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Sarcomas are a heterogeneous group of tumors that may have many etiologies. The incidence of histologic subtypes differs significantly between children and adults. The increase in incidence may be due to improved registry systems, diagnostic tools, and pathologic definitions. Environmental causes may contribute to increased incidence. Genetic alternations may play a role in sarcoma development. As a result of rapidly evolving genomic and proteomic technologies, increased knowledge of the oncogenic mechanisms underlying sarcomagenesis is being generated. Understanding the mechanisms involved in sarcomagenesis is rudimentary. Insight into the molecular basis of sarcoma inception, proliferation, and dissemination hopefully will lead to more effective therapies.

Classification and Pathology	483
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Soft tissue tumors are a heterogeneous group of benign and malignant processes. Some are assumed to be reactive; others are clearly neoplastic. Because of their rarity, they frequently pose diagnostic problems for surgical pathologists. Accurate diagnosis

of these tumors is enhanced by knowledge of the clinical features of the given lesions and, at times, by application of immunohistochemical and molecular techniques. In this article the lesions are described essentially in accordance with the World Health Organization classification.

Advanced Modalities for the Imaging of Sarcoma

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Dalia Fadul and Laura M. Fayad

There is a diversity of modalities available for the imaging of soft tissue and skeletal sarcomas. However, conventional radiography remains the first line imaging modality in the diagnostic work-up, as it provides superior spatial resolution for the evaluation of bone trabecular detail. This article focuses on the advanced imaging modalities of computer tomography, magnetic resonance imaging, and positron emission tomography in the evaluation of sarcomas.

Management of Extremity Soft Tissue Sarcomas

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Matthew T. Hueman, Katherine Thornton, Joseph M. Herman, and Nita Ahuja

This article provides an understanding of the evaluation, staging, and management of patients with extremity soft tissue sarcoma. Although there are straightforward guidelines to the management of patients with extremity soft tissue sarcoma, each patient presents with a unique tumor, and considerations for tumor control, functional outcome, and the toxicity of therapy must be considered. As is true for patients diagnosed with sarcoma at other anatomic sites, a multidisciplinary team approach streamlines care with attention to the complexities and intricacies of choosing and delivering optimal therapy.

Primary Breast Sarcoma

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Ying Wei Lum and Lisa Jacobs

Primary breast sarcomas are rare breast malignancies. This article describes the clinical management of the disease, including the controversies surrounding the pathologic classification, role of surgery, and adjuvant therapy.

Truncal Sarcomas and Abdominal Desmoids

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Jacqueline M. Garonzik Wang, and Steven D. Leach

This article gives a brief overview of the common histologic subtypes seen in truncal sarcomas and discusses fundamental diagnostic and treatment principles. It also provides a general review of abdominal desmoids. For both truncal sarcomas and desmoids, recurrence rates are high, and definitive

recommendations regarding optimal treating are lacking. A multidisciplinary approach to these entities therefore is critical to select appropriate therapeutic strategies for individual patients.

Management of Retroperitoneal Sarcomas

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Matthew T. Hueman, Joseph M. Herman, and Nita Ahuja

Retroperitoneal sarcomas present a therapeutic challenge based on their location, extent of invasion at diagnosis, and propensity for local recurrence. Surgical therapy remains the only potentially curative treatment option; however, even with aggressive surgical approaches, local recurrence remains a common type of failure. For patients who have high-grade lesions, distant metastatic disease may also limit survival. Optimizing disease control while minimizing the morbidity of therapy remains the primary goal of management. In this article, the authors describe the presentation, evaluation, and management of patients who have retroperitoneal sarcoma.

Management of Gastrointestinal Stromal Tumors

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Matthew T. Hueman and Richard D. Schulick

A gastrointestinal stromal tumor (GIST) is a rare mesenchymal malignancy of the gastrointestinal (GI) tract. Malignant GISTs were first defined as a separate entity from a collection of nonepithelial malignancies of the GI tract in the 1980s and 1990s based on pathologic and clinical behavior. The discovery of activating KIT mutations as a near-uniform occurrence in these tumors greatly influenced the classification [1] and revolutionized therapeutic management of these tumors. To meet the next challenges, newer tyrosine kinase inhibitors and targeted agents are being developed with the goal of providing improved response rates or alternative therapies for patients progressing on established agents. In this article, the authors describe the management of GISTs, concentrating on surgical management and targeted therapies.

Pediatric Soft Tissue Sarcomas

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David M. Loeb, Katherine Thornton, and Ori Shokek

Soft tissue sarcomas in children are rare. Approximately 850 to 900 children and adolescents are diagnosed each year with rhabdomyosarcoma (RMS) or a non-RMS soft tissue sarcoma (NRSTS). RMS is more common in children 14 years old and younger and NRSTS in adolescents and young adults. Infants get NRSTS, but their tumors constitute a distinctive set of histologies. Surgery is a major therapeutic modality and radiation plays a role. RMS is treated with adjuvant chemotherapy, whereas chemotherapy is reserved for the NRSTS that are high grade or unresectable. This review discusses the etiology, biology, and treatment of pediatric soft tissue sarcomas.

The Role of Radiation Therapy in the Management of Sarcomas 629
Aradhana Kaushal and Deborah Citrin

Sarcomas represent a heterogeneous, challenging, and rare group of tumors that present many management challenges. In this article, the authors concentrate on the radiotherapeutic management of sarcomas occurring in the most common locations: the extremities, the trunk, and the retroperitoneum. An overview of the current radiotherapeutic management of soft tissue sarcoma is presented in addition to a discussion of how surgical management may affect radiotherapeutic management. Finally, the authors describe current controversies surrounding the appropriate management of sarcomas with radiotherapy and describe ongoing studies and future areas of research.

Chemotherapeutic Management of Soft Tissue Sarcoma 647
Katherine Thornton

Soft tissue sarcomas are a heterogeneous group of connective tissue tumors, with more than 50 different subtypes. Given the heterogeneity, and the relative small numbers of patients, performing large adequately powered clinical trials in which one can glean any overall broad treatment decisions based on outcome is difficult at best. There is controversy on which chemotherapeutic agents to use in the adjuvant and metastatic settings, or even if to use chemotherapy in the adjuvant setting. In the metastatic setting, doxorubicin and ifosfamide have remained the standards of care for more than 20 years. This review discusses the data on chemotherapy for treatment of metastatic sarcomas and the utility of chemotherapy in the adjuvant and neoadjuvant settings. In addition, the utility of newer biologic agents in the treatment for sarcomas is considered.

Multidisciplinary Management of Metastatic Sarcoma 661
Katherine Thornton, Catherine E. Pesce, and Michael A. Choti

Soft tissue sarcomas comprise a heterogeneous group of malignancies of mesenchymal origin. Although sarcomas can arise virtually anywhere, the most common primary site is the extremity. The development of metastatic disease poses a major clinical problem because it is seldom amenable to a curative treatment. However, with careful and expert multidisciplinary team selection of patients with metastatic sarcoma—balancing probability of benefit with certain toxicity—a combined multimodality approach may provide hope to a select few for prolonged survival and even cure.

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