Successful En Bloc Resection of Complicated Giant Stomach Gastrointestinal Stromal Tumor in an Elderly Patient

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Patient: Female, 83-year-old

Final Diagnosis: Gastrointestinal stromal tumour

Symptoms: Abdominal discomfort • abdominal distension

Medication: —

Clinical Procedure: Colectomy • distal subtotal gastrectomy

Specialty: Surgery

Objective: Unusual clinical course

Background: Gastrointestinal stromal tumors (GISTs) are the most common mesenchymal tumors of the gastrointestinal tract and mostly affect the stomach. The size of the tumors vary, ranging from 0.6 cm to 25.5 cm, with a median size of 6.8 cm.

Case Report: We report a case of a giant GIST (25×18×8.5 cm) in an 83-year-old woman, which we believe is the largest reported GIST, in Borneo, Malaysia. She presented with gradually increasing abdominal distension with occasional discomfort on movement for 1 month. Computed tomography revealed a large multinodular enhancing mass measuring 10×20×22 cm with no clear plane with the posterior gastric wall, duodenum, and pancreas. We performed a distal gastrectomy and transverse colon segmentectomy, as the tumor was plastered to the mesentry of the transverse colon. Despite extensive surgery, she recovered well after surgery. Due to her advanced age and the tumor size, a tyrosine kinase inhibitor was not given owing to the possibility of adverse effects.

Conclusions: The management of GIST is complicated, especially for a huge GIST with local invasion. Despite the benefits of a tyrosine kinase inhibitor, the role of surgery in managing GIST is crucial, especially for patients with huge tumor size, advanced age, and local complications from the tumor.

Keywords: Borneo • Case Reports • Gastrointestinal Stromal Tumors • Stomach Neoplasms

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Background

Gastrointestinal stromal tumors (GISTs) are the most common mesenchymal tumors of the gastrointestinal tract, accounting for 85% of tumors [1,2]. Most GISTs that occur in the gastrointestinal tract are located in the stomach (53%), which are derived from interstitial cells of Cajal, whereas other GISTs are reported to be small intestine (32%) and colorectal (15%) [2-4]. The sizes of GISTs range from small to large lesions. GISTs are clinically asymptomatic until they reach a significant size [1]. GISTs in the stomach related to symptoms are a median size of 6 cm [5]. Clinically, symptomatic patients have a median tumor size of 8.9 cm, whereas tumors detected incidentally have a median size of 2.7 cm [6].

The most common presentation of a stomach GIST is upper gastrointestinal bleeding and gastric discomfort. Bleeding varies from life-threatening to chronic bleeding, which leads to anemia. Gastrointestinal bleeding is due to the high vascularity of the tumor and erosion of the mucosa of the gastrointestinal tract. However, spontaneous hemoperitoneum due to GIST rupture is relatively rare [7,8]. Most patients present with abdominal pain at the onset of tumor rupture [9].

For GISTs, the treatment strategy depends on tumor site, size, and evidence of metastasis. Surgery is the criterion standard in the management of large and non-metastatic GISTs. We present a case of the largest stomach GIST encountered in north Borneo, which was successfully removed with en bloc surgical resection, inclusive of the distal stomach and transverse colon segment.

Case Report

An 83-year-old woman presented with gradually increasing abdominal distension with occasional discomfort on movement for 1 month. No other gastrointestinal symptoms were present. Clinical examination revealed a well-defined transversely mobile lump over the epigastrium, extending to the pelvis and measuring 13×13 cm, which was firm and not tender. She was hemodynamically stable and anemic on presentation, with a hemoglobin level of 9.1 g/dL (reference range: 11-13 g/dL).

An abdominal ultrasound revealed a large tubular heterogeneous mass in the pelvis but was unable to determine the origin of the mass. Abdominal contrast-enhanced computed tomography (CT) revealed a large multilobulated enhancing mass of approximately 10×20×22 cm with a hypodensity component mainly at the central aspect (Figure 1). There was no clear plane with the posterior gastric wall, duodenum, and pancreas. Intraabdominal free fluid was seen at the perihpatic, perisplenic, right paracolic gutter, and pelvis, with a Hounsfield unit value of 26. Upper and lower gastrointestinal endoscopy revealed an external compression over the sigmoid colon. Clinical-radiological features suggested a stomach gastrointestinal stromal tumor.

