Case Report

Huge Gastrointestinal Stromal Tumor of the Jejunum Presenting as Bowel Obstruction: A Rare Presentation

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Keywords

Gastrointestinal tumor · Gastrointestinal stromal tumor · Jejunal tumor · Bowel obstruction · CD117 · CD34 · PDGFRA · Imatinib

Abstract

Gastrointestinal stromal tumors (GISTs) are mesenchymal tumors of the gastrointestinal tract arising from interstitial Cajal cells. A 54-year-old male patient without any comorbidities presented to the emergency department with complaints of abdominal distension, inability to pass flatus, and no motion for the past 2 days. Abdominal X-ray showed multiple air-fluid levels suggesting bowel obstruction. The patient was managed conservatively at first. Later, ultrasonography and contrast-enhanced computed tomography (CECT) of the abdomen were done, which revealed a mass lesion arising from the jejunum suggestive of GIST. The patient was taken up for exploratory laparotomy, and a tumor was found in the proximal jejunum around 10 cm from the duodenojejunal junction and encompassing the hepatic flexure of the transverse colon, with the omentum found adhered to the anterior surface of the lesion and distended proximal bowel loops. There was no evidence of mesenteric lymphadenopathy. The mass was resected along with the jejunal loop and the hepatic flexure of the colon, followed by end-to-end jejunojejunal anastomosis and end-to-end colocolic anastomosis. The patient's postoperative stay was uneventful. Imatinib therapy was started following histopathological confirmation and continued. The patient was followed up for 1 year postoperatively with CECT of the abdomen every 6 months, with no evidence of recurrence or any gastrointestinal symptoms.

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Introduction

Gastrointestinal stromal tumors (GISTs) are defined as mesenchymal tumors of the gastrointestinal tract expressing proto-oncogene protein CD117 [1]. They are the most common sarcomatous tumors of the gastrointestinal tract [2]. They were originally believed to be smooth muscle sarcomas and previously named as leiomyoma and leiomyosarcoma [1, 2]. They are now known to originate from interstitial Cajal cells (intestinal pacemaker cells) [2]. They are equally observed in males and females and can manifest at any age, more commonly in patients older than 50 years [1, 2]. Some cases in children and young adults have also been reported [3].

They can appear anywhere in the gastrointestinal tract – i.e., in the stomach (40–60%), jejunum and ileum (30%), duodenum (5%), and colon (15%) – and very rarely in the esophagus and appendix [2]. Extra-gastrointestinal tract GISTs have been reported in the omentum, mesentery, retroperitoneum, gallbladder, and urinary bladder [4, 5]. More than 95% of GISTs express CD117 (a c-Kit proto-oncogene) and 70–90% express CD34 (a human progenitor cell antigen) [2]. Along with c-Kit, PDGFRA mutations (in exon 18) are also seen. The c-Kit mutations are seen in exons 9 and 11 [6]. These tumors may sometimes stain positive for actin (20–30%), S100 (2–4%), and desmin (2–4%) [2].

Case Presentation

A 54-year-old male patient without any comorbidities presented to our emergency department with complaints of pain in the abdomen, abdominal distension, and complete obstipation for the past 2 days. The pain was sudden in onset, colicky in nature, diffuse, progressive for the past 2 days, and associated with 3–4 episodes of vomiting.

There was no associated history of fever or any urinary complaints. However, there was a history of 2–3 similar episodes in the past 2 years, which resolved spontaneously. There also was an associated history of loss of appetite for the past 2 months, but no history of weight loss, hematemesis, or melena.

An abdominal examination revealed abdominal distension with increased abdominal girth and hyperdynamic bowel sounds. A vague lump was palpable in the right lower abdomen with indistinct margins, noncompressible and firm in consistency. An abdominal X-ray was done, which confirmed the diagnosis of bowel obstruction. All routine laboratory investigations were unremarkable. Ultrasonography of the abdomen showed a well-defined, irregular, hypoechoic lesion of around 14 × 8.9 cm with a significant area of necrosis and peripheral vascularity. Multiple air foci were seen at the center of the lesion. The lesion was extending intraluminally into a small bowel loop, possibly arising from it. The features were suggestive of GIST/liver hemangioma.

