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Dedifferentiated Liposarcoma of Sigmoid Colon: A Case Report

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Case report

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Abstract

BACKGROUND: Primary liposarcoma of the colon with obstruction is a rare tumor, and patients with dedifferentiated liposarcoma of the sigmoid colon have not been reported in Pubmed for decades.

CASE PRESENTATION: We reported that a 75-year-old female patient was admitted to the hospital due to abdominal pain and bloating. The imaging examination revealed that the sigmoid colon liposarcoma was considered malignant. She was treated with pelvic mass resection + partial colectomy + intestinal fistula. Postoperative pathological immunohistochemical prompts Differentiated sigmoid colon liposarcoma.

CONCLUSIONS: We reported a rare case of dedifferentiated liposarcoma of the sigmoid colon, which is easily confused with atypical lipomatous tumors. Through this case, we hope to provide a reference for the diagnosis and treatment of dedifferentiated liposarcoma of the sigmoid colon.

Background

Soft tissue sarcoma (STS) arises from bone or extra-skeletal connective tissue in the limbs, retroperitoneum, head and neck, and subcutaneous tissue. Liposarcoma is the most common single soft tissue sarcoma, accounting for at least 20% of all sarcomas and more than 50% of retroperitoneal sarcomas[1].

Liposarcoma (LPS) is a group of malignant tumors composed of adipocytes with different degrees of differentiation and malignancy. The incidence is about 1% of all malignant tumors[2].Primary liposarcoma of the colon with obstruction is a rare tumor, The most common site of primary colonic sarcoma is the ascending colon and its mesenteric, followed by the rectum-sigmoid colon, and the transverse colon is rare. A total of 28 articles with "liposarcoma" and "colon" in the literature published from January 1980 to January 2020 were searched through Pubmed, including 4 cases of sigmoid colorectal[3,4,5,6].Only 4 cases suggest dedifferentiated liposarcoma,but not reported in line with dedifferentiated liposarcoma of sigmoid colon[7,8,9,10].We report a 75-year-old woman with dedifferentiated liposarcoma of the sigmoid colon.

Case Presentation

A 75-year-old woman was admitted at the Affiliated Hospital of Qinghai University because of "abdominal distension for 3 months, aggravated with abdominal pain for 1 week". Upon admission, the blood pressure was 150/80 mmHg, pulse rate was 68 bpm, and the SpO2 96%. The patient had a history of abdominal pain for 3 months, which was distended in nature and had intermittent attacks. The above symptoms were significantly worsened 7 days ago, accompanied by difficulty in eating. The color Doppler ultrasound on the abdomen in our hospital showed that the right iliac fossa mixed echo mass. No significant weight gain or loss during the disease.

Physical examination showed that the abdomen is distended, the lower abdomen can be palpable with a mass of about 10 cm in diameter, no abdominal muscle tension, light tenderness in the whole abdomen, obvious at the lower abdomen, but no rebound pain. The patient underwent a total hysterectomy 9 years ago and thyroidectomy 8 years ago both in our hospital, and the past medical history was normal. The patient denied any family history of tumors. Admission blood tests Included WBC12.69 × 109/L,Percentage of neutrophils 86.9%,Lymphocyte 0.63 × 109/L,Albumin 31.8 g/L,Glucose 10.2 mmol/L.Pelvic CT + MRI showed an Massive mixed-density foci at the entrance of the pelvis seems to be a liposarcoma;Fatty lesions in the lower abdomen and pelvis are considered malignant, and liposarcoma may invade the sigmoid colon (Fig. 1a,b,c).

After excluding relevant surgical contraindications, Surgery which included pelvic mass resection + partial colectomy + enterotomy under general anesthesia was performed on August 17, 2020. During the operation, the peritoneum and omentum are swollen, pelvic cavity closed, separated adhesions, a lump about 15 × 10 × 10 cm3 in size can be touched, fixed on the pelvic floor, containing about 500 ml of purulent fluid, unclear borders, adhesion with the posterior peritoneum, rectum, and part of the sigmoid colon close(Fig. 2a). When separate the adhesion between the intestine and the mass, the sigmoid colon appears to be ruptured, then carefully separate the adhesions, free the sigmoid colon, clamp and cut the purse string, end-to-end stapler anastomosis to the distal end, remove the ruptured intestinal tube, use a cutting stapler to make the left abdominal wall at the proximal end Ostomy (Fig. 2b).Use the ultrasonic scalpel to separates the adhesions along the edge of the tumor and removes the tumor completely (Fig. 2c).

