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Retroperitoneal Sarcoma: Two Case Reports

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Abstract: Case Report

Retroperitoneal liposarcoma is a rare mesenchymal tumor of the retroperitoneum, considered a diagnostic challenge due to the wide variety of clinical presentations, none of which are specific to it. Imaging can be predictive of its presence, but the definitive examination is anatomopathological with the existence of several subtypes impacting the prognosis. Complete resection remains the reference treatment even with a considerable risk of recurrence.

Keywords: Retroperitoneal Liposarcoma, Mesenchymal Tumor, Diagnostic Challenge, Imaging and Histopathology, Surgical Resection and Recurrence.

INTRODUCTION

Liposarcoma is a tumor originating from primitive mesenchymal cells differentiated into adipocytes, accounting for only 10% of soft tissue sarcomas and 15% of all sarcomas [1]. The most frequent location is the soft tissues of the limbs, but it can also be present in the abdominal region, particularly in the esophagus, stomach, and descending mesocolon, as well as the retroperitoneum [2, 3]. The latter is considered a rare location.

Although retroperitoneal liposarcomas can occur at any age, their peak incidence is around the sixth and seventh decades of life, with no gender or racial predilection [4].

Many cases are asymptomatic due to the vastness of the retroperitoneal space. Apart from incidental findings on imaging, clinical signs are not specific, namely lumbar pain, abdominal pain, those related to neighboring compression such as urinary signs related to ureteral compression or digestive signs related to colonic or small bowel compression [5].

The diagnosis mainly depends on the anatomopathological study, but imaging always retains its diagnostic place before the surgical act [6], particularly computed tomography or magnetic resonance imaging [7, 8]. The presence of a fatty component points toward the diagnosis.

After diagnosis, surgery is the recommended basic treatment for retroperitoneal liposarcoma; however, the extent of resection remains controversial [9].

The prognosis of retroperitoneal liposarcoma will obviously depend on the surgical approach and the histological subtype. The latter is associated with the site of the primary tumor. Due to the high rate of tumor

recurrence and the invasive nature of the tumor, secondary surgery, radio-chemotherapy, or targeted therapy may be necessary [10].

OBSERVATION

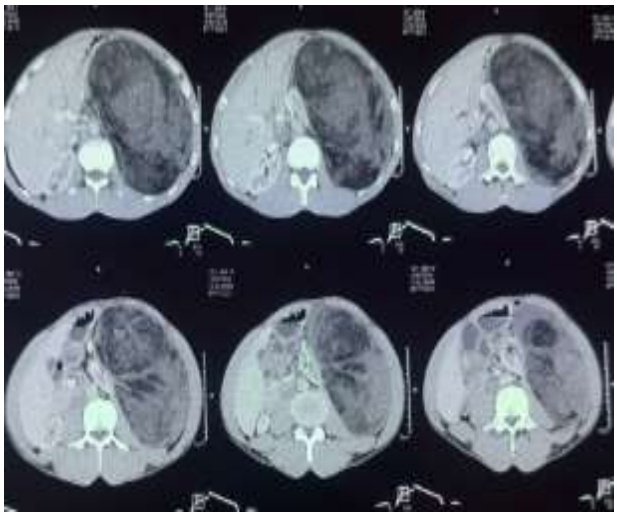
The first case:

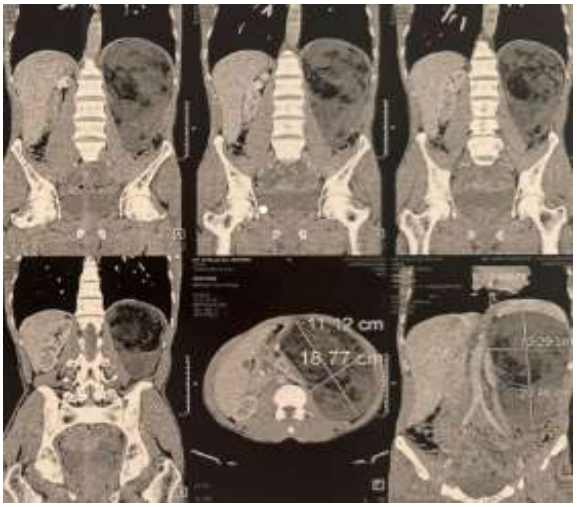
A 52-year-old patient, with no particular medical history, whose symptoms dated back to 1 year before his consultation with the appearance of an abdominal mass gradually increasing in size, causing him intermittent heaviness-type pain accentuated in the left flank and left lumbar fossa, without aggravating or relieving factors, without other digestive signs, particularly obstructive or extradigestive ones (neurological, muscular, or urinary), in a context of apyrexia and deterioration of general condition characterized by asthenia, anorexia, and unspecified weight loss.

