Multifocal intestinal stromal tumors with jejunal perforation and intra-abdominal abscess: Report of a case

Jejunal perforasyon ve karın içi abseye neden olan multifokal stromal intestinal tümör: Olgu sunumu

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A 70-year-old man with right-sided abdominal pain, inappetence, nausea, and bloating of one-week duration was admitted to our Emergency Department. The abdominal computed tomographic scan showed an abscess (55 x 66 mm) among the intestinal loops. After percutaneous ultrasonography-guided insertion of a drainage catheter into the abscess, pouchography showed that contrast medium had passed into the intestinal lumen via fine tract. The patient underwent surgery because of an intra-abdominal abscess caused by intestinal perforation. During laparotomy, a solid mass measuring 5 cm in diameter on the antimesenteric side of the jejunal wall and a perforation of approximately 0.5 cm were identified after exploration of the abscess cavity. In addition, 3 polypoid lesions (2 at the antimesenteric side of the jejunal wall, and 1 at the antimesenteric side of the ileal wall) were found incidentally. Resection and primary anastomosis of the perforated area and wedge resections of the polypoid lesions were performed. The results of pathologic examination of the all-surgical specimens indicated low-grade gastrointestinal stromal tumors. Since there was no metastasis, all the tumors were resected surgically and all were pathologically low-grade tumors, we did not use imatinib, the long-term results of which are not known at present. Neither complications nor evidence of recurrence were seen during the follow- up of 13 months. Stromal tumors of the small intestine are relatively rare and cause subtle clinical signs and symptoms. Their diagnosis is often delayed until complications develop. Even if complications occur, exact diagnosis is usually very difficult preoperatively.

Key words: Gastrointestinal stromal tumor, small intestine, intestinal perforation, multifocal location

Yetmiş yaşında erkek hasta bir haftadır karın sağ tarafında olan ağrı, iştahsızlık, bulantı ve karında şişkinlik şikayeti ile acil servise başvurdu. Abdominal bilgisayarlı tomografide ince barsak ansları arasında abse (55 x 66 mm) saptandı. Abse içine ultrasonografi eşliğinde perkütan drenaj katateri yerleştirildikten sonra çekilen poşografide kontrast maddenin ince bir trakt ile barsak lümenine geçtiği görüldü. Hasta intestinal perforasyon sonucu gelişen karın içi abse ön tanısıyla ameliyat edildi. Ameliyatta; jejunum duvarında, antimezenterik tarafta 5 cm boyutunda solid kitle saptandı ve abse kavitesi açıldığında yaklaşık 0.5 cm boyutunda jejunal duvarda perforasyon görüldü. Ayrıca rastlantısal olarak 3 adet polipoid lezyon (bunların ikisi jejunum duvarında antimezenterik tarafta diğeri ise ileum duvarında antimezenterik tarafta) bulundu. Perfore alana rezeksiyon ve ucuca anastamoz, diğer polipoid lezyonlara ise wedge rezeksiyon yapıldı. Tüm spesmenler patolojik inceleme sonunda düşük dereceli gastrointestinal stromal tümör tanısı aldı. Tüm lezyonlar cerrahi olarak çıkarıldığı ve düşük dereceli olduğu, metastaz olmadığı için uzun dönem etkisi bilinmeyen imatinib verilmedi. Hastanın 13 aylık takibinde komplikasyon ve rekürrens görülmedi. İnce barsak stromal tümörleri oldukça nadirdir ve klinik bulgu ve semptomları belirgin değildir. Bunların tanıları sıklıkla komplikasyonlar ortaya çıkana kadar gecikir. Ameliyat öncesi doğru tanı komplikasyonlar ortaya çıksa bile genellikle çok zordur.

