

A rare perforated gastrointestinal stromal tumor in the jejunum: A case report

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We herein report a 45-year-old Chinese male with a perforated jejunal gastrointestinal stromal tumor causing acute diffuse peritonitis and incomplete intestinal obstruction. At urgent laparotomy, a perforated tumor was found in the jejunum about 40 cm from the Treitz's ligament, and the upper small intestine was twisted 1080°. The clinical symptom of this case is extremely rare, and has not been reported in the literature previously.

Key words: Gastrointestinal stromal tumor, perforation, peritonitis, intestinal obstruction

Nadir görülen jejunumda perfore gastrointestinal stromal tümör vakası: Olgu sunumu

Burada, 45 yaşında Çinli erkek hastada jejunumda perforasyona, akut yaygın peritonite ve parsiyel intestinal obstrüksiyona neden olan gastrointestinal stromal tümör vakası sunulmuştur. Acil olarak gerçekleştirilen laparatomide, Treitz ligamanından yaklaşık 40 cm uzaklıkta jejunumda perfore tümör beraberinde proksimal ince barsaklarda 1080° torsiyon tespit edildi. Hastanın bu klinik bulguları oldukça nadirdir ve literatürde daha önce rapor edilmemiştir.

Anahtar kelimeler: Gastrointestinal stromal tümör, perforasyon, peritonit, intestinal obstrüksiyon

INTRODUCTION

Gastrointestinal stromal tumor (GIST) is the most common mesenchymal tumor arising from the GI tract (1-3). It is widely accepted that a GIST expresses the c-kit (CD117) oncoprotein or has a mutation in either the c-kit or the platelet-derived growth factor receptor- α , which is considered as a highly specific marker differentiating GIST from other mesenchymal tumors (4,5).

GIST is usually associated with abdominal pain, palpable mass, or GI bleeding, accompanied by fever, anorexia, weight loss, or anemia (6,7). However, GIST originating from the jejunum rarely causes perforation. Herein, we describe the extremely rare case of a perforated GIST in the jeju-

num causing acute diffuse peritonitis and incomplete intestinal obstruction, and we review the associated literature.

CASE REPORT

A 45-year-old Chinese male was admitted to the Department of General Surgery with paroxysmal left abdominal pain for 3 days, and persistent pain for 8 hours. There was no history of vomiting, fever, chill, jaundice, or GI bleeding. On admission, his vital signs (heart rate, blood pressure, respiratory rate, and body temperature) were stable. On physical examination, abdominal palpation revealed tenderness of the abdominal wall in the

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Figure 1. CR showed intestinal expansion in the right upper quadrant and several air-fluid levels in the upper abdomen.

middle and left lower quadrant, and a hard mass about 10 cm x 8 cm was palpable in the lower middle abdomen with minimal mobility. He denied any history of blood transfusion, alcohol abuse or medications. Laboratory data were normal, except the white blood cell (WBC) count was 18880/ μ L [normal range (NR): 4000-9000/ μ L]. The

liver and renal function tests were normal. Serological tests for hepatitis B and C viruses were negative, but the test for syphilis was positive. Computed radiography (CR) showed expansion of the small intestine in the right upper quadrant and several air-fluid levels in the upper abdomen (Figure 1). Computed tomography (CT) scan revealed an approximately 10 cm x 8 cm, non-uniformly enhanced, hard tissue mass with some air in the jejunum, partial adhesion between the tumor and bladder (Figure 2A), and expansion of the small intestine in the right middle abdomen (Figure 2B).

At urgent laparotomy, an approximately 10 cm x 8cm pedunculated solid tumor was found in the jejunum, which was 40 cm from the Treitz's ligament (Figure 3A). The tumor was perforated, with the hole measuring approximately 2 cm (Figure 3B), and the upper small intestine was twisted 1080° (Figure 3C). Partial resection of the jejunum was performed, and also partial resection of the bladder because of the partial adhesion (Figure 3D). Histological examination of the resected specimen revealed interlaced bundles of spindle-like tumor cells with low mitotic figures (less than 5 per 50 high-power field [HPF]). There were tumor cells in the subserosa, but no metastasis in the mesenteric lymph node (Figure 4A). Immunohistochemical results revealed that the tumor Vim, CD117, CD34, and α -smooth muscle actin (SMA)