At the clinical examination: the patient had a WHO score of 1, a BMI of 21, with the abdominal examination revealing the perception of an abdominal mass on the left flank, left lumbar fossa reaching the epigastrium, partially mobile, measuring approximately 20*20cm, without inflammatory signs in the area, of hard consistency, with the upper limit extending beyond the left costal margin. The examination of the other systems is normal, including that of the lymph node areas.

The patient underwent a thoraco-abdomino-pelvic CT scan which revealed the presence of a large left retroperitoneal mass, well-defined, oval in shape, heterogeneous, with a dual fleshy and fatty component measuring 20*19*12 cm in diameter.

This mass pushes the left kidney and adrenal gland inward and the pancreatic tail forward, with respect for the separation rim, all of which primarily suggests a retroperitoneal liposarcoma, without secondary locations.

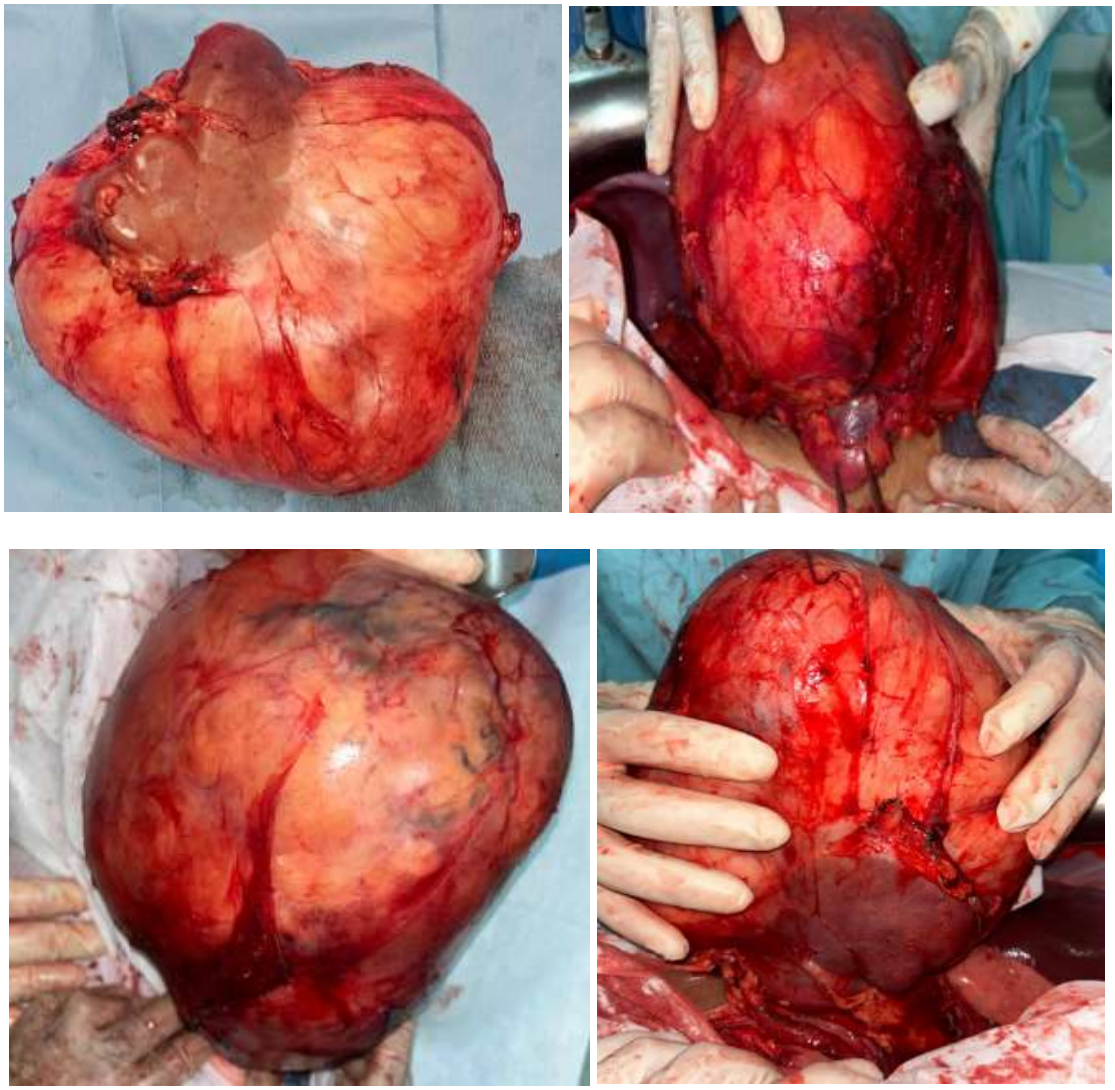




The biological assessment did not reveal any abnormalities, notably no inflammatory syndrome, anemia, or hypoproteinemia. The tumor markers were negative.

The patient was operated on in our institution with the exploration revealing: an enormous retroperitoneal mass originating from the left kidney, measuring approximately 35*30 cm, displacing the spleen as well as the tail of the pancreas, being cleavable.

The procedure involved a resection of the mass en bloc, including the left kidney. The postoperative course was straightforward except for the postoperative pain managed by analgesics. diuresis was maintained without notable complications.



