

CASE REPORT

Open Access



# Dedifferentiated liposarcoma with extensive cystic change causing significant diagnostic challenges: report of two cases and review of the literature

Viola Katharina Vetter<sup>1</sup>, Perparim Limani<sup>2,7</sup>, Falko Ensle<sup>3</sup>, Michelle Leanne Brown<sup>4,7</sup>, Lorenz Bankel<sup>5,7</sup>, Marco Matteo Bühler<sup>1</sup> and Chantal Pauli<sup>1,6,7\*</sup>

## Abstract

**Background** Retroperitoneal dedifferentiated liposarcoma is a rare, aggressive malignancy, characterized by high rates of recurrences and the potential for metastasis. On imaging, these tumors typically present as a solid mass with lipomatous and non-lipomatous components. Cystic changes of dedifferentiated liposarcomas is exceedingly rare and might pose significant diagnostic challenges, with only a few cases reported in the literature.

**Report of 2 cases** We here present two cases of retroperitoneal dedifferentiated liposarcoma with a rare cystic presentation in two female patients aged 51 and 62 years. Imaging revealed large perinephric cystic masses measuring up to 13.0 cm and 16.1 cm, respectively, with calcifications of the cyst wall observed in the second case. Differential diagnoses included cystic echinococcosis, mesenchymal neoplasms, and benign cystic lesions (e.g. endometrial cyst). Both patients underwent upfront compartmental en-bloc surgical resection of the tumor mass and the kidney after multidisciplinary tumor board (MDT) discussion. Macroscopically, the tumors were adherent to but sharply demarcated from the kidney. Histological examination of the first case revealed a small component of well-differentiated liposarcoma (WDLPS) adjacent to a large non-lipogenic sarcoma with a prominent whirling pattern, compatible with dedifferentiation. The second case demonstrated a spindle cell neoplasm with prominent osteosarcomatous heterologous differentiation. *MDM2* amplification was confirmed in both cases by molecular testing. No long-term follow-up data is available for either patient.

**Conclusion** In conclusion, these cases highlight the importance of recognizing unusual and extensive cystic changes of dedifferentiated liposarcoma, which can complicate the diagnostic work-up.

**Keywords** Liposarcoma, Cystic change, Retroperitoneal sarcoma, Dedifferentiated liposarcoma

\*Correspondence:

Chantal Pauli

chantal.pauli@usz.ch

Full list of author information is available at the end of the article



© The Author(s) 2025. **Open Access** This article is licensed under a Creative Commons Attribution-NonCommercial-NoDerivatives 4.0 International License, which permits any non-commercial use, sharing, distribution and reproduction in any medium or format, as long as you give appropriate credit to the original author(s) and the source, provide a link to the Creative Commons licence, and indicate if you modified the licensed material. You do not have permission under this licence to share adapted material derived from this article or parts of it. The images or other third party material in this article are included in the article's Creative Commons licence, unless indicated otherwise in a credit line to the material. If material is not included in the article's Creative Commons licence and your intended use is not permitted by statutory regulation or exceeds the permitted use, you will need to obtain permission directly from the copyright holder. To view a copy of this licence, visit <http://creativecommons.org/licenses/by-nc-nd/4.0/>.

## Background

Retroperitoneal liposarcoma is a rare and aggressive malignancy that accounts for over 45% of primary retroperitoneal sarcomas [1]. The World Health Organization classifies liposarcomas into five main histological entities: well-differentiated, dedifferentiated, myxoid, pleomorphic and myxoid pleomorphic liposarcoma. Within the retroperitoneum well-differentiated (WDLPS) and dedifferentiated liposarcoma (DDLPS) are the most relevant entities, while myxoid liposarcoma and pleomorphic liposarcoma are more common in the extremities [2, 3] and myxoid pleomorphic liposarcoma has a predilection for the mediastinum [4]. DDLPS is associated with a dismal oncological outcome due to its aggressive behavior, high rates of recurrences, and potential for metastasis [5]. DDLPS is defined as a WDLPS with progression and is characterized by the transition to a usually non-lipogenic high-grade sarcoma component. Both WDLPS and DDLPS are molecularly characterized by amplifications of *MDM2* and *CDK4* (12q14-q15) which are located on supernumerary ring and giant marker chromosomes [6]. On imaging DDLPS typically presents as a solid mass, often with evidence of both lipomatous and non-lipomatous components [7]. The occurrence of a cystic configuration in DDLPS is exceedingly rare, with only few reported cases in the literature. It might pose significant diagnostic challenges due to its atypical radiologic appearance and can significantly complicate the diagnostic process, potentially delaying appropriate treatment. Cystic lesions in the retroperitoneum raise a broad differential diagnosis ranging from benign cystic lesions, pseudocysts and cystic degeneration within a solid neoplasm [8].

Here, we describe two patient cases with a retroperitoneal dedifferentiated liposarcoma, both of which showed a rare extensive cystic change. These cases highlight the importance of recognizing cystic changes in DDLPS and contribute to expand the understanding of the diverse presentations of this aggressive and challenging malignancy.

