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·临床研究·

巨大腹膜后平滑肌肉瘤伴出血 1 例报告并文献复习

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摘 要

目的：探讨腹膜后平滑肌肉瘤的临床特点、影像学表现及治疗策略，以期对类似病例的诊治提供参考。

方法：回顾性分析收治的 1 例腹膜后平滑肌肉瘤患者临床资料，并复习相关文献。

结果：患者腹部胀痛伴贫血，影像学提示中下腹部恶性肿瘤伴出血，早期行液体复苏，进一步介入栓塞，超声引导下穿刺引流，避免腹腔隔室综合征的发生；复查待瘤体缩小，贫血纠正，行完整切除肿瘤，病理诊断为平滑肌肉瘤。术后随访 16 个月，患者恢复良好。

结论：腹膜后平滑肌肉瘤是一种罕见的恶性肿瘤，起病隐匿，易复发和转移，手术完整切除是其主要治疗方式，配合以化疗、放疗等手段综合治疗，可改善患者预后。

关键词 腹膜后肿瘤；平滑肌肉瘤；预后；病例报告

中图分类号：R735.4

Huge retroperitoneal leiomyosarcoma with hemorrhage: an analysis of one case and literature review

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Abstract

Objective: To investigate the clinical features, imaging manifestation and treatment strategies of retroperitoneal leiomyosarcoma, so as to provide reference for the diagnosis and treatment of similar cases.

Methods: The clinical data of one admitted patient with retroperitoneal leiomyosarcoma was analyzed retrospectively, and the relevant literature was reviewed.

Results: The patient had abdominal pain and anemia, and the preoperative imaging examination indicated a malignant tumor with hemorrhage in hypogastric region. The patient underwent fluid replacement therapy and interventional embolization first, and then ultrasound-guided puncture and drainage to avoid the occurrence of abdominal compartment syndrome. After tumor size reduction and anemia correction, the tumor was completely resected, and the pathological diagnosis was leiomyosarcoma. Postoperative follow-up was conducted for 16 months, and the patient had an uneventful recovery.

Conclusion: Retroperitoneal leiomyosarcoma is a rare malignant tumor, which has an insidious onset and is more likely to develop recurrence and metastasis. Complete surgical excision is the main treatment method, which in

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combination with multimodality therapy such as chemotherapy and radiotherapy can improve the prognosis of patients.

Key words Retroperitoneal Neoplasms; Leiomyosarcoma; Prognosis; Case Reports
CLC number: R735.4

腹膜后平滑肌肉瘤（retroperitoneal leiomyosarcoma）是来源于平滑肌细胞或可向平滑肌细胞分化的间叶细胞，女性多见，因其位置较深，起病隐匿，缺乏特异性症状，且呈侵袭性生长，患者因腹部症状就诊时肿瘤生长较大，肿瘤直径超过10 cm，常难以整块切除，易复发和远处转移，预后较差[1-2]。腹膜后平滑肌肉瘤临床少见，而合并出血病例更是非常罕见。本文总结1例巨大腹膜后平滑肌肉瘤合并瘤内出血患者诊治经验，为腹膜后平滑肌肉瘤的诊断及治疗提供参考。

1 病例资料

患者 男，49岁，因“腹部胀痛20 d”入院。查体：心率110次/min，血压110/54 mmHg（1 mmHg=0.133 kPa），贫血貌，腹部高度膨隆，上腹部叩诊腹部呈浊音，下腹部叩诊为实音，可触及明显包块，上腹部较软，下腹部质硬，推之无法活动，轻度压痛，移动性浊音阳性，肠鸣音遥远。辅助检查：CA-125 244.95 U/mL，CA19-9 52.4 U/mL，入院时血红蛋白94 g/L，呈现进行降低，最低时为78 g/L，凝血、生化、肝功正常。腹部增强CT（图1）：中下腹见团块样、囊实性混杂密度影，最大层面29 cm×15 cm，可见多发钙化影，明显不均匀强化，病变包绕右侧髂总及髂外静脉，考虑来源于间叶组织的恶性肿瘤。PET-CT提示：中下腹部巨大混杂密度高代谢肿块，考虑间叶来源的恶性肿瘤，未见明显的腹腔外转移（图2）。

