



A CASE OF GIANT RETROPERITONEAL LIPOSARCOMA BALA MURALI

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Abstract : Retroperitoneal liposarcoma accounts for about 0.1 per cent of all malignancies and 15 per cent of all sarcomas. The cell of origin is mesenchymal cells. They usually present with large size. Complete surgical resection with or without contiguous organ resection is the main treatment of choice. This is a case of giant retroperitoneal liposarcoma of size 21inc. x16 inc. weighing about 11kgs resected en bloc at first surgical attempt without any contiguous organ resection.

Keyword : RETROPERITONEAL LIPOSARCOMA, SURGERY, CHEMOTHERAPY, RADIOTHERAPY, RECURRENCE

INTRODUCTION:



Soft tissue sarcomas are rare and account for less than 1% of all newly diagnosed malignancies. One third of malignant tumors that arise in the retroperitoneum are sarcomas. Liposarcoma is the single most common soft tissue sarcoma and the most common retroperitoneal sarcoma. It accounts for at least 20% of all sarcomas in adults and up to 41% of all retroperitoneal sarcomas. Retroperitoneal liposarcomas (RPLS) grow slowly and silently. Only complete excision provides a hope of a cure, this is often difficult, especially in well differentiated subtypes because the margins are not grossly apparent thus often necessitating contiguous organ resection.^{1,6}

FIG 1 Pre operative picture showing mass of size 45cmx30cm occupying all quadrants of abdomen.

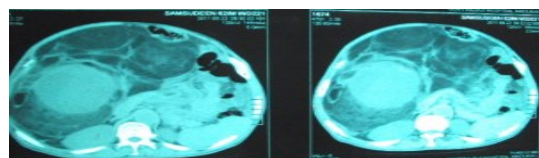
CASE REPORT

A 62 year old male presented with complaints of abdominal mass for 4 years which initially started around the right lumbar region and gradually progressed in size. On abdominal palpation, a mass of size 45cmx30cm occupying all quadrants of abdomen (Fig. 1.) which was hard to firm in consistency. There were no cutaneous lesions.

The routine blood investigations were found to be normal. Sonographic examination of the abdomen and pelvis showed a large ill-defined mixed echogenic mass lesion approximately 40x25 cms in right hypochondrium extending to right lumbar region and crosses the midline, displacing the liver superiorly. CT abdomen and pelvis showed a huge mass of size 35cmx28cms occupying entire right abdomen, multi locular with septa enhancing on contrast suggestive of liposarcomatous feature. The lesion is anterior and compressing right kidney displaces liver superiorly. Both kidneys were found to be normal. (Fig. 2,3). Metastatic work up was found to be normal.

Patient proceeded for elective laparotomy through midline abdominal incision and findings including a huge well encapsulated mass of size 21x16 inches occupying the entire right side of the retro peritoneum pushing the entire small bowel, ascending colon, and caecum towards left [Fig. 4]. Meticulous dissection was made all around the tumour and it was separated from ureters, inferior vena cava and aorta. Complete excision was done without any contiguous organ resection. (Fig 5,6,7,8)

The postoperative biopsy was reported as well differentiated liposarcoma showing lipocytes. The patient has been followed for three months till now clinically and radiologically without evidence of recurrence.



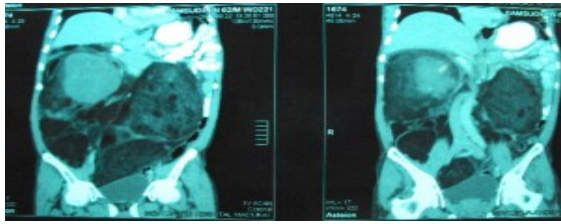


FIG 2 CT ABDOMEN SHOWING LESION ANTERIOR AND COMPRESSING THE RIGHT KIDNEY.

FIG 3 RETROPERITONEAL LESION DISPLACING THE LIVER ANTERIORLY. DISCUSSION

Soft tissue sarcomas are rare neoplasms. About one third of malignant tumours of retro peritoneum are sarcomas which commonly arise from the mesenchymal cells. Usually they occur in the sixth to seventh decade and there is no gender predisposition.¹ They are associated with various genetic syndromes like Li-fraumeni syndrome, Von Recklinghausen's disease. The majority of patients present with asymptomatic abdominal mass (80%), symptoms related to adjacent structures invasion or compression.

The most common histopathological variant of retroperitoneal sarcoma includes liposarcoma (41%) followed by leiomyosarcoma and malignant fibroushistiocytoma². Liposarcomas can be divided into well differentiated, myxoid, roundcell, pleomorphic variants. Well -differentiated liposarcoma is an adipocytic tumor with widened fibrous septa and enlarged, hyperchromatic atypi. Lipoblasts need not be present for the diagnosis of a well-differentiated liposarcoma. When the tumor contains an area of well-differentiated liposarcoma that abruptly changes to a solid, alipogenic area that measures more than a 10xpower field, the lesion is defined as dedifferentiated liposarcoma; usually, however, the alipogenic area is much larger and is visible both at gross inspection and radiologically³. Myxoid liposarcoma is composed of myxoid lobules with delicate plexiform vessels, predominantly monovacuolated and bivacuolated lipoblasts, and a paucity of round to stellate stroma cells. The stromal cells may coalesce and form sheets of round cells. These sheets constitute the round cell component of round cell liposarcoma. The pleomorphic variants are usually aggressive & frequently metastasise⁵.

