

# Multifocal Giant Retroperitoneal Dedifferentiated Liposarcoma with dual heterologous dedifferentiation– a diagnostic and therapeutic challenge

## Abstract

Dedifferentiated liposarcoma (DDL) with divergent dedifferentiation is uncommonly reported with variable incidences. Liposarcomas, particularly those manifesting dedifferentiation, occur in the retroperitoneum. They grow into large size, present at very late stages, and can have a poor clinical outcome. Here we present a rare case of recurrent dedifferentiated liposarcoma with dual heterologous differentiation with osteosarcoma and Chondrosarcoma in an elderly male who had initially presented with inguinal hernia. A large mass was occupying the entire abdomen, confirmed on CECT occupying the left side with loss of fat planes with the left kidney and spleen. During exploratory laparotomy, a large retroperitoneal mass occupying the whole abdomen from the left side of the retroperitoneum pushing the left and transverse colon with the mesocolon, the ureter to left side across midline was found, which was excised with the kidney preserved. Grossly multiple large fragments of tumor masses were received, ranging in size from 15 to 40 cm in their most significant dimension and weighing more than 25kgs. Extensive sampling from all the tumor masses was done for histopathology, which revealed well-differentiated liposarcoma having juxtaposed heterologous differentiation into chondrosarcomatous and osteosarcomatous areas. A final diagnosis of recurrent multifocal giant retroperitoneal dedifferentiated liposarcoma with dual heterologous differentiation with osteosarcoma and ChondrosarcomaChondrosarcoma was made. Despite recurrent multifocal tumors, the radiological information, extensive excision, detailed grossing, and histopathology helped in the diagnosis.

**Keywords:** Sarcoma, Osteoid, ChondrosarcomaChondrosarcoma, Spindle cells

## Introduction

Dedifferentiated liposarcoma (DDL) is a relatively uncommon yet long-known entity.<sup>[1,2]</sup> With divergent dedifferentiation showing heterologous elements, its incidence has been documented very variably.<sup>[2,3]</sup> Here we present a rare case of recurrent multifocal giant dedifferentiated liposarcoma with dual heterologous differentiation with osteosarcoma and ChondrosarcomaChondrosarcoma in an elderly male. A 60-year male with hypertension and obesity, presented with distension of the abdomen for 4-5 months. There was a history of left inguinal hernia for 2 years, for which surgery was performed. Physical examination revealed a distended abdomen with a large mass occupying the entire core. Left inguinal hernia scar was seen. Contrast Enhanced Computed Tomography(CECT) of the abdomen showed large retroperitoneal sarcoma on the left side. Fat planes with the left kidney and spleen were lost (**Figure 1a-1c**). Non-Contrast CT scan of the thorax did

not show any pulmonary metastasis. A provisional clinicopathologic diagnosis of giant retroperitoneal liposarcoma with the multifocal disease was made.

Exploratory laparotomy and retroperitoneal mass excision were done. Intra-operatively, a large retroperitoneal mass occupying the whole abdomen, probably arising from the left side of the retroperitoneum pushing the left and transverse colon with the mesocolon, the ureter to left side across midline. The left kidney was not seen, completely encased by the mass. The left ureter was dilated and stretched. The mesocolon was adherent to the mass, possibly suggesting the mass was arising from the left retroperitoneum postero-lateral to the left psoas muscle. The large mass was completely dissected off the kidney along with the renal capsule and the left kidney was preserved (**Figure 1d-1f**). Multiple lipomatous lesions ranged in size from 4 cm to 40cm in the most significant dimension,

