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Dedifferentiated liposarcoma (DDLPS) in the rectum: A case report

Wei Li, Qi Liu, Yu Mu, Kai Zhang and Tongjun Liu

Abstract

Dedifferentiated liposarcoma (DDLPS) is a rare subtype of liposarcoma with a poor prognosis. This current case report describes a rectal DDLPS in a 68-year-old Chinese male that presented with lower abdominal pain and weight loss. Computed tomography and magnetic resonance imaging were undertaken to evaluate the tumour. The patient underwent radical resection of the rectal tumour, sigmoid colostomy and partial ureterectomy. The tumour was positive for mouse double minute 2 by immunohistochemistry. The patient healed well but refused chemotherapy postoperatively for economic reasons. The tumour recurred and metastasized 4 weeks after the operation. After relevant treatment, the patient's condition deteriorated and he died of shock, metabolic acidosis, hyperlactataemia and acute renal failure. The case report also reviews the literature in terms of the clinical diagnosis, treatment and pathological characteristics of rectal DDLPS with the aim of improving the level of diagnosis and treatment.

Keywords

Dedifferentiated liposarcoma, rectum, management, surgery, case report

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Introduction

Liposarcoma (LPS) is a type of soft tissue sarcoma that is made up of adipose-derived cells with varying degrees of differentiation and atypia.[1] It accounts for approximately 20% of all soft tissue sarcomas.[2] The extremities (especially the thighs) and buttocks are the most common sites for LPS,

Department of Colorectal and Anal Surgery, The Second Hospital of Jilin University, Changchun, Jilin Province, China

Corresponding author:
Tongjun Liu, Department of Colorectal and Anal Surgery, The Second Hospital of Jilin University, 218 Ziqiang Street, Changchun, Jilin 130000, China.
Email: tongjunliu@163.com

but they can also appear in the retroperitoneal space.[1] Primary LPS is less common in the abdominal cavity and primary LPS of the rectum is even rarer.[3] Dedifferentiated LPS (DDLPS) is a distinct subtype of LPS that shows a morphological transition from the atypical lipomatous tumour (ALT) or well-differentiated LPS (WDL) to high-grade sarcoma.[4] Surgical resection is the most effective treatment, while radiotherapy and chemotherapy have ineffective curative effects.[3] More effective targeted therapies are required.

This current case report describes a patient with an aggressive giant DDLPS of the rectum. Although there are no guidelines for the treatment of rectal LPS, a previous report described in detail the systemic therapeutic options available.[5]Radical surgical resection along with systemic treatment is almost certainly required.

Case report

In July 2020, a 68-year-old male from northeast China presented to the Department of Colorectal and Anal Surgery, The Second Hospital of Jilin University, Changchun, Jilin Province, China with 6-month history of lower abdominal intermittent colic and weight loss of 5 kg. The pain had worsened in the previous week and the frequency of bowel movements had increased. To be more specific, he defecated more than 10 times per day and there was mitigation of the pain after each defecation. He had already undergone a computed tomography (CT) scan in Meihekou Central Hospital, Meihekou, Jilin Province, China, which identified an exogenous rectal mass with a diameter of up to 4 cm. The tumour was located on the left side of the middle section of the rectum. Moreover, signs of invasion of the peritoneal inflection and the left seminal vesicle gland were found. A digital rectal examination at a distance of 4 cm from the anus revealed an externally pressurized neoplasm.

The patient was admitted to the Department of Colorectal and Anal Surgery, The Second Hospital of Jilin University for further clarification of the diagnosis and to determine more information about the tumour. The patient underwent a colonoscopy and an enhanced magnetic resonance imaging (MRI) scan of the rectum. A raised lesion on the left rectal wall approximately 5 cm from the anus (Figure 1a) was observed during the colonoscopy. Its long diameter was approximately 4 cm and its short diameter was approximately 2 cm. The lesion was hard and immovable. The mucosa of the tumour was smooth and its colour was the same as the surrounding mucosa. Enhanced MRI scans suggested a rectal stromal tumour (Figures 1b–1f). The left side of the middle rectum was isointense on T1-weighted images and moderately hyperintense on T2-weighted images (Figures 1b and 1d). Diffusion-weighted imaging (DWI) revealed a strong signal, while the enhanced scan revealed uneven enhancement (Figure 1c). The tumour, with a maximum diameter of 48 mm, was located in the middle-left portion of the rectum, 34 mm from the anorectal ring. There was no clear distinction between the left seminal vesicle gland and the tumour (Figures 1e and 1f). There was no evidence of pelvic or other organ metastases and no swollen or suspected metastatic lymph nodes were found on MRI.

