

**Case Report:**

**Retroperitoneal lipomatosis with sarcomatous change: A case Report**

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**Abstract:**

Retroperitoneal lipomatosis is a benign condition involving overgrowth of mature fat tissue in extraperitoneal compartment of abdomen with incidence is 1.7 case per 1,00,000 population. It observes more commonly in male. However very few cases have been reported in Indian population. Herewith an adult male with 50 years, came with fullness and distension of abdomen associated with anorexia and lower abdominal pain. He was advised USG abdomen, which shows diffuse hyperechoic lesion in abdomen displacing bowel loops. Few anechoic areas seen within lesion on left side. No significant vascularity seen. Liposarcoma from retroperitoneum is relatively uncommon, our case is associated with diffuse peritoneal lipomatosis, few active nodules on PET CT, it was resistant to chemotherapy and size was almost near to that of giant liposarcoma. However during surgery it was removed in toto and no obvious invasion of adjacent organs seen which was also evident on histopathology of tumor which showed well differentiated variant without dedifferentiation. Follow up US/CT shows areas of persistent lipomatosis in peritoneal cavity with few focal areas of altered density suggestive of fat necrosis.

**Keywords:** Retroperitoneal lipomatosis, diffuse hyperechoic lesion, Liposarcoma

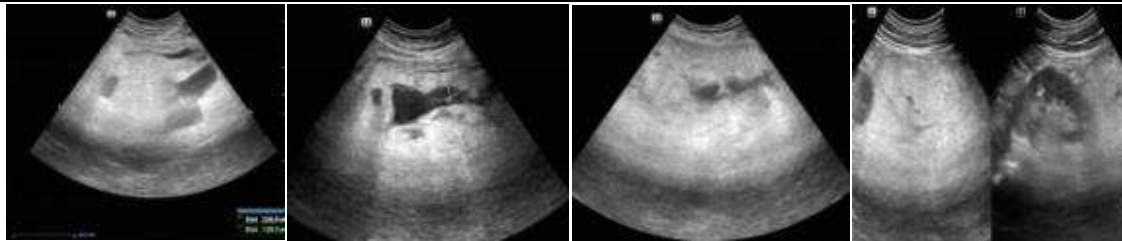
**Background:**

Retroperitoneal lipomatosis is a benign condition involving the overgrowth of mature fat tissues in the extraperitoneal compartment of abdomen. It predominantly affects the perirectal and perivesical regions. <sup>1</sup>Retroperitoneal lipomatosis is a benign condition involving overgrowth of mature fat tissue in extraperitoneal compartment of abdomen with incidence is 1.7 case per 1,00,000 population. It observes more commonly in male. However very few cases have been reported in Indian population. Herewith an adult male with 50 years, came with fullness and distension of abdomen associated with anorexia and lower abdominal pain.

**Case report:**

Clinically he has diffuse distension of abdomen.

He was advised USG abdomen, which shows diffuse hyperechoic lesion in abdomen displacing bowel loops. Few anechoic areas seen within lesion on left side. No significant vascularity seen.



USG shows diffuse hyperechoic lesion in abdomen with few anechoic areas, no vascularity.

CT abdomen and pelvis was advised.

Thin axial CT scan of the abdomen and pelvis was performed without and with administration of intravenous contrast. Sagittal and coronal reconstructions were obtained.

Diffuse and extensive noncapsulated fatty areas seen in peritoneal cavity with few thin septae within. It is displacing bowel loops to periphery and right side of abdomen. A large thinly capsulated heterogeneous but predominantly fatty density lesion seen on left side of retroperitoneum in relation with lower pole of left kidney. It shows some soft tissue density areas within. This lesion measures 24x20x19.6cm. It is displacing left kidney superiorly.

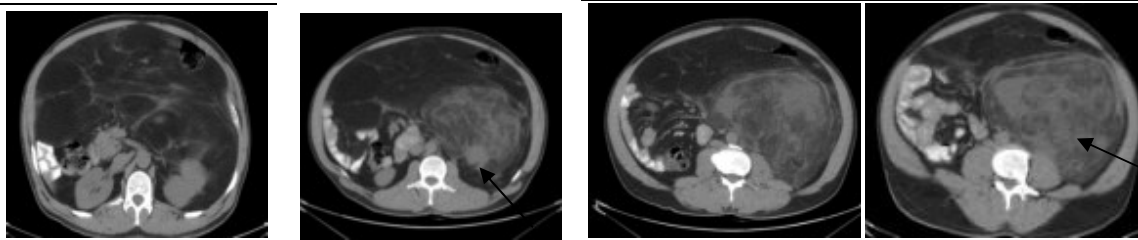
No evidence of abnormal enhancement in these areas. No evidence of calcification.

