A Rare Case of Acute Abdomen Caused by Perforation of Jejunal Gastrointestinal Stromal Tumor (Gist): A Case Report and Literature Review

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Case Report

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Abstract

Background: Gastrointestinal stromal tumor (GIST) is the most common mesenchymal neoplasm of the gastrointestinal system and it accounts for between 1-3% of all gastrointestinal malignancies. GIST arises most commonly from the stomach in 60-70% of cases. The clinical presentation of acute abdomen, due to GIST perforation, was a particularly rare described complication of this disease. Our case report aimed to present a case of acute abdomen in an elderly female patient, due to jejunal GIST perforation.

Case presentation: A 76-year-old female patient presented with acute abdomen to the emergency surgical unit University Clinical Center of Kragujevac. Computed tomography (CT) scan with intravenous contrast showed pneumoperitoneum, the presence of a 75 x 35 mm irregular fluid collection in the left lower quadrant of the abdomen with air inclusions and signs of small intestine perforations. The patient underwent an emergency laparotomy. Intraoperative findings showed the acute inflammatory reaction of the peritoneum with free bowel contents in the abdominal cavity due to jejunal tumor perforation. Partial resection of the small intestine with tumor was performed followed by intestine anastomosis. Histopathological examination of a specimen revealed a completed resected jejunal gastrointestinal stromal tumor.

CONCLUSIONS: GIST arises from the small intestine in 25-30% of cases, and jejunum in 10% of cases. Perforation of GIST and acute diffuse peritoneal inflammation was rare complication of the disease. Emergency laparotomy and complete resection of tumors are essential.

1. Background

Gastrointestinal stromal tumor (GIST) is the most common mesenchymal neoplasm of the gastrointestinal system and it accounts for between 1-3% of all gastrointestinal malignancies. GIST originates from the interstitial cells of Cajal, which are part of the autonomic nervous system of the gastrointestinal system and they are like pacemaker cells that regulate gastrointestinal motility (1). GIST is a benign tumor in 70-80% of cases. GIST arises most commonly from the stomach (60-70%), followed by the small intestine (20-25%), colon, rectum, esophagus, appendix, omentum, and retroperitoneum (2). Metastatic GIST was found in nearly 50% of cases, with the most common site of metastasis in the liver (65%) followed by the peritoneum (21%) and lymph nodes. GISTs are the most common in persons between the age of 40 to 70 and with similar frequency in men and women (1).

GISTs are diagnosed incidentally due to silent behavior. Intestinal bleeding is the most common symptom in about 40% of GIST cases, while perforation is rarely noticed in GISTs. The clinical presentation of gastrointestinal stromal tumors is rare like abdominal surgery emergencies such as massive gastrointestinal hemorrhage, intestinal obstruction, and tumor perforation. Also, in the literature reviews, acute abdomen due to GIST perforation in the peritoneal cavity is rare (3). The most frequent symptoms of GIST are anemia, weight loss, gastrointestinal bleeding, abdominal pain, and mass-related symptoms (1).

The definitive diagnosis of GIST is based on histopathological examination. CD117 protein (c-kit proto-oncogenic product) protein, a tyrosine kinase growth factor receptor, is the most important and specific immunohistochemical marker for GIST diagnosis (4). Morphologically GISTs can be classified into spindle cell type (70%), epithelioid cell type (20%), and mixed cell type (10%) (1).

The gold standard of GISTs treatment is surgical therapy (completed resection), with a 5-year-survival rate between 48-80% (1).

Here we present a case of a patient with jejunal GIST perforation into the peritoneal cavity with consequential acute diffuse peritonitis.

2. Case Presentation

A 76-year-old female patient presented to the emergency surgical unit University Clinical Center of Kragujevac with acute abdominal pain and abdominal distension. The pain started suddenly, a few hours before admission. The patient had normal bowel habits and no nausea or vomiting complaint. She had a past medical history of urinary bladder carcinoma, arterial hypertension, a stent in the coronary circulation, and a penicillin allergy. On admission, the patient's vital signs (heart rate, blood pressure, respiratory rate, and body temperature) were stable. Physical examination revealed abdominal distension, generalized tenderness, and guarding.

