

Case Report

A case presentation on rare case of retroperitoneal liposarcoma

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Received: 19 July 2023

Revised: 21 August 2023

Accepted: 01 September 2023

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ABSTRACT

Retroperitoneal liposarcoma is a rare biologically heterogeneous tumor that present considerable challenges due to its size and deep location. As consequences, the majority of the patients with high grade rpls will develop locally recurrent disease following surgery, and this constitutes the cause of death in most patients. Symptoms are usually nonspecific and they do not appear until the tumor becomes very large. Early diagnosis is difficult as there is an absence of specific clinical presentations. A painless abdominal lump that enlarges in a long period of time in an adult is the most common history of patient. Metastasis at initial presentation is uncommon. Surgical exploration is needed for the final pathological diagnosis. Case presentation: we report a case of 56 year old male patient who underwent a complete surgical excision of pleomorphic retroperitoneal liposarcoma. Retroperitoneal liposarcomas are rare soft tissue sarcoma. They often cause minimal or no symptoms and can reach a significant size, growing undetected in retroperitoneal space before invading or compressing surrounding organs, eventually leading to clinical symptoms. Complete surgical excision is the mainstay of treatment. Whenever possible, macroscopically complete resection should be aimed at, often requiring en bloc removal of adjacent structures such as the abdominal wall, psoas, or paravertebral muscles.

Keywords: Retroperitoneal liposarcoma, Soft tissue sarcoma, Abdominal lump, Surgical excision

INTRODUCTION

Liposarcomas are neoplasms of mesodermic origin derived from adipose tissue and correspond to 10-14% of all soft tissue sarcomas. They represent <1% of all malignant tumors.^{1,2} The most frequent subtypes are liposarcoma (41%), leiomyosarcoma (28%), malignant fibrous histiocytoma (7%), fibrosarcoma (6%) and tumors of the peripheral nerve sheath (3%).³ Retroperitoneal liposarcomas alone comprise 0.07-0.2% of all neoplasias.⁴ Approximately 85% of these are malignant, with soft-tissue sarcomas representing 35% of this group. Liposarcoma is the most frequent histopathological variety of the retroperitoneum.² It presents with inherent characteristics in relation to its deep localization and slow expansive growth. Average diameter of the tumor is 20-25 cm with a weight of 15-20 kg.⁴ There is compromise of the adjacent organs in up to

80% of the cases.^{4,5} Surgery is the gold standard for treatment of liposarcoma. Retroperitoneal liposarcoma is a distinct clinical entity that requires a more aggressive surgical approach, including multiple resections or multiorgan resection with recurrences. There is a low incidence of distant metastasis (7%) compared to other histological subtypes that range from 15 to 34%.⁶ The objective of this study is to report a case of giant retroperitoneal sarcoma.

CASE REPORT

A 56 year old Hindu married male patient presented with left sided abdominal lump since 2 months. Patient underwent abdominal CT scan for developing anorexia and weight loss that showed 11×16×20 cm sized lesion involving left renal-infrarenal compartment of retroperitoneum and extra-peritoneum. Abdominal lump which

was not associated with any other complaints. no relevant past medical or surgical history present.

Investigation

All blood investigations were within normal limits. Patient was diabetic and was adequately controlled over insulin dosage.

USG abdomen-approx. 12×17 cm sized solid cystic lesion without internal vascularity extending upto left lumbar region from left chondriac region.

MDCT scan of abdomen with pelvis -ct morphology of heterogeneously enhancing large lobulated soft tissue density with lobulated cystic area mass lesion involving left renal- infrarenal compartment of retroperitoneum and extra-peritoneum suggests possibility of retroperitoneal malignant lesion / sarcoma appears likely.