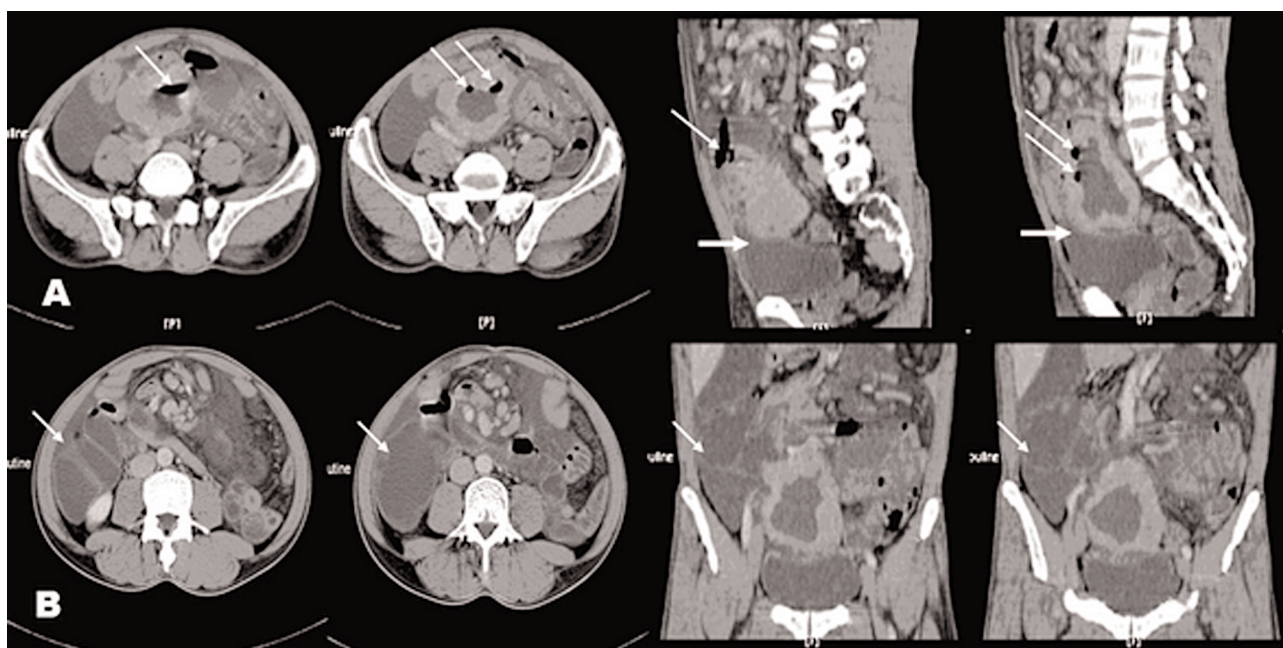


Figure 2. A. CT images at four different levels demonstrated an approximately 10 cm x 8 cm, non-uniformly enhanced, hard tissue mass with some air (white arrows) adjacent to the jejunum and adhesion between the tumor and bladder (bold arrows). B. CT scan at four different levels revealed intestinal expansion (white arrows) in the right middle abdomen.

