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Case report

Gastrointestinal stromal tumor in ruptured Meckel's diverticulum located in jejunum caused myelosuppression for a short time: A case report

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ABSTRACT

Introduction and importance: Gastrointestinal stromal tumor (GIST) in Meckel's diverticulum (MD) is rare but it seems to be a common phenomenon that GIST triggers MD perforation or rupture; the exact mechanism is unclear. In addition, the location of GIST in perforated or ruptrued MD is most common in ileum, rarely in jejunum. We herein report a GIST in ruptured MD Located in jejunum and severe peritoneal cavity infection leads to myelosuppression.

Case presentation: A female patient was admitted to our hospital with "abdominal pain". Physical examination and laboratory tests revealed that the patient was in shock and myelosuppression. Abdominal X-ray photograph and computed tomography indicated perforation of digestive tract. Laparotomy revealed rupture of MD located 90 cm from the Treitz ligament and a tumor was also found in the MD. As the condition is critical, the MD was excisioned from its root and the small bowel gap was closed and repaired. Laboratory indicators showed that myelosuppression was removed 24 h after operation. The pathological findings established a GIST in the MD. The patient was discharged on postoperative day 5 without significant complications.

Clinical discussion: A GIST in ruptured MD Located in jejunum caused the severe peritoneal cavity infection and myelosuppression In a short time, as seen in this case. Failure to recognize the severity of the disease and delay in treatment will endanger the life of the patient.

Conclusion: GIST in MD Located in jejunum is very rare, and the rupture of the MD can be life-threatening at any time.

1. Introduction

Gastrointestinal stromal tumor (GIST) is rare neoplasms of the gastrointestinal stromal tract associated with high rates of malignant transformation [1].GIST arise from the interstitial cells of Cajal, which are specialized network-forming cells distributed in the smooth muscle wall of the digestive tract with a pacemaker role; three identified and mutually exclusive mutations provide the growth stimulation of GIST by affecting c-KIT, PDGFRA, and BRAF genes and are found in 85%, 5%, and less than 1% of cases, respectively [2]. GIST occur more frequently in middle-aged men, with peak incidence in the sixth decade of life. While any portion of the gastrointestinal tract can be affected, they are usually detected in the stomach (60–70%), the percentage of GIST originating in the small intestine is about 20–30% and the remaining 5–10% occur elsewhere in the gastrointestinal tract (e.g., oesophagus, colon, rectum, omentum, and peritoneum) [3].

Meckel's diverticulum (MD) is the most common congenital abnormality of the gastrointestinal tract; it is an outpouching of the mid-ileum that develops as a result of an incompletely obliterated omphalomesenteric, or vitelline, duct. Embryologically, MD is caused by failure of closure of the vitelline duct at the fifth week of fetal growth [4]. The incidence of MD in the general population is about 1% [5], common complications presenting in adults include bleeding, obstruction, diverticulitis, and perforation [6].

Tumors within MD are rare, with a reported incidence of 0.5% to 3.2%, these tumors are commonly benign, like leiomyomas, angiomas, and lipomas, and the majority of malignant neoplasms are adenocarcinoma, sarcoma, and carcinoid tumors, with few being GIST [7]. The

Abbreviations: GIST, gastrointestinal stromal tumor; MD, Meckel's diverticulum.

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location of GIST in MD is most common in ileum, rarely in jejunum [8,9] and myelosuppression caused by ruptured MD with a GIST has not been reported. We herein report a case of GIST in ruptured MD, and more importantly, the MD was located in the jejunum and the ruptured MD resulted in severe peritoneal cavity infection and myelosuppression within an hour.

This work has been reported in line with the SCARE criteria [10].

2. Case presentation

A 72-year-old female patient who was suffering from severe abdominal pain for half an hour and a history of high blood pressure was brought to our hospital by ambulance. At admission, her general condition was poor, with a pulse rate of 120 bpm, blood pressure 80/40 mmHg. On a physical examination she showed muscular defense over the whole abdomen. The laboratory test results on admission were as follows: red blood cell count, 4.76×10^{12} cells/L; hemoglobin 161 g/L; leukocyte count, 2.45×10^{9}/L; platelet count, 80×10^{9}cells/L; super c-reactive protein, 35.6 mg/L; and neutrophil percentage 89.1%.(Table 1). Peritonitis due to perforation of digestive tract was diagnosed based on the free air and dilated loop of the small bowel found on Abdominal X-ray photograph and computed tomography (see Fig. 1); so we performed emergency surgery. Laparotomy revealed rupture of the MD located 90 cm from the Flexor ligament (see Fig. 2a, b) and Interintestinal, pelvic effusion was found. Because the patient was in shock, enterotomy and anastomosis was not performed. The MD were completely resected from the root of the diverticulum, and then the small intestine gap was sutured (see Fig. 2c, d). A mass adjacent to the ruptured rip was touched in the MD (see Fig. 2e). Peritoneal cavity irrigation and drainage were performed, and the abdominal cavity was closed in layers.

