

一例 45 cm 腹膜后去分化脂肪肉瘤报告

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摘要: **目的** 探讨腹膜后去分化脂肪肉瘤(DDLPS)的临床特点、诊断过程、手术治疗及围手术期管理,以提高对该巨大罕见肿瘤的认识,为临床诊治提供参考。**方法** 本文报道北京协和医院泌尿外科 2025 年 9 月收治的一例 57 岁男性患者。回顾性分析此例患者电子病历资料,包括就诊记录、住院病历及辅助检查结果,患者住院期间的治疗。**结果** 患者门诊就诊时查体扪及左侧腹部巨大包块,影像学显示肿瘤推挤脾脏、胰腺体尾部、左肾及左肾上腺移位,同时压迫左侧输尿管导致上尿路积水。肿瘤完整切除后测量最大径达 45 cm,重约 12.5 kg。病理荧光原位杂交(FISH)检测示鼠双微体 2(MDM2)信号扩增,提示 DDLPS 诊断。患者术后恢复良好,术后 11 d 出院,术后 1 个月随访一般情况可。**结论** 该病例影像学表现复杂,伴多形性钙化者较为罕见;对于此类巨大腹膜后肿物,应结合影像学、病理学及 MDM2 扩增检测明确诊断,并通过充分术前评估和完整手术切除以改善患者压迫症状及预后。

关键词: 去分化脂肪肉瘤;腹膜后肿瘤;罕见病;鼠双微体 2(MDM2)扩增

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One case report of a 45 cm retroperitoneal dedifferentiated liposarcoma

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Abstract: **Objective** To investigate the clinical characteristics, diagnostic process, surgical treatment and peri-operative management of retroperitoneal dedifferentiated liposarcoma (DDLPS) and to improve awareness of this type of rare giant tumor while providing a reference for future clinical diagnosis and treatment. **Methods** A case of a 57-year-old male patient admitted to the Department of Urology, Peking Union Medical College Hospital in September 2025. The patient's electronic medical records were retrospectively reviewed, including outpatient and inpatient records, laboratory and radiology results, and treatments during hospitalization. **Results** Physical examination revealed a giant palpable mass in the left abdomen. Radiology imaging showed significant displacements of the spleen, pancreas, left kidney, and left adrenal gland by the tumor and a compressed left ureter, resulting in upper urinary tract hydronephrosis. After en bloc resection, the tumor measured up to 45 cm in maximum diameter and weighed approximately 12.5 kg. Pathological fluorescence in situ hybridization (FISH) demonstrated amplification of murine double minute 2 (MDM2), supporting the diagnosis of DDLPS. The patient recovered uneventfully after surgery, was discharged on postoperative day 11, and remained in generally good condition at the 1-month follow-up. **Conclusions** The case demonstrated complex imaging features, since DDLPS with polymorphic calcification is relatively rare. For retroperitoneal masses of this size, diagnosis should be established based on a combination of

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imaging findings, pathological examination, and MDM2 amplification testing. Adequate preoperative assessment and complete surgical resection are also essential for relieving compression-related symptoms and improving prognosis.

Key words: dedifferentiated liposarcoma; retroperitoneal tumor; rare disease; murine double minute 2 (MDM2) amplification

脂肪肉瘤主要病理亚型包括高分化脂肪肉瘤 (well-differentiated liposarcoma, WDLPS)、黏液型脂肪肉瘤 (myxoid liposarcoma, MLPS)、去分化脂肪肉瘤 (dedifferentiated liposarcoma, DDLPS) 等^[1]。其中, WDLPS 约占 30%~33%, DDLPS 约占 14%~20%, 后者以腹膜后占位最常见, 恶性程度、局部侵袭性和复发风险较高^[2-3]。因其生长位置深、症状隐匿, DDLPS 发现时常已生长至较大体积, 平均直径可达 30 cm, 40 cm 以上也曾见文献报道^[4]。近期加拿大安大略省一项回顾性研究显示, DDLPS 累积患病率约为 1.995/10 万, 年发生率约为 0.465/10 万, 男性略多于女性, 多见于中老年人^[5]。典型 DDLPS 同时包含脂肪性以及非脂肪性成份, 约 10% 的病例可出现异源性分化, 但骨样分化及其罕见^[6]。现将北京协和医院收治的一例直径 45 cm, 重达 12.5 kg 的巨大腹膜后 DDLPS 患者的临床资料、影像学表现、手术治疗及病理结果进行总结, 以期为提高该病的临床识别和诊疗水平提供参考。