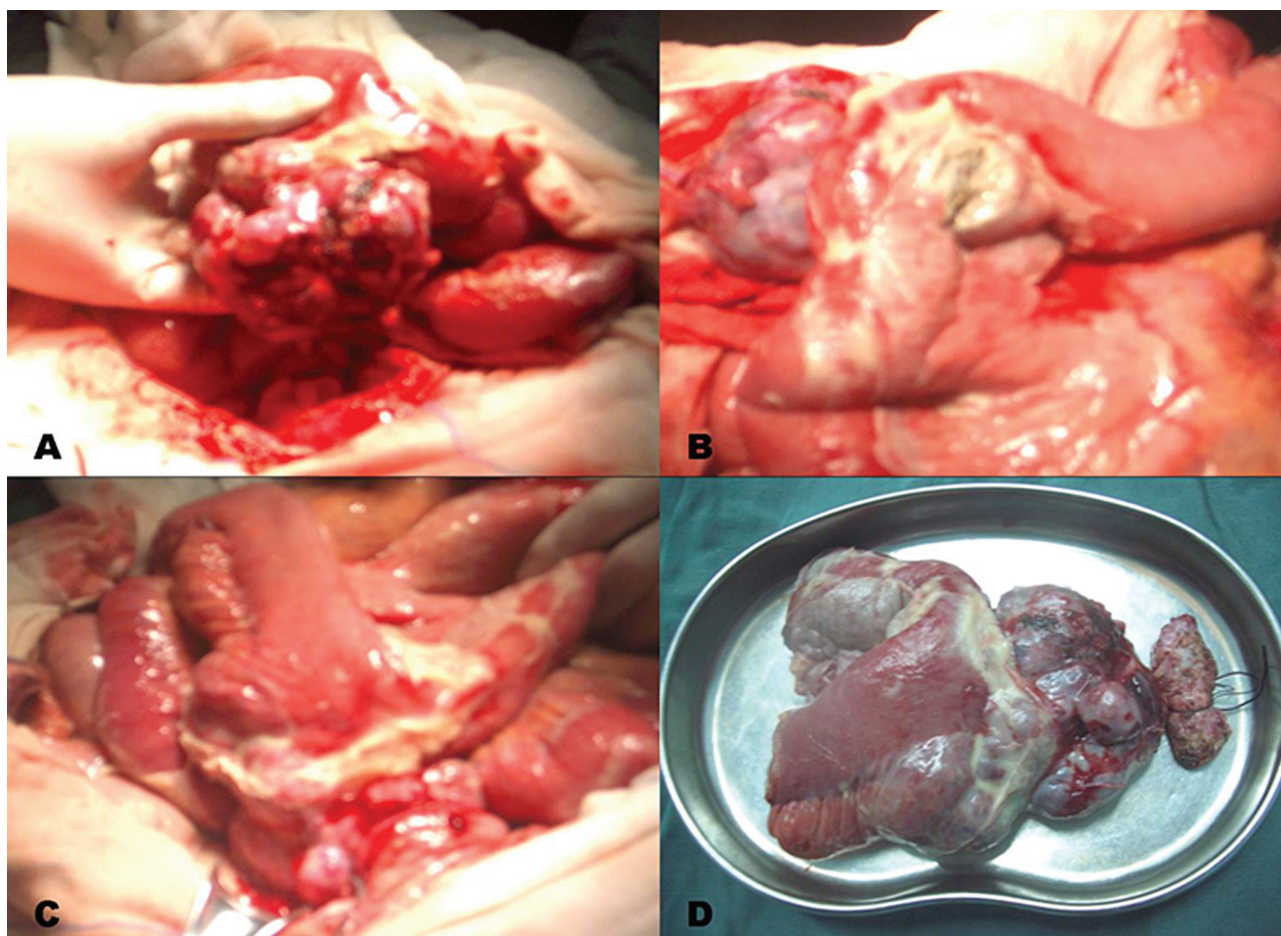


Figure 3. A. An approximately 10 cm x 8 cm pedunculated solid tumor was found in the jejunum, 40 cm from the Treitz's ligament. B. The tumor was perforated in the jejunum, with a hole measuring approximately 2 cm, and the GI contents effused via the hole. C. The intestinal obstruction caused upper small intestine twisting of 1080°. D. Partial resection of the jejunum was performed as well as partial resection of the bladder because of the partial adhesion between the tumor and bladder.

were positive, desmin and S-100 protein were negative, and the Ki-67 was <5% (Figure 4B). The tumor was diagnosed as GIST according to the above-mentioned results. The postoperative course was uneventful.

DISCUSSION

GIST is a rare visceral sarcoma arising in the GI tract, and usually occurs in the stomach (60-70%) or small intestine (25-35%). Colon, rectum, appendix (together 5%) and esophagus (2-3%) are rare sites (8). Velasco *et al.* (9) reported that the jejunal GIST accounts for approximately 10% of all cases. There are also sporadic reports of GISTs arising from the omentum, mesentery or retroperitoneum, unrelated to the GI tract, but most of GISTs in these sites are from gastric or intestinal metastases.

GISTs are usually associated with abdominal pa-

in, palpable mass, or GI bleeding, accompanied by fever, anorexia, weight loss, or anemia (6,7). Approximately 10-30% of patients with GISTs may be asymptomatic (10). However, GISTs originating from the jejunum rarely cause perforation, and also rarely cause acute diffuse peritonitis.

An extensive review of the English-language literature regarding GISTs revealed only one report about perforation of the tumor (11). In our case, the perforated tumor was transmural growth, and there were specific intraluminal mucosal changes in the jejunum. The perforated tumor communicated with the jejunum, and then the GI contents effused via the hole of the tumor causing acute diffuse peritonitis. The perforated tumor also induced incomplete intestinal obstruction causing the upper small intestine to twist 1080°. These atypical clinical symptoms are extremely rare, and have not been reported in the literature previously.