The patient was managed conservatively and was relieved of the obstruction within 3 days. Later, upper gastrointestinal endoscopy was done, which was unremarkable. Contrast-enhanced computed tomography (CECT) of the abdomen (Fig. 1) revealed an ill-defined, heterogeneously enhancing, lobulated intraperitoneal mass lesion of approximately 9.3 × 15 × 12 cm in the right lumbar and umbilical region arising from the wall of a jejunal loop. The lesion had a small endoluminal and a large extraluminal component. A large central nonenhancing area of necrosis was noted within the lesion. Multiple air foci were also noted within the lesion, suggestive of ulceration.

The mass was observed to displace small bowel loops and showed loss of the fat plane at the hepatic flexure of the colon. Anteriorly, the lesion extended into the greater omentum, and posteriorly it was abutting the third part of the duodenum, and the infrarenal inferior vena cava. The fat plane at the abdominal aorta, portal vein, bilateral renal vessels, and right ureter was maintained. The lesion was displacing the body and tail of the pancreas, the anterior surface of the spleen, and the left adrenal gland. The features were suggestive of GIST/small bowel lymphoma.

The patient was taken up for exploratory laparotomy, and a tumor was found in the proximal jejunum around 10 cm from the duodenojejunal junction and encompassing the hepatic flexure of the transverse colon (Fig. 2). The lump was observed to compress the jejunal bowel loops. The proximal bowel loop was

Fig. 1. Contrast-enhanced computed tomography image showing a jejunal gastrointestinal stromal tumor (arrow) with a large central nonenhancing area of necrosis noted within the lesion. Multiple air foci are also visible within the lesion, suggestive of ulceration.

Fig. 2. Intraoperative picture showing a large tumor involving the jejunum and colon. Top arrow: the jejunal loop from which the lesion is arising. Bottom arrow: colonic wall adhered to the lesion.

Color version available online

dilated. The omentum was found to adhere to the anterior surface of the lesion. There was no evidence of mesenteric lymphadenopathy.

The mass was resected along with the jejunal loop and the distal ascending and hepatic flexure of the colon (Fig. 3), followed by end-to-end jejunojejunal and colocolic anastomosis. The patient's postoperative stay was uneventful. Histopathological examination revealed a lesion of around $18.5 \times 14.5 \times 7$ cm. The cut surface of the tumor was gray-white and fleshy with necrotic and hemorrhagic areas.

Microscopic examination (Fig. 4) showed a submucosal, diffusely infiltrative tumor reaching up to the lamina propria and ulcerating into the mucosa. The large nodules of the tumor were separated by fibrovascular-to-fibrocollagenous septae. The nodules showed whorls and nests, as well as a focal organoid and storiform pattern of spindle-shaped-to-epithelioid tumor cells. The tumor cells had oval-to-round, mildly hyperchromatic nuclei. At some places, giant tumor cells were seen. Mitotic activity was 11–12/5 mm². Focally myxoid change, perivascular hyaline change, and areas of necrosis were also observed.

Color version available online

Fig. 3. Resected gastrointestinal stromal tumor specimen, with the right arrow showing the cut colonic side and the left arrow showing the cut jejunal side.

Color version available online

Fig. 4. Photomicrograph of an H&E-stained section showing large nodules of the tumor separated by fibrovascular-to-fibrocollagenous septae. The nodules show whorls and nests, as well as a focal organoid and storiform pattern of spindle-shaped-to-epithelioid tumor cells. The tumor cells have oval-to-round, mildly hyperchromatic nuclei. At some places, giant tumor cells are visible. Focally myxoid change, perivascular hyaline change, and areas of necrosis are also visible.

The tumor was infiltrating the serosal aspect of the colon, but was not penetrating the serosa. The omentum was tumor free. The features were suggestive of GIST of mixed type. Immunohistochemistry showed SMA positivity, DOG1 positivity, focal CD117 positivity, and CD34 negativity. The histopathological examination revealed an R0 resection.

Imatinib therapy was started. The patient was followed up for 1 year postoperatively with CECT of the abdomen every 6 months. There was no evidence of recurrence during follow-up. No specific gastrointestinal symptoms were seen.

