Huge retroperitoneal liposarcoma with renal involvement requires nephrectomy: A case report and literature review

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Abstract. A 60-year-old female visited Guihang Guiyang Hospital (Guiyang, China). She presented with abdominal pain in the right side for the previous 2-months, with a touchable mass identified for the previous 1-month. Computed tomography with magnetic resonance imaging revealed a huge mass in the right abdomen. The diagnosis of well-differentiated retroperitoneal liposarcoma with renal involvement was made. During surgery, the tumor was removed, including the fatty renal capsule; however, the kidney was preserved. It is currently debatable whether resection of adjacent organs is required to obtain the negative margins. Conventional viewpoints advise that multi-organ resection is required in order to obtain the negative-margin. However, even if an R0 resection is achieved, the local recurrence rate remains markedly high. Additionally, the complications of organ resection have more impact on patients. Radiotherapy and chemotherapy are an important adjuvant method for these patients. In conclusion, retroperitoneal liposarcoma is a rare disease with a high rate of recurrence. Complete resection is the predominant treatment; however, combined resection of adjacent organs must be considered.

Introduction

Retroperitoneal liposarcoma is a rare malignant tumor with an incidence of 2.5/1,000,000 individuals (1). Although the incidence rate is very low, early diagnosis is often difficult since symptoms appear only when the tumor becomes very large. Retroperitoneal liposarcoma is classified into five subtypes, according to the World Health Organization Classification: i) Well-differentiated; ii) dedifferentiated; iii) myxoid cell; iv) pleomorphic; v) mixed-type (2). The kidney is the most frequently invaded organ. Surgical resection is the predominant treatment method for this disease, and negative margins must be achieved to improve survival rate (1,3,4). It remains controversial as to whether resection of adjacent organs extends the 5- or 10-year survival rate. Additionally, the 10-year survival rate is not prolonged following multi-organ resection (5-8). Considering the complications of multi-organ resection, whether nephrectomy or other organ resection is required remains to be elucidated. The present study reported a case with retroperitoneal liposarcoma managed at Guihang Guiyang Hospital (Guiyang, China), and also performed a literature review on presentation, management and prognosis of this patient.

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Case report

A 60-year-old female presented to Guihang Guiyang Hospital with abdominal distension in the right side for the previous 2-months, with a touchable mass identified for the previous 1-month. The clinical symptom was just abdominal distension and no significant history relevant to this case was determined. The physical examination indicated a 10x15 cm flexible mass located in right side of the stomach. This mass lacked tenderness and exhibited an unclear boundary. The laboratory examinations, including assessment of tumor markers, routine blood, urine and stool tests, revealed no clear abnormalities. Computed tomography (Fig. 1) and magnetic resonance imaging (Fig. 2) demonstrated a huge mass in the right abdomen. The diagnosis was well-differentiated retroperitoneal liposarcoma. No evidence was found to support distant metastasis, with the exception that it is possible that the tumor had invaded the kidney.

During surgery, it was revealed that the mass originated from the right fatty renal capsule and the kidney was covered by the carcinoma. The patient received complete resection of the liposarcoma and the right kidney was preserved. The carcinoma covering the kidney was removed. Renal fascia was intact and no sign of invasion was visible. The renal periphery was completely bare, including the renal portal, the ureter and the inferior vena cava. The total mass was 20x15x10 cm in size (Figs. 3 and 4).

Hematoxylin and eosin staining of tissue shows that retroperitoneal well-differentiated liposarcoma, with negative margins, full of lipocytes. It was confirmed that the carcinoma had invaded the fatty renal capsule (Fig. 5). Immunohistochemical analysis was performed at the Department of Pathology, Guihang Guiyang Hospital. The

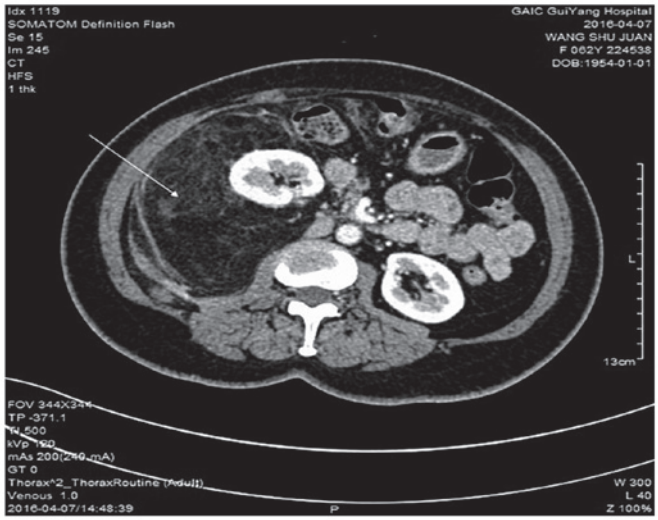

Figure 1. Contrast-enhanced computed tomography image. The images demonstrated a giant mass in the right abdomen. This was highlighted by the right kidney moving forward.