Laparoscopic surgery was undertaken after the reviewing the results of the MRI examinations. This revealed an irregular-shaped tumour in the rectum at the peritoneal fold, whose left part was closely related to the combination of the bladder and the ureter as a suspected invasion. As a result of the intraoperative diagnosis of rectal malignant tumour, radical resection of the rectal tumour, sigmoid colostomy and partial

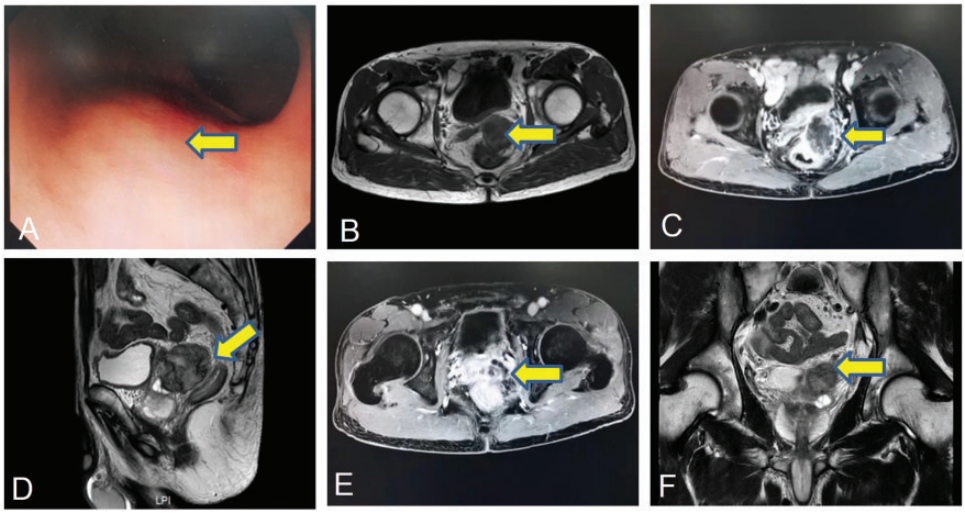

Figure 1. Imaging examinations of a 68-year-old male that presented with a 6-month history of lower abdominal intermittent colic and weight loss of 5 kg: (a) colonoscopy images of the tumour; (b) T1-weighted magnetic resonance imaging (MRI) in the axial position; (c) enhanced MRI in the axial position; (d) T2-weighted MRI in the sagittal position; (e) enhanced MRI in the axial position demonstrating invasion of the left seminal vesicle and (f) enhanced MRI in the coronal position demonstrating invasion of the left seminal vesicle. The yellow arrows show the location of the tumour. The colour version of this figure is available at: http://imr.sagepub.com.

ureterectomy were performed. After the mass was completely unshackled and excised, the specimen was sent for rapid pathological examination, which revealed that it was a rectal stromal tumour. It was finally identified as a DDLPS by pathological examination with positive or focally positive immunohistochemical staining for H-caldesmon, desmin, Ki67, transcription factor binding to IGHM enhancer 3 (TFE3), cyclin-dependent kinase 4 (CDK4) and mouse double minute 2 (MDM2) (Figure 2). Notably, metastasis was discovered in one of the 16 lymph nodes around the mesenteric artery. The patient healed well but refused chemotherapy postoperatively for economic reasons. Although his physicians attempted to persuade him to undertake appropriate systemic treatment, he was satisfied that the surgery had resolved his complex and rare disease and asked to be discharged.

