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Diagnostic and Therapeutic Challenges of Multifocal Giant Retroperitoneal Dedifferentiated Liposarcoma with Dual Heterologous Differentiation

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Abstract

Dedifferentiated liposarcoma (DDL) with unusual forms of tissue differentiation is rarely seen and varies in presentation. These tumors often arise in the retroperitoneal area, where they tend to grow silently to a large size, appear late, and have a poor prognosis. We describe a rare case of an elderly male who initially presented with what appeared to be an inguinal hernia. Further evaluation revealed a large abdominal mass. CT imaging showed that the tumor occupied most of the left side of the abdomen, with blurred boundaries between it and the left kidney and spleen. During surgery, a large retroperitoneal mass was found pushing the colon and ureter across the midline. The tumor was removed, and the left kidney was preserved. The specimen contained multiple tumor fragments ranging from 15 to 40 cm in size, weighing over 25 kg in total. Histological analysis revealed well-differentiated liposarcoma alongside two distinct types of heterologous differentiation—areas resembling both chondrosarcoma and osteosarcoma. Based on these findings, the final diagnosis was a recurrent, multifocal, giant retroperitoneal DDL with dual heterologous differentiation. Despite the tumor's recurrence and complex features, detailed imaging, complete surgical resection, and thorough pathological evaluation were crucial in confirming the diagnosis.

Keywords: Retroperitoneal sarcoma, Dedifferentiation, Osteosarcoma, Chondrosarcoma, Tumor histology

Introduction

Dedifferentiated liposarcoma (DDL) is a relatively rare but well-established subtype of liposarcoma [1, 2]. In certain cases, it can show divergent dedifferentiation, including heterologous elements such as bone or cartilage formation, though reports of such presentations vary significantly in frequency [2, 3].

We describe a rare case of an elderly male who initially presented with what appeared to be an inguinal hernia.

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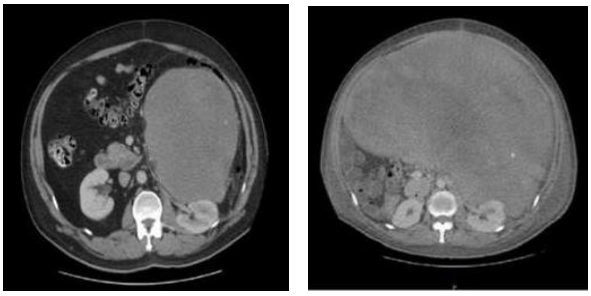
We report an unusual case of recurrent, multifocal, giant DDL with dual heterologous differentiation—specifically featuring both osteosarcomatous and chondrosarcomatous components—in a 60-year-old male patient. The patient, with a known history of hypertension and obesity, presented with progressive abdominal distension over a 4 to 5-month period. He also had a history of left inguinal hernia, previously treated surgically two years prior.

On physical examination, his abdomen was markedly distended, and a large intra-abdominal mass was palpable. A healed surgical scar from the hernia repair was noted in the left inguinal region. Contrast-enhanced computed tomography (CECT) revealed a large retroperitoneal mass on the left side of the abdomen with obliteration of the fat planes between the mass and adjacent organs, namely the left kidney and spleen (**Figure 1**). A non-contrast CT of the chest showed no evidence of pulmonary metastases. Based on clinical and radiological findings, a provisional diagnosis of giant multifocal retroperitoneal liposarcoma was made.

An exploratory laparotomy was performed, and the retroperitoneal mass was surgically excised. Intraoperatively, the tumor was found to occupy the majority of the abdominal cavity, likely originating from the left retroperitoneal region. It displaced the transverse and left colon, mesocolon, and ureter across the midline. The left kidney appeared fully encased within the mass and was not initially visible. The ureter was stretched and dilated, and the mesocolon was tightly adherent to the tumor, suggesting a possible origin near the left psoas muscle.