The anatomopathological examination of the specimen had macroscopically identified an encapsulated, intact tissue mass weighing 3.5 kg and measuring 37*35*14cm. The left kidney was adherent to this mass.

The overall appearance was in favor of a grade 2 pleomorphic retroperitoneal liposarcoma according to the FNCLCC, with an estimated 20% presence of tumor necrosis, without vascular emboli. The surgical resection margins are clear, remaining 1 mm from the tumor capsule, which is intact. The tumor remained 1 mm from the renal capsule with a separation rim, and the ureteral and renal vascular margins were clear.

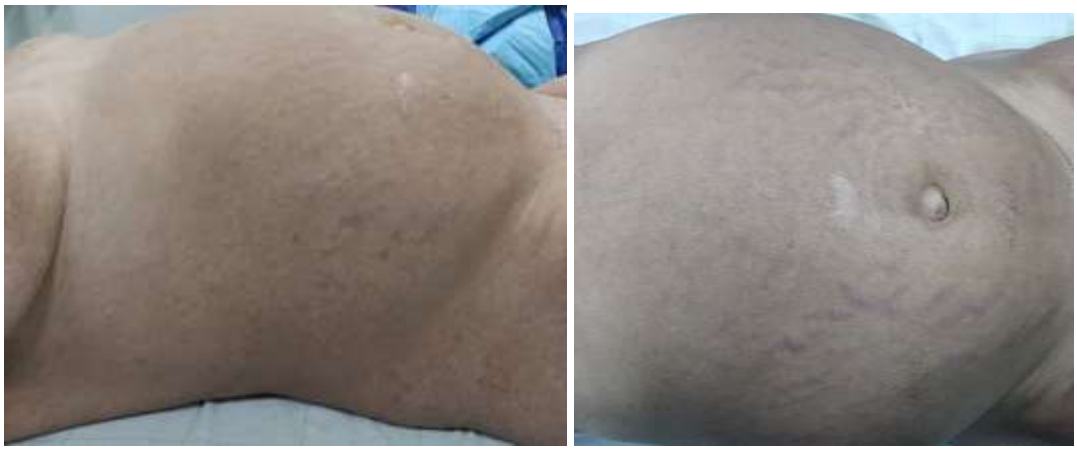
The proposed pTNM stage is: pT4Nx Mx (according to the AJCC, 8th edition). The patient was kept under surveillance with simple follow-up without notable tumor recurrence.

The 2nd case:

This is a 36-year-old patient, who had a cesarean section 2 years ago, with no other notable medical history, who presented with an abdominal swelling that had been developing for 18 months, located on the right flank extending to the right iliac fossa, with progressive enlargement, causing a heavy type of pain with intermittent food vomiting and chronic constipation without digestive hemorrhage or extra-digestive signs in a context of afebrile condition and deterioration of general health.

The clinical examination found a patient in an altered general condition with signs of malnutrition.