**Findings suggestive of diffuse intra-abdominal lipomatosis as described.**

Thinly capsulated predominantly fat density lesion with heterogeneous appearance from left side of retroperitoneum suggest possibility of liposarcomatous lesion.

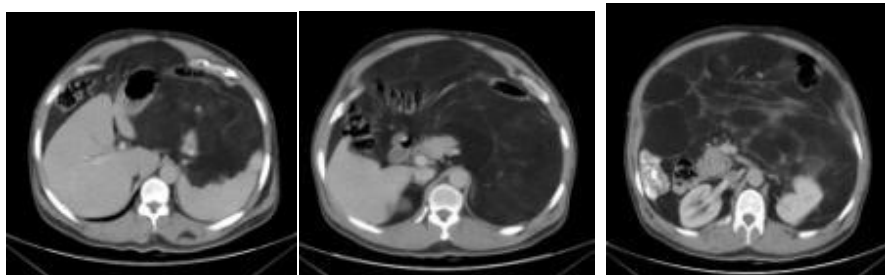
Rest of the organs show unremarkable features.

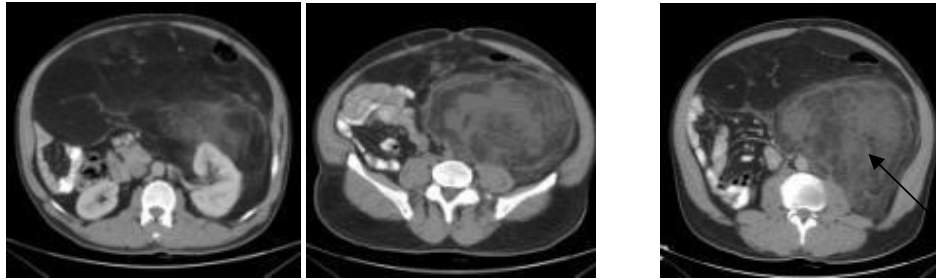
**Plain CT abdomen and pelvis.**



Diffuse lipomatosis seen in all sections with a focal altered density lesion in relation with left kidney.

