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CASE REPORT

Recurrent Adult Intraabdominal Undifferentiated High-Grade Pleomorphic Sarcoma Infiltrated the Descending Colon: a Case Report and Review of the Literature

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Introduction

Undifferentiated pleomorphic sarcoma (UPS), which was previously known as malignant fibrous histiocytoma (MFH), was first described in 1961 by Kauffman as a histiocytic tumor with storiform growth in children [1]. On the new 2013 WHO classification of soft-tissue tumors, a new class of "undifferentiated/unclassified sarcoma" was created, with the deletion of MFH replaced by UPS and attributed to this category. UPS is a type of malignant sarcoma that occurs most frequently in patients aged between 50 and 70 years [2], accounting for 28% of all soft-tissue sarcomas [3]. The major locations for UPS are the lower (49%) and upper extremities (19%), while tumors of the retroperitoneum and abdominal cavity account for 16%, and those of trunk account for 9% of the cases [4]. The histological morphology varies, but the classic form is composed of spindle-shaped and round cells. The prognosis for UPS is very poor because of its strong chances of regional invasiveness and distant metastasis. We report a rare case of recurrent intraperitoneal UPS. The patient was brought to our department because of locally recurrent intra-abdominal UPS 1 year after the operation. He underwent en-bloc resection of the tumor along with left hemicolectomy in our department. However, only 1 month after he was discharged from our hospital, we were deeply shocked that the tumor recurred and was larger than before.

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Case Report

A 43-year-old male presented in November 2016 because of left abdominal pain and a palpable intra-abdominal mass for 1 month. No weight loss or other symptoms were noted. Patient once underwent exploratory laparotomy for complete removal of intra-abdominal tumor in 2005, 2011, and 2015 in local hospital. Although the details of these surgeries were unknown, the pathology from local hospital showed fibrous histiocytoma.

Physical examination revealed a huge, firm palpable, and painful mass in the left part of the abdomen. The vital signs of patient were stable. Carcinoembryonic antigen (CEA) and carbohydrate antigen 19-9 (CA19-9) were all within normal range. Abdominal computerized tomography (CT) (Fig. 1) showed a huge heterogeneous mass with irregular margins in the left upper quadrant. There were no clear boundaries between the mass and descending colon. Colonoscopy (Fig.2) showed luminal narrowing but no space-occupying lesions. Based on these results, a recurrence of abdominal fibrous histiocytoma was suspected.

An exploratory laparotomy was performed in December 2016, in which we saw a large mass sized 15.0 × 12.0 cm, located in the left middle part of abdomen that infiltrated the descending colon and retroperitoneum with unclear boundaries. There was no distant metastasis and peritoneal seeding. A few enlarged lymph nodes were detected at the pericolic adipose tissue. We performed a left hemicolectomy along with en-bloc resection of the tumor and lymph node.

Macroscopically, the resected specimen measured 15 × 20 × 10 cm and was hard in texture with partial necrosis and unclear demarcation between the mass and serosal surface of colon. The cut surface of the mass was yellow, rufescent, and partially whitish. No cancer cells seen microscopically at the

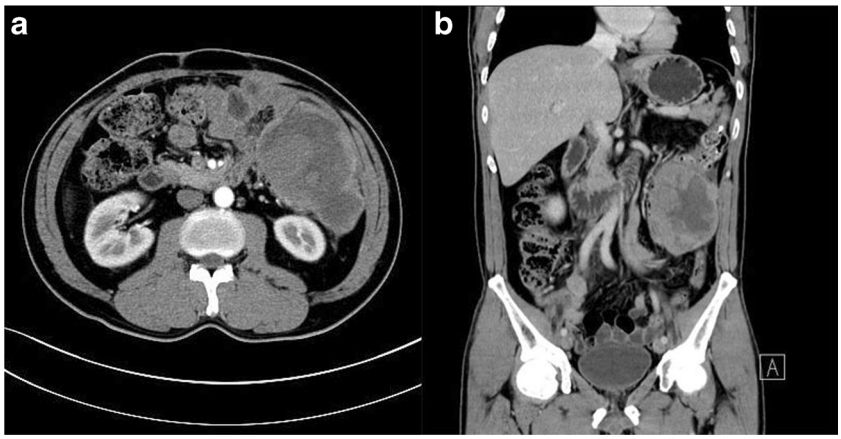

Fig. 1 Abdominal enhanced CT scan shows a heterogeneous and poorly defined mass (measuring 10.8 × 7.2 × 11.2 cm) in the left abdomen

