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Case report

Gastrointestinal stromal tumor of the small bowel complicated by torsion: A case report

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ABSTRACT

Introduction: Gastrointestinal stromal tumors (GISTs) are mesenchymal tumors that originate from the gastrointestinal tract wall. Approximately 20–30 % of GISTs originate from the small intestine. GISTs of the small intestine generally present with a palpable mass, distention, and abdominal pain and may exhibit acute abdomen at the onset. Herein, we describe a rare case of a pedunculated GIST of the small intestine complicated by torsion.
Presentation of case: A 69-year-old woman presented with lower abdominal pain. Abdominal contrast-enhanced computed tomography showed a 73 × 62 × 57-mm^3 tumor in the pelvic cavity with enhanced margins and reduced contrast. It was presumed that the tumor had caused hemorrhagic infarction. Emergency laparotomy was performed, and the pedunculated tumor was found to be twisted 360° clockwise at the pedicle with hemorrhage and necrosis due to torsion. We performed partial resection of the small intestine including the tumor. Histopathological examination revealed tightly arranged spindle-shaped cells with hemorrhage, congestion, and inflammatory cell infiltration. Immunohistochemical staining showed positivity for CD34, CD117, and DOG1.
Conclusions: Torsion of a pedunculated small intestine GIST, although very rare, requires emergency surgery and should be recognized as a cause of acute abdomen in patients with GIST. Immediate surgery is mandatory if torsion of a small intestinal GIST is suspected because the GIST or intestine may become necrotic owing to hemorrhagic infarction.

1. Introduction

Gastrointestinal stromal tumors (GISTs) are malignant mesenchymal tumors that can develop in any region of the gastrointestinal tract, and 20–30 % of these tumors occur in the small intestine [1]. GISTs are often asymptomatic but can present with abdominal pain or distention after they grow to a relatively large size. Furthermore, a small intestinal GIST may manifest with acute abdomen due to intraperitoneal hemorrhage, intestinal obstruction, gastrointestinal perforation, or intussusception [2]. This report describes a rare case of an extraluminal GIST with pedicle torsion that manifested with an acute abdomen.

This case report has been reported in line with the SCARE Criteria [3].

2. Presentation of case

A 69-year-old woman was referred to our hospital with a 2-day history of lower abdominal pain. She had no remarkable medical history. On admission, her body temperature was 36.8 °C. Physical examination revealed a slightly distended abdomen with direct and rebound tenderness in the lower right quadrant.

Blood test results revealed a white blood cell count of 14,870/μL (90.4 % neutrophils) and a C-reactive protein level of 3.79 mg/dL. Other blood parameters, biochemical examinations, and coagulation tests were normal. No abnormalities were found regarding tumor markers, including carcinoembryonic antigen (2.9 ng/mL), carbohydrate antigen (CA) 19–9 (5.8 U/mL), and CA 125 (10.1 U/mL).

Abdominal ultrasonography (US) showed a 73 × 52-mm^2 tumor in the pelvis, with a mixture of high and low echogenicity. Blood flow was not observed inside the tumor with color Doppler US (Fig. 1a, b). Abdominal contrast-enhanced computed tomography (CT) revealed a 73 × 62 × 57-mm^3 tumor in the pelvic cavity. It was in contact with the small intestine, which was thickened and edematous. The margins of the tumor were enhanced like a capsule, and the contrast effect was reduced inside the tumor (Fig. 2a, b).

