Case Report

Giant retroperitoneal liposarcoma: A very rare case report

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ABSTRACT

Soft-tissue sarcomas are very rare entity, seen in <1% of all malignancies. Sarcoma accounts for about one-third of all retroperitoneal malignancies. Management of retroperitoneal sarcoma is challenge for surgeons because of its rare presentation and no standard guidelines for it. A 65-year-old male presented to surgery out-patient department with the complaints of abdominal distention and pain in abdomen since 3 years. On imaging, he was diagnosed as case of retroperitoneal tumor and managed by radical surgery including resection of the tumor with left kidney, spleen, and descending colon. Postoperative period was uneventful.

Key words: Giant retroperitoneal sarcoma, liposarcoma, retroperitoneal tumor

INTRODUCTION

Retroperitoneal sarcomas (RPSs) are very rare tumors, constituting 1-2% of all solid tumors.[1] These malignant tumors arise from mesenchymal cells and are usually located in muscle, fat, and connective tissues.[2] However, it has been observed rarely in the gastrointestinal system. Liposarcoma accounts for more than 20% of all sarcomas in adults and up to 41% of all RPSs.[3,4]

Imaging studies plays a huge role in management of RPSs as they are present in anatomically challenging sites. Imaging provides the surgical pathway for resection as it tells about the local extent, lymph node status and distant metastasis. They are usually diagnosed in their advance stage. Surgery is the main stay of management as neoadjuvant chemo and radiotherapy are inferior to the radical surgery. In tumors that have been adequately excised, the final outcome depends on the tumor grade.[5] We are reporting a rare case of giant RPS in a 65-year-old male, which was managed by radical resection.

Access this article online

Quick Response Code:

Website: www.ccij-online.org

DOI: 10.4103/2278-0513.142672

CASE REPORT

A 65-year-old male presented with the complaints of abdominal pain and distention since 3 years. History of weight loss was present. On examination, a large, firm mass was palpable occupying whole of the abdomen. All laboratory investigations were normal.

Contrast-enhanced computed tomography (CECT) abdomen revealed large hypo dense fatty attenuation septated mass lesion. The mass is found to be displacing left kidney and colon to the right crossing the midline. Multiple enhancing nodular masses with septa are noted within the mass lesion [Figure 1]. On laparotomy huge retroperitoneal tumor encasing left kidney, spleen, mesentry of splenic flexure and descending colon with no lymphadenopathy was found. *En bloc* resection of the tumor with left kidney, spleen, splenic flexure and descending colon followed by colocolic (transverse-sigmoid) stapler anastomosis was done.

Gross examination of specimen revealed large, lobulated soft tissue mass measuring 40 cm × 30 cm and weighing 10.6 kg. Histopathology examination revealed well-differentiated liposarcoma [Figures 2 and 3]. Postoperative period was uneventful and patient was discharged on day 11.

DISCUSSION

The overall incidence of RPS is 0.3–0.4% per 100,000 of the population, and usually patient present in their fifth to sixth

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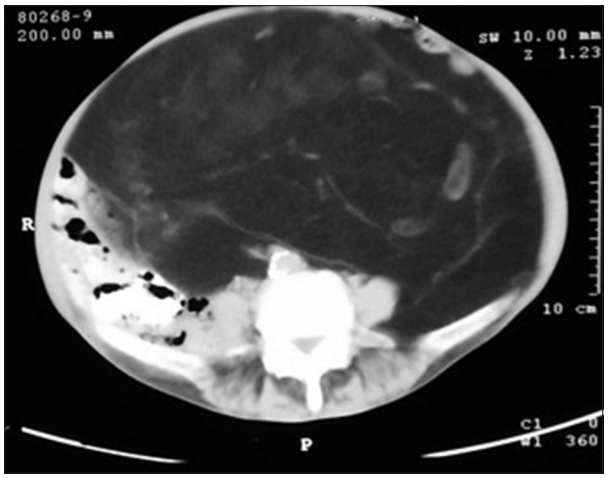

Figure 1: Contrast-enhanced computed tomography abdomen image

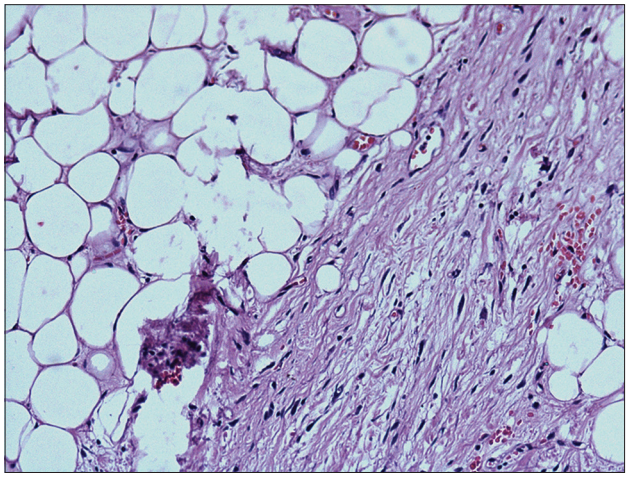

Figure 2: Histopathology-lobules of fat with presence of atypical round to spindle cells in the septa

