Society of Medical Education & Research

2024, Volume 4, Issue 2, Page No: 1-5
ISSN: 3108-4834

Archive of International Journal of Cancer and Allied Science

A Diagnostic and Therapeutic Dilemma: Giant Multifocal Retroperitoneal Dedifferentiated Liposarcoma with Dual Heterologous Components

Suyeon Kim[1], Hyunsik Bae[2*], Hyun-Soo Kim[1*]

[1]Department of Pathology and Translational Genomics, Samsung Medical Center, Sungkyunkwan University School of Medicine, Seoul 06351, Republic of Korea.
[2]Pathology Center, Seegene Medical Foundation, Seoul 04805, Republic of Korea.

***E-mail** ✉ hyunsik.bae@samsung.com

Abstract

Dedifferentiated liposarcoma (DDL), exhibiting divergent heterologous differentiation, is a rare entity with a variable reported incidence. These tumors typically arise in the retroperitoneum, often attaining substantial size and presenting at advanced stages, thereby posing significant diagnostic and therapeutic challenges. We report a rare case of recurrent DDL with dual heterologous differentiation—osteosarcomatous and chondrosarcomatous—in an elderly male initially presenting with symptoms mimicking an inguinal hernia. Contrast-enhanced CT revealed a massive abdominal mass occupying the left retroperitoneal space, with obliteration of fat planes adjacent to the left kidney and spleen. Exploratory laparotomy identified a large tumor displacing the left and transverse colon along with the mesocolon and ureter across the midline. The mass was surgically excised while preserving the left kidney. Gross examination revealed multiple tumor fragments, ranging from 15 to 40 cm in size, with a cumulative weight exceeding 25 kg. Extensive histopathological sampling revealed a well-differentiated liposarcoma with areas of heterologous differentiation, exhibiting features of both chondrosarcoma and osteosarcoma. A final diagnosis of recurrent multifocal giant retroperitoneal DDL with dual heterologous differentiation was established. Despite the tumor's multifocality and recurrence, comprehensive radiological assessment, meticulous surgical excision, and detailed pathological evaluation were critical in achieving an accurate diagnosis.

Keywords: Dedifferentiated liposarcoma, Retroperitoneal sarcoma, Osteosarcoma, Chondrosarcoma, Heterologous differentiation, Spindle cells

Introduction

Dedifferentiated liposarcoma (DDL) is a relatively rare yet well-recognized malignancy [1, 2]. When it demonstrates divergent dedifferentiation with heterologous elements, its incidence becomes even more variable and infrequently reported in the literature [2, 3]. We present a rare case of recurrent, multifocal, giant retroperitoneal DDL in an elderly male, exhibiting dual heterologous differentiation—osteosarcomatous and chondrosarcomatous components.

Access this article online https://smerpub.com/

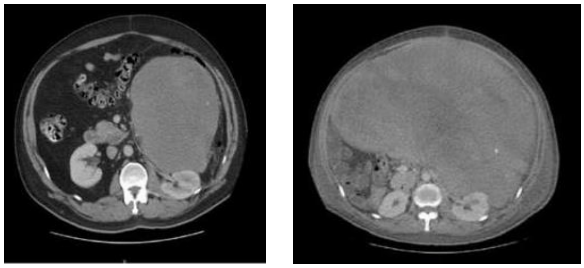
Received: 27 April 2024; **Accepted:** 09 August 2024

How to cite this article: Kim S, Bae H, Kim HS. A Diagnostic and Therapeutic Dilemma: Giant Multifocal Retroperitoneal Dedifferentiated Liposarcoma with Dual Heterologous Components. Arch Int J Cancer Allied Sci. 2024;4(2):1-5. https://doi.org/10.51847/5JnC3jAkZz

Case reports

A 60-year-old male with a medical history of hypertension and obesity presented with progressive abdominal distension over 4–5 months. He had previously undergone surgery for a left inguinal hernia diagnosed two years earlier. On physical examination, a markedly distended abdomen was noted with a large palpable mass occupying the central abdomen. A surgical scar from the prior inguinal hernia repair was evident. Contrast-enhanced computed tomography (CECT) of the abdomen revealed a large retroperitoneal mass on the left side with obliteration of fat planes between the tumor, left kidney, and spleen **(Figure 1)**. A non-contrast CT scan of the thorax showed no evidence of pulmonary metastasis. Based on clinical and radiological findings, a provisional diagnosis of a giant retroperitoneal liposarcoma with multifocal involvement was made.