Histopathology showed: macroscopic view: a pile of grayish pink irregularly shredded tissue, with a total volume of 18.5 × 14.5 × 7.0 cm. The multi-faceted incision and the cut surface has Fine texture. A section of the excised intestine is 6.5 cm long, Excise a section of intestine with a length of 6.5 cm, with circumferences of 2.5 cm and 2.0 cm at both ends.no obvious masses and nodules were seen in the intestinal mucosa, and a 5 × 3 × 2 cm hard zone was seen on the serosal surface of the intestine. Pathological diagnosis: Combined with immunohistochemistry, it is consistent with dedifferentiated liposarcoma with extensive necrosis; chronic inflammation of colonic mucosa, tumor invasion can be seen under the serous membrane. Immunohistochemical results: S100 (-), STAT6 (-), SMA (-), Des (partially weak +), CD34 (weak +), CD31 (vascular +), Fli-1 (partial +), Ki67 (50%), P53 (-), CDK4 (+), CD117 (-), Dog-1 (-), ER (-), MDM2 (+) (Fig. 4).

This disease needs to be differentiated from gastrointestinal stromal tumor and angiosarcoma. Immunohistochemical examination: immunohistochemical staining showed positive reaction of MDM2, CDK4, Ki67 (Fig. 4). This result confirmed the histological diagnosis of dedifferentiated liposarcoma.

Discussion

Liposarcoma is less common than other types of tumors, and the incidence is only 1% of malignant tumors[2].The latest classification of the World Health Organization divides liposarcoma into five

categories: (1) well-differentiated or atypical lipomas, including adipocytes, sclerosis and inflammatory subtypes; (2) mucinous; (3) highly differentiated; (4) Polymorphism; (5) Dedifferentiation[11].Among them, dedifferentiated subtypes are less common than other types. Dedifferentiated liposarcoma is characterized by histological coexistence in differentiated areas of well-differentiated liposarcoma and non-lipoma[12].Fluorescence in situ hybridization (FISH) and immunohistochemistry can both show the amplification of CDK4 and MDM2 genes on chromosome 12[13,14], among which CDK4 is a member of the cyclin D kinase family and is involved in cell cycle regulation[15,16], Both provide growth advantages for tumor cells. Positive immunohistochemical staining helps differentiate dedifferentiated liposarcoma from other types of liposarcoma [17,18,19],About 90% of dedifferentiated liposarcoma of the colon shows the oncogene MDM2 Overexpression[20], postoperative immunohistochemistry in this patient also showed MDM2 (+), CKD4 (+).

The clinical manifestations of colon liposarcoma are very different, but the common points are abdominal pain and bloating. Some reports show that colon liposarcoma also manifests as abdominal masses, gastrointestinal bleeding, and intussusception. The patient has no obvious symptoms in the early stage, and it is usually difficult to diagnose. The patient's condition is usually serious when clinical symptoms appear as the tumor enlarges and compresses adjacent organs. According to our report, the case we describe is the first case reported in the literature related to partial intestinal obstruction. The patient had no symptoms in the early stage, and only went to the doctor when abdominal pain and bloating occurred as the enlarged mass pressed the intestine. Colon liposarcoma usually originates in the serosal and subserous layers of the colon, or in the surrounding fatty tissue, and then they form masses of completely different sizes and weights. According to reports, the size of the dedifferentiated liposarcoma is between 2 and 80 cm. The size of the tumor in this patient is 15 × 10 × 10 cm.

Abdominal X-ray, ultrasound, CT and MRI are most commonly used to diagnose colon liposarcoma radiology. On CT, adipose tissue (-97HU), soft tissue (32HU) and tissues with high edge density can be observed. On MRI, adipose tissue, soft tissue, and fibrous cords can be observed in dedifferentiated liposarcoma. Consider the undifferentiated high-grade type of dedifferentiated liposarcoma as fibrosarcoma. The signal strength on the T1-weighted image is from low to medium, and the signal strength on the T2-weighted image is from medium to high. Biopsy is the most reliable way to diagnose retroperitoneal tumors.

