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Volvulus of Small Bowel Caused by GISTs: Case Report and Literature Review

Muhaned Alhassan[1], Ayman Barghash[1], Hatem Al-Saadi[2]

[1]Department of General Surgery, Sohar Hospital, Sohar, Oman
[2]Sohar Hospital, Sohar, Oman
Email: hondalhassan@gmail.com

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Abstract

Introduction: Gastrointestinal stromal tumors (GISTs) originating from the interstitial cells of Cajal and GISTs are the most frequent mesenchymal tumors in the gastrointestinal tract (20% to 30% occur in the small intestine). Most small-intestinal neoplasms are asymptomatic. The most common mode of presentation is partial small bowel obstruction, with associated symptoms of colicky abdominal pain, abdominal distention, nausea, and vomiting. Obstruction can be caused by luminal narrowing by the tumor itself or intussusception. **Case Presentation:** We presented an 85-year-old lady presented with acute intestinal obstruction and CT abdomen with contrast revealed, a large well defined solid mass lesion, enhanced in the arterial phase and inseparable from the jejunal loops and it measures 09 × 08 × 07 cm, likely GISTs with secondary volvulus of the small bowel. Emergency laparotomy with complete resection of the tumor and end-to-end anastomosis of jejunum was done. **Discussion:** Small-intestinal GISTs should be treated with segmental intestinal resection. If the diagnosis is known prior to resection, wide lymphadenectomy can be avoided as GISTs are rarely associated with lymph node metastases. GISTs are resistant to conventional chemotherapy agents. Imatinib (Gleevec) is a tyrosine kinase inhibitor with potent activity against the tyrosine kinase KIT and is used in those with metastatic disease and as an adjuvant chemotherapeutic agent.

Keywords

GISTs, Small Intestine, MRI, CT Scan, Imatinib, Tyrosine Kinase Inhibitor

1. Introduction

The specific features of GISTs are their mutation of proto-oncogene KIT, a re-

ceptor tyrosine kinase. The interstitial cells of Cajal normally express KIT, these make it the cell of origin for GISTs. KIT expression is assessed by staining the tissues for CD117 antigen, which is part of the KIT receptor, and present in 95% of GISTs [1].

GISTs are the most frequent mesenchymal tumors in the gastrointestinal tract, and they accounts for seven to 20 per million per year [2] [3]. They can occur anywhere in the GI tract, with the stomach accounting for 50% to 60% of cases, the small intestine for 20% to 30%, the colon or rectum for 5% to 10%, the esophagus for <5%, and the peritoneum and mesentery for <1% [4].

Because small intestinal neoplasms are usually rare, they are difficult to detect in early images. As a result, they are often missed and delayed in diagnosis [5]. GISTs of the small intestine are currently considered more invasive than GISTs of the same size in the stomach, and their incidence has been rising in the past few years, a phenomenon that some scholars attribute to advances in radiology and endoscopy techniques, as well as improved physician awareness [6] [7].

Small intestine GISTs predominantly affect people from 40 to 70 years of age and in patients of different ages, the distribution of small intestine GISTs is roughly the same regardless of gender [7].

Clinical manifestations of GISTs are depended on tumor size, location, and invasion of mucous membranes. The most common clinical presentation is abdominal pain and gastrointestinal bleeding. Although it can be symptomatic and symptoms of metastasis it has been reported [8].

