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Lipoma Like Liposarcoma

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INTRODUCTION

Soft tissue sarcomas are rare, unusual neoplasms accounting for <1% adult human cancers. They occur frequently in the extremity, followed by the trunk, retro peritoneum, head and neck respectively.

Most Retroperitoneal tumors malignant and one third are soft tissue sarcomas.

Amongst these, liposarcomas, malignant fibrous histiocytomas, leimyosarcomas are common.

Here, we are discussing a case report of our patient who presented with a retroperitoneal liposarcoma weighing 5 kgs.

CASE PRESENTATION

54 year old female patient presented with chief complaints of abdominal distension for 20 days. She had no other associated complaints. On examination, there was a vague mass extending from the suprapubic region to the epigastrium which did not move on respiration. Per rectal and per vaginal examinations were normal.

INVESTIGATIONS

Blood investigations were found to be within normal limits.

USG abdomen showed a fat attenuated lesion occupying the abdominopelvic region.

CECT whole abdomen showed- A large heterogeneous fat density mass (32x22x16cm), occupying almost the entire abdomen, displacing the adjacent bowel loops peripherally, displacing the kidneys and pancreas posteriorly. No areas of calcification/ encasement of infiltration of vessels or nerves seen within.

F/S/O Retroperitoneal liposarcoma.

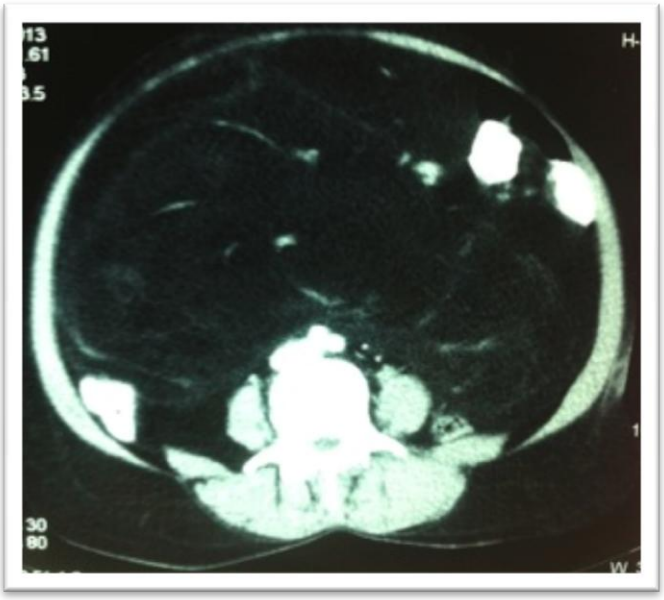

Fig -1 Axial view of the tumor occupying the entire abdomen

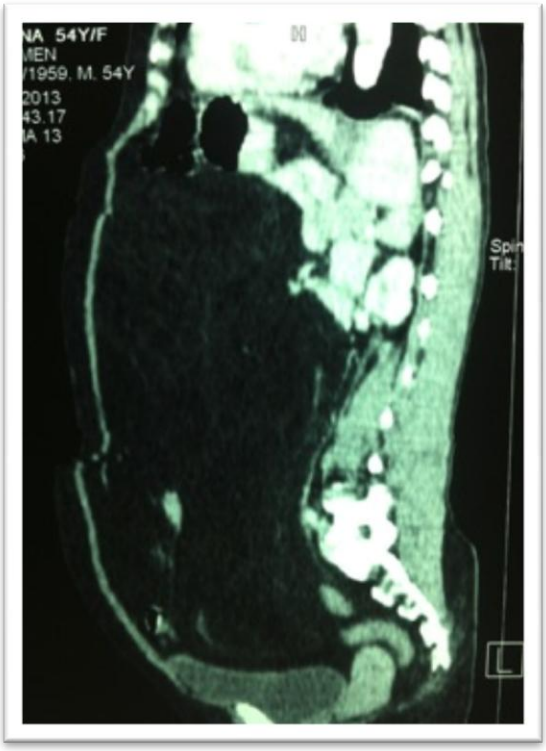

Fig -2 Sagittal view of the tumor occupying the entire abdomen.