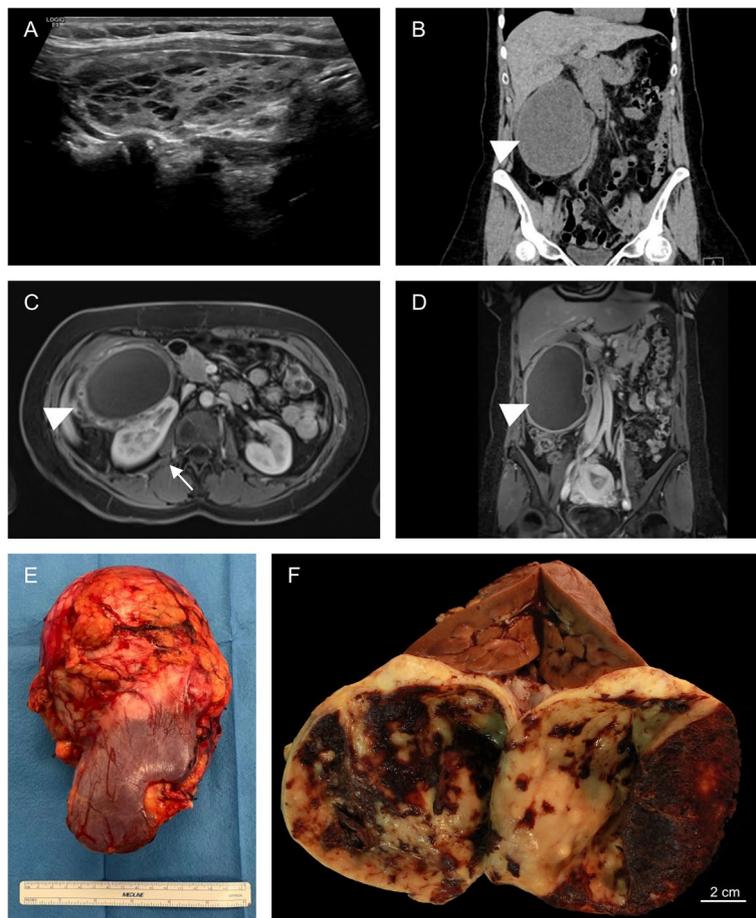
## Report of two cases

### Case 1

The first case was a 51-year-old female patient who presented at the emergency department with a two-week history of an increasing swelling of the abdomen and right upper quadrant abdominal pain. Apart from a benign thyroid condition and a history of endometriosis, the patient had no pre-existing medical history. On examination, there was tenderness on palpation of the right hemi-abdomen. Sonographically, a well-defined lesion measuring 13 × 11 × 8 cm with a hyperechoic internal structure was observed in the right mid-abdomen

(Fig. 1 A). MRI (magnetic resonance imaging) revealed a large, complex cystic mass located below the liver, anterolateral to the right kidney and right psoas muscle. The cystic content appeared inhomogeneous and slightly hyperintense on fat-saturated T1-weighted images, suggesting a protein-rich or hemorrhagic content. The wall of the cyst was irregular, up to 1.5 cm thick, with diffusion restriction and moderate contrast enhancement and contained smaller cysts within the cyst wall. A CT (computed tomography) scan confirmed an irregular cystic lesion without calcification (Fig. 1 B-D). The radiological differential diagnoses included a retroperitoneal hydatid cyst / cystic echinococcosis and a mesenchymal neoplasia. In addition, due to the patient history of endometriosis and the potential hemorrhagic content on imaging, an endometriotic cyst was discussed. Serologic tests for *Echinococcus* (*E. granulosus* and *E. multilocularis*) by ELISA (enzyme-linked immunosorbent assay) and Western Blot were negative. Still, due to the significant risk of cell dissemination in the event of a potential Echinococcosis diagnosis, a biopsy was not performed. Instead, at the tumor board a primary resection of the mass and the adherent right kidney was decided. Macroscopically, the resection specimen consisted of a nephrectomy with an adherent, smoothly delineated and encapsulated cystic mass of up to 11 cm in diameter (Fig. 1 E). The surface appeared smooth and glistening with sparsely adherent fatty tissue. The cyst was filled with hemorrhagic fluid, the inner wall appeared irregularly nodular with reddish-brown deposits (Fig. 1 F). The cyst wall was up to 1.5 cm thick and showed a tan-yellow cut surface.

Histologic examination revealed a DDLPS, consisting of a large, dedifferentiated sarcoma component with rare foci of necrosis and an adjacent small component of WDLPS (Fig. 2 A-H). The wall of the cystic mass showed a prominent nodular growth pattern with a striking variation in cellularity and tumor cell morphology. In addition to the large pseudo-cystic space, many of the cellular nodules of the dedifferentiated liposarcoma exhibited central degenerative change and small pseudocyst formation (Fig. 2 E). Cytological atypia ranged from relatively bland spindle cells to highly pleomorphic tumor cells. In some areas a prominent storiform to whorled growth pattern was observed (Fig. 2 G). Brisk mitotic activity was seen (9 mitoses/2 mm<sup>2</sup>). The inner aspect of the pseudo cystic space showed prominent stromal sclerosis, adherent hemorrhagic material and hemosiderin deposits (Fig. 2 F). No epithelial cyst lining, endometriosis components or parasites could be identified. By immunohistochemistry the tumor cells were strongly positive for MDM2, CDK4 (Fig. 2 I–J) as well as CD10 and partially positive for GLUT1 and Claudin1. Negative markers included SMA, desmin, CD34, STAT6, DOG1 and



**Fig. 1** Radiologic findings and macroscopic images of case 1: **A** Ultrasound image depicting a well-defined lesion with a hyperechoic internal structure. **B** Coronal CT image showing a cystic lesion (arrowhead) below the liver. **C** Axial, and **D** coronal T1-weighted contrast-enhanced MRI images of the lesion (arrowhead) with close proximity to the kidney (arrow). **E** Photograph of the fresh nephrectomy specimen with the adherent mass **F** Photograph of the opened specimen after formalin fixation showing a large cystic space with red deposits and an irregular wall