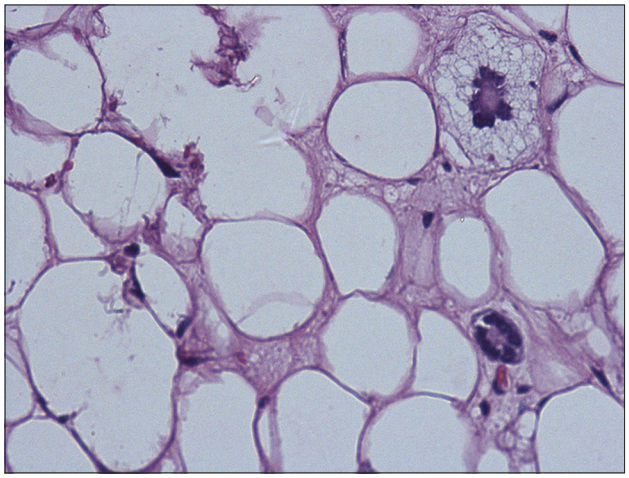

Figure 3: Histopathology (×40) - Presence of lipoblast within tumor

decade of life.[6] Primary tumors of the retroperitoneum can be of many types: Neoplasms arising in the kidney, adrenal gland, retroperitoneal lymph nodes (malignant lymphomas). The retroperitoneal soft-tissue sarcomas are also presented in this location, as liposarcoma, lipoma, malignant fibrous histiocytoma, leiomyosarcoma, rhabdomyosarcoma, fibrosarcoma, vascular tumors, peripheral nerve tumors, synovial sarcoma, etc., World Health Organization has divided liposarcoma into five subtypes according to the differentiation:

1. Well-differentiated
2. Dedifferentiated
3. Myxoid
4. Pleomorphic
5. Mixed type.[7]

Retroperitoneal sarcoma most commonly presents as an asymptomatic abdominal mass in 80% of cases because tumors smaller than 5 cm usually remain silent. 20% of cases present with nonspecific signs and symptom, e.g. vague abdominal discomfort or pain, weight loss, and early satiety whereas some cases may present with intestinal obstruction or bleeding. They can also present as lower extremity swelling or pain.[8]

Contrast-enhanced computed tomography abdomen is very helpful in preoperative planning and demonstrates relationship of tumor to the nearby structures and vascular invasion. Sensitivity of CT scan in low grade lesion is 100% and there is virtually no role of preoperative treatment of these lesions. It also helpful in detection of synchronous and metastatic lesion in liver.[9]

Optimal management of resectable RPS is complete surgical removal with tumor free margin, which sometimes requires *en bloc* resection of adjoining structures. Most commonly resected structures are kidney, large bowel, ureter etc.

Similar case reports of giant RPSs have been reported in literature. Morandeira *et al*.[10] have reported a case of giant RPS in a 63-year-old woman. She presented with the complaints of abdominal mass, edema lower extremity and cahexia. CECT abdomen revealed heterogeneous mass with necrosis pushing intestine. *En bloc* resection of tumor with right nephrectomy, appendectomy, hysterectomy was done.

Prognosis depends on grade of the tumor, completeness of resection and multifocality of disease. Hassan *et al*. reported series of 97 patients of RPS with complete resection in 78% of cases with 5 years survival in 51% cases and 5 years recurrence in 44% cases.[11]

Local recurrence is the main cause of failure, ranging from 40% to 80%, respectively.[4] About 75% of sarcoma-related deaths involve uncontrolled local recurrence.[4,12]

Follow-up is by CECT scans at every 6 months intervals to rule out any recurrence since most recurrence are seen within 2 years of initial surgery.[13]

Radical surgery provides the cure and is standard of care in RPSs. The concept of "compartmentalization" that applies to limb sarcomas is difficult to extrapolate directly to RPS.[14] It's difficult to get microscopic margins in huge RPS. No effective chemotherapy exists to influence survival in patients with RPS.

CONCLUSION

No standardized guidelines have been established for its treatment because too small a number of cases have been reported, but surgical resection was considered the treatment of choice in our case. These tumors require an aggressive surgical approach. A larger number of cases will be necessary to determine whether additional treatment such as chemotherapy and radiotherapy is necessary or effective in liposarcoma of colon.

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Cite this article as: Mohil RS, Jethwani U, Singh GJ, Arora J, Saroha R, Verma R, *et al*. Giant retroperitoneal liposarcoma: A very rare case report. Clin Cancer Investig J 2014;3:548-50.

Source of Support: Nil, **Conflict of Interest:** None declared.