



Case report

Jejunal gist simulating a uterine myoma: A case report

Maria Gonzalez-Vivo^{a,b,*}, Ander Zugazaga^{b,c}, Josep Maria Dedeu Cusco^{a,b},
Dolores Naranjo-Hans^{b,d}, Anna Casajoana^{b,e}, Laura Carot^{a,b}

^a Department of Gastroenterology, Hospital del Mar, Barcelona, Spain

^b IMIM (Hospital del Mar Medical Research Institute), Barcelona, Spain

^c Department of Radiology, Hospital del Mar, Barcelona, Spain

^d Department of Pathology, Hospital del Mar, Barcelona, Spain

^e Section of Gastrointestinal Surgery, Hospital del Mar, Barcelona, Spain



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ABSTRACT

Introduction: A small bowel gastrointestinal stromal tumor (GIST) is a rare neoplasm of the gastrointestinal tract. The manifestation of bleeding is a diagnostic challenge and could present as a life-threatening situation that needs urgent intervention.

Presentation of case: 64-year-old woman consulted for episodes of melena and anemia. The upper and lower endoscopies were not diagnostic. Capsule endoscopy (CE) revealed a probable jejunal hemangioma, however double-balloon enteroscopy and magnetic resonance imaging (MRI) did not show any intestinal nodule but MRI show a pelvic mass apparently related to the uterus confirmed by a gynecologist. Even so, the patient returned with melena, and a contrast-enhanced computed tomography (CT) scan again identified a pelvic mass, highlighting that its vascularization drained into the superior mesenteric territory and seemed to invade the jejunum, with active bleeding, suspicious for jejunal GIST. A laparotomy was performed to remove the jejunal mass. Histopathology and immunohistochemical studies confirmed the diagnosis.

Discussion: Bleeding is a common symptom in small bowel GISTs but its diagnoses could be difficult because its location. In most cases, gastroscopy and colonoscopy are not useful and CE or imaging studies are necessary to find the cause of bleeding. Moreover, it has recently proved that bleeding is a prognostic risk factor because it is related to tumor rupture and tumor invasion of blood vessels.

Conclusion: In this case, bleeding caused by small bowel GIST was misdiagnosed in endoscopic procedures and the clinical management was delayed. CT angiography was the most effective investigation to detect the source of bleeding.

1. Introduction

Gastrointestinal stromal tumors (GISTs) are the most common mesenchymal tumors of the GI tract [1]. Although previous studies have shown that up to 30 % of patients are asymptomatic, GISTs may be identified clinically because of symptoms including gastrointestinal (GI) bleeding, and abdominal pain due to tumor-induced mass effect [2]. The diagnosis of bleeding small intestine GISTs is challenging due to difficult approach by conventional endoscopy. Computed tomography (CT) angiography could help in demonstrating the tumor as well as the presence of active hemorrhage. We present a case in this scenario in a

tertiary hospital. This case has been reported in line with SCARE criteria [3].

2. Presentation of case

A 64-year-old woman, with high blood pressure and diabetes, presented to our emergency department by ambulance in October of 2020 after syncopal episode. The patient denied vomiting, hematochezia/melena or weight loss. On examination, she was afebrile with a blood pressure of 134/76 mmHg and heart rate of 103 bpm. Her abdominal examination was unremarkable and digital rectal examination revealed

Abbreviations: Capsule endoscopy, CE; Computed tomography, CT; Endoscopic ultrasound, EUS; Gastrointestinal, GI; Gastrointestinal stromal tumor, GIST; Magnetic resonance imaging, MRI.

* Corresponding author at: Department of Gastroenterology, Hospital del Mar, Barcelona, Spain.

E-mail address: mgonzalezvivo@psmar.cat (M. Gonzalez-Vivo).

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the presence of melena. Initial investigations revealed a drop of hemoglobin from previous 13.9 to 5.5 g/dl. She was promptly resuscitated with fluid therapy and blood transfusion, and an early upper endoscopy was performed. This procedure did not reveal any lesion that could cause bleeding. Patient was admitted in the gastroenterology ward. Colonoscopy was also normal. During hospitalization, stools were normal and hemoglobin remains stable after blood and iron transfusion and patient was discharged to continue study on outpatient clinic.