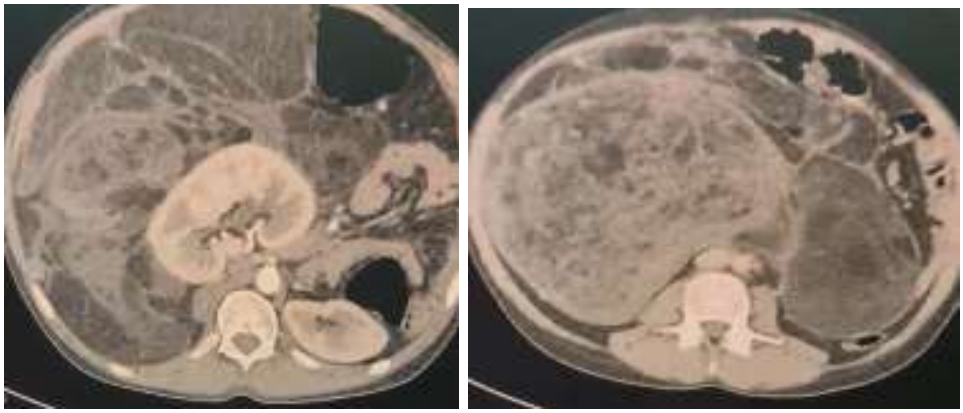
Her WHO score was 2 and her BMI was 17. The abdominal examination revealed fire-needle scar and Pfannenstiel incision, as well as the presence of a mass occupying the right flank, the right iliac fossa extending toward the right hypochondrium with lumbar contact. The rest of the clinical examination was unremarkable.



The patient underwent an abdominal CT scan, which demonstrated a large multiloculated retroperitoneal mass containing both solid and fatty components, with pseudocystic areas, measuring 20 × 24 × 33 cm. The mass encases the right kidney and its vascular pedicle, displacing them toward the midline.

Topographically, the mass is anteriorly and laterally in contact with the right anterior and lateral abdominal wall, and posteriorly abuts the psoas muscle without a clear fat plane. Medially, it contacts the subrenal abdominal aorta and the inferior vena cava, the latter remaining well opacified. The mass also displaces the intestinal loops and colonic frame and is in contact with the duodenopancreatic block.

Superiorly, it reaches the liver, and inferiorly, it extends toward the bladder. The overall radiologic appearance is highly suggestive of a retroperitoneal liposarcoma.

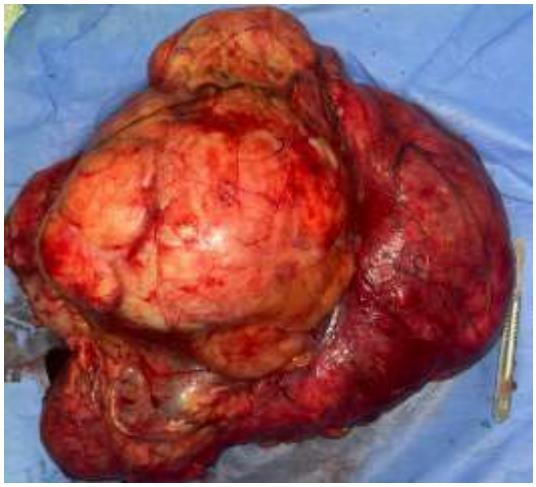


The preoperative staging assessment did not demonstrate any secondary lesions in the hepatic, osseous, or thoracic regions. Serum tumor markers were within normal limits.

The patient subsequently underwent surgical management in our department. Intraoperative exploration revealed a massive, multilobulated solid–cystic retroperitoneal mass measuring approximately 40 × 50 cm.

The tumor displaced the right colon and duodenum, and completely enveloped the right kidney along with its vascular pedicle, as well as the right fallopian tube, with no identifiable dissection plane. Superiorly, the tumor extended to the level of the inferior vena cava, where a distinct cleavage plane was present. No peritoneal effusion, hepatic metastases, or peritoneal carcinomatosis nodules were observed.

The procedure involved a monobloc resection of the mass, including the right kidney and the right fallopian tube without any breach.



The postoperative course was simple without complications. At the anatomopathological examination:

The macroscopic examination revealed a piece weighing 7050 g, measuring 45*40*12cm, encompassing the right kidney and the right fallopian tube.

The microscopic examination concluded with a well-differentiated liposarcoma, lipoma-like grade I (low grade). The postoperative course was straightforward without tumor recurrence.

DISCUSSION

Retroperitoneal liposarcoma (RPLPS) is a malignant tumor that develops in the retroperitoneal space from mesenchymal tissue [11], particularly in perirenal adipose tissue [12], accounting for only 0.1% of all adult cancers [13].

