CASE REPORT

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# Well-Differentiated Liposarcoma as a Case: The Competence of Surgical Treatment was Questioned

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**ABSTRACT** Sarcomas are malignant tumors arising from skeletal and non-skeletal connective tissue, as well as the peripheral nervous system. Sarcoma is detected in the majority of patients who have a primary retroperitoneal, extra-visceral, unifocal soft tissue mass. Liposarcoma and leiomyosarcoma are the most prevalent histologic types of retroperitoneal sarcoma in adults. Most retroperitoneal liposarcomas are known as low to mild grade tumors. Well differentiated (low grade) liposarcomas are the most prevalent kind and have no potential to metastasize. Atypical lipomatous tumors are defined as those that develop on the body wall, trunk, or extremity. However, the same tumors are called well-differentiated liposarcomas due to their tendency for local recurrence and located in the retroperitoneum, mediastinum, and spermatic cord. In our case, we aimed to present a case that occupies most of the intra-abdominal volume, has associated gastrointestinal system complaints, and eventually was diagnosed as well-differentiated liposarcoma following surgical treatment.

**Keywords:** Adipocytes; cell differentiation; neoplasm; liposarcomas; retroperitoneal space

Retroperitoneal sarcomas are often asymptomatic unless they invade or compress adjacent tissues. At the time of admission, the majority of cases have large masses with an average diameter of 15 cm.[1] The first approach as a radiological evaluation is contrast-enhanced computed tomography (CT) of the abdomen and thorax. It can provide information about the histological type and the extent of the mass. Magnetic resonance (MR) imaging is used to assess the local tumor spread in patients who are eligible for preoperative radiation.[2] The role of fluorodeoxyglucose (FDG) and positron emission tomography (PET)/CT in the evaluation of initial stating remain unclear and therefore it is not routinely used for this purpose.

Certain criteria have been determined for the patients who cannot undergo resection due to specific radiological features which include the presence of advanced vascular infiltration, the detection of peritoneal implants, the presence of distant metastases with no chance of curative surgical resection, and the presence of mesenteric pedicle infiltration, primarily superior mesenteric artery, and spinal cord involvement.[3] A biopsy can be taken in the case of an uncertain diagnosis or a neoadjuvant treatment plan is made.[4] Recent studies demonstrate that percutaneous core needle biopsy is safe and has no effect on local recurrence and survival.[5] It is uncommon to see seeding in the biopsy tract.[6] In addition, if radiological imaging reveals evidence of lipoma or well-differentiated liposarcoma in the retroperitoneal mass lesion and the patient refuses preoperative treatment, as in our case, surgical treatment can be the first choice.

## CASE REPORT

Informed consent was obtained from the patient in this case report.

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A 45-year-old female patient was evaluated with complaints of abdominal pain, indigestion, nausea, vomiting, and difficulty in defecation. A large mass lesion encompassing the entire abdomen was identified on palpation. CT scan revealed a mass lesion that occupies the majority of the intra-abdominal volume and originates in the right posterior pararenal area, which has vascular branches and heterogeneous enhancement in some areas measuring 37*24*17 cm in size (Figure 1). The mass lesion was determined to exhibit a heterogeneous distribution of contrast enhancement and did not show considerable invasion of the surrounding tissue and vascular structure. According to the decision of the multidisciplinary oncology meeting, an exploratory laparotomy was performed, and the mass was excised en-bloc (Figure 2). The pathological examination revealed that the surgical margins were clean, and the mass with a diameter of 42*28*24 cm was a well-differentiated liposarcoma, including areas of necrosis and no sign of invasion. Cyclin-dependent kinase 4 (CDK4) and mouse double minute 2 (MDM2) gene amplification was detected by fluorescent in situ hybridization. PET/CT imaging performed following the surgery revealed no evidence of systemic disease. An increase in intensity was observed in the lower right side of the abdomen, starting from the lower pole of the kidney and leaning on the psoas major muscle, which was accompanied by slight FDG uptake, as well as giving the impression of continuity with the lateral infrarenal fatty planes (Figure 3). Abdominal MR imaging was performed approximately four months after surgery. Although there was an appearance consistent postoperative changes, no evidence of a residual mass was observed (Figure 4). Based on the findings, chemotherapy or radiotherapy was not sug-

