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Case Report *Diagnostic Radiology*

Cystic retroperitoneal dedifferentiated liposarcoma: A case report

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ABSTRACT

Liposarcoma is the most common primary retroperitoneal sarcoma in adults. We report the case of an 86-year-old male who presented to the emergency department with frequent falls and unexplained weight loss that was found to have a cystic retroperitoneal dedifferentiated liposarcoma. Initial computed tomography revealed a large heterogeneous complex cystic hypoenhancing lesion in the left retroperitoneum. Subsequent magnetic resonance imaging demonstrates a multilocular cystic mass with microscopic lipid content, diffusion restriction, and enhancing nodular soft-tissue components. Histologic examination of the tissue sample following biopsy is consistent with cystic retroperitoneal dedifferentiated liposarcoma. Further management was not pursued due to the patient's advanced age and frailty.

Keywords: Retroperitoneal neoplasms, Sarcoma, Liposarcoma, Hemorrhage

INTRODUCTION

Liposarcoma is a malignant tumor of adipose tissue and is the most common primary retroperitoneal sarcoma. Liposarcomas can be histologically divided into four subtypes including well-differentiated liposarcoma, myxoid liposarcoma, pleomorphic liposarcoma, and dedifferentiated liposarcoma.[1] The most common of these seen in the retroperitoneum are well-differentiated liposarcoma and dedifferentiated liposarcoma. Dedifferentiation liposarcomas are high-grade sarcomas and are more clinically aggressive than the well-differentiated subtype.[2] Cystic change in dedifferentiated liposarcoma is a very rare phenomenon. To date, only a few cases of cystic retroperitoneal dedifferentiated liposarcomas have been reported.[3-5] Herein, we describe a case of cystic retroperitoneal dedifferentiated liposarcoma complicated by hemorrhage.

CASE REPORT

An 86-year-old male presented to the emergency department complaining of acute shortness of breath as well as background unexplained weight loss over several months. The patient also reported recent high-impact falls which led to a right femoral subcapital fracture that was treated by right hip hemiarthroplasty in the preceding few months. Clinical assessment revealed peripheral edema and hypoxia but no fever, night sweats, or constitutional symptoms. Laboratory assessment showed increased serum alkaline phosphatase and D-dimer levels. While hemoglobin has been chronically low for months, there was no acute decline to suggest recent

hemorrhage. The remaining blood work including white cell count, neutrophil count, and C-reactive protein was normal. The initial chest radiograph was unremarkable.

A contrast-enhanced computed tomography (CT) scan of the abdomen and pelvis was obtained which showed a large heterogeneous complex cystic hypoenhancing lesion in the left retroperitoneum posterior to the left kidney measuring 13 × 12 × 24 cm and demonstrating mixed density of 9–32 Hounsfield units [Figure 1]. The mass displaces the left kidney and encases the left renal vasculature and collecting system at the hilum leading to mild hydronephrosis. While the appearance of the mass is non-specific on CT, given the recent history of multiple falls, a complicated chronic hematoma was considered in the differential at the time. Subsequent magnetic resonance imaging (MRI) demonstrates heterogeneous T2-weighted hyperintensity suggestive of multi-cystic areas within the mass [Figure 2]. Multiple areas of T1-weighted hyperintensity are also seen on the T1 fat saturated sequence suggestive of hemorrhagic content [Figure 2]. Chemical shift T1 imaging [Figure 3] demonstrates associated microscopic lipid content as denoted by signal loss on the out-of-phase image compared to the in-phase image in the superior cystic component of the mass [Figures 3a and 3b]. There is diffusion restriction in the periphery of the solid components of the mass [Figure 3d]. The solid components show avid enhancement in the post-contrast fat saturated T1 [Figure 4].