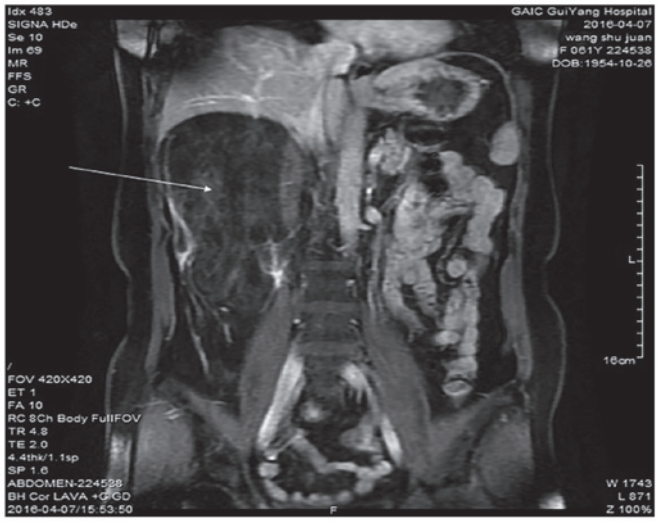

Figure 2. Nuclear magnetic resonance imaging of the patient. The image demonstrated a huge mass in the retroperitoneal space.

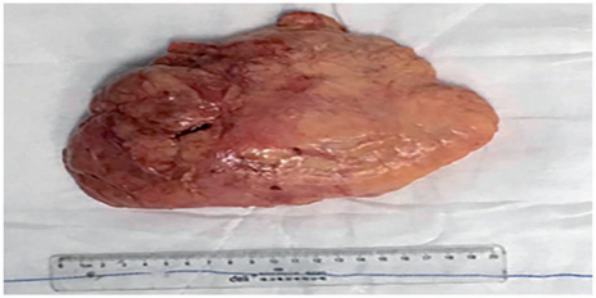

Figure 3. Macroscopic image of the resected tumor. The liposarcoma measured 20x15x10 cm in size.

mass was positive for CD63 and CD163 (data not shown). The patient recovered well and was transferred to the Oncology Department 2 weeks after surgery for chemotherapy and

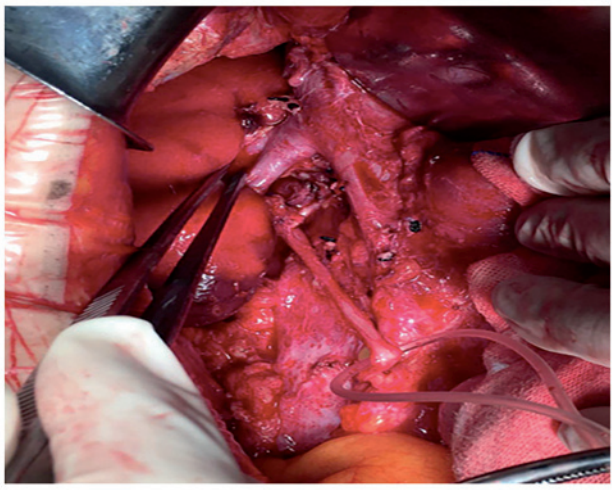

Figure 4. Surgical image. The renal periphery was completely bare, including the renal portal, the ureter and the inferior vena cava.

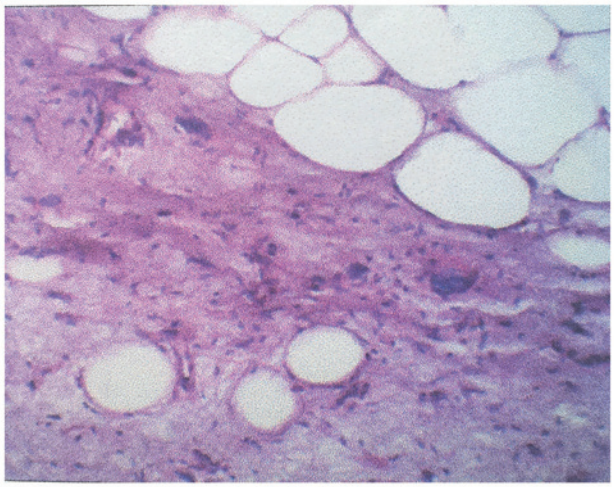

Figure 5. Hematoxylin and eosin staining of the tissue. Staining with hematoxylin and eosin revealed retroperitoneal well-differentiated liposarcoma, full of lipocytes (magnification, x400).

radiotherapy. No tumor recurrence was observed at the 6-month follow-up.