Anahtar kelimeler: Gastrointestinal stromal tümör, ince barsak, barsak perforasyonu, multifokal yerleşim

INTRODUCTION

Gastrointestinal stromal tumors (GISTs) are the most common mesenchymal tumors of the gastrointestinal tract in adults (1). The clinical manifestations of GISTs of the small intestine include abdominal pain, a palpable intra-abdominal mass, vomiting, weight loss, and occult gastrointestinal bleeding. Because those symptoms can result from any of several causes, GISTs are often diagnosed after complications such as hemorrhage, obstruction, and perforation have occurred (2, 3). Multifocal GISTs are uncommon (1, 3). We report the rare case of a patient with multifocal GISTs of the small intestine who presented with an abscess secondary to intestinal perforation.

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CASE REPORT

A 70-year-old man with right-sided abdominal pain, inappetence, nausea, and bloating of one-week duration was admitted to our Emergency Department and was subsequently hospitalized. His medical history was unremarkable except for the repair of a left-sided inguinal hernia. Physical examination revealed diffuse abdominal guarding with rebound tenderness. Upright plain abdominal radiographs showed a few air-fluid levels. The results of laboratory testing performed when the patient was admitted to the Emergency Department revealed the following values: hemoglobin, 11.1 g/dl (normal: 13.5-18 g/dl); leukocyte count, 10,000/mm^3 (76% polymorphonuclear cells); and C-reactive protein, 145 mg/L (normal: 0-10 mg/L). An abdominal computed tomographic scan showed an abscess (55 x 66 mm) in the right upper quadrant of the abdomen (Figure 1). The results of colonoscopic examination were within normal limits. Insertion via ultrasonographic guidance of a catheter into the abscess resulted in the immediate drainage of a purulent material followed by intestinal contents. Pouchography showed that a fine tract of contrast medium had passed into the intestinal lumen. The intestinal contents that drained from the abscess via the catheter suggested the presence of an abscess caused by the perforation of the intestinal diverticulum, and surgery was planned.

During laparotomy, a jejunal loop 5 cm distal from the ligament of Treitz was identified and was found to be covered with greater omentum and an abscess cavity. At the antimesenteric side of the jejunal wall, a solid mass measuring 5 cm in diameter and a perforation approximately 0.5 cm in diameter were identified after the exploration of the abscess cavity. In addition, 3 polypoid lesions (2 measured as 1 and 2 cm in diameter at the antimesenteric side of the jejunal wall, and 1 measured as 1 cm in diameter at the antimesenteric side of the ileal wall) were found incidentally. Resection and primary anastomosis of the perforated area and wedge resections of the polypoid lesions were performed (Figures 2, 3). The patient's postoperative course was uneventful, and he was discharged on the tenth postoperative day. The results of pathological and histological examinations of the surgical specimens showed no mitotic activity per 50 high power fields and the surgical margins were intact (Figure 4). The results of immunohistochemical stains were strongly positive for smooth muscle actin (SMA) antibody, 50% positive for S-100 protein, 30% positive for CD117 antibody, and negative for desmin and CD34. The pathologic diagnosis of all the lesions was similar to low-grade

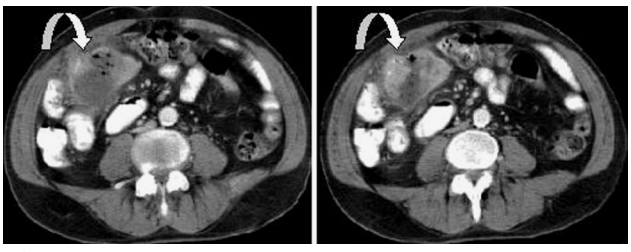

Figure 1. Axial post contrast computed tomographic image shows an abscess (55 x 66 mm in size) in the right upper quadrant of the abdomen (curved arrows).