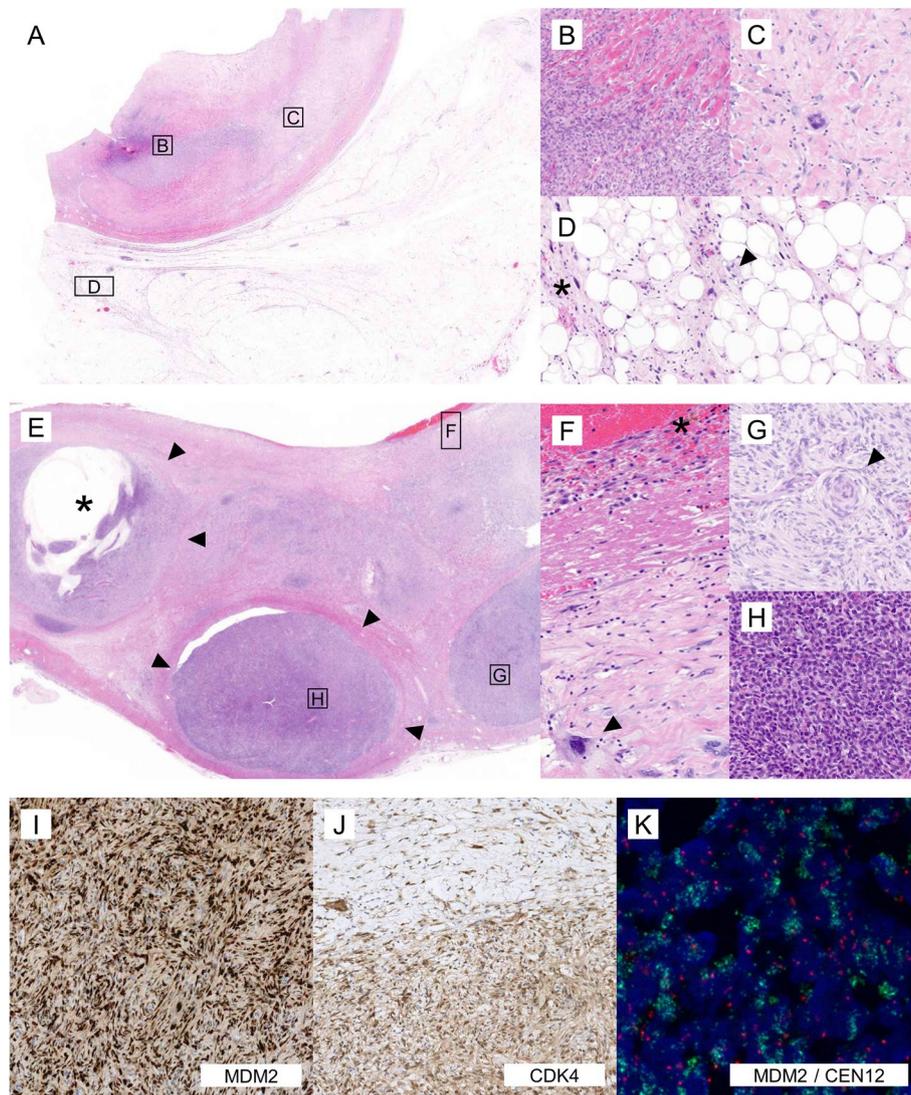
ALK. Ki67 staining showed an irregular proliferation rate of up to 25%. Fluorescence in-situ hybridization (FISH) analysis of the *MDM2* gene locus 12q15 (ZytoLight SPEC MDM2/CEN12 dual color probes, Zytovision GmbH, Bremerhaven, Germany) showed an amplification of the *MDM2* gene supporting the diagnosis of a dedifferentiated liposarcoma (Fig. 2 K).

By histology, the dedifferentiated liposarcoma component was entirely resected, while the completeness of the WDLPS resection could not be reliably assessed. In the postoperative tumor board regular follow-up using imaging (MRI) was recommended. Neoadjuvant radiation or chemotherapy was not pursued. Five months after resection, there was no radiologic evidence of recurrence.

## Case 2

The second case was a 62-years-old female who presented with a subjective swelling of the abdomen and a palpable mass below the right costal arch. Sonographically, a

cystic lesion located between the liver and the right kidney could be identified. It showed a hypoechoic center with an inhomogeneous hyperechoic rim and curvilinear calcifications. A CT scan (Fig. 3 A) revealed a cystic lesion of up to 12 cm in diameter in close proximity to the right kidney with coarse calcifications of the cyst wall. Initial radiological differential diagnoses included a hydatid cyst/ cystic echinococcosis, a peritoneal duplication cyst, a hamartoma or a hematoma. The patient reported contact with several dogs and other pets as well as the existence of foxes in close proximity. Hence, in spite of a negative serologic testing for *Echinococcus*, a clinical concern for an echinococcosis was raised by the primary care team. After specialist consultation, MRI (Fig. 3 B and C) was performed which confirmed a progressively enlarging retroperitoneal well-defined mass with compression and displacement of the right kidney anteriorly (13.3×12.9×16.1 cm). Centrally, the mass was hyperintense on T2-weighted and T1-weighted images

