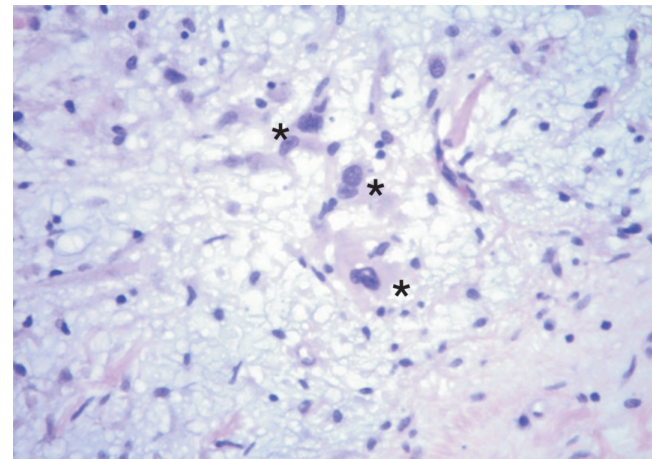
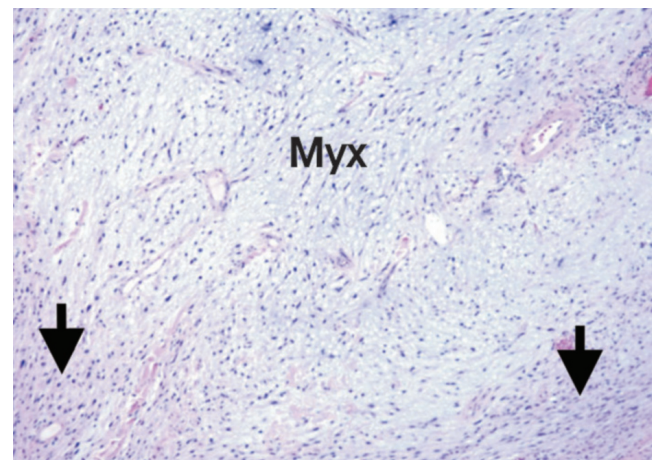
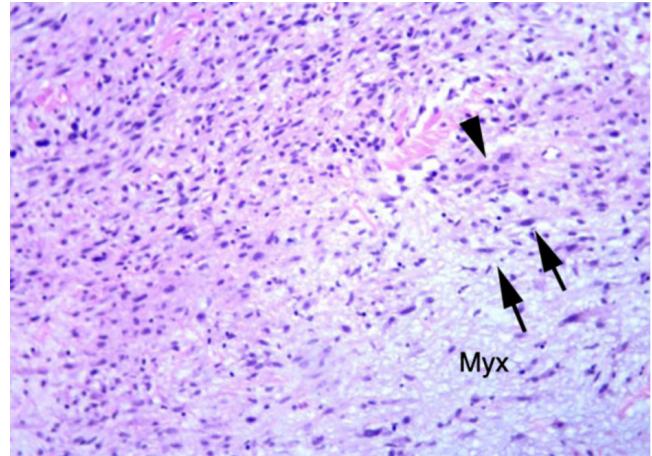


Figure 3. Histopathology. A. Medium-power magnification view of spindle-cell sarcoma demonstrating an area of myxoid morphology (Myx) and a mixed spindle-cell (arrows) and polygonal-cell (arrowhead) population. Hematoxylin and eosin stain. Original magnification, 200x. B. Low-power view of spindle-cell sarcoma with a large central area of myxoid matrix (Myx). Spindle cells at corners (arrows) show a swirling arrangement of cells with parallel nuclei. Hematoxylin and eosin stain. Original magnification, 100x. C. High-power view highlighting markedly abnormal nuclei (asterisks), which have variation in nuclear size and shape, irregular nuclear outlines, and hyperchromasia characteristic of a high-grade sarcoma. Hematoxylin and eosin stain. Original magnification, 400x.

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Three years following the amputation, the patient is free of local tumor recurrence or metastatic disease.

Discussion

The spindle-cell sarcoma group represents approximately half of adult soft-tissue sarcomas, and includes several histologic types (2, 4). These tumors can be a diagnostic challenge for radiologists and surgical pathologists, with molecular and immunohistochemical staining representing a key diagnostic tool (9). Included in the spindle-cell sarcoma group are malignant fibrous histiocytomas (the most common type), synovial sarcomas, leiomyosarcomas, malignant peripheral nerve-sheath tumors, fibrosarcomas, and unclassified tumors (2-5). These tumors are known to have an aggressive nature, with recurrent and metastatic behavior reported and corresponding to tumor grade (1, 4). Their immunophenotype and microscopic structure are consistent with myofibroblastic differentiation (1). Prognosis for these tumors may depend on expression of cell cycle markers such as cyclin E and p27Kip1 (3).

The spindle-cell sarcoma group of tumors has a variety of clinical presentations. Malignant transformation from a low- to high-grade tumor over many years can occur (10). Leiomyosarcoma arising from vascular wall mimicking venous thrombus has been reported (6). Rarely, malignant transformation of osteochondroma to spindle-cell sarcoma has been reported (11). Other clinical presentations, such as one mimicking Dupuytren's contracture nodules and a large tumor of small-bowel mesentery, have been noted (12-13).

As with the other soft-tissue tumors, the primary imaging modality is MRI, which allows characterization of the lesion based on signal characteristics on the precontrast T1W and fluid-sensitive sequences, size, and pattern of enhancement on the T1W fat-suppressed postcontrast sequences (5, 7-8, 14). In fact, MRI is superior to CT in defining the local extent of the tumor, which is important for treatment planning, while PET/CT imaging may assist in determining metabolic activity of a lesion (8). Like many other locally invasive malignant soft-tissue tumors, the spindle-cell sarcoma group may have heterogeneous appearance on MRI and CT, with avid postcontrast enhancement (8, 14). Ultimately, multiple imaging modalities may provide optimal assessment of these lesions, with biopsy representing the key step in diagnosis (14).

In summary, the soft-tissue tumor in the popliteal fossa/calf of an elderly female patient reported in this case represents the immunophenotype of undifferentiated spindle-cell sarcoma that on initial presentation mimicked a benign venous angioma.

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