

Proximal jejunal GIST associated with massive gastrointestinal bleeding. Case report

*GIST en yeyuno proximal asociado a hemorragia masiva de tubo digestivo.
Reporte de caso*

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Abstract

Introduction: Gastrointestinal stromal tumors (GIST) are rare, reported incidence is between 10 to 15 cases per million of habitants. They are usually located in the stomach (56%), small intestine (32%), colon-rectum (6%), and esophagus (<1%). Its symptoms include nausea, vomiting and abdominal fullness; 30% are asymptomatic. Incidental finding during abdominal surgery or imaging studies is common. Resection with negative margins is the standard treatment. **Case report:** A 69-year-old female patient who debuted with massive digestive tract bleeding, requiring surgical treatment. A tumor was detected at jejunum compatible with a GIST.

Keywords: Gastrointestinal Stromal Tumors. Jejunum. Small Intestine. Hemorrhage. Tumor.

Resumen

Introducción: Los tumores del estroma gastrointestinal (GIST) son poco frecuentes, con una incidencia de 10 a 15 casos por millón de habitantes. Suelen localizarse en el estómago (56%), el intestino delgado (32%), el colon-recto (6%) y el esófago (< 1%). Sus síntomas incluyen náusea, vómito y plenitud abdominal; el 30% son asintomáticos. Es común su hallazgo incidental durante una cirugía abdominal o en estudios de imagen. La resección con márgenes negativos es el tratamiento estándar. **Caso clínico:** Mujer de 69 años que debuta con hemorragia masiva de tubo digestivo, requiriendo tratamiento quirúrgico. Se detecta un tumor de yeyuno compatible con GIST.

Palabras clave: Tumores del estroma gastrointestinal. Yeyuno. Intestino delgado. Hemorragia. Tumor.

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Introduction

Gastrointestinal stromal tumors (*GIST*, gastrointestinal stromal tumor) are rare tumors of the GI tract, originating from mesenchymal tissue¹. Most studies report a clinical incidence of 10-15 cases per million people².

GISTs are typically located in the stomach (56%), followed by the small intestine (32%), the colon-rectum (6%), and the esophagus (< 1%). Occasionally, they can affect the omentum, mesentery, and peritoneum. Initial symptoms may include nausea, vomiting, and abdominal fullness, although 30% of cases are asymptomatic; however, incidental detection of small, asymptomatic lesions during abdominal surgery or radiological and endoscopic studies is common^{1,2}.

Immunohistochemical staining for the CD117 antigen is positive in 95% of GISTs and is considered a cornerstone for diagnosis³.

Resection with negative margins remains the standard treatment for GIST patients. The anatomical location of the tumor is a well-documented prognostic factor after radical surgery, with tumors in the small intestine having a worse prognosis than gastric tumors but a better prognosis than colorectal tumors, with extraintestinal locations being the least favorable⁴.

Case report

A 69-year-old woman with a past medical history of total abdominal hysterectomy 35 years ago for uterine fibroids and left renal cyst resection 13 years ago, without apparent complications. She reports multiple red blood cell transfusions due to anemia of unknown origin. She has no other relevant history for her current condition.

She presented with a clinical course characterized by rectal bleeding without accompanying symptoms, prompting a diagnostic workup. Colonoscopy showed the rectum and sigmoid colon with abundant clots and 12 uncomplicated diverticula in the sigmoid colon; the rest of the study was unremarkable. At that time, she was diagnosed with lower GI bleeding due to self-limiting diverticular disease. However, she continued to have a clinical course characterized by multiple melena-like stools, requiring hospitalization under the gastroenterology service for a diagnostic and therapeutic workup. An intestinal transit study was performed, with no abnormalities, and antegrade enteroscopy revealed a 7 mm thickened mucosal

