Archives of Surgical Research | Case Report

Huge Retroperitoneal Liposarcoma: Old Beast in Modern Era

Iqra Imtiaz, Ahmad Kaleem

IMPORTANCE Retroperitoneal liposarcomas are amongst the commonest soft tissue sarcomas in the retroperitoneum. They can reach enormous sizes before presentation to a hospital. Contrast-enhanced CT scan is of utmost importance in estimating the location, extent, and depth of the malignancy. An image-guided biopsy can be of great help in the planning of treatment plans and in clinically equivocal cases. Complete Ro resection is of paramount importance in treating this aggressive disease entity. Radiotherapy and chemotherapy in the pre-operative stage can help in sensitive tumors. Molecular therapies are in the trial phase to strengthen treatment options. Here we present an interesting case, discuss its management and provide a literature review.

KEYWORDS Retroperitoneal liposarcoma, Enbloc resection,

HOW TO CITE Imtiaz I, Kaleem A. Huge Retroperitoneal Liposarcoma: Old Beast in Modern Era. *Archives of Surgical Research*. 2022, 3 (1):49-52. https://doi.org/10.48111/2022.01.09

Case Report

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https://doi.org/10.48111/2022.01.09

Soft tissue sarcomas constitute 1% of all adult malignancies.10-15 % of these are in the retroperitoneum. Liposarcomas are the most common variant comprising 20% of all soft tissue sarcomas and 50% of retroperitoneal variety. Most of these sarcomas do not have a clearly identified cause with possible etiology including genetic changes and exposure to ionizing radiation and chemical substances[1].

The median age at diagnosis is 56 years with equal gender distribution or slight female preponderance. They have been associated diabetes mellitus in 25% of cases. According to ESMO, only 10% of these malignancies metastasize but these are probably one of the largest neoplasms in the human body. Amongst the four types of retroperitoneal liposarcomas, the well-differentiated subtype is the most common variant of RPS (46%), followed by the myxoid (28%), dedifferentiated (18%), and pleomorphic subtypes (8%). They are generally discovered as abdominal mass (66-80%) or vague abdominal pain as they can grow for a long time in an expandable retroperitoneal space. The symptomatology can vary from gastrointestinal, urological, and neurological compressive symptoms[2].

The most common genetic mutations seen in retroperitoneal liposarcomas are MDM2/CDK4 amplification (12q13-15), presence of FUS-DDIT3 fusion gene, HMGA2 mutation, FRS2 amplification and downregulation of genes such as LIPE, PLIN and PLIN2[3]. Mortality rates for patients with liposarcoma range from 1% to 90%, and recurrence rates range from 5% to 83% depending on the histologic subtype and location[4]. Contrast enhanced CT scan abdomen, chest and MRI abdomen plus pelvis are the most invaluable investigations in these malignancies to determine origin, extent of local and distant organ involvement[5].

The differential diagnoses of retroperitoneal tumors are: lymphomas, testicular carcinomas and germ cell tumors, among others[6]. Surgical resection with negative margins of these tumors is the mainstay of treatment. Radiotherapy and chemotherapy are planned with multidisciplinary meetings[7].

CASE REPORT

A 54-year-old male presented to us with eight months history of gradual onset large abdominal swelling with significant unintentional weight loss. He had complaints of back pain but no history of altered bowel or urinary habits, fever or night sweats with itching and lumps in the body. There was no family history of such swellings or tumors of digestive, respiratory or biliary tracts. Family history for soft tissue malignant tumors was negative. He did not have any history of exposure to ionizing radiations.

The patient had a medical history of diabetes mellitus, hypertension, and ischemic heart disease with good compliance and control. On abdominal examination, there was a large abdominal swelling 30 x 25 cm, occupying most of the right hypochondrial, lumbar and iliac regions with prominent overlying abdominal veins. The swelling was firm in consistency and not moving with respiration. It was not falling forwards in the knee-elbow position. There was no evidence of free fluid or organomegaly in the abdomen. Normal bowel sounds were heard to the left of the umbilicus. Lymphadenopathy was not present. There was no evidence of varicocele, hernial orifices were intact and testicular examination was unremarkable. The distal neurovascular status was intact, and examination of spine was unremarkable. The chest examination was unremarkable, and jaundice was not present. Right ankle edema was present. His laboratory investigations were unremarkable except Hb of 10.3g/dl. A contrast enhanced CT scan of abdomen and pelvis revealed a large 27 x 24 x30 cm heterogeneous mass seen originating from the right retroperitoneal space, displacing the right kidney anteriorly and medially, filling almost the entire abdominopelvic cavity with displacement of the bowel loops on the left side. Inferiorly, the mass was seen reaching up to the level of the lower sacral piece approximating the distended urinary bladder. There were fatty areas with enhancing septae and soft tissues in the mass. A core biopsy of the mass confirmed CT scan impression of retroperitoneal sarcoma with positive CD34 and S-100 cells. Clinical TNM stage was IIIB (T4, N0, M0). After anesthesia clearance for surgery, enbloc resection of retroperitoneal liposarcoma with right nephrectomy of grossly involved kidney was carried out.

