Gastrointestinal Stromal Tumor of the Small Bowel Complicated by Torsion: A Case Report

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Case report

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Abstract

Background: Gastrointestinal stromal tumor (GIST) is a mesenchymal tumor originating in the wall of the gastrointestinal tract, and 20-30% of these tumors originate from the small intestine. The clinical manifestations of GIST of the small intestine generally include a palpable mass, distention, and abdominal pain and sometimes include acute abdomen due to ileus, bleeding, perforation, and intussusception at onset. Here, we describe a rare case of a pedunculated GIST of the small intestine complicated by torsion.

Case presentation: A 69-year-old woman was referred for lower abdominal pain. Abdominal contrast-enhanced computed tomography showed a 73×62×57-mm tumor in the pelvic cavity with an enhanced margin and reduced contrast effect inside of it. T2-weighted magnetic resonance imaging showed a hyperintense, demarcated, lobulated tumor. The diffusion-weighted image showed hyperintensity, and the apparent diffusion coefficient value revealed diffusion restriction, indicating malignancy. T1-weighted images showed a low intensity tumor; contrast-enhanced images showed hypointensity inside of the tumor and slight hyperintensity around its edges. It was presumed that the tumor had caused hemorrhagic infarction. Emergency laparotomy was performed, and the pedunculated tumor was found to be twisted 360 degrees clockwise at the pedicle with hemorrhage and necrosis due to torsion. We performed partial resection of the small intestine including the tumor. Histopathology revealed tightly arranged spindle-shaped cells with hemorrhage, congestion, and inflammatory cell infiltration. Immunohistochemical staining showed positivity for CD34, CD117, and DOG1.

Conclusions: The present report describes a very rare case in which pedicle torsion of an extra luminal expanding GIST of small intestine, although there were some reports of small intestine volvulus caused by GIST of small intestine as a rare condition. The torsion of pedunculated GIST of small intestine should be recognized as an emergency condition of GIST, and we report that characteristic findings of this condition were acquired with CT and MRI which was possibility useful for the diagnosis.

Background

Gastrointestinal stromal tumor (GIST) is one of the malignant mesenchymal tumors that can develop in all regions of the gastrointestinal tract, and 20–30% of the tumors originate from the small intestine[1, 2]. A GIST is often asymptomatic, but can present as abdominal pain or distention after growing 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[3]. The present report describes a very rare case of an extraluminal GIST with pedicle torsion that manifested with acute abdomen. Moreover, we report characteristic features of this condition based on contrast-enhanced computed tomography (CT) and magnetic resonance imaging (MRI) findings, which may be useful in the diagnosis of similar cases.

Case Presentation

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

Blood test results indicated a white blood cell count of 14,870/ μ L (90.4% neutrophils) and a C-reactive protein level of 3.79 mg/dL. No abnormal findings were reported from the other blood counts, biochemical examinations, and coagulation tests. No abnormalities were found in the tumor markers: 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 tumor in the pelvis, with a mixture of high and low echogenicity. Blood flow was not observed inside the tumor with color Doppler US, and the ovary was unclear (Fig. 1A, B). Abdominal contrast-enhanced CT revealed a 73×62×57-mm tumor in the pelvic cavity. It had contact with the small intestine, which was thickened with edema. The margin of the tumor was enhanced like a capsule, and the contrast effect was reduced inside of it, similar to that in an abscess (Fig. 2A, B). There was ascites but no findings of free air or ileus. MRI demonstrated a 5×5×7-cm solid tumor showing moderate intensity, of which the surface was clearly demarcated and lobulated in the pelvis on a T2-weighted image (Fig. 3A, B). Diffusion-weighted imaging (DWI) showed a clearly hyperintense tumor, and the mean apparent diffusion coefficient (ADC) value was low (1.15×10^{-3} mm²/s), indicating diffusion restriction; thus, malignancy was suspected (Fig. 4). On T1-weighted imaging, the tumor was heterogeneous and had a low intensity, but on contrast-enhanced T1-weighted imaging, the inside of the tumor was hypointense and there was slight hyperintensity around the edges; hence, it was presumed the tumor caused hemorrhagic infarction (Fig. 5A–C).

