

Gastrointestinal bleeding and intussusception due to gastrointestinal stromal tumor (GIST)

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Abstract

Background: Small bowel tumors comprise 25% of gastrointestinal (GI) neoplasms, of which only between 0.2 and 1% correspond to gastrointestinal stromal tumors (GIST). GI bleeding is the most common presentation of GIST, being responsible in 1% of the cases. This type of neoplasm can also be the origin of an intussusception, which is an infrequent process during adult age, representing only 5% of all intestinal obstructions.

Clinical case: We report a case of a patient who arrived at the emergency department due to abdominal pain and lower GI bleeding. The diagnostic process was not conclusive. After an exploratory laparotomy, the diagnosis was GI bleeding and intussusception due to GIST.

Conclusions: Surgery must be the last diagnostic and therapeutic resource, but it is sometimes necessary to localize bleeding and intestinal obstruction site.

Key words: gastrointestinal bleeding, melena, intussusception, gastrointestinal stromal tumor.

Introduction

Gastrointestinal stromal tumors (GIST) are a rare cause (<1%) of gastrointestinal (GI) bleeding.¹ It is not often possible to localize the origin of the bleeding because of the difficulty in accessing lesions of the small intestine, with surgery being the last appropriate option, both diagnostic as well as therapeutic.²

Intussusception in adults is an uncommon process (5% of intestinal obstructions), and in 90% of cases is due to a parietal organ lesion with neoplastic etiology in 65% of cases, with adenocarcinoma being the most common histology. Treatment of choice is resection of the affected bowel segment, and there is controversy about the extent of the resection to be performed.³

We present the case of a 55-year-old female with lower GI bleeding secondary to jejunal GIST, which produced an intussusception. Due to its low frequency, adequate diagnosis is

imperative when being faced with lower GI bleeding of uncertain origin.

Clinical Case

The 55-year-old female patient admitted to a personal history of bronchial hyperactivity and duodenal polypectomy due to an isolated picture of upper GI bleeding. Upon her arrival to the emergency room she was pale, had periumbilical pain and was hemodynamically stable. Upon evaluation in the emergency room her CBC showed 3.67×10^6 erythrocytes/ μl ; hemoglobin, 10.8 g/dl; hematocrit, 31.4%; platelets, 136,000/ μl ; and leukocytes, 6400/ μl (67.1%). Upper and lower GI endoscopies, ultrasound, CT, scan and splachnic angiography were performed. All were found to be within normal limits. It was decided to conduct exploratory laparotomy because of a new episode of rectal bleeding and hemodynamic instability. Intussusception of a segment of intestine over a tumor of 5 cm in diameter with a hemorrhagic appearance was verified at the level of the small intestine, at a distance of 120 cm from the ileocecal valve (Figure 1). The resected tumor showed mucosal erosion (Figure 2), positivity for c-KIT (CD117) on immunohistochemical study, negative for CD34, specific for smooth muscle actin, desmin and S100 protein. The final pathological diagnosis was a low-grade malignant GIST. In the absence of metastasis and low degree of malignancy of the lesion, it was decided to follow the patient clinically without resorting to adjuvant treatment.

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Discussion

Small bowel tumors represent 0.3% of all malignancies¹ with the age range of onset being between 65 and 74 years. The most common location is at the level of the ileum (29.7%) followed by duodenum (25.4%) and jejunum (15.3%).¹ There are no conclusive figures on the incidence of various malignant neoplasms among different series because sometimes carcinoids are established as the most frequent (33%) followed by adenocarcinoma (30%) and lymphomas (16%),¹ whereas others place in order of frequency adenocarcinomas to carcinoid tumors (35-50% and 20-40%), respectively.²

On the contrary, there are similarities in terms of less common malignancies, i.e., GIST, which are estimated at 11-13%.^{1,2} Small bowel tumors are characterized by nonspecific symptoms that together with the difficulty of diagnosis encourage tumor growth. At the time of diagnosis, the tumor will generally be found in advanced stages.¹ The diagnostic technique of choice is computed tomography (CT). Other procedures such as ultrasound, endoscopy, intestinal capsule or positron emission tomography (PET) may also be used.⁴

GISTs are neoplastic processes derived from neoplastic interstitial cells of Cajal, located at the GI myenteric plexus level, which may appear along the entire digestive tract. GISTs are the most common mesenchymal neoplasms of the GI tract.^{4,5} Studies show that between 52 and 95% of cases are c-KIT (CD117) positive, whereas 35% of the situations that do not show c-KIT mutation have a mutation at the level of PDGFRA.^{2,4-6} The incidence of GIST has shown an increase in recent years, affecting >90% of cases in patients >40 years of age (mean: 55-60 years).¹

