

## Gastrointestinal Bleeding Secondary to Jejunum GIST, Report Case

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### ABSTRACT

The small intestine is by far the most difficult area to study from an endoscopic point of view, since traditional endoscopy only covers up to the 2<sup>nd</sup> portion of the duodenum and colonoscopy up to the terminal ileum. On rare occasions we find it necessary to use non-traditional endoscopic equipment to be able to study this middle area of the small intestine. Gastrointestinal stromal tumor is one of the neoplastic differential diagnoses found at this level. Given the advancement of genetics associated with immunohistochemistry, we have been able to better evaluate and characterize these lesions of mesenchymal origin. A case of a jejunal gastrointestinal stromal tumor with an initial presentation of melena is presented. Where together gastroenterology, surgery and pathology, the mentioned final diagnosis was reached.

### Introduction

Gastrointestinal stromal tumors (GIST) are the most common mesenchymal tumors of the gastrointestinal (GI) tract, yet they account for 1-3% of GI tumors. The possible cellular origin is from pluripotent stem cells programmed to differentiate into Cajal cells, the pacemaker of the GI tract. Among the most important findings that has been discovered is the proto-oncogene *c / kit*, whose gain in

functions has been shown to be present in GIST since 1998 [1]. We present a case of GIST from its initial clinical presentation as melena, until its surgical resection and histological diagnosis.

### Case Report

75 years old male, married, teacher, native and resident of Guatemalan rural area. History of left inguinal hernioplasty 15

years ago and alcoholism abandoned 10 years ago. Consultation with a history of intermittent melena of one month of evolution, associated with asthenia, adynamia, and loss of 20 pounds. Clinically with preserved functional status, generalized paleness, heart rate 105 bpm, P / A 110/75 mmHg, respiratory rate 14 bpm, cardiopulmonary without alterations, abdomen without palpable masses, painful left upper quadrant, digital rectal examination without melena. Laboratories showed hypochromic microcytic anemia with hemoglobin (Hb) at 6.6 g / dl. Transfusion therapy is started. Endoscopy and colonoscopy without relevant diagnostic findings. Abdominal tomography showing intraluminal mass from the small intestine, possibly jejunum measuring 7.48 x 4.95 x 13 cm (Figure 1), without distant disease. Therefore, a double balloon-assisted enteroscopy (Fujifilm EN-580T) is performed, approximately 80 cm from the dental arch, an exophytic lesion is evidenced, ulcerated with bleeding in layers with an approximate diameter of 6 cm (Figure 2), from which biopsies are taken which were reported as GIST. Laparotomy with small bowel resection was performed, intraoperative findings revealed an exophytic mass of approximately 10cm x 10cm x 5cm with multiple adhesions to the abdominal wall and transverse colon, located 20cm from the duodenum-jejunal angle (Figure 3). Small bowel resection was performed with meticulous dissection of the capsule. A primary lateral-lateral anastomosis was performed without lymphadenectomy. Abdominal cavity and surrounding organs were examined to rule out metastasis.

### Pathological Description

Enteric segment measuring 18x 17x 4.5 cm with a smooth surface, gray in color. A nodular lesion measuring 14 x 17 x 4.5 cm is identified. With a whitish surface and involves 90% of the intestinal mucosa (Figure 4).

### Microscopic

In the histological sections evaluated corresponding to the case, a neoplasm of mesenchymal origin formed by proliferation of spindle cells with fibrillar eosinophilic cytoplasm with central fusiform nucleus with fine granular chromatin without marked atypia is observed. 14 mitoses are counted in 50 high-power fields (40x). Said cells are arranged in bundles randomly (Figure 5).