The patient returned to the emergency department of The Second Hospital of Jilin University 4 weeks after his discharge from the hospital complaining of intestinal blockage for 1 week. After 17 days of conservative therapy, no substantial improvement was seen with the intestinal obstruction catheter. Acute renal failure and hyperkalaemia were observed 1 day after the catheter was used and the patient began dialysis. After 2 days of dialysis, an emergency exploratory laparotomy was performed and widespread intraabdominal metastases were discovered. The peritoneal omental, visceral wall was evident, with dark red, grey nodules dispersed throughout. A massive tumour with an uneven form that was brittle and bleeding was fixed in the pelvic cavity and adjacent anatomical systems were invaded. The small intestine in the pelvic cavity expanded in a closed loop showing evidence of intestinal

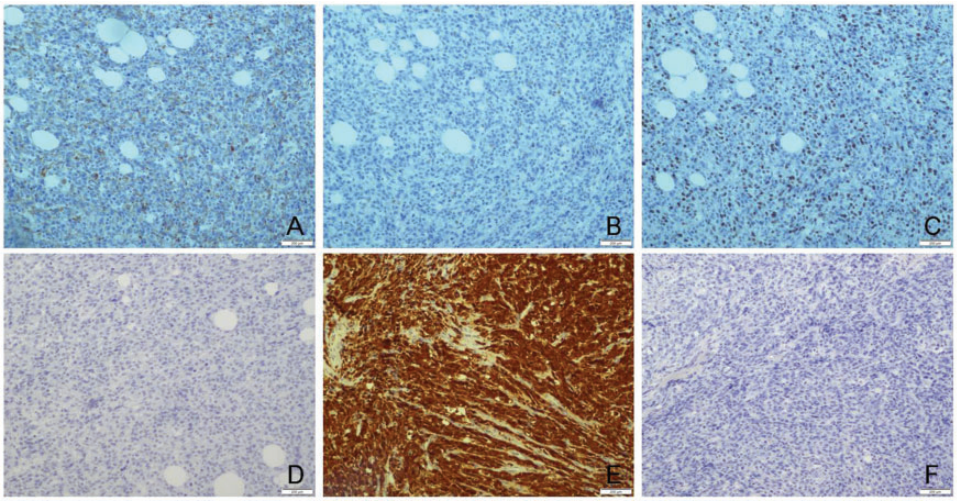

Figure 2. Representative photomicrograph images of tumour specimens from a 68-year-old male that presented with a 6-month history of lower abdominal intermittent colic and weight loss of 5 kg showing positive immunohistochemical staining for the following proteins: (a) H-caldesmon; (b) desmin; (c) Ki67; (d) transcription factor binding to IGHM enhancer 3 (TFE3); (e) cyclin-dependent kinase 4 (CDK4) and (f) mouse double minute 2 (MDM2). The colour version of this figure is available at: http://imr.sagepub.com. Scale bar 200 μm.

wall oedema. There was weak peristalsis and the necrotic bowl was dark purple in colour. The pelvic mass invaded the distal jejunum 60 cm distant from the Treitz ligament and the intestine was narrow. In the lumen proximal to the invaded intestine, the water sac of the intestinal obstruction catheter could be touched. The intestinal portion of the small intestine continued to the pelvic cavity at 50 cm from the distal side of the stenosis and the local anatomical connection could not be determined. After partial intestine resection and anastomosis, the obstruction was relieved. The patient was transferred to the intensive care unit for mechanical ventilation and blood purification after the operation. After 2 days of relevant treatment, the patient's condition deteriorated and he died of shock, metabolic acidosis, hyperlactic acidaemia and acute renal failure.

The study protocol was approved by The Ethics Committee of the Second Hospital of Jilin University (no. 2022064). The patient in this case report provided verbal informed consent for publication of this report. The reporting of this case conforms to CARE guidelines.[6]

Discussion

The term 'dedifferentiation' was first used in the medical literature in 1971 and it was applied to the progression of low-grade sarcomas to high-grade sarcomas, including low-grade osteosarcoma, chondrosarcoma and fibrosarcoma.[7] In 1979, the notion of dedifferentiation was further expanded to include LPS.[8] DDLPS accounts for approximately 20% of all LPS and it is typically characterized as the conversion of ALT/WDL to non-fatty sarcoma components.[2] Occasionally, the dedifferentiated and transformed components can be homologous pleomorphic LPS.[9] DDLPS, which is seldom observed in children and teenagers,

is more common in middle-aged and elderly individuals without a preference for a particular sex.[2] Dedifferentiation of ALT/WDL can occur in any area of the body and the proportion of dedifferentiation is approximately 10%.[1] DDLPS is characterized by an aggressive clinical behaviour with a high capacity for local recurrence and metastasis.[2]