Her abdomen was explored and an intraperitoneal tumor measuring 25×15 cm, originating from the body of the stomach, was found. There was also 2 L of hemorrhagic fluid in the peritoneal cavity. It was a heavily vascularized intraperitoneal tumor with significant adhesions with the lateral peritoneal wall and base of the liver. Adhesiolysis was done using electrocautery and blunt dissection. There were dense adhesions between the tumor and segment of the transverse colon, resulting in a distal gastrectomy and segmental transverse colon resection (Figure 2). She underwent a 5-h surgery with an estimated blood loss of 1 L. The patient recovered well and was discharged on day 9 after surgery.

Pathological examination of the specimen revealed a multilobulated tumor of 255×185×85 mm, weighing 3670 g, with the main tumor attached to the distal stomach. The specimen showed a circumscribed lesion composed of plump to epithelioid neoplastic cells arranged in diffuse sheets and nests. The neoplastic cells (Figure 3A, 3B) showed round to oval, vesicular nuclei, variably prominent nucleoli, and eosinophilic cytoplasm with ill-defined cell borders. There were also scattered bizarre-looking and multinucleated giant tumor cells noted in the specimen with focal myxoid change and tumor necrosis. There were aberrant forms of mitosis, >10 per 5 mm². The lesion invaded the serosa of the stomach, where the border between the lesion and stomach appeared irregular and infiltrative. The muscularis propria and subserosa of the transverse colon were involved as well. Immunohistochemistry (Figure 3C, 3D) on most of the neoplastic cells was positive for CD117. The overall pathological features were suggestive of a GIST with a prognostic category of 6b (high risk). Molecular typing was not done because it is not available in our center. Thus, we were unable to rule out a wild GIST.

Due to the patient’s advanced age and the size of her GIST, the kinase inhibitor imatinib might have been associated with an even worse adverse effect for our patient; therefore, it was not given. We followed up our patient monthly after her discharged and planned for a surveillance CT and upper gastrointestinal endoscopy at the 6-month follow-up. During the course of her follow-up, she has been well and has shown no symptoms of recurrence such as bleeding, obstruction, or any abdominal mass.

Discussion

Our patient presented with worsening abdominal distension for 1 month and was found to have an enormous GIST tumor.
of the stomach causing external compression to the sigmoid colon from the lower endoscopy. The average size of a stomach GIST on presentation is 8.78 cm [10]. Tumor size is one of the factors that contribute to the clinical presentation of symptoms. A population-based study showed that patients who present with clinical symptoms have a mean tumor size of 8.9 cm, whereas asymptomatic patients have a mean tumor size of 2.7 cm [11]. These clinical symptoms include overt bleeding, tumor rupture, pain due to mucosal ulceration, abdominal mass, anorexia, weight loss, refractory hypoglycemia, and external palpable mass [12,13]. Our patient had clinical symptoms with a tumor size of 25.5 cm.

GIST is a mesenchymal tumor that originates from the interstitial cells of Cajal, which are located within the muscle layers of the gastrointestinal tract. Therefore, GISTs commonly originate from the muscularis propria layer of the gastrointestinal tract. GISTs often demonstrate an exophytic growth pattern toward the peritoneal cavity, and patients usually remain clinically asymptomatic until the tumor reaches a large size [14].

**Figure 1.** An axial view of the contrast-enhanced computed tomography scan showing heterogenous lesion (A) (white arrow) arising from the posterior aspect of the stomach (black arrow) with (B) areas of hypodensity. The lesion which was seen from the (C) coronal and (D) sagittal view.
Large GISTs can invade the adjacent organs from the origin of the tumor and have a high potential for distant metastases. Surprisingly, in our patient, there was a local invasion to the transverse colon but no evidence of distant metastasis to other organs, although the tumor was so huge.

