Massive Gastro Intestinal Bleeding Revealing an Ileal GIST: A Case Report and Literature Review

Yassine Y1*, Rajih D1, Izaabel R2, Nacir O1, Lairani F1, Errami AA1, Oubaha S1, Samlani Z1, Krati K1

1Gastroenterology Department, Mohamed VI Hospital Center, Marrakech, Morocco
2General Surgery Department, Mohamed VI Hospital Center, Marrakech, Morocco

Abstract

Gastrointestinal stromal tumors (GISTs) are the most common malignant subepithelial lesions of the gastro-intestinal tract, they can be difficult to diagnose and have a varied clinical outcome. We report the case of a 70-year-old man who presented a recurrent and severe gastrointestinal bleeding revealing an ileal GIST. Upper GI endoscopy and colonoscopy showed no abnormalities, meanwhile abdominal CT scan revealed the tumor. The patient underwent surgery and the diagnosis of an ileal GIST was confirmed by histopathological evaluation.

Keywords: (GISTs), gastro-intestinal tract, diagnose, tumor.

INTRODUCTION

Gastrointestinal stromal tumors (GISTs) are rare mesenchymal neoplasms of the gastrointestinal tract. They are most commonly located in the stomach (50-60%) followed by small intestine (30-40%), colorectal (5%), and esophagus (1.6%) (Joensuu, 2006). These tumors can be asymptomatic or have various clinical presentations such as abdominal pain, bleeding, vomiting, abdominal mass or weight loss. Although bleeding is a frequent symptom seen in GISTs, significant acute bleeding is an unusual finding (Woodall et al., 2009). Here we report the case of an ileal GIST in a 70-year-old man, revealed by a recurrent and severe GI bleeding.

CASE PRESENTATION

A 70-year-old man, with no prior medical history, was referred to our department for the management of recurrent massive melaena and fatigue in the past month. Clinical examination was normal except for a pale skin color and a right iliac fossa pain. Heart rate was 115 beats per minute and blood pressure 100/58 mmHg. Blood tests showed severe anemia with hemoglobin of 3.8 g/dL (MCV 84 fL). Other biochemical parameters and liver function tests were normal. On the second day of admission, an upper GI endoscopy and colonoscopy with ileal intubation were performed and showed no abnormalities. Abdominal computed tomography (CT) revealed an 87x62 mm heterogenous mass at the distal ileum with exophytic growth, Figure 1 and 2).

Figure 1: Axial contrast-enhanced CT portal venous phase image shows a 8.8x6.3 cm tumor of the distal ileum with exophytic growth

Figure 2: Axial contrast-enhanced CT image shows an 8x6, 5 cm tumor of the distal ileum with heterogeneous enhancement
Laparotomy was then performed, showing a huge mass measuring 8x7 cm arising from the distal ileum, with a prevalent extramural growth and adhering to the caecum (Figure 3). No other intra peritoneal abnormalities were detected. An ileocecal resection removing the mass en-bloc with an end-to-end ileocolic anastomosis were performed without tumor rupture.

Figure 3: Resection specimen showing an extramural appearance of the ileal tumor

Pathology reported a high-grade ulcerated GIST with classic spinal cells with clear resection margins. Immunohistochemistry was positive for c-kit (CD117) and CD34 immunomarkers. Ki67 proliferation marker labels 10% of the nuclei. Mitotic activity was 18/50 on high-power fields. The post-operative course was complicated by the occurrence of an anastomotic leakage for which the patient underwent another surgery and was admitted to the intensive care unit where he passed away due to a septic shock.

**DISCUSSION**

Gastrointestinal stromal tumors (GISTs) are the most common malignant subepithelial lesions (SEls) of the gastro-intestinal tract. They originate from the interstitial cells of Cajal (Akahoshi et al., 2018). These cells, which are located in the myenteric plexus within the muscle layer, are pacemaker cells that cause gut peristaltic contractions (Joensuu, 2006). GISTs occur anywhere along the gastrointestinal tract but are most common in the stomach (50–60%) and the small intestine (30–35%), which was the location in our case, and less frequent in the colon and rectum (5%) and the oesophagus (<1%). GISTs found elsewhere within the abdominal cavity, usually in the omentum, mesentery, or the retroperitoneum (<5% of all GISTs), are referred to as extra-gastrointestinal tract tumours, or E-GISTs (Corless et al., 2011; Husain et al., 2023; Joensuu et al., 2012). GI bleeding is one of the most common presentations and occurs due to the erosion of GI tract lumen. It is seen in about 30% of patients. They may present with haematochezia, hematemesis, melena or unexplained anemia (Blanke et al., 2005).

