# A huge retroperitoneal mass histologically diagnosed as well differentiated liposarcoma-a case report

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**Abstract:**
A 45 year old female presented with huge abdominal swelling (subsequently diagnosed as a retroperitoneal mass (12kg) by ultrasonogram and CT scan), gradual weight loss and respiratory distress for last one year. She had also complaints of anorexia, epigastric pain, and hiccough for one year. Her retroperitoneal mass was resected and sent for histopathological examination which was later diagnosed as well differentiated liposarcoma.

**Key words:** Liposarcoma, retroperitoneal mass

## Case report

A 45 years old menstruating housewife hailing from Kapasia, Gazipur was admitted to a private clinic with the complaints of gradual swelling of abdomen, weight loss and respiratory distress for last one year. At first the swelling was in epigastric region and small in size , then the swelling increased in size gradually over one year and involved all region of abdomen and flanks. She had associated loss of appetite, dyspepsia, heart burn, gradual weight loss for one year. She mentioned that the problems got worse day by day and the swelling increases in size rapidly for last two months leading to mechanical distress of breathing manifested by tachycardia and breathlessness on lying position. and weakness of both lower limbs. She is normotensive, non diabetic, non asthmatic, afebrile and her blood group was B+ve . On examination the abdominal lump was very large in size involving whole abdomen and flanks. It was nontender, lobulated and firm in consistency. It was slightly mobile up & downwards. Other abdominopelvic organs were not palpable. Patient is ill looking, cachectic & severely anaemic. Respiration was mechanic. Hematological investigations, liver function tests, screening for HBV and HCV were normal except her Hb was 9.2gm/dl and ESR over 100mm/1st hour. USG of whole abdomen showed hugely enlarged homogenous area occupying whole abdomen and also homogeneous area 180-190mm in left upper abdomen separated from left kidney and spleen. Para aortic lymph nodes were enlarged. CT scan of whole abdomen report mentioned a large teratodermoid lesion in abdomen. Patient was prepared for surgery after taking all precautions and preoperative checkup and on 9/9/08 she was operated and the lump removed.

## Peroperative findings

Abdomen was opened by long midline incision. After opening the abdomen a huge multi lobulated, solid mass arising from the posterior retro peritoneal wall (L1-L5) extending from lesser sac downwards to the pelvis pushing transverse colon, descending colon, stomach and pancreas forwards. The mass was firmly adherent with surrounding structure particularly the spleen. The huge mass was excised except a small portion of tissue surrounding the mesenteric great vessels. Spleen was also removed with the mass. After proper hemostasis abdomen was closed in layers keeping a drain tube in situ. The excised mass was 12kgs in weight.The patient received . 3 units of whole blood during and after operation.The patient was given Injection Ceftriaxone 15gm/BD and injection Pneumovax along with pain killers and sedatives.

## Postoperative recovery

Post operative recovery was uneventful. The patient improved dramatically within 10 days and was discharged with proper medication and advice for follow up after 6 months and advised not to do heavy work for this period.

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## Histopathology of the mass:

*Gross:*

Specimen consists of multiple oval grayish yellow pieces of tissue, all are covered with capsule. The largest one measuring 18 X15 X 12 cm. The cut surfaces of the tumors are homogenous and yellowish in color with areas of hemorrhage and necrosis. 12 blocks are embedded (Figure-3).Resected Spleen was unremarkable grossly and weigh 500 gms.4 blocks are embedded.

## Microscopic Examination:

Multiple sections from the submitted tumor show fibrofatty and fibrocollagenous tissue. These reveal many atypical cells within the fibrous strands and adipose tissue lobules. A few lipoblasts are seen arranged in lobules, sheets and nests. Areas of myxoid degeneration are also noted. In some areas the tumor cells are arranged in fibrosarcomatous pattern. Mitosis is infrequent. A diagnosis of well differentiated liposarcoma was made (Figure1,2). Sections from spleen show congestion. No tumor extension is seen.

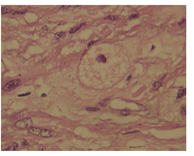

**Figure 1.** Liposarcoma. Showing Lipoblast (H&E X40)

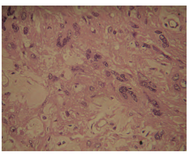

**Figure 2.** Photomicrograph of Liposarcoma (H&E X40)

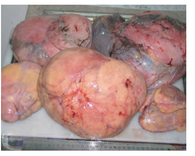

**Figure 3.** Gross specimen of the retroperitoneal mass. Variable sizes of mass, grayish yellow in colour and encapsulated.

## Review of literature:

Liposarcoma is one of the most common sarcomas of adulthood and appear in 40- 60's,uncommon in childhood. It usually arise in deep soft tissues of proximal extremities and retro peritoneum and notorious for developing into large tumors. It has also been reported in foot 2.