The patient underwent exploratory laparotomy and excision of the retroperitoneal mass. Intraoperatively, a massive tumor was found occupying the entire abdomen, likely originating from the left retroperitoneum. It displaced the left and transverse colon along with the mesocolon and shifted the ureter across the midline. The left kidney was not initially visible, as the mass entirely enveloped it. The left ureter appeared stretched and dilated. The mesocolon was adherent to the tumor, suggesting its origin was in the posterolateral retroperitoneum near the left psoas muscle. The mass was meticulously dissected from the kidney while preserving the renal capsule, and the left kidney was spared **(Figure 1)**. Multiple lipomatous tumor nodules, ranging from 4 to 40 cm in the most significant dimension, were excised and submitted individually for histopathological evaluation.



a) b)

Figure 1. Contrast-enhanced CT scans of the abdomen (a) revealed a massive retroperitoneal tumor on the left side, with indistinct fat planes separating it from the left kidney and spleen; follow-up imaging after surgical resection (b) confirmed the absence of residual disease.

Gross examination yielded five sizeable, well-encapsulated tumor fragments. The largest specimen measured 40 × 25 × 10 cm, and the smallest 15 × 15 × 10 cm, with a cumulative weight exceeding 25 kilograms **(Figure 2a)**. On cut section, most of the tumor displayed a yellow, greasy, lobulated appearance. Two mid-sized fragments (~18 cm) showed discrete firm-to-hard areas measuring up to 7 cm in diameter **(Figure 2b)**. Representative tissue samples were obtained from lipomatous, firm, and calcified regions for microscopic analysis.

Histopathological examination of the yellow, fatty areas confirmed well-differentiated liposarcoma **(Figure 3a)**. Other sampled regions exhibited dedifferentiated areas with distinct heterologous differentiation into chondrosarcoma and osteosarcoma, accounting for approximately 10% of the total tumor volume **(Figures 3b–3f)**. The tumor was graded as FNCLCC Grade 3, with tumor necrosis estimated at 5%.

The final diagnosis was a recurrent, multifocal, retroperitoneal dedifferentiated liposarcoma exhibiting dual heterologous elements—chondrosarcomatous and osteosarcomatous. The patient recovered postoperatively and was discharged with a recommendation for adjuvant chemotherapy, which he declined. At 17 months post-resection, surveillance imaging revealed a recurrent mass in the left anterior perinephric region, measuring 14.6 × 16.9 × 20.5 cm, encroaching upon the renal hilum and vessels **(Figure 4)**. By 26 months, the lesion had progressed to a size of 22 × 35 × 29 cm. The patient was subsequently lost to follow-up.

a) b)

Figure 2. (a) Gross examination revealed five large, encapsulated tumor fragments, the largest measuring 40 × 25 × 10 cm and the smallest 15 × 15 × 10 cm. The capsule appeared intact in all fragments. Serial sectioning showed a predominantly yellow, homogeneous, and solid appearance. (b) Medium-sized pieces, approximately 18 cm in the most significant dimension, exhibited firm regions measuring 6 × 5 × 4 cm and bony regions measuring 7 × 7 × 5 cm.