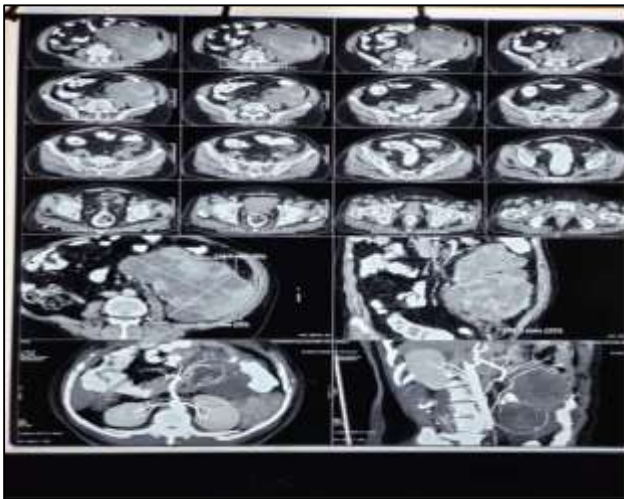


Figure 1: CT abdomen showing retroperitoneal lesion.

Operative findings-two multilobulated mass measuring approximately 10×10×14 cm and 17×15×10 cm was excised and sent for histopathology reports.



Figure 2: Intra-op photo of a lesion of 10×10×14 cm size delivered out of the abdomen.



Figure 3: Intra-op photo of another lesion of 17×15×10 cm delivered out of abdomen.

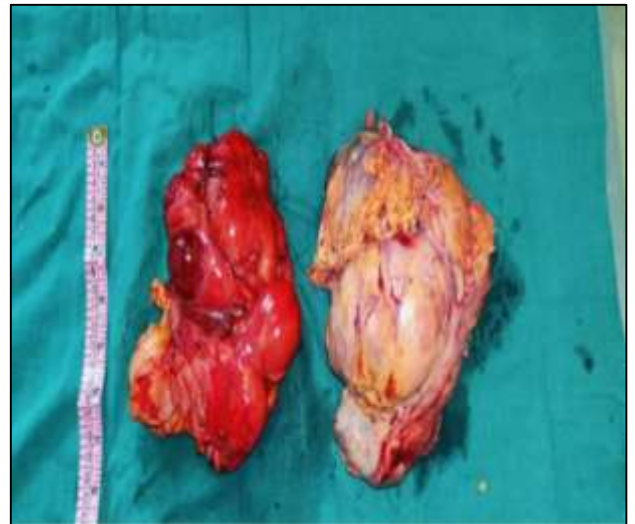


Figure 4: Measurement of both the lesion with measuring tape.

Histopathology

SP-1) received multilobulated mass measuring 14×11.5×8.5 cm. Cut surface is yellowish to greyish soft and shiny. Partially embedded. SP 2) received multilobulated mass measuring 17×15×9.5 cm. Cut surface is yellowish to greyish soft and shows multiple cyst measuring from 1.0 cm to 6.5 cm filled with jelly like material and hemorrhagic material. Partially embedded.

Microscopic examination

Spindle cell sarcoma, pleomorphic with focal myxoid changes (both specimen) note: morphology resembles pleomorphic liposarcoma.

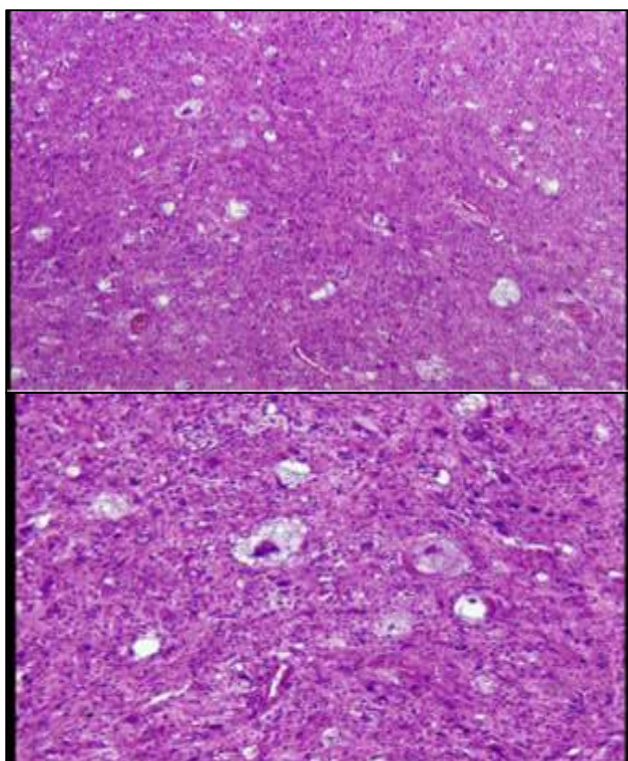


Figure 5: Microscopic examination of the lesion.

