CASE REPORT

A giant retroperitoneal liposarcoma encasing vital organs: A case report

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Abstract

Liposarcoma is the most common primary malignant tumour of the retroperitoneum. As the retroperitoneum is a large space, these tumours can grow significantly before diagnosis. In this case, a 67-year-old male was admitted to a hospital with the complaint of weight loss and abdominal distension. A palpable mass was detected on physical examination, the A scan of this showed it to be retroperitoneal, heterogenous, with multiple septations, calcifications and solid components. It was removed successfully via midline incision. Liposarcoma is derived from adipose tissue and is a soft tissue sarcoma. In most cases, retroperitoneal liposarcoma presents as a painless mass, evident on physical examination. These tumours can go undetected for long periods, as symptoms usually arise only when the mass becomes significantly large. Symptoms include abdominal pain and constipation. Surgical resection is the mainstay of treatment for liposarcoma. Special care must be taken to preserve abdominal organs, which may be difficult considering the size of the mass, especially in resource-limited environments.

Keywords: Liposarcoma, retroperitoneal liposarcoma.

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Introduction

Liposarcoma is the most common soft tissue sarcoma, and up to 20% mesenchymal malignancies are liposarcomas.[1] These tumours can be classified into 3 subgroups: (1) well-differentiated liposarcoma (including adipocytic, sclerosing, inflammatory, spindle-cell, and dedifferentiated variants), (2) myxoid and round cell (poorly differentiated myxoid) liposarcoma (MLPS), and (3) pleomorphic liposarcoma (PLPS).[1] Among these, the two largest subgroups include well-differentiated (WDL) and dedifferentiated (DDL) liposarcomas.[2] These occur in middle-aged to older adults, and WDL can progress to DDL, which is known for its aggressive behaviour and poorer prognosis.[2]

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Giant retroperitoneal liposarcoma with a dimension of 30cm or more is extremely rare. Only 13 such cases are available in the PubMed database.[3] In this article, we discuss the case of a giant, retroperitoneal, dedifferentiated liposarcoma encasing the whole right kidney, measuring 56*51*25cm. Consent for publication was obtained from the patient and his family.

Case Report

A 67 years old male presented to the outpatient department of Jinnah Hospital Lahore on 22nd January 2024 with the presenting complaint of increasing abdominal girth for the past one year. It was associated with 20kg weight loss. The abdominal distension developed gradually and was associated with difficulty in daily life activities. On physical examination, the patient was vitally stable. He was alert and oriented to time, place and person and was lying comfortably on bed. Pallor and pedal oedema were visible. Abdominal examination showed gross distension and a firm, mobile mass was felt on palpation. Other systems were unremarkable. Baseline investigations were normal except severe anaemia. Ultrasound showed peritoneal lipomatosis more asymmetric towards right side. There were a few soft tissue echogenicity areas in the mesentry on the right side of the midline, the largest two measured 10.2 x 7.3 cm and 10.5 x 7.4 cm. These findings raised concern for a sarcomatous aetiology, and liposarcoma was suspected.

A CT scan revealed a large, irregular, retroperitoneal mass with heterogeneous enhancement. The mass was predominantly fatty in attenuation and measured approximately 40x26cm, located in the right hemiabdomen with an exophytic bulge. Multiple enhancing septations were noted, along with several solid components and one calcific focus.

The patient underwent an exploratory laparotomy and the resection of his tumour was done. The procedure was performed via a midline incision under general anaesthesia. The whole tumour mass was identified and mobilized. The right kidney was found completely encased by the tumour. During operation right sided renal pedicle was identified and ligated. The aorta and inferior vena cava were spared from the tumour and haemostasis was secured as shown in figure 1. After resection the abdomen was washed with warm saline and drain was placed. The

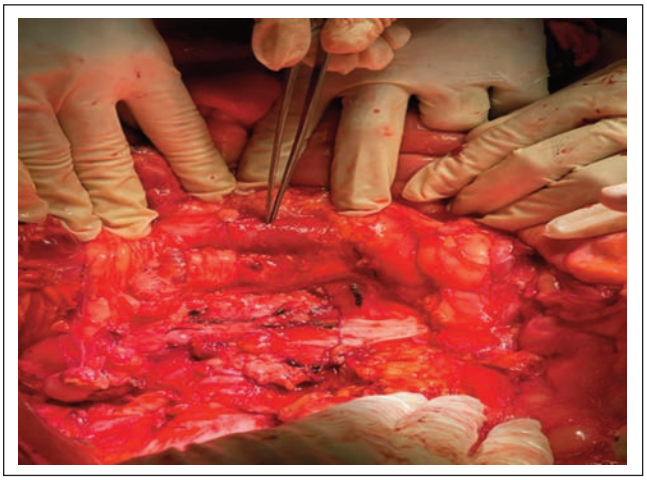

Figure-1: During surgery the aorta and inferior vena cava were spared from the tumour.