TREATMENT

We planned on an exploratory laparotomy for the patient. DJ stenting was done. Following which, under general anesthesia, via a midline scar, the abdomen was opened. Intra operatively, a 50x30x15cm mass, arising from the retroperitoneum, from the root of the mesentry, displacing the bowel anteriorly, posteriorly and left laterally. The mass was not infiltrating any surrounding structures. Ligating the feeding vessels, we excised the mass. A drain was placed and abdomen was closed. The patient recovered well. Diet was started on pod 2, drain was removed on pod 4 and she was discharged on pod 7. DJ stents were removed after 2 weeks.

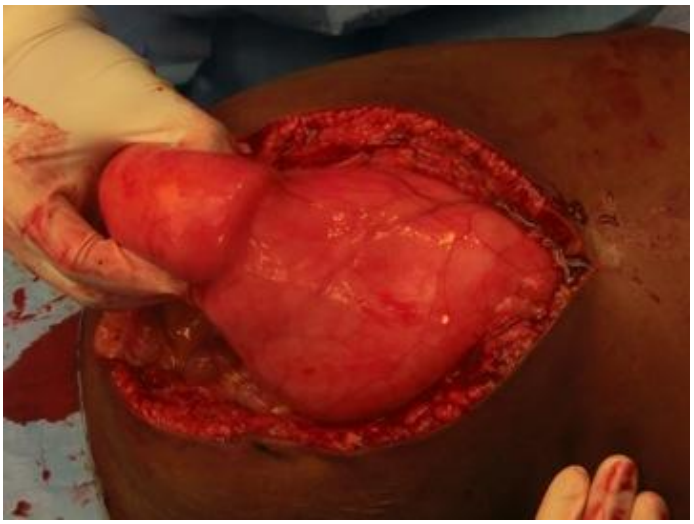

Fig 3-The Intra operative picture of the retroperitoneal tumour

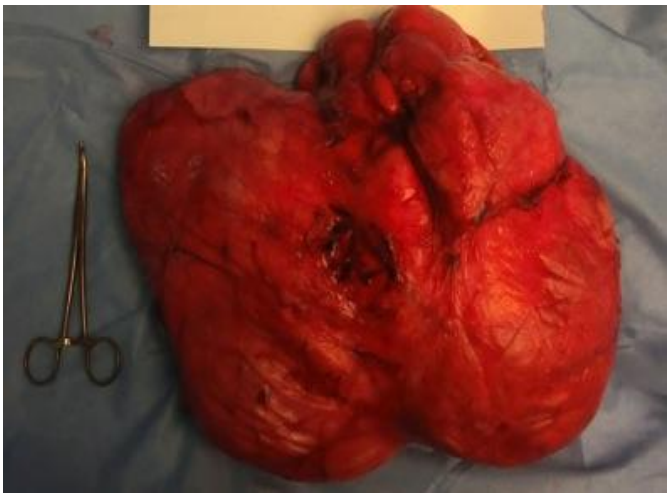

Fig -4 Liposarcoma- the excised specimen weighing 5 kgs

PATHOLOGY

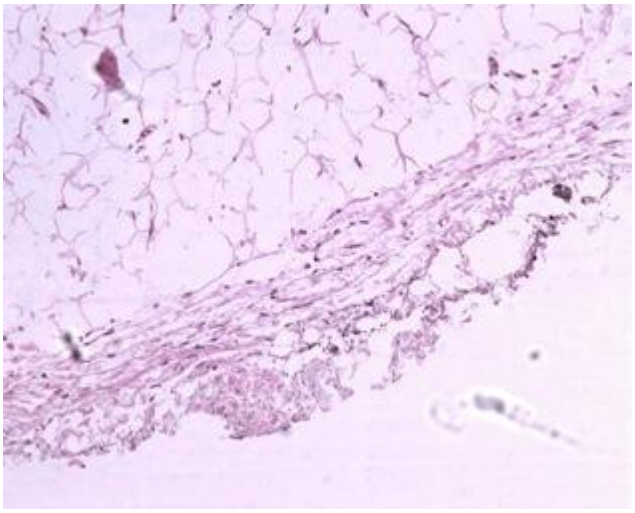

Fig -5 Histopathology -hematoxylin and eosin sections showing lobules of Mature adipocytes with a few cells resembling lipoblasts