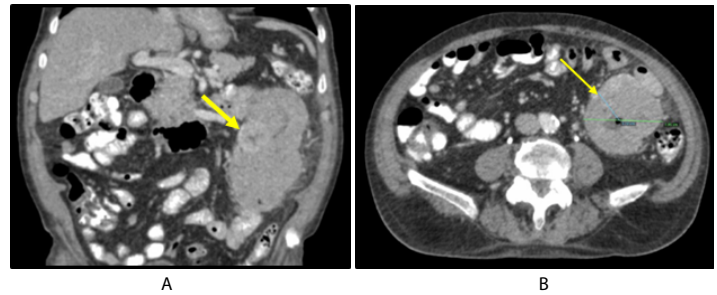
### Immunohistochemistry (Figure 6; Table 1)

Using the WINDOW BENCHMARK GX AUTOMATED complex method, immunohistochemical studies are performed with the following antibodies: CD 117 (EP10), DOG1 (SP31) and CD34 (QBend10) with the following results shown in Table 1.

With all the previously described findings, we reached the final diagnosis of a high-grade Gastrointestinal Stromal Tumor (GIST) with a high risk of metastasis.

**Table 1:** Immunohistochemistry.

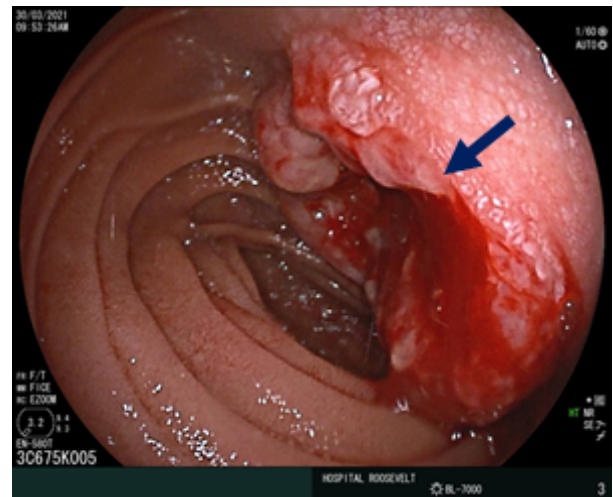
Antibody	Case	Control
CD 117	Positive in neoplastic cells	Positive
DOG 1	Positive in neoplastic cells	Positive
CD 34	Positive in blood vessels	Positive



**Figure 1:** Abdominal tomography.

A. Coronal section. B. Axial section. Circumferential thickening of the small intestine (Yellow arrow) of 7.48 x 4.95 x 13 cm in diameter, involving the jejunum in its left lateral aspect.

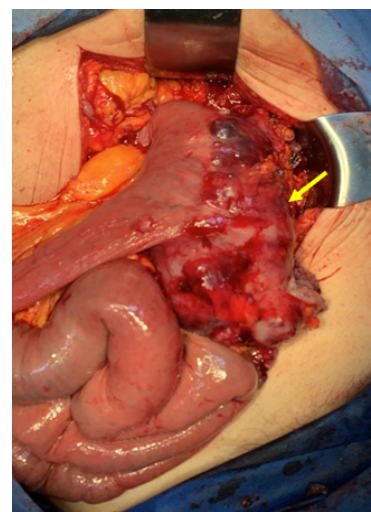
Source: Radiology Archive, Hospital Roosevelt.



**Figure 2:** Double balloon-assisted enteroscopy (Fujifilm EN-580T).

Exophytic, ulcerated lesion with oozing bleeding, with 6 cm approximate diameter (Blue Arrow).