lesion in the proximal jejunum with color change and another raised, friable, ulcerated lesion, 20 mm in diameter, 10 cm distal to the former one. Biopsies were taken, with subsequent profuse arterial bleeding. A consultation was made with our service during the procedure, but no emergency surgical procedure was required due to cessation of bleeding following the placement of 2 hemoclips, with hospital observation. During her hospitalization, she had another episode of lower GI bleeding with copious bloody stools and signs of hypovolemic shock, along with a drop in hemoglobin down to 4.3 g/dL. Due to massive hemorrhage and signs of hemodynamic instability, treatment was initiated with red blood cell transfusion, and surgical treatment with exploratory laparotomy was performed, identifying a 4 cm × 2 cm hypervascularized tumor in the proximal jejunum, located 20 cm from the Treitz angle, non-perforated (Fig. 1), with no free fluid or other relevant findings. An intestinal resection of 10 cm was performed 10 cm from the Treitz angle, with mechanical side-to-side entero-enterostomy. The specimen was sent for definitive histopathological study. During surgery, red blood cell transfusion was administered, with a postoperative hemoglobin level of 8.4 g/dL.

After surgery, the patient was admitted to the hospital ward, dependent on vasoactive amines, with norepinephrine at a dose of 0.08 µg/kg/min, and was kept under observation in a joint management with gastroenterology and digestive surgery. With adequate clinical progression and an increase in hemoglobin to normal levels, she was discharged 8 days after surgery and scheduled for follow-up in the outpatient clinic.

She returned to the outpatient digestive surgery clinic with a histopathology report concluding a low-grade 4 cm × 3 cm × 2.5 cm GI stromal tumor (*GIST*), with no necrosis and negative tumor margins, with < 5 mitoses in 50 high-power fields. Hematoxylin and eosin staining (Fig. 2) and immunohistochemistry were performed, with positive DOG1 and CD117 markers (Fig. 3); CKAE1/AE3, S100, and smooth muscle actin tested negative.

She was referred for oncology evaluation, with no need for adjuvant treatment due to the R0 tumor resection and absence of metastatic disease in extension studies, so she remains under medical monitorization.

Discussion

GISTs located in the jejunum are rare, representing 0.1% up to 3% of all GI tumors. Some cases present

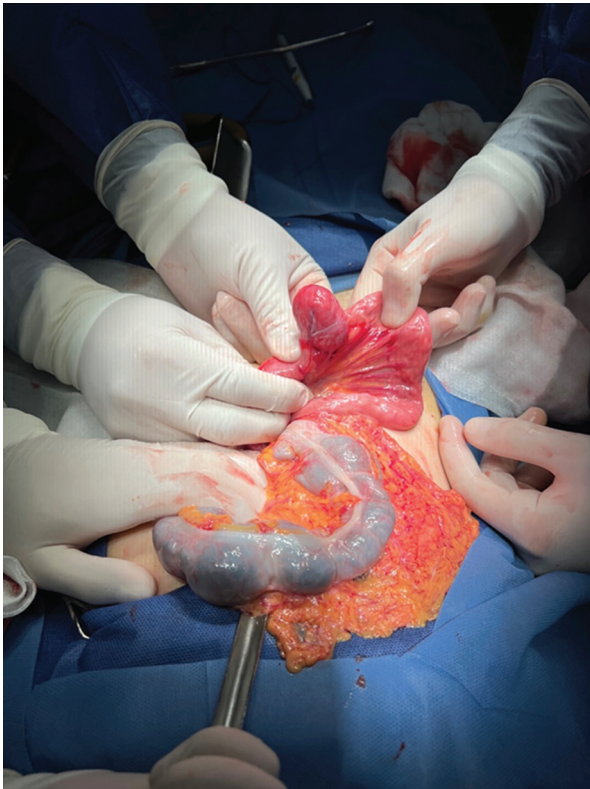


Figure 1. Jejunal GIST during exploratory laparotomy.

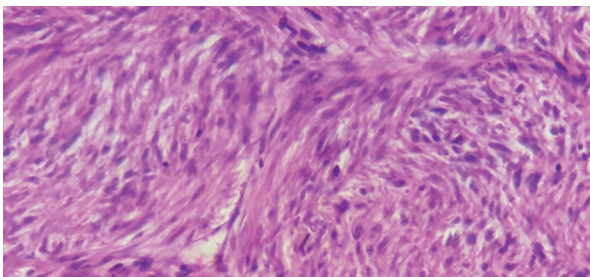


Figure 2. Hematoxylin-eosin staining at 40× magnification. Spindle-shaped neoplastic cells arranged in a syncytial pattern with elongated nuclei and eosinophilic cytoplasm.

with melena, hematemesis, and anemia due to recurrent bleeding. Acute GI bleeding is a common presentation in medical practice and may constitute a life-threatening emergency^{3,5}.

