In this review, the risk factors of an unplanned excision of soft-tissue sarcoma and the implications of non-oncologic resection are discussed. Although soft-tissue sarcoma remains a rare disease, many studies have shown the deleterious effects of unplanned excision, including decreased recurrence-free survival and increased morbidity. Once discovered, sarcomas should be referred to expert centers for further management, which often entails re-excision, radiation, and/or chemotherapy treatment. Although much still needs to be learned about this complex disease, a multidisciplinary approach including surgeons, medical oncologists, radiologists, and pathologists is paramount to its successful treatment.

The differential diagnosis of a soft tissue mass is broad, and an appropriate imaging workup is crucial to accurate identification. Additionally, imaging plays a critical role in soft tissue sarcoma (STS) staging and monitoring for disease progression. In this article, we discuss the different imaging modalities and their utility in the workup and surveillance of STS.

The potentially curative treatment of sarcoma is negative margin wide resection, the clinical tumor with an en bloc margin of surrounding tissue potentially contains microscopic tumor. Planned margins should be 1 to 2 cm but can be less for oncologically equivalent barrier tissues or to preserve an adjacent critical structure. Tumor spillage should be avoided. The role of radiation and/or chemotherapy should be discussed before surgery, as there are potential benefits to preoperative administration. An isolated local recurrence is potentially curable. Amputation is rarely necessary and should only be pursued after other limb salvage treatment options have been considered.
Radiation Therapy for Soft Tissue Sarcoma: Indications, Timing, Benefits, and Consequences
Kilian E. Salerno

Radiation therapy is an integral component of local management with oncologic resection for soft tissue sarcoma. In patients at an increased risk or local recurrence, radiotherapy is indicated to improve local control. Sequencing of radiotherapy and resection should be determined by multidisciplinary input before treatment initiation. For most patients, preoperative delivery of radiation therapy is preferred. In patients initially thought to be at low risk for local recurrence and found to have unexpected adverse pathologic features at resection, postoperative radiation therapy is indicated. The use of radiation therapy for retroperitoneal sarcoma is controversial; when used, preoperative delivery of radiation is recommended.

Plastic Surgery Reconstruction of Sarcoma Resection Defects: Form and Function
Zachary E. Stiles, Robert F. Lohman, and Gary N. Mann

Surgical wide resection is the mainstay of treatment of sarcomas, but the advent of multimodality therapy has improved outcomes and the rates of limb-sparing resection. Often, wounds are unable to be closed primarily and require plastic surgical reconstruction. Following adequate oncologic resection, reconstruction should focus on maintaining functional and esthetic outcomes with minimal postoperative complications. Reconstruction methods range from simple techniques such as skin grafting and local rotational flaps all the way to more complex procedures such as free flaps. The reconstructive surgeon is an integral member of the multidisciplinary team and should be actively involved in treatment planning.

Retroperitoneal Sarcomas: Histology Is Everything
Michael K. Turgeon and Kenneth Cardona

Retroperitoneal sarcomas (RPS) are a rare subset of soft tissue sarcoma that are composed of only a few histologic subtypes, each with a distinct tumor biology, clinical presentation, preferred treatment strategy, recurrence risk, and surveillance plan. In the modern era of precision medicine, our understanding of the implications of subtype tumor biology and anatomic location has led to a more nuanced, histology-specific approach to therapy, including surgery, neoadjuvant radiation therapy, and/or chemotherapy. This article provides a summary of recent updates to the management of RPS.

Sarcoma Pulmonary Metastatic Disease: Still a Chance for Cure
Mark Hennon

Tumors of soft tissue origin are not common but are increasing in incidence. Given the rare and heterogeneous nature of the disease, deciding on an effective treatment approach to the patient can be challenging. Approximately 20-50% of patients with sarcoma will develop metastases to the lung via hematogenous spread. Despite improvements in systemic therapy options for patients with metastatic disease to the lung, surgical resection of metastases is often the preferred option in patients who are safe surgical candidates. Clearance of metastatic disease with surgical
resection has been proven to be cost-effective and can improve chances for long term survival. Deciding on who may benefit from surgical resection is best achieved in a multidisciplinary setting.

