

Retroperitoneal liposarcoma: A case report

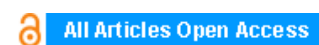
**M Sandra Jacob, Shirali Patel, Harvey Sasken, Yomayra Perez,
Valerie Katz, Mark Ingram**

ABSTRACT

Introduction: We report an interesting case of a 67-year-old female presented with symptomatic cholelithiasis and was found to have an occult retroperitoneal sarcoma on work up.

Case Report: A 67-year-old female was referred to the surgery clinic by the gynecology service for symptomatic cholelithiasis. On examination, she was moderately obese with mild right upper quadrant tenderness and a reducible incisional hernia. An abdominal ultrasound revealed cholelithiasis and a left retroperitoneal flank mass. She was referred for abdominal computed tomography scan and magnetic resonance imaging scan which revealed a large left retroperitoneal cystic mass adherent to the left kidney. She underwent en-bloc resection of retroperitoneal tumor, cholecystectomy, and repair of incisional hernia. Her postoperative course was uneventful and she continues to do well without adjuvant chemoradiation. **Discussion:** One-third of malignant tumors located in the retroperitoneum are sarcomas. The median age of presentation occurs in the sixth decade. As with our patient complete surgical resection is the optimal treatment for patients. The addition of adjuvant radiation therapy to surgical resection is associated with both a reduced risk of local recurrence and a longer recurrence-free interval, but it does not improve overall survival.

Conclusion: The review of the literature emphasizes that the management of retroperitoneal sarcomas consists of complete resection of the tumor with adjuvant radiotherapy (if the tumor is high grade) combined with surveillance for early liposarcoma detection of recurrence or metastases.



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Keywords: Sarcoma, Liposarcoma, Retroperitoneal tumor

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INTRODUCTION

Soft-tissue sarcomas are relatively rare with approximately 8,600 new cases annually and represent less than 1% of all newly diagnosed malignancies in the United States. Retroperitoneal sarcomas are malignant tumors arising from mesenchymal cells, which are usually located in muscle, fat, and connective tissues. One-third of malignant tumors located in the retroperitoneum are sarcomas, and approximately 15% of soft tissue sarcomas arise in the retroperitoneum [1]. According to the World Health Organization (WHO), soft-tissue liposarcomas are categorized into five distinct histological subtypes: well-differentiated, dedifferentiated, myxoid, pleomorphic and mixed type. Retroperitoneal sarcomas have varying clinical courses depending on their histological subtype and grade [1, 2].

The pathologic diagnosis of liposarcoma rests on the identification of lipoblasts in a milieu of supporting

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histomorphologic features. The well-differentiated liposarcoma is a low-grade neoplasm which can present as five histological variants: lipoma-like, sclerosing, inflammatory, spindle cell and liposarcoma with meningothelial whorls. The treatment of choice is complete surgical excision. According to Stoeckle et al., there are no survival benefits of adding adjuvant radiotherapy at this time for a resected well-differentiated retroperitoneal liposarcoma [3].

CASE REPORT

A 67-year-old female was presented to the gynecologist for screening Pap smear. The patient complained at that time of right upper quadrant abdominal pain. Her past medical history was significant for hypertension, asthma, and hyperlipidemia which were well-controlled and there was no significant family history. She had a previous midline scar from a total abdominal hysterectomy with bilateral salpingo-oophorectomy for fibroid. On examination, her abdomen was very obese (BMI 44.8) with right upper quadrant tenderness. No mass was palpated and she had a reducible incisional hernia. Abdominal ultrasound revealed a large heterogeneous left flank mass and cholelithiasis. Origin of the mass was uncertain, computed tomography (CT) scan was recommended for further assessment. She was subsequently referred to surgery for management and imaging studies. The CT scan of abdomen showed a large retroperitoneum mass with displacement of the retroperitoneal organs (Figure 1A–B). The origin and blood supply of the mass could not be determined on the CT scan and magnetic resonance imaging (MRI) scan was recommended, revealing a large complex retroperitoneal cystic mass adherent to the left kidney which extended from the splenic hilum inferiorly to the left lower abdomen (Figure 2).

