

Acute Abdomen Syndrome Caused by a Perforated Gastrointestinal Stromal Tumor – A Case Report

Taner Kivilcim¹, Fatih Altintoprak^{2,*}, Feyyaz Onuray¹, Serbulent Gokhan Beyaz³, Muzaffer Yildirim⁴ and Osman Nuri Dilek²

¹Department of General Surgery, Sakarya University Research and Educational Hospital, Turkey

²Department of General Surgery, Sakarya University Faculty of Medicine, Turkey

³Department of Anesthesiology, Sakarya University Faculty of Medicine, Turkey

⁴Department of Pathology, Sakarya University Faculty of Medicine, Turkey

Abstract: Gastrointestinal stromal tumors (GIST) are frequently observed in the stomach and intestines. Although they may be presented with various symptoms depending on their location and diameter, acute abdomen syndrome is a rare presentation form for GIST. A 54-year-old male patient was evaluated with the complaint of sudden onset abdominal pain at the urology clinic. In examination; fever (38.5 °C), abdominal distension and peritoneum irritation signs were detected. A mass that contained localized necrotic areas thought to have mesentery origins and intraabdominal free-fluid were detected on abdominal computed tomography. The patient was operated with the diagnosis of acute abdomen syndrome. In operation; extensive intestinal content in the abdomen, a perforated tumoral mass which was 7x5 cm in diameter, and anti-mesenterically localized at the ileum segment were detected. Segmental ileum resection-end ileostomy operation was performed, and the patient was discharged as uneventful on post-operative tenth day. As a result of a histopathological analysis, a GIST (<5/50 mitotic growth) was detected. By means of this finding, we want to remind that GISTs may be presented with acute abdomen symptoms due to perforation, though rarely.

Keywords: Stromal tumor, Perforation, Acute abdomen syndrome.

INTRODUCTION

Gastrointestinal stromal tumours (GIST) are the most common mesenchymal tumours of the gastrointestinal system. Many gastrointestinal soft tissue neoplasms, previously classified as different sub-groups, are today classified as GIST on the basis of molecular and immunohistological features. GISTs occur anywhere along the gastrointestinal tract and are believed to originate from the interstitial cells of Cajal [1]. These tumors account for 0.1–3% of all gastrointestinal cancers, and they represent up to 20% of small bowel malignancies [2]. Clinical symptoms, which depend on tumour size and location, are usually non-specific. In daily practice, presentation with acute abdomen symptoms is rare. In this paper, we have reported a perforated GIST case who had admitted with acute abdomen symptoms.

CASE REPORT

A 54-year-old male patient hospitalized in the urology clinic for the diagnosis of a mass adjacent to the urinary bladder was evaluated for complaints of

abdominal pain and nausea-vomiting, which suddenly began 8 hours prior. At the time of the first evaluation, the patient was pale, with tachycardia and fever (38.5 °C). On examination revealed abdominal distention, generalized tenderness, and signs of peritoneal irritation. Laboratory examinations revealed leukocytosis (17.500/mm³) and anemia (Hb < 8.0 gr/dL). The patient had Type II Diabetes Mellitus and his medical history revealed that he occasionally experienced abdominal pain for the previous year and was found to have anemia during the investigations performed for this complaint. The findings of the endoscopic examinations of the lower and upper gastrointestinal tracts performed to investigate the etiology of anemia were normal. On abdominal graphy revealed air-fluid levels. Abdominal computed tomography (CT) examination revealed extensive intra-abdominal free fluid and a 7x5 cm mass with a necrotic center, which was laterally adjacent to the urinary bladder and believed to originate from the ileum (Figure 1). The patient underwent emergency surgery with the diagnosis of acute abdomen syndrome. During abdominal exploration, extensive intestinal content in the abdominal cavity and all intestinal segments until the terminal ileum were dilated. A 7x5 cm mass originating in the anti-mesenteric border in the ileal segment at 50 cm proximal to the ileocecal valve was discovered, and a perforation occurred at this location (Figure 2). Segmental ileum resection-end ileostomy

*Address correspondence to this author at the Department on General Surgery, Sakarya University Faculty of Medicine, Turkey; Tel: +905335483415; Fax: +902642550000; E-mail: fatihaltintoprak@yahoo.com

This case has been presented at ESS 2012 ISTANBUL, XVI. Annual Meeting of the European Society of Surgery, 2012, Istanbul, Turkey, as poster presentation.