因该血红蛋白进行性降低，肿物占据整个腹腔，出现呼吸困难及进食后呕吐的情况，首先行DSA造影，右下腹巨大不规则异常肿瘤染色区，其内见迂曲血管影，肠系膜上下动脉、右侧第2~5腰动脉及右侧髂内动脉均有分支供应（图3），超选滋养动脉，应用碘化油栓塞，然后辅以明胶海绵。栓塞完成后待血红蛋白稳定后行腹腔内及肿物囊内穿刺。充分引流，引流液为陈旧的血性液体，共计引流9 000 mL，未查到阳性脱落细胞，肿瘤囊性部分体积明显缩小（图4），同时穿刺实性肿物病理结果为：穿刺组织内可见大量梭形细胞。纠正营养状态及贫血，保护重要脏器功能，充分备血，预置右侧输尿管支架，准备异体髂血管及人工血管，在多学科会诊及保障下行手术治疗。

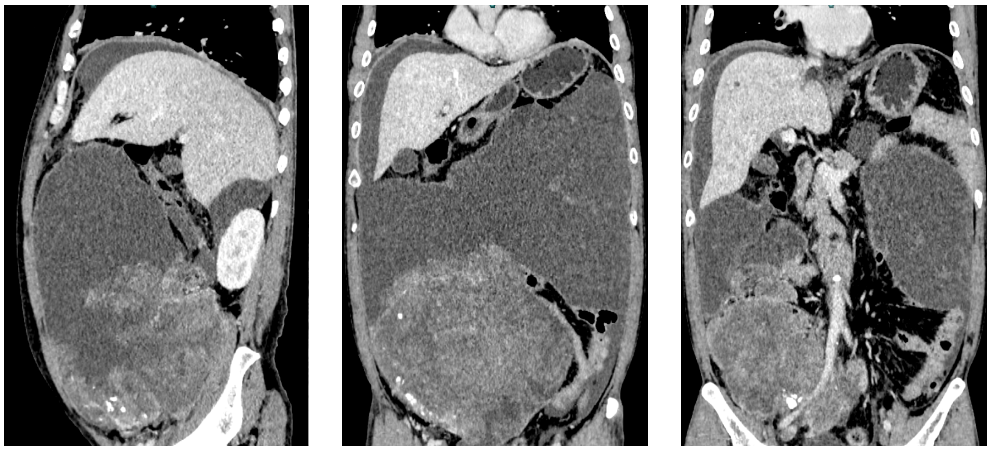


图 1 增强 CT 重建图显示混杂密度团块，上至肝下缘，下到盆腔，实性部分动脉期呈现不均匀强化

Figure 1 Enhanced CT reconstruction showing mixed density mass ranging from the inferior margin of the liver to the pelvic cavity, with heterogeneous enhancement of the solid part during arterial phase