The case was presented at the multidisciplinary tumor board. The recommendation was to proceed with surgery first. Neoadjuvant chemotherapy was not recommended as there was no tissue diagnosis. After discussion with the patient and her family, she underwent an exploratory laparotomy with en-bloc resection of the retroperitoneal tumor. At surgery, there was a left retroperitoneal mass as per, adherent to the left kidney, but separate from the spleen, pancreas, and colon. The mass was resected en-bloc with the kidney. The gallbladder was removed for chronic cholelithiasis which was symptomatic and her incisional hernia was repaired. Her postoperative course was uneventful.

The mass was well circumscribed and globular composed of fleshy homogeneous yellow tan tissue. The tumor weighed 1670 grams and measured 30x25x15 cm (Figure 3). Routine tissue stain demonstrated a well-differentiated liposarcoma, characterized by its hypocellularity, nuclear pleomorphic atypia and delicate vascularity. The tumor was composed of myxoid stroma with increase

microvasculature and atypical adipocytes. Some of these adipocytes have a single vacuole and others demonstrated a floret giant cell configuration (Figure 4). An atypical lipoblast was demonstrated in (Figure 5). This large cell had an enlarged nucleus, irregular in shape, with variably clumped chromatin and the cytoplasm contained numerous vacuoles.

The stroma was variably fibrillar with areas of abundant ground substance. A mild inflammatory infiltrate was present with a significant quantity of plasma cells. Final pathology revealed the tumor to be a low grade, well-differentiated, stage T2bG1NoMo retroperitoneal liposarcoma. The medical and radiation oncologists suggested observation and interval follow-up for surveillance.

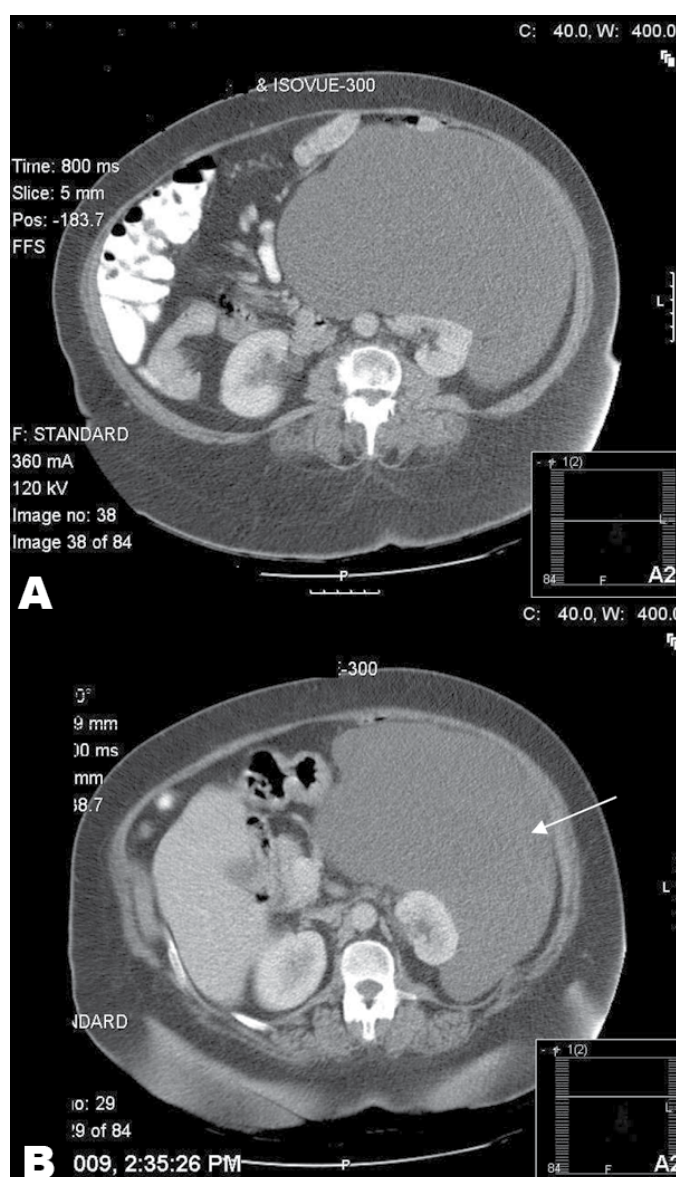


Figure 1: (A, B) Computed tomography scan showing a large retroperitoneal mass adherent to the left kidney.

