

Case Report

Giant retroperitoneal well-differentiated liposarcoma - a case report

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Abstract: Well-differentiated abdominal liposarcoma is a rare cancer of adipocyte origin. Due to their retroperitoneal location, these tumours can become very large and grow to a considerable size before symptoms appear. A 50-year-old woman was diagnosed with a large retroperitoneal tumour. After excluding contraindications for surgery, surgery was performed. The tumour was found to be closely related to multiple organs, and compression was obvious. It was completely excised, and postoperative pathology revealed a well-differentiated liposarcoma. Abdominal and retroperitoneal liposarcomas are rare and are locally progressive at the time of presentation. Surgery is the primary treatment method, and complete resection is the goal of surgery. Extended resection to reduce local recurrence remains an area of research, as does a balance between the incidence and recurrence rates. Given the rarity of these tumours, treatment at an experienced centre is the key to obtaining the best outcome.

Keywords: Giant liposarcoma, retroperitoneal, well-differentiated, surgery

Introduction

Abdominal well-differentiated liposarcoma (also known as atypical lipoma [1]) is a rare cancer of adipocyte origin that is estimated to occur in 1 per 100,000 people every year. The 5-year overall survival rate of the cancer is 71-96%. Due to the high local recurrence rate, the survival rate drops to approximately 50% after 10 years, becoming the most common cause of disease-related death [2, 3]. Because of their retroperitoneal location, these tumours can become very large and grow to a considerable size before symptoms appear. This usually leads to critical vascular supply and abdominal visceral involvement before the diagnosis and affects the complete removal of these tumours.

Liposarcomas can be categorised into four types. The two most common types of retroperitoneal liposarcomas are low-grade well-differentiated liposarcomas (WDLPS) and high-grade dedifferentiated liposarcomas (DDLPS). These two variants have the molecular characteristics of MDM2 gene amplification, which distinguishes them from other retroperitoneal tumours. In retroperitoneal soft tissue sarco-

mas, WDLPS and DDLPS account for 40% of cases. Other less common sarcomas histologically include leiomyosarcomas, undifferentiated pleomorphic sarcomas, solitary fibromas, desmoplastic small round cell tumours, and malignant peripheral nerve sheath tumours.

WDLPS account for approximately 45% of all liposarcomas and are usually locally invasive but rarely metastatic. The presence of dedifferentiated components greatly affects the overall prognosis. Compared with DDLPS, WDLPS have a lower metastatic rate, local recurrence rate, and overall death risk of 1/6 [4, 5]. These two types of liposarcomas are unique to liposarcomas and are commonly found in the same tumour juxtaposed with each other. While this report focuses on WDLPS, in practice, surgeons often encounter components of both types. The main complications of these lipomas are mass, compressive symptoms and cachexia. Surgery is the main treatment method for abdominal WDLPS, and the quality of surgery is critical for any potential opportunity of cure. Surgery is usually considered when the diagnosis is clear on imaging or after pathological confirmation [6]. After surgery, WDLPS have a 30-60% risk of

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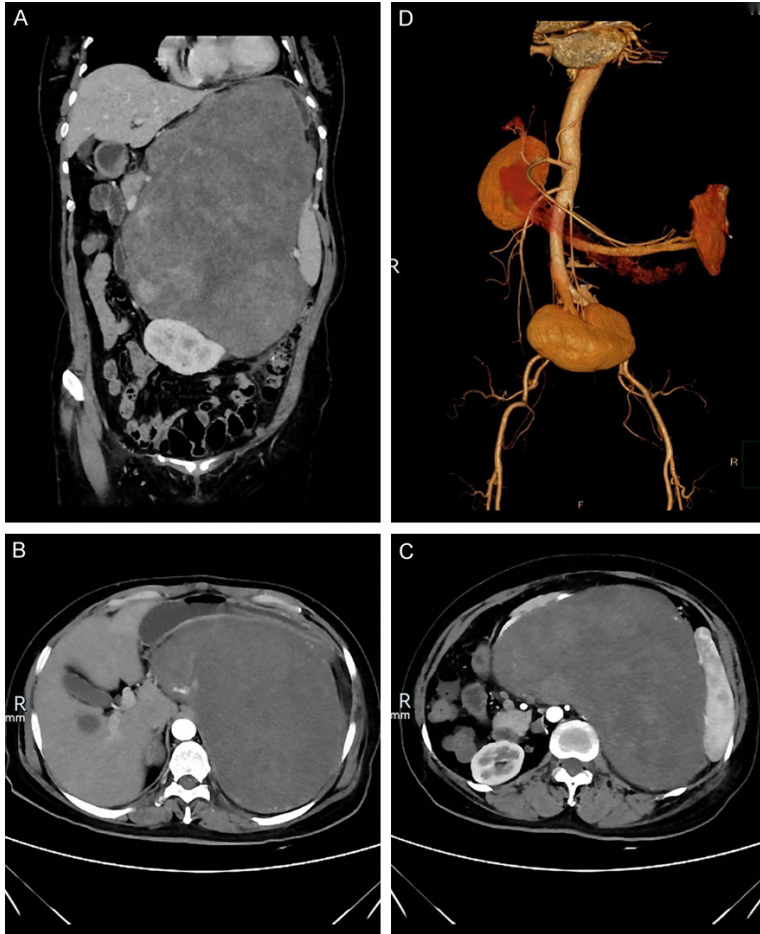