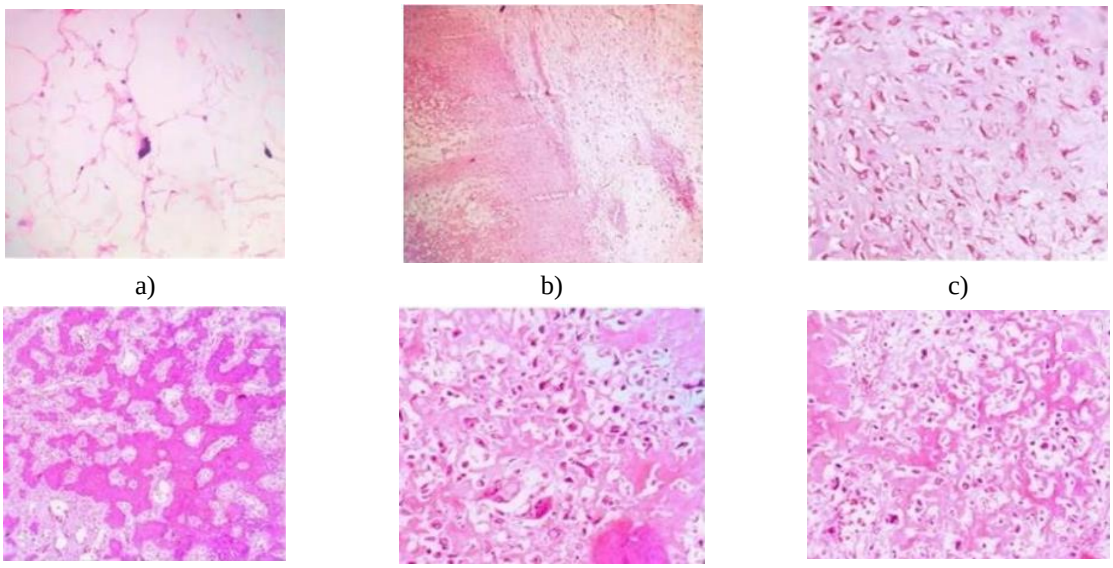


Figure 3. (a) Microscopic section from a greasy region showing well-differentiated liposarcoma characterized by lipoblasts within a fatty matrix (H&E, 400×). (b) An abrupt transition between hypocellular, well-differentiated areas and hypercellular, dedifferentiated regions is noted (H&E, 100×). (c) Section from a glistening area reveals chondrosarcomatous differentiation with increased cellularity, pleomorphic chondrocytes, irregular nuclear chromatin, and occasional prominent nucleoli within a chondroid matrix (H&E, 400×). (d–f) Sections demonstrate varying degrees of osteogenic differentiation. Well-differentiated tumor areas with bone formation (d) are interspersed with regions of malignant spindle cells producing lacy osteoid (e, f) (H&E, 400×).

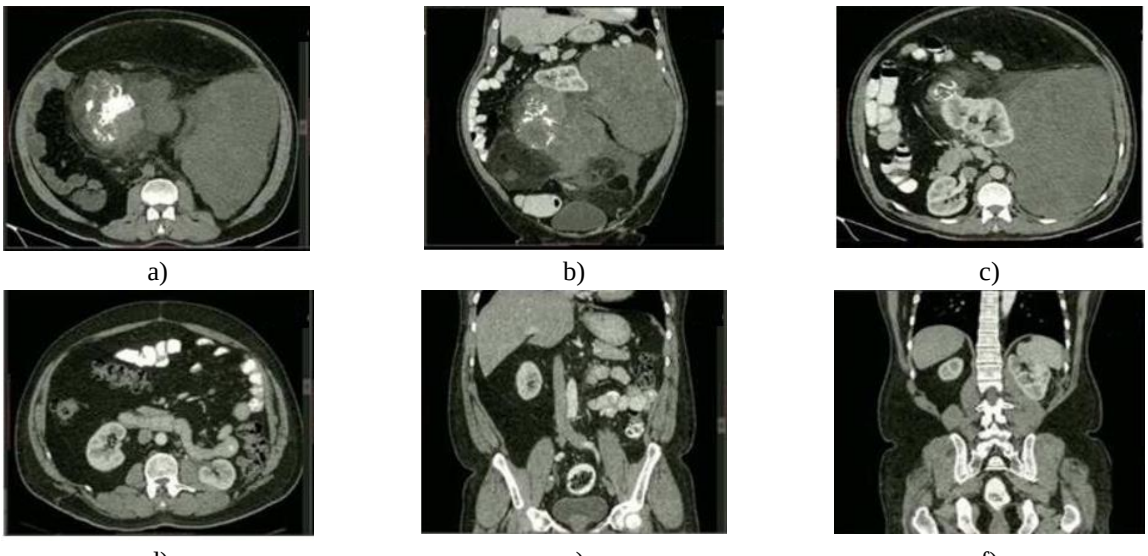


Figure 4. CT scan at 17 months post-surgery shows a recurrent mass in the left anterior perinephric space, extending to the anterior abdominal wall and displacing the pancreatic tail and colon. At 26 months postoperatively, the recurrent mass had significantly increased in size.

Results and Discussion

The term *dedifferentiated liposarcoma* (DDL) was first introduced by Evans in 1979. It refers to a tumor that exhibits a morphological transition from an atypical lipomatous tumor or well-differentiated liposarcoma (WDL) to a non-lipogenic sarcoma, which can be either low- or high-grade [1]. The retroperitoneum is the most

frequent anatomical site of occurrence, and tumors in this region can display a broad spectrum of histological differentiation. Current understanding recognizes the potential for dedifferentiation to occur toward either homologous or heterologous lineages. However, divergent heterologous differentiation is rare, with reported prevalence ranging from 3.87% to 44% across studies [2, 3]. Among these, myogenic differentiation is most commonly observed, while osteochondromatous and angiosarcomatous elements are less frequently reported [2].

Typically, DDL presents as an abdominal mass that is either asymptomatic or causes symptoms due to compression of adjacent structures. Rarely, as seen in the present case, it may initially present as an inguinal hernia—an atypical clinical manifestation that has been infrequently documented in the literature [4]. Radiologically, the identification of fat-containing components is a helpful diagnostic clue for liposarcoma [5]. The presence of non-lipomatous areas larger than 2 cm on imaging may suggest dedifferentiation [4]. While the average reported size of DDLs is approximately 17.5 cm [3], the tumor in this case qualifies as a *giant* liposarcoma. To date, only 19 cases of giant retroperitoneal liposarcoma have been reported in the English literature, with only eight exhibiting dedifferentiation [6–10].