Due to the few reported colonic liposarcoma in the literature, a 2017 literature review [21] concluded that radical resection of the tumor is still considered the first-line treatment for all retroperitoneal liposarcoma. Thorough surgical resection is an important factor affecting the prognostic survival rate and recurrence rate [22]. However, in many cases, the resectability is limited due to the tumor's size or the invasion of surrounding organs or adhesion to large blood vessels, and the average complete clearance rate is 50% [23]. For poorly differentiated sarcomas with a mass larger than 10 cm or incompletely resected cases, radiotherapy or other chemotherapy can be considered. Although the value of perioperative radiotherapy is still controversial, emerging evidence supports its role in the treatment of retroperitoneal sarcoma [24]. In

addition, doxorubicin-based adjuvant chemotherapy has shown marginal efficacy in local resection, distant recurrence, overall recurrence, and overall survival in locally resectable STS. The addition of ifosfamide to the doxorubicin-based regimen further improves these benefits [25].

Many variables are considered to be prognostic factors for survival of primary retroperitoneal liposarcoma. Compared with well-differentiated histology, the dedifferentiated liposarcoma subtype is associated with a six-fold increase in the risk of death ($p < 0.0001$). The 3-year local recurrence rate of retroperitoneal dedifferentiated liposarcoma is 83%, and the distant recurrence rate is 30% [26].In addition, tumor size (5–15 cm vs ≤ 5 cm, $p = 0.03$; or > 15 cm vs ≤ 5 cm, $p < 0.0001$), non-acral primary site ($p = 0.0016$) and histological high disease grade ($p = 0.001$) was identified as a statistically significant poor independent prognostic factor, and recurrence during follow-up was the highest risk factor for STS-specific mortality ($p < 0.0001$) [27].Positive microsurgical margins increase the risk of local recurrence, distant recurrence, and disease-related death: since 72% of patients with positive margins have no recurrence, other treatments after tumor removal should be carefully considered in these patients [28].After definite treatment, the median time to recurrence of high-grade retroperitoneal sarcoma is less than 5 years [29]. In our case, the dedifferentiated liposarcoma has been completely removed, and the surgical margin is disease-free under the microscope.

Conclusion

In short, although sigmoid colon liposarcoma is rare, it should be considered in the differential diagnosis in the case of partial sigmoid colon obstruction. Surgical treatment is still the first-line treatment for retroperitoneal liposarcoma, because of its extremely high postoperative recurrence rate. Surgical resection needs to be as precise and thorough as possible. If necessary, this treatment should be combined with radiotherapy and chemotherapy. We hope that the medical record report can provide clinicians with new views on the diagnosis and treatment of this rare disease.

Declarations

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Additional Information

Declaration of Competing Interest

The authors declare no conflicts of interest.

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Ethical approval

Ethical approval was not required in the treatment of the patient in this report.

Consent

Written consent has been received from the subject.

Availability of data and materials

The datasets used and/or analyzed during the current study are available from the corresponding author on reasonable request.

Consent for publication

Written informed consent for publication was obtained from the participant.

Author Contributions

Xuepeng Mei and Xiaobin Chen contributed equally to this work. Xuepeng Mei and Xiaobin Chen: study design, data collection, analysis, and manuscript writing. Ying Zhou: data collection and manuscript review. Haining Fan,Zhixin Wang,Yichong Haijiu Wang: data collection and analysis.