1 临床资料

1.1 一般资料

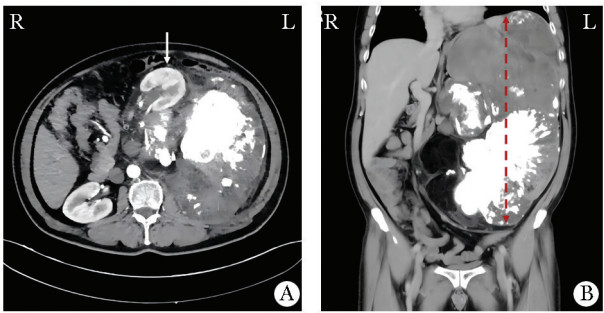
患者男性, 57 岁, 2025 年 10 月因“发现左侧腹膜后肿物半年”收治入院。患者诉偶有腹痛、腹胀不适, 否认肉眼血尿、发热、恶心呕吐等症状。查体可见腹部明显膨隆, 左侧腹部可扪及巨大包块, 质硬, 无明显压痛; 双侧输尿管走行区无压痛, 膀胱区无充盈。既往高血压病史 4 年余, 规律服药后血压控制可, 余既往史、个人史及家族史无特殊。

1.2 检查

入院后完善相关实验室检查, 血常规示白细胞计数 7.26×10^9 个/L, 中性粒细胞比例 0.73, 血红蛋白 131 g/L, 血小板 261×10^9 个/L; 神经元特异性烯醇化酶 (neuron-specific enolase, NSE) 17.4 ng/mL, 立位肾素活性 0.42 ng/mL/h, 血儿茶酚胺及其代谢产物水平未见明显异常。

术前行 CT 尿路成像 (computed tomography urography, CTU) 及正电子发射断层显像/计算机体

层成像 (positron emission tomography/computed tomography, PET/CT) 检查, 提示左侧腹膜后可见巨大混杂密度肿物, 上缘导致左膈抬升, 下缘达骨盆入口, 病灶内可见多发不规则高密度影、液性低密度区及脂肪密度影, 范围约 23 cm×19 cm×33 cm。增强扫描可见实性成分不均匀强化, 实性部分放射性摄取增高, 最大标准摄取值 (SUV_{max}) 为 28.2, 未见明确转移及淋巴结受累。占位推挤脾脏、胰腺体尾部、左肾及左肾上腺移位, 并压迫左侧输尿管腹段, 导致局部管腔狭窄, 上游输尿管及左侧肾盂肾盏扩张积水 (图 1)。术前肾动态显像提示左肾未见有效灌注, 右肾功能保留良好, 肾小球滤过率 (glomerular filtration rate, GFR) 为 74 mL/(min·1.73 m²)。



A. tumor markedly displaced the left kidney to the right upper abdomen; White arrow indicated the left kidney; B. tumor expanded from the subdiaphragmatic region to the pelvis; Red dashed arrows indicated the superior and inferior border of the tumor.