Retrospectively, no abnormalities were found in the ovaries; thus, torsion of the pelvic malignant tumor with ischemic necrosis, especially a small intestine tumor, was the most likely diagnosis based on the results of contrast-enhanced CT and MRI.

First, emergency laparotomy was performed by gynecologists on the same day based on the presumptive diagnosis of ovarian tumor torsion. Bloody ascites was observed in the abdominal cavity during laparotomy. A fist-sized dark red mass was found in the pelvis. The tumor was recognized as a pedunculated tumor on the antimesenteric side 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, and it was twisted 360 degrees clockwise at the pedicle. The tumor was hemorrhagic and necrotic due to torsion (Fig. 6A, B). We performed partial resection of the small intestine including the tumor. No postoperative complications occurred, and the patient was discharged.

The tumor was a well-defined reddish-brown, uneven tumor measuring 7.5×5.5×4.5 cm. The cut surface was a gray/red solid tumor with phyllodes (Fig. 7A, B). Microscopic examination after hematoxylin-eosin staining revealed tightly arranged spindle-shaped cells that were accompanied by changes in blood flow, such as hemorrhage, congestion, and inflammatory cell infiltration (Fig. 8A–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. 9A–F). The mitotic index was $< 5/50$ high-power fields ($\times 400$), and the Ki-67 value was 5–10%. Based on these findings, the patient's 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 discharge, she was started on oral imatinib mesylate therapy daily (dose, 400 mg). She is currently being followed up, and at 1-year postoperatively, she has had no recurrence.

Discussion And Conclusions

In 1983, Mazur and Clark proposed the name “stromal tumor” to distinguish it from other smooth muscle gastrointestinal tumors[4]. GIST originates in the interstitial cells of Cajal that regulate autonomous contraction of the gastrointestinal tract[5]. The incidence of GIST is 6–14 cases per million people in the United State of America and Europe[6] and approximately 16–22 cases per million in Asia[7]. Although they account for only 0.1–3% of all gastrointestinal malignancies, GISTs are the most common mesenchymal tumors of the gastrointestinal tract[8]. They can occur anywhere in the gastrointestinal tract, with the stomach accounting for 50–60% of cases; the small intestine, 20–30%; the colon or rectum, 5–10%; the esophagus, $< 5\%$; and the peritoneum and mesentery, $< 1\%$ [9]. However, it has been reported that small intestinal GISTs account for 8.4% of small intestinal malignancies[10].

Based on histological classification, GISTs can be divided into the spindle cell (70%), epithelioid cell (20%), and mixed subtypes[11, 12]. They are usually positive for c-KIT (CD117), CD34, and DOG1[13]. DOG1 was discovered in 2004 and has been strongly positive in various histological types of GIST[14]. DOG1 refers to a monoclonal antibody against the chloride channel protein expressed in GISTs. DOG1 was positive in 87–94% of GIST cases, whereas CD117 and CD34 were reported to be positive in 74–95% and 60–70% of GIST cases, respectively[15, 16]. It is considered that the diagnostic rate can be increased by combining CD117 and DOG1 in immunohistochemical staining when evaluating for GISTs.

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 relationship between the tumor and surrounding tissues. It causes various symptoms such as a palpable abdominal mass, fullness, nausea, vomiting, and abdominal pain[6]. 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[17–21]. In addition, small intestinal volvulus caused by GIST manifests as acute abdomen. Generally, small intestinal volvulus is caused by 1) mid-gut volvulus due to abnormal intestinal rotation and fixation; 2) postoperative adhesions, Meckel diverticulum, tumor, intestinal duplication, hernia, or diverticulitis; or 3) an unknown cause[22]. There are reported cases of small intestinal volvulus caused by GIST, including 18 cases in Japanese literature and 4 cases in English literature[23–26]. The average age of patients with GIST was 67 years, and there were