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Figures

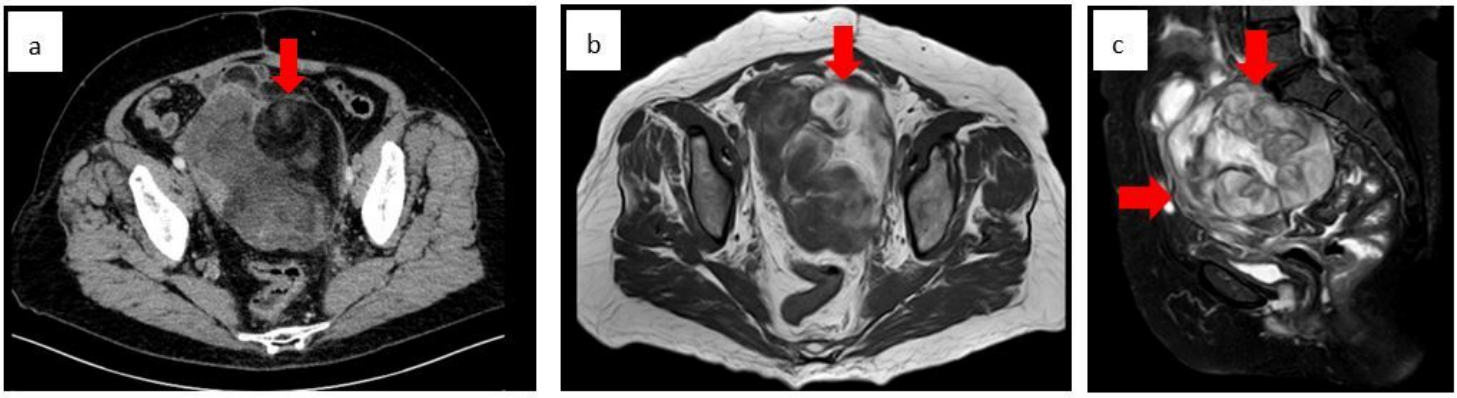


Figure 1

Pelvic CT scan,coronal plane (Fig. 1a) showing a massive mixed density foci at the entrance of the pelvis(red arrows).Pelvic MRI scan,coronal (Fig. 1b) and axial plane (Fig. 1c) showing an 12 cm mass located in the Sigmoid colon (red arrow).



Figure 2

During the operation, the mass was closely adhered to the posterior peritoneum, rectum, and part of the sigmoid colon(Fig. 2a),remove the ruptured intestinal tube, use a cutting stapler to make the left abdominal wall at the proximal end Ostomy(Fig. 2b),complete removal of liposarcoma invading the sigmoid colon area(Fig. 2c).

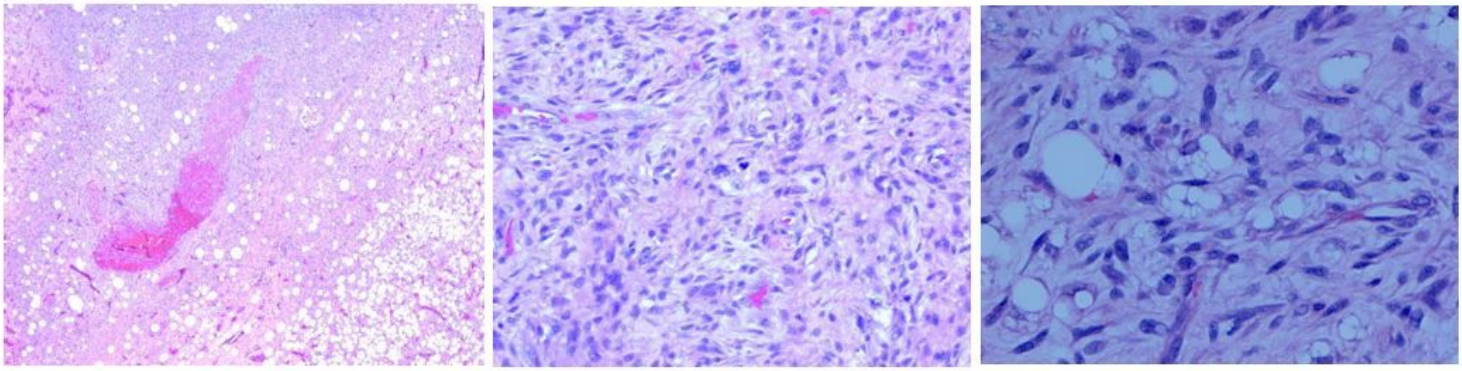


Figure 3

The boundary between the liposarcoma area and the dedifferentiated area is relatively clear (Fig. 3A) (H&E×20). A large number of spindle cells can be seen in the dedifferentiated area (Fig. 3B)(H&E×200). Adipocytes visible under the microscope (Fig. 3C)(H&E×400) (red arrow).