Emergency laparotomy was performed the same day based on the

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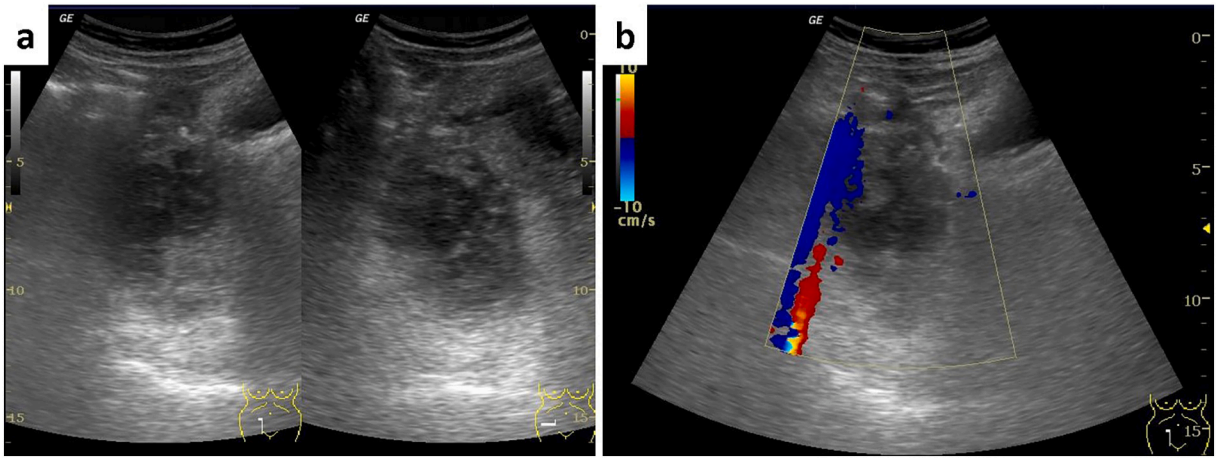

Fig. 1. Abdominal ultrasonography findings.
(a) Abdominal ultrasonography shows a 73 × 52 mm^2 tumor in the pelvis, which has a mixture of high and low echo densities.
(b) No blood flow is observed inside the tumor on color Doppler ultrasonography.

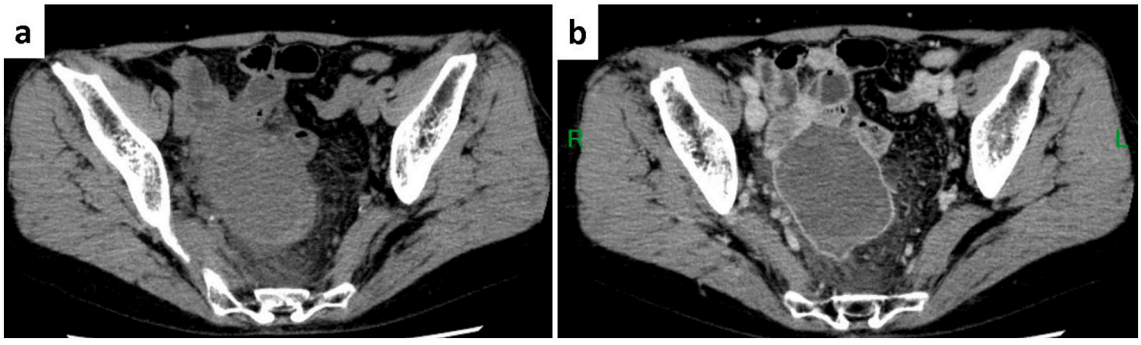

Fig. 2. Computed tomography findings.
(a) Abdominal computed tomography (CT) shows a 73 × 62 × 57 mm^3 low-density tumor in the pelvic cavity.
(b) Contrast-enhanced CT shows an enhanced tumor margin but no contrast effect inside it.