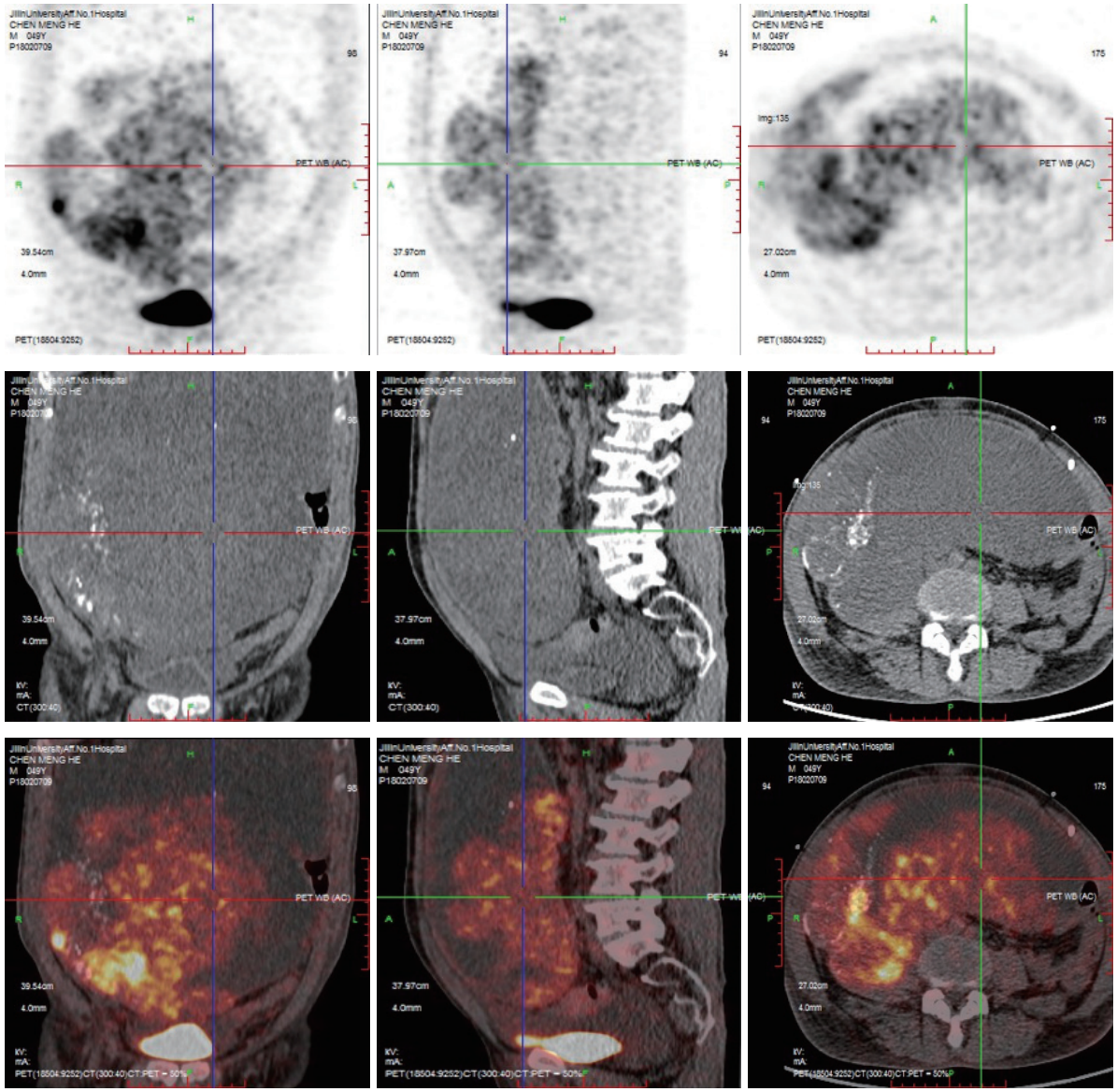


图 2　PET-CT 显示中下腹部巨大混杂密度团块，放射性高摄取病灶范围 20.2 cm×19.3 cm×20.7 cm，SUV 值 4.8~9.6

Figure 2　PET-CT showing the mixed density mass in the lower-middle part of the abdomen, and a high uptake lesion with a scope of 20.2 cm×19.3 cm×20.7 cm and SUV value from 4.8 to 9.6

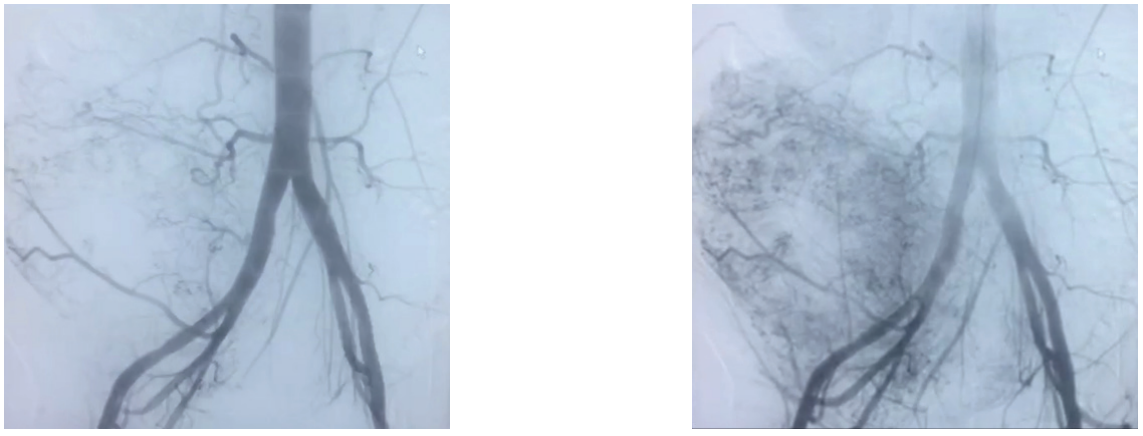


图 3　造影显示供应肿瘤的动脉

Figure 3　Angiography demonstrating the arteries supplying the tumor

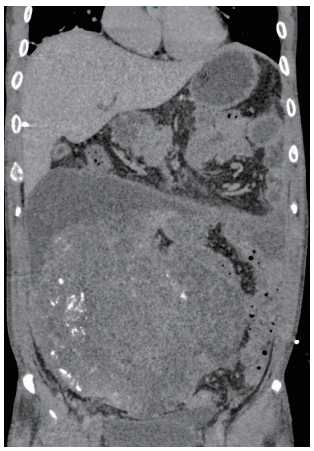


图 4 肿瘤及腹腔穿刺后 CT 重建（术前最大直径 29 cm，术后最大直径 24 cm）

Figure 4 CT reconstruction after tumor and abdominal puncture (preoperative maximum diameter of 29 cm, postoperative maximum diameter of 24 cm)

