SPINDLE CELL SARCOMAS

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KEYWORDS

• Synovial sarcoma • Malignant peripheral nerve sheath tumor • Fibrosarcoma
• Inflammatory myofibroblastic tumor • Myofibrosarcoma • Leiomyosarcoma
• Spindle cell rhabdomyosarcoma • Endothelial neoplasms

ABSTRACT

Information is presented on the pathology of spindle cell sarcomas. Synovial sarcoma, malignant peripheral nerve sheath tumor, fibrosarcoma, inflammatory myofibroblastic tumor, low-grade myofibrosarcoma, leiomyosarcoma, spindle cell rhabdomyosarcoma, and endothelial neoplasms are discussed in terms of an overview of the tumor, microscopic and gross features, diagnostic techniques, genetic markers, differential diagnosis, clinical details, and prognosis.

OVERVIEW

Spindle cell sarcomas constitute a large and diverse morphologic category of tumors composed of elongated cells with rounded, ovoid, or tapered nuclei and variable amounts of cytoplasm, disposed in a variety of patterns with a range of appearances modified by stromal fibrosis or myxoid accumulation. They occur in soft tissue, bone, or viscera; represent a variety of cell lineages; and exhibit a range of behavior so that accurate diagnosis and grading are essential for correct clinical management. Because of their common cell morphology and limited patterns, they can be difficult to distinguish from each other and from some reactive lesions.

The clinical history is frequently contributory, and subtle morphologic clues are present in most cases. Their identification and interpretation, however, require experience as well as familiarity with the application of ancillary diagnostic methods, including immunohistochemistry and, increasingly, molecular genetic techniques.

Spindle cell sarcomas include synovial sarcoma, malignant peripheral nerve sheath tumor, leiomyosarcoma, fibrosarcoma, rhabdomyosarcoma, inflammatory myofibroblastic tumor, and angiosarcoma. In the abdomen, gastrointestinal stromal tumor, dedifferentiated liposarcoma, and follicular dendritic cell sarcoma enter the differential diagnosis. It should also be remembered that spindle cell tumors of nonmesenchymal lineage can occur in many anatomic locations and mimic sarcomas. These include carcinoma, especially in relation to epithelial structures, and viscera, melanoma, and some lymphoreticular tumors, notably follicular dendritic cell sarcoma.

SYNOVIAL SARCOMA

OVERVIEW

Synovial sarcoma was first described in detail, as a "primitive sarcoma of synovial joints," more than a century ago by Lejars and Rubens-Duval, who illustrated both glandular and spindle cell components.[1] The successive introduction of diagnostic techniques has established synovial sarcoma as a translocation-associated sarcoma with variable epithelial differentiation that is not related to synovium and only very rarely arises in any joint. The most common presentation is in a young adult as a deep soft tissue mass around or adjacent to a large joint, usually the knee, but it can occur at any anatomic site,[2] including superficial soft tissues, viscera, and head and neck. It can be initially indolent with the patient describing gradual enlargement for many years, but very small examples (smaller than 1 cm) are sometimes encountered, especially in the distal extremities.[3] Radiologic techniques reveal variably sized foci

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Key Features
Synovial Sarcoma

- Can occur in any location but is most common around the knee.
- Origin within joints is extremely rare.
- Nuclear pleomorphism is almost always lacking.
- Biphasic tumors have glandular epithelial areas with mucin secretion.
- Monophasic spindle cell tumors have fascicles of uniform short spindle cells with scanty cytoplasm.
- Focal calcification is a typical feature and some tumors show ossification.
- Poorly differentiated synovial sarcoma is often a small round cell tumor.
- A hemangiopericytic pattern is commonly seen in all variants.
- Expression of CK, EMA, CD99, and S100 protein is focal and variable.
- bcl2 is diffusely expressed and CD34 is absent.
- There is consistent expression of TLE1 (nuclear).
- Specific t(X;18)(p11.2;q11.2) chromosomal rearrangement is a diagnostic feature.

of calcification in many cases. Rarely, synovial sarcoma arises following radiation therapy.[4–6]

GROSS FEATURES

Synovial sarcoma is typically circumscribed but not encapsulated, with a soft, white-tan cut surface. Cyst formation and hemorrhage are sometimes seen, with focal necrosis in poorly differentiated examples. Spontaneous total infarction is a rare phenomenon.

MICROSCOPIC FEATURES

Biphasic synovial sarcoma has an epithelial and a spindle cell component. The epithelial component can be glandular or adenopapillary (with neoplastic spindle cells rather than collagen in the papillary core), or form solid nodules or cords of cuboidal cells with oval vesicular nuclei, moderate amounts of cytoplasm, and distinct cell membranes. Rare variations include large, dilated glands with scanty spindle cell component, and widespread keratinizing squamous metaplasia.

The spindled component can be sparse, predominant, or occur alone as monophasic spindle cell synovial sarcoma (which is encountered more frequently than typical biphasic synovial sarcoma). The spindle cells (**Fig. 1**) have typically uniform, relatively small, short spindle or oval pale-staining nuclei, the edges of which can appear to overlap

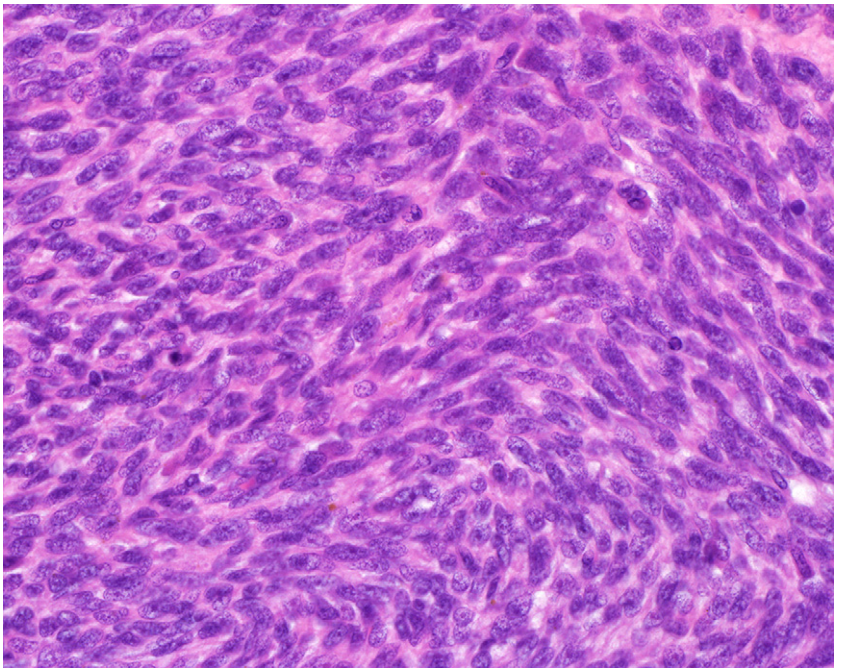

Fig. 1. Synovial sarcoma. The monophasic spindle variant is composed of fascicles of uniform short spindle cells, with nuclei that appear to overlap. Note the mast cells (hematoxylin-eosin [H&E], original magnification ×100).