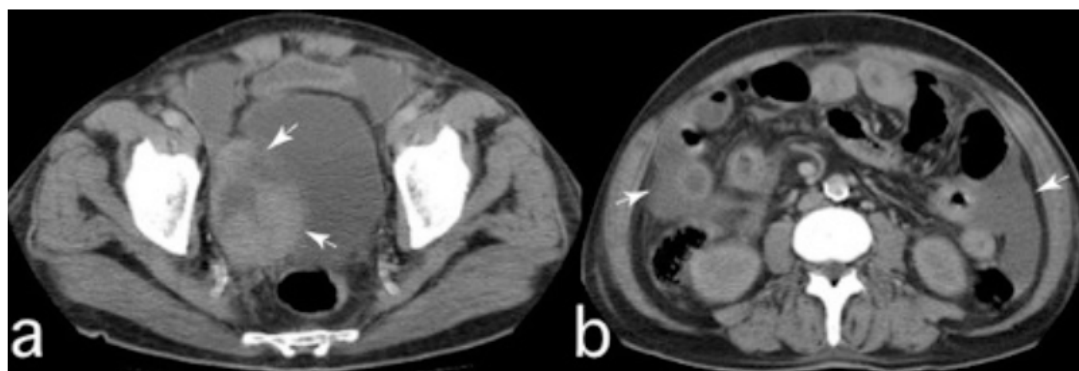


Figure 1: Abdominal CT examination; **a)** pelvic sections showing a mass (arrows) laterally adjacent to the right margin of the urinary bladder with partially smooth margins and central necrotic areas, which, on first sight, suggests an origin in the bladder wall. **b)** Abdominal CT sections of the same patient showing the presence of free fluid between the segments of the small intestine and in bilateral paracolic areas (arrows).

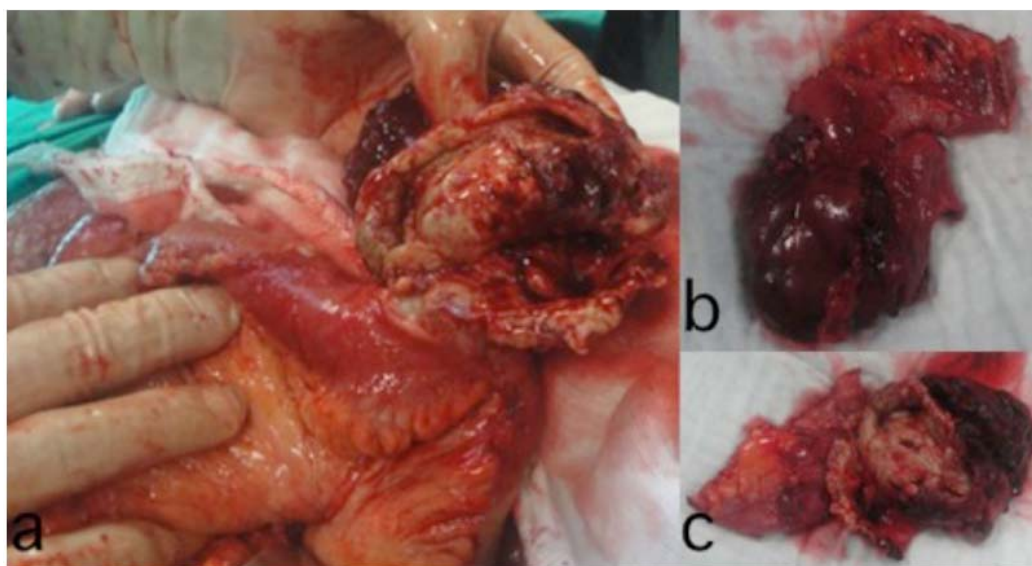


Figure 2: Intra-operative view; **a)** Showing a perforated tumor mass originating from the anti-mesenteric border of the ileum, **b** and **c)** Anterior and posterior view of the mass after resection.

operation was performed and the patient was discharged uneventfully on postoperative 10th day. Histopathological analysis revealed a GIST (mitotic activity <5/50) (Figure 3). The patient is currently in the 9th month of his follow-up and has no problems.

DISCUSSION

In a 2005 United States report, the distribution of small bowel cancer histology was: 44% carcinoid, 33% adenocarcinoma, 15% lymphoma, and 7% gastrointestinal stromal tumor (GIST) [3]. GISTs are most commonly observed in the stomach and small intestine in the gastrointestinal system. Their clinical signs vary depending on their anatomical location and size. Tumors with small diameters are usually asymptomatic. Large tumors localized in the small intestine, however, are commonly present with signs of

intestinal obstruction and palpable intra-abdominal masses [4]. Despite the rarity, GIST presented with acute abdomen symptoms have been reported in literature [5,6]. At the time of the first evaluation, the patient in the current study had abdominal distention and signs of peritoneal irritation. The patient had no history of clinical complaints suggesting intestinal obstruction, despite the presence of a mass of approximately 7x5 cm in diameter, likely due to the growth of the mass towards the outside of the lumen.

