

## Retroperitoneal dedifferentiated liposarcoma with osteosarcomatous dedifferentiation

Vinh N. Nguyen, MD; Sarah Shaves, MD; and Janet Winston, MD

We present a case of a dedifferentiated liposarcoma with osteosarcomatous degeneration in a 46-year-old male. The mass, palpated during a routine physical exam, was analyzed with dynamic, contrast-enhanced CT and CT-guided core-needle biopsy before surgical resection.

### Case report

A 46-year-old Caucasian male initially saw his primary care physician for a routine physical exam. During the visit, an obvious large palpable mass was discovered in the central-to-left abdomen. The patient reported initially noticing the mass about six months before but was otherwise asymptomatic. A thorough review of systems was negative for constitutional symptoms, abdominal pain or back pain, nausea/vomiting, or changes in bowel habit. The patient's past medical, surgical, and family history were likewise noncontributory. There was no history of radiation treatment, anticoagulation therapy, or recent trauma.

A CT scan of the abdomen and pelvis demonstrated a large, heterogeneously enhancing retroperitoneal mass measuring 8.4 x 16.5 x 18.4 cm (Fig. 1A). The tumor was centered within and expanded the left perirenal space inferior to the left kidney. Given its large size, there was effacement of the anterior pararenal space, with anterolateral displacement and compression of the mid and distal descending colon. There was also extension past the midline, with the superior mesenteric artery and vein draped along its medial margin. The anterior and posterior renal fascias

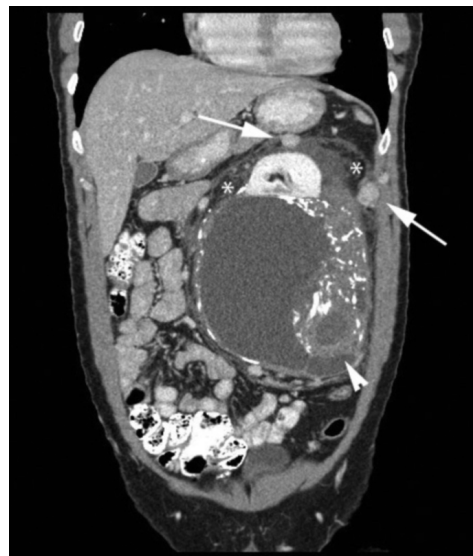


Figure 1A. 46-year-old male with a left retroperitoneal mass. Portovenous-phase, contrast-enhanced coronal image demonstrates extensive flocculent calcification and mass effect on the left kidney. Multiple enhancing thick septations (arrowhead), nodules (arrows), and fat-density lobules (\*) are scattered along the periphery of the lesion.

were poorly delineated, with multiple small, enhancing satellite nodules extending into the adjacent pararenal space (Fig. 1B). Superiorly, the lesion extended into the left upper quadrant, resulting in anterior and medial displacement of the left kidney and pancreatic tail. The mass was closely opposed to multiple retroperitoneal structures; however, there was no gross parenchymal invasion. There were extensive areas of curvilinear and flocculent mineralization along the periphery of the main lesion, with central areas

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Drs. Nguyen and Shaves are in the Department of Radiology at the Eastern Virginia Medical School, Norfolk VA. Dr. Winston is in the Department of Pathology at Sentara Norfolk General Hospital, Norfolk VA. Contact: Dr. Nguyen at [vinhnguyen@gmail.com](mailto:vinhnguyen@gmail.com).

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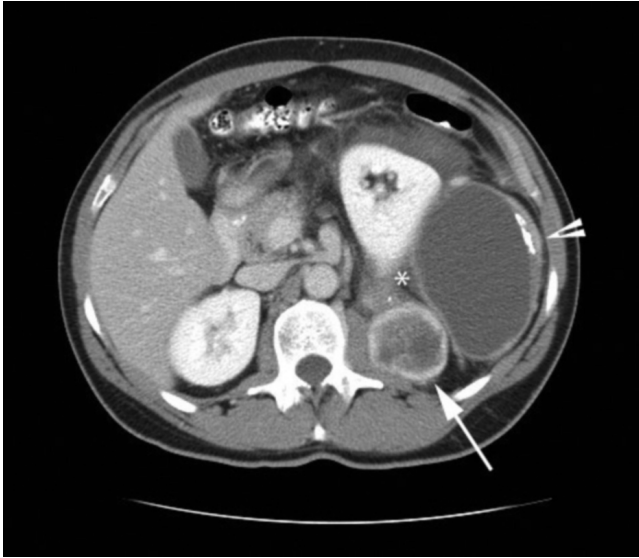