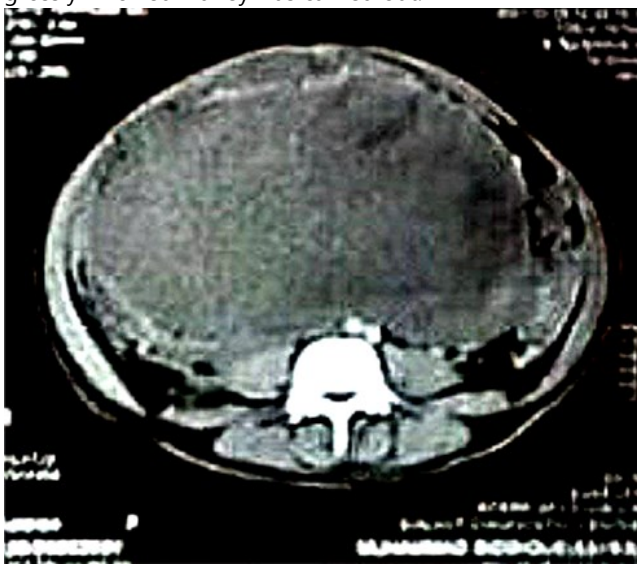

Figure 1: CT scan abdomen showing large retroperitoneal tumor displacing bowel loops

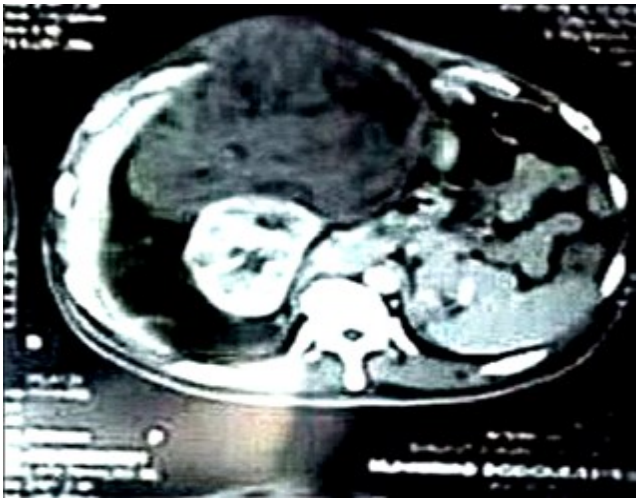

Figure 2: A CT scan image (upper slice) showing tumor displacing right kidney.

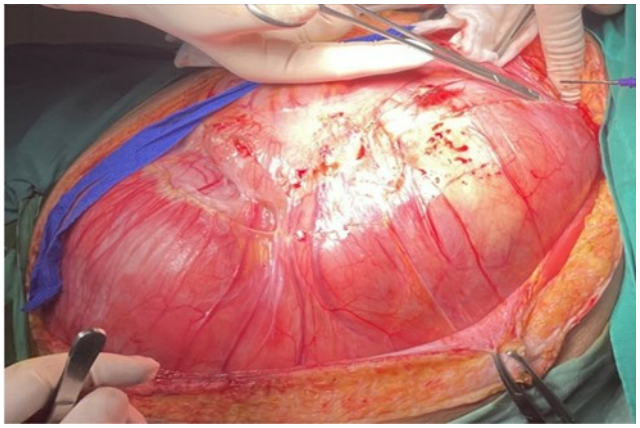

Figure 3: Intraoperative view of huge retroperitoneal sarcoma.

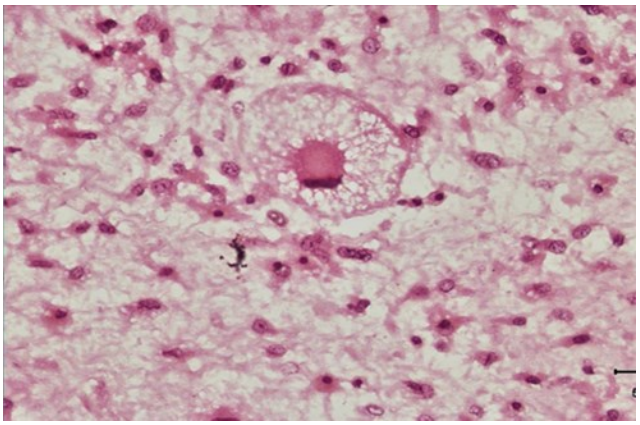

Figure 4: Microscopy reveals many lipoblasts with a central nucleus and vacuolated cytoplasm in a loose myxoid stroma.

