CASE REPORT

GIANT RETROPERITONEAL LIPOSARCOMA: IMAGING AND LITERATURE

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ABSTRACT: Liposarcoma is the most common retroperitoneal sarcoma among the common three retroperitoneal sarcoma that is liposarcoma, leiomyosarcoma and malignant fibrous histiocytoma. Liposarcoma accounts around 40% of such cases. Liposarcoma has got unique property that they frequently attain large size before they diagnosed due to painless slow growth. Here we report a case with an extremely large well differentiated retroperitoneal liposarcoma that occupying entire abdomino-pelvic cavity with characteristic computed tomographic findings and review of the relevant literature.

KEYWORDS: Retroperitoneal Sarcoma, Liposarcoma, Computed Tomography.

INTRODUCTION: Retroperitoneal liposarcoma is the most common malignant tumor of mesenchymal origin where mesenchyma abnormally differentiates into bulk of adipose tissue. Retroperitoneal liposarcomas are remarkable because of their frequently large size (>10-20 cm) when they diagnosed. They are among the largest tumors of the body.^[1] Mesenchymal retroperitoneal sarcomas constitute mainly liposarcoma (40-41%), leiomyosarcoma (28-30%), malignant fibrous histiocytoma (7-16%), and rest are others.^[2] Most liposarcoma occur in deep soft tissue, with slow expansive growth, while lipomas are frequently from superficial soft tissue.^[3] Most common grade is well differentiated and rare variety is pleomorphic round cell. Early surgery is the best way to treat but with the use of neoadjuvant radiation therapy may be able to decrease the rate of onset of local recurrence of tumor after surgical removal.^[4] The main purpose of this study is to report a case of huge abdomino-pelvic advanced retroperitoneal liposarcoma which was unresectable on the basis of computed tomographic study with review of relevant literature.

CASE REPORT: A 56 years old woman presented with progressive enlargement of abdomen since 3 years ago and recent onset of frequent constipation, abdominal pain and low grade persistent fever. There is also relative progressive weight loss in the form of muscular wasting and unduly enlarged abdomen.

On clinical examination, patient's all vital signs are unremarkable with visible fullness of entire abdomen. Palpable lump was noted having lobulated contour but full margin could not be delineated.

Her laboratory examinations were also unremarkable except low haemoglobin (6.9gm%). Patient was referred to radiology department for further evaluation by whole abdominal sonography. Ultrasonography showed large soft tissue mass extending from epigastrium to pelvis. The mass was diffusely echogenic and lobulated associated with displaced hydronephrotic right kidney and compressing retroperitoneal vessels. For proper characterization and extension of large mass a complimentary computed tomography (CT) of the abdomen was done on 16 slice multidetector scanner without contrast, with contrast and delayed phase.

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Non-contrast, contrast enhanced, sagittal and coronal reformatted scans (Fig. 1, 2, 3, & 4) demonstrating large mass entirely occupying the abdominal cavity displacing the retroperitoneal structure dorsally and bowel loops are sandwiched and trapped by mass. The mass was measuring around 59x25x32 cm, having multiple lobules with distinct zone of fat and intervening thick septa particularly in lower abdomen and pelvic cavity while other locules having solid soft tissue density with indistinct fat component. The mass appeared adhered with bowel loops, retroperitoneal vasculatures, muscles and having large size right parietal wall metastatic mass. There was also adherence with pelvic structures and anterior bladder wall associated with multiple retroperitoneal lymphnodes. Thus the lesion was reported as advanced giant retroperitoneal liposarcoma on the basis of fat density, and was inoperable. To know the histologic type ultrasound guided biopsy was performed and a well-differentiated type liposarcoma with components of non-lipogenic elements was reported.

Since tumor was in advanced stage, it was considered inoperable. She was referred to oncology department for further assessment. She was advised to start with chemotherapy, include Adriamycin and Ifosfamide for at least six cycles to know the clinical and radiological response. She and her attendant were also made aware about the nature of disease, and its prognosis, as well cost and side effect of chemotherapy as they belonged from low socio-economic strata, and told to come next week for chemotherapy schedule. But unfortunately she was lost to follow-up treatment.

