Section: Surgery

Case Report

LARGE ASYMPTOMATIC RETROPERITONEAL LIPOSARCOMA CASE REPORT AND LITERATURE REVIEW

P Balaji[1], Vidya Lakshmi[2], Siddharth D[3], Partheeban Balasundaram[4], Nitish R Jayaharan[5], Eaasvar JC[5]

[1]*Director of General Surgery and Minimal Invasive Surgery, SRM Institute of Medical Science, Vadapalani, Chennai, Tamil Nadu, India*
[2]*Consultant, Department of General Surgery and Minimal invasive surgery, SRM Institute of Medical Science, Vadapalani, Chennai, Tamil Nadu, India*
[3]*Senior Consultant, Department of General Surgery and Minimal invasive surgery, SRM Institute of Medical Science, Vadapalani, Chennai, Tamil Nadu, India*
[4]*Pathologist, SRM Institute of Medical Science, Vadapalani, Chennai, Tamil Nadu, India*
[5]*Junior Resident, Department of General Surgery and Minimal invasive surgery, SRM Institute of Medical Science, Vadapalani, Chennai, Tamil Nadu, India*

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Corresponding Author:
Dr. P Balaji,
Director of General Surgery and Minimal invasive surgery, SRM Institute of Medical Science, Vadapalani, Chennai, Tamil Nadu, India.
Email: drpbalaji15@gmail.com

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ABSTRACT

Background: Retroperitoneal liposarcomas are uncommon malignant adipocytic neoplasms that can attain considerable size before producing significant symptoms. Well-differentiated liposarcoma (WDLPS) is characterized by mature adipocytic differentiation and has a low metastatic potential but a substantial propensity for local recurrence. Complete macroscopic resection remains the cornerstone of treatment and may require en-bloc resection of adjacent organs when the tumour is locally extensive[1-4,9]

Case Report: A 69-year-old man presented with abdominal discomfort and fullness for 2 months. Ultrasonography demonstrated a large heterogeneous abdominal mass, and fine-needle aspiration cytology suggested an atypical adipocytic neoplasm. Contrast-enhanced computed tomography revealed a large predominantly fat-attenuating retroperitoneal mass extending into the pancreatico-gastric region, left adrenal and perirenal compartments, and central mesentery, with close involvement of the distal pancreas, spleen and left kidney. The patient underwent exploratory laparotomy with compartmental excision of the tumour and en-bloc multivisceral resection comprising distal pancreatectomy, splenectomy, left nephrectomy and adrenalectomy. The resected tumour measured 23 × 21 × 14 cm on pathological examination. Histopathology confirmed well-differentiated liposarcoma, FNCLCC grade 1, with pathological stage pT4 and regional nodal status not assigned. The postoperative course was uncomplicated, with satisfactory recovery and preserved renal function.

Conclusion: Giant retroperitoneal WDLPS can remain relatively indolent clinically despite extensive local growth and involvement of adjacent organs. Complete macroscopic en-bloc resection remains the principal treatment for localized disease. In selected patients, multivisceral resection may be necessary to achieve adequate oncological clearance. Long-term surveillance is essential because of the significant risk of local recurrence[1-4, 13,14]

Keywords: retroperitoneal liposarcoma; well-differentiated liposarcoma; multivisceral resection; distal pancreatectomy; splenectomy; nephrectomy; retroperitoneal sarcoma.

INTRODUCTION

Liposarcomas are malignant mesenchymal neoplasms characterized by adipocytic differentiation and represent an important group of adult soft-tissue sarcomas. They comprise several histological subtypes with distinct molecular

characteristics and biological behaviour. Well-differentiated liposarcoma and dedifferentiated liposarcoma have a particular predilection for the retroperitoneum and are commonly associated with amplification of the MDM2 and CDK4 genes.[1-10]

Retroperitoneal liposarcomas frequently remain asymptomatic until they reach a considerable size because of the large potential space within the retroperitoneal compartment. Patients may present with nonspecific symptoms such as abdominal fullness, discomfort, distension, early satiety or a palpable abdominal mass.[9-14] WDLPS generally has a low propensity for distant metastasis in the absence of dedifferentiation; however, local recurrence is common and represents the principal oncological challenge.[3,4,9,14]

Contrast-enhanced CT and magnetic resonance imaging are important for defining tumour extent and its relationship to major vessels and adjacent organs.[5,6,15] Histological diagnosis is preferably established using image-guided core biopsy, with molecular testing such as MDM2 amplification analysis used when required for confirmation.[9,15]

For localized retroperitoneal sarcoma, complete macroscopic surgical resection is the primary treatment.[1-3] Because of the tendency of retroperitoneal liposarcomas to envelop or involve adjacent structures, resection may require removal of contiguous organs.[1-4,13] En-bloc resection without tumour rupture or fragmentation is preferred when technically feasible.[12] We present a case of a giant primary retroperitoneal WDLPS involving multiple adjacent visceral organs that was successfully managed with extensive en-bloc multivisceral resection.