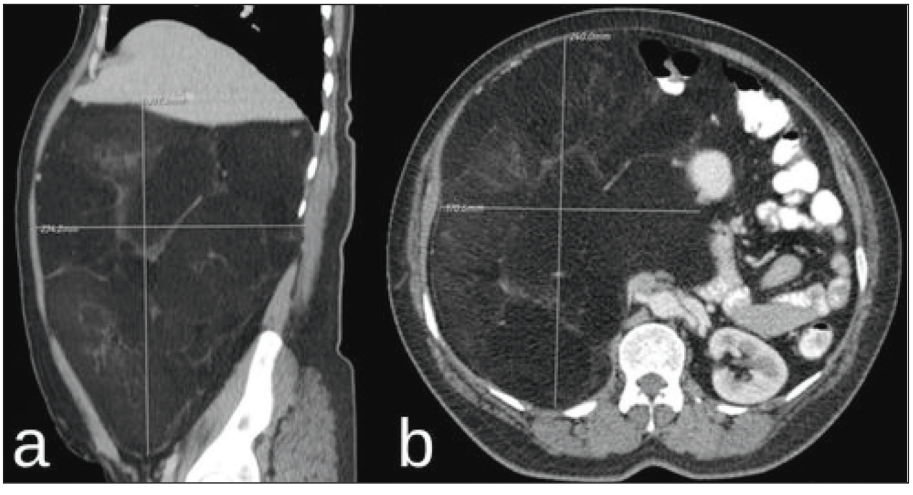


**FIGURE 1:** Massive lesion originating in the right posterior pararenal region, occupying the majority of the intra-abdominal volume. The mass lesion was determined to exhibit a heterogeneous distribution of contrast enhancement and did not show considerable invasion of the surrounding tissue and vascular systems.

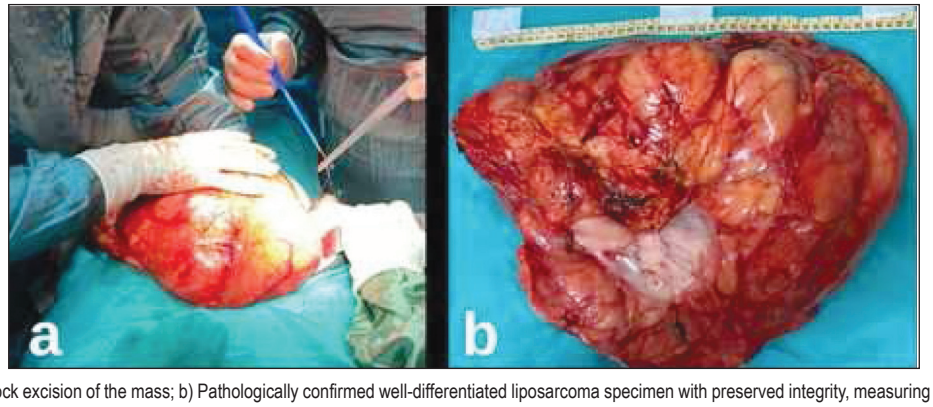


**FIGURE 2:** a) En-block excision of the mass; b) Pathologically confirmed well-differentiated liposarcoma specimen with preserved integrity, measuring 42*28*24 cm in diameter.