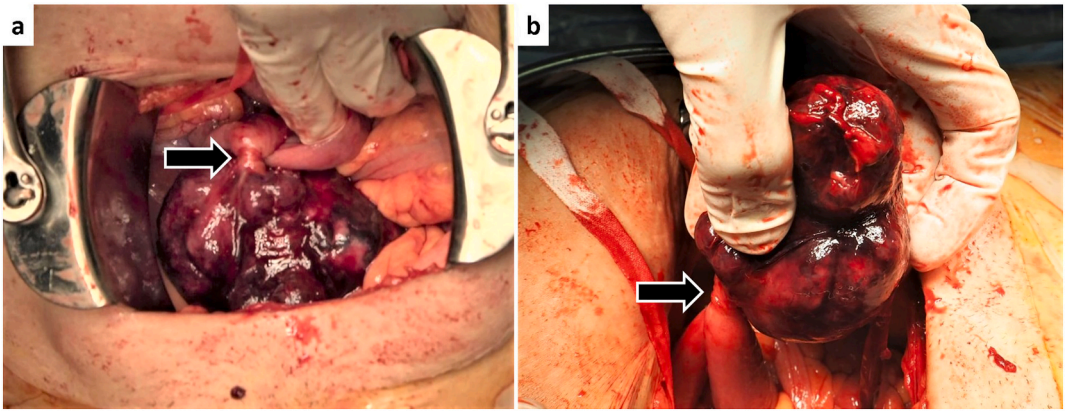

Fig. 3. Intraoperative findings.
(a, b) The pedunculated tumor originates from the contralateral mesentery of the small intestine, 190 cm from the ligament of Treitz on the anal side and 250 cm from the ileocecal valve on the oral side; it is twisted 360° at the pedicle (arrow) and shows features suggestive of hemorrhagic necrosis. No torsion was observed in the small intestine, to which the tumor was attached, and no evidence of intestinal obstruction, bleeding, or necrosis was present. There was no evidence of tumor infiltration into surrounding organs and lymph node metastasis in the small mesentery.

presumptive diagnosis of tumor torsion with ischemic necrosis. Bloody ascitic fluid was observed in the abdominal cavity during laparotomy. A fist-sized, dark red, uneven tumor was found in the pelvis. The tumor was pedunculated on the antimesenteric side of the small intestine, 190 cm from the ligament of Treitz and 250 cm from the ileocecal valve, and it was twisted 360° clockwise at the pedicle. The tumor was hemorrhagic and necrotic due to torsion. No torsion was observed in the part of the small intestine to which the tumor was attached; no evidence of intestinal obstruction, bleeding, or necrosis was observed. There was no evidence of tumor infiltration into surrounding organs and lymph node

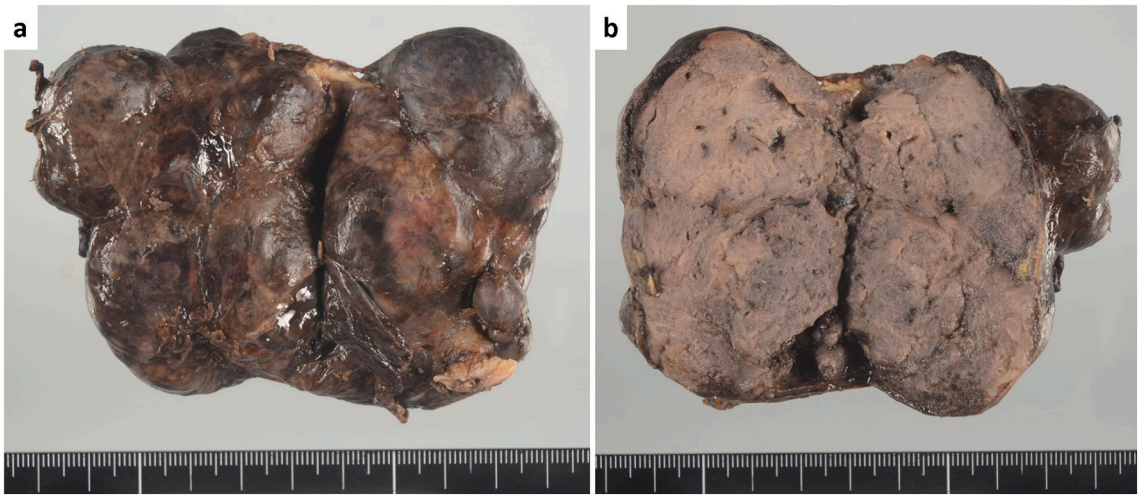

Fig. 4. Gross findings after formalin fixation.
(a). The 75 × 55 × 45-cm^3 tumor is well-defined, reddish-brown, uneven, and elastically hard.
(b)The cut surface shows a gray/red solid tumor with phyllodes and internally, the tumor is heterogeneous and solid after formalin fixative is applied.

