Synovial sarcoma

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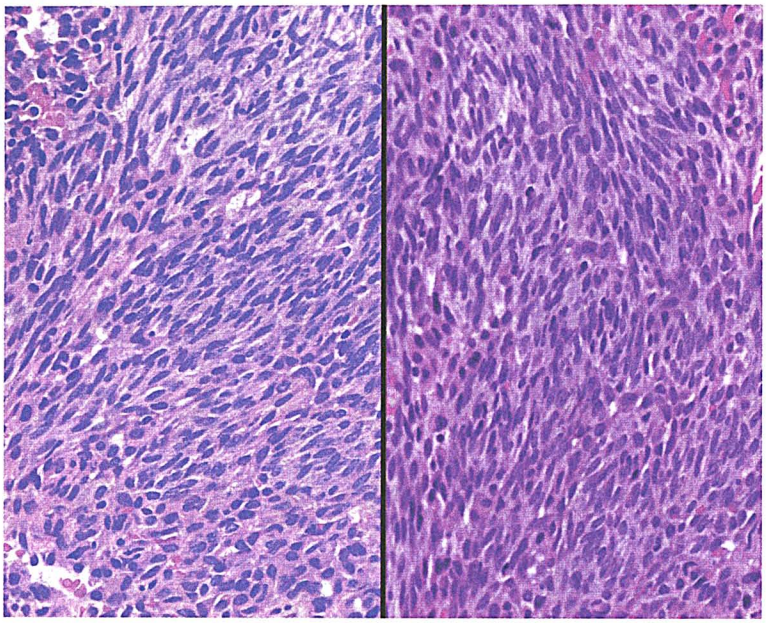

Figure 1. This monophasic SS is made up of densely packed and relatively uniform spindle cells. The cells have very little cytoplasm, and their borders are indistinct. Special studies are required to confirm the diagnosis.

Synovial sarcoma (SS) is a unique tumor that usually involves the large joints. Approximately 10% of these tumors develop in the neck, oropharynx, hypopharynx, and larynx. Despite the name, these tumors neither originate from synovium nor express synovial differentiation. SS typically presents in young adults (median age: 25 yr). The male-to-female ratio is 3:1. Symptoms are site-specific but tend to be nonspecific. SS typically appears as a solitary, painless mass, occasionally accompanied by dyspnea or hoarseness. The diameter of the lesion ranges from 1 to 12 cm. The cut surface is yellow, firm, whorled, gritty, and friable; cyst formation with hemorrhage or mucoid material is often seen.

Histopathologically, SS is either monophasic (epithelial *or* spindle cells) or biphasic (epithelial *and* spindle cells). The most common pattern is the monophasic spindle cell type, which is characterized by short, streaming fascicles of relatively uniform spindle cells with spindled nuclei and indistinct cell borders (figure 1). In biphasic tumors, the epithelial cells are arranged in solid nests, pseudoglands, or papillary structures (figure 2). Their shapes vary from cuboidal to tall columnar. Mitotic activity is easily identified, but it is not increased. Approximately one-third of all synovial sarcomas exhibit focal calcification. Poorly differentiated tumors are characterized by necrosis and increased mitotic figures. Either epithelial or mesenchymal mucin may be identified in each component of a biphasic tumor. Immunohistochemistry is necessary to confirm a diagnosis of SS. Both spindle and epithelial cells express low- and high-molecular-weight cytokeratins and epi-

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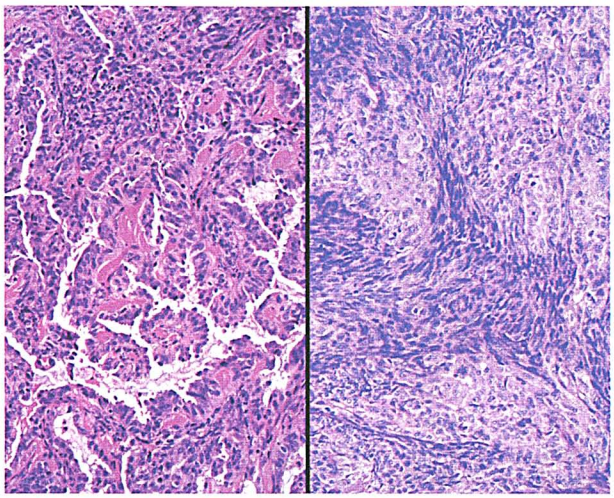

Figure 2. This biphasic SS exhibits areas of gland formation and papillary structures that are traversed by streaming fascicles of smaller, darker spindle cells.

thelial membrane antigen, but only spindle cells express vimentin and bcl-2.

SS has a unique balanced and reciprocal translocation: t(X;18)(p11.2;q11.2). This finding is not seen in other neoplasms. The histologic differential diagnosis is predicated on whether the tumor is biphasic or monophasic; it includes adenocarcinoma, spindle cell carcinoma, fibrosarcoma, malignant peripheral nerve sheath tumor, glomangiopericytoma (hemangiopericytoma), leiomyosarcoma, malignant melanoma, epithelioid sarcoma, and angiosarcoma.

Recurrence rates as high as 25% have been reported, usually within 2 years. Another 25% of cases involve metastasis, usually to the lung, bone, or lymph nodes. The 5-year survival rate is approximately 66%. A more favorable clinical outcome is suggested by a young age at diagnosis, small tumor size (<5 cm), extensive calcification, and complete surgical removal. Therefore, meticulous attention to surgical margins and multimodality therapy (radiation and chemotherapy) will help achieve the best overall outcome.

Suggested reading

Dei Tos AP, Dal Cin P, Sciot R, et al. Synovial sarcoma of the larynx and hypopharynx. Ann Otol Rhinol Laryngol 1998;107:1080-5.

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