Figure 1. Partial images of CT scan. A-C. There was macrosomia in abdominal cavity. It can be seen that the kidney and other organs are displaced, and the spleen and other organs are compressed. D. Vessels such as the abdominal aorta are squeezed and displaced.

5-year local recurrence. Patients may undergo multiple surgeries in an attempt to protect them from the cancer recurrence. Postoperative complications include incision infection, postoperative bleeding, damage to adjacent organ function, and haemodynamic disturbances. Scars before surgery, distorted normal anatomy, and/or restricted access to the abdomen may limit the chances of complete resection. Since surgery remains the most effective treatment, the quality and scope of initial surgery are the subject of in-depth research on this particular disease. Because of the low risk of metastasis, adjuvant chemotherapy or radiotherapy has a limited role in this cancer type.

The rarity of tumours limits the number of meaningful randomised controlled trials that evaluate therapeutic algorithms [7]. Often,

studies combine different sarcoma subtypes, although their underlying biology is very different. Therefore, it cannot be underestimated that these patients need to be treated by multidisciplinary teams at high-volume centres.

Case description

A 50-year-old woman from Zhejiang, China, visited our outpatient clinic because of abdominal distension. She felt that her abdominal circumference gradually increased within 1 month, abdominal distension increased, and she occasionally had laboured breathing. Abdominal CT performed in the outpatient department revealed a large solid space-occupying lesion in the abdominal cavity. She was then hospitalised for further treatment.

The patient was concerned about the abdominal distension and enlarged abdominal circumference when she visited the doctor. Physical examination revealed a huge mass in the left abdomen, up to the xiphoid process and down to

the level of the umbilicus, with hard texture, poor mobility, and no obvious tenderness. The gastric origin of the tumour was considered to be large. Further contrast-enhanced CT examination of the whole abdomen indicated that the huge mass was of retroperitoneal origin and that the liposarcoma might be large (**Figure 1A**). Imaging data also showed that the tumour caused displacement of the abdominal organs (**Figure 1B, 1C**). The space in the stomach was compressed such that there was not enough room for food, which was mainly why the patient often experienced bloating. The diaphragm was squeezed up; therefore, she struggled to breathe. The spleen was significantly compressed, down to near the level of the umbilicus. The left kidney was displaced anteriorly and inferiorly to the lower abdomen, reaching the front of the bifurcation of the abdominal

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Figure 2. Gross appearance of the tumor. The initial measurement was about 35 cm in length and about 8 kg in mass.

aorta to the common iliac vessels, with obvious pressure. The abdominal aorta and other blood vessels (**Figure 1D**), small intestine, colon, etc., showed different degrees of rightward and downward displacement. The results of the preoperative routine blood, stool, biochemistry, coagulation function, and tumour marker tests were all unremarkable. A routine urine test revealed weakly positive occult blood. Thus, although various organs of the patient were obviously oppressed, their functions had not been significantly affected.

After the preoperative preparation was perfected, surgical resection was performed and a left abdominal paramedian incision was made. Combination of blunt and sharp separation was made. The transverse colon and descending mesangium were freed, and the tumour was further exposed. As the pancreas was raised, its body and tail were freed together with the spleen and pulled to the outside of the abdominal cavity for protection. Continued dissection showed no obvious boundary between the tumour and part of the gastric wall; therefore, a part of the gastric wall was removed. Blood vessels such as the superior mesenteric artery,

