Surgical Treatment of Retroperitoneal Leiomyosarcoma With Adjuvant Radiotherapy

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INTRODUCTION

Although leiomyosarcoma is among the most common nonepithelial retroperitoneal tumors along with fibrosarcoma, liposarcoma, and malignant lymphoma, it only constitutes 5% to 15% of all retroperitoneal tumors. About 70% of leiomyosarcomas are found in the retroperitoneal space.[1] Extensive surgical resection for achieving complete removal and providing negative surgical margins is the treatment of choice. This is also the most important factor in preventing local recurrence.[2-4] We present 2 cases with retroperitoneal leiomyosarcomas invading the inferior vena cava and the ureter. Preoperative workup was made for complete resection and achieving negative margins. Adjuvant radiotherapy was administered following surgical therapy to decrease the risk of local recurrence.

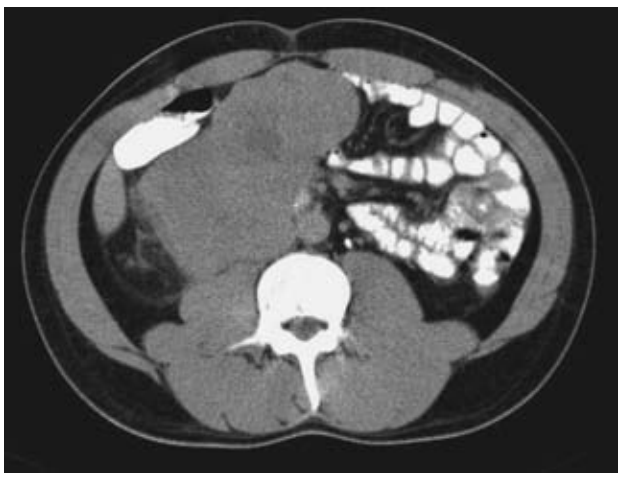

Figure 1. Preoperative CT showing large retroperitoneal mass on the right side in case 1.

CASE REPORT

Case 1

A 31-year-old man presented with abdominal discomfort and weight loss. Physical examination revealed a palpable right upper quadrant mass and a 12-cm retroperitoneal mass was confirmed by computed tomography (CT). The lesion was surrounding the anterior part of the inferior vena cava (IVC; Figure 1). The right pelvicalyceal system and the proximal ureter were dilated and the testicles were normal. Testis tumor markers (β-human chorionic gonadotropin and α-fetoprotein) were measured which were in their reference ranges. The CT-guided biopsy revealed no malignancy and probable diagnosis was schwannoma. Surgical exploration was performed and the mass invading the mesothelium of the ascending colon, ureter, and the IVC was detected. Dissection of the IVC was performed by a vascular surgeon and the mass was removed with a patch of the IVC. Intraoperative frozen section analysis revealed no malignancy, but the final histopathology examination confirmed a grade 2 leiomyosarcoma with negative surgical margins (Figure 2). Thorax CT scan showed no abnormality. Adjuvant radiotherapy was administered with 50.4 Gy/28 fractions.

Twelve months later, a 2-cm spot of liver recurrence was detected and then resected. Subsequently, chemotherapy

