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Giant retroperitoneal liposarcomas presenting as potbelly: A rare mimicker and surgical challenge

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Abstract
Retroperitoneal liposarcoma (RPLPS) is a rare malignant mesenchymal tumor arising from adipose tissue, often presenting late due to its deep anatomical location and indolent growth. We report two cases of large, well-differentiated retroperitoneal liposarcoma (WD-RPLPS) in young male patients aged 47 and 36 years, respectively. Both presented with abdominal distension and imaging revealed giant fat-density retroperitoneal masses measuring 35×30×25 cm and 30×28×17 cm. One patient underwent PET-CT evaluation, demonstrating mild-to-moderate heterogeneous FDG uptake within a predominantly fatty lesion. Both patients underwent complete surgical excision under general anesthesia. Gross and microscopic findings demonstrated mature adipocytes with atypical spindle cells and fibrous septations, while immunohistochemistry confirmed strong nuclear MDM2 positivity, consistent with well-differentiated retroperitoneal liposarcoma (WD-RPLPS). Postoperative recovery was uneventful in both cases. These cases highlight the diagnostic importance of advanced imaging and confirmatory immunohistopathology in identifying WD-RPLPS. Complete surgical resection remains the cornerstone of curative treatment, particularly given the tumor's tendency to attain massive size prior to detection. Early recognition and multidisciplinary management are vital to optimize outcomes in this uncommon malignancy.

Keywords: Well-differentiated retroperitoneal liposarcoma, surgical excision, immunohistochemistry, murine double minute 2 (MDM2)

Introduction
Liposarcoma, commonly referred to as soft tissue sarcoma (STS), is the most prevalent subtype of sarcomas that arise in the retroperitoneum, accounting for several no. of all cases [1]. Liposarcomas mostly originates from adipocytes, which are fat cells and it is predominantly found in the extremities (52%) followed by the retroperitoneum (12-40%) and perirenal fat (35%) [2]. Retroperitoneal liposarcomas (RPLPS) are uncommon primary malignant mesenchymal tumors that arise from adipose cells located in the retroperitoneal area and these tumors are still tough to diagnose and treat [3]. The identification of RPLPS is largely dependent on pathological findings, with computed tomography (CT) or magnetic resonance imaging (MRI) being crucial in the majority of presurgical diagnoses [4]. Due to its position in the retroperitoneal space and indolent growth pattern, the disease often presents at an advanced stage with a large mass, making surgical management technically challenging for both the surgeon and the patient [5].
Here we present a two rare cases of male patients diagnosed with retroperitoneal liposarcoma, treated by undergoing retroperitoneal liposarcoma excision under general anesthesia.

Case Report
Case 1
A 47-year-old male presented with complaints of abdominal pain and progressive distension for one month (Figure 1). He was admitted to our department for further evaluation and management. There was no relevant family history of malignancy or other comorbid conditions. On clinical examination, the patient's vital parameters were stable, and baseline laboratory investigations were within normal limits.
A whole-body positron emission tomography-computed tomography (PET-CT) scan was performed from the vertex to the mid-thigh following intravenous administration of 7.5 mCi of 18F-fluorodeoxyglucose (FDG), with contrast-enhanced CT images acquired subsequently.

No abnormal metabolic activity was observed in the brain, head and neck, chest, or skeletal system. However, imaging of the abdomen and pelvis demonstrated a large, oval, fat-density mass with smooth, lobulated margins and internal septations occupying much of the abdominal and pelvic cavities. Multiple ill-defined enhancing areas of soft tissue with mild-to-moderate heterogeneous FDG uptake were noted within the lesion. It originated from the left retroperitoneum, extending across the midline into the right lower quadrant. The mass caused a significant mass effect, compressing and displacing adjacent structures-pushing the left kidney and colon medially, compressing the pancreas and stomach superiorly, and exerting downward pressure on the urinary bladder. The lesion measured approximately 35×30×25 cm, with moderate FDG uptake in its solid upper portion (Figure 2).

After completing all preoperative assessments, the patient underwent surgical excision of the retroperitoneal mass under general anesthesia with epidural analgesia. Intraoperatively, mesenteric structures were uninvolved. The mass, measuring 35×30×25 cm and weighs 11.45 kg and it was completely mobilized and removed along with its capsule (Figure 3). The left ureter was displaced posteriorly, and the vascular supply from the left common iliac artery was ligated. Hemostasis was achieved, a drain was placed and the wound was closed in layers. The postoperative recovery was uneventful.