Shock was always present with low blood pressure, and fast heart rate during the operation. The anesthesiologist administered antishock therapy, including Liquid infusion and the use of vasoactive drugs. The patient was admitted to the intensive care unit after surgery, laboratory indicators on 24 h after operation are as follows: leukocytes count, 18.32×10^{9}/L, platelets count, 362×10^{9}/L, super-C-reactive protein,322.8 mg/L,(Table 1); Showing that myelosuppression resulted from severe infection was removed. Blood pressure can be maintained after evacuation of vasoactive drugs 48 h postoperatively, and was transferred to the general ward.

Table 1
The laboratory test results on admission and 24 h after operation (NRV: normal reference value).

	On admission	On 24 h after operation
red blood cell count (NRV: 4.3–5.8cells/L)	4.76×10^{12}	4.15×10^{12}
Hemoglobin (NRV: 130–175 g/L)	161	146
leukocyte count (NRV:3.5–9.5cells/L)	2.45×10^{9}	18.32×10^{9}
platelet count (NRV:125–350cells/L)	80×10^{9}	362×10^{9}
super c-reactive protein (NRV:<8 mg/L)	35.6	322.8
neutrophil percentage (NRV: 40–70%)	89.1	71.6
direct bilirubin (NRV:0–7umol/L)	8.4	7.3
Indirect bilirubin (NRV:2–19umol/L)	18.1	19.6
Albumin (NRV:40–55 g/L)	42.9	35.6
Alanine aminotransferase (NRV:9–50 U/L)	39	47
Creatinine (NRV:57–111umol/L)	98.9	72.4
Urea (NRV:3.6–9.5 mmol/L)	4.9	5.1

Regarding the pathological fndings, the MD ruptured at the head position and the length of the rupture rip was about 4 cm; a solid mass measuring 2.5 × 2.5 × 2 cm in size was located at head of the MD adjacent to the ruptured rip; haematoxylin and eosin staining showed proliferation of spindle-shaped cells (see Fig. 3a), and immunohistochemical staining revealed that the tumor was positive for CD117, Dog-1, SMA and Vimentin, with approximately mitotic count of 2/50 per 50 high-power fields (Fig. 3b).Therefore, the patient was diagnosed with a low-risk GIST of the jejunum. The patient was discharged on post-operative day 5 without complications. When the diagnosis was confirmed by histology, the patient immediately received imatinib mesylate therapy. He is currently under follow-up without recurrence or peritoneal dissemination.

3. Discussion

MD is the most common congenital abnormality of the gastrointestinal tract. In the majority of patients, it is asymptomatic and some of them may present with vague nonspecific gastrointestinal complaints such as early satiety, nausea, and vomiting and very few present in an acute setting [11]. MD is reported to occur in 2% of the population, usually found within 40 to 100 cm of the ileocecal valve [12]. MD complications include common complications such as hemorrhage, bowel obstruction, and diverticulitis; uncommon complications include conditions such as enterolith formation, perforation, and neoplasm [13]. Amongst those, which present in an acute setting, bleeding from the GI tract is the most common symptom [14]. The perforation of MD is usually secondary to diverticulitis, gangrene, and peptic ulcer [15], but it can also be induced by trauma [16] and foreign body [17,18]. Symptomatic MD such as gastrointestinal bleeding, Meckel diverticulitis, perforation, and bowel obstruction can all be treated by resection of symptomatic MD. GIST accounts for less than 1% of all gastrointestinal malignancies, their most common location being the stomach [19].

The association of GIST with MD is a known entity; it accounts for about 12% of the tumors associated with MD [20]. Recently, the literature has reported that GIST are the most common tumors responsible for perforation of a MD [21].The exact mechanism is unclear and may be related to tumor necrosis, making ulcer for the pressure of tumor, bowel obstruction and tumor invasion into the muscularis propria replacing the gut wall [22].

We report a 72-year-old woman with a GIST in ruptured MD Located in jejunum. In addition, myelosuppression due to severe abdominal infection were caused within 1 h.

Compared to previous reports, one difference: the diverticulum is located in the jejunum; another difference: the abdominal cavity infection was more serious. Previous literature revealed that the perforation or rupture of MD with GIST mostly occurred in ileum. The occurrence in jejunum is rare. There was only one report [23] that was similar to ours. This is a 46-year-old male patient who was admitted to the hospital with diffuse peritonitis, laparotomy revealed a rupture at the neck of a 12 cm MD in size located in jejunum. Immunohistochemistry of the resected specimen confirmed a GIST about 7.0 × 6.5 cm in size in the MD. In contrast, the patient in our case was older, had smaller volume of diverticulum and stromal tumor, and less bleeding after rupture, but the degree of peritoneal cavity infection was more serious, reaching the degree of myelosuppression. Regrettably, objective laboratory indicators of infection were not mentioned in this case report. Therefore, it is inferred that the peritoneal cavity infection caused by rupture of MD with GIST in jejunum has more serious consequences but it can also be due to personal factors, such as the patient's constitution or diet. Further observation of similar cases is needed.