In a retrospective review of 32 cases of primary GIST of the small bowel, Zhou et al showed that most cases were asymptomatic at presentation and the most frequent presenting symptom was GI bleeding (Zhou et al., 2018). The degree of hemorrhage varies from asymptomatic chronic to massive life-threatening. However, significant acute bleeding, such as the presentation of our patient, is an unusual finding (Husain et al., 2023). Other symptoms can be seen, such as abdominal pain, distension, and discomfort due to a tumor-induced mass effect. More rarely, obstructive syndromes or organ perforations might be a revealing form (Tran et al., 2005). Some GISTS can be difficult to detect by endoscopy due to their exophytic growth pattern, which was the case in our patient. CT scan provides the basis for diagnosis and staging in most stromal tumors. It exhibits exophytic growing hypervascular tumors and enhance inhomogeneously (Michael King, 2005). EUS can be useful in the diagnosis of gastric or rectal GISTs since it distinguishes the different layers of the wall. Capsule endoscopy is a safe and painless method for mucosal imaging of the small bowel, in case of no obstruction. However, double balloon endoscopy enables endoscopic inspection of the entire small bowel with the ability to take biopsy samples and with the potential to administer localized therapy (Zhou et al., 2018). Biopsy is not mandatory prior to surgery, except to rule out differential diagnoses like lymphoma or other malignant or benign neoplasms (Beham et al., 2012).
GISTs are usually well circumscribed and surrounded by a pseudocapsule. Microscopically, GIST cell morphology is usually spindle-shaped but some consist of rounded cells (epithelioid type, 20%) or a mixture, but they can also be pleomorphic (Joensuu, 2006; Miettinen et al., 2002). Large tumors often show cystic degeneration or central necrosis. 95% of GISTS stain positively for the KIT protein (the CD117 antigen, an epitope of the KIT tyrosine kinase) (Tryggyvason et al., 2005). An isoform of protein kinase C, PKC-h, is highly expressed and constitutively phosphorylated in many GISTS, and PKC-h may be useful for identifying KIT-expression-negative GISTS. They also express CD34 (70%) and smooth muscle actin SMA (30–40%) (Blay et al., 2004). Joensuu suggested in 2008 and risk stratification for patients with GIST. They can be classified as very low risk, low risk, intermediate risk, and high risk based on tumor diameter, mitotic count and primary location of tumor (H, 2008). Malignant potential of GISTS ranges from small lesions with a benign behavior to aggressive sarcomas, and prognosis is usually based on tumor location, size, and mitotic activity(Miettinen et al., 2002). In a prospective cohort including 200 patients diagnosed with GISTS, Ronald et al. found out that a tumor size of 10 cm carried a recurrence relative risk of 2.5 (confidence interval 1.2–5.5), and tumor diameter could also predict disease-specific survival in patients with primary disease who undergo complete gross resection (DeMatteo et al., 2000). Other factors suggested to be associated with an adverse outcome include presence of tumor necrosis, high cellularity and marked pleomorphism, a high S-phase fraction, presence of telomerase activity, and presence of KIT exon 11 deletion mutation.(Fletcher et al., 2002).

From a therapeutic perspective, surgery is the standard treatment for localized GISTS (Kang et al., 2012). The tumor should be removed en-bloc with its pseudocapsule to yield an adequate resection margin. The optimal width of tumor-free margin has not been defined. Regional lymph node resection is of unproven value, since GISTS rarely give rise to lymph node metastases. Low and intermediate risk GISTS do not require adjuvant treatment, whereas high risk GISTS with mutations sensitive to Imatinib are usually treated with three years of Imatinib (Schafer et al., 2017). (ESMO/European Sarcoma Network Working Group, 2014) . Imatinib is also considered as the standard treatment of metastatic GIST. Approximately 65–70% of patients achieve a partial response, and another 15–20% have stable disease (van Oosterom et al., 2001).

**CONCLUSION**

GISTS are encountered infrequently. In this report, we presented the case of an ileal GIST with active and massive bleeding, which is seen rarely in clinical practice. Intestinal GIST should be kept in mind in patients with GI bleeding of unknown origin. Diagnosis can be supported by endoscopic or imaging techniques, including CT and MRI, with the definitive diagnosis made by histopathology and immunohistochemistry.

**REFERENCES**