Gross pathological examination revealed a pseudo capsulated, yellow, soft, fatty mass with focal cystic and myxoid changes. Microscopy showed mature adipocytes in a fibrous stroma with scattered atypical spindle cells, multinucleated stromal elements, and chronic inflammatory infiltrates. Immunohistochemistry demonstrated MDM2 positivity in atypical spindle cells, confirming the diagnosis of well-differentiated retroperitoneal liposarcoma (WDLS) (Figure 4). The tumor was negative for CD99, LCA, Desmin, NKX2.2, Sall4, pan CK and S100, with preserved INI-1 expression and a MIB-1 proliferation index of 20-30%. The patient recovered well and was discharged in stable condition on prescribed medications.

Case 2

A 36-years-old male presented with complaints of lump in abdomen not associated with pain, not associated with bowel and bladder symptoms (Figure 5). Upon physical examination, the baseline vital signs were found to be stable and all laboratory investigations fell within normal ranges. Contrast-enhanced computed tomography (CECT) scan of the abdomen and pelvis demonstrated a large, well-defined, oval, fat-density mass with lobulated borders and internal septations extending throughout the abdominal and pelvic cavities. The mass appeared to originate from the retroperitoneum, mainly occupying the left upper and lower quadrants, crossing the midline and extending into the right lower quadrant. It exerted a significant mass effect, compressing the left hemi colon and kidney, displacing the small bowel and other peritoneal organs to the right and causing mild superior indentation of the pancreas and stomach, as well as inferior pressure on the urinary bladder. The lesion measured approximately 30×28×17 cm and occupied as solid upper region (Figure 6).

After completion of preoperative evaluation, the patient underwent surgical excision of the retroperitoneal mass under general anesthesia with epidural analgesia. Intraoperatively, mesenteric structures were preserved. A solid retroperitoneal mass measuring 30×28×17 cm and weighing 9.5 kg, consistent with liposarcoma, was completely resected. The vascular supply arising from the left common iliac artery was ligated. The mass was carefully mobilized circumferentially and excised along with its capsule (Figure 7). Hemostasis was achieved, a drain was positioned and the incision was closed in layers.

The resected specimen was sent for histopathological evaluation. Gross examination revealed a pseudo capsulated mass with a yellow, soft, fatty cut surface showing focal cystic and myxoid-like degeneration. Microscopy sections A-M, MS1-MS13 demonstrated variably sized adipocytes within thickened fibrous septa, scattered atypical spindle cells and occasional multinucleated stromal cells. Areas of chronic inflammation, hemosiderin-laden macrophages, infarction, calcification and fat necrosis were also noted, suggestive of well-differentiated retroperitoneal liposarcoma. Immunohistochemistry revealed MDM2 positivity in a subset of spindle cells, consistent with well-differentiated liposarcoma (WDLS), (Figure 8) and was negative for CD99, LCA, Desmin, NKX2.2, Sall4, pan-CK and S100. Finally the patient recovered sucessfully and was discharged in haemodynamically stable condition with medications.

Discussion

Liposarcoma (PLS) is an uncommon cancer, representing 0.07% to 0.2% of all tumors, with around 12% to 40% found in retroperitoneal area. In these type of the cases, majority of patients are aged below 60 years [6]. In this case report, one patient was around 47 years, reported abdominal pain and distension for one month and another patient was 36 years presented with complaints of lump in abdomen not associated with pain. Retroperitoneal liposarcoma (RLPS) is often asymptomatic in its early stages. As the tumor enlarges, patients may develop abdominal pain, distension, discomfort or a palpable mass, these symptoms generally related to tumor size and compression of surrounding structures [7].

Computed tomography (CT) and magnetic resonance imaging (MRI) are the conventional modalities used for diagnosing, staging and monitoring retroperitoneal liposarcoma (RPLS); however, they provide limited information about tumor biology. Conversely, 18F-fluorodeoxyglucose positron emission tomography combined with computed tomography (18F-FDG PET/CT) integrates anatomical detail with metabolic data, offering enhanced insight into the evaluation of well-differentiated liposarcoma [8]. In our case, one patient underwent a whole-body PET-CT scan (vertex to mid-thigh) after receiving 7.5 mCi of 18F-FDG, revealing a large, fat-density, oval-shaped mass in abdominal and pelvic measuring approximately 35×30×25 cm and the second patient underwent contrast-enhanced computed tomography (CECT) in which 30×28×17 cm fat-density, oval-shaped mass was observed in abdominal and pelvic region.

Complete surgical excision continues to be the definitive and potentially curative treatment for retroperitoneal liposarcoma [9]. In this case report, two patients underwent surgical excision of retroperitoneal masses under general anaesthesia. In two patients the solid masses arising from the left retroperitoneum was identified and confirmed as well-differentiated retroperitoneal liposarcoma. Similarly,

Fiaschetti V *et al*. described a 2 cases of well-differentiated retroperitoneal liposarcoma that occupied one side of the abdomen. The tumor was successfully removed through surgical approach [10].