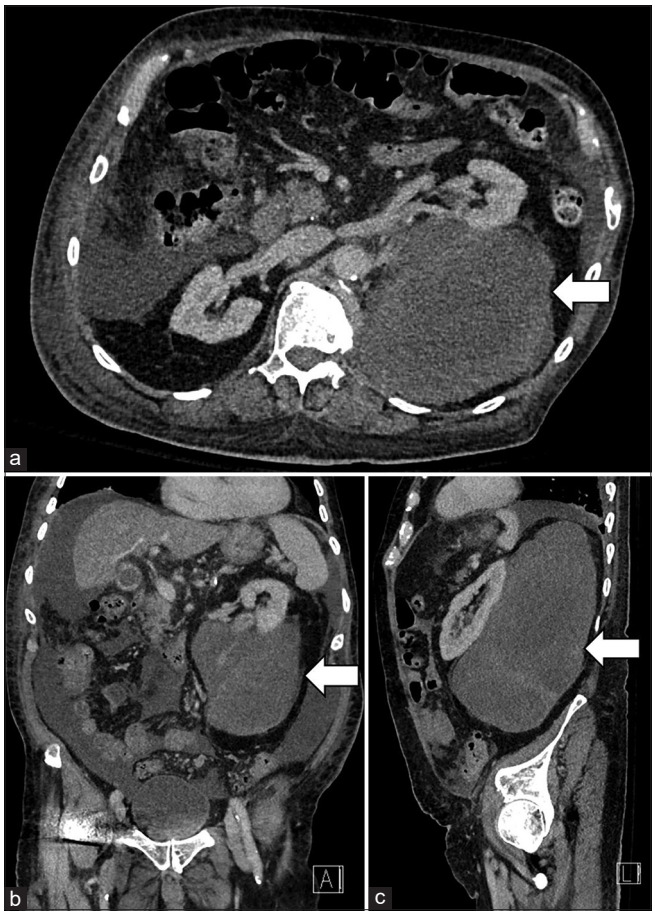

Figure 1: An 86-year-old male with cystic retroperitoneal dedifferentiated liposarcoma who presented with unexplained weight loss and frequent falls. Axial (a), Coronal (b), and Sagittal (c) contrast enhanced computed tomography (CT) images of the abdomen and pelvis in the portal phase demonstrate a large heterogenous complex cystic lesion in the left retroperitoneum (arrows). Differential considerations on CT included a complex hematoma, abscess, or a cystic tumor.

The findings on CT and MRI are most consistent with a large left retroperitoneal cystic neoplasm with internal hemorrhage of differing ages. Given the signal loss seen on the out-of-phase image [Figure 3b arrow], the first differential diagnosis considered is liposarcoma, followed by lymphoma or peripheral nerve sheath tumor. The patients underwent ultrasound-guided biopsy with subsequent pathology confirming a diagnosis of cystic retroperitoneal dedifferentiated liposarcoma. Given the advanced age and frailty of the patient, further management of this mass was not pursued.

DISCUSSION

Retroperitoneal sarcoma is rare with a mean incidence of 2.7/million though accounts for approximately 12–15% of all soft-tissue sarcomas.[6] They are frequently incidental findings

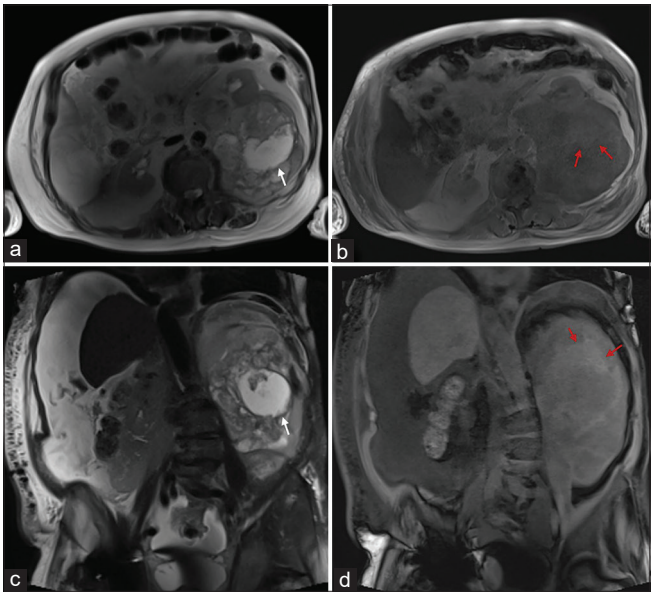

Figure 2: An 86-year-old male with cystic retroperitoneal dedifferentiated liposarcoma who presented with unexplained weight loss and frequent falls. Axial T2 (a), Axial T1 (b), Coronal T2 (c), and Coronal fatsaturated T1 (d) Images demonstrate heterogeneous mixed T2 signal intensity with foci of T2 high signal intensity throughout the lesion consistent with cystic components (white arrows). Multiple areas of T1 high signal suggesting hemorrhage of different ages (red arrows).