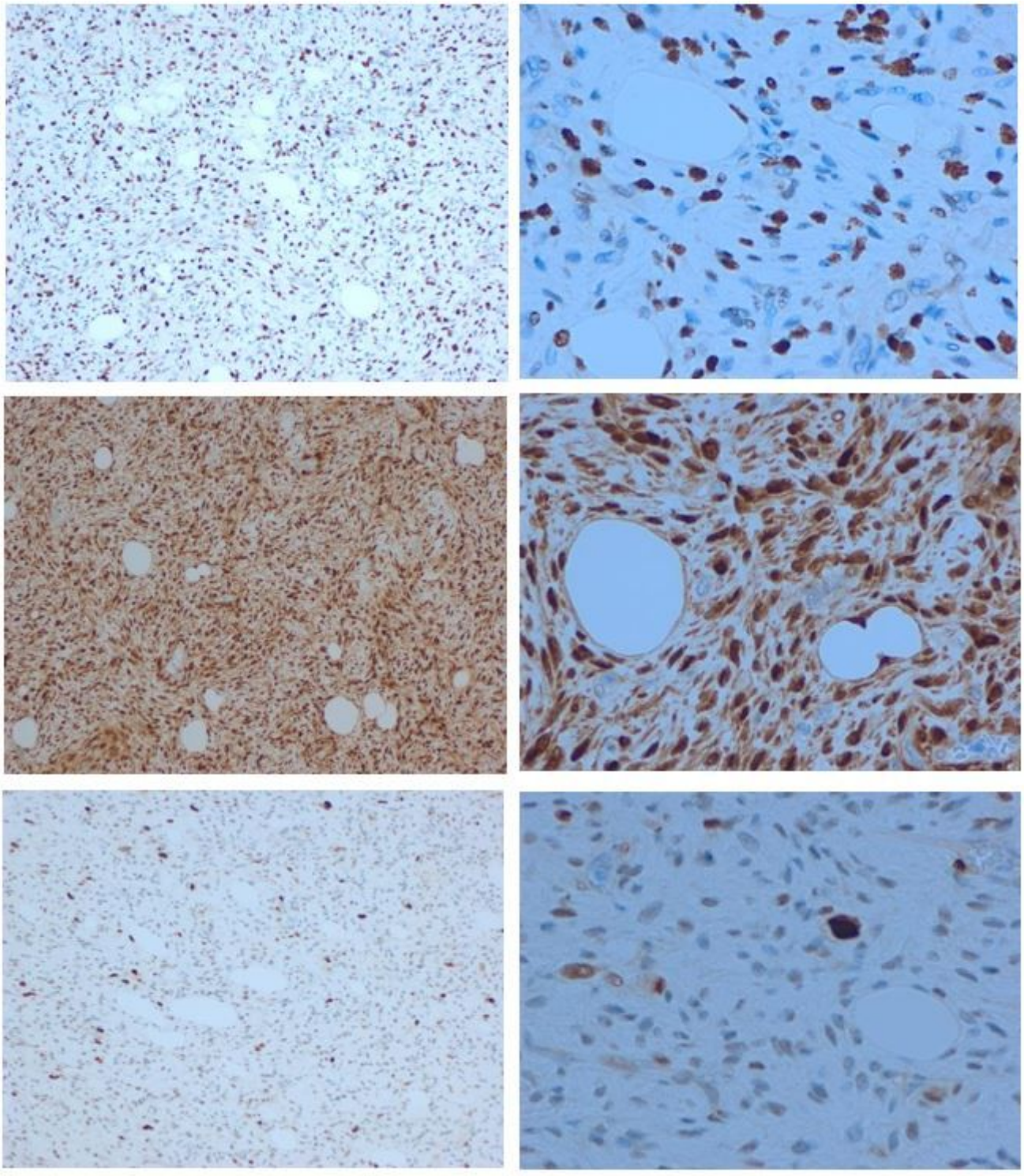


Figure 4

Immunohistochemical examination: immunohistochemical staining showed positive reaction of MDM2, CDK4, Ki67.

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