Gastrointestinal bleeding associated with mucosal ulceration is a common finding in GISTs [2,7]. Twenty percent of the cases have complaints of non-specific abdominal pain, nausea and loss of appetite associated with the intra-abdominal mass. Ten percent of the patients have metastatic disease at the time of presentation [7]. The history of the current patient

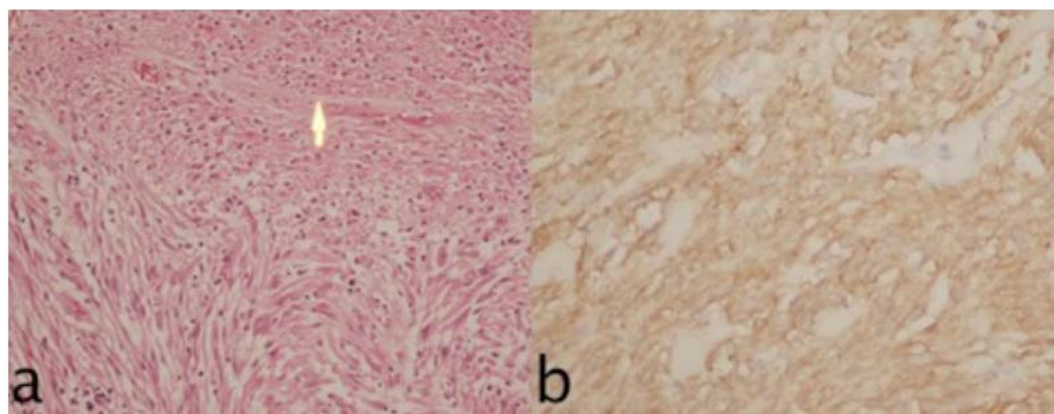


Figure 3: Microscopic view; **a)** (H&E stain x20) Spindle tumor cells with eosinophilic cytoplasm. Necrosis is seen in the area shown with arrows. **b)** (x40) CD 117-positive tumor cells.

revealed abdominal discomfort and anemia for the previous year and no weight loss. The endoscopic examinations performed to investigate the etiology of anemia revealed normal results due to the ileal localization of the mass.

Radiological and endoscopic methods are used in the diagnosis of GISTs. Gastrointestinal endoscopy, endoscopic ultrasonograph, abdominal CT, magnetic resonance imaging and FDG-PET are methods that may be used for diagnosis. Abdominal CT is commonly used for preoperative diagnosis, staging, surgical planning, and treatment follow-up. In general, GISTs are seen as intraluminal or extraluminal masses with smooth contours and homogenous internal structures. Large lesions, however, may appear as masses with irregular contours and a heterogeneous internal structure [8]. The abdominal CT examination of the current patient revealed a lesion with a smooth contour and central necrosis, which was localized in the ileum.

The diagnosis of GIST can only be made by histopathological and immunohistochemical examinations. However, it should be noted that preoperative biopsy is not recommended for surgically resectable masses. The rate of KIT (CD117) positivity is 95% in GISTs [9]. In rare cases with KIT negativity (5%), the experience of the evaluating pathologist is very important along with other immunohistochemical markers (CD34, SMA, S-100, Desmin). In this case, extensive cytoplasmic and membrane staining with CD117 was observed in the tumor cells.

Today, the standard treatment modality for non-metastatic GIST is total surgical excision. Achieving R0 resection (absence of residual disease) is the most important factor affecting treatment outcomes (disease-free interval and survival) [10]. Frozen-section

examination can be used in negative surgical border evaluation. Attention should be paid to not perforate the tumor during operation. This could cause neoplastic spread, and is a factor that increases the risk of the disease recurring, and decreases life expectancy. In the current case, a frozen-section examination was not required since the patient already had tumor perforation during the operation.