Most DDLPS tumours are located in the space between the posterior peritoneum and the soft tissue of the pelvis.[2] The extremities, as well as the spermatic cord or paratesticular region, are typical locations. [2] The thoracic cavity, mediastinum and head and neck are among the uncommon sites (such as the larynx or oesophagus).[5] In this current case, the patient was a 68-year-old Chinese male and the tumour was located in the rectum. There have been just a few instances of this tumour type in the rectum. For example, a previous report described a case of rectal DDLPS that was treated with Hartmann's operation in 2012.[10] Most DDLPS occur in the retroperitoneum and extremities, with other anatomical sites occasionally reported, such as the small bowel mesentery, the colon and the sigmoid mesocolon.[10] Notably, a previous report described a 75-year-old male with a DDLPS originating from the descending colon.[11]

In terms of symptoms, the presentation of DDLPS is usually abdominal pain or the syndromes generated from pressure on the surrounding tissues.[2]However, LPS in the retroperitoneal area of the pelvis may grow for a long period without causing any symptoms. In this current case, due to the impact on the rectum, the patient had experienced intermittent colic in the lower abdomen for 6 months and had lost 5 kg in weight. Such symptoms are quite common and unspecific. As a result, several diagnostic methods are needed to identify the tumour.

A variety of clinical examinations can be undertaken to diagnose DDLPS. LPS has heterogeneous, solid and hypoechoic ultrasound manifestations.[12] LPS should be suspected when the tumour has a heterogeneous structure and a relatively low echo of blood vessels.[13] In addition to ultrasonography, CT and MRI can differentiate fat from other soft tissue components with higher precision.[13] Ultrasound imaging is less helpful for the diagnosis of LPS than a CT scan.[13] MRI gives accurate information about the tumour position, but it cannot completely identify the type of tumour. In this current case, the tumour was isointense on T1-weighted images mixed with mildly hyperintense on T2-weighted images. DWI showed a high signal and the enhanced scan showed uneven enhancement. Positron emission tomography-CT scans are also useful in cases of recurrence. Imaging examination can be used to determine the extent of the tumour and the infiltration of surrounding organs, but the final diagnosis depends on pathological examination.[12,13] The surgeon needs to prepare to undertake a radical resection as the imaging examinations might not show the full extent of any invasions. The tumour was quite complex in this current case. The preoperative CT and MRI scan findings suggested tumour invasion in the left seminal vesicle gland, but the ureter was also found to be involved during surgery.

The histology of DDLPS often demonstrates ALT/WDL components converted into non-adipose-derived tumour components. When viewed under a microscope, the two components are generally clearly separated, with a sharp transition between the two. The amplification and overexpression of the 12q14-15 region is consistent with increased levels of MDM2 and CDK4 proteins, so the immunohistochemical staining of MDM2 and CDK4 (both show diffuse nuclear staining) is extremely

useful in the diagnosis of DDLPS.[2] MDM2, CDK4 and p16 are found in both well-differentiated and dedifferentiated LPS components.[2] The sensitivity of MDM2 and p16 levels is quite high, but they lack specificity and may be found in several different high-grade sarcomas.[14]When it comes to diagnosing DDLPS, the use of MDM2, CDK4 and p16 together can improve specificity.[14] MDM2 is an oncogene that promotes the degradation of the tumour suppressor protein p53 and it is regarded as the most important gene in the tumorigenesis of WD/DDLPS because it is amplified and expressed in almost all DDLPS cases.[14]