In both patients, immunohistochemistry of the resected specimens demonstrated MDM2 positivity. In the second case, additionally the biopsy confirmed a well-differentiated liposarcoma with an FNCLCC grade of G1, finally a well-differentiated retroperitoneal liposarcoma was confirmed in both cases. Well-differentiated retroperitoneal liposarcomas are typically indolent, low-grade tumors characterized by slow progression. Due to their gradual growth and deep retroperitoneal location, they often reach substantial size before becoming clinically apparent, which can complicate surgical management and increase operative risk [11]. According to the world health organization (WHO) histological classification, liposarcomas are categorized into five major types like well-differentiated (also termed atypical lipomatous tumor), dedifferentiated, myxoid, round cell, and pleomorphic variants. In the updated 2020 WHO classification, this system was simplified into four principal subtypes like atypical lipomatous tumor/well-differentiated liposarcoma, dedifferentiated liposarcoma, myxoid liposarcoma and pleomorphic liposarcoma [12].

Biopsy remains the gold-standard method for confirming well-differentiated liposarcoma (WDLPS), particularly when evaluating retroperitoneal masses. Immunohistochemistry plays a critical role by identifying characteristic protein markers within the sampled tissue, thereby supporting the diagnosis and distinguishing WDLPS from other morphologically similar lesions [13]. Murine double minute 2 (MDM2) is an oncogene that is commonly amplified in well-differentiated retroperitoneal liposarcoma (WDLS), The amplification of MDM2 represents a significant genetic change in WDLS, often regarded as one of the initial occurrences in tumor progression. It is involved in the regulation of the cell cycle by marking the tumor suppressor protein p53 for degradation, which results in uncontrolled cell growth [14].

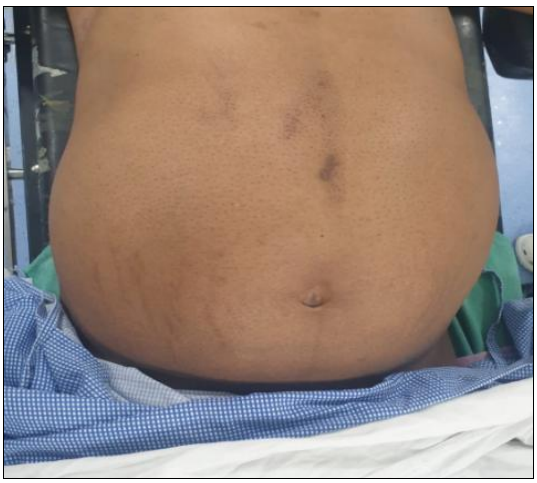

Fig 1: Frontal view of the abdomen highlighting the significant distension caused by the retroperitoneal tumor.

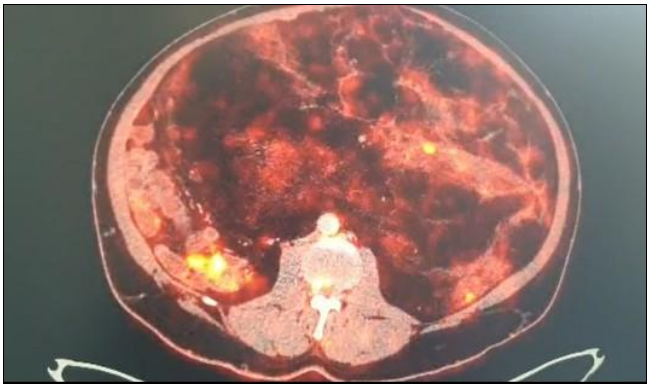

Fig 2: PET-CT scan of abdomen and pelvis revealed a large, oval-shaped, fat-density mass lesion around 35 × 30 × 25 cm.

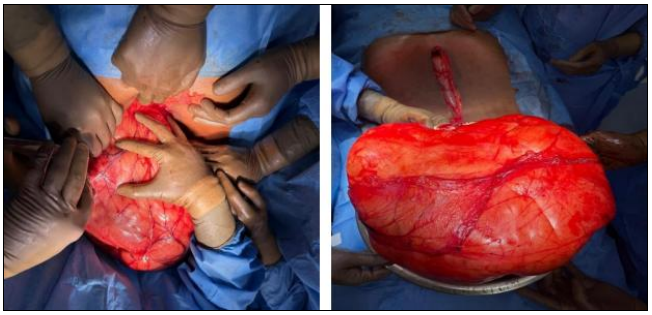

Fig 3: Intraoperative image showing dissection of a large encapsulated retroperitoneal tumor, along with the gross appearance of the excised mass displaying its size and prominent vascularity.