图 1 患者术前 CT 影像

Fig 1 Patient's preoperative CT images

1.3 诊断

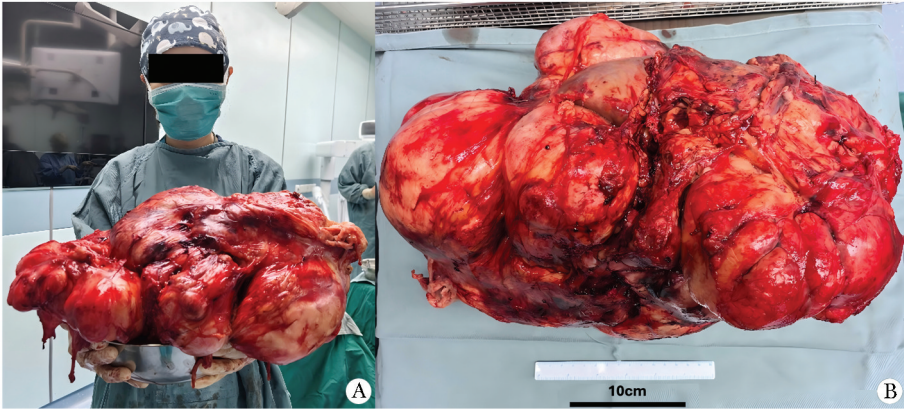
术后病理组织学检查提示左腹膜后肿物为去分化脂肪肉瘤 (dedifferentiated liposarcoma, DDLPS), 进一步行荧光原位杂交 (fluorescence *in situ* hybridization, FISH) 检测, 结果显示鼠双微体 2 (murine double minute 2, MDM2) 信号呈点簇状分布并存在信号扩增, 约 60% 的细胞检出 MDM2 信号扩增。结合肿瘤部位、影像学特征、组织病理学表现及 MDM2

扩增结果,最终诊断为左侧腹膜后去分化脂肪肉瘤,恶性程度偏高。

1.4 治疗

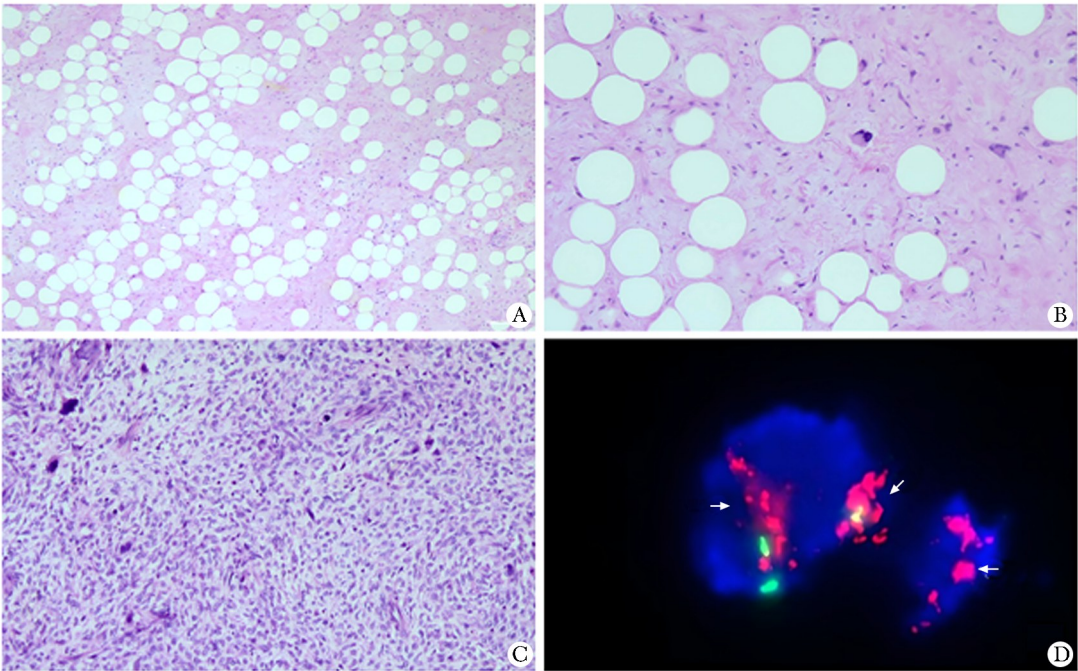
充分评估手术风险后,患者术前行肿瘤供血血管(左肾动脉)栓塞以减少术中出血。经剑突至左下腹弧形切口行开腹探查见左侧腹膜后巨大肿物,