The results of laboratory testing performed when the patient was admitted to the Emergency Department revealed the following values: hemoglobin 134 g/L (normal range, 110-157 g/L), leukocyte count 7.9 x 10^9/L (normal range, 3.7-10.0 x 10^9/L) with 79.10% of polymorphonuclear cells, hyperglycaemia with the value of 10.0 mmol/L (normal range 3.8-6.1 mmol/L), increased C-reactive protein concentration of 59.5 mg/L (normal range, 0.0-5.0 mg/L), decreased sodium concentration with the value of 133 mmol/L (normal range, 137-147 mmol/L) and normal creatinine, blood urea nitrogen, pancreatic amylase, lipase, and potassium level.

Abdominal plain radiography, on admission, showed distention of the bowel without free intraperitoneal air. Abdominal ultrasonography was normal. Computed tomography scan with intravenous contrast revealed pneumoperitoneum and a 75 x 35 mm irregular fluid collection located at the level of the pelvic inlet with air inclusions and with signs of small intestine perforations (Fig. 1).
The patient underwent an emergency laparotomy. Intraoperative findings showed acute diffuse peritonitis with significant contamination of the peritoneal cavity by purulent fluid mixed with enteric content. The tumor was located 150 cm from the Treitz ligament. The size of the tumor was 35 mm with a perforation of tumor size about 10 mm in diameter. Segmentary jejunal resection, including the mass lesion, with clear macroscopical margins, was conducted which was followed by a side-to-side handsewn small intestine anastomosis and irrigation drainage.

The post-operative gastrointestinal motility and oral intake were normal. On the fifth postoperative day, the patient developed basal bilateral pneumonia; we consulted a pulmonologist and chest surgeon and administered antibiotics. On the 17th postoperative day, the patient was discharged in good general condition.

Figure 2A showed a histopathological examination of the specimen that revealed a jejunal gastrointestinal stromal tumor, mixed cell type composed of predominately spindle cell morphology and partly palisaded-vacuolated morphology, with transmural infiltration of the jejunum and high malignancy potential (mitotic rate of more than 5 mitosis per 50 high-power field (HPF), and tumor size of 35mm) (Table 1). The immunohistochemical staining revealed that the tumor cells were positive for CD117, DOG1, and vimentin, while negative for S100 protein, smooth muscle actin (SMA), CD99, CD34, LCA (Figs. 2B-2I). Ki-67 proliferation index was at approximately 25% (Fig. 2J). A total of 7 lymph nodes were harvested and none of the lymph nodes was involved by the tumor. Surgical margins were confirmed as clear, and without lymphovascular invasion and perineural invasion. Based on all listed facts the tumor was classified as pT2N0M0 per the American Joint Committee on Cancer's (AJCC) TNM classification. The GIST was determined to be staged IIIb in accordance with the TNM classification and the high-grade mitotic rate (AJCC staging) (Table 1) (5).
<table>
<thead>
<tr>
<th>Risk of malignancy</th>
<th>Size of tumor (cm)</th>
<th>Mitotic counts (/50 high power fields)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Very low</td>
<td>&lt; 2</td>
<td>&lt; 5 / 50</td>
</tr>
<tr>
<td>Low</td>
<td>2–5</td>
<td>&lt; 5</td>
</tr>
<tr>
<td>Intermediate</td>
<td>&lt; 5</td>
<td>6–10</td>
</tr>
<tr>
<td></td>
<td>5–10</td>
<td>&lt; 5</td>
</tr>
<tr>
<td>High</td>
<td>&gt; 5</td>
<td>&gt; 5</td>
</tr>
<tr>
<td></td>
<td>&gt; 10</td>
<td>Any counts</td>
</tr>
<tr>
<td></td>
<td>Any size</td>
<td>&gt; 10</td>
</tr>
</tbody>
</table>

TNM staging of GISTs (5)

<table>
<thead>
<tr>
<th>T</th>
<th>N</th>
<th>M</th>
</tr>
</thead>
<tbody>
<tr>
<td>T0 = no evidence of tumor</td>
<td>N0 = no regional lymph node metastasis or unknown</td>
<td>M0 = no distant metastasis</td>
</tr>
<tr>
<td>T1 = ≤ 2cm</td>
<td>N1 = regional lymph node metastasis</td>
<td>M1 = distant metastasis</td>
</tr>
<tr>
<td>T2 = 2.1cm–5.0cm</td>
<td>N0</td>
<td>M0</td>
</tr>
<tr>
<td>T3 = 5.1cm–10cm</td>
<td>N0</td>
<td>M0</td>
</tr>
<tr>
<td>T4 = &gt;10cm</td>
<td>M0</td>
<td>M0</td>
</tr>
</tbody>
</table>