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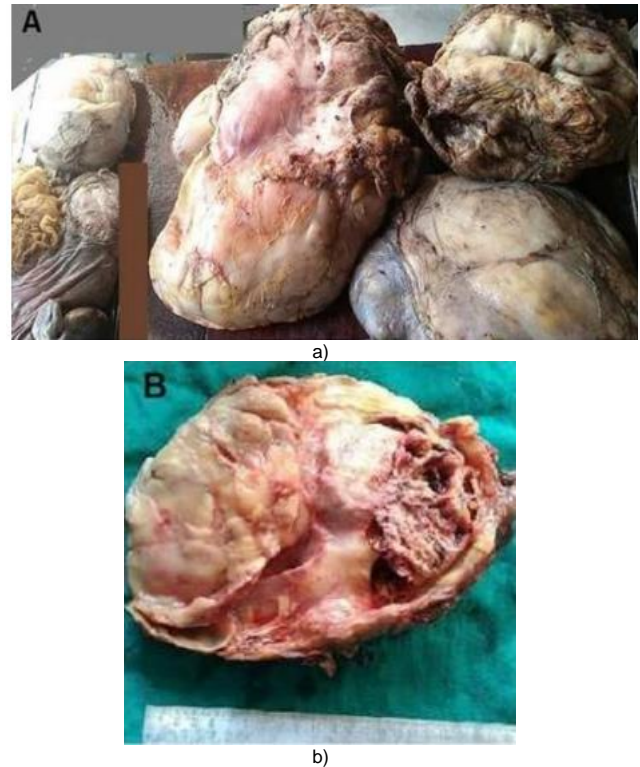
which was excised and sent separately for histopathology.



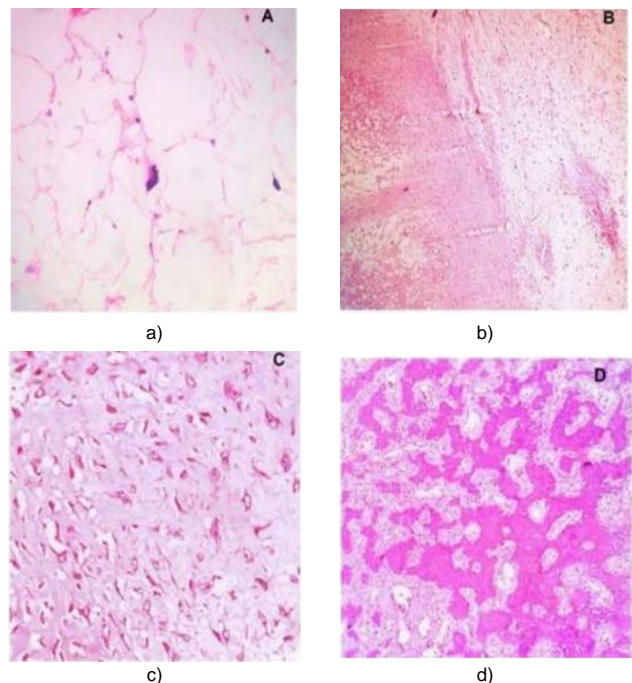
**Figure 1.** Pre-operative Contrast-Enhanced Computed Tomography (A-C) of abdomen showed large retroperitoneal sarcoma abutting left kidney. Fat planes with left kidney and spleen were lost.; (D-F) Post-operative imaging showing free of disease.

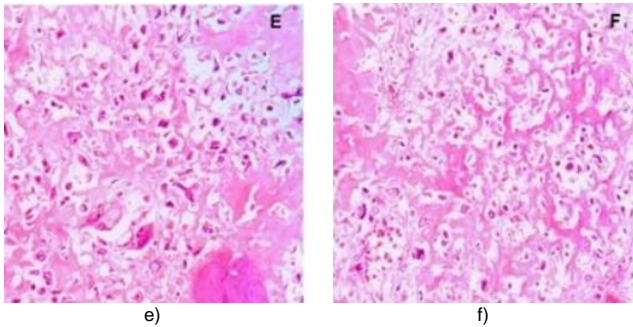
Grossly five large fragments of encapsulated tumor were received largest of which measuring 40x25x10cm and the smallest measuring 15x15x10cm and weighing about >25kgs (**Figure 2a**). Serial slices showed a predominantly yellow, homogenous, and solid tumor. However, two of the medium-sized fragments measuring 18cm in the greatest dimension showed firm to hard areas measuring 7cm in the most significant dimension (**Figure 2b**). Multiple sections from the greasy, firm, and bony areas were studied. Sections from greasy areas showed features of well-differentiated liposarcoma (**Figure 3a**). Sections from the firm and bony areas showed well-differentiated liposarcoma, having juxtaposed heterologous differentiation into chondrosarcomatous and osteosarcomatous areas (~10% of total tumor) (**Figure 3b-3f**). Histologic grade was high (grade3), with areas of necrosis amounting to around ~5%. A final diagnosis of recurrent multifocal giant retroperitoneal dedifferentiated liposarcoma with dual heterologous differentiation with osteosarcoma and ChondrosarcomaChondrosarcoma was made. The patient was discharged after post-operative recovery and advised for adjuvant chemotherapy. He refused the same and developed a recurrent mass after a disease-free period of 17 months, measuring 14.6 x 16.9 x 20.5 cm (**Figure 4**). The recurrent lobulated solid mildly enhancing mass was in the region of the left anterior perinephric space, abutting the renal hilum &