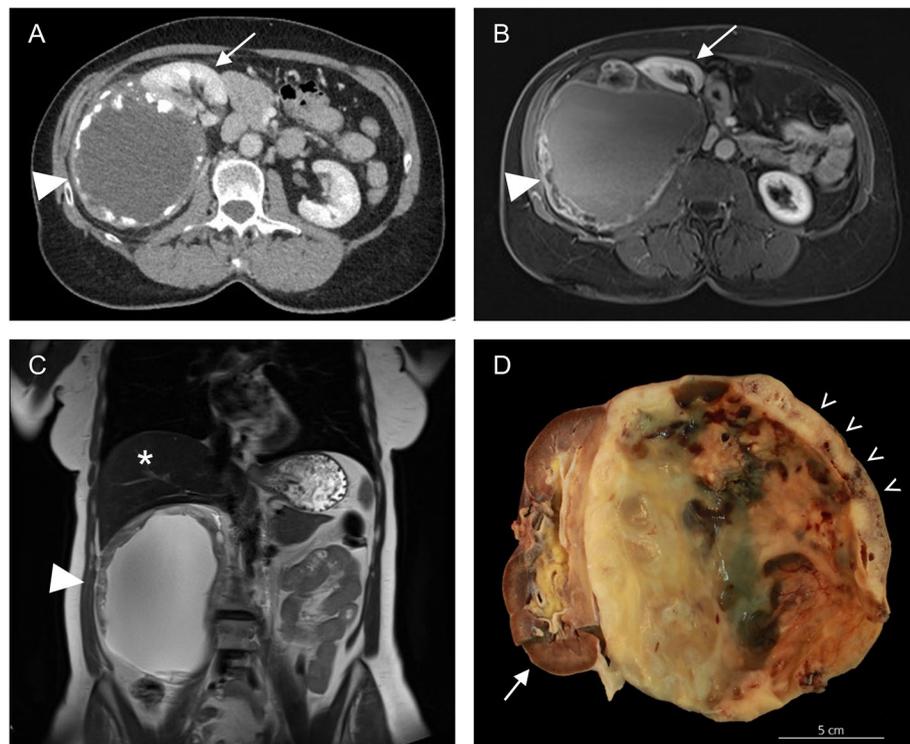


**Fig. 2** Histology (A-H) and ancillary studies (I-K) of case 1: **A** Scanning magnification of the tumor showing an abrupt transition between the well-differentiated liposarcoma component (bottom right) and the dedifferentiated liposarcoma component (top left). **B, C** Higher magnification of the dedifferentiated component with areas of higher cellularity (B) and lower cellularity with stromal hyalinization and interspersed pleomorphic tumor cells (C). **D** Higher magnification of the well-differentiated liposarcoma component showing adipocytes of varying size, atypical hyperchromatic stromal cells (asterisk) and occasional lipoblasts (arrowhead). **E** Scanning magnification of the wall of the cystic space showing a prominent nodularity (arrowheads) with formation of smaller pseudocysts (asterisk). **F** Higher magnification of the inner aspect of the pseudocystic space showing adherent blood and hemosiderin (asterisk), fibrin, stromal sclerosis and pleomorphic tumor cells (arrowhead). **G, H** Higher magnification of the tumor nodules showing relatively bland spindled cells with prominent whirling (G) and highly cellular areas with oval to round tumor cells (H). **I, J** Immunohistochemical stainings showing strong nuclear positivity for MDM2 (I) and CDK4 (J). **K** MDM2 / CEN12 FISH showing amplified clusters of green *MDM2* signals, red signals: centromere 12

with sedimentation. In addition, it showed coarse, wall-adherent calcifications. Moreover, diffuse and nodular areas that enhanced with contrast and showed diffusion restriction could be identified. By imaging, the etiology remained unclear, but the suspicion of a mesenchymal neoplasm (e.g. a dedifferentiated liposarcoma or a neuronal neoplasm) was raised. Because of the contrast enhancement an echinococcal cyst seemed less likely. A

biopsy was not performed due to the cystic nature of the lesion. Instead, a direct primary resection was favored. Intraoperatively the tumor was adherent to the kidney, so that an en bloc resection including a right-sided nephrectomy was performed.

Macroscopically, the resection specimen weighed 1790g and revealed a large cystic mass measuring up to 18 cm in diameter, adherent to the kidney (Fig. 3 D). The



**Fig. 3** Radiologic findings and macroscopic images of case 2: **A** Axial contrast-enhanced CT and **B** T1-weighted MRI images showing a cystic lesion (arrow head) with calcifications of the cyst wall adjacent to the right kidney (arrow). **C** Coronal T2-weighted MRI image depicting the lesion (arrowhead) below the liver (asterisk). **D** Photograph of the bisected nephrectomy specimen (arrow) with the adherent cystically configured tumor exhibiting an irregularly thickened, partially bone-hard wall structure (arrow heads)