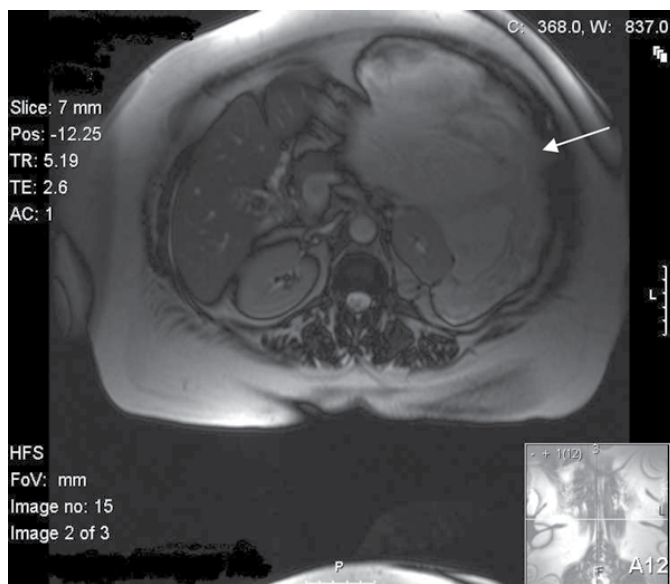


Figure 2: Magnetic resonance imaging scan showing a large retroperitoneal mass adherent to the left kidney.

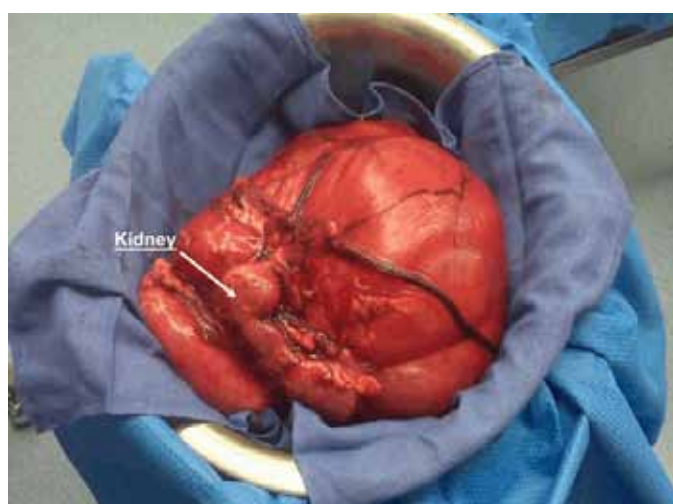


Figure 3: Retroperitoneal tumor with left kidney, globular, well-circumscribed mass which is covered by a smooth surface with displayed prominent vasculature.

DISCUSSION

Retroperitoneal tumors are an extremely heterogeneous group of neoplasms, 85% of which are malignant. Liposarcomas constitute between 45–55% of retroperitoneal masses [4]. Age at presentation is younger compared with most other malignancies, with many being diagnosed

Between 54–65 years of age [5]. There is an equal male/female ratio [1]. The distribution of soft tissue sarcomas by anatomic site can be found in an article by Lawrence et al. [6].

Retroperitoneal sarcomas present 80% of the time as an asymptomatic abdominal mass. Symptoms can

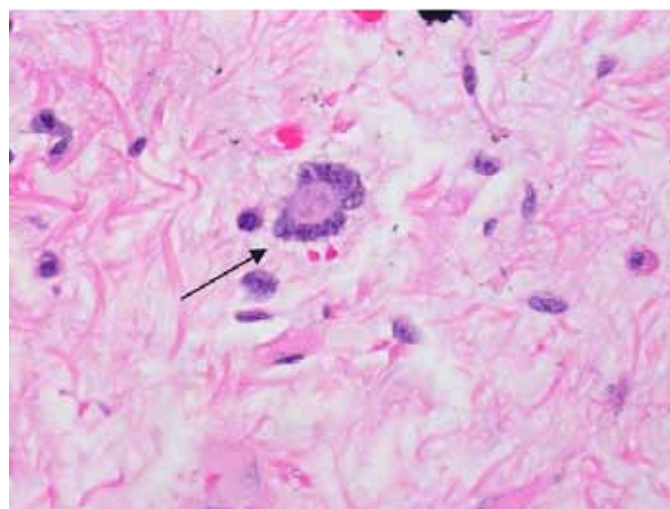


Figure 4: Photomicrograph showing a floret giant cell configuration.