resection margin. Microscopic examination revealed malignant tumor with abundant necrotic tissue that gave rise to spindle cell sarcoma as a possible different diagnosis. High power view of tumor cells showed marked pleomorphism with bizarre nuclei (Fig.3). In the immuno-histochemical studies, vimentin, CD31, CK-pan, CK-L, and Fli-1 were positive in the tumor cells, while desmin, SMA, CD117, CD34, HMB-45, S-100, DOG-1, EMA, ERG, CD21, CD23, and CD35 were negative. Ki-67 proliferation index was about 60% (Fig. 4).The final histopathologic diagnosis was high-grade undifferentiated pleomorphic sarcoma, and four pericolic lymph nodes showed no tumor metastasis.

Our multi-disciplinary team (MDT) suggested that postoperative radiation might be a good option after considering that this patient presented with retroperitoneal and descending colon invasion. However, there are no clear guidelines regarding adjuvant radiotherapy of UPS in the colon. Besides, the patient refused further adjuvant chemotherapy and radiotherapy. The postoperative course was uneventful and the patient was quickly discharged. However, only 1 month after he was discharged from our hospital, he returned to our department because of abdominal pain and distention. Contrast-enhanced abdominal computed tomography (CT) showed a large heterogenous and poorly circumscribed mass in the left and lower abdomen again (Fig.5). Pathology of CT-guided mass puncture showed malignant tumor with local necrosis, suspected to be recurrence of UPS (Fig.6). We were deeply shocked that the tumor recurred so quickly and was larger than before. The patient did not receive any further operation in our hospital.

Discussion

Undifferentiated pleomorphic sarcoma (UPS) is a sarcoma of mesenchymal origin affecting soft tissues of the body, particularly the extremities and retroperitoneum, yet it has been reported in almost all parts of the body. [5] UPS is divided into four histological types: storiform-pleomorphic, myxoid, giant cell, and inflammatory subtypes [2, 4]. The histological morphology varies, but the classic form is composed of spindle-shaped and round histiocytes arranged in storiform pattern as in our case described. Although the cells of origin are currently unknown, gene expression profiling and functional analysis suggest that mesenchymal stem cells may be the precursors of UPS [6, 7].

Most patients with UPS do not have typical clinical manifestations; they present to the hospital late because of the relative absence of early clinical symptoms. If symptomatic, patients often suffer from abdominal distension, altered