Subsequently, capsule endoscopy (CE) in January 2021 revealed probable jejunal hemangioma (Fig. 1a). Then, double-balloon enteroscopy did not reach the jejunal lesion and magnetic resonance image (MRI) did not show any intestinal nodule but pelvic mass was found apparently related to the uterus (Fig. 2). Patient was sent to Gynecology, that confirmed the presence of 8-cm subserous myoma by transvaginal ultrasound.

After that, patient came to emergency department in December 2021 with melena and anemia. At that time, an additional upper endoscopy was performed and showed two Forrest III small antral ulcers without any recent hemostatic sign. During the hospitalization, patient remained hemodynamically stable, but hemoglobin was dropping and therefore patient needed blood transfusion almost every day. CE was repeated and revealed the same lesion in jejunum with adherent clot and the presence of blood near the lesion (Fig. 1b). Thus, CT angiography was performed.

Pelvic mass was again found but notably its vascularization drained to the superior mesenteric territory and it seemed to invade jejunum, with active bleeding through jejunum (Fig. 3). In light of this situation, jejunal GIST with active bleeding was suspect. Few hours later, midline laparotomy was performed. Intraoperative findings revealed an exophytic lesion measuring 10 cm approximately 2 m proximal to the ileocecal valve (Fig. 4). Small bowel resection and side-to-side anastomosis were performed. She recovered uneventfully and was discharged.

The histopathological report (Fig. 5) confirmed the diagnosis of GIST arising from the jejunum (immunohistochemistry staining positive for cKIT). The long axis measured 80 mm, with a mitosis index of 1 per 50 high power field (HPF), with no evidence of tumor rupture. The final TNM stage was IB (T3 N0 M0), and classified as intermediary risk. Mutational study was performed and *KIT* exon 11 mutation was detected. No mutations were identified in *PDGFRA* gen. Close surveillance is undergoing.

3. Discussion

GISTs are malignant mesenchymal tumors that originate from the interstitial cells of Cajal and contain spindle cells. They are rare tumors with significant variations in reported incidence (around 8 cases per million per year) due to different diagnostic criteria over the decades