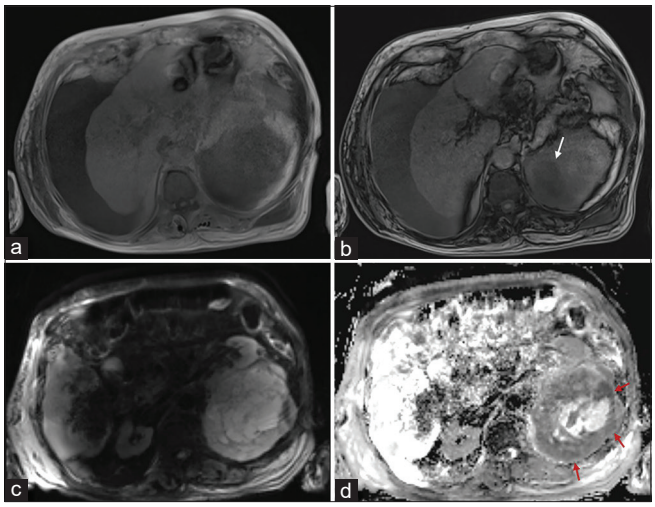

Figure 3: An 86-year-old male with cystic retroperitoneal dedifferentiated liposarcoma who presented with unexplained weight loss and frequent falls. T1 in-phase (a) and T1 out-of-phase (b) images as well as diffusion-weighted imaging (c) and apparent diffusion coefficient map (d) demonstrate foci of microscopic fat in the superior portion of the retroperitoneal mixed cystic mass (white arrow), and restricted diffusion in the periphery of the retroperitoneal heterogenous mixed cystic mass (red arrows).

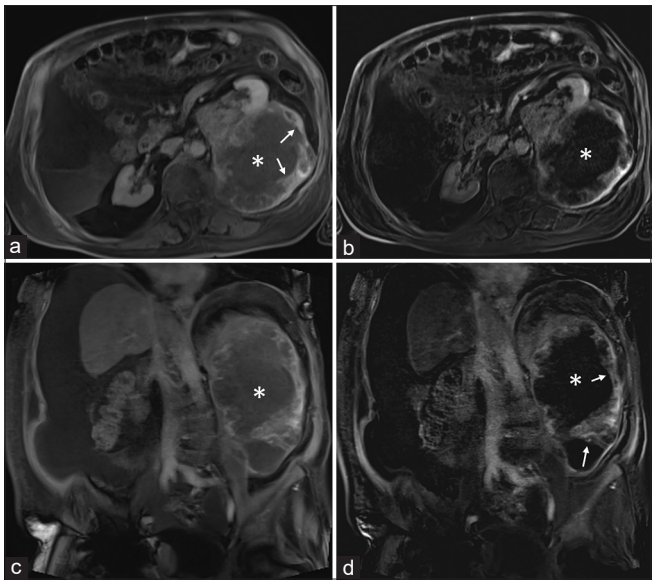

Figure 4: An 86-year-old male with cystic retroperitoneal dedifferentiated liposarcoma who presented with unexplained weight loss and frequent falls. Post contrast axial fat saturated T1 (a) and subtraction (b) as well as post contrast coronal fat saturated T1 (c) and subtraction (d) images demonstrate enhancement of the thick irregular peripheral walls of the mass and its thick internal septations (white arrows). No enhancement of the predominantly central cystic components (asterisk).

on imaging for non-related symptoms or diseases. Liposarcoma is the most common primary retroperitoneal sarcoma with well-differentiated and dedifferentiated liposarcoma being the two predominant subtypes.[1] Dedifferentiated liposarcoma is a high-grade sarcoma demonstrating more aggressive behavior compared to well-differentiated liposarcoma with the propensity for local recurrence and metastasis.[2] The characteristic imaging appearance of dedifferentiated liposarcoma is a mass with coexisting fatty and non-fatty tissue, often by a focal nodular non-lipomatous component >1 cm in size. Calcifications can be seen and are a sign of poor prognosis.[7]