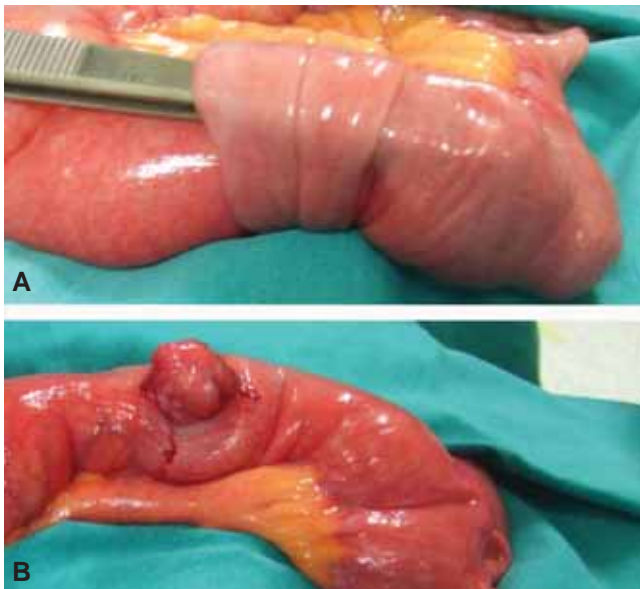


Figure 1. (A) Jejunum-jejunal intussusception. (B) Tumor at the level of the antimesenteric border.

In some series the highest prevalence occurs in males,¹ and other series show equal prevalence between genders. As for the location of these tumors, they are more frequent at the level of the stomach (50%), followed by the small intestine (25%), colon and rectum (10%) and may also appear in mesentery, retroperitoneum, omentum, pelvis, pancreas, liver and gallbladder.^{1,2,6,7} Fletcher et al. established that the lifetime risk of GIST depends on tumor size and mitotic index, although other prognostic factors have been proposed for these neoplasms invading the mucosa such as tumor necrosis and high cellularity.^{2,4}

The primary form of presentation is GI bleeding. Treatment of choice is surgical resection with adequate margins and without lymphadenectomy because metastatic development is through blood (liver and lungs) and peritoneal.

Because these types of neoplasms are resistant to adjuvant treatments of chemotherapy and radiotherapy, proper identification of GISTs is crucial since the discovery of a specific treatment for the inhibition of tyrosine kinase (imatinib mesylate).⁸ This is currently widely used for metastatic and unresectable GIST because it inhibits proliferation and promotes apoptosis, with cure rates of 54% and symptomatic palliation in inoperable patients (90%).^{4,9} Tzen et al. in a review of 17,858 GI lesions showed that CD117-negative GISTs may be underdiagnosed unless a specimen detection of KIT and PDGFRA genes is done, noting a higher annual incidence figures when diagnosis is made jointly using immunohistochemistry and mutation study.⁹

Intussusception or intestinal invagination is a rare entity in adults (<1% of all obstructions), generally secondary to a tumor that acts as cephalic traction of the invagination.³ Its usual clinical presentation is a mechanical ileus of acute or subacute presentation, with the diagnosis made intraoperatively on multiple occasions.

Intussusception is the most common cause of intestinal obstruction in childhood and is of idiopathic etiology. It is probably related to intestinal motility disturbances and appears most often in the ileocolic segment at the level of the Bauhin valve. Intussusception in adults is usually secondary to organic processes of the abdominal wall (90%), with adenocarcinoma being the



Figure 2. Surgical specimen showing ulceration of the intestinal mucosa.

most common etiologic factor.⁹ Among the benign processes, the most common causes are lipomas, inflammatory lesions of the intestinal wall and Meckel's diverticulum. CT, colonoscopy, plain abdominal x-ray, barium enema and ultrasound are methods that can be effective for diagnosis, with the possible need to resort to laparotomy for definitive diagnosis.^{3,10} Treatment is surgical and consists of resection of the affected intestine, avoiding the simple manual reduction of the invagination from the most minimal suspicion of malignancy.¹⁰

In conclusion, we propose that proper treatment of GIST requires a multidisciplinary approach that should include oncological surgery and a thorough study, both pathological and molecular, as this will determine the appropriate adjuvant treatment.

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