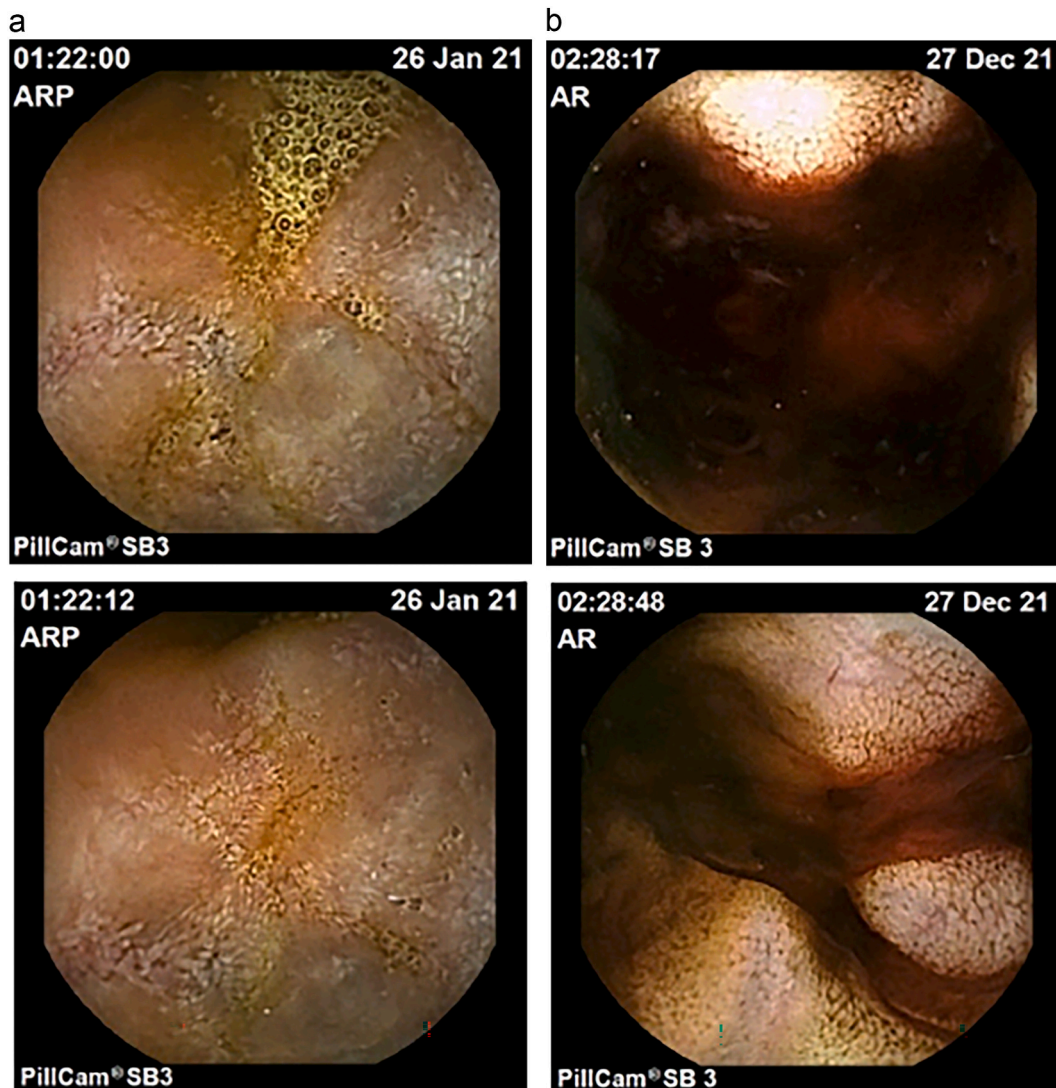


Fig. 1. Capsule endoscopy studies during follow-up. (A) In January 2021 CE showed dubious hemangioma in jejunum. (B) In December 2021 CE revealed same lesion with adherent clot and blood nearby.

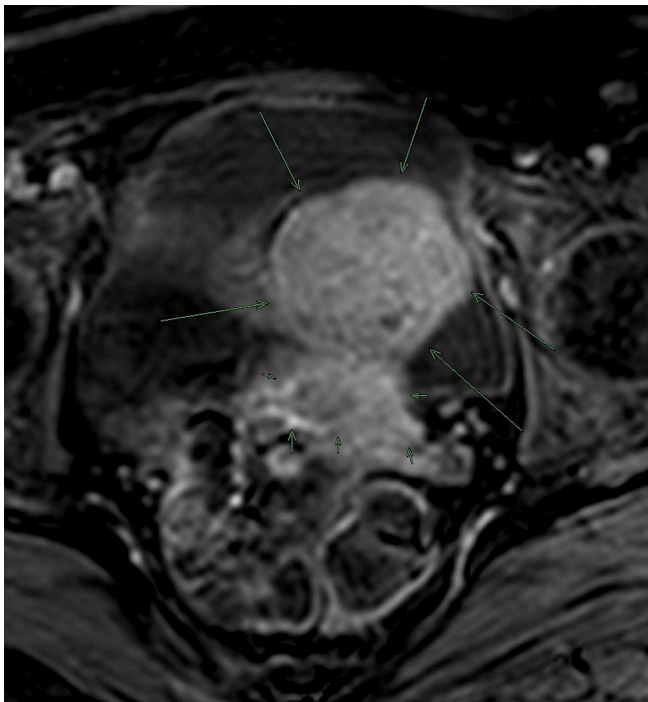


Fig. 2. MRI found a pelvic mass (long arrows) probably depending on the uterus (short arrows).

besides the fact that the smallest GISTs normally are not diagnosed histologically [2,4].

GISTs appear in the small intestine up to 36 %, being the second most frequent location after the stomach [5]. The estimated mean age at diagnosis is in the sixth decade [6] with a wide range of symptoms depending on site, size and growth pattern. It has been reported that patients with GI bleeding constitute approximately 23–33% [1,9] especially when the GIST is located in the small intestine due to narrower intestinal wall [7]. Besides, when the tumor volume is large, it is more likely to develop ulcers or break through the intestinal cavity. In other studies, the microvessel density and vascular endothelial growth factor were higher in tumors with tumor diameter ≥ 5 cm [8].

