

# Primary retroperitoneal liposarcoma: a rare case report

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## Abstract

Primary retroperitoneal liposarcoma (PRPLS) is a rare malignant tumor with a low incidence. A 34-year-old female patient presented to our department with abdominal pain, nausea, and vomiting for 2 days. Abdominal computed tomography (CT) indicated a huge mass between the liver and kidney, with a clear boundary and measuring approximately 202 mm × 155 mm × 106 mm. The mass was considered a retroperitoneal lipoma or liposarcoma. The entire tumor was completely resected without auxiliary injury, and histopathology of the resected specimen indicated liposarcoma. The patient recovered well and was discharged from our department on the 6th postoperative day. No signs of relapse were seen during 1-year of follow-up. PRPLS is rare and without obvious symptoms in the early stage. CT plays a vital role in the diagnosis of PRPLS, and surgical resection is considered the most suitable treatment. Radiotherapy and chemotherapy might also be treatment options to improve the overall survival of PRPLS patients.

## Keywords

Primary retroperitoneal liposarcoma, diagnosis, surgery, radiotherapy, chemotherapy, computed tomography, histopathology, malignant

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## Background

Liposarcoma is a rare malignant tumor with a low incidence rate that accounts for less than 1% of all malignancies.<sup>1</sup> According to the morphological characteristics and cytogenetics, sarcoma can be mainly divided into liposarcomas,

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leiomyosarcomas, and malignant fibrous histiocytomas,<sup>2</sup> among which, liposarcoma is the most common, accounting for 41% of all sarcomas.<sup>3</sup> Liposarcoma can occur in any region where fat is present, and approximately 30% of the tumors occur in the retroperitoneal cavity; 35% originate from perirenal fat.<sup>4</sup> Liposarcoma manifests clinically as a painless, progressively enlarging mass; thus, nearly half of retroperitoneal liposarcomas have a diameter of more than 20 cm when diagnosed.<sup>5</sup> Primary retroperitoneal liposarcoma (PRPLS) occurs mainly in patients aged 60 to 70 years, with no sex predominance.<sup>6</sup> In the present study, we report a 34-year-old female patient with PRPLS who successfully underwent surgical resection. In addition, the clinical and pathological characteristics, diagnosis, and treatment methods of PRPLS were summarized.

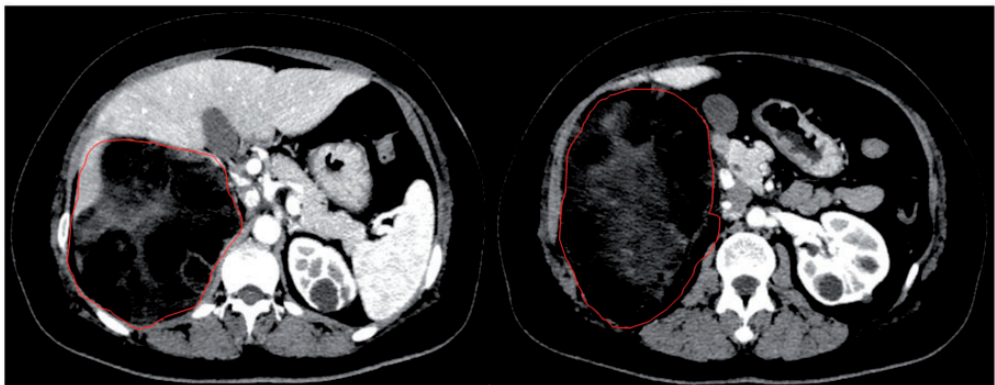
### Case presentation

A 34-year-old female patient presented to our department with abdominal pain, nausea, and vomiting for 2 days. She complained mainly of intermittent gas pains in the upper abdomen without excruciating pain but accompanied by nausea and

vomiting. The vomitus was the stomach contents, and the gas pain was not relieved after vomiting. She sought further diagnosis and treatment at our hospital.