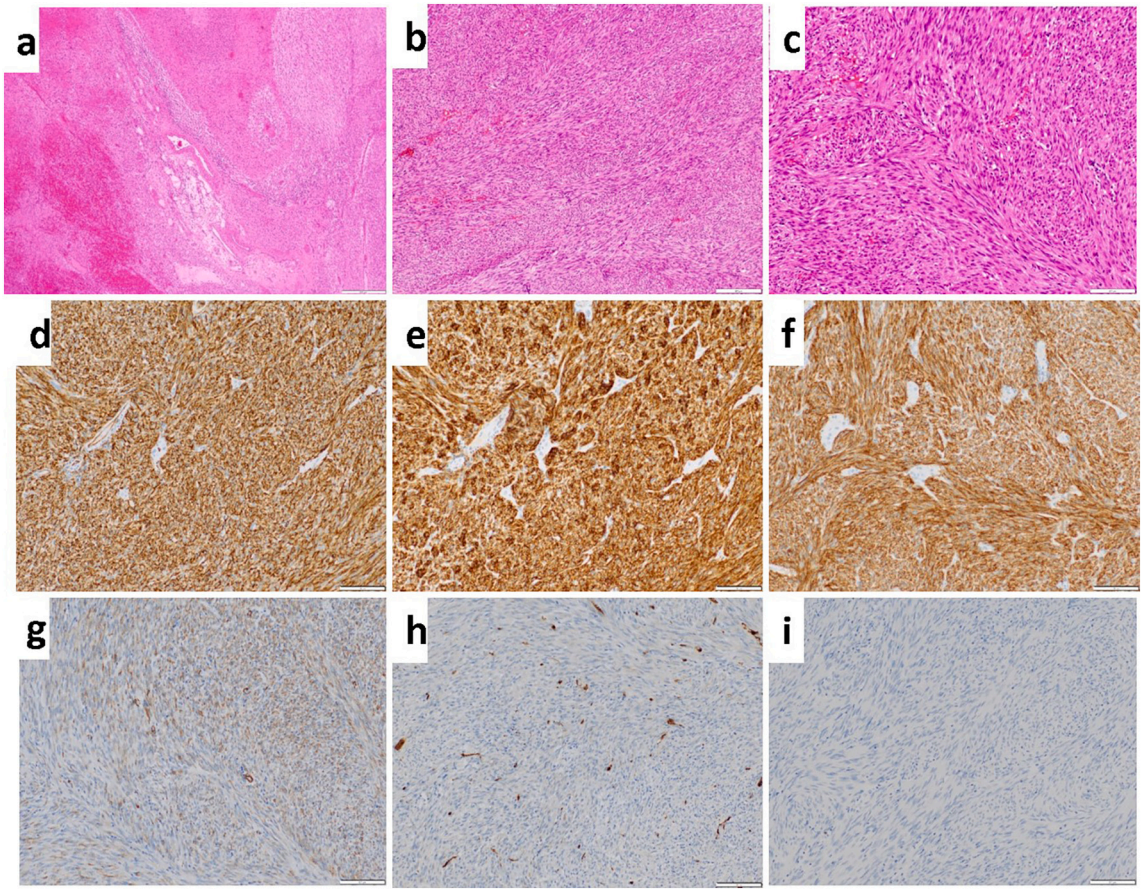

Fig. 5. Histopathological findings.
1) Hematoxylin and eosin staining showing tightly arranged spindle-shaped cells (b, c) that are accompanied by hemorrhage, congestion, and inflammatory cell infiltration (a). The scale bars in (a), (b), and (c) indicate 500 μm, 200 μm, and 50 μm, respectively.
2) Immunohistochemical analysis showing tumor cells stained positive for (d) CD34, (e) CD117 (C-kit), and (f) DOG1; weakly positive for (g) α-smooth muscle actin; and negative for (h) S200 and (i) desmin. The scale bar indicates 100 μm.

metastasis in the small mesentery. (Fig. 3a, b). We performed segmental resection of approximately 10 cm of the small intestine including the tumor with end-to-end anastomosis. No postoperative complications occurred, and the patient was discharged.