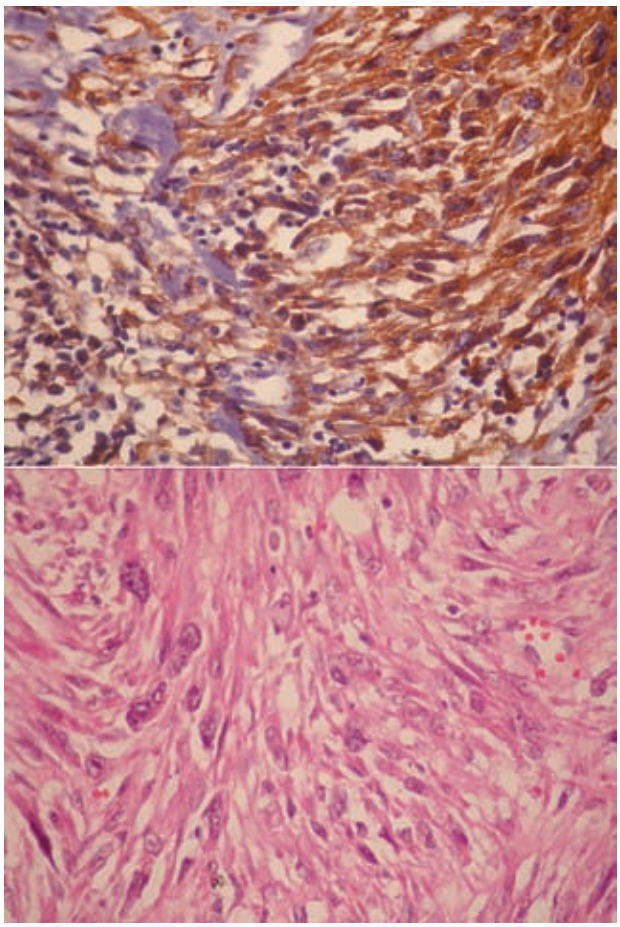

Figure 2. Top, Tumor cells were revealed by positive staining with smooth muscle actin in case 1 (smooth muscle actin, × 400). **Bottom,** Cellular pleomorphism and cellularity of the tumor in case 1 (hematoxylin-eosin, × 400).

with ifosfamide, adriamycin, and dacarbazine was instituted. The patient was without any new recurrences during the 24 months' follow-up.

Case 2

A 52-year-old woman presented to our center with the chief complaint of right lumbar pain and nausea. Tenderness of the right costovertebral angle was detected on physical examination. On CT scan, we detected a 30-mm mass in the right retroperitoneal area anterolateral to the IVC. The mass was compressing the ureter causing mild hydronephrosis (Figure 3). The result of thorax CT was normal. Following dissection of the stented ureter, the mass was removed.

Histopathology examination revealed high-grade leiomyosarcoma with negative surgical margins (Figure 4). Adjuvant radiotherapy was administered

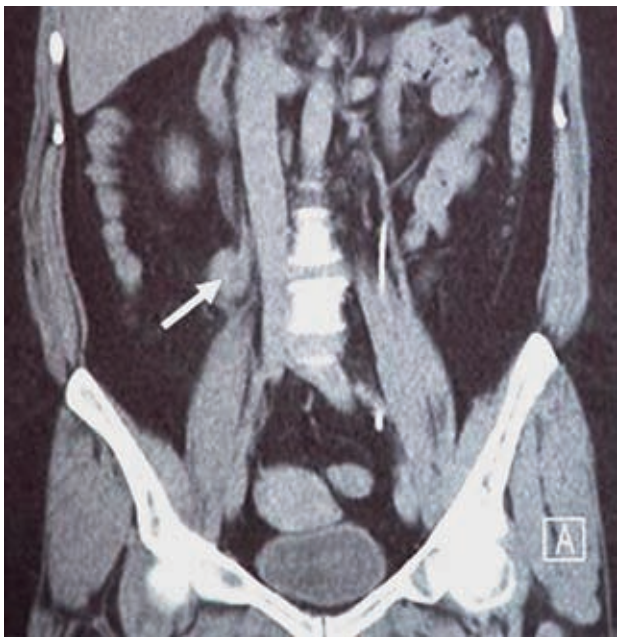

Figure 3. Preoperative CT showing retroperitoneal mass on the right side in case 2 (coronal view).