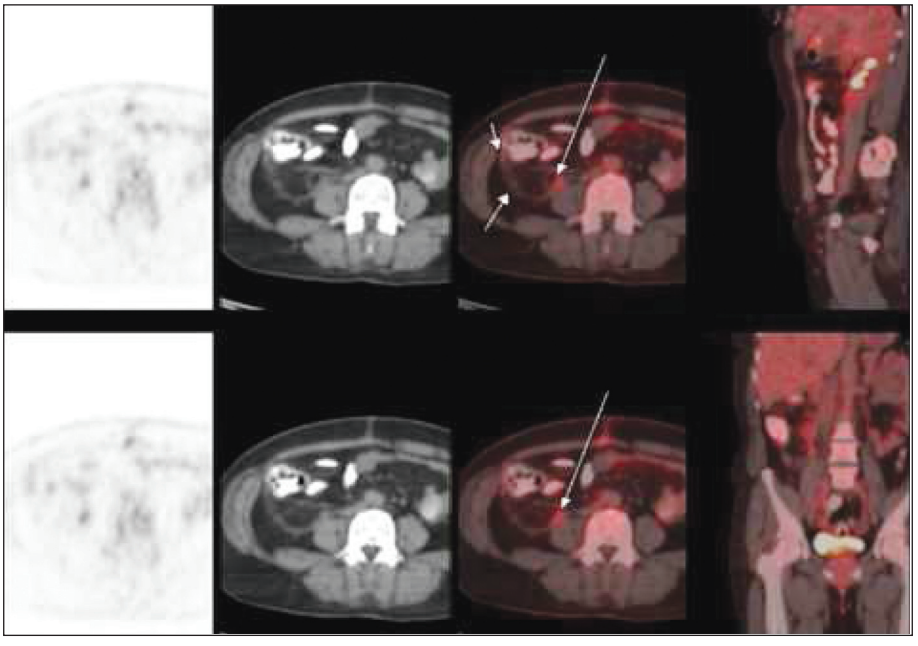

**FIGURE 3:** Postoperative positron emission tomography/computed tomography imaging at the fifth week. An increase in intensity was observed in the lower right side of the abdomen, starting from the lower pole of the kidney and leaning on the psoas major muscle, which was accompanied by slight fluorodeoxyglucose uptake, as well as giving the impression of continuity with the lateral infrarenal fatty planes. The findings were reported as suspicious for residual malignant processes following surgery.

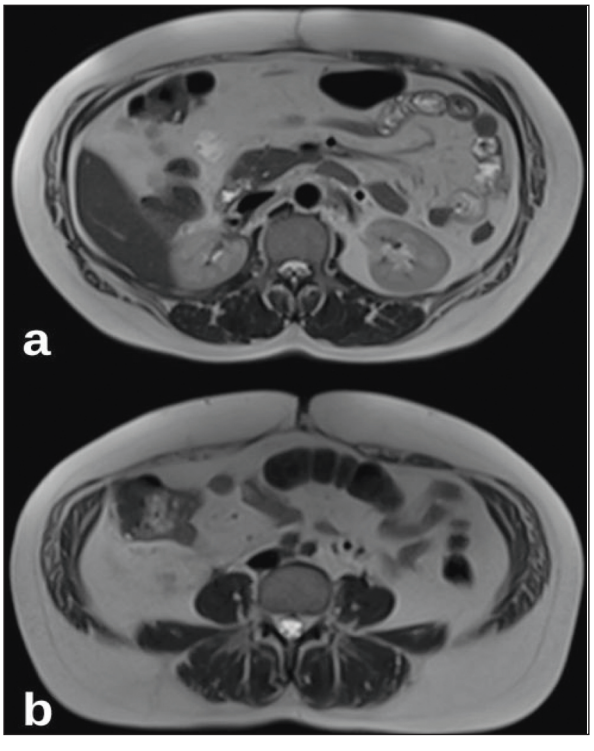


**FIGURE 4:** Postoperative magnetic resonance imaging at the 16th week, which shows no evidence of a residual lesion, the inflammation from the procedure had subsided, and the fat planes were clear.

gested for the patient, and a follow-up decision was made.

