Primary retroperitoneal liposarcoma with extensive ossification
A case report
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Abstract
Rationale: Primary retroperitoneal liposarcoma, which originates from mesenchymal tissues, can rarely present with extensive ossification.

Patient concerns: A 41-year-old male patient presented with a chief complaint of discomfort around the waist for 2 months.

Diagnoses: Computerized tomography (CT) and magnetic resonance imaging suggested a lesion of approximately 5.6 × 5.1 × 8.7 cm in front of the psoas major muscle, which was considered to be a mesenchymal or neurogenic tumor.

Interventions: The hard mass was removed by laparotomy, and the pathological investigation revealed that this was an atypical lipomatous tumor/well-differentiated liposarcoma, with extensive ossification.

Outcomes: The patient was discharged from the hospital after surgery. There was no sign of reoccurrence after 1 year of follow-up.

Lessons: Retroperitoneal liposarcomas with extensive ossification are rare tumors that can present with nonspecific symptoms, and are difficult to diagnose. CT is the most common imaging technique, and surgical resection has been considered to be the most effective treatment. This rare case can be challenging for diagnosis and treatment.

Abbreviations: CT = computerized tomography, MRI = magnetic resonance imaging, PRPLS = primary retroperitoneal liposarcoma.

Keywords: diagnose, ossification, primary retroperitoneal liposarcoma, surgical resection, well-differentiated liposarcoma.

1. Introduction
Retropertioneal sarcoma is a rare type of malignant tumor, which accounts for approximately 15% of all sarcomas.[1,2] An incidence rate of 0.3% to 0.4% per 100,000 population has been reported from a US-based disease registry.[1,3] Retroperitoneal sarcomas can be liposarcomas, or leiomyosarcomas or malignant fibrous histiocytomas, and out of these, liposarcomas are the commonest.[2] These liposarcomas generally occur in the extremities or retroperitoneum.[3,4] Primary retroperitoneal liposarcoma (PRPLS) has a higher incidence within the age group of 60 to 70 years old, with no gender predominance.[5] We present a rare 41-year-old male patient with a sclerotic type retroperitoneal well-differentiated liposarcoma, and extensive ossification.

2. Case report
A 41-year-old male patient presented with a chief complaint of discomfort in the flanks for the last 2 months. The pain was intermittent and undiffused, and there was no relief on taking rest. Up to this presentation, the patient has been healthy with no related symptoms. The patient was hospitalized for further diagnosis and treatment. It was found that the routine examination and tumor markers were within the normal range. On magnetic resonance imaging (MRI), a lesion was observed in front of the psoas major muscle. This lesion could be a mesenchymal tumor or neurogenic tumor. Furthermore, computerized tomography (CT, Fig. 1) identified a right retroperitoneal space that occupied the lesion, which was approximately 5.6 × 5.1 × 8.7 cm in size.

Based on the preoperative medical history, physical signs and auxiliary examination, it was diagnosed as a case of posterior peritoneal space-occupying lesion. However, the nature of the tumor needed to be confirmed by pathological examination after resection. Thus, a retroperitoneal mass resection was planned. After careful separation in front of the tumor, it was observed that the tumor lifted the right ureter, and the right ureter continued to move downward, entering the tumor from the middle of the tumor and passing out from the bottom of the tumor. Since the tumor was severely adhered to the right kidney, the right ureter left a crevasse of approximately 0.3 cm when this was moved away from the tumor. However, considering that the
reseption of the tumor might have affected the ureteral stent implantation, the gauze was used to isolate this as the operator continued the separation. There was a gap between the tumor and blood vessels, such as the inferior vena cava, without obvious adhesion. After completely separating the tumor from the inferior vena cava and right iliac artery, the posterior wall of the tumor and psoas muscle could be closely observed. Then, the tumor was completely removed. Next, the urology surgeon performed ureteral stent implantation, and no leakage of urine or bleeding was found in the right ureter. The tumor (Fig. 2) had a lobular mass of approximately 8.2 × 5.5 × 4.2 cm. On the surface of the mass, there was a suspicious bone tissue of approximately 2.0 × 1.5 × 0.9 cm. Postoperative pathological (Fig. 3) evaluation revealed that it was a hardening type atypical lipomatous tumor/well-differentiated liposarcoma, with extensive ossification. There was no metastatic invasion in the vessels, nerves, and surrounding lymph nodes. The immune-histochemical results revealed the following: CD34(–); CD99(±); Desmin(–); SMA (part+); ki-67(5%+); STAT6(+); Bcl-2(–); CD117(–); S-100(diffusion+). CD34, SMA, Desmin, CD99, S-100 protein, and Ki-67 (cell proliferation markers) are often used in the diagnosis of soft tissue tumors. S-100 protein has a positive expression in well-differentiated and mucinous PRPLS, but has a negative expression in undifferentiated and polymorphous/mixed PRPLS. STAT6 has a positive expression in isolated fibrous tumors. K-67 is a recognized nuclear antigen that is specifically correlated to cell proliferation, and is mainly used to determine cell proliferation activity. The clinical diagnosis of PRPLS was
established. Then, the patient was discharged from the hospital after a week (July 14, 2017). There was no report of relapse on the follow-up survey after 6 months and after 1 year.