The 75 × 55 × 45-mm^3 tumor was well-defined, reddish-brown, uneven, and elastically hard. The cut surface showed a gray/red solid tumor with phyllodes and internally, the tumor was heterogeneous and solid after the application of formalin fixative (Fig. 4a, b). Microscopic

examination after hematoxylin-eosin staining revealed tightly arranged spindle-shaped cells that were accompanied with changes in blood flow, such as hemorrhage, congestion, and inflammatory cell infiltration (Fig. 5a–c). Immunohistochemical staining was performed to characterize the tumor cells. The tumor cells showed positive staining for CD34, CD117 (C-kit), and DOG1; they were weakly positive for α-smooth muscle actin but were negative for S200 and desmin (Fig. 5d–i). The mitotic index was <5/50 high-power fields (×400), and the Ki-67 value was 5–10 %. Based on these findings, a diagnosis of torsion and strangulation necrosis of the GIST of the small intestine was confirmed, and her condition was classified as high risk based on the prognostic classification.

After the patient was discharged, she was started on oral imatinib mesylate therapy (400 mg daily). She is currently being followed up using CT, and she has had no recurrence for 1 year postoperatively.

3. Discussion

In 1983, Mazur and Clark proposed the name "stromal tumor" to distinguish it from other smooth muscle gastrointestinal tumors [4]. GISTs originate from interstitial cells of Cajal that regulate autonomous contraction of the gastrointestinal tract. The incidence of GIST is 6–14 cases per million people in the United States of America and Europe [5] and approximately 16–22 cases per million in Asia [6]. Although they account for only 0.1–3 % of all gastrointestinal malignancies, GISTs are the most common mesenchymal tumors of the gastrointestinal tract. They can occur anywhere in the gastrointestinal tract; the stomach accounts for 50–60 % of cases, the small intestine for 20–30 %, the colon or rectum for 5–10 %, the esophagus for <5 %, and the peritoneum and mesentery for <1 % of cases. However, small intestinal GISTs account for 8.4 % of small intestinal malignancies [7].

Small intestinal GISTs often develop with non-specific symptoms after they become relatively large. Clinical symptoms are primarily due to tumor diameter, tumor rupture, and the relationship between the tumor and surrounding tissues. It causes various symptoms, such as a palpable abdominal mass, fullness, nausea, vomiting, and abdominal pain. Although uncommon, small intestinal GISTs may cause gastrointestinal bleeding, which manifests as hematemesis, anemia, and acute abdomen due to intestinal obstruction, perforation of the tumor, or intussusception [8–11]. In addition, small intestinal volvulus caused by GIST manifests as acute abdomen. Generally, small intestinal volvulus is caused by mid-gut volvulus due to abnormal intestinal rotation and fixation, postoperative adhesions, Meckel diverticulum, tumor, intestinal duplication, hernia, or diverticulitis, or an unknown cause [12]. There are reported cases of small intestinal volvulus caused by GIST, including 18 cases in Japanese literature and four cases in English literature [13–16]. The average age of patients with GIST was 67 years, and there were seven male and 11 female patients. Tumors with a diameter ≥10 cm (median diameter, 9.5 cm) were found in 10 cases, and many tumors were relatively large. Volvulus of the small intestine varied from 120° to 720°, and necrosis of the small intestine was observed in six cases. The "whirl sign," which is seen when the intestinal tract is rotated around the superior mesenteric artery, was a characteristic finding on CT, magnetic resonance imaging (MRI), and US, and it was observed in many reported cases of small intestinal volvulus caused by small intestinal GIST. Regarding its pathology, as the tumor diameter increases in extramural small intestinal GISTs, the small intestine may become twisted as the tumor rotates. Alternatively, if the tumor has poor mobility in the abdominal cavity, the tumor may become an axis, and small intestine volvulus may occur. Of these previous reports, two cases, one in Japanese and one in English literature, involved a volvulus with small intestinal GIST torsion [14]. Torsion of the small intestinal GIST is reportedly caused by rotation of the pedunculated GIST attached to the small intestine at the pedicle, and this causes secondary volvulus of the small intestine. However, in our case, small intestinal GIST torsion developed without a volvulus of the small intestine. In this case, the tumor base, continuous with the intestine, was thin, and the tumor was relatively large and highly mobile; hence, it was speculated that the pedicle of the small intestinal GIST became twisted. Small intestinal volvulus caused by GIST is usually accompanied by intestinal obstruction, but small intestinal GIST torsion alone without intestinal obstruction has been reported. Additionally, small intestinal GIST torsion itself can cause blood flow disorders, including necrosis, which is different from cases involving small intestinal volvulus as well.

