Retroperitoneal Sarcomas

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KEYWORDS

• Retroperitoneal sarcoma • Retroperitoneal liposarcoma • Radiation • Chemotherapy • Surgery

KEY POINTS

- Retroperitoneal sarcomas are a rare disease, and 5-year survival rates vary from 12% to 70%.
- Although surgery represents the mainstay of treatment, the emphasis is on extended resections to achieve negative margins.
- In the main, patients eventually succumb to metastatic disease, so, in order to treat these patients better, there needs to be better adoption of multimodality therapies both in the neoadjuvant and adjuvant settings.
- Because this disease is rare, multi-institutional collaborations are the only way advances can be made.

INTRODUCTION

Retroperitoneal sarcomas are rare tumors and make up a small subset of all sarcomas; approximately 15%. They tend to grow and can be sizable, and thus patients can present with very large tumors. Most patients present with an abdominal mass and abdominal pain, although most symptoms are nonspecific. The most common subtypes are liposarcoma and leiomyosarcoma. Clinicians should also consider other primary retroperitoneal tumors, including lymphomas, germ cell tumors, and testicular cancer, as a differential diagnosis. The role for surgery in the treatment of retroperitoneal sarcomas remains the gold standard compared with other modalities of therapy. Chemotherapy has limited efficacy, and radiation can be limited by toxicity to adjacent intra-abdominal structures.[1,2] The challenge is often that multivisceral resections are needed to clear disease. The authors advocate patients should be managed at a center specializing in a multimodality treatment, given poor survival rates caused by difficulty with resectability and high recurrence rates.[3]

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Most patients present with an abdominal mass, early satiety, increase in abdominal girth, or abdominal pain.[4] Evaluation should include a complete history and physical examination with abdominal imaging (either computed tomography [CT] or MRI) to determine the extent of disease. Chest imaging should be included to complete staging. Approximately 11% of patients present with metastatic disease.[1,4] Staging of retroperitoneal sarcomas is based on the American Joint Committee on Cancer staging system. The authors generally advocate preoperative core biopsy of the lesion. If imaging is consistent with a large lipoma versus well-differentiated liposarcoma, we generally defer biopsy and plan for surgical resection (**Figs. 1** and **2**). Gene expression analysis is being used by pathologists to help distinguish liposarcoma subtypes; murine double minute-2 (MDM2) is overexpressed in liposarcomas.[5] Preoperative biopsy allows for discussion and consideration for a clinical trial or for neoadjuvant therapy. Patients with retroperitoneal sarcomas should be discussed at a multidisciplinary sarcoma tumor board.

SURGERY

Surgery is the mainstay of treatment of retroperitoneal sarcomas and is the only potentially curative treatment. As for many different types of cancers, the ability to remove tumors with negative margins is one of the most important prognostic factors relating to survival. Singer and colleagues[6] confirmed that incomplete resections and contiguous organ resections were independent prognostic factors for survival. Unique histologic subtype also played a role in prognosis. Dedifferentiated histologic subtypes and the need for contiguous organ resection were associated with an increased risk for local and distant recurrence.[6] The practice of multivisceral resections in order to obtain negative margins is imbedded in surgical teachings; however, what extent of surgical resection is considered too much?

Complete resection is the most important prognostic factor for prolonged survival. There is noted to be a high rate of recurrence up to 48% of patients and complete excision occurs less than 70% of the time.[7,8] Multivisceral en bloc resection may be necessary in up to 83% of patients in order to achieve negative margins.[9] The kidney, colon, adrenal, pancreas, bladder, and spleen are the most common organs resected.[1,10]

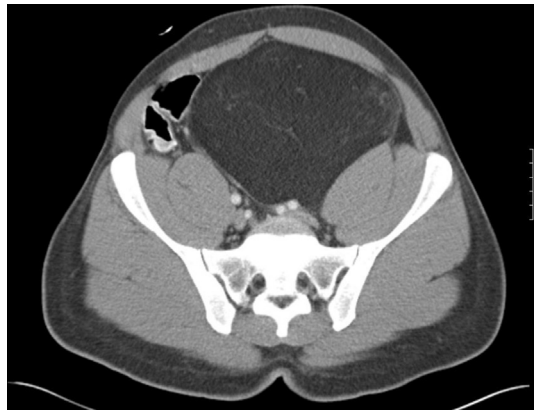

Fig. 1. CT of the abdomen showing a well-differentiated retroperitoneal liposarcoma.