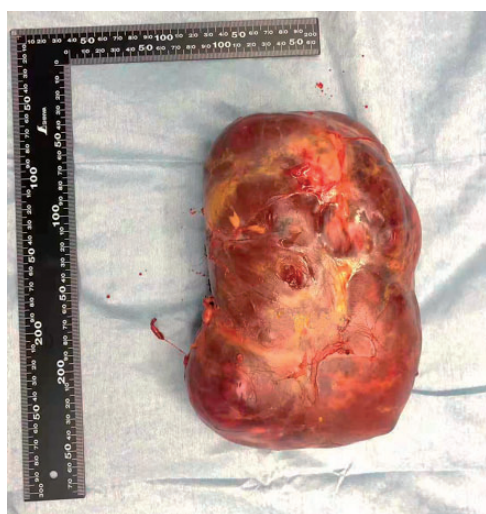
Her medical history showed that she had undergone a cesarean section 5 months prior to presentation. She denied a history of hypertension, diabetes, coronary heart disease, blood transfusions, or drug and food allergies. A physical examination revealed that the abdomen was flat, with a 20-cm transverse surgical scar in the lower quadrant. In the right upper abdomen, at the level of the umbilicus, a smooth and hard abdominal mass measuring 18 cm × 11 cm was palpable. Blood biochemical examination indicated only an elevated white blood cell count ( $13.7 \times 10^9/L$ , normal range:  $3.5\text{--}9.5 \times 10^9/L$ ). Tumor markers were within their respective normal ranges, and electrocardiography and chest radiographic findings were normal. Abdominal computed tomography (CT) indicated a huge mass in the space between the liver and kidney, with a clear boundary, and measuring approximately 202 mm × 155 mm × 106 mm (Figure 1).

Based on the medical history and physical examination and CT findings, the clinical team considered the huge mass as



**Figure 1.** Abdominal computed tomography (CT) images showing a huge mass in the space between the liver and kidney, with a clear boundary. The areas outline in red indicate a primary retroperitoneal liposarcoma (PRPLS).

a retroperitoneal lipoma or liposarcoma. However, histopathological examination is the “gold standard” for an accurate diagnosis. Retroperitoneal resection of the tumor was performed, during which, a 15-cm incision at the right side of the costal margin and a 10-cm incision on the midline of the upper abdomen were made. The tumor was located behind the peritoneum, with a diameter of approximately 20 cm and a complete envelope, and the adjacent organs, such as the stomach, pancreas, and kidney were compressed. There was a clear space between the tumor and nearby blood vessels, such as the abdominal aorta and superior mesenteric artery. The entire tumor was completely resected along the outer edge of the tumor envelope, without auxiliary injury. Rapid intraoperative pathological analysis confirmed negative resection margins. The gross tumor measured 20 cm × 10 cm × 10 cm in size and had a complete envelope and smooth surface (Figure 2). Histopathology demonstrated that the mass was composed of polygonal or short spindle cells with obvious cell

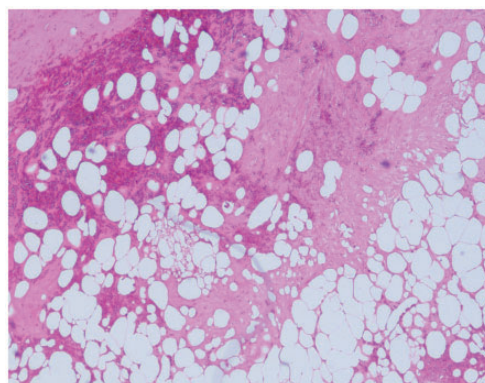


**Figure 2.** The gross tumor measured 20 cm × 10 cm × 10 cm in size, with a complete envelope and smooth surface.

atypia, clear nuclear division, visible tumor giant cells, and necrotic tissue. In addition, the tumor contained large amounts of fatty tissue (Figure 3). Based on the histopathology, the mass was considered a PRPLS. The patient recovered well and was discharged from our department on the 6th day postoperative day. She declined further postoperative treatment, and there were no signs of relapse after 1-year of follow-up.

## Discussion and conclusions

Liposarcoma originates from primitive mesenchymal cells and is composed mainly of adipocytes.<sup>7</sup> Liposarcomas often occur in the peritoneum, limbs, arms, and abdomen, and often invade deep soft tissues.<sup>8</sup> Liposarcomas can be divided into five types pathologically: highly differentiated, myxoid, polymorphic, round cell, and dedifferentiated.<sup>9</sup> The prognosis of well-differentiated and myxoid liposarcomas is good. Most PRPLSs are easy to define, with an intact envelope that consists of a thin layer of flat tumor cells formed by tumor growth pressure. The tumor usually has distinct lobes, and each lobe is round or oval in shape.<sup>10</sup>



**Figure 3.** Histopathology (hematoxylin and eosin staining × 100) indicating that the tumor was a primary retroperitoneal liposarcoma (PRPLS).