Figure 1B. 46-year-old male with a left retroperitoneal mass. Portovenous-phase, contrast-enhanced axial image demonstrates extensive flocculent calcification with multiple enhancing thick septations (arrowhead). A dominant heterogeneously enhancing nodule (arrow) abuts the left psoas muscle. The left kidney is displaced anteriorly and cephalad. A central island of fat-density tissue (\*) is wedged between the left kidney and the enhancing satellite nodule.

of low attenuation suggestive of necrosis (Fig 1C). Of note were multiple discrete foci with fat attenuation that were scattered peripherally along the lateral and inferior margins of the mass (Fig 1D).

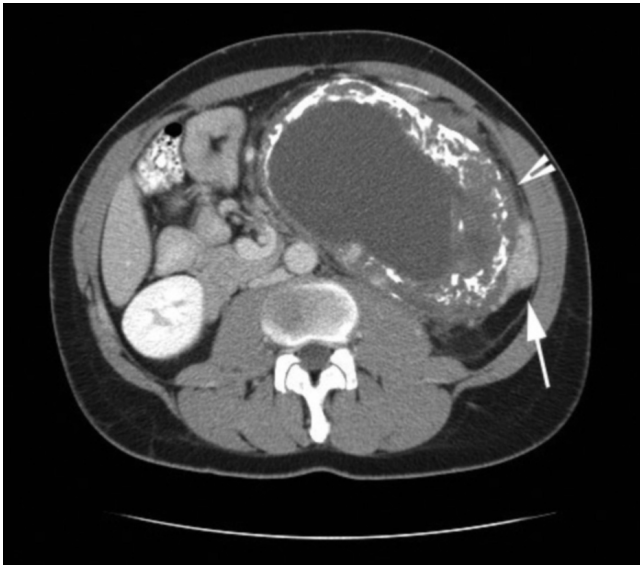


Figure 1C. 46-year-old male with a left retroperitoneal mass. Portovenous-phase, contrast-enhanced axial image demonstrates extensive flocculent calcification and mass effect on the left kidney. Enhancing thick septations (arrowhead) and a satellite nodule (arrows) are present along the periphery of the lesion.

Next, a CT-guided percutaneous core biopsy was obtained using an 18G needle, with samples containing spindle cells without overt evidence for malignancy. However, given the discordant findings by biopsy and the lesion's appearance on imaging, the patient was counseled regarding the high likelihood of malignancy; he subsequently elected to proceed with definitive surgery.



Figure 1D. 46-year-old male with a left retroperitoneal mass. ROI measurement of a central fat-attenuation lobule.

At time of surgery, a large left retroperitoneal mass with multiple smaller satellite lesions was resected en bloc. Given the mass's intimate association with adjacent structures, the left kidney and left colon were sacrificed, along with portions of the left psoas muscle. At the conclusion of the operation, there was no evidence of additional disease within the surgical bed. The patient had an unremarkable postoperative course with quick return of bowel function. He was discharged on POD 8.

On gross pathologic review, the specimen measured 22 cm in greatest dimension; cut sections revealed a large cystic cavity containing hemorrhagic fluid (Fig 2). Evaluation of the specimen margins demonstrated that the tumor abutted but did not invade the left kidney or attached portions of the left colon. Multiple surrounding satellite nodules measured up to 5.5 cm in size. On cut section, these had a solid, white-tan surface with foci of hemorrhage and cystic degeneration.

Microscopic examination of the tumor displayed only small residual foci of well-differentiated liposarcoma, with a predominance of dedifferentiation including foci of osteoid formation (Figs. 3, 4). The walls of the cystic cavity showed extensive osseous metaplasia. Immunohistochemical stains showed the lesional cells to be positive for MDM2 and CDK4.

