Multimodality Management of Soft Tissue Tumors in the Extremity

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KEYWORDS

• Soft tissue sarcoma • Extremity sarcoma • Limbs • Resection • Radiotherapy

KEY POINTS

- Work-up for an extremity mass suspicious for a soft tissue sarcoma includes cross-sectional imaging with an MRI and a core biopsy done in line with the planned incision.
- The standard for treatment of extremity soft tissue sarcomas is limb-sparing surgery with a margin of 1 to 2 cm. Overlying fascial layers (ie, muscular fascia, femoral sheath, periosteum) are often barriers to tumor extension and are acceptable margins when major neurovascular or bony structures are in close proximity.
- Rates of local and distant recurrence vary by histologic subtype. These differences inform surgical margins and the use of chemotherapy and radiation.
- Radiation therapy is used to decrease rates of local recurrence in high-risk tumors. Neoadjuvant (vs adjuvant) radiation can minimize side effects to nearby joints and normal tissues, but is associated with increased rates of wound complications and has equivalent rates of local control.
- Use of adjuvant chemotherapy is controversial. Neoadjuvant chemotherapy should be routinely prescribed for high-risk, chemosensitive subtypes (ie, Ewing sarcoma and rhabdomyosarcoma). It can be selectively prescribed for moderately chemosensitive subtypes based on other risk factors, such as size.

INTRODUCTION

Soft tissue sarcoma (STS) is a term referring to approximately 100 different subtypes of cancer.[1] These diseases are rare, and as a group are diagnosed in only approximately 12,000 patients in the United States each year.[2] Although STS is identified in

Disclosures: The authors have no disclosures to report.
This work was supported by the Memorial Sloan Kettering Cancer Center Core Grant (P30 CA008748) and the Kristen Ann Carr Fund.
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Surg Clin N Am 96 (2016) 977–992
http://dx.doi.org/10.1016/j.suc.2016.05.001 **surgical.theclinics.com**
0039-6109/16/$ – see front matter

any site within the body, 40% are located in the extremities, and multimodality treatment is used to manage patients with localized disease.[3] What combination of surgery, radiation, and systemic treatment is best for a particular patient depends on histologic subtype, which is diagnosed using a combination of cross-sectional imaging, microscopy, and molecular diagnostics. The most common histologies in the extremity are liposarcoma, undifferentiated pleomorphic sarcoma, myxofibrosarcoma, and synovial sarcoma (Fig. 1). Each carries a different risk for distant metastases and local recurrence; for example, dermatofibrosarcoma protuberans (DFSP) carries a higher long-term risk of local recurrence than leiomyosarcoma, but its risk of metastasis is much lower. We present an algorithm for diagnosis and treatment of STS, highlighting modifications that should be made based on the biologic behavior of specific histologic subtypes.

CLINICAL PRESENTATION AND DIAGNOSIS

Most patients eventually diagnosed with an STS present with a painless mass. More than 90% of painless masses are benign lesions, such as lipomas. Therefore, sarcomas are sometimes initially diagnosed as lipomas, resulting in a delay in the correct diagnosis. In general, lipomas tend to be softer, be in a subcutaneous location, have a history of prolonged stability, and be uniformly mobile with no overlying skin changes.

In contrast, STS may be firm, deep, enlarging over time, multifocal, and associated with neovascularization of the overlying skin. Radicular pain or swelling in the distal extremity related to underlying neurovascular involvement may reflect locally advanced disease. A history of nearby trauma may be reported by the patient, but it is unclear that trauma can initiate STS development. More likely, trauma brings attention to a preexisting mass.

IMAGING THE PRIMARY TUMOR

Work-up of an extremity mass begins with cross-sectional imaging for all but the smallest superficial lesions (<2–3 cm), which can be managed with an excisional

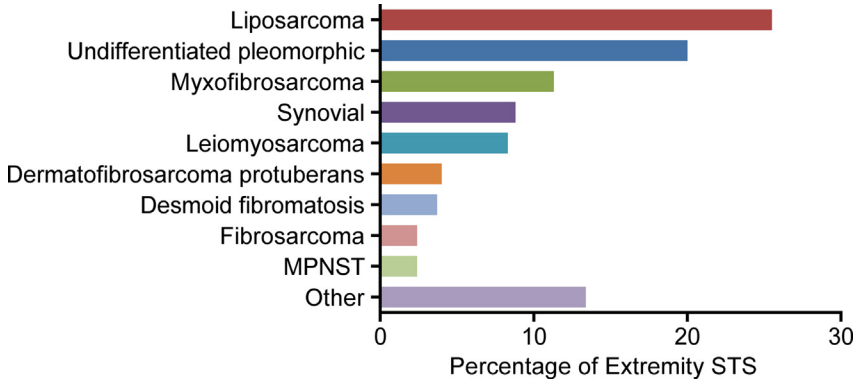

Fig. 1. Histologic distribution of primary extremity soft tissue sarcomas (n = 3103). The data are derived from all surgically resected soft tissue sarcomas followed prospectively at Memorial Sloan-Kettering Cancer Center between 1980 and 2014. Tumors previously designated as malignant fibrous histiocytoma are denoted as undifferentiated pleomorphic sarcoma. Histologic subtypes that represented ≤2% of all cases are grouped as other. MPNST, malignant peripheral nerve sheath tumor.