7 male patients and 11 female patients. Tumors with a diameter of 10 cm or more (median diameter, 9.5 cm) were found in 10 cases, and many tumors were relatively large. Volvulus of the small intestine varied from 120 degrees to 720 degrees, and 6 cases had necrosis of the small intestine. The “whirl sign,” which is seen when the intestinal tract is rotated around the superior mesenteric artery, was a characteristic finding on CT, MRI, and US, and it was observed in many reported cases of small intestinal volvulus caused by small intestinal GIST. Regarding 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 can become an axis, and small intestine volvulus may occur. Of these previous reports, 2 cases, 1 in Japanese and 1 in English literature, involved a volvulus with small intestinal GIST torsion[24]. It is considered that torsion of the small intestinal GIST is 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 volvulus of the small intestine. In this case, the tumor base continuous with the intestine was thin, the tumor was relatively large, and the tumor was highly mobile, so 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. In addition, small intestinal GIST torsion itself has caused blood flow disorders including necrosis, which is different from cases involving small intestinal volvulus as well.

CT is an essential imaging 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, but as the tumor grows and becomes more malignant, internal heterogeneity, necrosis, and cystic change can appear[6, 27]. As in our case, if a GIST has torsion and ischemic necrosis, the tumor will have no contrast effect inside and may be seen only as a cystic lesion, similar to an abscess on CT.

Since MRI usually has a long imaging time, it is easily affected by peristaltic movement of the gastrointestinal tract. Furthermore, since the spatial resolution is not high, there is a limit to the visualization of small lesions. However, MRI can clearly reveal the relationship with surrounding organs because of its high contrast resolution. Moreover, MRI performed with various sequences is more accurate than CT for identifying the morphologic features of the tumor components, such as fluid, mucin, and fibrous portions, as well as the presence of bleeding, edema, and infarction. On MRI, GISTs often show a moderate to low intensity on T1-weighted images, and high to moderate intensity on T2-weighted images. Typically, small GISTs are round and have a strong and uniform contrast effect in the arterial phase. Large GISTs are often lobular and heterogeneous, their contrast effect is mild with a heterogeneous gradual enhancement pattern, and they frequently exhibit intratumoral cystic changes similar to CT findings. DWI is particularly sensitive to the microscopic movement of water. GISTs often show diffusion restriction on DWI and a low ADC value, which reflect the higher cellularity seen as common findings of a malignant tumor. DWI in combination with ADC may be useful for diagnosing small intestinal GIST and assessing its malignancy[6, 28]. In our case, T2-weighted MRI showed a heterogeneous solid tumor of moderate intensity. DWI showed diffusion restriction, which indicated a

malignant tumor. Furthermore, contrast-enhanced T1-weighted MRI findings suggested hemorrhagic ischemic changes. The tumor was in contact with the edematous small intestine, and these findings suggested torsion of a malignant tumor of the small intestine. Since the small intestinal GIST had undergone ischemic necrosis due to torsion, it was revealed only as a mass with no blood flow on Doppler US and as a cystic mass on contrast CT similar to an abscess. If the patient had been diagnosed with an abscess, she would have received conservative treatment with drainage or antibiotics. The tumor was suspected to have torsion based on MRI findings, and thus, the patient underwent an operation. MRI has restrictions and limitations for causes of acute abdomen such as torsion of a small intestinal GIST; thus, CT is usually prioritized, although it takes longer to perform than MRI. Torsion of a small intestinal GIST may be preoperatively diagnosed by combining US, CT, and MRI. Furthermore, in our case, torsion of an ovarian tumor was suspected before laparotomy, but both ovaries were found to be normal retrospectively on MRI. In several reported cases of women with acute abdomen due to GIST, gynecological diseases such as torsion of an ovarian cyst or tubo-ovarian abscess were suspected, and laparotomy was performed by gynecologists[21]. In the case of acute abdomen due to GIST in women, MRI may be useful for confirming both ovaries are normal and excluding gynecological disorders as differential diagnoses in addition to evaluating the tumor.