GIST are mesenchymal tumors that originate from the interstitial cells of Cajal. The majority of GIST are positive for the CD117, Dog-1, SMA and Vimentin [24]. The most reliable prognostic factors for risk classification are the size of the primary tumor and the mitotic index, which measures the proliferative activity of the cells [25]. The

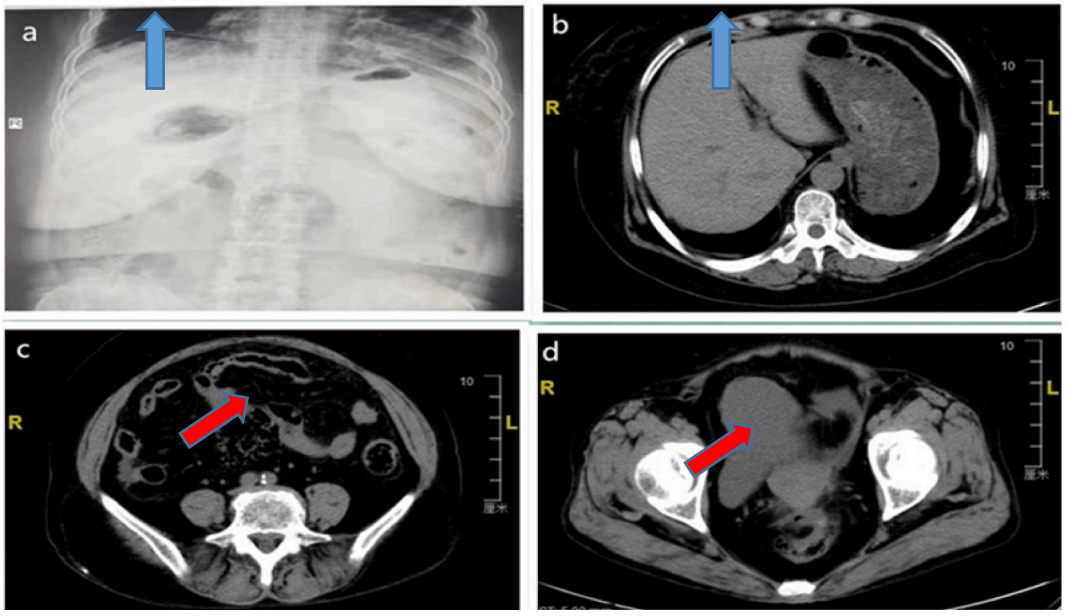

Fig. 1. X-ray photograph and computed tomography findings. Free air is found in the abdominal cavity (the blue arrow). Intestinal dilatation and peritoneal effusion are seen inside the abdominal cavity (the red arrow).