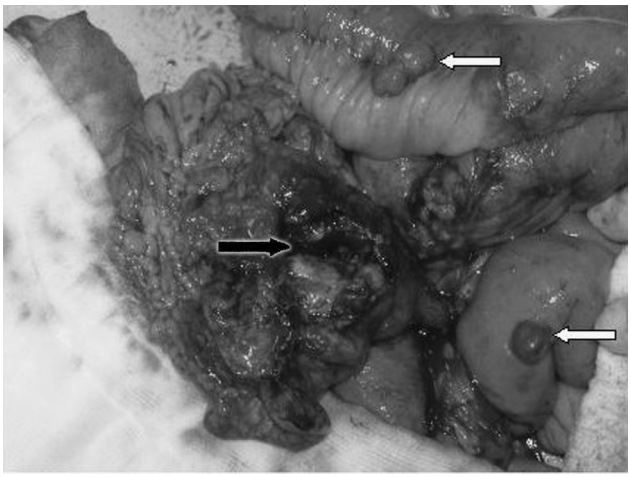

Figure 2. An abscess cavity surrounded by greater omentum and intestine loops (black arrow) and polypoid lesions at the antimesenteric side of the jejunum are demonstrated (white arrow).

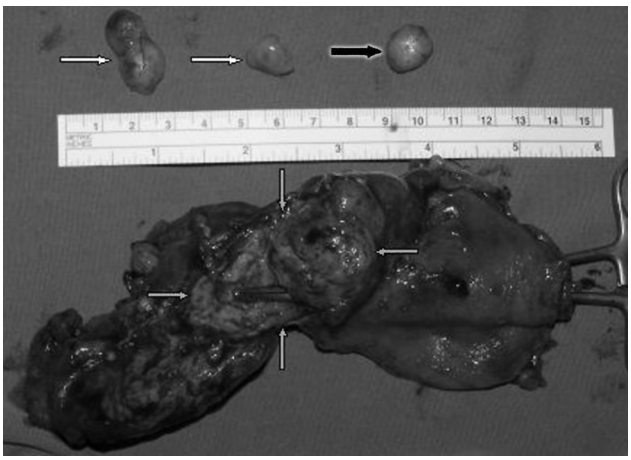

Figure 3. Resected materials. Polypoid lesions: Two are resected from the jejunal wall (white arrows), and the other from the ileal wall (black arrow). Gray arrows indicate the large tumor and perforated area of the jejunum.

GISTs. The patient consulted with an oncologist, and follow-up without treatment was planned. Abdominal computed tomography was performed on the 13th month of the follow-up and the patient remained healthy and without evidence of recurrence.

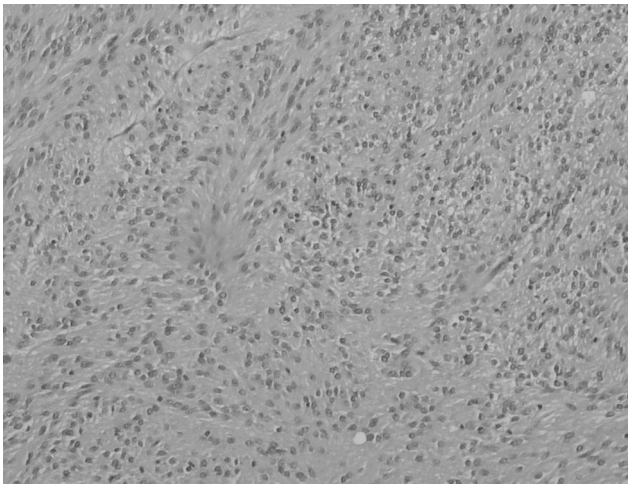

Figure 4. Pathologic specimen of the mass; no mitotic activity on the stromal tumor (H&E, x 100).

DISCUSSION

Gastrointestinal stromal tumors are the most common mesenchymal tumors of the gastrointestinal tract in adults. The incidence of GISTs in all gastrointestinal tumors is 0.1%-3% (1). GISTs occur primarily in those older than 40 years (peak incidence occurs in the sixth and seventh decades of life), and they are slightly more common in men (4). GISTs of the small intestine develop (in descending order of frequency) in the jejunum, ileum, and duodenum (1).

