Case report

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

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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 intaperitoneal 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 × 62× 57-mm tumor in the pelvic cavity with enhanced margins and 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 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 clinical and imaging findings. Abdominal contrast-enhanced computed tomography showed a 73 × 62 × 57-mm 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).

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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.
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 present. There was no evidence of tumor infiltration into surrounding organs and lymph node metastasis in the small mesentery.

Fig. 1. Abdominal ultrasonography findings.
(a) Abdominal ultrasonography shows a 73 × 52 mm² 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 mm³ 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.
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-mm³ 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
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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 the gastrointestinal tract, the incidence of GIST becomes higher, and its prognosis is worse. Therefore, early diagnosis and treatment are essential.

Small intestinal GISTs are relatively rare and only a few cases have been reported. According to our literature review, 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 the gastrointestinal tract, the incidence of GIST becomes higher, and its prognosis is worse. Therefore, early diagnosis and treatment are essential.

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

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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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