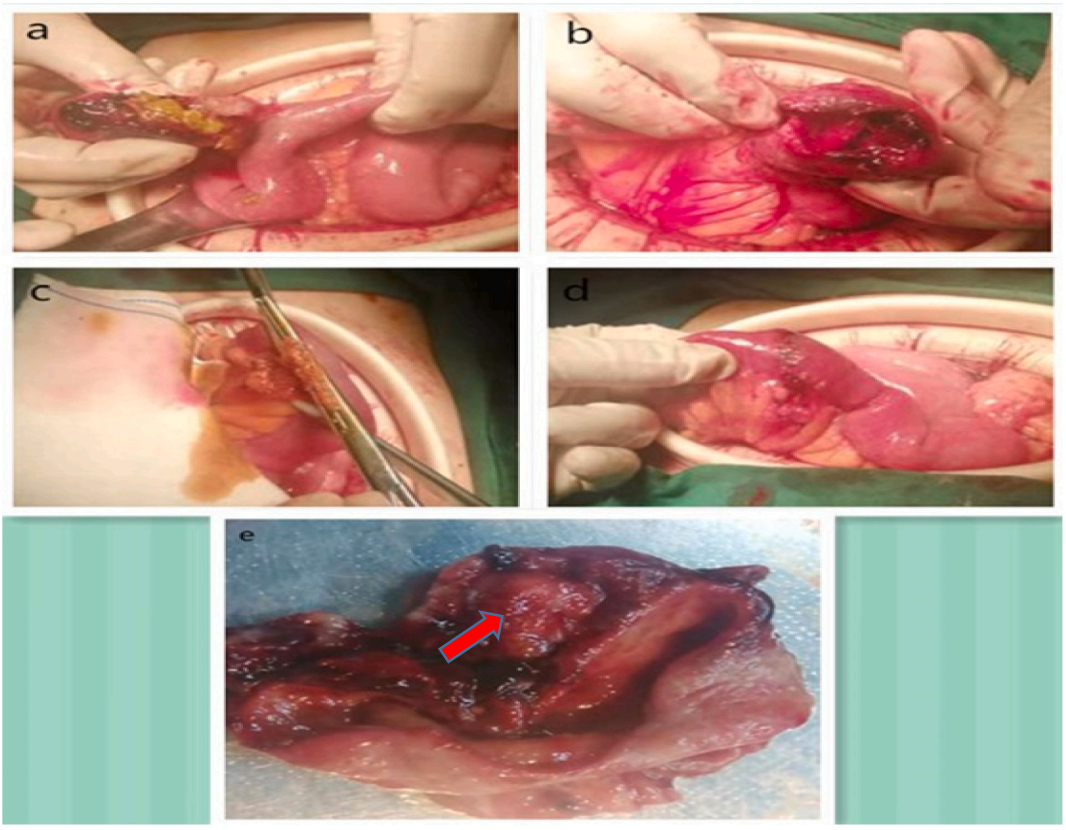

Fig. 2. Intraoperative and postoperative findings.
(a and b) Laparotomy revealed rupture of the MD located 90 cm from the Flexor ligament. View from the side and front. (c and d) The MD was completely resected from the root of the diverticulum, and then the small intestine gap was sutured. e. A mass was touched in the MD close to the rupture rip (the red arrow).

approaches to treating GIST are resection of primary low-risk tumors and resection plus imatinib for high-risk primary or metastatic tumors. According to Johnsue's classification [26], tumor rupture merits assignment to the high-risk category, and any patient presenting with a perforated GIST should receive imatinib, regardless of the mitotic count, as there is a possibility of tumor cells being disseminated into the peritoneal cavity after perforation. However, the patient we reported was in a state of shock during the operation. In order to shorten the operation

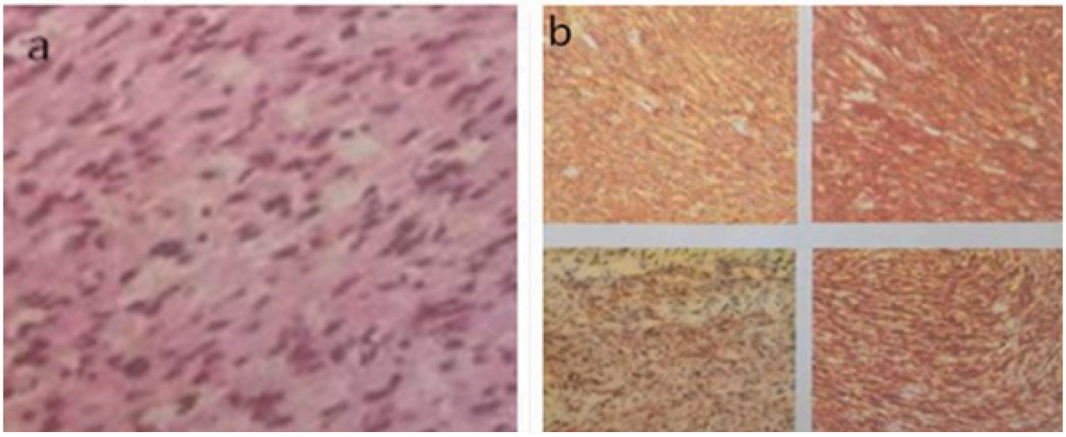

Fig. 3. Immunochemistry findings.
(a) Haematoxylin and eosin staining showing proliferation of spindle-shaped cells.
(b) Immunohistochemistry reveals that the tumor is positive for CD117 (+++), Dog-1 (+++), SMA (+), Vimentin (+++).

time and reduce the damage, the enterectomy was not performed, diverticulotomy alone was performed, and imatinib was given after the operation. The patient was well after one and a half years follow-up.

4. Conclusion

GIST in MD Located in jejunum is very rare. In particular, severe peritoneal cavity infection arising from the rupture of the diverticulum can be life-threatening at any time.

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Ethical approval

The study is exempt from ethnical approval from our institution.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

Fengjuan Li collected case details, literature search and helped to draft the manuscript. Guoqun Jia participated in treatment planning of the patient draft the manuscript. All authors read and approved the final manuscript.

Registration of research studies

Not applicable.

Guarantor

Guoqun Jia.

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Not commissioned, externally peer-reviewed.

Other relevant information

The case report complies with SCARE Guidelines [10].

Declaration of competing interest

The authors have no conflicts of interest.

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