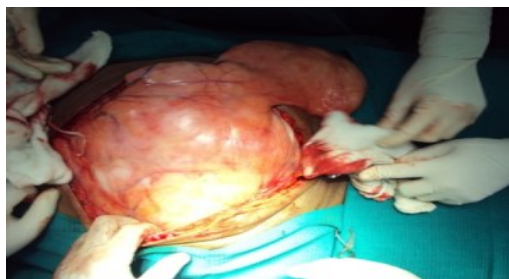


FIG. 4 Mass pushing entire small bowel,ascending colon,ceacum towards left

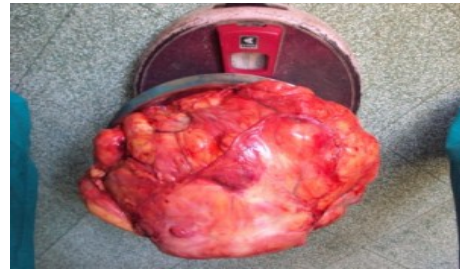


FIG.5. EXCISED SPECIMEN MEASURING 21X16INCHES

FIG. 6. RESECTED SPECIMEN WEIGHING 11 KGS

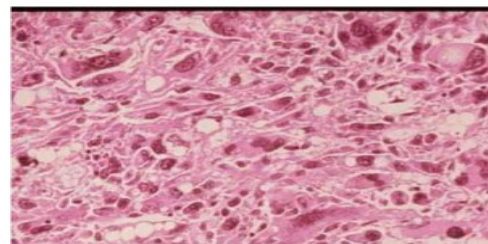
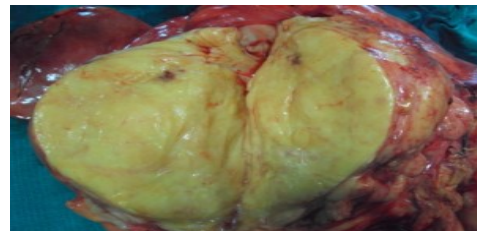


FIG 7 CUT SECTION OF THE SPECIMEN SHOWING LIPOMATOUS AREAS

FIG 8 MICROSCOPIC PICTURE SHOWING LIPOCYTES AND ATYPICAL NUCLEI SUGGESTIVE OF RETROPERITONEAL LIPOSARCOMA.

Well-differentiated liposarcomas contain mature fatty elements with imaging characteristics that may be indistinguishable from those of normal fat¹⁰. This normal fatty appearance may represent greater than 75% of the volume of a lesion, a finding that explains why the tumor might be mischaracterized as lipoma. At MR imaging, liposarcomas appear predominantly bright with T1-weighted sequences, iso intense with T2-weighted sequences, and dark with fat saturation (Fig 7b). At CT, the lesions demonstrate fat attenuation. Well-differentiated liposarcomas are usually round or lobulated, displacing or surrounding normal structures. They frequently contain septa, as well as occasional nonadipose, solid-appearing regions. These nonfatty areas are hypointense relative to skeletal muscle on T1-weighted images and iso- to hyperintense on T2-weighted images. The solid-appearing regions are poorly defined, with no clear demarcation between them and fat. Calcifications or ossification within a liposarcoma have proved to be a sign of poor prognosis, often indicating dedifferentiation.^{9,10}

Preoperative biopsy is often not necessary. However, a tissue diagnosis is made from core needle or open-incision biopsy specimens in cases in which therapy would be altered.

Such cases include a suspected lymphoma or germ cell tumour (both of which respond to chemotherapy), a tumour for which preoperative irradiation or chemotherapy is planned, or an apparently unresectable tumour.

Surgery remains the cornerstone therapy for both primary and recurrent retroperitoneal liposarcomas. Although the goal is complete resection with tumour-free margins, large tumour size and involvement of critical structures limit the resectability of these tumours and make precise delineation of the status of their margins. Complete surgical resection frequently requires en bloc resection of adjacent viscera kidney (36%) followed by colon(22%), spleen (10%),pancreas(9%), small bowel, stomach(6%) and IVC(3%).²

The median survival following complete primary resection is 103 months and following incomplete primary resection is around 18 months⁴. Complete resection rates ranges from 62%-86%.The primary pattern of treatment failure is local recurrence which occurs usually within 2 years. Local recurrence rate ranges from 32%-82%⁶. The predictor of overall survival is tumour grade and tumour size >15cm.The predictor of recurrence is tumour size and vascular involvement.

The addition of adjuvant radiotherapy to surgical resection is associated with reduced risk of local recurrence, longer recurrence- free interval but there is no impact on overall survival⁷.The advantages of preoperative radiotherapy are the gross tumour volume is readily definable for accurate treatment planning. The tumour displaces the radiosensitive viscera outside the radiotherapy field. The radiation dose believed to be biologically active is lower in the preoperative setting. There will be no adhesions or tethering of bowel to tumour bed.^{7,8}

The role of chemotherapy in retroperitoneal liposarcoma is experimental. There is no survival benefit^{3,9}. Most of the local recurrences occur within 2 years post operatively. Patients with first local recurrence with no metastases can be taken up for reexploration and excision. Median survival after local recurrence in patients following resection is 60 months and those without resection is 20 months

⁸. Distant metastases depend on tumour grade, gross & microscopic margins of resection. Metastases occur hematogenously to lung and liver. In pulmonary metastases the median survival duration is 6-12 months. In hepatic metastases survival rates are less than those observed for resection of pulmonary metastases¹. Surveillance is mainstay after the treatment with the goal of early detection of local recurrence, hepatic, pulmonary metastases by physical examination and imaging.⁴

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