## Pathophysiology

Liposarcoma is a lipogenic tumor of large deep-seated connective tissue spaces. Fusion proteins created by chromosomal abnormalities are key components of mesenchymal cancer development. An abnormality of band 12q13 has been associated with the development of liposarcomas. The most common chromosomal translocation is the FUS-

CHOP fusion gene, which encodes a transcription factor necessary for adipocyte differentiation. The well differentiated variant is relatively indolent, myxoid is intermediate and round cell & pleomorphic variants are aggressive and frequently metastasize. All types recur locally and often repeatedly unless adequately excised. The most important microscopical finding is the presence of lipoblasts. Lipoblasts- mimic fetal fat cells contain round clear cytoplasmic vacules of lipid that scallop the nucleus





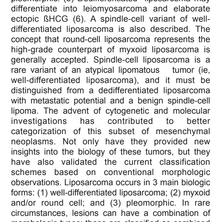

**Figure 4.** Ultrasonogram (USG) finding of abdomen of the patient with retroperitoneal mass showing entanged homogenous aea occupying whole abdomen.

The development of a liposarcoma from a preexisting benign lipoma is rare. Most cases arise de novo. Liposarcomas most frequently arise from the deep-seated stroma rather than the submucosal or subcutaneous fat. The most recent World Health Organization classification of soft tissue tumors recognizes 5 categories of liposarcomas: (1) well differentiated, which includes the adipocytic, sclerosing, and inflammatory subtypes; (2) dedifferentiated; (3) myxoid; (4) round cell; and (5) pleomorphic. (3) Dedifferentiated liposarcoma may differentiate into leiomyosarcoma and elaborate ectopic ßHCG (6). A spindle-cell variant of well-differentiated liposarcoma is also described. The concept that round-cell liposarcoma represents the high-grade counterpart of myxoid liposarcoma is generally accepted. Spindle-cell liposarcoma is a rare variant of an atypical lipomatous tumor (ie, well-differentiated liposarcoma), and it must be distinguished from a dedifferentiated liposarcoma with metastatic potential and a benign spindle-cell lipoma. The advent of cytogenetic and molecular investigations has contributed to better categorization of this subset of mesenchymal neoplasms. Not only have they provided new insights into the biology of these tumors, but they have also validated the current classification schemes based on conventional morphologic observations. Liposarcoma occurs in 3 main biologic forms; (1) well-differentiated liposarcoma; (2) myxoid and/or round cell; and (3) pleomorphic. In rare circumstances, lesions can have a combination of morphologic types; these are classified as combined or mixed-type liposarcomas 4.

The anatomical distribution of liposarcoma appears to be partly related to the histologic type. Well-differentiated liposarcoma tends to occur in deep soft tissues of both the limbs and the retroperitoneum. Myxoid and/or round-cell liposarcomas and pleomorphic liposarcomas have a striking predilection for the limbs, and dedifferentiated liposarcoma occurs predominantly in the retroperitoneum. Although any liposarcoma subtype occasionally arises in the subcutis, involvement of the dermis appears to be exceedingly rare. The overall 5-year survival rate of patients with deep high-grade liposarcoma is less than 50%. Metastases are common, especially in poorly differentiated liposarcomas. The lungs and the liver are the most common sites of metastasis. Primary cutaneous liposarcoma has an indolent course. No association with race or geography is known. Liposarcomas are slightly more common in males than in females 1.The mean patient age at onset is 50 years. Although liposarcomas account for about 17% of all soft tissue sarcomas, they are involved in only 4% of childhood soft tissue sarcomas. Cases of liposarcoma are reported in young adults and teenagers, but cases in children are rare.

Patients usually note a deep seated mass in their soft tissue. Only when the tumor is very large do symptoms of pain or functional disturbances occur. Retroperitoneal tumors may present themselves with signs of weight loss and emaciation and abdominal pain. These tumors may also compress the kidney or ureter leading to kidney failure.

Incidence/ Prevalence. Most frequent in middle-aged and older adults (age 40 and above), liposarcomas are the most common of all soft-tissue sarcomas. Annually 2.5 cases occur per million population. The prognosis varies depending on the site of origin, the type of cancer cell, the tumor size, the depth, and proximity to lymph nodes. Metastases are common. The 5 year survival rate for a high-grade liposarcoma is less than 50%.

## Conclusion

The most common disease of a retroperitoneal mass is fibrosarcoma. However Liposarcoma should also come as a differential diagnosis and histo-pathological examination is a must to come to a conclusive diagnosis. Early diagnosis will help to define better treatment plan in these patients.





## Reference

1. Rosai and Ackerman's Surgical Pathology.Editor.Juan Rosai.9th Edition.Publisher.Mosby Elsevier 2004, pp2280
2. Luka Brcic, Antonija Jakovcevic, Lovorka satalja valetic Dubravko onlic, and sven sciaerth. Pleomorphic lipooacroma of the foot a case report. Diagn Pathol. 2008; 3: 15. Published online 2008 April 16. doi: 10.1186/1746-1596-3-15.
3. Liposarcoma-Wikipedia, the free encyclopedia http://en.wikipedia.org/wiki/Liposarcoma
4. Liposarcoma : Article by Robert A Schwartz. www.emedicine.com/derm/topic856.htm
5. Michael J. Russell,Frederick L. Flynt, Allyson L. Harroff, and Oluwole Fadare, Dedifferentiated Liposarcoma of the Retroperitoneum with Extensive Leiomyo sarcomatous Differentiation and B Human Chorionic Gonadotropin Production
Sarcoma. 2008; 2008: 658090. Published online 2008 March 12. doi: 10.1155/2008/658090
6. Robbins & Cotrans, Pathologic Basis of Disease, editor. Kumar V, Abbas AK and Fausto N.7th edition, 2004, pub.WB Saunders Ltd, USA,pp1318