Based on morphological characteristics and cytogenetic abnormalities, liposarcomas are classified into five types [14, 15], each presenting specific genetic mutations:

1. Well-differentiated liposarcoma;
2. Dedifferentiated liposarcoma;
3. Myxoid/round cell liposarcoma;
4. Pleomorphic liposarcoma;
5. And, more recently, myxoid-pleomorphic liposarcoma.

Well-differentiated liposarcoma is the most common subtype, generally characterized by slow growth but with a high rate of local recurrence and resistance to both radiotherapy and chemotherapy. Dedifferentiated liposarcoma represents a more aggressive progression of the former, marked by greater invasiveness, a higher likelihood of metastasis, and an overall poorer prognosis [16, 17].

Pleomorphic liposarcoma is the most aggressive subtype, exhibiting a complex karyotype and marked genetic heterogeneity, which explains its unfavorable prognosis [18].

The diagnosis of retroperitoneal liposarcoma is particularly challenging due to the wide range of nonspecific clinical signs, or because it is discovered incidentally during routine clinical examination or imaging performed for unrelated reasons. In the two cases we reported, both patients presented with progressive abdominal distension associated with intermittent pain, which delayed medical consultation, especially in the absence of other specific symptoms.

Local invasion or compression of adjacent organs accounts for the polymorphic clinical presentation, while the large retroperitoneal space explains the delayed onset of symptoms.

Computed tomography (CT) is the reference imaging modality for diagnosing

retroperitoneal liposarcoma, as it allows precise characterization of the tumor, its size, exact location, and anatomical relationships with adjacent organs [19, 20]. It is considered both a diagnostic and a surveillance tool.

In the two cases reported, CT imaging was sufficient for diagnosis, clearly identifying the tumor location, size, and relationships with surrounding structures.

Magnetic resonance imaging (MRI) may also be used for diagnostic purposes, as it provides improved delineation of tumor margins and better assessment of invasion of adjacent organs [21].

Histological subtypes can be differentiated based on imaging characteristics, but the definitive diagnosis is established only through histopathological examination [22]. Currently, no consensus exists in the literature regarding the necessity of percutaneous biopsy before treatment of a suspected retroperitoneal sarcoma. In the retroperitoneum, neoplasms are far more likely to be liposarcomas than benign lipomas, so it is generally preferable to consider the lesion malignant and proceed directly to surgical excision without prior biopsy.

Biopsy should be reserved for patients considered for preoperative radiotherapy and/or chemotherapy, patients with unresectable tumors, and patients presenting with hematogenous metastases [23].

Surgery remains the cornerstone of treatment, primarily consisting of tumor resection with a margin of surrounding healthy tissue [24].

Several factors influence the prognosis of patients with retroperitoneal liposarcoma, including tumor size, resection margins, and histologic grade.

As previously mentioned, histologic subtype is an important prognostic factor, particularly regarding local recurrence, distant metastasis, and mortality [25].

Tumor size is also a prognostic factor, especially for the development of distant metastases and overall survival. Patients with large tumors (>20 cm) appear to have a significantly worse prognosis compared to those with smaller tumors (<20 cm) [26].

Resection margin status also influences prognosis and significantly increases the risk of local recurrence [27]. Therefore, surgical resection should be complete and sometimes extended to adjacent organs to maximize the likelihood of achieving negative margins. This extended resection has been associated with lower local recurrence rates compared to standard resection [28], although it may be linked to higher morbidity.

For unresectable tumors, preoperative radiotherapy and/or chemotherapy may help reduce tumor size and facilitate resection [29].

Some studies have shown that preoperative radiotherapy does not significantly improve recurrence-free survival compared with surgery alone and may increase adverse effects [30]. Chemotherapy is reserved for metastatic or inoperable tumors, with limited efficacy [31].

CONCLUSION

Retroperitoneal liposarcoma is an insidious tumor often diagnosed late due to its deep location and lack of early symptoms. Management remains a major therapeutic challenge. Complete surgical resection with negative margins is the cornerstone of treatment and the primary factor influencing survival. Local recurrences are common, and tumor size and histologic subtype are important prognostic factors.

Several multimodal approaches, combining surgery, radiotherapy, chemotherapy, and targeted therapy, aim to improve prognosis and reduce morbidity and mortality.

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