## DISCUSSION

Neoplasms in the retroperitoneal region often do not present symptoms before reaching a certain size. About 80 percent of these tumors are malignant and liposarcomas play a significant role in this group.[7,8] The tumor, node, metastasis approach is used to stage retroperitoneal liposarcomas. The most recent edition includes distinct T stage and prognosis stage classification for retroperitoneal sarcomas.[7] The size of the tumor was not found to be a prognostic factor. In addition, nodal or distant metastases are not detected in most resectable retroperitoneal sarcoma in patients presenting with large masses.[8,9] In our case, the biopsy was not considered for the patient who presented with gastrointestinal symptoms and radiological findings consistent with lipoma-well-differentiated liposarcoma; instead, surgical treatment was decided. The lesion was diagnosed post-operatively, its size was measured as greater than 40 cm according to the pathology report, and there was no evidence of systemic metastasis with infiltration regional structures, which is consistent with the literature.

Liposarcomas constitute 15-20% of soft tissue sarcomas and have four subtypes: well-differentiated,

dedifferentiated, mixed, and pleomorphic.[10,11] Well-differentiated liposarcomas are characterized as slowly growing tumors that do not have metastatic potential and have a low local recurrence rate. As a result, they have excellent clinical outcomes following a successful complete excision. Radiotherapy and chemotherapy are ineffective against liposarcoma. Histologically, it consists of single, enlarged, pleomorphic adipocytes with hyperchromatic nuclei intersected by fibrous septa. Cytogenetically, they have a supernumerary ring and giant rod chromosomes, which consist of amplified segments of 12q13-15 containing a number of cancer-associated genes involved in carcinogenesis. The most extensively researched of them is MDM2, which is an E3 ubiquitin protein ligase that functions as a major negative regulator of p53 and is amplified in approximately 100 percent of patients. The other is CDK4, a critical regulator of G1/S cell cycle checkpoints, which is amplified by MDM2 in over 90 percent of patients.[12,13] In our patient, both amplifications were positive.

The main curative treatment for localized liposarcoma is wide local excision with open surgical margins. However, the width of the resection margin of the retroperitoneal liposarcoma remains unclear. According to some authors, extensive resections should be performed in order to reduce the rate of local recurrence.[14] Additionally, unresectable local and/or metastatic liposarcomas are associated with poor prognosis. According to Bonvalot et al. preoperative radiotherapy was found to be not beneficial in the treatment of retroperitoneal sarcoma.[15] The role of systemic chemotherapy in soft tissue sarcomas at an early stage is controversial. In our case the tumor was easily removed from the surrounding soft tissues during the operation, and no evidence of invasion was observed. However, imaging technologies that provide objective information are required for such patients, similar to the logic of staining with indocyanine green, which is used for different purposes during surgery. The presence of a residual lesion requires reoperation, systemic chemotherapy, or radiotherapy, the efficacy of which is controversial. In our case, a suspicious appearance in the form of a residual mass in the excision area was identified on PET/CT performed for systemic screening during the fifth postoperative week. It could not be differentiated from postoperative inflammatory changes. This circumstance complicated the process of preparing a treatment strategy. Radiotherapy was ruled out due to the size, pathological characteristics, and placement of the mass. After waiting for four months, MR imaging was performed to ascertain the need for additional surgical intervention. The MR imaging revealed no evidence of a residual lesion, the inflammation from the procedure had subsided, and the fat planes were clear. In such instances, it is critical to choose the appropriate imaging modality following surgery and the timing for the procedure. The timing for the optimal imaging should be identified.

A decision regarding the surgical and perioperative management of retroperitoneal liposarcoma should be determined on a case-by-case basis by a multidisciplinary specialist team. Maximum effort should be made to reduce false positivity and negativity rates in imaging methods.

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### *Conflict of Interest*

*No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.*

### *Authorship Contributions*

***Idea/Concept:*** *Sami Açar;* ***Design:*** *Sami Açar;* ***Control/Supervision:*** *Sami Açar, Murat Api;* ***Data Collection and/or Processing:*** *Erman Çiftçi;* ***Analysis and/or Interpretation:*** *Sami Açar, Murat Api;* ***Literature Review:*** *Sami Açar, Erman Çiftçi;* ***Writing the Article:*** *Sami Açar;* ***Critical Review:*** *Murat Api;* ***References and Fundings:*** *Sami Açar;* ***Materials:*** *Sami Açar.*

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