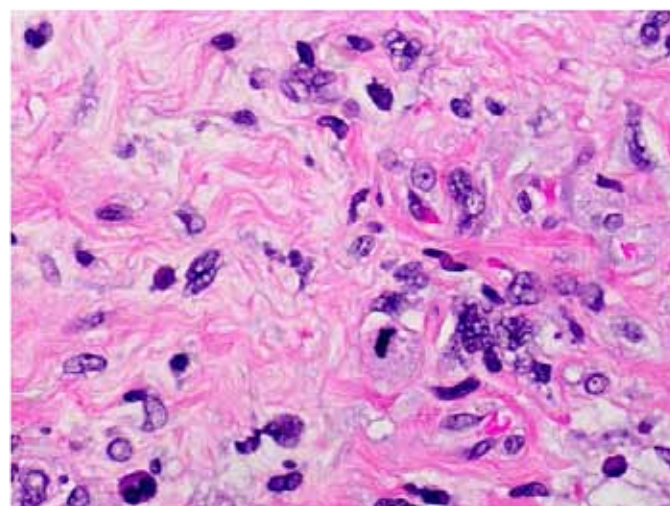


Figure 5: Photomicrograph showing atypical lipoblasts present with occasional mitoses within a mixture of myxoid and fibrillar stroma.

also be related to mass effect or local invasion which may lead to pain, gastrointestinal obstruction, feelings of early satiety, and weight loss. In addition, neurologic and muscular skeletal symptoms are referred to the lower extremities [7].

Histopathologic variety is the main prognostic factor. Five histologic types are recognized. Well differentiated liposarcoma represents around 30% like our case and has the best prognosis. The myxoid type is the most frequent liposarcoma, constituting around 50% of all tumors. It has a less favorable progression, as it often recurs early. The pleomorphic, round cell and undifferentiated types display the worst prognosis [4].

After a physical examination CT scan provides an excellent understanding of the relationship between nearby structures and is critical to preoperative planning. A patient presenting with a palpable abdominal mass,

should be have a high-resolution, thin-cut CT scan with intravenous and oral contrast since these images allow for further distinction between intra-abdominal and retroperitoneal structures. This allows a discussion of the need for biopsy if indicated, the operative plan, and the preparedness of the operative team, as well as a discussion with the patient regarding the risks and benefits. The differential diagnosis includes a primary neoplasm arising from a retroperitoneal visceral structure (e.g., pancreas, adrenal glands, kidneys, and duodenum), a retroperitoneal sarcoma, a lymphoma, or a metastatic lesion [5].

The optimal treatment for patients with localized, resectable retroperitoneal sarcomas is surgery with gross and microscopically negative margins. Complete surgical resection frequently requires en-bloc resection of adjacent viscera [8]. The kidney was the most frequently resected organ (36%) followed by segmental resection of the large bowel, spleen, and pancreas [9].

The addition of adjuvant radiation therapy to surgical resection is associated with both a reduced risk of local recurrence and a longer recurrence-free interval. However, it does not improve overall survival. Studies have demonstrated the advantages of preoperative radiotherapy in the management of marginally resectable retroperitoneal sarcomas. The benefits of pre-operative radiation are multiple [3, 10]. It allows for the gross tumor volume to be readily definable for accurate treatment planning. Moreover, the tumor displaces radiosensitive viscera. Thus, no adhesions and tethering of bowel to the tumor bed can occur and the tumor is treated in situ.

Another treatment modality is intra-operative radiotherapy (IORT) which is targeted to a specific region allowing for maximum doses of radiation to the tumor bed. Studies show that IORT improves tumor control in the field. However, it does not influence recurrence-free or overall survival rates [9, 10].

CONCLUSION

The review of the literature emphasizes that the management of retroperitoneal sarcomas consists of complete resection of the tumor followed by adjuvant radiotherapy reduce local recurrence but does not affect overall survival and combined with surveillance for early detection of recurrence or metastases. Imaging studies are essential for proper preoperative planning and allow assessment of respectability prior surgery; preoperative radiotherapy can be considered in patients with questionably resectable tumors. Contrast-enhanced computed tomography scan and magnetic resonance imaging were valuable aids in our case. The patient should be closely followed with regular physical examinations and imaging studies such as chest X-rays and computed tomography scans. Our patient continues to follow-up for surveillance and is doing well.

Author Contributions

M Sandra Jacob – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Shirali Patel – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Harvey Sasken – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Yomayra Perez – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Valerie Katz – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Mark Ingram – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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