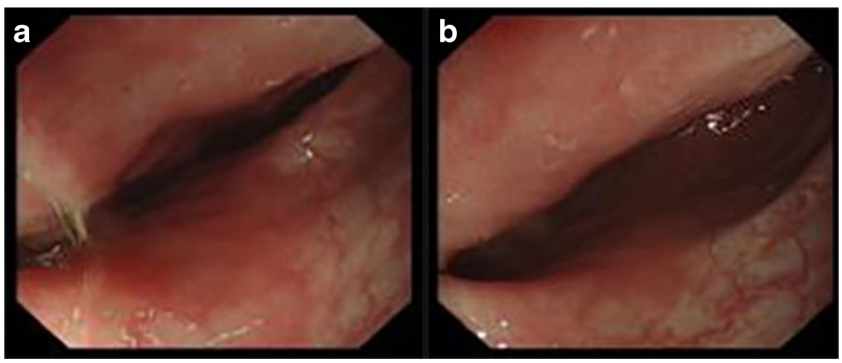

Fig. 2 Colonoscopy showed luminal narrowing but no space-occupying lesions

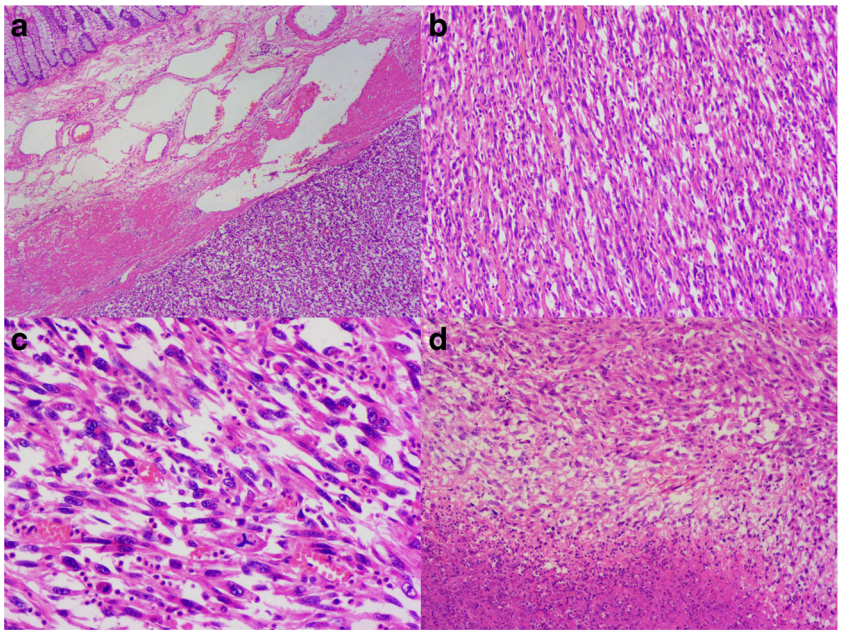

Fig. 3 Hematoxylin-eosin stains. **a** Showed tumor cells infiltrated the left colon that reached the muscular layer of colon. (HE, ×40). **b** Revealed the tumor was composed of a mixture of rounded spindle cells in a storiform pattern, mixed with numerous acute and chronic inflammatory cells. (HE, ×100). **c** Revealed significant atypia and pleomorphism present in tumor cells. Obvious nucleolus and pathological mitotic phase were present. (HE, ×200). **d** Showed tumor cells with large necrotic areas. (HE, ×100)

bowel motions, abdominal discomfort, and occasionally weight loss and anemia. Due to the lack of specific clinical presentation or images or tumor markers, it is difficult to make the preoperative diagnosis of UPS. The tumor is often large at presentation since it usually goes unnoticed for a long time [8–10]. For localization, diagnosis and staging of retroperitoneal tumors, CT scan is the most commonly utilized imaging modality. The CT imaging of UPS usually demonstrates a large, lobulated and nodular heterogeneous mass. Calcifications may be seen, and lesions may be hemorrhagic, have areas of myxoid degeneration or necrotic change [8, 11]. There may also be invasion of other organs. In our patient, the tumor infiltrated the retroperitoneum and descending colon. As regards to the histological and immuno-histochemical staining results, it was reported that MFH frequently expresses vimentin, actin, CD68, alpha 1-antitrypsin, and alpha 1-antichymotrypsin, and often laminin mRNA. [10] In this case, vimentin, CD31, CK-pan, CK-L, and Fli-1 were positive. There is neither reproducible immunophenotype nor any pattern of protein expression that would allow a more specific sub-classification [7]. Usually, as in our case, only vimentin is convincingly positive.

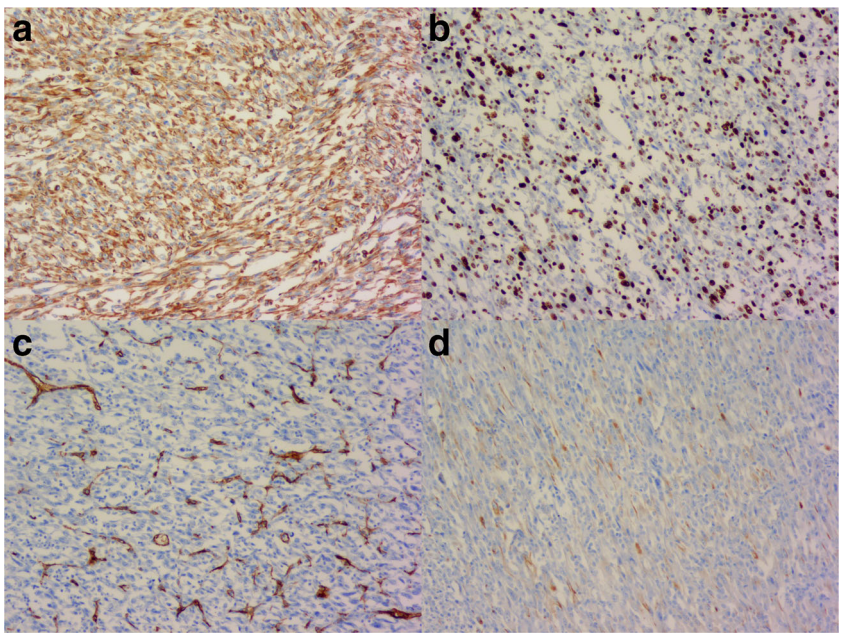

Fig. 4 Immunohistochemical stains. **a** Showed vimentin was positive in the cytoplasm of the tumor cells (×100). **b** Showed Ki-67 proliferation index was about 60% (×100). **c** Showed CD34 was negative (×100). **d** Showed CK-L was negative (×100)