3. Discussion
Liposarcomas are the commonest type of retroperitoneal sarcomas.\[1,6\] Generally, patients with sarcomas present late, since the retroperitoneal area provides a potential space for the growth of tumors without producing any symptoms.\[2\] Most of these patients may remain asymptomatic or present with nonspecific symptoms since these sarcomas can grow large without pressing on any organ.\[3\] However, some patients with retroperitoneal sarcomas can present with gastrointestinal symptoms such as vomiting and/or nausea, when the sarcoma is pressing the gastrointestinal tract, while others can have uremia, or hydronephrosis or nephropelitis, when pressure has been placed on the kidneys. Similarly, patients can present with increased frequency of micturition or urgency when the urinary bladder is affected, and pressure on the nerves can present as neural syndromes.\[4\] The above-mentioned patient presented to the hospital with the only complaint of discomfort in the flanks, regardless of the growing size of the sarcoma.

The abovementioned case was diagnosed by CT and MRI. Ultrasound examination was used to screen the retroperitoneal sarcomas since it can accurately determine the location, size, and relationship of the tumor with the surrounding blood vessels. An ultrasound examination is easy to perform, has a high penetration rate, and is economical. Furthermore, the machine is portable. However, it is difficult to differentiate low-differentiated liposarcomas from other types of retroperitoneal tumors.\[5\] CT is the most common imaging technique for the identification, localization, and staging of retroperitoneal sarcomas.\[6\] CT can help in clarifying the anatomical location, size, and possible origin of the tumor, its relation with the adjacent viscera, nerves and vessels, and the presence or absence of metastasis.\[7\] MRI is one of the best techniques for evaluating retroperitoneal tumors.\[8\] MRIs are required to further investigate the level of invasion of the tumor, determine the source of the tumor, and identify the neurovascular or muscle invasion.\[9\] MRI combined with enhanced-CT can help in the differential diagnosis of various histological subtypes of liposarcomas.\[10\] Along with radiological investigations, pathological, and immunohistochemical investigations form the gold standard for the diagnosis of PRPLS. Percutaneous needle biopsy has low accuracy for the diagnosis of dedifferentiated retroperitoneal liposarcomas.\[11\]

The prognosis of retroperitoneal liposarcomas depends on the extent of differentiation, histologic subtype, margin of resection, gross resection of the tumor, and need for contiguous organ resection.\[6,10\] Liposarcoma specific nomograms help in determining the prognosis of patients. As compared to other retroperitoneal sarcomas, liposarcomas can be graded. A low-grade malignant PRPLS shows a clear boundary, and is lobulated, mostly fat dense, and without calcification. On the other hand, a highly malignant tumor shows a blurring border, has nodular separation, and may have calcification, but with less fat content.\[11\] The presence of ossification or calcification is a poor prognostic sign.\[2\] Retroperitoneal dedifferentiated liposarcomas are known to be associated with 83% local recurrence and 30% distant recurrence rates.\[6\] Univariate and multivariate analyses have shown that histologic subtype and contiguous organ resection are independent prognostic factors for both local and distant recurrence. However, age, gender, tumor burden, margins, nephrectomy, and sclerosing subtype are not prognostic factors for local recurrence.\[10\] The above-mentioned case had a well-differentiated tumor, and there was no invasion of the surrounding areas. However, ossification was present, which is a poor prognostic sign. There has not been any sign of recurrence or any other sign of poor prognosis after 1 year of surgery. However, it is notable that the local recurrence of liposarcomas is difficult to detect since it is difficult to differentiate small recurrent liposarcomas from normal retroperitoneal fat.\[2\] Regular follow-up is required to detect its local or distant reoccurrence.

The patient underwent complete surgical resection, which was considered to be the most effective treatment.\[2,11–13\] Chemo-therapy and radiotherapy does not have known definitive effects on PRPLS.\[11\] According to general practice, for the present case, 1 to 2 cm of the negative area at the edge of the tumor was removed along with tumor resection.\[14\] Since the metastasis was not evident, the draining lymph nodes and adjacent organs were not resected. Studies have shown that the complete resection of tumors visible to the naked eye improves the prognosis.\[10,15,16\] Furthermore, a retrospective analysis revealed that complete surgical resection led to a 3- and 5-year survival rate of 87% and 49%, respectively.\[11\] In addition, the resection of adjacent organs helps in preventing the local spread.\[17\] Palliative resection is beneficial in case of tumor invasion of the surrounding organs.

There are a limited number of case reports of retroperitoneal liposarcoma with ossification. Among the published cases, the present patient was the youngest. Furthermore, all cases were dedifferentiated retroperitoneal malignant liposarcomas, whereas the present case had a well-differentiated histopathologic picture.\[18–22\]

The patient provided informed consent for the publication of this case.

Author contributions
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