AJCC staging of jejunal GISTs (5)

<table>
<thead>
<tr>
<th>AJCC Stage</th>
<th>TNM</th>
<th>Mitotic rate</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>T1 or T2</td>
<td>Low (≤ 5/50 HPF)</td>
</tr>
<tr>
<td></td>
<td>N0</td>
<td></td>
</tr>
<tr>
<td></td>
<td>M0</td>
<td></td>
</tr>
<tr>
<td>II</td>
<td>T3</td>
<td>Low (≤ 5/50 HPF)</td>
</tr>
<tr>
<td></td>
<td>N0</td>
<td></td>
</tr>
<tr>
<td></td>
<td>M0</td>
<td></td>
</tr>
<tr>
<td>IIIa</td>
<td>T1</td>
<td>High (&gt; 5/50 HPF)</td>
</tr>
<tr>
<td></td>
<td>N0</td>
<td></td>
</tr>
<tr>
<td></td>
<td>M0</td>
<td></td>
</tr>
<tr>
<td></td>
<td>T4</td>
<td>Low (≤ 5/50 HPF)</td>
</tr>
<tr>
<td></td>
<td>N0</td>
<td></td>
</tr>
<tr>
<td></td>
<td>M0</td>
<td></td>
</tr>
<tr>
<td>IIIb</td>
<td>T2</td>
<td>High (&gt; 5/50 HPF)</td>
</tr>
<tr>
<td></td>
<td>N0</td>
<td></td>
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<tr>
<td></td>
<td>M0</td>
<td></td>
</tr>
<tr>
<td></td>
<td>T3</td>
<td>High (&gt; 5/50 HPF)</td>
</tr>
<tr>
<td></td>
<td>N0</td>
<td></td>
</tr>
<tr>
<td></td>
<td>M0</td>
<td></td>
</tr>
<tr>
<td></td>
<td>T4</td>
<td>High (&gt; 5/50 HPF)</td>
</tr>
<tr>
<td></td>
<td>N0</td>
<td></td>
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<tr>
<td></td>
<td>M0</td>
<td></td>
</tr>
</tbody>
</table>
The patient consulted with an oncology council for malignancies of the digestive tract, and follow-up without treatment was planned. Abdominal computed tomography was performed on the 12th month of the follow-up and the patient was without evidence of recurrence and systemic dissemination of the disease.

3. Discussion And Conclusions

GIST is the most common mesenchymal neoplasm of the gastrointestinal system that occurs in adults more than 40 years of age, where they peak between 60 and 65 years. Males are affected more than females without any geographic or ethnic relation (2). About 60–70% of GISTs occur in the stomach, 25–35% in the small intestine, and 10% in the jejunum (6). The patient's presentation of a perforated GIST of jejunum is rare. A few cases have been reported in the English medical literature associated with the perforation of GIST in the jejunum (Table 2) (7–12).
Table 2
Summary of perforated gastrointestinal stromal tumor (GIST) of jejunum in the english medical literature