renal vessels. The mass further increased to 22 x 35 x 29 cm after 26months of surgery. However, the patient was lost to follow-up.

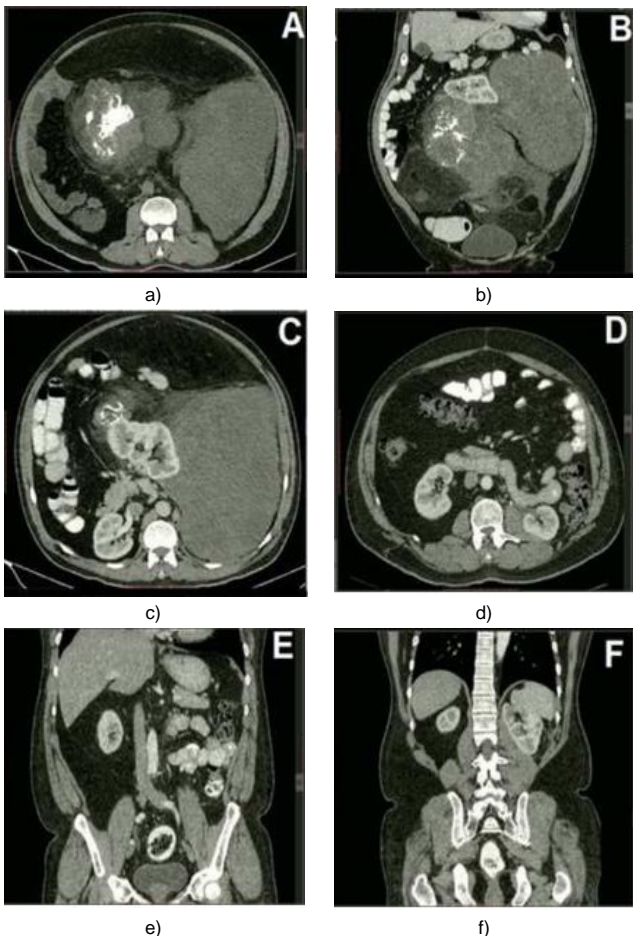


**Figure 2.** A Grossly five large fragments of encapsulated tumor were received largest of which measuring 40x25x10cm and smallest measuring 15x15x10cm. Capsule appears intact in all fragments. Serial cut section shows predominantly yellow, homogenous, solid tumor. B. medium sized fragments measuring 18cm in greatest dimension show firm areas measuring 6x5x4cm and bony areas measuring 7x7x5cm.





