## **GAINT RETROPERITONEAL LIPOSARCOMA: A RARE CASE REPORT**

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**INTRODUCTION:** Retroperitoneal liposarcoma is a rare malignancy arises from perirenal fat comprises of 0.02 to 0.2% of all malignancies and 10-20% of all soft tissue sarcomas.<sup>[1,2,3]</sup> Usual age presentation is 5-6<sup>th</sup> decade of age with slight male predominance.<sup>[4]</sup> These tumors are usually large, in 20% of patients the tumor is more than 10cms size.<sup>[5,6]</sup> The liposarcoma may have weight and dimension variable; those over 20 kg are called "giant liposarcomas" and are extremely rare.<sup>[1,5]</sup> Complete surgical resection [R0] is the only way for survival advantage.<sup>[7,8,9]</sup> We report a case of huge retroperitoneal liposarcoma, which of size of 30><25><20cms and 8kg weight. Few cases have been reported of this size in literature.<sup>[4, 10]</sup>

**CLINICAL DETAILS:** A 60 yrs old female presented with progressive distension of abdomen since 2 yrs. On examination firm, lobulated mass felt all quadrants of abdomen, there is dull note on percussion. On knee elbow position the mass not falling forward, lower border of mass not felt. All routine blood investigations are within normal limits. US abdomen reveals single hyper echoic mass found occupying all parts of abdomen. A CECT abdomen shows a 30><25><20cms heterogenous soft tissue dense mass filling the whole part of abdomen and pelvis, pushing the right kidney anteromedially and bowels laterally left of abdomen.

An US guided core biopsy revealed a well differentiated liposarcoma.

Patient was subjected for surgery, on laparotomy there was massive fatty tumor occupying all parts of abdomen pushing the small bowels to left side and engulfing right kidney but not infiltrated. A meticulous dissection performed and most of the mass excised after carefully separating from the adherent bowels and right kidney, which was preserved. Post op period was uneventful. Patient was discharged on 7<sup>th</sup> post op day.

Patient was followed up to 2yrs with regular 6 monthly interval, CECT abdomen was our choice of imaging for follow up. There was no evidence of recurrence.

The post-operative biopsy report came as a well differentiated liposarcoma, with sclerosing variant, margins were negative.

**DISCUSSION:** Liposarcoma is derivative of primitive or embryonal lipoblastic cells, arising from perirenal fat is very uncommon.<sup>[1,2,3]</sup> They grow slowly and silently with unimpending growth and attains enormous size, symptoms are due to pressure effects and complications are late.<sup>[11,12,9]</sup> Haematogenous spread is rare, lung is the most common organ for metastasis.<sup>[11,12,9]</sup>

There are three histological types described<sup>[13,14,15]</sup>

Atypical Myxoid [most common] Pleomorphic Atypical - well differentiated and dedifferentiated.

Well deferentiated -- sclerosing, spindlecell, adipocytic, and inflammatory, low grade – local infiltrative and low metastatic potential.

Undifferentiated and pleomorphic are high grade, with biological aggressiveness and metastatic potential.<sup>[11,12,9]</sup>

CECT abdomen is the investigation of choice, but sometimes MRI would help in doubtful diagnosis and in assessing recurrences, along with satellite deposits. Percutaneous biopsy does not yield accurate diagnosis especially in dedifferentiated.<sup>[5,6,16,17,18]</sup> The resection of a retroperitoneal sarcoma of remarkable size is a challenge for the surgeon owing to the anatomical site, to the absence of an anatomically evident vascular-lymphatic peduncle that makes it hard to obtain safe margin and to the adherences with the contiguous organs and with the great vessels. Therefore the retroperitoneal liposarcoma shows a high rate of local recurrence after surgical excision. Actually, the complete surgical (R0) resection represents the only possibility of radical treatment, in fact as reported in a study<sup>[15]</sup> carried out on 177 patients with retroperitoneal liposarcoma operated with curative intent, the percentage of patients disease free at 3 and 5 years was 73% and 60% respectively.<sup>[9,10,11,12]</sup>

Surgery with wide excision is the treatment of choice. If the excision is incomplete, to palliate, tumor can be debulked.<sup>[7,8]</sup> Recurences are common, re-excision sometimes considered, but will not have survival advantage.<sup>[10]</sup> Chemotherapy is not effective, but radiotherapy has limited role as postop treatment but again there is no survival advantage.<sup>[9, 19]</sup> Radiotherapy would cause more damage to abdominal viscera after surgical debulking. As any other soft tissue sarcoma the prognosis depends upon margin status and histological grade. Mortality is due to recurrence and repeated surgery, bowel resections and its morbidity.<sup>[9,19]</sup>

Actually the overall survival at 5-years reported in literature for the various histological subtypes well differentiated, myxoid/round cell, undifferentiated and pleomorphic, ranging from 90%, 60 to 90%, 75% and 30 to 50%, respectively.<sup>[9]</sup> the overall 5yrs survival is 54%.

Follow up usually with CT abdomen and pelvis with CXR]. Following surgical resection, the 50 - 100% of liposarcomas recur from residual tissue, which is the primary cause of death.<sup>[10]</sup>

High grade – every 4 months for first 3yrs and there after every 6 months for next 2 yrs. Low grade – every 6 months for 5-6yrs and every one year for next 2-3 yrs.

**CONCLUSIONS:** Retroperitoneal liposarcomas are rare malignancies, CECT abdomen is the investigation of choice. Surgery is the main modality of treatment, tumor grade and resectional status are important for prognosis. Chemo and radiotherapy has limited role and mortality is due recurrences. Patient is kept under regular follow up.

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