DISCUSSION: Retroperitoneal liposarcoma is the most common histological subtype of retroperitoneal sarcoma accounting for around 40% and mostly occurs in middle age group (5th decade onward). Liposarcomas mostly occur in deep soft tissue while lipoma more commonly from superficial soft tissue.^[3] Retroperitoneal liposarcomas originate from primitive mesenchymal cells and not from adipocytes. Conceptually liposarcomas are divided into three subgroups: first group includes well-differentiated and dedifferentiated, in second group myxoid and round cells and in third group pleomorphic histology.^[3]

Liposarcoma has unique property of being large size when it is diagnosed and mostly having a mean diameter of 20cm. Local recurrence is frequent after resection and responsible for patient's morbidity and mortality. Metastases mostly involving liver and lung, accounts for less than 10% while with other retroperitoneal sarcomas metastases are more common.

Well-differentiated liposarcoma is the most common subtype and together with dedifferentiated subtype accounting 35-40% of liposarcomas. Well differentiated liposarcoma usually large multilobular and confused with lipoma except large size and numerous thick septa, gelatinous zones or punctate hemorrhage.^[3]

Well differentiated liposarcoma closely resemble renal angiomyolipoma in imaging. But angiomyolipoma can be differentiated on the basis of presence of renal parenchymal defect and thicker vascular pedicles supplying the tumor compare with liposarcomas.^[5] Well differentiated liposarcomas are mostly non-metastasizing mass of low grade histology compared to other subtypes, but local recurrence is very high (upto 100%).^[6] Histologically dedifferentiated subtype has same character as well-differentiated type but it has non-lipogenic components appearing as a high grade fibrosarcoma.^[3]

Myxoid liposarcoma is second most common subtype occurs in younger age groups (5th decade) and on histology shows myxoid matrix containing mucopolysaccharides with small amount

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of mature fats and large pool of extracellular mucoid material. Prognosis is good compared with dedifferentiated histology.

Pleomorphic and round cell are rare subtypes of retroperitoneal liposarcomas. They are heterogeneous and non-fatty tumors and indistinguishable from other soft tissue masses on imaging. The pleomorphic and round cell subtypes are highly malignant with a tendency towards local recurrence and metastasis.

Ultrasound rarely represents a specific diagnosis because there is considerable overlap in the sonographic appearances of retroperitoneal tumors, with most lesions being large and heterogeneous. Increased echogenicity within the retroperitoneal mass may be due to extensive fatty matrix, calcification, increased vascularity or hemorrhage. CT will often give a better overview of a retroperitoneal masses or liposarcomas.

On CT well differentiated liposarcoma contains similar in attenuation to macroscopic fat.^[7] On T1 and T2-weighted MR imaging liposarcomas are isointense to subcutaneous fat with loss of signal intensity on fat suppressed imaging sequence,^[7] but have subtle differences in the attenuation or signal intensity compared with normal abdomino-pelvic fat and helps in identification of tumor recurrence after resection.^[8] Well-differentiated liposarcoma demonstrates little or no contrast enhancement due to deficient vascularity. However thickened septa or minor nodular elements may enhance homogeneously.^[7] Area of necrosis and calcification uncommon. While dedifferentiated liposarcomas on CT and MRI imaging, have areas with attenuation and signal intensity characteristic of fat, like well-differentiated liposarcoma but they have areas of nonfatty masses with higher attenuation and low signal intensity on T1-weighted and high signal intensity on T2-weighted.^[9] In 30% cases calcification or ossification may be seen.^[9]

Myxoid variety on CT and MRI shows low attenuation, low signal intensity on T1-image and very high signal intensity on T2-image relative to muscle.^[10] Foci of fat may be seen in the myxoid mass. Myxoid subtype enhances appropriately on contrast because of significant vascularity. Pleomorphic and round cell liposarcomas on CT do not show fat component and similar to muscle attenuation and contain necrosis, hemorrhage and variable enhancement.^[7] On MRI they look heterogeneous on T1 and T2-weighted imaging and commonly reveal foci of intratumoral hemorrhage and necrosis.

Surgical resection is the gold standard method of treatment especially well-differentiated subtype. Complete tumoral resection along with adherent structures if any can only be the best method of treatment to improve the patient survival. But local recurrence is very common. Neoadjuvant radiation therapy may be able to decrease the rate of onset of local recurrence of tumor after surgical resection.