<table>
<thead>
<tr>
<th>Authors</th>
<th>Year</th>
<th>Age</th>
<th>Sex</th>
<th>Symptoms</th>
<th>Location (distance from Treitz’ ligament)</th>
<th>Size (cm)</th>
<th>Intraoperative findings</th>
<th>Mitotic count in high-power fields (HPF)</th>
<th>Ki67%</th>
<th>Treatment+ adjuvant therapy (duration)</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Karagülle et al. (8)</td>
<td>2008</td>
<td>70</td>
<td>M</td>
<td>Right-sided abdominal pain, inappetence, nausea, and bloating</td>
<td>Jejunumum (5cm)</td>
<td>5</td>
<td>Abscess</td>
<td>0/50</td>
<td>N/A</td>
<td>SR</td>
<td>13 months ANED</td>
</tr>
<tr>
<td>Ku MC et al (7)</td>
<td>2010</td>
<td>33</td>
<td>F</td>
<td>Acute abdominal pain</td>
<td>Jejunum (multiple tumors)</td>
<td>6.5×5.3×3.9 (largest one)</td>
<td>Peritonitis</td>
<td>N/A</td>
<td>N/A</td>
<td>SR</td>
<td>N/A</td>
</tr>
<tr>
<td>Feng et al. (10)</td>
<td>2011</td>
<td>49</td>
<td>M</td>
<td>Paroxysmal than persistent left abdominal pain</td>
<td>Jejunumum (40cm)</td>
<td>10x8</td>
<td>Peritonitis</td>
<td>&lt; 5/50</td>
<td>&lt; 5</td>
<td>SR</td>
<td>N/A</td>
</tr>
<tr>
<td>Memmi et al. (9)</td>
<td>2012</td>
<td>59</td>
<td>M</td>
<td>Acute abdominal pain</td>
<td>Jejunumum (150cm)</td>
<td>12x10x9</td>
<td>Peritonitis</td>
<td>7/50</td>
<td>8</td>
<td>SR</td>
<td>N/A</td>
</tr>
<tr>
<td>Shoji et al. (11)</td>
<td>2013</td>
<td>61</td>
<td>M</td>
<td>Sudden abdominal pain and nausea</td>
<td>Jejunumum (40cm)</td>
<td>9x7</td>
<td>Peritonitis</td>
<td>0/50</td>
<td>1</td>
<td>SR + Imatinib (36 monts)</td>
<td>36 months ANED</td>
</tr>
<tr>
<td>Misawa et al. (12)</td>
<td>2014</td>
<td>70</td>
<td>M</td>
<td>Symptoms of fever and abdominal pain</td>
<td>Jejunumum (near Treitz’s ligament)</td>
<td>10 × 10</td>
<td>Peritonitis</td>
<td>N/A</td>
<td>26</td>
<td>SR + Imatinib</td>
<td>12 months ANED</td>
</tr>
<tr>
<td>Alessiani et al. (3)</td>
<td>2014</td>
<td>82</td>
<td>M</td>
<td>Fever, vomiting, diarrhea and diffuse abdominal pain</td>
<td>Jejunumum (10cm)</td>
<td>7 × 5</td>
<td>Peritonitis</td>
<td>16/50</td>
<td>15</td>
<td>SR + Imatinib</td>
<td>6 months ANDE</td>
</tr>
<tr>
<td>Sato et al. (6)</td>
<td>2017</td>
<td>74</td>
<td>M</td>
<td>Vomiting and abdominal pain</td>
<td>Jejunumum (100cm)</td>
<td>14</td>
<td>Peritonitis</td>
<td>N/A</td>
<td>N/A</td>
<td>SR + Imatinib (3 monts)</td>
<td>22 months</td>
</tr>
<tr>
<td>Menesesa et al. (5)</td>
<td>2020</td>
<td>46</td>
<td>M</td>
<td>Left upper quadrant pain with fevers and chills</td>
<td>Jejunumum (10cm)</td>
<td>13×6×7.5</td>
<td>Peritonitis</td>
<td>&lt; 5/50</td>
<td>10</td>
<td>SR</td>
<td>N/A</td>
</tr>
<tr>
<td>Al-Swaiti et al. (2)</td>
<td>2020</td>
<td>59</td>
<td>M</td>
<td>Severe generalized abdominal pain</td>
<td>Jejunumum (mild)</td>
<td>11 × 9</td>
<td>Peritonitis</td>
<td>8/50</td>
<td>N/A</td>
<td>SR + Imatinib</td>
<td>N/A</td>
</tr>
<tr>
<td>Our case</td>
<td>2022</td>
<td>76</td>
<td>F</td>
<td>Acute abdominal pain and abdominal distention</td>
<td>Jejunumum (150cm)</td>
<td>25</td>
<td>Peritonitis</td>
<td>&gt; 5/50</td>
<td>25</td>
<td>SR</td>
<td>12 months ANED</td>
</tr>
</tbody>
</table>

**Abbreviations:** SR, segmental jejunal resection; N/A, not available; ANED, alive with no evidence of disease
It is important to note that a GIST expresses the c-kit (CD117) oncoprotein which is a mutated receptor of tyrosine kinase (13). Another mechanism for how GISTs become malignant is by an oncogenic mutation in platelet-derived growth factor receptor-α (14).

The primary GISTs symptoms are variable according to the site and size of the tumor. Approximately one-third of patients with GIST are asymptomatic. The most common symptom of jejunal GIST is vague, nonspecific abdominal pain accompanied by early satiety or abdominal fullness (20–50% of cases). Gastrointestinal bleeding is present in half of the patients with jejunal GIST due to pressure necrosis by tumor and ulceration of the overlying mucosa (1). Acute abdominal symptoms were more frequent in patients with jejunal and ileal than in gastric GISTs (15).