GI bleeding has been reported in nearly 25% of jejunal GISTs; however, it is unusual for them to present with massive GI hemorrhages requiring emergency intervention^{5,6}. In our case, the patient had massive bleeding, an uncommon presentation of this tumor, requiring emergency surgery during which a proximal jejunal tumor was detected.

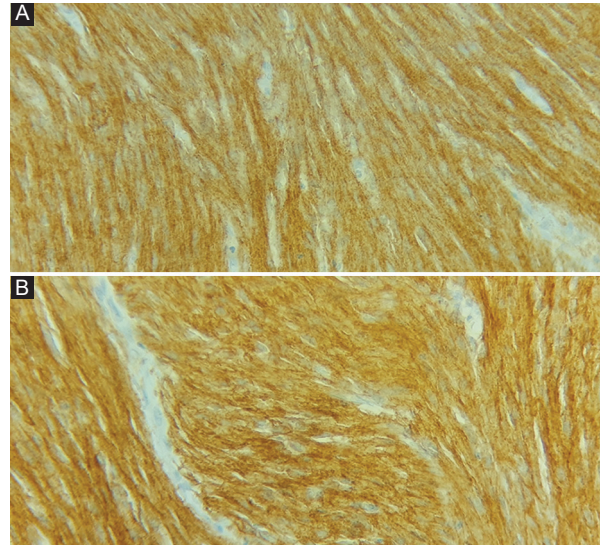


Figure 3. Immunohistochemistry showing positivity for DOG1 (A) and CD117 (B).

Upper and lower endoscopy remains the first-line study for GI bleeding. Bleeding originating from the small intestine remains a diagnostic challenge due to the inaccessibility of this region with conventional endoscopy. Additional imaging modalities are necessary when upper and lower endoscopies are negative for bleeding data, including capsule endoscopy, coronary computed tomography angiography (CCTA), IV or oral contrast CT, enteroscopy, and magnetic resonance enterography³.

Microscopically, most GISTs show 3 main histologic subtypes: spindle cell type (the most common, 70%), epithelioid type (20%), and mixed type (10%). Immunohistochemistry is necessary for differentiation^{6,7}.

GISTs are managed with various treatment modalities, including surgical resection, targeted medical therapies, chemotherapy, and radiotherapy; however, surgical resection is preferred due to the poor response of other modalities⁶. Surgical resection is the treatment of choice for any potentially resectable GIST > 2 cm, if the tumor is localized and symptomatic or if malignancy data are available on endoscopic ultrasound⁸. Postoperative surveillance alone is recommended if an R0 tumor resection (negative microscopic margins) is achieved¹. In our case, complete surgical resection of the lesion was performed, with a histopathology report of spindle cell GIST with tumor-free margins, which was considered a successful treatment, and the patient was kept under surveillance only.

The prognosis for GISTs depends on tumor size, anatomical location, imaging findings, metastasis, and tumor rupture⁸. Numerous studies suggest that immunohistochemical expression is related to prognosis⁹. Fletcher's risk stratification is an established method for classifying the risk of recurrence¹⁰.

Tyrosine kinase inhibitors are useful in the treatment of GISTs as neoadjuvant preoperative therapy for 6 to 12 months for cytoreduction of large tumors to make them operable. If tyrosine kinase inhibitors are considered as part of the treatment plan, tumor mutation analysis is highly recommended. KIT gene mutations are found in approximately 80% of GISTs¹¹⁻¹³.

Conclusions

GISTs represent a low-incidence condition, especially those located in the jejunum and those presenting with massive GI bleeding. Due to their aggressive and nonspecific presentation, it is important to be knowledgeable about this type of tumor to establish an early diagnosis and provide timely treatment, as resection with clear margins improves patient survival.

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Conflicts of interest

None declared.

Ethical disclosures

Protection of human and animal subjects. The authors declare that no experiments were performed on humans or animals for this study.

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

Right to privacy and informed consent. The authors declare that no patient data appear in this article.

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