Surgical resection is widely recognized as the basic treatment modality for DDLPS.[2,11] There is disagreement over the systemic treatment for DDLPS since there is no clear consensus. However, studies have indicated that systemic treatment coupled with surgery can decrease the chance of recurrence in DDLPS patients.[5,15] Systemic treatment options have traditionally been restricted to cytotoxic chemotherapy drugs such as doxorubicin, ifosfamide, gemcitabine and docetaxel, which have been demonstrated to be effective in unselected groups of patients with soft tissue sarcomas.[5] Recently, the tyrosine kinase inhibitor pazopanib has been shown to provide clinical benefit for patients with DDLPS.[5] Moreover, potential targeted therapies such as anti-CDK4 therapy, anti-MDM2 therapy and the checkpoint inhibitors require more research.[5]

In conclusion, this current case report describes a patient with an aggressive giant DDLPS of the rectum. It also reviews the literature in terms of the clinical diagnosis, treatment and pathological characteristics of rectal DDLPS with the aim of improving the level of diagnosis and treatment.

Declaration of conflicting interest

The authors declare that there are no conflicts of interest.

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ORCID iD

Tongjun Liu https://orcid.org/0000-0001-5712-4838

References

1. Weiss SW and Rao VK. Well-differentiated liposarcoma (atypicallipoma) of deep soft tissue of the extremities, retroperitoneum, and miscellaneoussites. A follow-up study of 92 cases with analysis of the incidence of "dedifferentiation". *Am J Surg Patho* 1992; 16: 1051–1058.
2. Thway K. Well-differentiated liposarcoma and dedifferentiated liposarcoma: An updated review. *Semin Diagn Pathol* 2019; 36: 112–121.
3. Mastrangelo G, Coindre JM, Ducimetière F, et al. Incidence of soft tissue sarcoma and beyond: a population-based prospective study in 3 European regions. *Cancer* 2012; 118: 5339–5348.
4. Jo VY and Doyle LA. Refinements in Sarcoma Classification in the Current 2013 World Health Organization Classification of Tumours of Soft Tissue and Bone. *Surg Oncol Clin N Am* 2016; 25: 621–643.
5. Gahvari Z and Parkes A. Dedifferentiated Liposarcoma: Systemic Therapy Options. *Curr Treat Options Oncol* 2020; 21: 15.

6. Gagnier JJ, Kienle G, Altman DG, et al. The CARE guidelines: consensus-based clinical case reporting guideline development. *Headache* 2013; 53: 1541–1547.
7. Dahlin DC and Beabout JW. Dedifferentiation of low-grade chondrosarcomas. *Cancer* 1971; 28: 461–466.
8. Evans HL. Liposarcoma: a study of 55 cases with a reassessment of its classification. *Am J Surg Pathol* 1979; 3: 507–523.
9. Mariño-Enríquez A, Fletcher CD, Dal Cin P, et al. Dedifferentiated liposarcoma with "homologous" lipoblastic (pleomorphic liposarcoma-like) differentiation: clinicopathologic and molecular analysis of a series suggesting revised diagnostic criteria. *Am J Surg Pathol* 2010; 34: 1122–1131.
10. Tsuruta A, Notohara K, Park T, et al. Dedifferentiated liposarcoma of the rectum: a case report. *World J Gastroenterol* 2012; 18: 5979–5981.
11. Serafini L, Lauro A, Eusebi LH, et al. Dedifferentiated Liposarcoma of the Descending Colon: A Case Report and Review of the Literature. *Dig Dis Sci* 2020; 65: 1643–1651.
12. Li Z, Zhou L, Zhao L, et al. Giant paratesticular liposarcoma: A case report and review of the literature. *Mol Clin Oncol* 2018; 8: 613–616.
13. Patel NG. Rajagopalan A and Shrotri NS. Scrotal liposarcoma – a rare extratesticular tumour. *JRSM Short Rep* 2011; 2: 93.
14. Yamashita K, Kohashi K, Yamada Y, et al. Prognostic significance of the MDM2/HMGA2 ratio and histological tumor grade in dedifferentiated liposarcoma. *Genes Chromosomes Cancer* 2021; 60: 26–37.
15. Yamamichi G, Nakata W, Yamamoto A, et al. Liposarcoma of the spermatic cord associated with scrotum lipoma: A case report and review of the literature. *Urol Case Rep* 2018; 17: 114–116.