PRPLS, which occurs mainly in patients aged 60 to 70 years, is rare and without obvious symptoms in the early stage. Accurate diagnosis and complete resection are vital for the treatment of PRPLS. In the present study, a 34-year-old female patient with PRPLS successfully underwent surgical resection. The clinical and pathological characteristics, diagnosis, and treatment methods of PRPLS were also summarized for the reference of other researchers.

PRPLS has no obvious symptoms in the early stage, and a prominent feature is a huge abdominal mass with mild symptoms.<sup>10,11</sup> Similarly, retroperitoneal uterine leiomyoma also presents with non-specific symptoms, but patients with these tumors always experience pain during intercourse.<sup>12</sup> When diagnosed, PRPLS is often in the late stage, and the corresponding symptoms appear only when the adjacent organs are compressed or when tumors gradually increase in size.<sup>13</sup> When the gastrointestinal tract is compressed, patients may feel fullness, anorexia, nausea, vomiting, diarrhea, abdominal pain, bloating, and pain related to constipation or defecation pain.<sup>13,14</sup> In addition, pressure on the kidney may cause hydronephrosis, and pressure on the bladder may cause frequent urination and urgency.<sup>15</sup> Pressure into the chest from a large tumor may lead to breathing difficulties.<sup>16</sup>

Ultrasonography, CT, and magnetic resonance imaging (MRI) are especially important because the early clinical signs of liposarcoma are not obvious. Ultrasonography is used mainly to determine the size and number of tumors and has the advantages of non-invasiveness and low cost. CT is often the first auxiliary examination for PRPLS. CT has a certain value in judging the pathological type and malignant degree of PRPLS, but confirming these features depends mainly on the histological components of the tumor.<sup>17</sup> Recent improvements in CT have led to

higher resolution and clear images showing the size, location, and scope of liposarcomas and their relationship with surrounding organs. MRI can identify soft tissue diseases with important diagnostic significance for tumor invasion of structures, such as the abdominal aorta or inferior vena cava.

Complete surgical resection with negative margins is the main treatment for PRPLS. In a single-center study of 500 patients with PRPLS, patients who underwent complete surgical resection with negative margins had a median survival of 103 months. However, the median survival in patients who did not undergo complete resection was only approximately 18 months.<sup>18</sup> If it is difficult to remove the tumor completely in the advanced stage, tumor reduction surgery should still be performed to reduce the symptoms of compression, prolong survival time, and improve quality of life.<sup>19</sup> For patients with recurrent liposarcomas, reoperation should be performed.

Radiotherapy, including preoperatively, intraoperatively, and postoperatively, can be used to treat PRPLS to improve quality of life and tumor-free survival.<sup>20</sup> Although well-differentiated and slow-growing PRPLS is relatively sensitive to radiotherapy, this modality is not a substitute for surgery. The combination of intraoperative radiotherapy, surgical resection, and postoperative external irradiation can control the patient's condition effectively.<sup>21</sup> However, it is still controversial whether adjuvant radiotherapy can improve the survival rate of PRPLS patients owing to the lack of sufficient clinical evidence.

Adjuvant or neoadjuvant chemotherapy is not a standard treatment method for PRPLS patients; however, it might be an option when the tumor is unresectable or insensitive to radiotherapy.<sup>22</sup> Although there are no specific chemotherapeutic drugs for liposarcoma, some combined

chemotherapy agents possess significance for micrometastatic liposarcoma.<sup>23</sup>

Radiotherapy and chemotherapy for PRPLS have not provided definitive effects. Therefore, tumors that recur post-surgery are recommended to be treated as soon as possible. Regular postoperative follow-up is required for PRPLS patients. Scientists should focus on the molecular mechanisms of liposarcoma to develop new targeted drugs for PRPLS. Surgery combined with targeted drugs, radiotherapy, or chemotherapy might reduce tumor recurrence and improve the prognosis of PRPLS patients.

In the present study, we reported a young female patient with PRPLS. In addition, the clinical and pathological characteristics, diagnostic, and treatment methods of PRPLS were summarized.

#### Availability of data and materials

All data generated or analyzed are included in this published article.

#### Authors' contributions

SW wrote the first version of the article. XH and SYL revised the article. XH, SYL, GMX, and JNL participated in the conception and design of the study and drafting the article. JNL supervised the study and reviewed the article critically for intellectual content. All authors reviewed and approved the final version of the article.

#### Declaration of conflicting interest

The authors declare that there is no conflict of interest.


#### Ethics approval and consent to participate

This study was approved by the Ethics Committee and Institutional Review Board of the Second Hospital of Jilin University, Changchun, China. Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

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