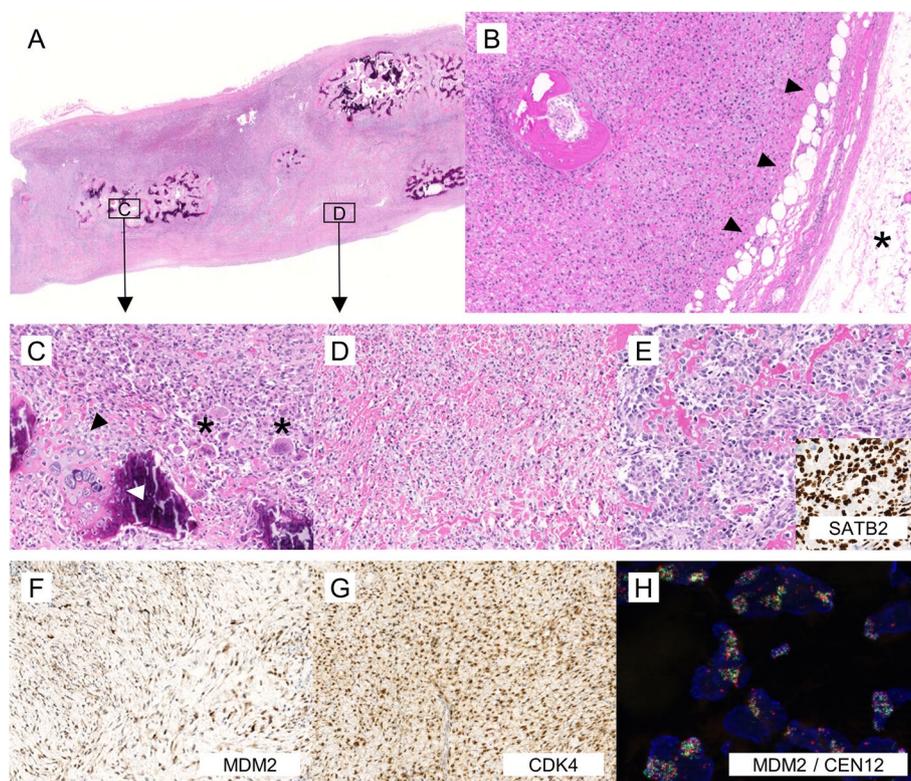
surface and inner lining of the cyst appeared predominantly smooth with some focal hemosiderin deposits. The cyst wall varied in thickness from 0.1 and 1.4 cm, displaying a tan to brown color and, in some areas, a solid, gritty consistency.

Histologically, the wall of the cystic tumor consisted of a spindle cell neoplasia displaying a heterogeneous morphology with a marked variation in cellularity and cellular atypia, ranging from relatively inconspicuous spindle cells to markedly atypical tumor cells (Fig. 4 A-E). Some areas showed a low cellularity with ample amounts of loose to collagenized stroma, while other regions were highly cellular. Neoplastic bone formation, including osteoid and disorganized trabeculae of woven bone, was abundant, corresponding to the calcifications visible in imaging studies (Fig. 4 A, B and E). In addition, small foci of neoplastic cartilage and osteoclast-like giant cells could be identified (Fig. 4 C). At the periphery of the tumor small foci of adipocytes were visible (Fig. 4 B). By immunohistochemistry, the tumor cells were positive for *MDM2*, *CDK4* (Fig. 4 F and G) and *GLUT1*. Areas of osteosarcomatous differentiation showed a strong positivity for *SATB2* (Fig. 4 E inset). FISH analysis of the *MDM2* gene locus 12q15 (Fig. 4 H) showed

several, partially weak *CEN12* signals and dense clusters of uncountable *MDM2* signals corresponding to a *MDM2* amplification. The increased numbers of *CEN12* signals were interpreted as multiple faint alphoid 12 signals, as previously described in the literature [9]. Next generation sequencing (OncoPrint Comprehensive Plus, Thermo Fisher Scientific, Waltham MA, USA) confirmed an amplification of *MDM2* (12q15, 103.17 copies) and *CDK4* (12q14, 52.9 copies). In addition, an amplification of *STAT6* (12q13.3, 17.7 copies) was found. Due to the detection of the *MDM2* amplification and the retroperitoneal location of the tumor a dedifferentiated liposarcoma with heterologous differentiation (osteosarcomatous/ chondrosarcomatous) was diagnosed. The resection of the dedifferentiated liposarcoma was very narrow but complete by histology. In the postoperative multidisciplinary tumor board, close follow-up care was recommended. One year after resection, there was no evidence of recurrence.

## Discussion

Dedifferentiated liposarcomas (DDLPS) are aggressive malignancies that account for a significant proportion of retroperitoneal sarcomas [1]. Cystic changes of DDLPS



**Fig. 4** Histology (A–E) and ancillary studies (F–H) of case 2: **A** Scanning magnification of the wall showing irregular ossifications and calcifications and a variation of cellularity. **B** Periphery of the tumor with sparse intratumoral adipocytes (arrowheads) and adjacent regional adipose tissue (asterisk). **C, D** Higher magnification of A showing (C) areas of high cellularity with foci of cartilaginous differentiation (arrowheads) and osteoclast-like giant cells (asterisk) and (D) areas of lower cellularity with relatively bland small tumor cells in a myxoid and collagen-rich stroma. **E** Area of heterologous osteosarcomatous differentiation with markedly atypical tumor cells producing osteoid and SATB2 expression by immunohistochemistry (inset). **F, G** Immunohistochemical stainings showing strong nuclear positivity for *mdm2* (F) and *cdk4* (G). **H** MDM2 / CEN12 FISH showing uncountable green signals (*MDM2*) and increased red signals (CEN12)

is exceedingly rare and can cause diagnostic challenges. We here report two patients with a retroperitoneal dedifferentiated liposarcoma showing an unusual cystic configuration that complicated the pre-operative diagnostic work-up due to their atypical radiological presentation. Radiologically, dedifferentiated liposarcomas characteristically show both lipomatous and non-lipomatous components [7]. Cystic retroperitoneal masses account for neoplastic and non-neoplastic lesions. The radiological differential diagnosis of a cystic retroperitoneal mass is broad and may include cystic change in a solid neoplasm, in particular cystically degenerated benign peripheral nerve sheath tumors, mucinous cystadenoma, cystic mesothelioma but also peritoneal pseudocysts, developmental cyst and cystic lymphangiomas have to be considered [8, 10, 11]. Cystic degeneration in retroperitoneal liposarcomas is very uncommon and only few case reports describing this phenomenon exist in the literature (Table 1) [11–18]. The reported cases affected both male and female patients between 60 and 86 years of age.