Failure to adequately sample tumor specimens can result in the missed detection of foci of dedifferentiation and underdiagnosis of DDL [5]. Regardless of the degree of maturity or type of osseous tissue identified, the bone formed in DDL is considered neoplastic, arising from osteogenic differentiation of tumor cells. The level of cytologic atypia can vary from minimal to pronounced, ranging from heterotopic bone formation to high-grade osteosarcomatous transformation [5]. It is also critical to identify true lipoblasts within mature adipose areas and distinguish them from histologic mimics. In our case, the presence of widespread greasy regions with features of well-differentiated liposarcoma supported the initial diagnosis. However, meticulous and extensive sampling of the large tumor masses allowed for the detection of focal heterologous elements during gross examination. These elements were identified only through serial sectioning across all specimens.

This case is particularly notable for its dual heterologous differentiation into both osteosarcomatous and chondrosarcomatous components—an exceedingly rare finding. It is important to emphasize that DDL is far more common than primary extraskeletal osteosarcoma [5]. Dedifferentiation has been observed in approximately 10% of WDLs [11]. Both chondrosarcoma and liposarcoma are known to dedifferentiate into aberrant mesenchymal components occasionally [12].

Although immunohistochemical markers such as CDK4 and MDM2 are often used to support the diagnosis of DDL, they are not definitive when used in isolation. Various markers, including osteopontin, osteonectin, IDH mutations, SATB2, and galectin-1, have been investigated in osteosarcoma and chondrosarcoma; however, the diagnosis ultimately hinges on the histological identification of malignant osteoid and cartilaginous matrix. Therefore, immunohistochemistry is reserved for diagnostically challenging cases and was not essential in our diagnosis.

This case is unique in several respects: its unusual initial presentation as an inguinal hernia, multifocality, gross morphological heterogeneity, massive size, and the rare microscopic finding of dual divergent dedifferentiation. It highlights the crucial importance of thorough grossing and comprehensive histopathological evaluation in diagnosing rare variants of liposarcoma.

Acknowledgments: None

Conflict of Interest: None

Financial Support: None

Ethics Statement: None

References

1. Evans HL. Liposarcoma: a study of 55 cases with a reassessment of its classification. Am J Surg Pathol. 1979;3(6):507-23.
2. Henricks WH, Chu YC, Goldblum JR, Weiss SW. Dedifferentiated liposarcoma: a clinicopathological analysis of 155 cases with a proposal for an expanded definition of dedifferentiation. Am J Surg Pathol. 1997;3(3):271-81.
3. Rekhi B, Navale P, Jambhekar NA. Critical histopathological analysis of 25 dedifferentiated liposarcomas, including uncommon variants, reviewed at a tertiary cancer referral center. Indian J Pathol Microbiol. 2012;55(3):294-302.
4. Fiaschetti V, Castellani F, Croce G, Mondillo M, Amico A, De Majo A, et al. Incidentally discovered

well-differentiated retroperitoneal liposarcoma with inguinal canal herniation: report of 2 cases. Radiol Case Rep. 2017;12(2):308-12.
5. Gordhandas J, Lin G, Tipps AMP, Zare SY. Osteosarcomatous divergence in dedifferentiated liposarcoma presenting as a colonic mass. Case Rep Pathol. 2019;2019:8025103.
6. Zeng X, Liu W, Wu X, Gao J, Zhang P, Shuai X, et al. Clinicopathological characteristics and experience in the treatment of giant retroperitoneal liposarcoma: a case report and review of the literature. Cancer Biol Ther. 2017;18(9):660-5.
7. Herzberg J, Niehaus K, Holl-Ulrich K, Honarpisheh H, Guraya SY, Strate T. Giant retroperitoneal liposarcoma: a case report and literature review. J Taibah Univ Med Sci. 2019;14(5):466-71.
8. Carboni F, Valle M, Federici O, Covello R, Garofalo A. Giant primary retroperitoneal dedifferentiated liposarcoma. J Gastrointest Surg Off J Soc Surg Aliment Tract. 2019;23(7):1521-3.
9. Moyon FX, Moyon MA, Tufiño JF, Yu A, Mafla OL, Molina GA. Massive retroperitoneal dedifferentiated liposarcoma in a young patient. J Surg Case Rep. 2018;2018(10):rjy272.
10. Agrusa A, Di Buono G, Buscemi S, Randisi B, Gulotta L, Sorce V, et al. Dedifferentiated retroperitoneal large liposarcoma and laparoscopic treatment: Is it possible and safe? The first literature case report. Int J Surg Case Rep. 2019;57:113-7.
11. Dei Tos AP. Liposarcomas: diagnostic pitfalls and new insights. Histopathology. 2014;64(1):38-52.
12. Dey B, Chikhale M, Pradhan P, Barwad A, Badhe BA. Proximal epithelioid sarcomatous dedifferentiation in secondary chondrosarcoma in a known case of multiple osteochondromatosis. J Lab Physicians. 2017;9(03):214-6.