The curative treatment for GIST is radical resection, and it is essential to ensure a safe surgical excision margin without rupture[29–31]. There is no need for systematic lymph node dissection because of the low frequency of lymph node metastasis[32].

Regarding the completely resected GIST, several risk classifications are made according to the tumor size, mitotic index, primary site, and presence of tumor rupture, which greatly reflects the risk of recurrence[33–35]. According to the modified National Institutes of Health Consensus Criteria, a tumor size greater than 10.0 cm, more than 10 mitoses per 50 high-power microscopic fields, tumor diameter greater than 5.0 cm and mitotic count greater than 5, or tumor rupture is defined as a high estimated risk of recurrence[35]. Three-year adjuvant imatinib for patients with high-risk GIST who have undergone surgery helps improve both recurrence-free survival and overall survival[36, 37]. Recently, adjuvant imatinib therapy has been used in wider indications, including medium-risk tumors larger than 3 cm and primary tumors with rupture or perforation[36, 38].

Learning points of this case report are as follows: 1) 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 GIST cases; 2) torsion of a small intestinal GIST may show characteristic findings on CT and MRI; and 3) immediate surgery is mandatory if torsion of the small intestinal GIST is suspected because the GIST or intestine may become necrotic due to hemorrhagic infarction.

Abbreviations

GIST; gastrointestinal stromal tumor, CT; computed tomography, MRI; magnetic resonance imaging, DWI; diffusion-weighted imaging, ADC; apparent diffusion coefficient

Declarations

Ethics approval and consent to participate: Not applicable.

Consent to publish: Written informed consent was obtained from the patient for publication of this Case Report and any accompanying images. A copy of the written consent is available for review by the Editor of this journal.

Availability of data and materials: The datasets supporting the conclusions of this article are included within the article.

Competing interests: The authors declare that they have no competing interests.

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Authors' contributions: 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.

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Figures

Figure 1

Abdominal ultrasonography findings (A) Abdominal ultrasonography shows a 73×52-mm tumor in the pelvis, which has a mixture of high and low echo densities. (B) No blood flow is observed inside the tumor with color Doppler ultrasonography.

Figure 2

Computed tomography findings (A) Abdominal computed tomography (CT) reveals a 73×62×57-mm low-density tumor in the pelvic cavity. (B) Contrast-enhanced CT reveals an enhanced tumor margin but no contrast effect inside of it.

Figure 3

T2-weighted axial magnetic resonance imaging MRI reveals a heterogeneous tumor with slight hyperintensity and a clearly demarcated, lobulated surface in the pelvis. (A) Axial image, (B) coronal image

Figure 4

Diffusion-weighted magnetic resonance image showing hyperintensity of the tumor

Figure 5

T1-weighted magnetic resonance imaging findings T1-weighted magnetic resonance imaging demonstrates (A) a heterogeneous hypointense tumor inside and faint hyperintensity around the edges of

the tumor. (B, C) After injecting the contrast agents, the inside of the tumor has minimal contrast, and the surrounding capsular structure shows good contrast.

Figure 6

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 degrees at the pedicle (arrow) and shows hemorrhagic necrosis.

Figure 7

Gross findings (after formalin fixative application) (A) The 7.5×5.5×4.5-cm tumor is well-defined, reddish-brown, and uneven. (B) The cut surface is a white to gray/red solid tumor with phyllodes after the formalin fixative was applied.

Figure 8

Histopathological findings Hematoxylin and eosin staining reveals 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 μ m, 200 μ m, and 50 μ m, respectively.

Figure 9

Immunohistochemical analysis Tumor cells are stained positive for (A) CD34, (B) CD117 (C-kit), and (C) DOG1; weakly positive for (D) α -smooth muscle actin; and negative for (E) S200 and (F) desmin. The scale bar indicates 100 μ m.

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