剑突下到耻骨联合的正中开口，脐下2 cm处右侧横行切口到腋前线。灰白 色肿物占据整个腹腔，与腹膜间存在致密的间隙，沿间隙分离进入腹腔，肿物挤压腹腔内脏器，右下腹部与腹膜后组织固定，无法推动，先行去除大部分肿物便于处理重要血管，进一步探查，肿瘤完整包绕输尿管、股神经，与髂总动脉、髂外静脉呈180° 接触，沿着神经外膜、髂总动脉切除肿瘤组织，切除局部输尿管进行重建（图5），完整切除肿瘤。术中出血2 045 mL，术后恢复顺利，无并发症发生。病理结果为：平滑肌肉瘤（图6）。免疫组化Ki-67（40%+），SMA（+）。术后第7天出院，经多学科会诊后进行后续化疗。随访16个月，患者状态良好。

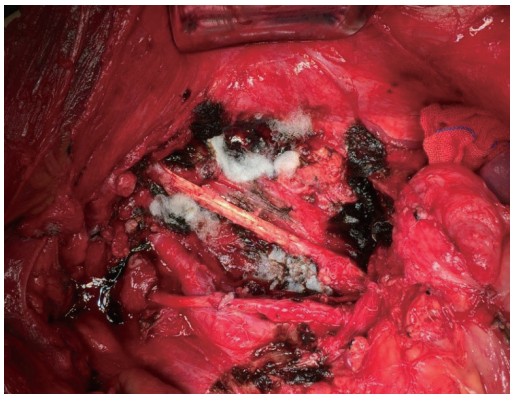
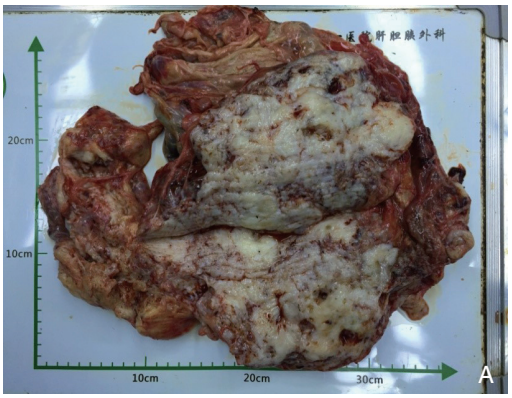


图 5 切除肿瘤后呈现的股神经，髂血管及输尿管的重建

Figure 5 Exposure of the femoral nerve after tumor resection, and reconstruction of the iliac vessels and the ureter



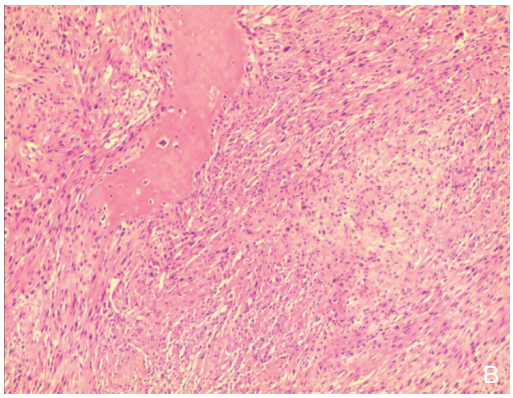


图 6 术后病理 A：大体病理标本；B：组织病理学示平滑肌肉瘤，伴大面积坏死、囊性变及局部钙化，核分裂象15个/10 HPF（HE ×100）

Figure 6 Postoperative pathology A: Gross pathological specimen; B: Histopathological result demonstrating leiomyosarcoma with extensive necrosis, cystic degeneration and partial calcification and mitotic count of 15/10 HPF (HE ×100)

2 讨论并文献复习

腹膜后肉瘤是一种罕见的恶性肿瘤，发病率约占软组织肉瘤的15%[3]，年均发病率为2.7例/百万人口[4]，其中腹膜后平滑肌肉瘤发病率仅为腹膜后肉瘤的15%~30%[5-9]。腹膜后平滑肌肉瘤是一种发生于腹膜后软组织的恶性肿瘤，其生长隐匿常无明显症状，多偶然发现，主要表现为巨大肿块，由于缺乏典型的临床特征，主要表现为肿物压迫引起的腹痛、腹胀，部分患者因肿瘤压迫或侵袭邻近器官可出现便频、血尿、下肢水肿及肌肉萎缩等症状，且肿瘤位于腹膜后，解剖位置深，局部空间较大，诊断时多为巨大肿瘤。Lewis等[10]报道500例确诊腹膜后平滑肌肉瘤的患者，