Jejunal GISTs seldom cause perforation and acute diffuse peritonitis (6). GISTs perforation lowers the five-year survival to 24%, probably due to the peritoneal dissemination of tumor cells (1). In the literature, three types of GIST rupture have been described: closed perforation (abscess type), hemoperitoneum leading to rupture of the hematoma capsule in the tumor (hemoperitoneum type), and free perforation (bowel perforation type) (6). The last type, bowel perforation type, that is the rarest type, may be caused by an obstruction with increasing intraluminal pressure or tumor erosion that leads to mural necrosis (1, 16). Other described mechanisms of perforation are tumor embolization of intestinal blood vessels and consequent ischemia, and replacement of the bowel wall by tumor cells followed by necrosis (8). However, GISTs originating from the jejunum rarely cause perforation.

Diagnostic procedures that include several examinations, like barium examination of the gastrointestinal system, computed tomography, and angiography, none of them can establish the 100% correct diagnosis of GIST (17). Notably, magnetic resonance imaging (MRI) brings better information than CT in preoperative settings. Approximately one-third of GISTs were incidental discovered (1).

Histopathological diagnosis of GIST depends on the morphological type of tumor cells and immunohistochemistry. According to morphology, there are three types of tumor cells: spindle cell type (70%), epithelioid cell type (20%), and mixed cell type (10%). Almost all of the GISTs are positive for c-kit (CD117) and DOG1, and 60–70% are positive for CD34 on immunohistochemistry (2, 18). Further, 30–40% of GISTs are positive for SMA, 10% positive for vimentin, and 5% positive for S100 protein (19, 20). All of the listed markers were tested for in this case.

Prognostic factors of possible malignant potential of GISTs include mitotic activity (more than 5 mitosis/50 HPF) and the size of the tumor (more than 5 cm). Ki67 index of more than 22% is the most powerful predictor of poor survival (6).

The treatment of GIST has undergone a substantial transformation over the last decades. Surgical resection is considered the only potentially curative option for patients with localized GISTs. About 85% of tumors can be completely resected, and the incidence of recurrence and metastasis after radical surgery is 50% (21). A negative margin is important to prevent the local recurrence of the tumor. Lymph node involvement is rare and lymphadenectomy is not typically indicated (2). Independent adverse prognostic factors are large tumor size, high mitosis count, non-gastric location, male sex, and especially emphasized intraoperative rupture which according to some reports is related to an almost 100% chance of postoperative recurrence (22). Adjuvant chemotherapy with imatinib, the inhibitor of tyrosine kinase, significantly increase overall survival in patients with advanced disease and markedly reduced recurrence after surgery (23).

In summary, we report a case of a female with a perforated GIST in the jejunum causing acute diffuse peritonitis. GIST is rare and its presentation such as spontaneous perforation is especially rare. Preoperative diagnosis of GIST is very difficult despite all diagnostic procedures and the suspicion of GIST is most often made during the surgery. Emergency surgery is the gold standard and should achieve radical resection. The clinical outcome is worse when this tumor presents with bowel perforation and peritonitis due to delayed diagnosis of the disease and peritoneal dissemination of tumor cells.

**Abbreviations**

GIST  
Gastrointestinal stromal tumor  
CT  
Computed tomography  
CD  
Cluster of differentiation  
HPF  
High-power field  
DOG1  
Discovered on GIST-1  
SMA  
smooth muscle actin  
LCA  
Leukocyte common antigen
Declarations

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Ethics approval and consent to participate

Not applicable.

Consent for publication

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 upon request.

Availability of data and materials

Not applicable.

Competing interests

The authors declare that they have no competing interests.

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Authors’ contributions
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None

References


Figures

(A) Axial view of the contrast-enhanced CT scan revealed an irregular fluid collection (75 x 35 mm in size) in the left lower quadrant of the abdomen (white arrows). (B) Coronal and (C) sagittal view identified the fluid collection located in the lower abdomen. CT, computed tomography.
A histological examination of the resected specimen revealed morphology of spindle tumor cells with high mitotic figures (more than 5 per 50 HPF) and somewhat palisaded-vacuolated morphology (Hematoxylin-eosin stain; original magnification x100). Immunohistological results revealed that the tumor CD117 (B), DOG1 (C), Vimentin (D) were positive, while S-100 protein (E), α-smooth muscle actin (SMA) (F), CD99 (G), CD34 (H), and LCA (I) were negative, and the Ki-67 (J) was 25% (original magnification x40).