Radiological diagnosis plays an important part in the diagnosis of a GIST [15]. GISTs are known to show a wide spectrum of radiological appearances depending on tumor size, imaging modality, site, and growth pattern of the tumor. Abdominal ultrasound usually finds a large mass in the abdomen that is heterogeneous and has frequent central necrosis. The site and origin of any intraabdominal mass are often undetectable by ultrasonography [16,17]. Because most GISTs appeared to be well-defined, extraluminal, or intramural masses, GISTs are better visualized on CT images. In the present case, the patient’s CT images showed a multilobulated mass with central hypodensity,

![Figure 2](image2.png)

**Figure 2.** Distal gastrectomy and segmental transverse colon specimen, weighing 3.67 kg, and the measurement is compared to the Deaver retractor.

![Figure 3](image3.png)

**Figure 3.** The histology at (A) ×10 and (B) ×40 magnification demonstrated neoplastic cells showing round to oval, vesicular nuclei, variably prominent nucleoli, and eosinophilic cytoplasm with ill-defined cell borders. Scattered, bizarre-looking, and multinucleated giant tumor cells are also noted in the specimen with focal myxoid change and tumor necrosis. (B) There are aberrant forms of mitosis, >10 per 5 mm². Positive immunohistochemistry staining of CD117 at (C) ×10 and (D) ×40 magnification.
likely representing central necrosis, which is usually reported in tumors sized ≥6 cm [18]. Ideally, to get the confirmation of the diagnosis of GIST, certain centers perform a tumor biopsy with endoscopic ultrasound-guided fine-needle aspiration. Because the GISTs of 10 cm or larger tend to have many larger vessels, they are prone to cause hemorrhage [19]. Biopsy was not taken before surgery for our patient because of the risk of hemorrhage in a large GIST tumor. Therefore, the diagnosis was made based on clinical features combined with radiological features, such as exogastric growth, diameter ≥5 cm, central necrosis, and presence of extension to other organs [20].

For large GISTs with no evidence of distant metastasis, surgery remains the treatment of choice. The goal of resection is complete macroscopic and microscopic resection (R0) without rupture of the pseudocapsule [21]. In our patient, the huge GIST originated from the distal stomach and invaded part of the transverse colon. Our upper gastrointestinal surgeon performed a distal gastrectomy and transverse colon segmentectomy to remove the huge GIST en bloc.

The US Food and Drug Association approved the use of tyrosine kinase inhibitors (TKIs) in the management of advanced GISTs, with imatinib as the first-line therapy, sunitinib as the second-line therapy, regorafenib as the third-line therapy, and ripretinib as the recently approved fourth-line therapy. The prognosis for advanced GISTs has improved dramatically since the emergence of TKI. Imatinib can either be used as neoadjuvant therapy to reduce tumor size or adjuvant therapy to prevent tumor recurrence among patients with high-risk features [22]. However, neoadjuvant therapy with imatinib was not given for our patient because the tumor was large, with a compressive effect on the colon. In addition, imatinib does cause adverse effects, such as periorbital edema, nausea, diarrhea, muscle cramps, muscles aches, joint pain, tiredness, hematologic toxicity, congestive heart failure, tumor lysis syndrome, hypothyroidism, and hepatotoxicity. Due to elderly age and the size of her GIST, imatinib might have been associated with an even worse adverse effect for our patient. Therefore, there is still a role for surgery in the management of GIST, despite the current recommendation of neoadjuvant therapy with TKI. Surgery is an important option for patients with large GISTs and the elderly. These groups of patients benefit from surgery because they can avoid the adverse effects of TKI and it can also prevent further complications from the GIST, such as hemorrhage, invasion to more adjacent organs, and distant metastasis.

Conclusions

The management of GIST is complicated, especially for a huge GIST with local invasion. Despite the benefits of TKI, the role of surgery in managing GIST is crucial, especially for patients with a large tumor size, advanced age, and local complications from the tumor.

Declaration of Figures’ Authenticity

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