70%直径>10 cm，23%直径在5~10 cm之间，而以破裂出血为首要表现的则较为罕见，病情凶险。腹膜后平滑肌肉瘤病情凶险，易侵袭、复发及转移，预后较差，在所有软组织肿瘤中生存率最低[11]。

随着目前影像技术的飞速发展，腹膜后平滑肌肉瘤的诊断方法越来越多，其中CT、MRI及超声技术被临床广泛应用[12]。CT是术前诊断对重要的检查方法。典型的CT表现包括腹膜后肿块，形态呈不规则椭圆形或分叶状，密度接近肌肉组织密度，大部分肿瘤内部密度不均匀，囊性变性较多，可伴坏死等，钙化及出血罕见；多有明确的包膜，界限多较清，出现临近器官侵袭时界限不明显；增强扫描可见明显强化；静脉期强化减弱，有明显延迟强化的特点[13]。MRI具有较高的软组织分辨率，对于有静脉造影禁忌证的患者，MRI可作为评估大血管浸润程度的首选。超声通常表现为较低回声、边界清晰的团块，团块内部可见血流信号[14]。超声检查定位准确度较高，能识别肿瘤对周围组织器官的浸润几压迫，对手术方式的选择具有指导意义。

病理学诊断是金标准。但为防止肿瘤的扩散和针头植入，不建议常规进行术前活检。对于不能手术，需要放疗或化疗的患者，可进行穿刺行病理学检查。

有文献[3, 15-16]报道，腹膜后平滑肌肉瘤术后仍有较高的远处转移率，但局部复发率较低。Gladdy等[17]对353例首次手术的LMS患者进行研究发现，腹部/腹膜后平滑肌肉瘤5年远处复发率为43%，其中肺是最常见的复发器官，其次是肝脏。多因素分析发现肿瘤>10 cm是局部复发和远处转移的危险因素。

疑似腹膜后肿瘤破裂出血者，应迅速进行液体复苏，保证重要器官的灌注，警惕腹腔内液体积聚引起的腹间隔室综合征，在血流动力学稳定情况下进行DSA造影，尽可能的寻找供血动脉，进行彻底栓塞以减小肿瘤体积，降低手术风险。由于腹膜后肿瘤毗邻脏器较多，常伴有重要血管的侵犯[18]，由于术区暴露欠佳，易造成血管损伤，而选择性栓塞肿瘤滋养动脉，可减少术中出血量，保证手术安全。栓塞完成后待血红蛋白稳定，进行腹腔内、肿物内的穿刺引流，降低腹腔内压力，改善重要脏器的血供，为进一步手术提供了操作空间。

手术完整切除肿瘤是腹膜后平滑肌肉瘤治疗的关键，也是预测局部复发和转移重要的因素[19]。然而肿瘤生长快速，常常累及重要的神经、血管，难以从周围组织中分离，甚至出现腹腔内及远处转移。腹膜后平滑肌肉瘤出现远处转移后其生存期相比与其他肉瘤更长，因此对于出现远处转移的患者，为进一步减少并发症，提高患者生存质量，手术治疗仍是必要的[20]。Gronchi等[21]报道腹膜后肿瘤完整切除率为54%~88%。累及大动脉的肿瘤尽量沿血管壁剥离肿瘤，而肿瘤局限侵犯大动脉，能完整切除者，可行大血管壁的切除并修复重建。而下腔静脉和髂总静脉受累时，首选沿血管壁剥离，如不能分开时可做静脉壁切除或静脉段的切除及血管修补或重建。股神经来自腰2~4，腰丛各支中最粗者，在髂凹内行走于腰大肌与髂腰肌之间。术中如遇股神经受累者，可沿其外膜进行分离，必要时可以劈开肿瘤。原本生存期较短的生物学特性决定如再丧失1条腿的运动及感觉功能得不偿失。但无论是何种情况，都建议积极的手术探查，只有探查才能最终确定是否可以完整切除。

笔者汇报1例巨大腹膜后巨大平滑肌肉瘤伴出血的病例的治疗过程，为类似病例的后续治疗提供一定的参考。早期的液体复苏，进一步的介入栓塞，超声引导下穿刺引流，避免腹腔隔室综合征的发生，而对于累及重要血管及神经者，仍建议积极手术探查，最终通过手术确认能否达到完整切除。

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