2. Case Presentation

We present an 85 years old lady, who had complained of colicky central abdominal pain for 4 days, associated with bilious vomiting and central abdominal distention but no constipation. She reported that there is a history of recurrent attacks of colicky abdominal pain. No history of previous abdominal surgery and no chronic illnesses. Abdominal examination revealed a distended soft abdomen with no guarding or tenderness, exaggerated bowel sound, and rectum was empty on digital rectal examination. Labarotory investigations including RFT, LFT, CBC, CRP and ABG were normal. CT abdomen with contrast revealed a large well defined solid mass lesion enhanced in arterial phase and inseparable from the jejunal loops and it measures 09 × 08 × 07 cm, likely GISTs with secondary volvulus of the small bowel. Emergency laparotomy was done with medline vertical incision, and the finding was a large mass involving antimesenteric part of med-jejunum with volvulus of loop of the jejunum and proximal small bowel dilatation (**Figure 1**). Complete resection of the tumor and end-to-end anastomosis of the jejunum was done (**Figure 2**). The patient was discharged home after one week of admission in good health condition. Histopathology results showed the features are consistent with GISTs, spindle cell morphology and no mucosal ulceration or invasion and free margins; therefore, the patient was referred to oncology evaluation and adjuvant therapy.

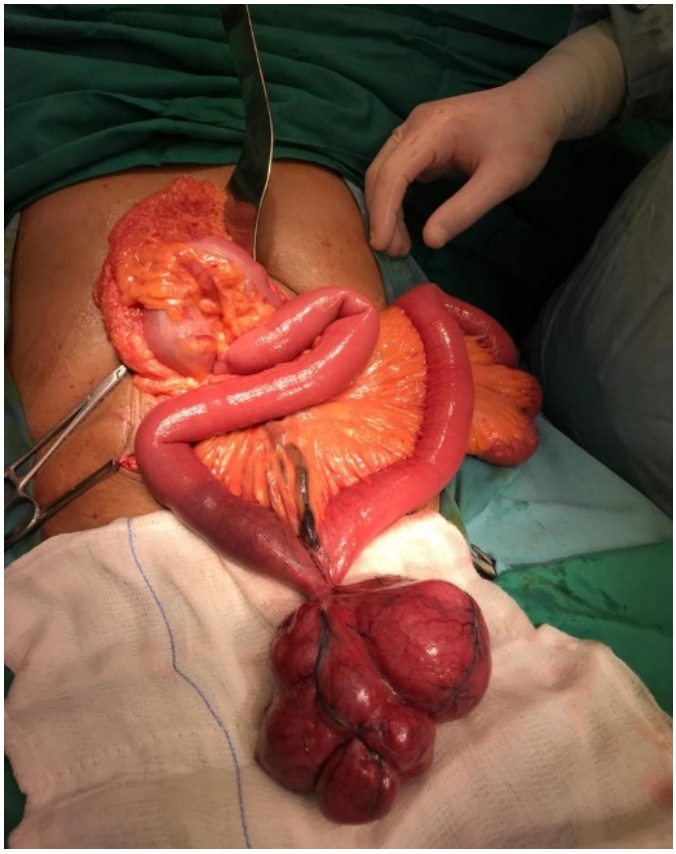

Figure 1. Large tumor obstructing the bowel.

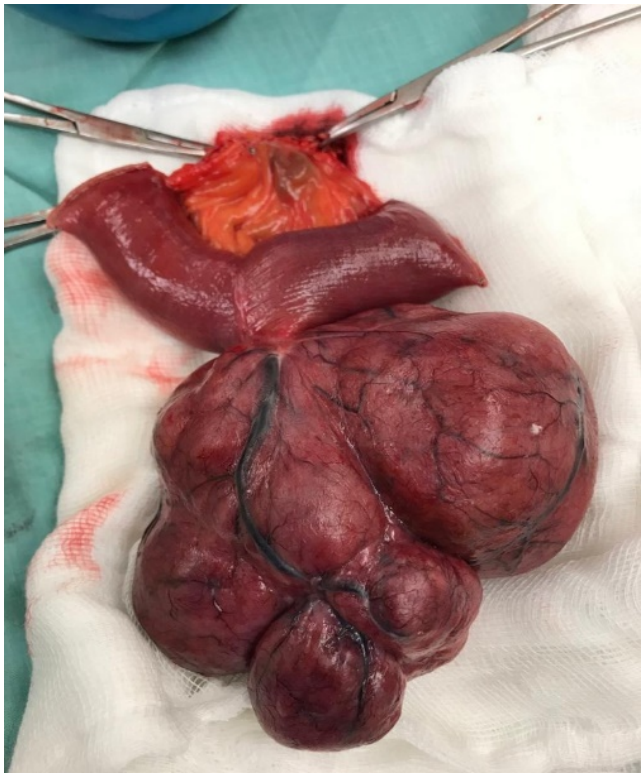

Figure 2. Tumor after bowel resection.