Discussion

GISTs vary considerably in their presentation and clinical course. Most GISTs are asymptomatic and diagnosed incidentally when conducting an abdominal radiological investigation or during surgery for another etiology. The symptoms depend on the size and location of the tumor. Symptomatic GISTs usually present with bleeding (hematemesis/melena), vague abdominal pain or discomfort, and weight loss. They may show intramural growth, very rarely leading to obstruction, or have intramural and extramural growth, leading to massive sizes [2]. Some patients with large GISTs may have externally palpable masses [7, 8]. Lymph node metastasis is extremely rare [1]. The most common kinds of metastasis are to the peritoneum and liver [1], whereas metastasis to the lung and bones has been reported in some cases [9]. Size and mitotic index are the best predictors of metastasis. The mitotic index is classified as low (<5 mitoses/50 HPFs) or high (>5 mitoses/50 HPFs) [2]. Patients are evaluated using upper gastrointestinal endoscopy, endoscopic ultrasound, and CECT of the abdomen and pelvis (to assess metastasis). Surgery is the primary modality of treatment.

GISTs are tumors with unpredictable biological behavior. Their diagnosis is confirmed by biopsy with immunohistochemistry positive for CD117, CD34, or PDGFRA. Some GISTs express positivity for DOG1 also. The mainstay of treatment is complete surgical resection. Tumors >2 cm in diameter should be resected. Tumors <2 cm in diameter with high-risk features like irregular borders, ulceration, etc., should also be resected, whereas without these features, these can be observed by repeated upper gastrointestinal endoscopy and endoscopic ultrasound every 6 months [2]. Smaller tumors can be treated by wedge resection, whereas larger tumors may require a gastrectomy/duodenectomy, etc. [1]. The aim of surgery should be to have an R0 resection. No lymph node dissection is required, as lymph node metastasis is rare. Recurrence rates are around 40%, and most patients demonstrate metastasis to the liver, while only one-third of the patients have isolated local recurrence. Long-term disease-free survival is around 50%. Gastric GISTs have a more favorable prognosis than other GISTs. The risk factors for malignancy and recurrence are tumors >10 cm and tumors having >5 mitoses/50 HPFs [2]. Most benign tumors have a low mitotic index (<5 mitoses/50 HPFs).

Adjuvant therapy with imatinib (a tyrosine kinase receptor inhibitor) is used to prevent recurrence following surgery, in unresectable cases, and with metastatic disease. It is effective in patients with GISTs who have mutations in exon 11 of the KIT gene. Patients having mutations in exon 9 of the KIT gene also may respond to imatinib, but only with higher doses, whereas patients without mutations in the KIT gene do not respond to imatinib [6]. The Scandinavian Sarcoma Group (SSG) XVIII trial established a postoperative therapy of 3 years with imatinib for high-risk GISTs [6]. Sunitinib (a tyrosine kinase inhibitor) is used for the treatment of imatinib-refractory GIST and in patients not tolerating imatinib [6]. Sunitinib targets multiple kinases, including VEGF receptors, PDGFRA, KIT, and FLT3. Other, newer drugs in development are sorafenib, dasatinib, nilotinib, and regorafenib [6]. FDG-PET is widely accepted now for preoperative screening of GISTs, as well as for detecting an early response to imatinib [6].

The optimal follow-up criteria for patients with GIST need further investigation. Very-low-risk GISTs may not require routine follow-up. For low-risk tumors, follow-up may be done by abdominal CT scanning or MRI every 6–12 months for 5 years. High-risk patients require follow-up with abdominal CT scanning or MRI every 3–6 months for 3 years during adjuvant therapy, and then, on cessation of adjuvant therapy, every 3 months for 2 years, then every 6 months until 5 years from stopping adjuvant therapy, and annually for an additional 5 years [10].

Conclusions

Bowel obstruction is a very common presentation at the surgery department. It can have varied etiologies, but GIST is a very rare etiology of bowel obstruction. With this case report, we wanted to emphasize the importance of GISTs as a rare cause of bowel obstruction which should be kept in mind when managing such cases. Complete surgical resection should be done and imatinib therapy started. Patients need to be followed up with CECT of the abdomen to look for any future recurrences.

Statement of Ethics

Informed and written consent was obtained from the patient for the contents of the manuscript to be published, including the images.

Disclosure Statement

The authors have no conflicts of interest to declare.

Author Contributions

M.P. Singh contributed substantially to the concept, design, and drafting of the work and gave final approval for the version to be published. T. Huda contributed substantially to the acquisition, interpretation, and critical analysis of the data and drafting of the work.

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