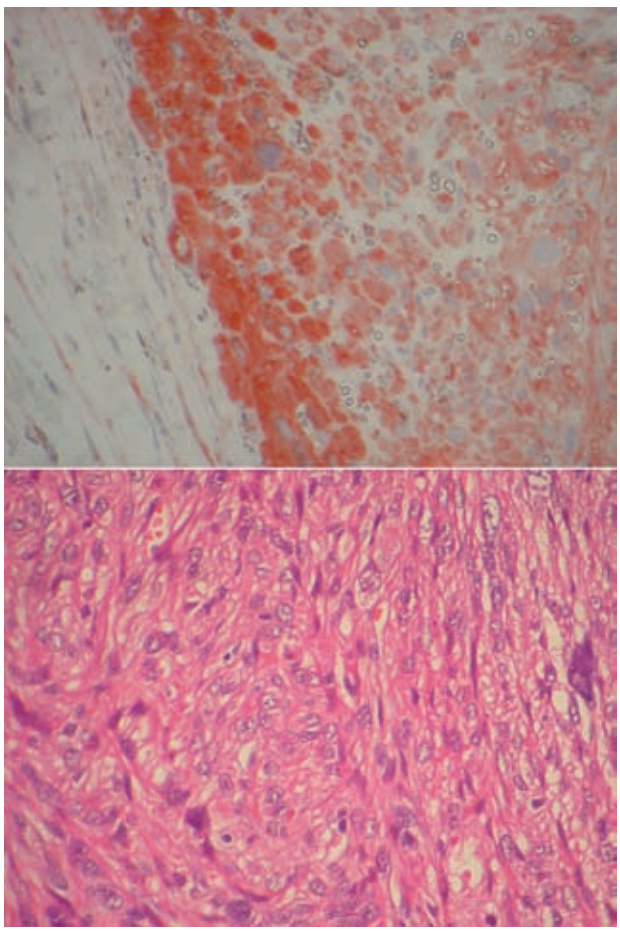

Figure 4. Top, Immunoreactivity of the tumor with smooth muscle actin in case 2 (smooth muscle actin, × 400). **Bottom,** Cellular pleomorphism and cellularity of the tumor in case 2 (hematoxylin-eosin, × 400).

with 50 Gy/25 fractions. No recurrence was detected during the 32 months' follow-up.

DISCUSSION

Sarcomas of the retroperitoneum and the urogenital tract grow slowly and typically remain asymptomatic until the tumor becomes evident as a large mass.[1] Imaging techniques, notably magnetic resonance imaging and CT, have improved permitting excellent visualization, evaluation, and preparation.[5] Preoperative histopathologic diagnosis of a retroperitoneal mass can help planning the operative procedure. However, interpretation of a retroperitoneal mass using needle biopsies may be difficult and inconclusive, as it was in our first case. Guz and colleagues published their experience with retroperitoneal neural sheath tumors. They performed preoperative CT-guided needle biopsies in 3 patients and all yielded inaccurate or inconclusive results.[6]

Complete resection often needs extended dissection which may include the vascular structures, kidneys, bladder, and gastrointestinal tract.[4,7] Management can need to be provided by a specialized team of surgeons. In case 1, although needle biopsy result was schwannoma, IVC dissection was made by a vascular surgeon to achieve negative surgical margins. In addition to incomplete resection, tumor grade is another prognostic factor predicting local recurrence and metastasis.[2,3] Different studies report recurrence rates ranging from 45% to 82% after complete resection.[7,8]

Extirpating the retroperitoneal leiomyosarcomas with negative margins cannot be accomplished due to some limitations on dissection which result in high incidence of local recurrence. It has been stated in many studies that in most patients, disease recurs locally within 3 years of definitive treatment of the primary tumor.[7] Some series have reported that approximately 80% to 87% of all local recurrences become evident within 2 years and 100% are detected within 3 years.[7,9] In a series of sarcomas of the retroperitoneum and the urogenital tract, local relapse was detected in 83% of the patients within 3 years.[7] Patients with retroperitoneal sarcomas should be followed closely especially for the first 3 years after the primary treatment. High incidence of the local recurrence remains as a major problem in long-term and follow-up, perhaps as long as 10 years, is mandatory.[10]

Radiation therapy seems to be favorable for local control. In a series of retroperitoneal sarcomas, 13 of 34 patients received high-dose postoperative radiotherapy which was found to have a significant favorable effect on recurrence.[11] The employment and dosage of radiotherapy have not been standardized and differ considerably in the literature.[4,12,13] In a study about leiomyosarcoma of the IVC, routine administration of postoperative radiotherapy with 45 Gy to 50 Gy was reported to be effective.[12] To decrease the risk of local recurrence, our patients underwent adjuvant radiotherapy with 50 Gy. Although follow-up was short, none of our patients had local relapse after 24 and 32 months.

Information on the value of chemotherapy is scarce. There are investigational studies concerning combined-modality therapy for retroperitoneal sarcomas.[4] In a case report, beneficial effect of neoadjuvant chemotherapy for complete resection of advanced leiomyosarcoma was depicted.[14] However, no study or report has offered a consistent advantage of chemotherapy.[4,12]

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