Predominant cystic change is uncommon in retroperitoneal liposarcoma; hence, radiologists may not consider primary cystic retroperitoneal liposarcoma in the differential diagnosis. To date, only a few case reports of predominantly cystic retroperitoneal liposarcoma have been published in the literature. In a 2017 report by Uchihashi *et al.*, the authors described an elderly female who presented with a large cystic dedifferentiated liposarcoma without a solid component.[3] An additional case of cystic dedifferentiated liposarcoma was previously described by Khoury *et al.* in the setting of prior chemoradiation therapy.[4] The radiologic diagnosis of our case was difficult due to its large cystic component and non-specific appearance on CT. Given the patient history of frequent falls leading to recent right total hip arthroplasty, the heterogeneous appearance of the mass on CT makes hematoma a plausible differential diagnosis on the initial CT examination. The diagnosis of primary retroperitoneal neoplasm and more specifically liposarcoma was only made possible following MRI characterization.

Dedifferentiated liposarcomas represents the conversion of well-differentiated components to non-fat-derived tumor components and requires both lipogenic well-differentiated liposarcoma and cellular non-lipogenic sarcoma. However, well-differentiated and dedifferentiated liposarcomas share similar underlying genetic and histological characteristics including common genetic abnormalities with amplified sequences arising from the long arm of chromosome 12 (12q13-15), which includes amplifications of CDK4 and MDM2 cell cycle oncogenes.[8] These genetic and histological similarities contribute to a challenge in morphological differentiation of these tumors on imaging. The previous studies have investigated imaging features such as morphology of fat and non-fat components in liposarcomas. As shown in [Table 1], Rajiah *et al.* summarized the imaging features of different retroperitoneal masses.[9] Other imaging features described in both atypical lipomatous tumor/well-differentiated liposarcoma includes large size of the lesion (more than 10 cm), thick irregular/nodular septa, non-adipose areas or nodules, and fat content involving <75% of the lesion. Other described imaging characteristics for distinguishing between well-differentiated and dedifferentiated liposarcomas include prominent foci of high signal on fluid sensitive sequences and septal enhancement, although these features are shown to have limited specificities.[10] To date, no combinations of imaging features

Table 1: Imaging features suggestive of a specific diagnosis for retroperitoneal masses.

Imaging feature	Diagnosis
Pure fat-containing mass	Lipoma, well-differentiated liposarcoma
Heterogenous mass with fat	Dedifferentiated liposarcoma, myelolipoma, angiomyolipoma
Fat-fluid level	Teratoma, well-differentiated liposarcoma
Fat with calcification, teeth, or fluid	Teratoma
Myxoid stroma	Myxoid liposarcoma, neurogenic tumor, myxoid malignant fibrous histiocytoma. Less common diagnosis: desmoid tumor, hemangiopericytoma, leiomyoma or leiomyosarcoma, rhabdomyosarcoma, malignant mesenchymoma.
Large mass, extensive necrosis, invasion of IVC	Leiomyosarcoma
Fluid-fluid level caused by hemorrhage	Paraganglioma
Extremely hypervascular	Paraganglioma, hemangiopericytoma
Moderately hypervascular	Myxoid malignant fibrous histiocytoma, leiomyosarcoma, other sarcoma
Hypovacular	Lymphoma, low-grade liposarcoma, benign tumor
T2 hypointenisty	Lymphoma, desmoid tumor, small round cell tumor, retroperitoneal fibrosis, Erdheim-Chester disease
Paravertebral mass	Neurogenic tumor
Paravertebral mass, high catecholamine levels, hypertension	Paraganglioma
Extension between normal structures, encasement without luminal compression	lymphangioma, ganglioneuroma, lymphoma (gloating aorta or CT angiogram sign)
Extension along normal structures	Paraganglioma, ganglioneuroma
Mantlelike mass around aorta of IVC	Lymphoma, retroperitoneal fibrosis, Erdheim-Chester disease
floating aorta or CT angiogram sign	Lymphoma
Soft-tissue mass with calcification	Malignant fibrous histiocytoma, teratoma, extraosseous chondrosarcoma/Ewing sarcoma, synovial sarcoma, dedifferentiated liposarcoma
Cystic mass with solid tumor enhancement	Myxoid liposarcoma, schwannoma, neurofibroma
Cystic mass with slow, progressive enhancement	Lymphangioleiomyoma, urinoma
Cystic mass with pulmonary cysts	Lymphangioleiomyoma
Cyst after trauma	
With high attenuation or intensity	Hematoma
With obstruction	Urinoma
Cyst wit negative attenuation, history of radical lymphadenectomy	Lymphocele
Multilocular cystic mass, calcification, elongated shape, crossing retroperitoneal compartments	lymphangioma
Cystic mass, calcification, no contrast enhancement, bone lesions, visceral involvement	Lymphangiomatosis
Cyst, history of pancreatitis, high amylase level	Pancreatic pseudocyst
Cyst in obese woman receiving hormonal therapy for menstrual irregularity	Mullerican cyst
Unilocular cyst in subdiaphragmatic space	Bronchogenic cyst
Multilocular cyst, thick septa, calcification, right lower quadrant	Pseudomyxoma retroperitoneal
Presacral unilocular cyst	Epidermoid cyst
Presacral multilocular cyst	Tailgut cyst
Multilocular perianal cyst with history of anal fistual	Perianal mucinous carcinoma

