CASE REPORT

Retroperitoneal liposarcoma: Case of a giant tumour inside the abdomen

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Abstract

Retroperitoneal liposarcomas are rare malignant tumours that often grow rapidly in size and become symptomatic late in the disease course, posing diagnostic and therapeutic challenges. Although abdominal imaging can reliably diagnose the tumour, definitive diagnosis is only possible through biopsy after surgical excision, which remains the primary treatment modality for these tumours. Long-term survival is primarily determined through histologic grade and post-resection tumour margins. We report the case of a 43-year-old male patient, seen at Dr Ruth KM Pfau Hospital Karachi, who underwent successful surgical excision for a well-differentiated retroperitoneal liposarcoma and had no radiographic evidence of local recurrence at both 3 and 12-month follow-ups.

Keywords: Retroperitoneal liposarcoma, surgical excision, tumour recurrence, tumour grade.

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Introduction

Liposarcomas are a specific type of soft tissue sarcomas, arising from the neoplastic growth of fat cells.[1] They can arise from fat cells anywhere in the body but have a predilection for the extremities.[2] The retroperitoneal origin of liposarcomas is less common, carries a poor prognosis, and poses diagnostic challenges due to two main factors: rarity and asymptomatic clinical course.[3] Due to abundant anatomic space, the tumour's locally invasive nature allows it to grow rapidly and symptoms do not typically manifest until the tumour reaches a large size or compresses other organs.[2] Once suspected on clinical grounds, imaging is a useful modality for diagnosis but biopsy remains the gold standard.[4] Like all soft tissue sarcomas, surgical excision is the mainstay of management, and other treatment modalities can be tailored according to individual cases.[3] Histologic grade and tumour margins after resection are the main prognostic indicators and determine the long-term survival of patients.[5]

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We present a case report of primary retroperitoneal liposarcoma in a 43-year-old male patient, seen in June, 2022, who was successfully treated with surgical excision. The aim of this report is centred around a discussion of the epidemiology, disease course, and management of primary retroperitoneal liposarcomas, along with a comparison of existing literature.

Case Report

Our case surrounds a 43-year-old male patient who was referred to our Surgical-IV department, Dr Ruth KM Pfau Hospital Karachi on 18th of May, 2022 with complaints of vomiting, abdominal distension/fullness, and dull abdominal pain for the past 3 months. His symptoms were progressive in nature and the chief concern was a massively distended abdomen, which was rapidly increasing in size over time.

Abdominal examination revealed a distended abdomen, with a centrally placed, inverted umbilicus, and an ill-defined mass approximately 30 x 20 cm in size, the major bulk of which was unilateral on the right side. On palpation, the mass was firm, non-tender, non-compressible, and slightly mobile along both the horizontal and vertical planes of the abdomen. No peripheral stigmata of chronic liver disease were noted. There was associated right-sided testicular swelling, consistent with a varicocoele on palpation.

Haematological and biochemical investigations were unremarkable. An ultrasound of the abdomen was first performed which demonstrated a huge, complex heterogeneous area at the epigastric region with multiple echogenic internal echoes. A subsequent whole-body CT scan with I/V contrast revealed a large, well-defined, heterogeneously enhancing retroperitoneal mass, measuring 26x16x20 cm in craniocaudal, transverse, and anteroposterior dimensions respectively. Mass effect without any direct infiltration was observed in the surrounding viscera (Figure 1). A few internal areas of fat attenuation (-79.83 HU) were observed along with tiny foci of calcification. Compression of the right testicular vein was also noted.

The combined clinical and radiographic picture suggested a pre-emptive diagnosis of retroperitoneal liposarcoma, and a surgical excision was planned as a diagnostic and

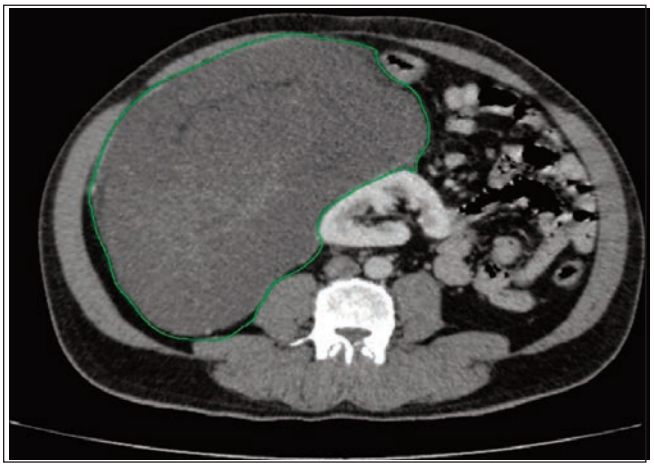

Figure-1: Axial-CT scan displaying a-giant tumour arising in the retroperitoneum (green overlay) with compressive mass effect on the right kidney.