The surgical team was able to carefully dissect the mass from the left kidney, preserving the renal capsule and the kidney itself (**Figure 1**). Multiple tumor nodules, ranging in size from 4 to 40 cm, were removed and submitted separately for histopathological evaluation.



a) b)

Figure 1. Pre-operative contrast-enhanced computed tomography (a) of the abdomen showed a large retroperitoneal sarcoma abutting the left kidney; fat planes with the left kidney and spleen were lost, and (b) post-operative imaging showing free of disease.

Five large, encapsulated tumor fragments were received for examination. The largest measured 40 × 25 × 10 cm, and the smallest was 15 × 15 × 10 cm, with the total tumor burden exceeding 25 kilograms in weight (**Figure 2a**). Serial sectioning revealed a predominantly yellow, solid, and homogeneous appearance characteristic of adipocytic tumors.

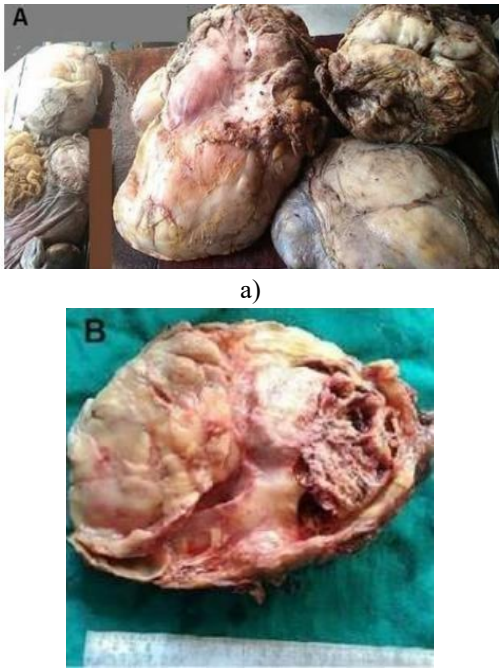
Among the fragments, two mid-sized masses (each up to 18 cm) displayed distinct firm-to-hard regions up to 7 cm in size (**Figure 2b**). Tissue samples were taken from greasy, firm, and calcified (bony) areas for histological evaluation.

Microscopic analysis of the greasy portions showed features consistent with well-differentiated liposarcoma (**Figure 3a**). In contrast, the firmer and bony sections exhibited areas of well-differentiated liposarcoma alongside heterologous elements, including regions of both chondrosarcomatous and osteosarcomatous differentiation. These heterologous components accounted for approximately 10% of the total tumor volume (**Figures 3b–3f**).

The tumor was graded as high-grade (grade 3), with approximately 5% necrotic areas observed. Based on the gross and microscopic findings, a final diagnosis was made: recurrent, multifocal, giant retroperitoneal dedifferentiated liposarcoma with dual heterologous differentiation—osteosarcoma and chondrosarcoma.

Following surgery, the patient recovered well and was discharged with a recommendation for adjuvant chemotherapy. However, he declined further treatment. After a disease-free interval of 17 months, imaging revealed a recurrent mass measuring 14.6 × 16.9 × 20.5 cm in the left anterior perinephric space, adjacent to the renal hilum and vessels (**Figure 4**).

Over the next 9 months (26 months post-surgery), the recurrent mass had grown further to 22 × 35 × 29 cm. Unfortunately, the patient was lost to follow-up thereafter.



a)

b)

Figure 2. Gross examination: (a) five large, encapsulated tumor fragments were received; the largest measured 40 × 25 × 10 cm and the smallest 15 × 15 × 10 cm; all fragments had intact capsules; serial sectioning revealed a predominantly yellow, homogeneous, and solid tumor, and (b) medium-sized fragments (up to 18 cm) exhibited firm areas measuring

6 × 5 × 4 cm and distinct bony regions measuring 7 × 7 × 5 cm.

