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Case Report

Small bowel gastrointestinal stromal tumor presenting as a life-threatening emergency

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ABSTRACT

Gastrointestinal stromal tumor (GIST) is a rare stromal neoplasm, which represents the most common mesenchymal tumor of the gastrointestinal tract. It is characterized by indolent clinical symptoms, although it can present as a life-threatening emergency. Here in, we present a case of primary small bowel GIST treated at our department. A 58-year-old male patient presented to our emergency department with a diffuse abdominal pain of acute onset. Patient presented with symptoms and signs of acute intestinal obstruction. Imaging studies revealed a mass at mid ileum, with multiple air fluid levels. Intra-operatively, a mass of 12×9 cm was found at mid ileum on antimesenteric border with 3600 volvulus of adjacent ileum. En bloc resection was performed. Histopathology report was suggestive of gastrointestinal stromal tumor.

Keywords: Gist, Peritonitis, Volvulus, Obstruction

INTRODUCTION

Gastrointestinal stromal tumor (GIST), first described by Mazur and Clark (1983), is the most common mesenchymal neoplasm of the GI tract; however, it accounts for less than 1% of all GI tumors.[1,2] It originates from the interstitial cells of Cajal, which are part of the autonomic nervous system of the intestine.[3] The majority of the lesions are benign with a possibility of 10-30% for malignancy.[4]

GISTs arise usually from the muscularis mucosa or propria layers and mostly have an exophytic pattern of growth. The estimated frequency of GIST tumors is 10-20/1 million population, and it occurs in patients at the sixth decade of life and can arise anywhere in the GI tract from the esophagus to the rectum with gastric being most common site.[4,5] Most of GIST tumors are asymptomatic and might present as painless lump in abdomen. However, GISTs (mainly tumors larger than 4 cm) may present as abdominal emergencies, including GI hemorrhage, usually due to pressure necrosis and ulceration of the overlying mucosa, intestinal obstruction, or perforation.

We describe case of small bowel GIST presenting as surgical emergency at our institute.

CASE REPORT

A 58-year-old male patient presented to emergency department complaining of acute abdominal pain. The pain lasted for 8 hours, started abruptly, and was described as sharp, acute, and being without radiation. He also had complaints of nausea, multiple episodes of bilious vomiting, and reduced appetite.

On physical examination upon admission, the patient's vital signs showed fever of 39.3°C and tachycardia of 110/min. An abdominal examination revealed abdominal distention, diffuse tenderness with guarding. No abdominal mass was palpated. Digital rectal examination was normal. A clinical diagnosis of acute intestinal

obstruction was made. There was bilious output on Ryle tube insertion. Complete blood count showed increased white blood cells of 12000, with 10% bands. Liver and kidney function tests were within normal limits. An upright abdominal X-ray were suggestive of multiple air fluid levels (Figure 1). A computed tomography scan revealed a cystic mass lesion of size approximately 12×8×7 cm (AP×TR×CC) arising from mid ileum causing upstream dilatation of bowel and collapsed distal bowel (Figure 2). He was admitted with a diagnosis of intestinal obstruction. patient underwent an exploratory laparotomy, during which a mass of 12×8 cm, located on antimesenteric border of mid ileum, 60 cm distal to Treitz ligament, was found, with a dilated proximal bowel with 360°C volvulus of ileum perpendicular to axis of mesentery (Figure 3). En bloc resection of the mass with end-to-end anastomosis between the proximal ileum and distal ileum was performed. Inspection of the rest of the GI tract and abdominal cavity did not reveal associated abnormalities. His postoperative course was uneventful, and the patient was discharged on day eight.

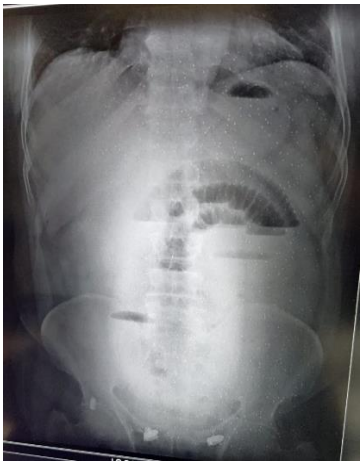

Figure 1: X-ray abdomen showing multiple air fluid levels.

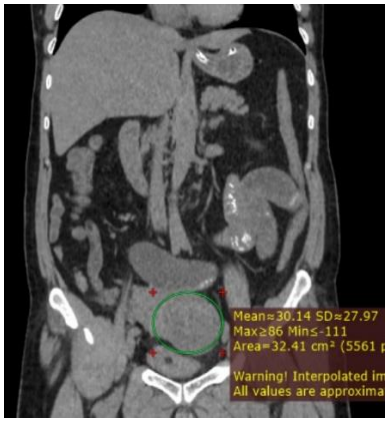

Figure 2: CT scan showing a mass at mid ileum.

The histopathological examination showed a GIST of 12×10 cm, made of spindle and epithelioid cells and invading the mucosa (Figure 4). Mitoses were <5 per 50 high power fields. Immunohistochemically, the tumor cells were positive for CD 117, DOG-1, CD34 and *H. caldesmon*, and negative for Desmin, SOX1, HMB45, and S100. Ki67 index was 2-5%. These findings are compatible with high risk for recurrence.