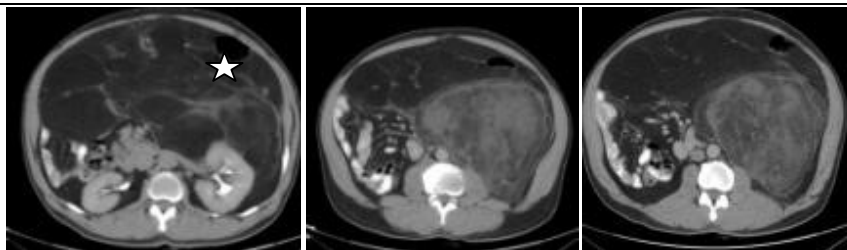
**CONTRAST CT**





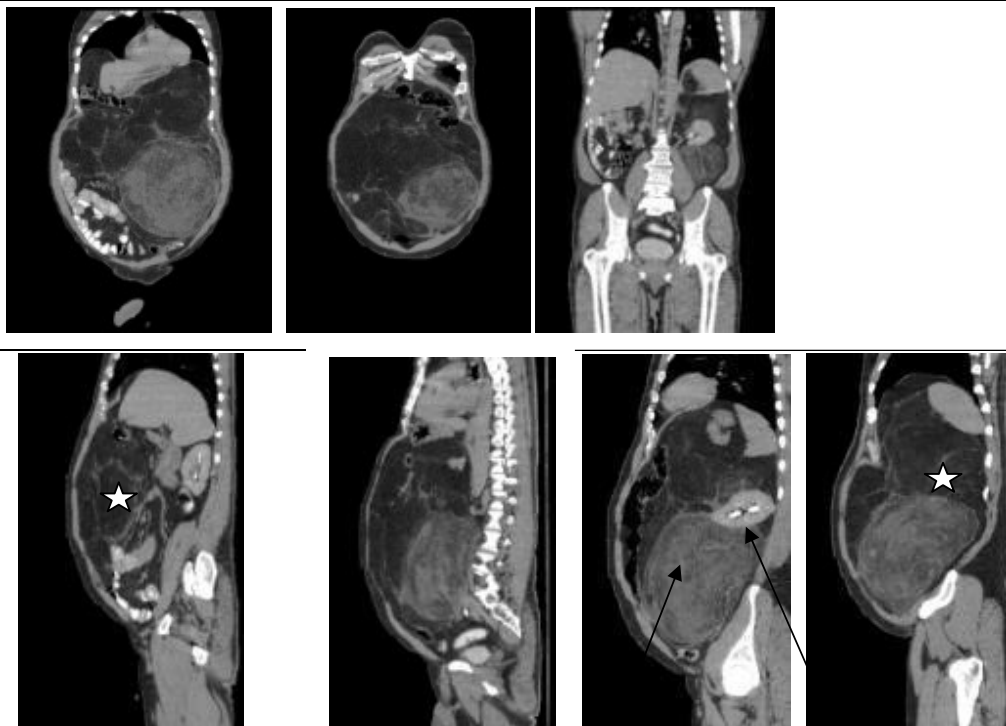
Contrast CT shows no significant enhancement in focal capsulated lesion.

CONTRAST ENHANCED DELAYED PHASE



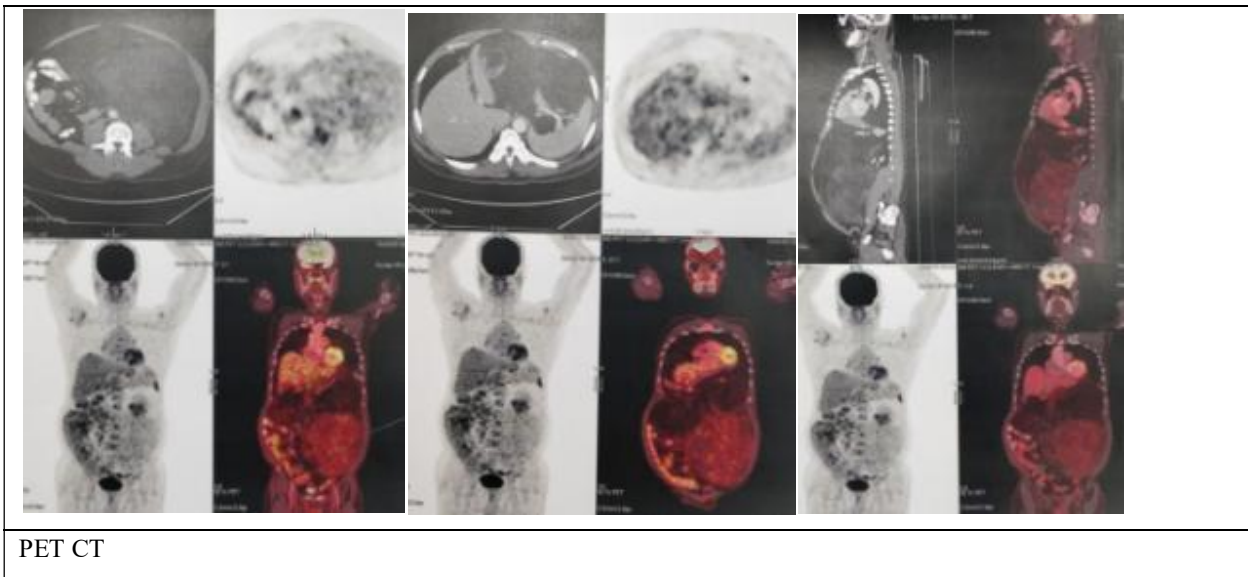
Lesion remains unenhanced, no areas of calcification.

SAGITAL AND CORONAL RECONSTRUCTIONS



Sagittal and coronal reformats show relation of capsulated lesion with left kidney (arrow) and diffuse lipomatosis (star).

PET CT was done and it shows abnormal minimal increased FDG uptake seen in heterogeneously enhancing diffuse and extensive soft tissue mass in left lumbar region without any calcification and soft tissue density areas within causing mass effect over adjacent bowel loops and retroperitoneal structures. (suv MAX 3.4). Abnormal focal increased FDG uptake seen in a few small soft tissue nodules within omental fat, largest of size 11x8mm. these findings suggest liposarcoma with soft tissue nodules suggestive of omental fat-liposarcomatous deposits.