Well differentiate dlipoma like Liposarcoma (retroperitoneum)

pT2NxcMX-grade 1

Mitotic rate- <1 mitoses/10hpf

Tumor necrosis- score 0

DISCUSSION

Retroperitoneal sarcomas are rare malignancies with an estimated incidence of 1000 cases per year in the United States. There are no anatomical compartments in the retroperitoneum, hence these neoplasms often grow to a large size and involve multiple adjacent structures before diagnosis. [1] Liposarcoma comprises 41% of the retroperitoneal tumors and about 20% of them are >10cms at diagnosis.[2]

Liposarcoma occurs most commonly in the extremities (52%), retroperitoneum (19%), and inguinal region (12%) [3]. They are of mesodermal origin and they attain large sizes despite their poor vascularization, weighing over 100 pounds. In literature, 18 and 42 kg liposarcomas have been recorded. [3]. They present with vague abdominal symptoms, weight loss , abdominal mass or obstructive symptoms.

Liposarcomas are subdivided into four well recognized subgroups based on morphology and cytogenic abnormalities: well differentiated, dedifferentiated, myxoid/round cell, and pleomorphic[3]

CT and MRI are the imaging modalities of choice for retroperitoneal tumors. They define the extent of the tumor and invasion into the adjacent structures. In suspicion of a malignancy, staging can be done with a chest x ray or a ct thorax for lung metastases and an ultrasound abdomen. Careful consideration of cross sectional imaging and history is they key in determining whether a biopsy is indicated of not. If preoperative systemic therapy or radiotherapy is deemed to be potentially useful, then a biopsy maybe done. [12]

For lesions that appear to be well differentiated retroperitoneal liposarcomas, as the sensitivity of CT is nearly 100%, there is virtually no role for preoperative biopsy. FNAC is indicated for recurrent tumors.

Primary management is the excision of the tumor with negative margins. In cases of infiltration, en bloc resection might include excision of the surrounding structures. The kidney is the most common structure to be excised. Laparoscopic surgery should be aggressively indicated in cases pre-operatively diagnosed as low-grade liposarcoma; however, the surgery should be meticulously indicated in cases of high-grade

neoplasms, like pleomorphic liposarcoma. Well-differentiated, small, and well-capsulated retroperitoneal liposarcoma's are the best candidates for a laparoscopic approach. [8]

Neither chemotherapy nor radiotherapy arecurative[13,14]. They have shown little efficacy in the treatment of retroperitoneal liposarcomas. There is no prospective randomized controlled trial confirming the potential benefit of radiotherapy that emerges from retrospective studies [11,15]. Adjuvant radiation is often not an option when given to large tumors due to the truncal location of retroperitoneal sarcomas and the doses required would be high. Adjuvant chemotherapy yields little benefit. In high-grade disease, administration of adriamycin and ifosfamide may yield partial responses in up to 50% of patients with increased overall survival; however, complete responses are seen in less than 10% of patients [7,11]

Locoregional recurrences remain the main cause of death in patients with retroperitoneal liposarcoma.[9,10]. Excision/ Debulking still remains the treatment of choice for recurrences. [6,9,10]

From the literature, the overall 5-year survival for well-differentiated subtypes is 90%, while 5-year survival for pleomorphic subtypes is only 30-50%. De-differentiated and myxoid/round cell subtypes have intermediate 5-year survival rates of 75% and 60-90%, respectively. Well-differentiated liposarcomas may recur locally, but metastatic potential is low. Pleomorphic liposarcomas have high metastatic potential, accounting for the decreased rate of survival[7]. Well differentiate dliposarcomas and myxoidliposarcomas have a good prognosis and their rates of metastasis are low compared to the other types of liposarcomas. [4,5]

Despite advances in medical care, the primary treatment of retroperitoneal liposarcomas is excision with negative margins. Post operative chemotherapy or radiotherapy depends on the surgical margin and the histopathology.

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