In many of the reported cases the pre-operative radiological differential diagnosis proved difficult and included dedifferentiated liposarcoma [11, 14, 18], peripheral nerve sheath tumor [11], lymphoma [11], a cystic renal neoplasm (renal cell carcinoma or angiomyolipoma) [12] and even benign lesions such as a hematoma [11]. Only three cases were biopsied prior to surgery [11, 13, 15].

Interestingly, in both our cases the initial clinico-radiological differential diagnosis included a hydatid cyst (cystic echinococcosis), potentially due to the proximity of the cystic mass to the liver in both cases and the patient history (contact or proximity to dogs, foxes and other pet animals) in the second case. Cystic echinococcosis is a rare and potentially underreported parasitic disease caused by *Echinococcus spp.* and is mainly transmitted in rural areas. Throughout Europe it has an estimated mean annual incidence of 0.64 cases per 100 000 people. In Switzerland incidence is lower with a mean annual incidence of 0.08 (1997–2020) but has recently increased to 0.16 (2017–2019) [19]. Retroperitoneal

**Table 1** Overview of case reports of cystic degeneration in liposarcomas

Publication	Age/Sex	Site	Size (cm)	Imaging modalities	Radiological differential diagnosis	Histological diagnosis	Ref
Moriyama et al. (2003)	77 m	r	n/a	CT, FDG-PET	malignant retroperitoneal tumor	DDLPS	[15]
Sorour et al. (2023)	86 m	r, pn	13×12×24	CT, MRI	chronic hematoma, liposarcoma, lymphoma, peripheral nerve sheath tumor	DDLPS	[11]
Komine et al. (2019)	60 m	r	23×21×15	CT, MRI, FDG-PET	"mucinous type of retroperitoneal sarcoma", liposarcoma	WDLPS with multilocular cysts	[14]
Van Haverbeke et al. (2017)	69 f	r	3 (recurrence)	MRI	recurrence of the known liposarcoma	DDLPS with heterologous osteosarcomatous and aneurysmal bone cyst-like morphology	[18]
Uchihashi et al. (2017)	67 f	r, pn	21×14×13	MRI	"radiological diagnosis [...] difficult"	DDLPS	[17]
Khoury et al. (2015)	72 f	r, pn	8.7 (primary), 20 (recurrence)	US, CT	n/a	DDLPS	[13]
Núñez et al. (2005)	67 f	mes	8.5×7.5	US, CT	n/a	"myxoid type liposarcoma"	[16]
Horiguchi et al. (2002)	71 m	pn	n/a	CT	cystic renal cell carcinoma, angiomyolipoma	DDLPS	[12]

**Abbreviations:** *f* female, *m* male, *r* retroperitoneal, *pn* perinephric, *mes* mesenteric, *CT* computed tomography, *FDG-PET* fluorodeoxyglucose positron emission tomography, *MRI* magnetic resonance imaging, *US* ultrasound, *DDLPS* dedifferentiated liposarcoma, *WDLPS* well-differentiated liposarcoma

hydatid cysts are uncommon but have been described in the literature [20, 21]. Radiological features that can aid in distinguishing Echinococcus cysts from other mesenchymal lesions include floating membranes, daughter cysts and vesicles. However, their appearance varies depending on the developmental stage of the parasite and confident diagnosis can be additionally complicated by internal hemorrhage and/or superimposed infection [22]. In the first case, the differential diagnostic considerations were further complicated by the patient's known history of endometriosis.

Corresponding to the typical radiology the histological hallmark of a DDLPS is the abrupt transition of a WDLPS to a non-lipogenic sarcoma. Sometimes, however, the WDLPS-component can be only focally present or even absent [23, 24]. The abrupt transition between WDLPS- and DDLPS-components could be observed in the first case where a small adjacent component of WDLPS was present. In the second case no component of WDLPS could be identified.

In addition to the unusual cystic configuration, both cases showed further peculiar morphological features. The first case exhibited areas with a prominent storiform to whorled architecture. Combined with a partial immunohistochemical expression of GLUT1 and Claudin1, these areas were reminiscent of a perineural-like differentiation. Notably, a neural-like or meningotheelial-like whorling pattern in DDLPS has previously been described in small case series in the literature [25, 26].

The second case histologically showed an osteosarcomatous heterologous differentiation. Heterologous differentiation including neoplastic bone formation is a well-described phenomenon in DDLPS [4]. However, to our knowledge, osteosarcomatous heterologous differentiation in combination with a cystic configuration has previously only been described in one case report by Van Haverbeke et al. [18]. Histological differential diagnoses of malignant spindle cell proliferations in the retroperitoneum include leiomyosarcoma, solitary fibrous tumor, inflammatory myofibroblastic tumor, malignant peripheral nerve sheath tumor (MPNST), synovial sarcoma and many others. A spindle cell neoplasm with bone formation raises the question of a MPNST with heterologous differentiation, ossifying synovial sarcoma, extraskelatal osteosarcoma [27–29] and the rare entity of a leiomyosarcoma with osteosarcomatous differentiation [30]. In contrast to DDLPS, all of these entities should not harbour amplifications of *MDM2* [31]. However, according to the literature primary extraskelatal osteosarcomas can harbor amplifications of *MDM2* and distinction from DDLPS may be difficult, particularly in the absence of a WDLPS component [32]. We here favor DDLPS with heterologous osteosarcomatous differentiation due to the retroperitoneal location and the presence of fat in the tumor periphery, while a primary extraskelatal osteosarcoma should remain a diagnosis of exclusion.