CASE REPORT

A 69-year-old man presented with a 2-month history of abdominal discomfort and progressive abdominal fullness. His medical history included hypertension, diabetes mellitus, dyslipidaemia and hypothyroidism, all controlled with regular medication.

On examination, he was haemodynamically stable. Abdominal examination revealed a large, firm, palpable mass occupying the epigastric and umbilical regions. Routine laboratory investigations were within acceptable limits.

Ultrasonography demonstrated a large encapsulated heterogeneous hyperechoic mass in the midline of the abdomen. Ultrasound-guided fine-needle aspiration cytology revealed an atypical adipocytic neoplasm.

Contrast-enhanced CT of the abdomen demonstrated a large heterogeneous predominantly fat-attenuating mass occupying the central retroperitoneum and extending into the pancreatico-gastric region. Superiorly, the lesion extended into the lesser sac and produced mass effect on the pancreas. Posteriorly, it extended around the left adrenal gland and reached the left Gerota's fascia, with mild extension toward the left lateral conal fascia. Inferiorly, the lesion extended toward the central mesentery and abutted adjacent small-bowel loops. The tumour was closely related to the distal pancreas, spleen and left kidney. Vascular branches supplying the lesion arose predominantly from the left gastric and splenic arterial territories, with venous drainage toward the splenic vein. The radiological findings were suggestive of a large retroperitoneal liposarcoma.

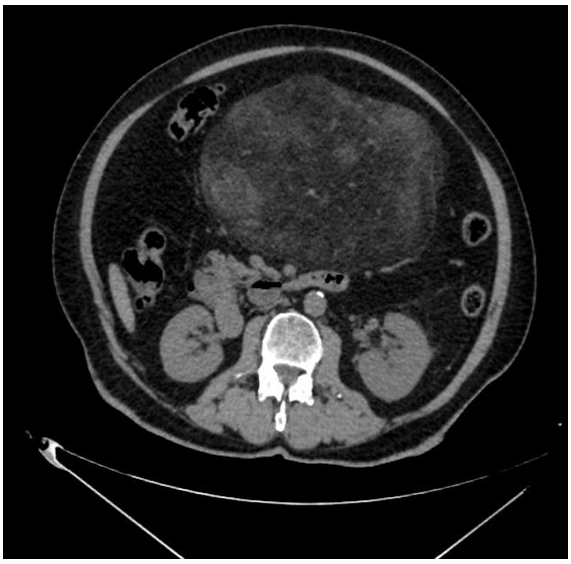

Figure 1: CT scan abdomen- Retroperitoneal liposarcoma

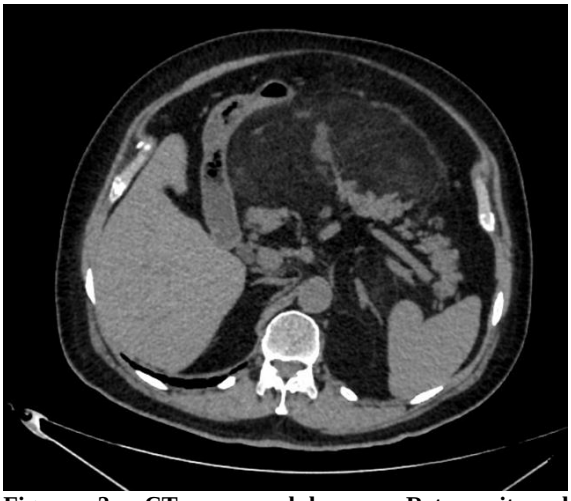

Figure 2: CT scan abdomen- Retroperitoneal liposarcoma involving the pancreas

Following multidisciplinary evaluation and anaesthetic and cardiac assessment, the patient underwent exploratory laparotomy with the intent of complete macroscopic tumour removal.

A midline laparotomy was performed and a large retroperitoneal tumour measuring approximately 25 × 20 × 15 cm, distal pancreas, spleen, left kidney, adrenal gland and perinephric tissue were removed en bloc and submitted for histopathological examination.