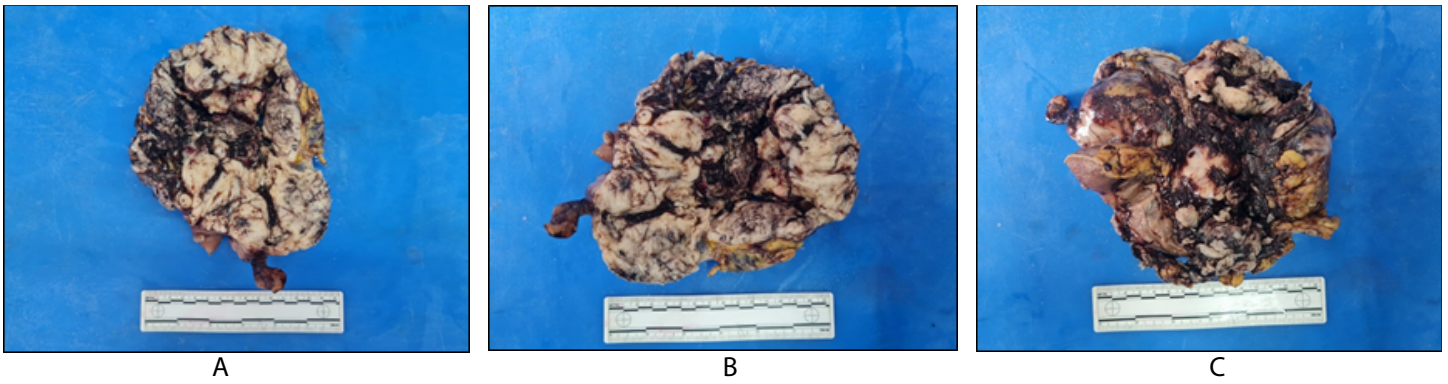
Source: Gastroenterology, Hospital Roosevelt



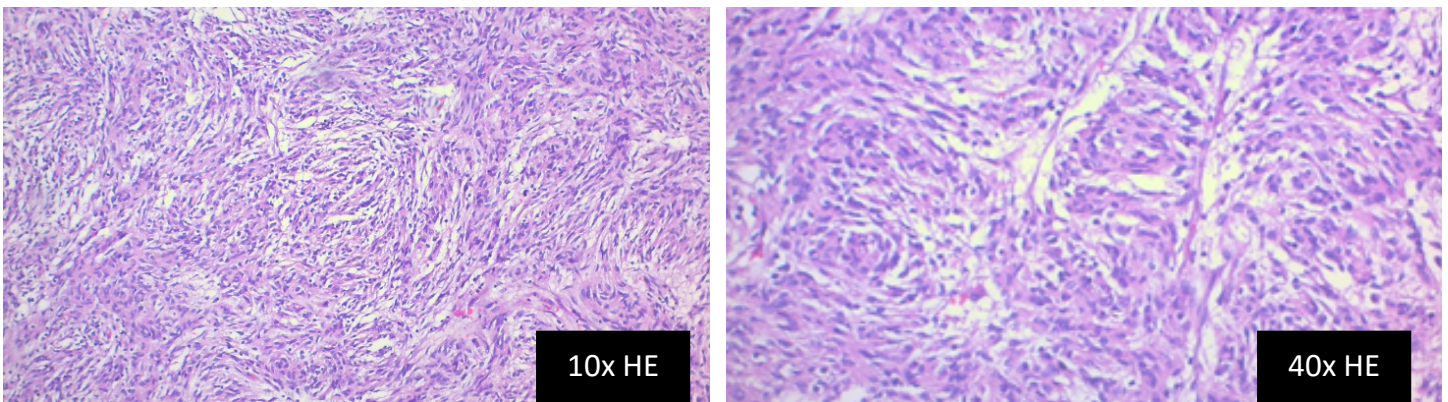
**Figure 3:** Exploratory Laparotomy.

Exophytic mass of 10 x 10 x 5 cm. approximately, localized 20 cm from the duodenojejunal angle (yellow arrow).

