

# HOSPITAL PHYSICIAN®

## GENERAL SURGERY BOARD REVIEW MANUAL

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## Soft Tissue Sarcoma I: Evaluation

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#### I. INTRODUCTION

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Soft tissue sarcoma (STS) is a malignant tumor that develops from connective tissue. Approximately 6000 new cases of STS are reported per year in the United States. More than 50% of patients with STS will die as a result of their tumor. Although sarcomas can arise in any anatomic site, about 50% of STSs occur in the extremities, 35% in the lower extremity and 15% in the upper extremity<sup>1</sup> (**Figure 1**). The other major areas affected are the viscera, retroperitoneum, and trunk.

In general, the keys to improved survival and limitation of morbidity in patients with STS are early recognition, diagnosis, and precise surgical treatment. The principles of management are based on tumor location, stage, and behavior. Combination surgery and radiation are used for limb and corporeal preservation to limit morbidity and optimize patient survival. Chemotherapy has had limited success and is used for treatment of metastatic disease or as part of a clinical trial.

This article is the first half (Part 1) of a review on STS. Part 1 of this review ("Soft Tissue Sarcoma I: Evaluation") focuses on etiology, pathology, and staging of sarcoma and describes how to initiate evaluation of patients with a soft tissue mass. A case patient is also presented to highlight features of managing STS. Part 2 will describe the basic concepts involved in managing patients with STS, including surgical resection, limb-sparing surgery, adju-

vant therapy, and follow-up; diagnosis and management of retroperitoneal sarcomas are also discussed. This review is not intended to be comprehensive; additional articles are provided in the reference section.

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#### II. ETIOLOGY

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The cause of STS is unknown. However, genetic mutations in mesenchymal stem cells may predispose these cells to develop malignant clones. The alteration in p53 and RB-1 has been seen in several sarcomas.<sup>2,3</sup> A genetic predisposition to STS has been seen in patients with neurofibromatosis (von Recklinghausen's disease),<sup>4,5</sup> Li-Fraumeni syndrome,<sup>6,7</sup> retinoblastoma, and familial polyposis coli.<sup>8</sup>

Cell mutations can result from ionizing radiation. Since 1922, osteogenic sarcoma has been known to result from radiation exposure.<sup>9</sup> Moreover, an increased incidence of malignant fibrous histiocytoma, angiosarcoma, and lymphangiosarcoma appear to occur after external-beam radiation used in cancer treatment.<sup>10</sup>

Chemical exposure has also been reported as a potential causal factor in STS. The chemical agents implicated include vinyl chloride, arsenic, chlorophenols, and phenoxyacetic acids. The role of dioxin (Agent Orange) and pesticides in the development of STS remains unclear.<sup>1</sup>