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The Surgical Management of a Spontaneous Rupture of a Giant Infected Gastrointestinal Stromal Tumor of the Jejunum

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Abstract **Case Report**

Gastrointestinal stromal tumor (GIST) is a rare tumor of the small bowel, which can be difficult to diagnose and has a varied clinical outcome. It is described for the first time by Mazur and Clark [1], is the most common mesenchymal neoplasm of the GI tract; however, it accounts for less than 1% of all GI tumors. It originates from the interstitial cells of Cajal, which are part of the autonomic nervous system of the intestine. The majority of the lesions are benign with a possibility of 20–30% for malignancy.

Keywords: Small bowel, gastrointestinal stromal tumor, diagnosis, prognosis, clinical outcome.

INTRODUCTION

Gastrointestinal stromal tumor (GIST), first described by Mazur and Clark [1], is the most common mesenchymal neoplasm of the GI tract; however, it accounts for less than 1% of all GI tumors. It originates from the interstitial cells of Cajal, which are part of the autonomic nervous system of the intestine. The majority of the lesions are benign with a possibility of 20–30% for malignancy. GISTs arise usually from the muscularis mucosa or propria layers and mostly have an endophytic pattern of growth.

We describe an emergency case of small bowel GIST treated at our department.

CASE REPORT

A 50-year-old man previously fit and well, presented with fever, pain and abdominal discomfort that had been gradually increasing over 2 days. There were no episodes of gastrointestinal bleeding. The blood pressure was 12/90 mmHg, the pulse 90 beats per minute, and the temperature 39°C.

Abdominal examination revealed tenderness and muscular defense and there was a huge solid mass palpable in the left lower quadrant of the abdomen. It was non-pulsatile, solid and immobile.

Blood tests showed a hemoglobin level of 8.7 g/dL, white blood cell count of 26000/mm3, and C-reactive protein of 400 mg/dL.

The abdominal scan demonstrated a large inhomogeneous mass, measuring 139.7x112.5mm, within the left iliac fossa. It was a well-circumscribed, lobulated mass which contained several areas of necrosis and abscess (Figure-1). The mass was suspected as a GIST of the jejunum accompanied by a giant abscess.

The patient was treated with intravenous administration of antibiotics. Subsequent to improvement in his clinical condition and laboratory tests, laparotomy was performed.

Intraoperative findings showed presence of two purulent collections, one on the Douglas and the other at the right iliac fossa, we also found an outgrowing 15 cm mass which was located in the meso of the proximal small bowel about 1 meter from the Treitz ligament with omentum and loops of small bowel densely adhering to and covering the mass. Segmental resection of the jejunum with the tumor was performed. The resection was extended from 80 cm to 1m20 of the jejunum carrying 40 cm of it. A jejuno-jejunal mechanical anastomosis was performed.

The postoperative course was uneventful and the patient was discharged on the fifth postoperative day.