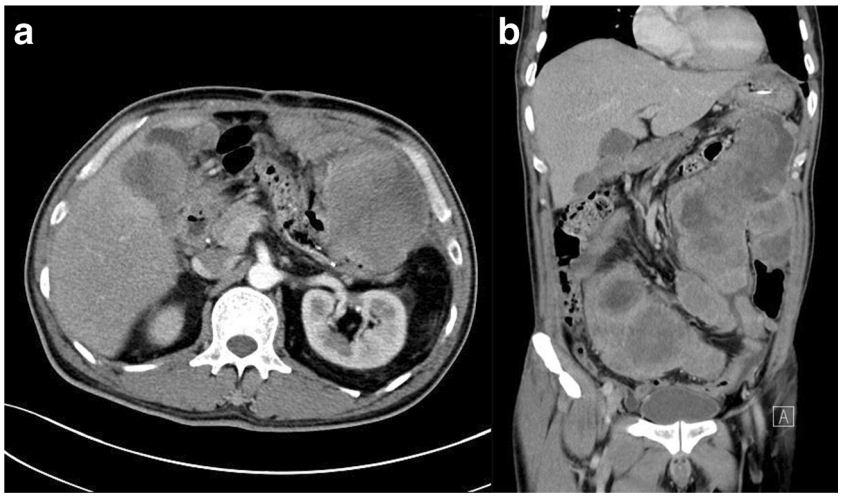

Fig. 5 Abdominal enhanced CT scan shows multiple heterogeneous and poorly demarcated circumscribed mass

The primary treatment of retroperitoneal UPS is en-bloc resection of tumor with free margins. However, the prognosis of UPS is very poor. Because of low incidence of UPS in abdomen, the prognosis associated with intraabdominal UPS is unclear, but most of the case reports suggest unfavorable outcomes, due to the propensity of UPS to invade contiguous organs and to recur locally [10]. Metastases are most frequently to the lung (82%) and lymph nodes (32%), followed by the bone and liver [4]. Recent studies suggest that some indicators such as p-STAT3, SOCS3, MET, and KIT may be a useful prognostic factor for UPS, whether correlated with a better or worse prognosis [12, 13]. The percentages of myxoid component and tumor size are useful predictors of prognosis in UPS. [14] In this case, there was no distant metastasis, and four lymph nodes around the tumor were negative.

Until now, there are no effective clinical chemotherapy options to choose. The efficacy of radiotherapy is well established in the adjuvant treatment of MFH of the extremities. The addition of radiation therapy (RT) to wide surgical excision for UPS has improved local control rates when compared with surgery alone [15]. However, the use of adjuvant radiotherapy is controversial in the abdominal cavity, and is not routinely used. In our case, the patient refused further adjuvant chemotherapy or radiotherapy and was discharged 1 week after operation. However, only 1 month later, pathology of CT-guided mass puncture showed recurrence of UPS. We were deeply surprised that the tumor recurred again so quickly. Our surgeons must be aware that there is a much higher relapse rate in UPS; therefore, a decision must be made on whether a more aggressive combination therapy or only surgical resection must be adopted to achieve "disease control."

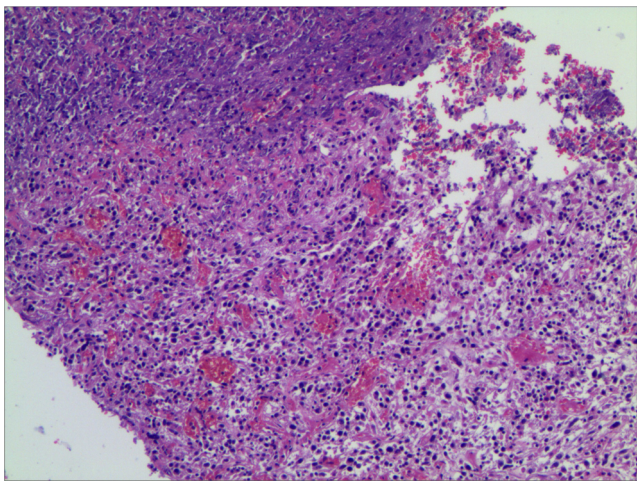

Fig. 6 Hematoxylin-eosin stains showed malignant tumor with local necrosis

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Compliance with Ethical Standards

Ethical Approval Not needed.

Conflict of Interest The authors declare that they have no conflict of interest.

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