Case Report

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Retroperitoneal liposarcoma of the intestine: a rare cause of acute abdomen

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ABSTRACT

Retroperitoneal liposarcoma is a rare malignant mesenchymal tumor with an incidence of 2.5 per million individuals. It is usually asymptomatic until its large enough to compress the surrounding organs, so its early diagnosis is difficult. In current case a 41 year old male patient with no significant past medical history presented to emergency department with sudden onset of abdominal pain in the left lumbar region. On examination, the abdomen was distended, guarding and rigidity present with tenderness to palpation in the left lumbar region, and no mass was palpated. Abdominal USG revealed a large heterogeneous mass in the left lumbar region. As the origin of the mass was uncertain, the patient was evaluated with CECT of the abdomen which revealed a retroperitoneal mass highly s/o liposarcoma. Given the suspicion of a liposarcoma, the patient underwent an abdominal exploration and there was a retroperitoneal mass of 12x8 cm adherent to the left colon. Wide resection of this mass was done along with left colectomy and the specimen was sent for examination. The histopathology study determined lipomatous tumor well differentiated retroperitoneal liposarcoma. Retroperitoneal liposarcoma is a malignant tumor whose treatment is fundamentally surgical. These tumors tend to be resistant to radiotherapy and/or chemotherapy. Among the most important prognostic factors related to survival is surgery with non-affected margins.

Keywords: Liposarcoma, Retroperitoneal, Surgery, Colectomy

INTRODUCTION

Retroperitoneal liposarcoma (RPLS) is the most commonly observed soft tissue sarcoma in the retroperitoneal space. Its age of presentation is between the fourth and sixth decade of life without finding differences between men and women. Liposarcoma is a common type of soft tissue sarcoma, which occurs most commonly in the extremities (52%), followed by the retroperitoneum (19%). 1,2 Retroperitoneal liposarcoma is a rare tumor with an incidence of 2.5 per million individuals. It is usually asymptomatic until it's large enough to compress the surrounding organs. It is often misdiagnosed due to atypical presentations and the absence The well-differentiated of symptoms.

liposarcoma is a low-grade neoplasm that can present as five histological variants: lipoma-like, sclerosing, inflammatory, spindle cell, and liposarcoma with meningothelial whorls. The treatment of choice is complete surgical excision. According to Stoeckle et al there are no survival benefits of adding adjuvant radiotherapy for a resected well-differentiated retroperitoneal liposarcoma.³

CASE REPORT

A 41 year old male patient with no significant past medical history presented to our emergency department with sudden onset of abdominal pain in the left lumbar region. On examination, the abdomen was distended,

guarding and rigidity present with tenderness to palpation in the left lumbar region, and no mass was palpated. Abdominal USG revealed a large heterogeneous mass in the left lumbar region. As the origin of the mass was uncertain, the patient was evaluated with CECT of the abdomen which revealed a retroperitoneal mass highly s/o liposarcoma. On abdominal exploration, there was a retroperitoneal mass of 12x8 cm adherent to the left colon. Wide resection of this mass was done along with left colectomy and the specimen was sent for histopathological examination. On histopathology examination: Lipomatous tumor well differentiated liposarcoma measuring 12x8x7.5 cm involving muscularis propria and serosa. Both colonic surgical margins of resection were free from tumor infiltration. The patient was discharged on the tenth day of hospital admission without incidences, currently requiring followup without receiving adjuvant treatment.

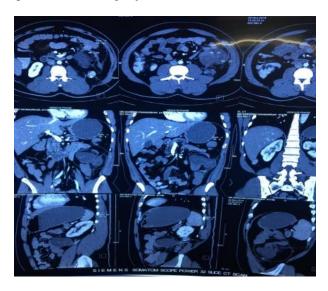


Figure 1: CECT showing retroperitoneal mass.



Figure 2: Retroperitoneal mass of 12x8 cm adherent to the left colon.

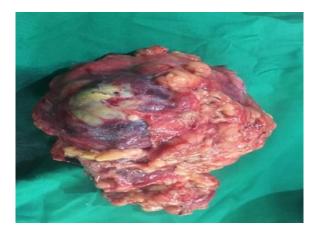


Figure 3: Gross specimen; 12x8x7.5cm and 1060 gms.

DISCUSSION

Retroperitoneal tumors are an extremely heterogeneous group of neoplasms, 85% of which are malignant. Liposarcomas constitute between 45-55% of retroperitoneal masses. One-third of malignant tumors located in the retroperitoneum are sarcomas with a median age of presentation in the sixth decade with an equal male/female ratio. Retroperitoneal sarcomas present 80% of the time as an asymptomatic abdominal mass. Symptoms can also be related to mass effect or local invasion which may lead to pain, gastrointestinal obstruction, feelings of early satiety, and weight loss. Besides, neurologic and muscular-skeletal symptoms are referred to the lower extremities.

World health organization identifies five histologic subtypes (well-differentiated, myxoid, round cell, dedifferentiated and pleomorphic). Histopathologic variety is the main prognostic factor. Well-differentiated liposarcoma represents around 30% like our case and has the best prognosis. The myxoid type is the most frequent liposarcoma, constituting around 50% of all tumors. It has a less favourable progression, as it often recurs early. The pleomorphic, round cell, and undifferentiated types display the worst prognosis.⁷

In general, well-differentiated RPLS may not infiltrate other organs or major intra-abdominal structures, whereas dedifferentiated RPLS frequently infiltrates and involves surrounding tissues. The optimal treatment for patients with localized, resectable retroperitoneal sarcomas is surgery with gross and microscopically negative margins. Complete surgical resection frequently requires en-bloc resection of adjacent viscera.⁸

Chemotherapy for retroperitoneal liposarcoma is not effective and radiotherapy has limited efficacy due to toxicity affecting intra-abdominal structures and showed validity only in case of high-grade malignancy by reducing local recurrence, but it does not improve overall survival. Nowadays only the complete surgical resection

remains the most important predictor of local recurrence and overall survival.

Unfortunately, retroperitoneal liposarcoma is almost always large at the time of diagnosis owing to its slow growth and vague symptoms. Preoperative evaluation is best accomplished with abdominal CT scanning; however, MRI may be useful in select circumstances where involvement of unresectable structures is suggested by the CT. The resection of a retroperitoneal sarcoma of remarkable size is a challenge for the surgeon owing to the anatomical site, to the absence of an anatomically evident vascular-lymphatic peduncle that makes it hard to obtain safe margin and to the adherence with the contiguous organs and with the great vessels. Therefore the retroperitoneal liposarcoma shows a high rate of local recurrence after surgical excision.

The prognosis of liposarcoma depends on the degree of differentiation, size, histological type, and tumor staging. The gold standard treatment is total surgical resection with free margins, which might be predictive of cure. However, if total resection is not possible, radiotherapy should be considered to decrease the recurrence of the disease. The overall survival at 5 years reported in the literature for the various histological subtypes well-differentiated, myxoid/round cell, undifferentiated and pleomorphic, ranging from 90%, 60 to 90%, 75%, and 30 to 50%, respectively.⁹

CONCLUSION

Liposarcoma a rare tumor located retroperitoneally is usually asymptomatic and is being diagnosed when they are large enough and produce compressive symptoms. Surgical resection is the mainstay of treatment despite its various size. The role of chemotherapy and radiotherapy is controversial due to its low sensitivity to these types of tumors.

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