Gastrointestinal Stromal Tumors and the General Surgeon 625
Ilaria Caturegli and Chandrajit P. Raut
Gastrointestinal stromal tumors (GISTs) are rare malignancies of the gastrointestinal tract but are the most common sarcoma. This review covers aspects of the care of patients with GIST relevant to surgeons. In particular, management of sub-2 cm GISTs, the utility of neoadjuvant and adjuvant therapy for primary GISTs, and indications for surgery in the setting of metastatic disease are discussed.

Lipoma and Its Doppelganger: The Atypical Lipomatous Tumor/Well-Differentiated Liposarcoma 637
Elliott J. Yee, Camille L. Stewart, Michael R. Clay, and Martin M. McCarter
Lipomatous tumors are among the most common soft tissue lesions encountered by the general surgeon. Shared history and clinical presentation make differentiation between benign lipomas and low-grade liposarcomas a diagnostic dilemma. This article reviews the epidemiology, clinical history, diagnostic workup, management, natural history, and surveillance of benign lipomas and atypical lipomatous tumors/well-differentiated liposarcomas. Although it is important that aggressive, potentially malignant atypical lipomatous tumors and liposarcomas be managed in a multidisciplinary, preferably high-volume setting, it is equally as important for the nonspecialist general surgeon to be familiar with lipoma and its doppelganger—the well-differentiated liposarcoma.

Dermatofibrosarcoma Protuberans: What Is This? 657
Gerardo A. Vitiello, Ann Y. Lee, and Russell S. Berman
Dermatofibrosarcoma protuberans (DFSP) is a rare, locally aggressive dermal-based sarcoma. Metastatic potential is extremely low, primarily in the setting of fibrosarcomatous transformation. DFSP is characterized by a t(17;22) (q22;q13) translocation that results in active PDGFB signaling. Surgical resection with negative margins (typically including the underlying fascia) is the potentially curative treatment. Delayed wound closure should be considered for cases requiring extensive resection or tissue rearrangement. Tyrosine kinase inhibitors, such as imatinib, have shown response rates of 50% to 60% in patients with locally advanced or metastatic disease. Radiation can be useful for residual or recurrent diseases.

The Evolving Management of Desmoid Fibromatosis 667
Katherine Prendergast, Sara Kryeziu, and Aimee M. Crago
Desmoid fibromatosis is a rare disease caused by genetic alterations that activate β-catenin. The tumors were previously treated with aggressive surgeries but do not metastasize and may regress spontaneously. For these reasons, in the absence of symptoms and when growth would not induce significant complications, active observation is considered first-
line therapy. When intervention is required, surgery can be considered based on anatomy and risk of postoperative recurrence, but increasingly nonoperative therapies such as liposomal doxorubicin or sorafenib are prescribed. Cryoablation, chemoembolization, and high-intensity focused ultrasound can also be used to obtain local control in selected patients.

Benign Neurogenic Tumors

Jeffrey M. Farma, Andrea S. Porpiglia, and Elaine T. Vo

Neurogenic tumors arise from cells of the nervous system. These tumors can be found anywhere along the distribution of the sympathetic and parasympathetic nervous system and are categorized based on cell of origin: ganglion cell, paraganglion cell, and nerve sheath cells. Ganglion cell-derived tumors include neuroblastomas, ganglioneuroblastomas, and ganglioneuromas. Paraganglion cell-derived tumors include paragangliomas and pheochromocytomas. Nerve sheath cell-derived tumors include schwannomas (neurilemmomas), neurofibromas, and neurofibromatosis. Most of these are benign; however, they can cause local compressive symptoms. Surgery is the mainstay of treatment, if clinically indicated. Nonetheless, a thorough preoperative workup is essential, especially for catecholamine-secreting tumors.