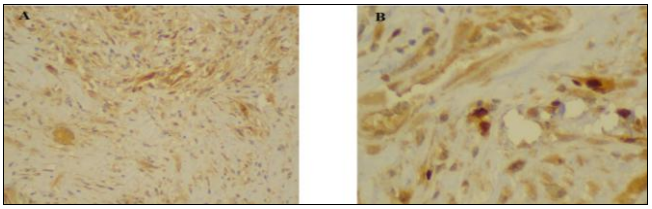

Fig 4: Immunohistochemical staining for MDM2 reveals nuclear positivity in tumor cells at (A) 20× and (B) 40× magnification, which supports the diagnosis of well-differentiated liposarcoma.

Fig 5: Clinical image showing a markedly distended abdomen with visible abdominal wall stretching and striae.

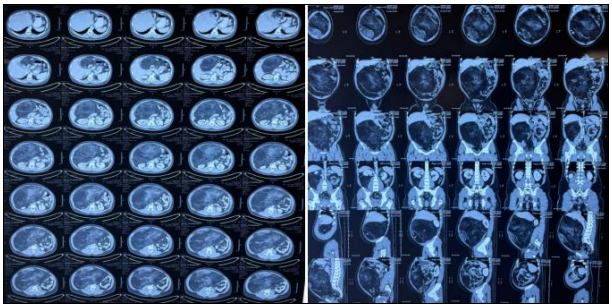

Fig 6: Contrast-enhanced CT scan of the abdomen demonstrating a large heterogeneous retroperitoneal mass with fat density components and internal septations, displacing surrounding visceral structures, suggestive of well-differentiated liposarcoma.

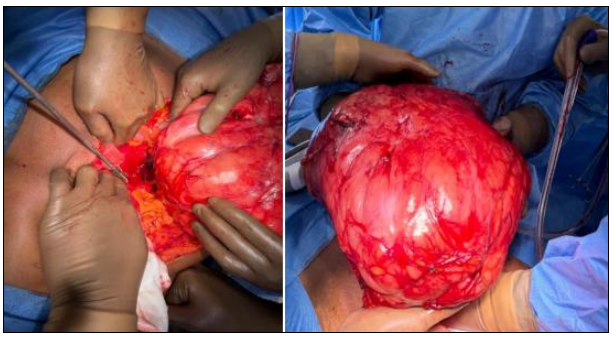

Fig 7: Intraoperative view of demonstrating careful dissection along the tumor capsule to separate it from surrounding structures and mobilization of the retroperitoneal tumor, emphasizing surgical technique for tumor resection.

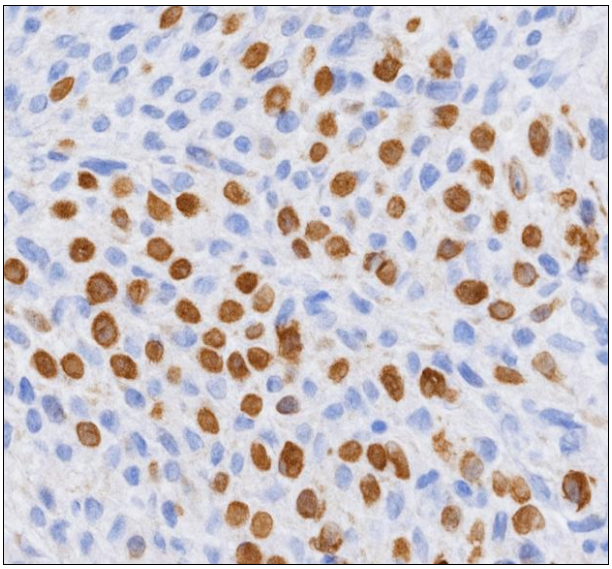

Fig 8: Immunohistochemical staining demonstrating MDM2 nuclear positivity in well-differentiated retroperitoneal liposarcoma, showing scattered tumor cell nuclei with strong brown nuclear staining for MDM2.

Conclusion

Retroperitoneal liposarcoma (RPLPS) is a rare tumor that often remains unnoticed until it attains a significant size, due to its deep-seated retroperitoneal location and slow progression. These two cases highlights the importance of thorough assessment, which includes contrast-enhanced CT and 18F-FDG PET/CT, to accurately delineate tumor burden and features. Surgical removal is the primary and most definitive approach for treating RPLPS. The combination of radiological insights with histopathology, tissue biopsy and immunohistochemical evaluation especially with markers like MDM2 enhances diagnostic certainty and assists in differentiating this tumor from other retroperitoneal tumors, ultimately promoting timely and effective treatment.

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Declarations

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Ethical approval: Not required.

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