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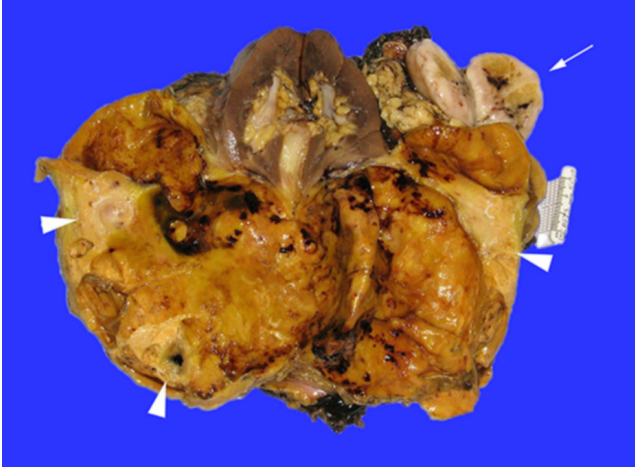


Figure 2. 46-year-old male with a left retroperitoneal mass. Bisected gross specimen, with the kidney along the superior margin of the mass. Soft-tissue nodules adjacent to the kidney (arrow) correspond to enhancing areas on CT. Multiple pockets of dense calcification are present along the periphery (arrowheads).

### Discussion

Liposarcomas are the most common primary retroperitoneal tumors, followed by leiomyosarcomas and malignant fibrous histiocytomas (MFH). Liposarcomas can be further classified into three subgroups: well-differentiated liposarcoma with or without dedifferentiation, myxoid and round cell liposarcoma, and pleomorphic liposarcoma (1). Within

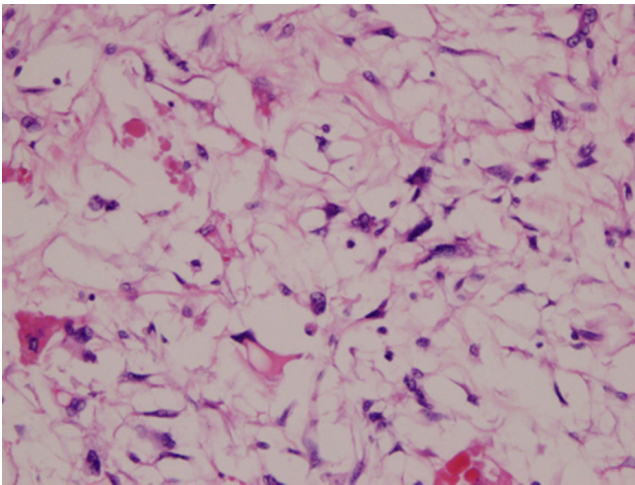


Figure 3. 46-year-old male with a left retroperitoneal mass. Well-differentiated liposarcoma with lipoblasts (hematoxylin-eosin, x40 magnification).

the well-differentiated subgroup, dedifferentiated liposarcoma (DDL) and atypical lipomatous tumor/well-differentiated liposarcoma (ALT-WDL) represent the most common subtypes. Classically, DDL are characterized by the presence of a well-differentiated lipomatous lesion juxtaposed with an area of high-grade dedifferentiation (2).

The dedifferentiated (DD) components have a highly variable histological appearance, with 90% of cases resembling MFH or fibrosarcomas, and a minority of cases containing components resembling rhabdomyosarcoma, myosarcoma, and osteosarcoma (2, 3, 4).

Our case of dedifferentiated liposarcomas with osteosarcomatous dedifferentiation represents an extremely rare entity with, to our knowledge, fewer than ten reported cases in the literature (2, 5-8). In general, DDL appears as a heterogeneous mass adjacent to variable amounts of mature fatty elements. The well-differentiated components are indistinguishable from normal fat by imaging, and thus follow fat characteristics on both CT and MRI. The appearance of the DD components are typically nonspecific, reflecting their pleomorphic histology. On CT, these lesions demonstrate similar to slightly decreased density when compared to skeletal muscle (4). On MRI, the lesions have low-to-intermediate signal on T1W sequences, with

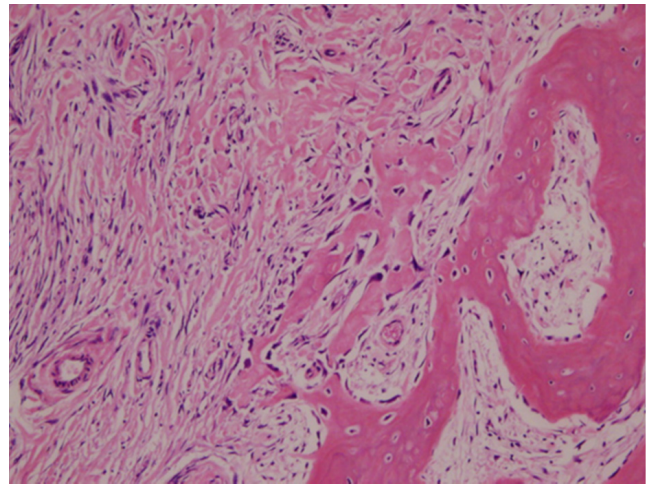


Figure 4. 46-year-old male with a left retroperitoneal mass. Dedifferentiated area with osteoid formation (hematoxylin-eosin, x40 magnification).