Patient was posted for chemotherapy.

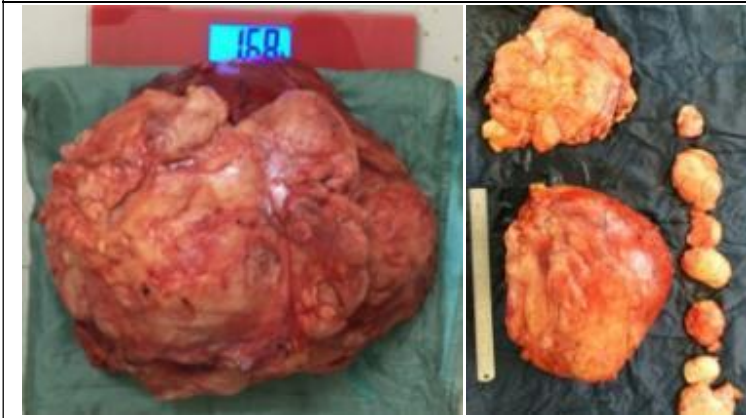
Four cycles of chemotherapy were done.

Follow up CT was done which show no change in mass and lipomatosis part as compared with previous report.

So the patient was subjected to surgery with possibility of left nephrectomy explained during surgery.

However during surgery the large capsulated mass was removed (almost 17 kg weight) with few other fat components and left kidney was spared.

**Post operative specimen**



**Discussion:**

In this rare case of liposarcoma, histopathology came as well differentiated liposarcoma, no areas of dedifferentiation. It was not invading perirenal fat so left kidney was spared during surgery. Retroperitoneum is most common site for liposarcomas(2), other being extremities, it can invade retroperitoneal structures and also project in peritoneal cavity. other common sites being extremities, intrathoracic and subcutaneous tissue. Although Liposarcoma was previously classified into five subtypes, recent developments in molecular biology and genetic mapping now divides in three major categories. The first category includes atypical lipomatous tumor (ALT), well-differentiated LPS, and dedifferentiated LPS (3). The second category includes myxoid LPS and round cell LPS. The third category is composed of pleomorphic LPS.

Well differentiated liposarcoma has more subtypes but radiologically no reliable criteria to classify(4). These have low grade malignant potential and do not metastasise unless areas of dedifferentiation are seen within (2). Our case was well differentiated liposarcoma associated with diffuse peritoneal lipomatosis. That means it contained highest amount of fat. No obvious areas of dedifferentiation were observed on histopathology as it was suspected on PET CT and contrast CT (enhancing nodules within mass).

Our case is different from Previously reported liposarcoma cases, as it is also associated with diffuse peritoneal lipomatosis along with a large focal liposarcoma mass. Also the focal mass can be labeled as giant liposarcoma by its dimensions and weight.

Other liposarcomas can have purely myxoid form, the pleomorphic and round cell varieties where soft tissue, myxoid, osseous, leiomyomatous or rhabdosarcomatous component with predominate. The presence of a large amount of homogenous fat suggests a low degree of malignancy, whereas extensive area of necrosis suggests rapidly growing aggressive course(2). CT is modality of choice and it is highly specific for fat component in the lesion. It can also detect other components like myxoid (low density areas), leiomyomatous (soft tissue density areas), osseous (calcification) within mass, also subtle density difference can be picked up. Also invasion of adjacent structures and planning for surgery can be done by prior CT. however MRI will be more specific for presence of fat within lesion(4) and to rule out invasion of vital structures like vessels (3).

Areas of dedifferentiation can occur in post operative cases and can be easily diagnosed by follow up CT. These are generally low grade masses and do not metastasise(5).Surgery is mainstay of treatment (5) with chemotherapy and radiotherapy as adjuvant. Recurrence is common, more with aggressive tumors.

**Conclusion:**

Liposarcoma from retroperitonem is relatively uncommon, our case is associated with diffuse peritoneal lipomatosis, few active nodules on PET CT, it was resistant to chemotherapy and size was almost near to that of giant liposarcoma .

However during surgery it was removed in toto and no obvious invasion of adjacent organs seen which was also evident on histopathology of tumor which showed well differentiated variant without dedifferentiation.

Follow up US/CT shows areas of persistent lipomatosis in peritoneal cavity with few focal areas of altered density suggestive of fat necrosis.

**References:**

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