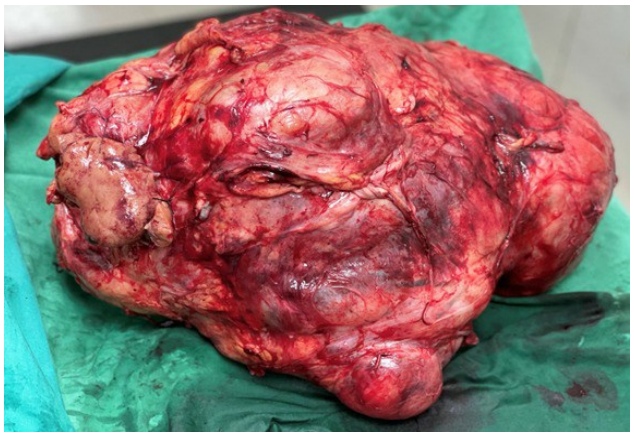

Figure 5: Cut section reveals solid tumor with variegated appearance with hemorrhagic and yellowish fatty areas.

Histopathology report revealed a huge (44x36x34cm) retroperitoneal myxoid liposarcoma with positive S-100 and p 53. No lymphovascular invasion, necrosis was identified. Pathological TNM was T4, Nx, Mx with grade 2 to 3.

The post-operative course remained uneventful, and patient was discharged for follow up (through imaging of chest, abdomen every 3-6 months for 2 years, then every 6 months for the next 2 years, then annually) and referred for further care to department of medical and radiation oncology.

DISCUSSION

The average annual incidence of retroperitoneal sarcomas is 2.7 cases per million population. It has 36-58% over all 5-year survival rate with a natural history characterized by late recurrence. Locoregional recurrence remains a frequent cause of death. Only 28% of patients do not experience recurrence in 5 years[8].

We found case report of retroperitoneal sarcoma of size 30cm in literature[9]. On the contrary, in our case, maximum size dimension of tumor was 44cm. Cases of local progression with colonic perforations have been reported which require implementation of judicious clinical workup strategies for prevention[10].

Liposarcomas have got a different behavior from other sarcomas in being indolent and death occurring principally from local progression than distant metastasis. Seventy percent of these require multivisceral resection, most commonly of kidney (32%) and colon (25%). Studies focusing on growth rate of these tumors have found that the ones with growth of 0.9cm per month had improved survival after aggressive resection of local recurrence[11].

In our case, we removed right kidney enbloc with the tumor because of its involvement. Liberal visceral enbloc resection has been able to achieve good local control[12].

The mainstay of management for liposarcomas is surgical resection with negative margins. Complete resection has been associated with a 5-year survival rate of 70%[13]. The post-operative margin status is single most important prognostic factor for disease free survival. One study reports 72 month median overall survival in primary retroperitoneal tumors, 28 months for recurrent cases and 10 months for metastatic disease[14].

The tumor may be deemed unresectable when it invades aorta or inferior vena cava, viscera, spinal cord, root of mesentery and peritoneal implants[15]. Perioperative radiotherapy, in particular pre-operative variety, has been able to lessen recurrence with no effects on distant metastasis and overall survival[16]. Chemotherapy, particularly in neoadjuvant form may be deployed in chemo sensitive tumors in which complete resection is uncertain[17]. Novel therapies such as CDK4/6 inhibition, PPAR-γ agonists, MDM2 inhibitors, PI3K and mTOR inhibitors, and inhibition of selective nuclear export inhibitors, multikinase inhibitors and immunotherapy are also in use in trial phases[18].

In a study, 1- year, 3- and 5-year survival rates have been reported to be 86%,66% and 57% respectively. The median overall survival is around 6 years. Important factors involved in predicting decreased survival are high grade, tumor rupture, gross disease and positive resection margins. Compartmental resection by high volume surgeons is of paramount importance in lowering recurrence rate to 3.29-fold compared with simple resection[19].

To conclude, retroperitoneal liposarcomas can reach enormous sizes before presentation. Enbloc compartmental resection is the way to go in curative approach to these malignancies.

ARTICLE INFORMATION Accepted for Publication: April 14, 2022,
Published Online: 00 00, 2022.
https://doi.org/10.48111/2022.01.00O

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Financial Support and Sponsorship: Nil.

Conflicts of Interest: There are no conflicts of interest

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