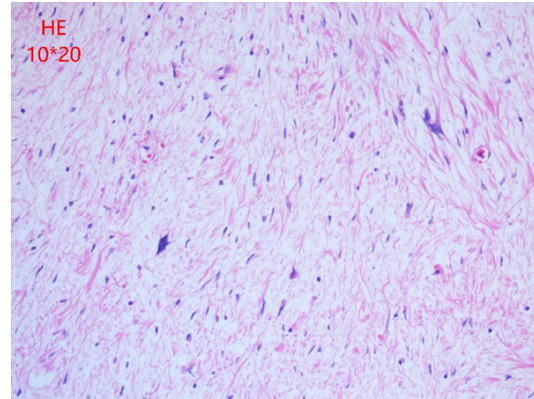


Figure 3. Images were obtained by H&E staining and light microscopy at 200 × magnification. It shows a well-differentiated liposarcoma.

superior mesenteric vein, portal vein, and abdominal aorta were closely attached to the tumour and strictly protected during tumour dissociation. The tumour was closely related to the diaphragm and separated from the capsule. It was completely excised and appeared yellowish white. On initial measurement, the tumour measured approximately 35 cm in length and weighed 8 kg (**Figure 2**). The patient was transferred to the ICU for transition after surgery, transferred to the general ward and recovered smoothly. Postoperative pathology showed that the large retroperitoneal tumour was a well-differentiated liposarcoma (**Figure 3**), and there were three conjoined primary foci with sizes of 31 × 25 × 10 cm, 5 × 3 × 2 cm, and 11 × 5 × 1.5 cm. The tumour showed CDK4, P16, and CD34 positivity, which was consistent with the diagnosis of liposarcomas.

The patient was referred to the ICU for monitoring after surgery and had no significant disturbance of consciousness after waking up from anaesthesia. ICU monitoring revealed that she was mentally sound during and had blood pressure instability, moderately or severe anaemia, hypoalbuminaemia, with a chief complaint of pain in the surgical area; thereafter, she was transferred back to the general ward on the 9th postoperative day. She was uneventfully discharged 2 weeks postoperatively and was followed up at the Department of Medical Oncology for regular chemotherapy; after chemotherapy, fatigue and myelosuppression developed, which improved after symptomatic treatment, and the patient successfully completed

eight courses of chemotherapy; no tumour recurrence or metastasis was found 1 year after surgery.

Discussion

The patient thought that she was only generally obese at first, so she had not come to medical attention until she developed obvious symptoms such as abdominal distension and chest tightness. This suggests that WDLPS does not easily cause symptoms until it grows large enough, and the authors believe that abdominal ultrasonography should be refined at least for patients with sudden signs of obesity, which can help in the early detection of possible WDLPS. Although WDLPS can be differentiated from other tumours by enhanced CT examination, there is no definite criteria for the identification of patients with WDLPS on imaging, expecting the progress of related techniques and theories. The current absence of indicators or markers in blood that directly reflect WDLPS awaits further development in relevant studies. The operation of this patient, although it appeared difficult, was finally completed smoothly, and the patient recovered uneventfully after the operation, suggesting that operation is still the primary treatment for this disease. As long as there is no infiltration of vital organs or vessels, complete resection of the tumour should be attempted, and should not be trapped for its size. Several courses of post-operative chemotherapy ended smoothly, and the patient had no obvious signs of tumour recurrence or metastasis, suggesting that post-operative chemotherapy should still be performed as a routine item when disease occurs.

Summary

This patient's lack of significance before symptomatic preoccupation with thinking about general adiposity illustrates the insidious nature of the disease, which is not easily detected when it is not sufficiently large. The smooth recovery after surgery suggests that surgery remains the preferred treatment method for these tumours, and it is more beneficial that experienced physicians perform the surgery so as to guarantee safety.

Abdominal and retroperitoneal liposarcomas are rare and locally progressive. Surgery is the mainstay treatment for these tumours, and

complete resection is the initial objective of surgery. Expanding resection to reduce local recurrence remains an area of research, and a balance between the morbidity and recurrence rates of such surgeries is required [6]. Given the rarity of these tumours, treatment at an experienced centre is key to achieving optimal outcomes.

Well-differentiated liposarcomas represent the pedigree of a single disease entity, which is characterised by abnormalities involving chromosome 12q13-15, especially high-level amplification of the MDM2 and CDK4 oncogenes [8]. WDLPS reshape mature fat morphologically and have recurrence locally, which is related to their location, especially the retroperitoneum; once metastasised, they exhibit an aggressive behaviour. Surgical resection is the primary treatment for these tumours. Conventional drug treatment is limited to patients with local recurrence or metastasis. However, further understanding of the genetic basis of this group of tumours may help identify targeted therapies, which can be incorporated into multimodal therapeutic approaches.

Disclosure of conflict of interest

None.

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