Synovial Sarcoma With Rhabdoid Features

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A 13-year-old white girl presented with a soft-tissue mass on the volar aspect of her proximal right forearm of 1-month duration. Magnetic resonance imaging showed a 6-cm soft-tissue mass with no bone involvement. A diagnosis of synovial sarcoma was rendered on needle biopsy. The patient received 6 months of chemotherapy with ifosfamide and doxorubicin hydrochloride with no significant response. An above-elbow amputation showed an ovoid, well-defined, firm mass measuring $4.1 \times 3.1 \times 2.2$ cm in the deep soft tissue. The mass had gray-pink, fleshy, and homogeneous cut surfaces. No areas of hemorrhage, necrosis, or cystic change were noted. Microscopically, the tumor showed features of a typical biphasic synovial sarcoma with well-formed epithelial glands adjacent to malignant spindle cells and numerous mast cells (Figure 1). The spindle cell areas showed extensive sclerosis with ropy, thick, osteoid-like collagen and microcalcifications. Hemangiopericytoma-like vascular areas and focal cystic change were present. A focus of poorly differentiated, briskly mitotic (20 mitoses per 10 high-power fields) cells was noted. A loosely cohesive area of round cells with rhabdoid morphology was observed along with the typical biphasic areas (Figure 1). These rhabdoid cells were polygonal with abundant cytoplasm containing eosinophilic hyaline inclusions or globules and eccentric vesicular nuclei (Fig. 2). The intracytoplasmic inclusions were positive for vimentin, cytokeratin (AE1:AE3), and epithelial membrane antigen (EMA) by immunohistochemistry. Ultrastructurally, the tumor cells showed characteristic paranuclear whorls of intermediate filaments consistent with rhabdoid differentiation (Fig. 3).

Synovial sarcoma is a tumor of predominantly young adults and teenagers. It has a predilection for extremities and tends to occur in the vicinity of large joints, especially in the knee region. Tumors in extremities constitute 83% of all cases, with 60% in lower and 23% in upper extremities. Contrary to its name, it does not involve the joint or show any evidence of differentiation toward synovial cells. Approximately 20% of synovial sarcoma tumors contain poorly differentiated areas.1 Synovial sarcoma with rhabdoid features is rare. Machen et al2 studied 34 cases of synovial sarcoma of the extremities and demonstrated that features associated with metastatic disease included rhabdoid morphology, poorly differentiated areas, grade-3 nu-
clei, mitotic figures greater than 10 per 10 high-power fields, increasing age, tumor size of 5 cm or more, and location in the lower extremity. Thus, the presence of areas of rhabdoid and poor differentiation in the current tumor favors poor prognosis and may be the cause of minimal response to chemotherapy.

The differential diagnosis includes other tumors with rhabdoid features, for example, melanoma, mesothelioma, meningioma, lymphomas, sarcomas (endometrial stromal sarcoma, rhabdomyosarcoma, leiomyosarcoma, myxoid chondrosarcoma, and desmoplastic small round cell tumor), and carcinoma (transitional cell carcinoma, colorectal adenocarcinoma, renal cell carcinoma, Merkel cell carcinoma, and vulvar carcinoma). The mesenchymal areas in synovial sarcoma have characteristic thin, closely packed spindle cells alternating with thick, almost osteoid-like collagen, calcifications, and hemangiopericytoma-like areas that help differentiate it from other sarcomas. Additionally, unlike other sarcomas, tumor cells in synovial sarcoma express cytokeratin and epithelial membrane antigen. In children, it is important to distinguish tumors with rhabdoid cells from pure rhabdoid tumors, both renal and extrarenal. The latter show predominant rhabdoid morphology and lack the clear lines of epithelial or mesenchymal differentiation seen in synovial sarcoma.²

References