Small sized tumors (< 2 cm) are usually asymptomatic, and are discovered incidentally (5). Crosby and colleagues (1) reported that the most common signs or symptoms of GISTs include abdominal pain (74%), abdominal mass (72%), gastrointestinal bleeding (44%), partial or complete small bowel obstruction (44%), weight loss (16%), and fever or abscess (14%). The diagnosis of GISTs of the small intestine is often made after complications such as hemorrhage, obstruction, or perforation (2, 3). The symptoms and signs of GISTs are not disease-specific, so that about 50% of them already have metastases at the time of diagnosis, usually to the liver or the peritoneum (3, 4, 6-8). The pathogenesis of perforation due to the malignant tumors of the small bowel is unclear. The possible perforation mechanisms include replacement of the bowel wall by tumor cells followed by necrosis, ischemia of the intestine due to tumor embolization, and increased intraluminal pressure caused by obstruction (2).

None of the diagnostic procedures, such as barium examination of the gastrointestinal tract, computed tomography, or angiography, can establish the correct diagnosis with 100% certainty (5, 9-11). Chao and colleagues (2) reported that the preoperative diagnosis of the perforation of the small intestine was 8% due to malignant tumors and 4% due to multiple primary GIST lesions without metastasis. Multiple GISTs of the small intestine can also develop in individuals with neurofibromatosis type 1 (3). Our patient had no signs or symptoms of neurofibromatosis type 1, and the diagnosis was made after an intra-abdominal abscess caused by perforation had been identified.

Although the main prognostic factor is the mitotic count, considering the possibility of metastasis or recurrence, GISTs can be classified as low- or high-risk tumors (12-14) (Table 1). The positivity for KIT protein (CD117 antigen) is always shown in the immunohistochemical examination of GISTs, while the positivity regarding other markers varies (3, 6, 12, 15, 16) (Table 2).

The surgical excision of the tumor is the treatment of choice for GISTs. All the tissues that are infiltrated and the tumors should be completely resected. However, systemic lymph node dissection is not recommended by many authors (17, 18). 48-

Table 1. GISTs classification by Fletcher et al. (14)

Risk of malignancy	Size of tumor (cm)	Mitotic counts (/50 high power fields)
Very low	< 2	< 5 / 50
Low	2 - 5	< 5
Intermediate	< 5	6 – 10
	5 - 10	< 5
High	> 5	> 5
	>10	Any counts
	Any size	> 10

Table 2. Proportion of positivity of GISTs for various immunohistochemical markers

Marker	Positivity
KIT (CD117 antigen)	100%
CD34	70%
SMA	20-30%
S-100	10%
Desmin	<5%

65% five-year survival is reported in complete surgical resection. The cases in which only partial resection must be performed are in large tumors, when there is palliation or control of symptoms or in complications such as compression of adjacent organs, hemorrhage, or pain (6).

When the tumor presents with symptoms or signs such as perforation, multifocal localization or metastatic lesions, the prognosis is poor. The five-year survival is 0% in patients with metastatic tumors or multifocal tumors, whereas patients with localized or locally advanced tumors have 46% five-year survival. In cases of tumor perforation, the five-year survival is 24%, probably due to peritoneal dissemination (1). In our patient, complete resection was performed despite the multiple locations of the primary tumor.

Gastrointestinal stromal tumors have poor response to conventional chemotherapy (<10%), whereas in cases of intraperitoneal hemorrhage, when the exact location of the tumor is shown, or for analgesic purposes, radiotherapy can be used (6, 12, 18).

STI571, which has been recently named as imatinib, has been found to be a powerful selective inhibitor of tyrosine kinases and the receptors of platelet-derived growth factor receptor and c-kit. Imatinib has been shown in clinical studies to be the first effective treatment for non-resectable or metastatic GISTs. However, long-term results are still not extracted due to the short period of its use (6). Since our patient did not have any metastasis, complete surgical resection of the tumors was performed and all were reported as low-grade tumors in pathologic examinations. Imatinib was not used since the long-term results are not known.

In conclusion, we have limited treatment options in GISTs. Complete surgical resection without extensive lymph node sampling is still the choice of the primary treatment. Small intestine GISTs cause subtle clinical signs and symptoms; their diagnosis is often delayed until complications develop. Even if complications occur, exact diagnosis is usually very difficult preoperatively. It should be kept in mind in the differential diagnosis of patients presenting with unexplained intra-abdominal perforation and/or abscess.

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