Discussion

Liposarcoma is one of the most common soft-tissue sarcomas that frequently occurs between the ages of 40 and 70 years (9). Since retroperitoneal liposarcoma has no characteristic symptom, the complaints of patients are predominantly associated with direct invasion or compression of other adjacent organs. No significant laboratory abnormalities are observed in the earlier stages and the tumor often has to grow to a larger size before diagnosis can be made. Such tumors are often identified using a diagnostic modality, including computed tomography or magnetic resonance imaging. R0 resection of retroperitoneal liposarcoma had been suggested as the only way to cure the patients (2,8,10).

In order to achieve the R0 resection, contiguous organ resection, including nephrectomy, is always performed when

the intraoperative histological results are dedifferentiated, myxoid/round cell, pleomorphic or mixed-type (10). A recent report demonstrates that with R0 resection, the 5-year survival for well-differentiated subtypes is 90%, while 5-year survival for pleomorphic subtypes is only 30-50%. Dedifferentiated and myxoid/round cell subtypes have 5-year survival rates of 75% and 60-90%, respectively (11). Since it is difficult to detect the retroperitoneal liposarcoma from adjacent normal fat, even if resection of the contiguous organ is performed, the rate of local recurrence is notably high, particularly in high-grade subtypes (12). In order to obtain the free margin, although the single or multi-organ resection may improve clinical outcome, the complication and the quality of life must be considered. The long-term survival rate has been confirmed in a subset of patients after R2 resection (5), particularly with good histological characteristics, including the well-differentiated type. At a 6-month follow-up of the present case, no signs of metastasis or recurrence were observed. Previously, more and more investigations and case reports are inclined to preserve-organ surgery combined with neoadjuvant chemotherapy and radiotherapy to improve the clinical outcome (12-14). It is of great surprise that a new drug, Eribulin mesylate (Halaven), has been approved to treat non-resectable or metastasized liposarcoma (15). Recent research, including 228 patients with retroperitoneal liposarcoma, undergoing surgery in Russia, revealed that the 10-year survival rate was 26% following combined organ surgeries, compared with 35% following surgery on patients not requiring multi-organ resection in dedifferentiated, myxoid cell, pleomorphic and mixed-type cases (7). Low-grade liposarcoma is an expansively growing tumor and so invasion to other organs is relatively rare. Surgical excision with a wide margin will increase the adverse effects, particularly on intestines. Adjacent organ resection is often not necessary since the invasion potential of these tumor types is low (11). Therefore, the present study suggested that nephrectomy must only be performed if required to accomplish a complete gross resection R0 in others histological subtypes. Numerous previous studies have shown that histological subtype, incomplete resection, contiguous organ resection and increasing age are markedly associated with mortality from this tumor. However, tumor burden and nephrectomy were not associated with disease-specific survival (10). Therefore, according to the intraoperative histological subtype, combined organ resection may not be required if confirmed as a well-differentiated tumor. In addition, it is important to have intraoperative histological confirmation on obtained free margins. R0 resection is a gold-standard for retroperitoneal liposarcoma. However, if free margins cannot be achieved by combined organ resection, the decision should be made as to whether combined organ resection can be performed, particularly in low-grade tumor types. Maybe R1 or R2 resection combined with neoadjuvant chemotherapy and radiotherapy are a novel therapeutic method (5,13). Therefore, each aspect of the tumor histological characteristics, the quality of life of the patients, the complications of organ-resection, the benefit of the R0 resection and the long-team survival rate must be considered.

In conclusion, it is unnecessary to performed aggressive surgery, including organ resection, for the majority of well-differentiated retroperitoneal liposarcomas. However, the other subtypes of tumor may require expanding resection, including nephrectomy, in order to obtain negative margins according to the intraoperative histological outcome. A novel therapeutic method, including R1 or R2 resection combined with neoadjuvant chemotherapy and radiotherapy can be performed. However, multi-center, large sample investigations must be performed to further confirm this hypothesis.

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