3. Discussion

Small intestinal GISTs account for 20% - 30% of gastrointestinal stromal cell tumors and the most common site of small bowel is jejunum [3], as in our case presentation.

The diagnosis of small bowel GISTs is usually late as the result of nonspecific symptoms and difficulty in detecting the tumor in early images, which is required a high incidence of suspicion from the clinician [4].

The nature of progression and invasion of small bowel GISTs is more than the stomach GISTs [5] [6]. The presentation in our case was the same with aggressive large obstructing tumor occlude hall lumen of the jejunum (**Figure 1** & **Figure 2**).

Most small-intestinal neoplasms are asymptomatic. The most common mode of presentation is partial small bowel obstruction, with associated symptoms of colicky abdominal pain, abdominal distention, nausea, and vomiting. Obstruction can be caused by luminal narrowing by the tumor itself or intussusception. Hemorrhage is the second most common mode of presentation and physical examination is usually nonsignificant. However, up to 25% of patients with all small-intestinal malignancies including GISTs are reported to have a palpable abdominal mass [7] [8].

The diagnosis of small intestine GISTs is usually challenging and it is often neglected clinically. It is characterized by low morbidity, common clinical symptoms, and nonspecific imaging characteristic features. However, the clinical history and examination, imaging examinations and endoscopic examinations can help to reach the diagnosis [9].

The diagnostic modalities for small bowel GIST included ultrasound, CT scan, CT angiography, and MRI. Furthermore, an abdominal ultrasound scan is the low sensitivity of no more than 26% in the diagnosis of small intestine GISTs [10].

CT scan can be used for the detection, localization, staging, surgical planning, and in the evaluation of response to therapy, including monitoring postoperative follow-up in patients with GIST [11].

CTA is used for the diagnosis of small bowel tumors with GI bleeding with a reported diagnostic sensitivity of 90.9% for small bowel GIST [12].

MRI is good for soft tissue contrast and is also an effective imaging modality when differentiating small bowel GIST from other small bowel tumors [13].

Small-intestinal GISTs should be treated with segmental intestinal resection. If the diagnosis is known prior to resection, wide lymphadenectomy can be avoided as GISTs are rarely associated with lymph node metastases [8] [13].

Medical therapy includes tyrosine kinase inhibitors (TKIs), which are usually given as the treatment of metastatic and unrespectable GISTs. TKI is given in patients with metastatic and unresectable GISTs as definitive therapy [14].

Adjuvant therapy is given after surgery and continued for 3 years to decrease the incidence of recurrence of GISTs with a high risk of recurrence rate [14].

4. Conclusions

The patients who presented with chronic nonspecific abdominal pain needed to

be investigated with more advanced radiological investigations to roll out progression of internal pathology (like tumors).

Diagnosis of small intestine GISTs is challenging because of slow progression, the lack of specific symptoms, and the lack of specific modalities of investigation for small bowel tumor and it requires a high index of suspicion.

The treatment of localized small bowel GISTs is usually surgical with segmental resection and bowel anastomosis.

Tyrosine kinase inhibitor (imatinib) is the chemotherapy drug of choice for adjuvant and neoadjuvant treatment for GISTs.

The prognosis of a patient with GISTs depends on early detection, size, site, mitotic division, and metastasis of the tumor.

Right to Privacy and Informed Consent

The authors declare that no patient data appear in this article.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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