占据左侧腹腔及腹膜后间隙,与周围组织关系密切,左肾被肿瘤完整包绕。肿瘤推挤邻近脏器移位,肠管虽受明显推挤,但未见明确侵犯。遂行腹膜后巨大肿物切除术及左肾切除术。手术历时约4 h,术中出血约1 500 mL。术后标本测得肿瘤最大径约45 cm,重量约12.5 kg(图2)。



A. size comparison of the resected tumor to surgeon; B. resected mass had an intact capsule, with a soft peripheral component and a central calcified region.

图2 肿瘤大体表现
Fig 2 Gross appearance of the resected giant tumor



A. low-power view($\times 40$) of the well-differentiated liposarcoma component; B. high-power view($\times 100$) of the well-differentiated liposarcoma component; C. high-power view($\times 100$) of the dedifferentiated liposarcoma component; D. fluorescence *in situ* hybridization for MDM2($\times 1\,000$) demonstrates MDM2 gene amplification, supporting the diagnosis of dedifferentiated liposarcoma; White arrows indicated the punctate cluster-like distribution of MDM2 signals.

图3 肿瘤病理组织学切片及荧光原位杂交
Fig 3 Histopathological and molecular *in situ* hybridization of the tumor

表 1 患者术前及术后实验室指标变化

Table 1 Changes in preoperative and postoperative laboratory parameters

category parameters	abbreviation	pre-op	post-op d1	post-op d2	post-op d7
liver function/nutrition					
albumin/(g/L)	Alb	38	33 ↓	31 ↓	37
alanine aminotransferase/(U/L)	ALT	10	9	9	16
total bilirubin/(μmol/L)	TBil	15. 1	22. 9 ↑	20. 4	15. 3
direct bilirubin/(μmol/L)	DBil	4. 0	10. 0 ↑	7. 7 ↑	5. 1
kidney function					
creatinine/(μmol/L)	Cr(E)	58 ↓	83	79	70
urea/(mmol/L)	Urea	4. 0	6. 2	6. 2	6. 6
potassium(mmol/L)	K	4. 5	—	4. 2	5. 0
sodium/(mmol/L)	Na	141	—	139	139
corrected calcium/(mmol/L)	cCa	2. 24	2. 20	2. 24	2. 27
hemorrhage					
red blood cell/(/L)	RBC	4. 90 ×10 ¹²	2. 41 ×10 ¹² ↓	2. 41 ×10 ¹² ↓	2. 85 ×10 ¹² ↓
hemoglobin/(g/L)	HGB	131	68 g/L ↓	69 g/L ↓	80 g/L ↓
hematocrit/%	HCT	42. 8	19. 9 ↓	20. 5 ↓	24. 3 ↓
platelet/(/L)	PLT	261 ×10 ⁹	139 ×10 ⁹	174 ×10 ⁹	348 ×10 ⁹
inflammation					
white blood cell/(/L)	WBC	7. 26 ×10 ⁹	9. 95 ×10 ⁹ ↑	12. 69 ×10 ⁹ ↑	6. 45 ×10 ⁹
neutrophil percentage(%)	NEUT%	73. 0	86. 0 ↑	90. 1 ↑	71. 3
absolute neutrophil count/(/L)	NEUT#	5. 30 ×10 ⁹	8. 55 ×10 ⁹ ↑	11. 44 ×10 ⁹ ↑	4. 60 ×10 ⁹
lymphocyte percentage/%	LY%	19 ↓	6. 9 ↓	6. 3 ↓	19. 1 ↓
C-reactive protein/ high-sensitivity C-reactive protein(mg/L)	CRP/hsCRP	—	—	138. 04 ↑	51. 51 ↑
coagulation/thrombotic risk prothrombin time/sec					
international normalized ratio	PT	12. 1 s	12. 8 ↑	11. 3	12. 2
activated partial thromboplastin time/sec	INR	1	1. 03	0. 92	0. 98
fibrinogen/(g/L)	APTT	28. 1 s	35. 5 ↑	31. 9	28. 6
D-dimer/(mg/L FEU)	Fbg	4. 38 ↑	3. 32	4. 86 ↑	3. 76 ↑
myocardial injury	D-Dimer	2. 36 ↑	—	4. 34 ↑	9. 34 ↑
creatine kinase(U/L)	CK	—	235 ↑	207 ↑	26
myoglobin/(μg/L)	Myo	—	466 ↑	81	33
high-sensitivity cardia troponin I/(ng/L)	hs-cTn I	—	8	4	<2. 5
N-terminal pro-B-type natriuretic peptide/(pg/mL)	NT-proBNP	—	86	400 ↑	280 ↑
post-abdominal surgery					
amylase/(U/L)	AMY	—	945 ↑	200 ↑	89
lipase/(U/L)	LIP	—	27	37	74 ↑