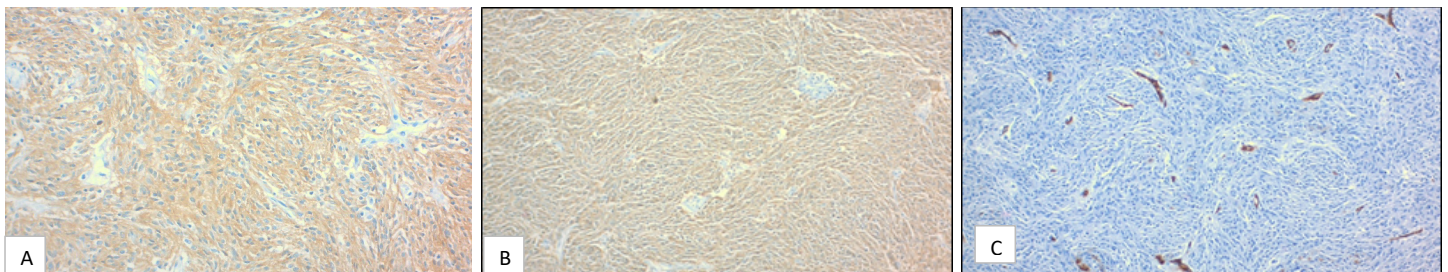
Source: Operating Room, Hospital Roosevelt.



**Figure 4:** Jejunal mass macroscopy.  
Source: Pathology, Hospital Roosevelt.



**Figure 5:** Jejunal mass microscopy.  
Mesenchymal neoplasm composed of spindle cell proliferations with a swirling pattern and interlaced bundles.  
Source: Pathology, Hospital Roosevelt.



**Figure 6:** Immunohistochemistry.  
A: DOG1 Intense cytoplasmic positive in neoplastic cells; B: CD117 Intense cytoplasmic positive in neoplastic cells; C: CD34 Negative for neoplastic cells.  
Source: Pathology, Hospital Roosevelt.

## Discussion

GISTs were first described by Mazur and Clark in 1983 [2] are rare cancers, with an estimated unadjusted incidence of 1.5 / 100,000 / year [3]. It is believed that GISTs arise from interstitial cells of Cajal or their stem cell precursors, the average age is 60 to 65 years, above 40 years its frequency is higher [4]. Comparing with our patient, this makes him out of the average, since he presented over 70 years.

The most common sites of GIST origins are stomach (60% -70%), small intestine (25% -35%), the least frequent place is in the jejunum region corresponding to 0.04%, esophagus (2 % -3%) and

rarely in the colon, rectum, or appendix (5%) [5]. This places our case within the less common locations, within the 0.04% situation in the jejunum.

The most common symptoms of GIST include upper gastrointestinal bleeding and anemia, while larger tumors can present with abdominal pain / discomfort and a palpable mass. Small bowel GISTs can remain silent for a prolonged period before presenting with an acute event such as hemorrhage or rupture [3], the case presented consulted with intermittent melena and anemia, for which the usual endoscopy protocol was followed and then colonoscopy. Finally ending with the diagnosis by enteroscopy.

The hemorrhagic pictures are produced by the growth and friction of the contralateral mucosa.

GISTs present as incidental lesions and range from those measuring just a few millimeters in diameter to large tumors greater than 35 cm, although their average size at diagnosis is 5 cm [6]. The radiological diagnosis of GIST is similar to that of other digestive tract tumors. In several studies, GISTs appear as submucosal lesions and in ultrasound studies as hypoechoic masses that, when large, can displace neighboring structures and show cystic, necrotic, or hemorrhagic areas. On computed tomography, tumors appear as well-circumscribed exoluminal masses that, after contrast, show heterogeneous enhancement, especially large tumors, which may have necrotic-hemorrhagic areas or degenerative components [7]. Our tomographic study showed intraluminal exophytic lesion, covering almost the entire intestinal lumen. No obvious distant disease. Ninety percent of primary GISTs may have mutations in the KIT (80%) or PDGFRA (10%) genes, and 10% have no mutation in both gene [8]. The mutations found in GISTs mainly affect the exons that encode the functional domains of the KIT and PDGFRA receptors. Among the main types of mutation, we find the following: deletions, point mutations, duplications, insertions and complex mutations [7]. (a scheme proposed to estimate the risk of metastasis in these lesions, based on tumor size and mitotic count is shown in Table 2 [9] and Table 3 document the modified NIH classification suggested by Joensuu combining the advantages of the NIH criteria (tumor size and mitosis) and AFIP (tumor location) together with the additional rupture factor [10]. In both classifications, our case corresponds to high risk, for size > 10 cm, rupture and 14/50 HPF mitosis.