CT is an essential test for the diagnosis of GIST. Contrast-enhanced CT can identify the tumor and metastasis, as well as evaluate the tumor's size, relationship to surrounding organs, and contrast effect. On CT images, small GISTs are relatively uniform inside and are usually enhanced from the arterial phase. However, as the tumor grows and becomes more malignant, internal heterogeneity, necrosis, and cystic change can appear [5,17]. As in our case, if a GIST has torsion and ischemic necrosis, the tumor may have no contrast effect inside and may be seen as a cystic lesion on CT.

The curative treatment for GIST is radical resection, which is essential to ensure a safe surgical excision margin without rupture. There is no need for systematic lymph node dissection because of the low frequency of lymph node metastasis.

In patients with elective symptoms of GIST, diagnosis using CT, 18F-fluorodeoxyglucose positron emission tomography, MRI, and when applicable, endoscopic US-fine needle aspiration is common, and resection surgery should be considered. Intraperitoneal hemorrhage, intestinal obstruction, gastrointestinal perforation, and intussusception are known causes of acute abdomen in patients with GIST. Additionally, volvulus or torsion of GISTs should be recognized as a cause of acute abdomen in patients with GIST, and such cases require emergency surgery. GISTs often become necrotic due to hemorrhagic infarction; therefore, radial resection should be performed.

Within the completely resected GIST, several risk classifications can be made according to tumor size, mitotic index, primary site, and presence of tumor rupture, which sharply reflects the risk of recurrence. According to the revised National Institutes of Health Consensus Criteria, tumor size >10.0 cm, >10 mitoses per 50 high-power microscopic fields, tumor diameter >5.0 cm mitotic count >5, and tumor rupture pose a high risk of recurrence [18]. Three-year adjuvant imatinib for high-risk GIST patients who have undergone surgery helps improve recurrence-free and overall survival. Recently, adjuvant imatinib therapy has been used for wider indications, including medium-risk tumors >3 cm in size and primary tumors with rupture or perforation [19].

Regular CT scans or MRIs should be considered for follow-up after complete resection. The 5-year recurrence-free survival varies greatly (60–95 %) depending on the recurrence risk, and the recurrence rate increases after completing adjuvant chemotherapy. Therefore, there is no consensus regarding the follow-up interval or period, and the risk of recurrence, number of years after surgery, and presence or absence of adjuvant chemotherapy should be considered [20].

4. Conclusions

In summary, torsion of a small intestinal GIST, although very rare, requires an emergency operation, and it should be recognized as one of the causes of acute abdomen in patients with GIST; and immediate surgery is mandatory if torsion of a small intestinal GIST is suspected because the GIST or part of the intestine may become necrotic due to hemorrhagic infarction.

Provenance and peer review

Not commissioned, externally peer-reviewed.

Consent to publish

Written informed consent was obtained from the patient for the 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.

Ethical approval

Ethical approval has been exempted by our institution because this is a case report and no new studies or new techniques were carried out.

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Guarantor

The guarantor for this case report is Jun-ichi Yoshizawa.

Research registration number

Not applicable.

CRediT authorship contribution statement

JY contributed to the conception and design of the Case Report and drafted the manuscript. JY performed the resection of GIST. JY, TS, TI, KF, and AN treated the patient after surgery. All authors have read and approved the final version of this manuscript.

Declaration of competing interest

None.

Data availability

The datasets supporting the conclusions of this article are included within the article.

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