intermediate-to-high signal on T2W sequences and variable enhancement (1, 4). In a review of imaging characteristics of 20 retroperitoneal DDLs, Tateishi et al reported that the lesions were typically lobulated in appearance, with a high propensity for invading the pararenal spaces. In 16/20 cases (80%), there were sharp borders between the lipomatous and nonlipomatous components, with the remainder demonstrating a gradual transition. Enhancing septations were present in 90% of the lesions, which correlated to fibrous bands containing collagen fibrils on the pathologic specimens (9). Calcifications are present in up to 32% of liposarcomas, and are best evaluated with CT (10). In the reported cases of DDL with osteosarcomatous dedifferentiation, the lesions typically contain multiple areas of dense mineralization (5, 6, 8).

Given their protean appearance on both histological analysis and imaging, DDLs can be easily confused with

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other primary retroperitoneal mesenchymal tumors. However, recent work by Binh et al. has shown that immunohistochemical stains for MDM2 and CDK4 can be useful to differentiate DLLs from other poorly differentiated sarcomas. In their series, DLLs stained positive for MDM2 and CDK4 antibodies (97% and 92%, respectively, with sensitivity and specificity of 97% and 92% for MDM2 and 83% and 95% for the CDK4 protein) (11).

In our case, the primary diagnostic considerations would include other intrinsic retroperitoneal tumors such as leiomyosarcoma, malignant fibrous histiocytoma (MFH), neurogenic tumors, and primary germ-cell tumors. Although there is great overlap in the radiographic appearance of the solid components, the combination of discrete fat lobules, thick septations, and calcium is highly suggestive of liposarcoma. Teratomas can have a similar appearance but are typically midline lesions, and the patient would likely have elevated human chorionic gonadotropin (HCG) and alpha-fetoprotein (AFP) levels (12). Additionally, teratomas present either in the first 6 months of life or early adulthood, while liposarcomas typically present in older adults (5th-7th decades) (1).

Common benign etiologies (to include retroperitoneal hematoma, seroma, and lymphocele) should also be considered. These lesions are typically homogeneous with fluid density, though a fluid-fluid/hematocrit level may be present in the case of a hematoma. Given the patient's benign clinical presentation and the lack of a trauma or prior surgical history, these diagnoses were considered less favorable. A retroperitoneal abscess can have an aggressive appearance with loss of fat planes and gas; however, the patient did not present with clinical or laboratory signs for infection.

The treatment of choice is wide local excision, but unfortunately complete resection with wide margins is usually not achievable, given the typical large tumor burden at presentation. Henricks et al reported a high recurrence rate (47%), with 34% of the patients eventually succumbing to their disease. Distant metastasis occurred in 17% percent of patients, with sites involving the brain, lung, liver, and bone. The Tateishi series reported a mortality rate of 15%, and local recurrence in 30% of patients. Additional treatment with radiation and chemotherapy remains controversial, and is primarily influenced by histologic grade, size, and location of the tumor (1, 4).

In summary, dedifferentiated liposarcoma with osteosarcomatous dedifferentiation is a rare entity that can have a highly variable imaging appearance. Given the substantial overlap of imaging findings with other soft-tissue sarcomas, biopsy is warranted for a definitive diagnosis. Additional immunohistochemical stains are also frequently useful for further lesional stratification. Furthermore, this case demonstrates the role of imaging in the management of retroperitoneal tumors. Given that the majority of retroperitoneal masses are malignant and that their typical presentation is insidious, a negative biopsy result should be appraised with high clinical suspicion. In our case, the patient was counseled regarding these discordant imaging findings

and was subsequently treated with surgical resection. The patient was referred to an outside facility for adjuvant therapy; however, a CT scan four months post surgery demonstrated no signs of residual disease. His prognosis remains very guarded, considering the thin surgical margins and the historical recurrence and survival rates.

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