**Table 2:** Proposed stratification to define risk of aggressive behavior.

Risk Classification	Size (cm)	Mitotic Count (x /50 HPF)
Very low risk	< 2	<5/50 HPF
Low Risk	2 a 5	<5/50 HPF
Intermediate Risk	< 5	6-10/ 50 HPF
	5 a 10	<5/50 HPF
High Risk	>5	>5/50 HPF
	>10	Any mitotic range
	Any size	>10/50 HPF

The classification cell where the presented patient belongs is highlighted. Source: [9]

**Table 3:** NIH modified by Joensuu- Risk Stratification.

Risk Category	Tumor Size (cm)	Mitotic Count (x /50 HPF)	Primary tumor site
Very low risk	< 2	< 5	Any
Low Risk	2.1 – 5.0	<5	Any
Intermediate Risk	< 5	6-10	Any
	5 a 10	<5	Any
High Risk	Any	Any	Tumor rupture
	>10	Any	Any
	Any	>10	Any
	5.0	>5	Any
	< 5.0	>5	Non gastric
	5.1-10	<5	Non gastric

The classification box where the presented patient belongs is highlighted. Source: [10]

Multidisciplinary treatment planning is necessary (involving pathologists, radiologists, surgeons, and medical oncologists, as well as gastroenterologists, nuclear medicine specialists, etc., as appropriate), such as that available in reference centers for sarcomas and GIST, and / or within referral networks that share multidisciplinary expertise and treat large numbers of patients annually [11].

Surgery remains a mainstay for a possible permanent cure even in the era of tyrosine kinase inhibitors. The indications for multidisciplinary neoadjuvant and adjuvant therapy can be individualized. Before neoadjuvant therapy, pathological diagnosis with biopsy samples is mandatory [8]. Due to its non-gastric location, surgical resection is recommended, regardless of the size or morphology of the tumor [12]. Because it is a high-risk phenotype, it is planned to start systemic therapy with imatinib. The optimal duration of treatment in these cases is unknown, given the uncertainty as to whether they should be considered to have essentially metastatic disease, but it should be at least 3 years, as in the case of high-risk resected GISTs [3]. Regorafenib, a multikinase inhibitor of KIT, PDGFR, and VEGFR is recommended after failure of high-dose imatinib and sunitinib, with a regimen of 160 mg daily for the first 3 weeks of each 4-week cycle [12].

Relapses occur most frequently in the liver and / or peritoneum (other sites of metastasis, including bone lesions and other sites, may be less rare throughout the course of metastatic disease treated with various lines of therapy). The mitotic rate probably affects the rate at which relapses occur. High-risk patients are routinely followed up with an abdominal CT scan or MRI every 3-6 months for 3 years during adjuvant therapy (with clinical follow-up). stricter due to the need to manage side effects of adjuvant therapy), unless contraindicated, then upon cessation of adjuvant therapy every 3 months for 2 years, then every 6 months up to 5 years from discontinuation of adjuvant therapy and annually for a further 5 years [11].

## Conclusion

We present the case of an elderly patient with gastrointestinal bleeding. It began with the baseline endoscopy and colonoscopy protocol; without showing source of bleeding. Image study shows a possible origin of the small intestine, for which we proceed to double-balloon assisted enteroscopy, with which we found a neoformative lesion in the jejunum, with pathological findings of high-risk GIST. Despite being a patient with preserved functional status, complete surgical resection and CT scans without evidence of distant tumor activity; with high probabilities of relapse and / or distant disease, therefore a candidate for systemic therapy.

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