IHC reports

Desmin [D33 Dako]-positive in few cells S100 [SE2E2 Biogenex]-positive in few cells, MDM-positive in few cells.

Diagnosis was retroperitoneal liposarcoma.

Patient was discharged on post operative day 6 without any postoperative complications and its histopathology reports showed pleomorphic liposarcoma.

DISCUSSION

Retroperitoneal tumor predominantly originates from fat, loose connective tissue, fascia, muscles, lymphatic tissue or residual embryonic tissue, of which 80% is malignant.

Liposarcoma is the most frequent histological type of retroperitoneal sarcoma, corresponding to 41% of these tumors.^{4,5} It has been reported that 20% of the tumors are >10 cm at the time of diagnosis; however, few cases of retroperitoneal liposarcomas exist that can be considered as giant.^{3,4,6} Clinically, these tumors tend to present with diffuse abdominal pain accompanied by anorexia and weight loss and increase in abdominal girth. The most characteristic sign is a painless abdominal mass detected in ~78% of the cases. Abdominal symptomatology is due to compression of the organs, similar to that reported with the present case.^{3,4} It is clear that our patient presented all the signs and symptoms specific for these tumors due to the size of the abdominal mass.

Overall, liposarcoma is the most frequent soft tissue sarcoma subtype and represents 45% of all retroperitoneal sarcoma; it is composed of three histologic varieties: well-differentiated and dedifferentiated liposarcoma, pleomorphic liposarcoma, and myxoid/round cell liposarcoma, listed in order of decreasing frequency. Well-differentiated and dedifferentiated liposarcomas more typically arise from the retroperitoneum versus the extremities, whereas the inverse is true for pleomorphic and myxoid/round cell liposarcoma. Compared with well-differentiated liposarcoma, the dedifferentiated variety has a worse prognosis, largely because of its much greater risk of distant metastasis compared with well-differentiated liposarcoma. Local recurrence is common in both types. The malignant behavior of well-differentiated and dedifferentiated liposarcomas is attributable to the amplification of chromosome 12q13-15, which accounts for the upregulation of mdm2 and CDK4. Both well differentiated and dedifferentiated retroperitoneal liposarcomas are often multifocal. Myxoid and round cells are descriptive terms.

Based on their histologic appearance. These liposarcoma varieties are characterized by distinct translocations such as FUS-DDIT3.

Located at t(12;16)(q13;p11) and more rarely EWSR1-DDIT3 located at t(12;22)(q13;q12). Multiple tumor-promoting pathways including met, ret, and PI3K/AKT are activated as a result of these translocations. Myxoid liposarcoma is unusual in its relative sensitivity to radiation and chemotherapy, resulting in a 10-year disease-specific survival of 87%. Considered a poorly differentiated form of the myxoid variety, the round cell variety has a worse outcome than myxoid liposarcoma, with metastasis developing in 21% of patients in one large series. Pleomorphic liposarcoma is another example of a poorly differentiated liposarcoma with a poor outcome.

The treatment of patients with retroperitoneal liposarcoma is more complex. The principal goal is a gross complete resection as incomplete gross resection is associated with an increased risk of mortality.⁷ Traditionally, retroperitoneal sarcoma has been treated by resection with a generous gross margin, with resection of organs and structures that are contiguous with or invading the tumor when feasible. More recently, some have advocated for a “complete compartmental resection,” which mandates the resection of adjacent organs, even if they are not directly involved with the tumor.⁸ Although it is controversial, the concept that “the resection is only as good as the closest margin” is an important one.