The pathophysiological mechanism of the cystic degeneration of liposarcomas remains unknown. In the

literature cystic changes due to hemorrhage or ischemia have been discussed [14, 17]. Both our cases showed hemosiderin deposition and at least focal stromal hyalinization with minimal necrosis indicating the possibility of an ischemic / necrotic process. In addition, both cases expressed GLUT1, which is a downstream-target of hypoxia-induced factor 1-alpha (HIF1-alpha) and is recognized to be involved in the cellular adaptation to hypoxia. Interestingly, GLUT1 expression has been reported in a broad variety of mesenchymal neoplasms [33].

In conclusion, we report two cases of retroperitoneal, perinephric dedifferentiated liposarcoma with prominent cystic degeneration which caused significant diagnostic challenges. Recognizing the existence of cystic change in dedifferentiated liposarcoma is of considerable importance in the diagnostic work-up of these lesions as this will guide patient management.

#### Abbreviations

CT	Computed tomography
DDLPS	Dedifferentiated liposarcoma
ELISA	Enzyme-linked immunosorbent assay
f	female
FDG-PET	Fluorodeoxyglucose positron emission tomography
FISH	Fluorescence in-situ hybridization
m	male
MDT	Multidisciplinary tumor board
MPNST	Malignant peripheral nerve sheath tumor
MRI	Magnetic resonance imaging
PET-CT	Positron emission tomography-computed tomography
US	Ultrasound
WDLPS	Well-differentiated liposarcoma

#### Acknowledgements

We would like to thank both patients to agree to share their patient history for educational purposes.

#### Authors' contributions

VKV: putting data together, figures, writing PL: performed surgeries of both patients, writing FE: radiology of both cases, writing MLB: therapy decision making, writing LB: oncologist input, writing MB: putting data together, writing CP: putting data together, writing.

#### Data availability

No datasets were generated or analysed during the current study.

#### Declarations

##### Ethics approval and consent to participate

The present study was conducted following regional/ cantonal and institutional guidelines and in compliance with the Helsinki Declaration and after approval by our cantonal ethical review board Zurich (BASEC- 2021–00417). Both patients have agreed and signed a general consent to allow the material used for research purposes by investigators from the University Hospital Zurich, Switzerland.

##### Competing interests

The authors declare no competing interests.

##### Author details

<sup>1</sup>Department of Pathology and Molecular Pathology, University Hospital Zurich, Zurich, Switzerland. <sup>2</sup>Department of Visceral Surgery and Transplantation, University Hospital Zurich, Zurich, Switzerland. <sup>3</sup>Diagnostic and Interventional Radiology, University Hospital Zurich, University of Zurich, Zurich,

Switzerland. <sup>4</sup>Department of Radiation Oncology, University Hospital Zurich, Zurich, Switzerland. <sup>5</sup>Department of Medical Oncology and Hematology, University Hospital Zurich, Zurich, Switzerland. <sup>6</sup>Medical Faculty, University of Zurich, Zurich, Switzerland. <sup>7</sup>Swiss Sarcoma Center, Comprehensive Cancer Center Zurich, University and University Hospital Zurich, Zurich, Switzerland.