**Figure 3.** A: Section from the greasy areas showed well differentiated liposarcoma with lipoblasts amidst fatty areas. (H&E, 400X) B. Section shows an abrupt transition between hypocellular well differentiated regions and dedifferentiated hypercellular regions juxtaposed with each other (H&E, 100X) C. Section from the glistening areas showing chondrosarcomatous differentiation with hypercellularity, atypical pleomorphic chondrocytes, irregular nuclear chromatin and few cells showing prominent nucleoli amidst the chondroid matrix. (H&E, 400X) D-F. Sections show varied degrees of bone formation. Well-differentiated areas with bone formation (D) are intermingled with areas showing plump malignant spindle cells with lacy osteoid formation (E,F). (H&E, 400X)



**Figure 4.** Recurrent mass in the left anterior perinephric space extending anteriorly upto parietal wall displacing pancreatic tail & colon anteriorly after 17months (a) which further increased after 26months (b)

‘Dedifferentiated liposarcoma’ (DDL) was first coined in 1979 by Evans. It has been defined as an entity with the morphological transition from an atypical lipomatous tumor/well-differentiated liposarcoma to a non-lipogenic sarcoma.<sup>[1]</sup> The latter may be low or high grade. At

retroperitoneum, a common site, dedifferentiation shows varied histomorphology. The current understanding says it can show dedifferentiation towards homologous or heterologous components. The occurrence of divergent differentiation is rare, and various studies have shown various prevalence ranging from 3.87-44%.<sup>[2,3]</sup> Among these most frequent is myogenic, and less common are osteochondromatous and angiosarcomatous differentiation.<sup>[2]</sup> It conventionally presents as an abdominal mass that is asymptomatic or with symptoms due to pressure on adjacent organs. As is seen in the present case, rare instances have been documented in literature wherein inguinal hernia is their inaugural presentation.<sup>[4]</sup> The presence of fat components in radiology imaging findings can be helpful in the diagnosis of liposarcoma.<sup>[5]</sup> When the non-lipomatous areas exceed >2cm, it suggests the possibility of dedifferentiation.<sup>[4]</sup> The documented mean size of the DDLs is 17.5cm while here the tumor was a giant liposarcoma.<sup>[3]</sup> To the best of our knowledge, only 19 cases of giant retroperitoneal liposarcomas were documented in English literature until only eight were dedifferentiated.<sup>[6-10]</sup> Inadequately sampled specimens, therefore, might lead to missing these areas and thereby under-diagnosing DDL.<sup>[5]</sup> Irrespective of the maturity and the morphological type of bone tissue formed in DDL, it is usually neoplastic. It is a consequence of osteogenic differentiation in tumor cells. The degree of atypia may be minimal to severe and may show heterotopic bone tissue to high-grade osteosarcoma.<sup>[5]</sup> It is also required to search for the lipoblasts with the mature adipose tissue and differentiate the former from its mimics. In this case, the widespread presence of surrounding, greasy areas with features of well-differentiated liposarcoma made us consider the same as a primary diagnosis. Despite the large size of the tumor, the extensive sampling of the tumor enabled us to recognize the heterologous elements on the gross examination itself. These were focal and required serial slicing through each large mass. Also, this case was unique in its dual differentiation in osteosarcomatous as well as chondrosarcomatous areas. Also, it is very important to note that DDL is far more frequent as compared to extraskeletal osteosarcoma.<sup>[5]</sup> Dedifferentiation has been recorded in about 10% of well-differentiated liposarcomas.<sup>[11]</sup> Chondrosarcomas and liposarcomas frequently showcase dedifferentiation into aberrant elements.<sup>[12]</sup> Though CDK4 and MDM2 positivity have been used to detect DDL, they alone are insufficient. Though numerous immunohistochemical markers have been studied in osteosarcoma and Chondrosarcoma including osteopontin, osteonectin and isocitrate dehydrogenase (IDH), SATB2, and galectin-1, the diagnosis is essentially based on the presence of malignant osteoid and cartilage. Hence, the use of immunohistochemistry is confined to difficult cases, unlike ours.

This case stands unique due to its inaugural presentation as inguinal hernia, multifocality, heterogeneous gross nature, giant expansive size, and rare unique microscopic findings revealing divergent dedifferentiation on histopathology. This rare entity requires extensive sampling and microscopic identification for the right diagnosis.

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### Conflict of interest

None.

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### Ethics statement

A written informed consent was taken from the patients.

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