are known to reliably differentiate between these lesions with definitive subtype diagnosis often requiring histological analysis following biopsy of the most suspicious portions of the lesion or even tumor resection.

Surgical resection remains the mainstay of curative therapy. Neoadjuvant or adjuvant chemotherapy could be considered in patients with large tumors (>5 cm) or high-grade round cell or pleomorphic subtypes. Neoadjuvant or adjuvant radiation therapy may be used for local control in large high-grade liposarcoma.[2]

CONCLUSION

We have presented an uncommon case of a predominantly cystic retroperitoneal dedifferentiated liposarcoma. While

liposarcoma is the most common primary retroperitoneal neoplasm, occurring in 33% of cases, they are most typically solid. Predominant cystic change is unusual in retroperitoneal liposarcoma; hence, why radiologists may not consider primary cystic retroperitoneal liposarcoma in the differential diagnosis of these retroperitoneal lesions. To date, only a few case reports of cystic retroperitoneal liposarcoma have been published in the literature. The diagnosis of primary retroperitoneal neoplasm and more specifically liposarcoma with a predominantly cystic component is only made possible following MRI characterization.

Declaration of patient consent

Patient's consent not required as patient's identity is not disclosed or compromised.

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Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Fletcher International Agency for Research on Cancer. WHO Classification of Tumours of Soft Tissue and Bone. Geneva: World Health Organization; 2013.
2. Dalal KM, Antonescu CR, Singer S. Diagnosis and management of lipomatous tumors. J Surg Oncol 2008;97:298-13.
3. Uchihashi K, Matsuyama A, Shiba E, Kimura Y, Ogata T, Yabuki K, *et al*. Retroperitoneal dedifferentiated liposarcoma with huge cystic degeneration: A case report. Pathol Int 2017;67:264-8.
4. Khoury M, Sim GC, Harao M, Radvanyi L, Amini B, Benjamin RS, *et al*. Multicystic dedifferentiated retroperitoneal liposarcoma: Tumour cyst fluid analysis and implications for management. BMJ Case Rep 2015;2015:bcr2015211218.
5. Horiguchi A, Oyama M. Perinephric liposarcoma mimicking cystic renal tumor. Nihon Hinyokika Gakkai Zasshi 2002;93:491-4.
6. Bonvalot S, Rivoire M, Castaing M, Stoeckle E, Le Cesne A, Blay JY, *et al*. Primary retroperitoneal sarcomas: A multivariate analysis of surgical factors associated with local control. J Clin Oncol 2009;27:31-7.
7. Francis IR, Cohan RH, Varma DG, Sondak VK. Retroperitoneal sarcomas. Cancer Imaging 2005;5:89-94.
8. Thway K. Well-differentiated liposarcoma and dedifferentiated liposarcoma: An updated review. Semin Diagn Pathol 2019;36:112-21.
9. Rajiah P, Sinha R, Cuevas C, Dubinsky TJ, Bush WH Jr., Kolokythas O. Imaging of uncommon retroperitoneal masses. Radiographics 2011;31:949-76.
10. Doyle AJ, Pang AK, Miller MV, French JG. Magnetic resonance imaging of lipoma and atypical lipomatous tumour/well-differentiated liposarcoma: Observer performance using T1-weighted and fluid-sensitive MRI. J Med Imaging Radiat Oncol 2008;52:44-8.

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