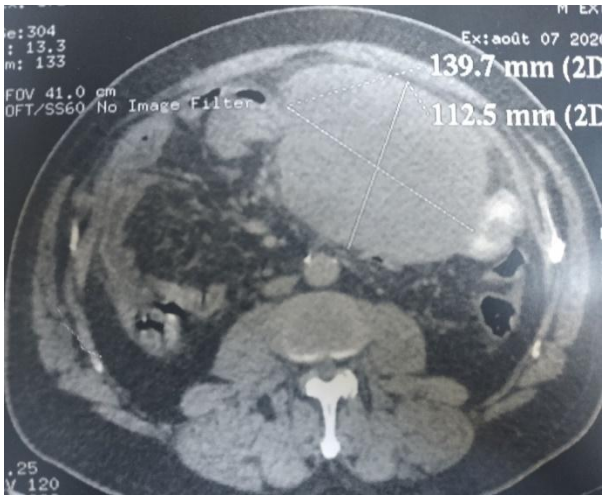

Fig-1: CT scan

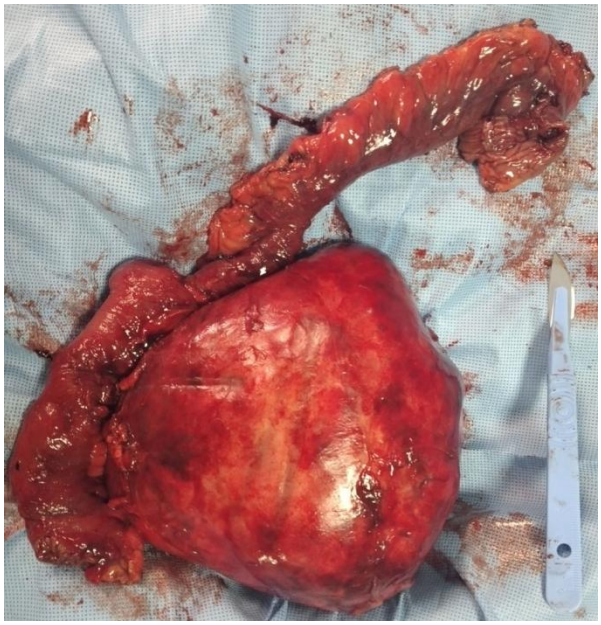

Fig-2: Giant infected gastrointestinal stromal tumor

DISCUSSION

Gastrointestinal Stromal Tumor (GIST) originates from the intestinal cells of Cajal, which constitute the autonomic nervous system of the intestine. Gastrointestinal stromal tumors (GISTs) are the most common mesenchymal neoplasms of the gastrointestinal tract. They constitute 1–2% of all gastrointestinal neoplasms but are the most common subtype of soft tissue sarcomas, accounting for 20–25%. Majority of these tumors are benign but a small percentage of them have malignant potential, especially intestinal GISTs. Most often GISTs occur in the stomach, followed by the small bowel and colon/rectum. In contrast to epithelial tumors, GISTs grow transmurally and submucosal. GISTs can be found with highly variable growth features including tumors with intraluminal, intra- or transmural, and pedunculated appearance [2].

Estimated incidence of GIST tumors is 10–20 cases per million, with the majority of cases in patients over 50 and mean age of diagnosis being around 64 years of age. GIST tumors usually present with a wide array of symptoms. The clinical presentation is very heterogeneous, depending on tumor site, size, and growth pattern. GISTs of the stomach is the group with the lowest rate of acute or emergency symptoms with 31%, followed by GISTs of the duodenum with 42%, whereas GISTs of the small bowel show acute symptoms in more than 50% of the cases and have an emergency surgery rate of almost 15%. Many patients are diagnosed accidentally, through screening examinations, or with latent, unspecific symptoms [3].

A few cases have been reported in the English medical literature associated with perforation or rupture of GIST in the small intestine. Bleeding into the peritoneal cavity due to a ruptured GIST can lead to acute abdominal pain presenting as a surgical emergency. In addition, only a few cases of GIST of the small intestine accompanied with abscess formation have been reported. In this case, GIST originating in the jejunum was invaded via a fistula, leading to central necrosis and spontaneous rupture into the peritoneal cavity [4].

High index of suspicion is warranted in patients having recurrent GI bleeding without obvious source on esophagogastroduodenoscopy and colonoscopy. Further imaging modalities including CT scan and MRI of abdomen can help in making the diagnosis [5].

CD117, the c-kit proto-oncogene protein, is a tyrosine kinase growth factor receptor and is the most specific and important immunohistochemical tissue marker for GIST. The use of histopathology and immunohistochemical staining together with other immunomarkers, including CD34, DOG1, smooth muscle actin (SMA), S100 protein, and desmin, have helped to distinguish GIST from other primary mesenchymal tumors of the GI tract [6].

Recommended treatment of choice for small bowel GIST is en bloc surgical resection with inclusion of pseudocapsule. Radical lymphadenectomy is not routinely indicated as lymph node involvement is less than 10%. Treatment with selective tyrosine kinase inhibitor is recommended in high risk cases to reduce risk of recurrence [7].

Large GIST tumors have a change of recurrence even after surgery though the rate of recurrence decreases if treated with imatinib mesylate.

CONCLUSION

Although GIST can arise in any portion of the GI tract, from the esophagus to the rectum, the small bowel is the second most common site of involvement, after the stomach. Small bowel GIST is one of the most common tumors of the small bowel. The diagnosis of GIST of the small bowel may be delayed for several reasons, including its relatively low incidence, nonspecific and variable symptoms, the wide spectrum of radiological appearances, intestinal thickening, and the presence of overlapping loops of intestine, which make imaging studies difficult; all of these lead to delayed or misdiagnosis of GIST of the small bowel.

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IRB and clinical trial registration approval are not required for case reports.
Constant of the use of medical data is obtained verbally from the patient.

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