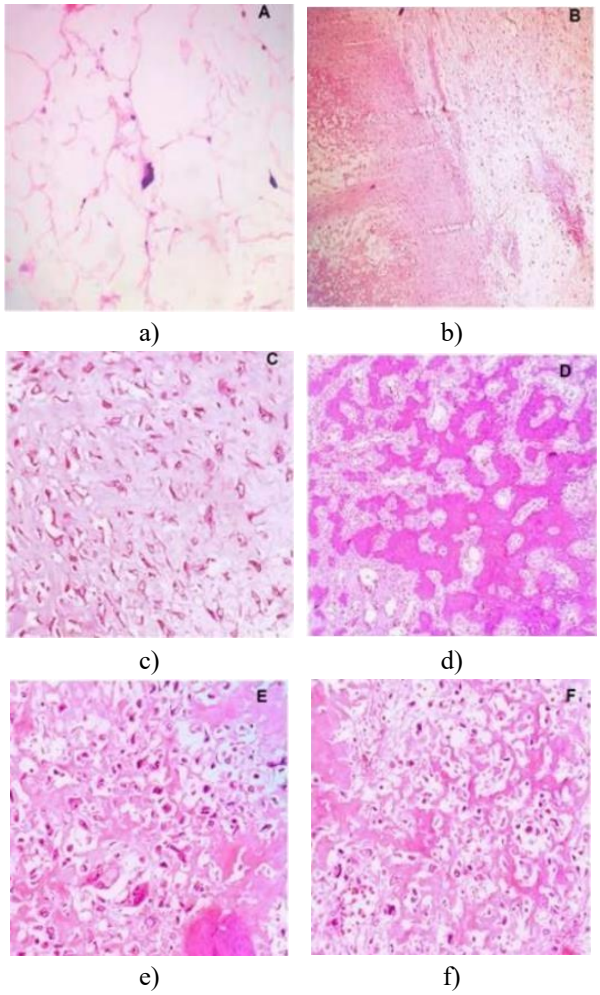


Figure 3. Histopathological features: (a) section from greasy tumor areas showing well-differentiated liposarcoma with lipoblasts within adipocytic tissue (H&E, 400×), (b) abrupt transition observed between hypocellular, well-differentiated regions and hypercellular, dedifferentiated regions (H&E, 100×), (c) glistening areas reveal chondrosarcomatous differentiation characterized by hypercellularity, atypical pleomorphic chondrocytes, irregular nuclear chromatin, and prominent nucleoli within a chondroid matrix (H&E, 400×), (d–f) sections demonstrate varied degrees of osteoid formation; well-differentiated areas with mature bone (d) transition to zones with malignant spindle cells and lacy osteoid matrix, indicating osteosarcomatous differentiation (e, f) (H&E, 400×).

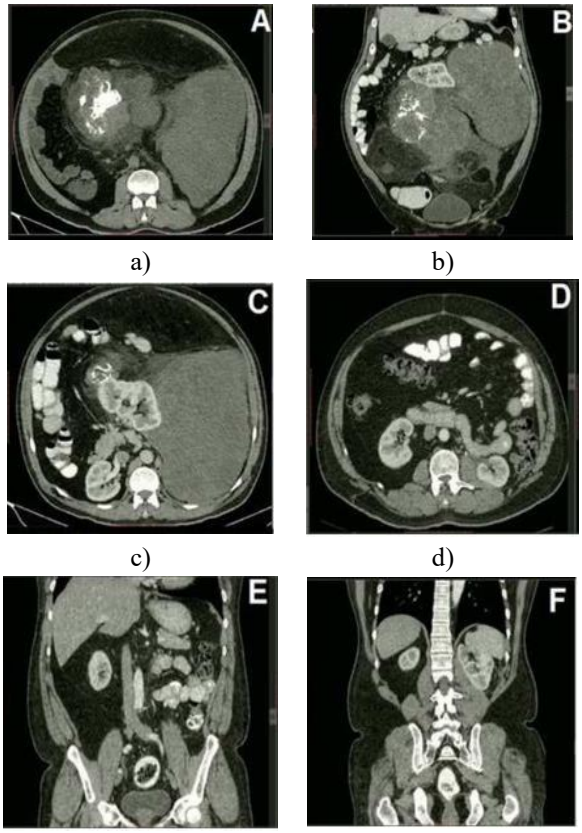


Figure 4. Imaging of tumor recurrence: CT image showing recurrent lobulated soft-tissue mass in the left anterior perinephric space, displacing the pancreatic tail and colon anteriorly, 17 months post-surgery; follow-up imaging after 26 months demonstrating significant progression of the recurrent mass.