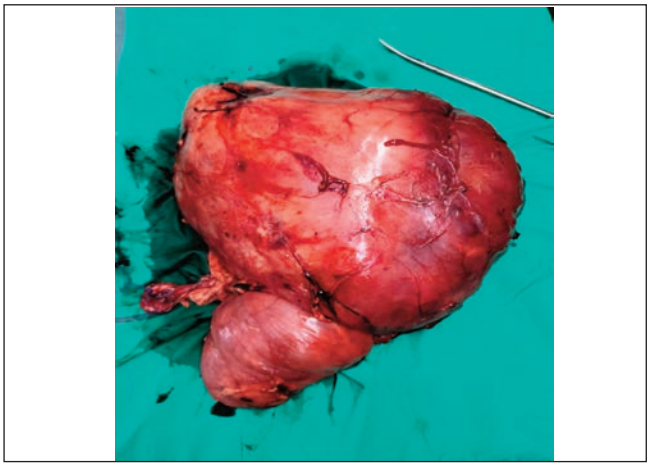

Figure-2: Excised retroperitoneal mass.

therapeutic strategy. A midline incision was made, and the mass was dissected and excised along its outer planes, carefully preserving the adjacent structures such as the right kidney's upper pole and avoiding auxiliary damage. The excised mass measured around 34x16x10 cm (Figure 2) and was sent for histopathological analysis. The result was positive for a well-differentiated liposarcoma containing chondroid, osseous, and rhabdomyoblastic elements. The background was primarily myxoid and sclerotic. Lipoblasts and atypical stromal cells were present both in the background and the vessel walls. Tumour margins were positive (<0.1 cm from the surface), and the patient was referred to our oncology department. Post-operative CT scans 3 and 12-month follow-ups did not demonstrate any evidence of tumour recurrence.

Discussion

Liposarcomas are the most common type of soft tissue sarcomas arising in the retroperitoneum.[6] About 35% of retroperitoneal liposarcomas originate from perirenal fat.[7] There is a predilection for the age group of 60-70 years, with males affected a little more than females.[5]

Besides the origin and location, another way to classify liposarcomas, is their histological features. Liposarcomas are broadly grouped into 4 histological categories namely:

Well-differentiated (further classified into lipomatous, sclerotic, and inflammatory).

De-differentiated

Pleomorphic

Myxoid/round cell

These histological subtypes have grades that are important prognostically. Well-differentiated tumours are typically low grade whereas dedifferentiated and pleomorphic tumours are classically high-grade. The middle continuum is formed by the mixed/round cell subtype which can be high-grade (round cell>5%) or low-grade (round cell<5 %). High-grade variants exhibit higher metastatic potential and increased local recurrence rates.[8] In the case of this patient, the tumour analysis indicated a well-differentiated subtype, which meant a good prognosis histologically. Tumours of similar grade have been previously described by Evola et al,[9] and Wang et al.[7]

CT imaging has a role in both assessment and grading of liposarcomas. Well-differentiated tumours tend to display a high attenuation of fat and minimal soft tissue components, calcifications, and nodular enhancing septations. Such tumours do not display the typical malignant characteristics of invasion and infiltration.[4] Similar findings were seen in this case, although unlike Evola et al,[9] areas of fatty attenuation were significantly lesser, an unusual finding in the context of the tumour's grade.

Surgical excision remains the primary mode of treatment for retroperitoneal liposarcomas, but the chief concern is that of local recurrence, which contributes to overall mortality.[5] Secondary removal of the recurrent tumour can be more challenging, due to the altered anatomical milieu caused by the primary surgery. There was failure to achieve negative microscopic margins in this case, which is associated with worse local recurrence and overall survival rates.[4] We did, however, manage to preserve other abdominal viscera and combined resection was not deemed necessary.

Local recurrence rates in well-differentiated tumours have been reported to be around 24% in a systematic review.[5] The review also suggests that positive tumour margins have a decreased overall survival and increased local recurrence rates in comparison to negative tumour

margins. Despite the failure to achieve a negative microscopic margin, we did not find radiographic evidence of early recurrence (<12 months) in this case.

Conclusions

Retroperitoneal liposarcomas are a rare clinical entity, with high rates of recurrence after surgical removal, which forms the mainstay of the treatment. Our case is among the few reported in our country, and most findings correlate with existing literature. Tumour recurrence is a common and important prognostic feature, and we did not find any radiographic evidence of local recurrence, which is a remarkable surgical outcome for retroperitoneal liposarcomas.

Informed consent: The patient gave a written informed consent for the publication of this case report.

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MAK: Study design, concept, literature search, data interpretation, writing the manuscript.
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