CONCLUSION: Retroperitoneal liposarcoma is the most common retroperitoneal mesenchymal tumor in middle age and accounting 40% of retroperitoneal sarcoma. Retroperitoneal liposarcomas have unique property of their large size (>20 cm). CT and MR are the most effective imaging procedures to know the extent of local and distant tumor invasion. Well-differentiated liposarcomas can be differentiated radiologically from other subtypes by their largely lipomatous appearance and malignant behaviour increases with degree of tumor's heterogeneity like loss of lipomatous matrix, necrosis and hemorrhage. For liposarcomas, wide surgical resection is the most effective way of treatment, preventing the recurrence and progression to another subtypes.

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REFERENCES:

1. Inoue K, Higaki Y, Yoshida H. Giant Retroperitoneal Liposarcoma. *Int J Urol.*, 2005; 12 (2): 220-222.
2. Lewis JJ, Leung D, Woodruff JM, Brennan MF. Retroperitoneal soft-tissue sarcoma: analysis of 500 patients treated and followed at a single institution. *Ann Surg.*, 1998; 228: 355-65.
3. Weiss SW, Goldblum JR: *Enzinger and Weiss's Soft Tissue Tumors*. St. Louis, Mosby. 2001.
4. Bossi A, De Wever I, Van Limbergen E, et al: Intensity modulated radiation-therapy for preoperative posterior abdominal wall irradiation of retroperitoneal liposarcomas. *Int J Radiat Oncol Biol Phys.*, 2007; 67: 164-170.
5. Israel GM, Bosniak MA, Slywotzky CM, et al: CT differentiation of large exophytic renal angiomyolipomas and perirenal liposarcomas. *AJR Am J Roentgenol.*, 2002; 179: 769-773.
6. Weiss SW, Rao VK: Well-differentiated liposarcoma (atypical lipoma) of deep soft tissue of the extremities, retroperitoneum, and miscellaneous sites: A follow-up study of 92 cases with analysis of the incidence of "dedifferentiation". *Am J Surg Pathol.*, 1992; 16: 1051-1058.
7. Kim T, Murakami T, Oi H, et al: CT and MR imaging of abdominal liposarcoma. *AJR Am J Roentgenol.*, 1996; 166: 829-833.
8. Francis IR, Cohan RH, Varma DG, et al: Retroperitoneal sarcoma. *Cancer Imaging.* 2005; 5: 89-94.
9. Tateishi U, Hasegawa T, Beppu Y, et al: Primary dedifferentiated liposarcoma of the retroperitoneum: Prognostic significance of computed tomography and magnetic resonance imaging features. *J Comput Assist Tomogr.*, 2003; 27: 799-804.
10. Neville A, Herts BR: CT characteristics of primary retroperitoneal neoplasms. *Crit Rev Comput Tomogr.*, 2004; 45: 247-270.

Fig. 1: Ultrasonography of abdomen shows large diffusely echogenic mass (fat tissue) encasing and displacing retroperitoneal major vessels.

Fig. 1

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Fig. 2a

Fig. 2b

Non-contrast CT scan (Fig. 2a&2b) abdomen showing large heterogeneous lobulated mass lesion with distinct zone of fat and thick fibrous bands (in pelvis), and solid soft tissue density that entirely occupying the abdominal cavity and displacing retroperitoneal structures postero-inferiorly. There is also adherence of mass with adjacent structures.

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Fig. 3a: (Sagittal reformatted CT scan images)

Fig. 3b: (Coronal reformatted CT scan images)

Fig. (3a & 3b): Demonstrating the large/giant retroperitoneal abdomino-pelvic mass from subdiaphragmatic to pelvis, having zones of fats and solid density lobules displacing the abdominal structures dorsally and superiorly. Bowel loops are compressed. Also a parietal mass (metastasis) seen at the level of umbilicus.

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Fig. 4a

Fig. 4b

Contrast CT scan of the abdomen (Fig. 4a & 4b) showing capsular enhancement of nonfatty solid density areas and thick fibrous bands of fatty zone in pelvic region. Also strongly enhancing parietal metastasis in right abdominal wall. The mass displacing and adherent with abdominal structures.

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