Changes in the patient’s major laboratory parameters before and after surgery for preoperative status and perioperative inflammatory response evaluation. Results were collected at four time points: preoperatively (pre-op) , postoperative day 1 (post-op d1) , postoperative day 3 (post-op d3) , and 1 week after surgery (post-op w1) ; “ ↑ ” indicated a value above the corresponding reference range, whereas “ ↓ ” indicated a value below the corresponding reference range; The units for each parameter were indicated in the table.

患者术后恢复良好,术后 6 d 可经口进食,术后 11 d 予以出院。术后 1 个月患者于本院泌尿外科门诊随访,一般情况可,未诉明显腹痛、腹胀等不适。

2 讨论

约 10%的DDLPS 病例可出现异质性分化,包括

横纹肌样、软骨-骨样分化或其他更为罕见的组织成分^[6]。此例肿瘤可见大面积及多形性的钙化,并混杂脂肪密度及囊性成分。肿瘤中央巨大骨化灶达 15 cm×13 cm×12 cm,且具有“爆米花样”钙化以及囊性分隔膜环形钙化等多种组织成分并存的异质性表现。上述特征在DDLPS 中较为罕见,同时也提示

肿瘤去分化及高度恶性的可能。2025 年发表的软组织肿瘤诊断指南中指出,肿瘤直径>10 cm、结节样非脂肪成分>1 cm、分隔厚度>2 mm 以及无定形或骨样钙化为 DDLPS 的重要影像学表现^[7]

DDLPS 复发率高、预后差,总体病死率接近 28%,5 年生存率约 49.4%,远处转移率约 15%~30%,局部复发率约 40%,也是影响患者长期生存的重要因素^[8]。因此,术中肿瘤 R0 切除(完整切除肿瘤)是降低复发风险的关键。对于巨大肿瘤,术前充分评估肿瘤供血并对主要供血动脉进行栓塞是提高手术安全性、肿瘤完整切除可能的关键。

由于该病局部复发风险较高,术后应对患者定期进行腹盆部增强 CT 或 MRI 以早期发现复发。当局部复发生长速度较快(>0.9 cm/月)将显著增加切除难度,患者预后差^[9]。肺实质是 DDLPS 较常见的远处转移部位,因此胸部 CT 同样应作为术前评估和术后随访的重要组成部分^[10]。此例患者术前评估影像中未见明确远处转移灶。目前,受限于 DDLPS 的罕见

性,关于系统药物治疗的循证医学证据仍较有限,辅助放射和化学治疗的获益暂不明确。基于 DDLPS 分子特征的 CDK4/6 抑制剂等靶向治疗正在开展临床研究,但现有结果仍处于初步阶段^[11]。