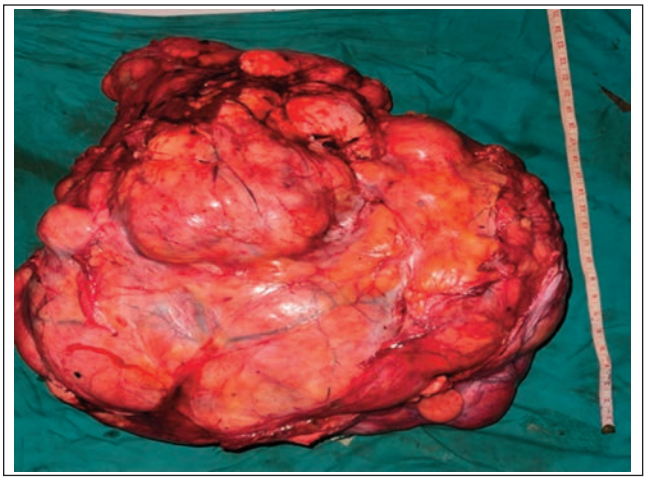

Figure-2: The gross specimen measuring 56x51x25 cm and 26kg in weight.

abdomen was closed with prolene 1/0 in a continuous fashion. The gross specimen measured 56x51x25 cm and weighed around 26kg as shown in figure 2. Histopathology of the specimen revealed malignant spindle cell neoplasm with prominent scattered pleomorphic cells, tumour giant cells and well differentiated adipose tissue at the periphery. The diagnosis was dedifferentiated liposarcoma and the soft tissue grading system, French National Federation of Cancer Centers (FNCLCC) showed grade 2 tumour. The patient was discharged on seventh post-operative day. However, he presented to the Emergency Department in shock on 21st post op day and despite all conservative management, he expired on the 22nd post op day.

Discussion

Well-differentiated (WDL) and dedifferentiated (DDL) liposarcomas together comprise the two largest subgroups of liposarcoma. Up to 10% of cases of WDL may progress to DDL, which have higher malignant potential and aggressive behaviour, leading to poorer prognosis.[2]

Several cases of giant liposarcomas have been described in the literature, for example, Xu C et al.[3] reported a case with dimensions of 37x32x26.5 cm, weighing 21 kilograms. Eren F et al.[4] presented a case of DDL measuring 40x30x6 cm, which had perforated the first part of the duodenum. A case report by Sung Don Oh et al.[5] described a liposarcoma which was 45 cm in its largest dimension and encompassed the entire left kidney. We have reported a case of DDL measuring 56x51x25 cm, weighing 26 kilograms, making it one of the largest known cases.

As discussed above, dedifferentiated liposarcomas, often develop in deep anatomical locations, such as the retroperitoneum or mediastinum. Although surgery is the main mode of treatment, complete excision can be challenging due to the tumour's tendency to encase vital structures. Radiotherapy may often be needed, especially in the radiosensitive myxoid/round-cell (MLPS) subtype.6 For unresectable or advanced tumours, chemotherapy may be considered, with doxorubicin and ifosfamide being the first-line drugs.[6]

For each subtype, prognosis depends on the grade and metastatic involvement. Successful complete resection of retroperitoneal liposarcoma may increase the 5-year survival rate from 16.7% to 58%.[7] To monitor recurrence, a follow-up CT scan every 3 months for the first 2 years, every 6 months for 2–5 years and annually thereafter is generally recommended.[7] Overall, liposarcomas can be deadly and each of its subtypes should be treated as different entities, with variable treatment options and prognoses.

Conclusion

Giant liposarcoma is a rare condition for which surgical management is the mainstay of treatment. Preoperative diagnosis is imperative in the optimal treatment of this condition. Adjuvant therapy may be needed to downstage the tumour in case of metastasis before surgery can be performed. Removal of the adjacent structures, such as a kidney or a part of the intestines may be necessary for complete resection of the lesion. Care must be taken to preserve organ function and subsequently, the quality of life of the patient.

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Author Contribution:

AA: Idea, drafting and reviewing.
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FM: Drafting, references and added citations.
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