This takes into account the relationships between vital structures on one side of the tumor and not resecting contiguous but uninvolved organs. Understanding of the patterns of retroperitoneal liposarcoma recurrence is important in planning the optimal approach. For patients

with well-differentiated retroperitoneal liposarcoma, a unifocal versus multifocal presentation does not appear to confer an adverse prognosis, but patients with dedifferentiated disease multifocality have a worse overall survival.⁹ Patients who develop recurrence after initial resection are likely to develop multifocal disease. This appears to be reflective of the tumor biology because an initial resection with positive margins does not appear to affect whether a patient develops a unifocal versus multifocal recurrence. The complete compartmental resection approach results in frequent multi-visceral resections, with the following organs resected in more than 50% of cases: spleen, pancreas, diaphragm, adrenal gland, and kidney.⁸ Proponents of a more traditional approach in which only tumor-contiguous organs are removed point out that 15% of patients who have recurrence after undergoing standard resection do so beyond the compartmental bounds of their initial tumor.⁹ These out-of-field recurrences are unlikely to have been prevented with an aggressive complete compartmental resection strategy, and patients who may eventually benefit from nephrotoxic systemic chemotherapy are adversely affected by a potentially unnecessary complete compartmental resection-related nephrectomy.

Although grossly incomplete resections are to be avoided, a margin-negative resection is not possible in some situations. At times, this can be predicted on the basis of the preoperative imaging, but at other times, the difficulty of the resection is not appreciated until during the operation. A single-institution retrospective study compared the outcome of patients with retroperitoneal liposarcoma who underwent an incomplete resection versus patients who underwent exploration and biopsy without tumor resection. Even incomplete resection provides a statistically significant improvement in survival compared with no resection, 26 versus 4 months. In addition, 75% of patients undergoing incomplete resection reported palliation of their presenting symptoms. In the setting of recurrent retroperitoneal liposarcoma, the rate of recurrent tumor growth is associated with prognosis. Patients whose recurrence grows less than 0.9 cm/mo benefitted from complete resection of the recurrence, whereas recurrent tumor growth of more than 0.9 cm/mo was associated with poor outcome.¹⁰ Palliative chemotherapy options are emerging for patients with unresectable recurrence who have already failed chemotherapy. A subgroup analysis of a randomized phase 3 trial comparing eribulin versus dacarbazine for either extremity or retroperitoneal liposarcoma showed that eribulin was associated with an improvement in overall survival (15.6 vs. 8.4 months). Based on these data, single-agent eribulin is approved in the palliative setting for patients with liposarcoma. Together, these observations contribute to the complexity of developing an individualized treatment plan for retroperitoneal liposarcoma.

According to “excellent local control with preoperative

radiation therapy, surgical resection, and intraoperative electron radiation therapy for retroperitoneal sarcoma”—thirty-seven patients (59%) underwent surg-RT and 26 (41%) had surgery alone. 51% of tumors were high grade and 36% of patients had locally recurrent disease. Final margin status was: r0 73%, r1 16%, r2 6%, and unknown 5%. Of those with r0 resections, 67% received surg-RT. Median follow-up was 45 months. The 5-year local control rate was 89% for surg-RT patients and 46% for surgery alone patients ($p=0.03$). On multivariate analysis, surg RT was the only variable associated with a lower risk of LR (hr 0.19; ci 0.05-0.69, $p=0.003$). The actuarial 5-year OS was 60% for patients receiving either surg-RT or surgery alone.¹¹

CONCLUSION

Patient was operated for exploratory laparotomy and complete excision of the lump was done for the management and to know the exact pathology. No chemotherapy or radiotherapy was given to patient. The postoperative course was uneventful. Patient has no complaints in follow ups.

Funding: No funding sources

Conflict of interest: None declared

Ethical approval: Not required

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Cite this article as: Patel SH, Patel AN, Malek TN, Kuswaha MR. A case presentation on rare case of retroperitoneal liposarcoma. *Int Surg J* 2023;10:1693-7.