本例患者术前影像及术中情况见肿瘤完整包绕左肾,阻断左肾血流灌注,压迫左侧输尿管。其次,此例肿瘤内部同时存在脂肪密度、液性低密度区、不规则高密度影及实性强化成分,并伴有多形性钙化。肿瘤中央大范围骨化灶,脂肪样成分与肾脏关系密切,局部呈膨胀性生长,且具有多发“爆米花样”钙化灶及囊性成分。如此复杂多变的影像学特征在脂肪肉瘤中并不常见,使本例具有较高的临床和教学价值(图 1)。且由于软组织肉瘤影像表现多样,早期症状缺乏特异性,约 30%在初诊时可被误诊^[2]。因此,应提高临床警惕性、加强影像学识别能力,并通过泌尿外科、影像科、病理科及肿瘤科等多学科协作制定个体化诊疗方案,以实现早期诊断、完整切除和患者预后改善。

参考文献:

- [1] Liu H, Wang X, Liu L, *et al.* Targeting liposarcoma: unveiling molecular pathways and therapeutic opportunities [J/OL]. *Front Oncol*, 2024, doi: 10.3389/fonc.2024.1484027.
- [2] Kyriazoglou A, Pagkali A, Kotsantis I, *et al.* Well-differentiated liposarcomas and dedifferentiated liposarcomas: systemic treatment options for two sibling neoplasms [J/OL]. *Cancer Treat Rev*, 2024, 125: 102716. doi:10.1016/j.ctrv.2024.102716.
- [3] Thway K, Jones RL, Noujaim J, *et al.* Dedifferentiated liposarcoma: updates on morphology, genetics, and therapeutic strategies [J/OL]. *Adv in Anat Pathol*, 2016, 23: 30-40. doi:10.1097/PAP.000000000000101.
- [4] Lee ATJ, Thway K, Huang PH, *et al.* Clinical and molecular spectrum of liposarcoma [J/OL]. *J Clin Oncol*, 2018, 36: 151-159. doi:10.1200/JCO.2017.74.9598.
- [5] Seung SJ, Wong A, Milan R, *et al.* Epidemiology, treatment patterns, survival, healthcare resource utilization, and costs of dedifferentiated liposarcoma (DDLPS) in Canada: a retrospective cohort study using administrative databases in Ontario [J/OL]. *Curr Oncol*, 2025, 32: 273. doi:10.3390/curroncol32050273.
- [6] Francis IR. Retroperitoneal sarcomas [J/OL]. *Cancer Imaging*, 2005, 5: 89-94. doi: 10.1102/1470-7330.2005.0019.
- [7] Masroori Z, Mirghaderi P, Haseli S, *et al.* Pictorial review of soft tissue lesions with calcification [J/OL]. *Diagnostics*, 2025, 15: 811. doi:10.3390/diagnostics15070811.
- [8] De Sanctis R, Giordano L, Colombo C, *et al.* Long-term follow-up and post-relapse outcome of patients with localized retroperitoneal sarcoma treated in the Italian sarcoma group-soft tissue sarcoma (ISG-STs) protocol 0303 [J/OL]. *Ann Surg Oncol*, 2017, 24: 3872-3879. doi:10.1245/s10434-017-6105-y.
- [9] Jolissaint JS, Raut CP, Fairweather M. Management of recurrent retroperitoneal sarcoma [J/OL]. *Curr Oncol*, 2023, 30: 2761-2769. doi:10.3390/curroncol30030209.
- [10] Haddox CL, Nathenson MJ, Mazzola E, *et al.* Phase II study of eribulin plus pembrolizumab in metastatic soft-tissue sarcomas: clinical outcomes and biological correlates [J/OL]. *Clin Cancer Res*, 2024, 30: 1281-1292. doi:10.1158/1078-0432.CCR-23-2250.
- [11] Haddox CL, Hornick JL, Roland CL, *et al.* Diagnosis and management of dedifferentiated liposarcoma: a multidisciplinary position statement [J/OL]. *Cancer Treat Rev*, 2024, 131: 102846. doi:10.1016/j.ctrv.2024.102846.