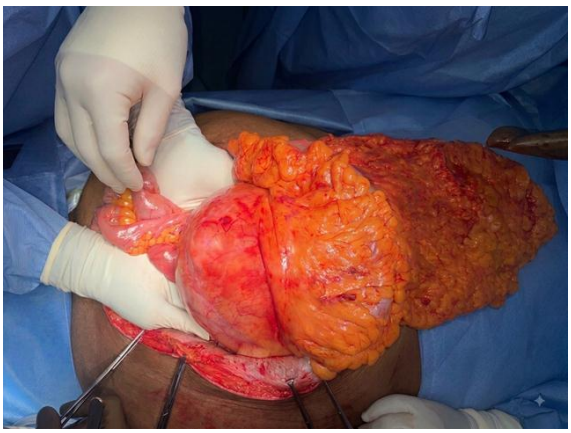

Figure 3: Retroperitoneal liposarcoma intraoperative image

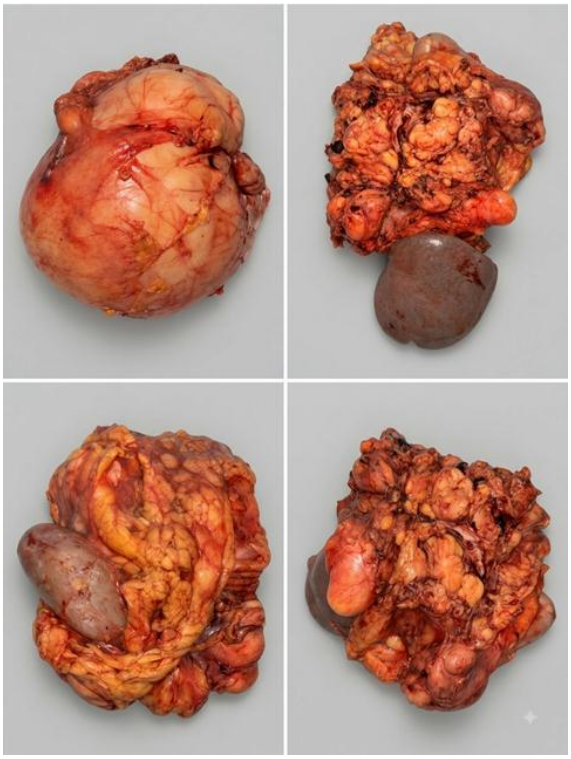

Figure 4: Post operative specimen including retroperitoneal liposarcoma, spleen, left kidney and distal pancreas

Histopathological examination demonstrated a well-differentiated liposarcoma, FNCLCC grade 1. The tumour measured 23.0 × 21.0 × 14.0 cm. The tumour encased and involved pancreatic tissue and encased the left kidney with associated adrenal tissue. The spleen showed vascular congestion. The reported pathological stage was pT4, pN not assigned.

The postoperative period was satisfactory. The patient received respiratory physiotherapy and incentive spirometry and was gradually advanced to oral feeding. Renal function remained stable following nephrectomy, with adequate urine output. The surgical wound remained healthy, and the drains were subsequently managed according to output. The patient was discharged in clinically stable condition and is on follow-up.

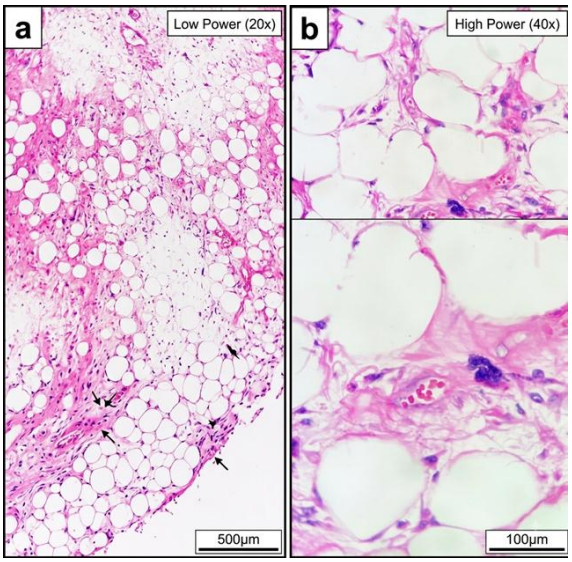

Figure 5: Histopathology

DISCUSSION

Retroperitoneal liposarcoma is one of the most frequently encountered histological subtypes of primary retroperitoneal sarcoma. These tumours may grow to an enormous size before becoming symptomatic because the retroperitoneal compartment can accommodate substantial tumour expansion.[9,10,12] In the present case, the patient presented only with abdominal discomfort and fullness despite having a tumour measuring more than 20 cm.