Discussion and Conclusions

The term dedifferentiated liposarcoma (DDL) was first introduced by Evans in 1979 and describes a tumor exhibiting a morphological transition from an atypical lipomatous tumor/well-differentiated liposarcoma (ALT/WDL) to a non-lipogenic sarcoma, which may be of either low or high histologic grade [1]. The retroperitoneum is a common site for DDL, where it displays diverse histomorphological patterns. Current understanding suggests that DDL can dedifferentiate into either homologous or heterologous elements, with divergent differentiation being relatively rare. The reported incidence varies widely, ranging from 3.87% to 44% across different studies [2, 3].

Among heterologous differentiation, myogenic elements are the most frequent, while osteochondromatous and

angiosarcomatous forms are less common [2]. Typically, DDL presents as an abdominal mass, often asymptomatic or causing symptoms due to pressure on adjacent structures. Rarely, as observed in the present case, DDL may initially present as an inguinal hernia—an atypical presentation reported only in a few cases [4].

Radiologically, the presence of fat components supports a diagnosis of liposarcoma. Non-lipomatous areas measuring greater than 2 cm raise suspicion for dedifferentiation [4]. The average reported size of DDLs is approximately 17.5 cm [3]; in this case, the tumor was considerably larger, qualifying as a giant liposarcoma. According to the literature, only 19 cases of giant retroperitoneal liposarcomas have been reported in English, of which just eight were dedifferentiated [5–10]. Under-sampling of tumor specimens may lead to a failure in identifying dedifferentiated components, thus underdiagnosing DDL [5]. Regardless of the degree of maturation or the morphological subtype of the osseous component in DDL, bone formation is generally considered neoplastic and arises from osteogenic differentiation within the tumor. The atypia in these areas can range from minimal to marked and may present as benign-appearing heterotopic bone or as high-grade osteosarcoma [5].

The identification of lipoblasts amid mature adipose tissue is critical and must be distinguished from their histologic mimics. In this case, the extensive distribution of greasy areas with features characteristic of well-differentiated liposarcoma was instrumental in establishing the primary diagnosis. Despite the tumor's massive size, meticulous and comprehensive sampling allowed for the identification of heterologous components during gross examination. These components were focal and could only be recognized through careful serial sectioning of each tumor mass.

Uniquely, this case demonstrated dual heterologous differentiation, featuring both osteosarcomatous and chondrosarcomatous elements. It is important to note that DDL is considerably more common than extraskeletal osteosarcoma [5], and dedifferentiation occurs in approximately 10% of well-differentiated liposarcomas [11]. Chondrosarcomas and liposarcomas are also known to undergo dedifferentiation into unusual mesenchymal elements [12].

Although immunohistochemical markers like CDK4 and MDM2 help identify DDL, they are not definitive. Other markers, such as osteopontin, osteonectin, SATB2, IDH mutations, and galectin-1, have been explored in osteosarcoma and chondrosarcoma. However, diagnosis remains reliant on identifying malignant osteoid or cartilage histologically. Therefore, immunohistochemistry plays a supporting role in challenging cases but was not required in our case.

This case is notable due to several rare features: initial presentation as an inguinal hernia, multifocal tumor growth, a heterogeneous gross appearance, extreme size, and the presence of dual divergent heterologous differentiation. Accurate diagnosis in such rare cases depends heavily on extensive sampling and detailed histopathological examination.

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Conflict of Interest: None

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Ethics Statement: None

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