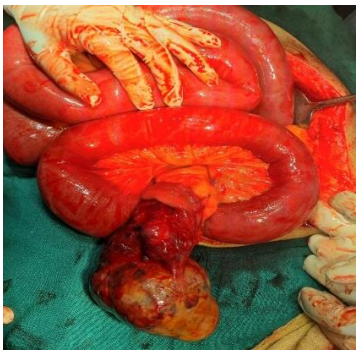

Figure 3: At laparotomy: mass at mid ileum with proximal dilated bowel.

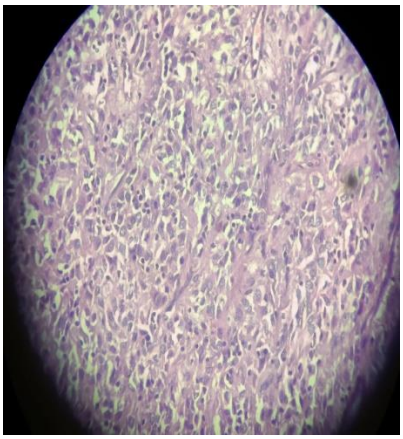

Figure 4: Histo-pathological examination made of spindle and epithelioid cells.

Post-operatively, the patient was offered adjuvant treatment with imatinib. During one years of postoperative follow-up the patient is asymptomatic, no signs of recurrence and compliant with follow ups and advised treatment.

DISCUSSION

GIST tumors are uncommon tumors, and high index of suspicion is warranted for diagnosis.[3,4,7] None of the diagnostic procedures, including computer tomography, ultrasound, barium examination, angiography, and magnetic resonance imaging, has a 100% certainty for diagnosis, and preoperative fine needle aspiration is not indicated due to the risk of tumor rupture and intraperitoneal seeding.[4,5,8] However, some recent studies have showed the importance of endoscopic ultrasound guided fine needle aspiration, with reported accuracy of 89%.[9] A prognostic classification, proposed by Fletcher et al depends on tumor size and mitotic counts, classifying GISTs into very low, low, intermediate, and high risk for

malignancy (Table 1).[10] There is slight male predominance.[7] These tumors are mainly sporadic, although familial forms with autosomal dominant inheritance have been documented.[3,7] GIST usually spreads by direct extension to adjacent structures and hematogenously to the liver, lung, and bone. Lymphatic metastasis is unusual.[11] GIST tumors have variable markers, including C-KIT (CD 117), DOG-1, CD 34, SMA, S100, and Desmin, of which, they are almost always positive for C-KIT and DOG 1.[3,5,12-14]

Table 1: Prognostic classification.[10]

Class	Size (largest dimension) (cm)	Mitotic count (HPF)
Very low risk	<2	<5/50
Low risk	2-5	<5/50
Intermediate risk	<5	6-10/50
	5-10	<5/50
High risk	>5	>5/50
	>10	Any mitotic rate

The most common site for GIST is the stomach (60-70%), followed by the small bowel (25-35%).[5] GISTs involving the esophagus, appendix, colon, and rectum are rare, and tumors arising from the omentum, mesentery, or retroperitoneum have been documented; but most of these were found to be metastatic from gastric or intestinal primaries.[7] Due to the nonspecific symptoms and signs, it is difficult to diagnose ileal GIST preoperatively. Although specific signs and symptoms are absent, most GISTs (70%) are symptomatic, mainly presenting with vague abdominal pain.[5,17] Other symptoms include nausea, vomiting, early satiety, and abdominal fullness. The remaining (30%) are asymptomatic and diagnosed incidentally. These latter tumors are usually small sized tumors (<2 cm).[3,5,12]

Although computer tomography is a viable imaging modality for patients suspected of having intra-abdominal GIST, magnetic resonance imaging modality provides a more accurate preoperative picture.[18] Prognostic factors include anatomic location of the primary tumor, age at presentation, histomorphology, molecular genetics, and immunohistochemistry, of which tumor size is the most important.[19] Surgical complete R0 resection has always been the main stay of treatment, with the conventional chemotherapy and radiation treatment having proven to be ineffective on GIST. The 40-90% of surgically treated patients experience recurrence of the disease. Another study on 127 patients with localized GISTs after complete resection showed a 5-year recurrence-free survival (RFS) rate of 63%.[21] GIST tumors have poor response to chemotherapy or radiotherapy, which is usually used in cases of hemorrhage or for analgesic purposes.[5,12,20] Post-operative use of imatinib, a tyrosine kinase inhibitor has shown to delay the recurrence and improve the survival rate.[22]

The aggressive behavior of the tumor is based on size, mitotic rate >2/50 hpfs, Ki 67>10%, cyst formation, necrosis and increased cellularity.[22] In our case, the microscopic examination of the specimen made of spindle and epithelioid cells and invading the mucosa (Figure 4). Mitoses were <5 per 50 high power fields. Immunohistochemically, the tumor cells were positive for CD 117, DOG-1, CD34 and *H. caldesmon*, and negative for Desmin, SOX1, HMB45, and S100 and ki67 index was 2-5%.

GIST presenting as a small bowel obstruction is reported in less than 10% of cases.[24] In our case gist was located on antimesenteric border of ileum causing volvulus of small intestine leading to small bowel obstruction.

CONCLUSION

Small bowel GIST is an uncommon tumor, with a small bowel obstruction being a rare initial presentation. In order to make the correct diagnosis a high index of suspicion is mandatory. The mainstay treatment for these emergent presentations of small bowel GIST is En bloc resection with a favourable clinical result achievable when the diagnosis is made in a timely manner.

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