WDLPS is characterized by mature adipocytic tissue containing atypical stromal cells and variable fibrous septa. In contrast to dedifferentiated liposarcoma, WDLPS generally has minimal metastatic potential when a dedifferentiated component is absent.[9,15] Its major clinical problem is local progression and recurrence.[3,4,9] The diagnosis may be supported by amplification of MDM2, with CDK4 amplification also frequently present.[9,15] Molecular confirmation is particularly useful when distinguishing WDLPS from benign lipomatous lesions or other adipocytic neoplasms.[9]

Cross-sectional imaging is essential for defining the anatomical extent of retroperitoneal liposarcoma and determining its relationship with adjacent organs and major vessels.[5,6,15] In this patient, CT demonstrated extensive involvement of the left upper retroperitoneum, lesser sac, perirenal compartment and central mesentery, with close involvement of the distal pancreas, spleen and left kidney. These findings were important in planning the extent of surgical resection.

Fine-needle aspiration cytology in this case demonstrated an atypical adipocytic neoplasm; however, FNAC has limitations in accurately subclassifying lipomatous sarcomas because it

provides limited architectural information. Histopathological assessment by an experienced sarcoma pathologist, supplemented by molecular testing when appropriate, is important for accurate classification.[9,15]

Surgery remains the principal potentially curative treatment for localized retroperitoneal liposarcoma.[1-3] Current consensus recommendations emphasize management within specialist sarcoma centres and advocate complete macroscopic resection while maintaining tumour integrity.[12] En-bloc removal of adjacent organs is appropriate when they are directly involved by the tumour or cannot be safely separated without compromising oncological clearance.[1-4, 13] Piecemeal tumour removal or rupture should be avoided because residual microscopic or macroscopic disease may increase the risk of recurrence.[12]

The present case demonstrates the extent of surgery that may occasionally be required for a giant retroperitoneal liposarcoma. The tumour involved the distal pancreas, spleen, left kidney and adrenal gland, necessitating distal pancreatectomy, splenectomy, nephrectomy and adrenalectomy as part of the en-bloc resection. Although such multivisceral surgery carries significant perioperative morbidity, it may provide the best opportunity for complete macroscopic tumour clearance in carefully selected patients.[12,13]

The role of radiotherapy in retroperitoneal sarcoma remains individualized. The STRASS trial did not demonstrate a significant benefit from routine preoperative radiotherapy for the overall population of patients with primary retroperitoneal sarcoma. However, selective preoperative radiotherapy may be considered in patients with a high risk of local recurrence, particularly certain patients with WDLPS or low-grade dedifferentiated liposarcoma. Routine postoperative radiotherapy is generally limited by the risk of radiation-related toxicity to surrounding abdominal organs.

Systemic chemotherapy has a limited role in localized WDLPS because these tumours are relatively resistant to conventional cytotoxic therapy.[9] Chemotherapy is generally considered in selected patients with unresectable, recurrent or metastatic disease, particularly when a dedifferentiated or other high-grade component is present.[9] Therefore, surgery remains the primary treatment in a patient such as ours with a localized, resectable, low-grade WDLPS.

The large size of the tumour and extensive local involvement represent important risk factors for recurrence despite the favourable FNCLCC grade.[3,4,13] WDLPS is particularly associated with repeated local recurrence, and recurrence can occur many years after the initial operation.[4,9,14] Consequently, prolonged surveillance with periodic cross-sectional imaging is essential.[9, 14] Follow-up should be individualized according to tumour biology, surgical margins and institutional sarcoma protocols.

CONCLUSION

Giant retroperitoneal well-differentiated liposarcoma may remain clinically silent despite extensive local growth and involvement of multiple adjacent organs. Accurate preoperative imaging and multidisciplinary surgical planning are essential in determining resectability and the required extent of surgery.[5,6]

Complete macroscopic en-bloc resection remains the cornerstone of treatment for localized retroperitoneal liposarcoma.[1-3] When the tumour involves or is inseparable from adjacent organs, multivisceral resection may be required to achieve adequate oncological clearance.[1-4, 13] In the present case, en-bloc resection of the retroperitoneal tumour with distal pancreas, spleen, left kidney and adrenal gland resulted in satisfactory postoperative recovery.

Although the low FNCLCC grade indicates favourable tumour biology with a low risk of distant metastasis, the large tumour size and retroperitoneal location confer a significant risk of local recurrence.[3,4,9,13] Long-term clinical and radiological surveillance is therefore essential.

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