Received: 12 November 2024 Accepted: 15 February 2025

Published online: 27 February 2025

#### References

- Schmitz E, Nessim C. Retroperitoneal Sarcoma Care in 2021. *Cancers*. 2022;14(5):1293.
- Downes KA, Goldblum JR, Montgomery EA, Fisher C. Pleomorphic liposarcoma: a clinicopathologic analysis of 19 cases. *Mod Pathol*. 2001;14:179–84.
- Kilpatrick SE, Doyon J, Choong PF, et al. The clinicopathologic spectrum of myxoid and round cell liposarcoma. A study of 95 cases. *Cancer*. 1996;77:1450–8.
- Alaggio R, Coffin CM, Weiss SW, et al. Liposarcomas in young patients: a study of 82 cases occurring in patients younger than 22 years of age. *Am J Surg Pathol*. 2009;33:645–58.
- 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;21:271–81.
- Sirvent N, Coindre JM, Maire G, et al. Detection of MDM2-CDK4 amplification by fluorescence in situ hybridization in 200 paraffin-embedded tumor samples: utility in diagnosing adipocytic lesions and comparison with immunohistochemistry and real-time PCR. *Am J Surg Pathol*. 2007;31:1476–89.
- Francis IR, Cohan RH, Varma DGK, Sondak VK. Retroperitoneal sarcomas. *Cancer Imaging*. 2005;5:89–94.
- Yang DM, Jung DH, Kim H, et al. Retroperitoneal cystic masses: CT, clinical, and pathologic findings and literature review. *Radiographics*. 2004;24:1353–65.
- Kashima T, Halai D, Ye H, et al. Sensitivity of MDM2 amplification and unexpected multiple faint alpha 12 (alpha 12 satellite sequences) signals in atypical lipomatous tumor. *Mod Pathol*. 2012;25:1384–96.
- Scali EP, Chandler TM, Heffernan EJ, et al. Primary retroperitoneal masses: what is the differential diagnosis? *Abdom Imaging*. 2015;40:1887–903.
- Sorour S, Bao B, Wilson MP, Low G. Cystic retroperitoneal dedifferentiated liposarcoma: A case report. *J Clin Imaging Sci*. 2023;13:22.
- Horiguchi A, Oyama M [Perinephric liposarcoma mimicking cystic renal tumor]. *Nihon Hinyokika Gakkai Zasshi*. 2002;93:491–4.
- Khoury M, Sim GC, Harao M, et al. Multicystic dedifferentiated retroperitoneal liposarcoma: tumour cyst fluid analysis and implications for management. *Case Reports*. 2015;2015:bcr2015211218.
- Komine C, Fukuchi M, Sakurai S, et al. Retroperitoneal Liposarcoma with Multilocular Cysts. *Case Rep Gastroenterol*. 2019;13:514–20.
- Moriyama K, Negishi T, Furubayashi N, et al. A Case of Cystic Retroperitoneal Dedifferentiated Liposarcoma Diagnosed by Percutaneous Image-Guided Biopsy. *Case Rep Oncol*. 2023;16:1460–5.
- Núñez Fernández MJ, García Blanco A, López Rodríguez A, et al. Primary mesenteric liposarcoma of jejunum: presentation like a cystic mass. *Minerva Med*. 2005;96:425–8.
- Uchihashi K, Matsuyama A, Shiba E, et al. Retroperitoneal dedifferentiated liposarcoma with huge cystic degeneration: A case report. *Pathol Int*. 2017;67:264–8.
- Van Haverbeke C, Van Dorpe J, Lecoutere E, et al. Dedifferentiated Liposarcoma of the Retroperitoneum With Heterologous Osteosarcomatous Differentiation and a Striking Aneurysmal Bone Cyst-Like Morphology. *Int J Surg Pathol*. 2017;25:374–8.
- Casulli A, Abela-Ridder B, Petrone D, et al. Unveiling the incidences and trends of the neglected zoonosis cystic echinococcosis in Europe: a systematic review from the MEME project. *Lancet Infect Dis*. 2023;23:e95–107.
- Slavu IM, Gheorghita V, Macovei Oprea AM, et al. Primary Retroperitoneal Hydatid Cyst: A Diagnostic and Treatment Conundrum. *Cureus*. 2024;16:e53842.

21. Yadav SK, Ruchal A, Gaurav B, et al. Retroperitoneal hydatid cyst challenging the diagnosis: Case report with review of literature. *Int J Surg Case Rep.* 2024;114:109106.
22. Zalaquett E, Menias C, Garrido F, et al. Imaging of Hydatid Disease with a Focus on Extrahepatic Involvement. *Radiographics.* 2017;37:901–23.
23. McCormick D, Mentzel T, Beham A, Fletcher CD. Dedifferentiated liposarcoma. Clinicopathologic analysis of 32 cases suggesting a better prognostic subgroup among pleomorphic sarcomas. *Am J Surg Pathol.* 1994;18:1213–23.
24. Thway K, Jones RL, Noujaim J, et al. Dedifferentiated Liposarcoma: Updates on Morphology, Genetics, and Therapeutic Strategies. *Adv Anat Pathol.* 2016;23:30–40.
25. Fanburg-Smith JC, Miettinen M. Liposarcoma with meningothelial-like whorls: a study of 17 cases of a distinctive histological pattern associated with dedifferentiated liposarcoma. *Histopathology.* 1998;33:414–24.
26. Nascimento AG, Kurtin PJ, Guillou L, Fletcher CD. Dedifferentiated liposarcoma: a report of nine cases with a peculiar neurallike whorling pattern associated with metaplastic bone formation. *Am J Surg Pathol.* 1998;22:945–55.
27. Guo A, Liu A, Wei L, Song X. Malignant peripheral nerve sheath tumors: differentiation patterns and immunohistochemical features - a mini-review and our new findings. *J Cancer.* 2012;3:303–9.
28. Winnepeninckx V, De Vos R, Debiec-Rychter M, et al. Calcifying/ossifying synovial sarcoma shows t(X;18) with SSX2 involvement and mitochondrial calcifications. *Histopathology.* 2001;38:141–5.
29. Chung EB, Enzinger FM. Extraskeletal osteosarcoma. *Cancer.* 1987;60:1132–42.
30. Yu S, Hornick JL. "Malignant Mesenchymoma" Revisited: A Clinicopathologic Study of Leiomyosarcomas With Osteosarcomatous Differentiation. *Am J Surg Pathol.* 2022;46:1430–5.
31. Choi JH, Ro JY. Retroperitoneal Sarcomas: An Update on the Diagnostic Pathology Approach. *Diagnostics (Basel).* 2020;10(9):642.
32. Yamashita K, Kohashi K, Yamada Y, et al. Primary extraskeletal osteosarcoma: a clinicopathological study of 18 cases focusing on MDM2 amplification status. *Hum Pathol.* 2017;63:63–9.
33. Ahrens WA, Ridenour RV 3rd, Caron BL, et al. GLUT-1 expression in mesenchymal tumors: an immunohistochemical study of 247 soft tissue and bone neoplasms. *Hum Pathol.* 2008;39:1519–26.

## Publisher's Note

Springer Nature remains neutral with regard to jurisdictional claims in published maps and institutional affiliations.