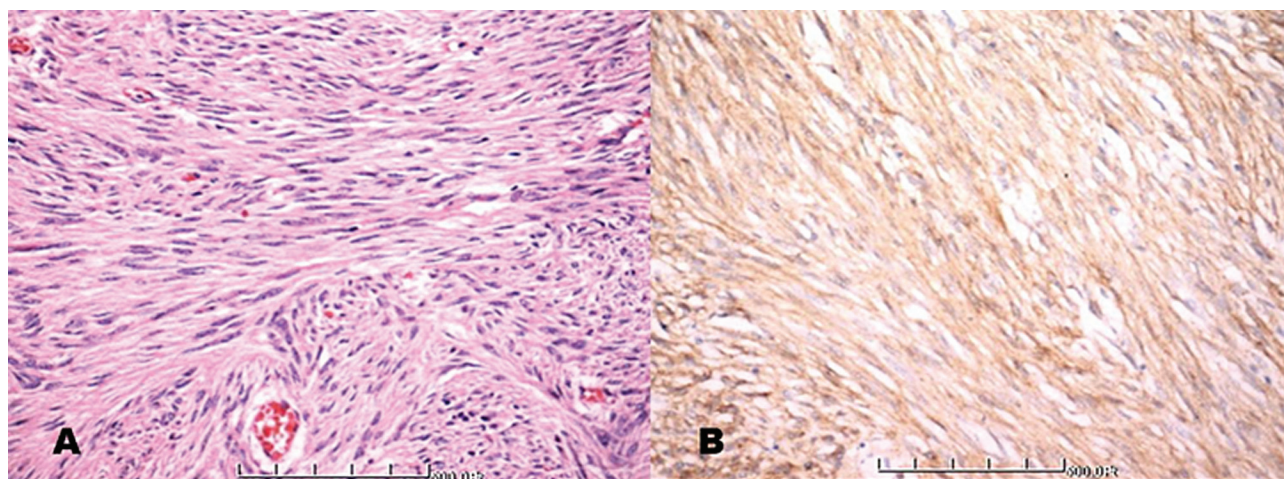


Figure 4. **A.** Histological examination of the resected specimen revealed interlaced bundles of spindle-like tumor cells with low mitotic figures (less than 5 per 50 HPF) (Hematoxylin-eosin stain; original magnification x200). **B.** Immunohistological results revealed that the tumor Vim, CD117, CD34, and α -smooth muscle actin (SMA) were positive, desmin and S-100 protein were negative, and the Ki-67 was <5% (Hematoxylin-eosin stain; original magnification x200).

It was difficult to diagnose a GIST because of the extremely rare clinical symptoms above, and it was also difficult to differentiate the tumor on images. CT scan is, however, more widely available and is currently the imaging modality of choice for patients with suspected abdominal mass of GISTs in the omentum and mesentery (12). For those with known or suspected GISTs, a dedicated magnetic resonance imaging (MRI) provides better information than CT in the preoperative staging workup (13). However, all of the relative usefulness of images depends on the site of the GISTs. In the absence of the typical imaging findings, it is difficult to make a decisive diagnosis, and the final diagnosis needs to rely on the pathological and immunological tests. Accordingly, in our case, we first considered the mass as a small intestinal tumor originating in the jejunum because of the well-enhanced characteristics of the tumor on CT images and the rare clinical symptoms. At the laparotomy, when we observed the perforated tumor communicating with the jejunum, we understood

the reason for the acute diffuse peritonitis. The contents of the tumor as well as the GI contents of the jejunum effused via the hole of the tumor, and the GI contents caused the acute diffuse peritonitis. The preoperative diagnosis was not clear, and the final diagnosis was based on the pathological and immunological tests. Optimal surgical treatment of GISTs entails complete removal of the tumor with clear surgical margins including the adjacent involved organs, because local and regional lymph node involvement is infrequent in GISTs (14,15). However, in our case, although peritoneal metastasis was not seen, we will pay attention to the possible recurrence of the tumor in view of its perforation.

In conclusion, we report the case of a male with a perforated GIST in the jejunum causing acute diffuse peritonitis. It should be kept in mind that if there is an abdominal mass with the symptom of acute diffuse peritonitis, the possibility of GIST should be considered because GIST can perforate and cause acute diffuse peritonitis.

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