GISTs are tumors with malignant potential. In general, small tumors (<2 cm) are considered benign, while the diameter, mitotic index, and localization of the tumor are important factors affecting prognosis. Tumors >5 cm in diameter and/or mitotic count >5/50 HPF mitoses are potentially malignant. GISTs are tumors that are resistant to radiotherapy and chemotherapy. Although it is possible to achieve surgical R0 resection in small tumors, there were no treatment options in cases with large tumors and metastatic tumors until the discovery of the efficacy of imatinib in GIST. Currently, imatinib is the treatment of choice in patients who are not candidates for surgery due to technical reasons (the presence of a large tumor or metastasis) or patient-related reasons (high surgical risk) [11,12]. Because the patient in this study had tumor perforation, imatinib treatment was administered during the postoperative period.

In conclusion, the localization and size of the tumor is important in the clinical presentation of GISTs. Although GISTs are generally associated with non-specific symptoms, it should be kept in mind that they may also present with acute abdomen symptoms by causing perforation, regardless of tumor size.

CONFLICT OF INTEREST

None.

REFERENCES

- [1] Kindblom LG, Remotti HE, Aldenborg F, Meis-Kindblom JM. Gastrointestinal pacemaker cell tumor (GIPACT): gastrointestinal stromal tumors show phenotypic characteristics of the interstitial cells of Cajal. *Am J Pathol* 1998; 152(5): 1259-69.
- [2] Blanchard DK, Budde JM, Hatch GF 3rd, Wertheimer-Hatch L, Hatch KF, Davis GB, *et al.* Tumors of the small intestine. *World J Surg* 2000; 24(4): 421-29.
<http://dx.doi.org/10.1007/s002689910067>
- [3] Bilimoria KY, Bentrem DJ, Wayne JD, Ko CY, Bennett CL, Talamonti MS. Small bowel cancer in the United States: changes in epidemiology, treatment, and survival over the last 20 years. *Ann Surg* 2009; 249(1): 63-71.
<http://dx.doi.org/10.1097/SLA.0b013e31818e4641>
- [4] Van der Zwan SM, DeMatteo RP. Gastrointestinal stromal tumor: 5 years later. *Cancer* 2005; 104(9): 1781-88.
<http://dx.doi.org/10.1002/cncr.21419>
- [5] Badger SA, Yousaf M, Campbell WJ. A case of a gastrointestinal stromal tumour presenting as acute abdomen. *Ir J Med Sci* 2005; 174(3): 84-5.
<http://dx.doi.org/10.1007/BF03169154>
- [6] Efremidou EI, Liratzopoulos N, Papageorgiou MS, Romanidis K. Perforated GIST of the small intestine as a rare cause of acute abdomen: Surgical treatment and adjuvant therapy. Case report. *J Gastrointest Liver Dis* 2006; 15(3): 297-9.
- [7] Bucher P, Villiger P, Egger JF, Buhler LH, Morel P. Management of gastrointestinal stromal tumors: from diagnosis to treatment. *Swiss Med Wkly* 2004; 134(11-12): 145-53.
- [8] Chourmouzi D, Sinakos E, Papalavrentios L, Akriviadis E, Drevelegas A. Gastrointestinal stromal tumors: a pictorial review. *J Gastrointest Liver Dis* 2009; 18(3): 379-83.
- [9] Dupart J, Zhang W, Trent JC. Gastrointestinal stromal tumor and its targeted therapeutics. *Chin J Cancer* 2011; 30(5): 303-14.
<http://dx.doi.org/10.5732/cjc.011.10062>
- [10] Valadao M, Linhares E. The role of the surgeon in the management of GIST. *Rev Col Bras Cir* 2009; 36(3): 261-65.
- [11] Joensuu H, Roberts PJ, Sarlomo-Rikala M, Andersson LC, Tervahartiala P, Tuveson D, *et al.* Effect of the tyrosine kinase inhibitor STI571 in a patient with a metastatic gastrointestinal stromal tumor. *N Eng J Med* 2001; 344(14): 1052-56.
<http://dx.doi.org/10.1056/NEJM200104053441404>
- [12] Blanke CD, Rankin C, Demetri GD, Ryan CW, von Mehren M, Benjamin RS, *et al.* Phase III randomized, intergroup trial assessing imatinib mesylate at two dose levels in patients with unresectable or metastatic gastrointestinal stromal tumors expressing the kit receptor tyrosine kinase: S0033. *J Clin Oncol* 2008; 26(4): 626-32.
<http://dx.doi.org/10.1200/JCO.2007.13.4452>

Received on 13-11-2012

Accepted on 05-12-2012

Published on 14-02-2013

[DOI: http://dx.doi.org/10.6000/1929-2279.2013.02.01.2](http://dx.doi.org/10.6000/1929-2279.2013.02.01.2)