Most gastric GISTs are detected as submucosal endoscopic lesions during ordinary upper endoscopy as a nonspecific smooth nodule covered with normal mucosa. Given that optical endoscopic examination provides insufficient information, endoscopic ultrasound (EUS) is a key test for differential diagnosis. In large nodules (> 2 cm), EUS-guided fine needle aspiration is recommended to obtain a conclusive diagnose by immunohistochemical analysis [9].

In cases of bleeding GIST, upper and lower GI endoscopy remained the first line investigation. However, small intestinal location is a diagnostic challenge as this region is inaccessible by conventional endoscopy. Facing that situation, CE and enteroscopy play a predominant role in the diagnosis and treatment of these lesions. However, in some cases the lesions are not visible or are not accessible and other imaging modalities such as CT- or MR- enterography are essential for diagnosis. In addition, these studies provide additional information about the cause of the bleeding, which helps to guide the appropriate intervention.

GIST on CT shows the characteristic of a well-defined soft tissue of relatively low density, which is homogenous. However, it is difficult to differentiate GIST of the small bowel and rectosigmoid or gynecological lesions based on imaging studies.

In fact, the role of image test in small bowel bleeding is increasingly established. In patients with obscure hemorrhage, comparison of CE with angiography demonstrated that immediate CE has superior yield, although long-term outcomes, including rebleeding and hospitalization

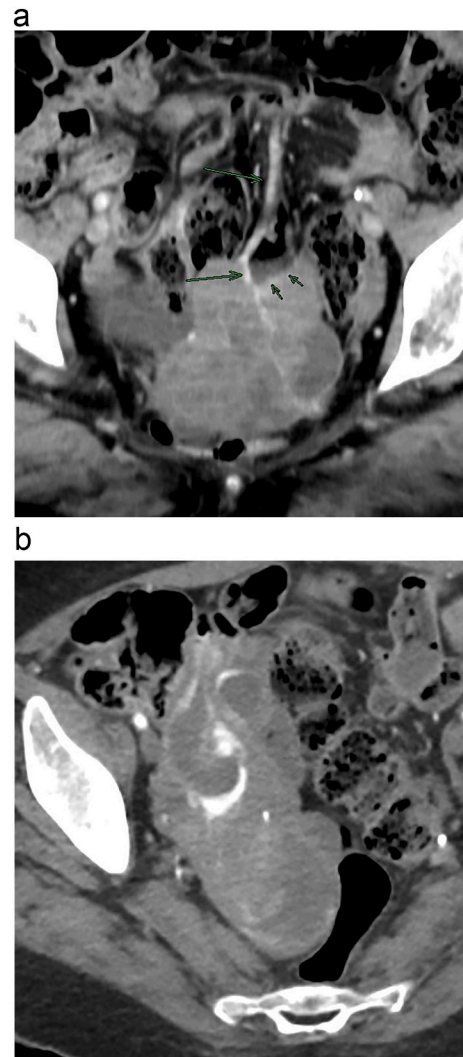


Fig. 3. CT angiography. (A) Pelvic mass with mesenteric venous drainage (long arrows); it seemed to invade jejunum (short arrows). (B) Active bleeding through jejunum.

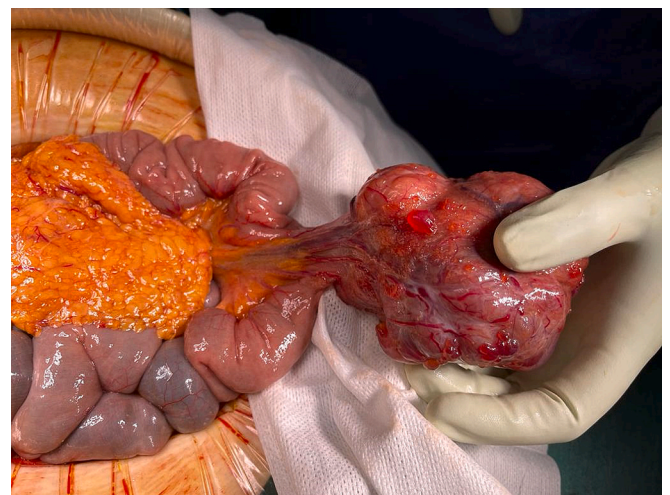


Fig. 4. Surgical piece of mass excised from the jejunum with its vascular pedicle.

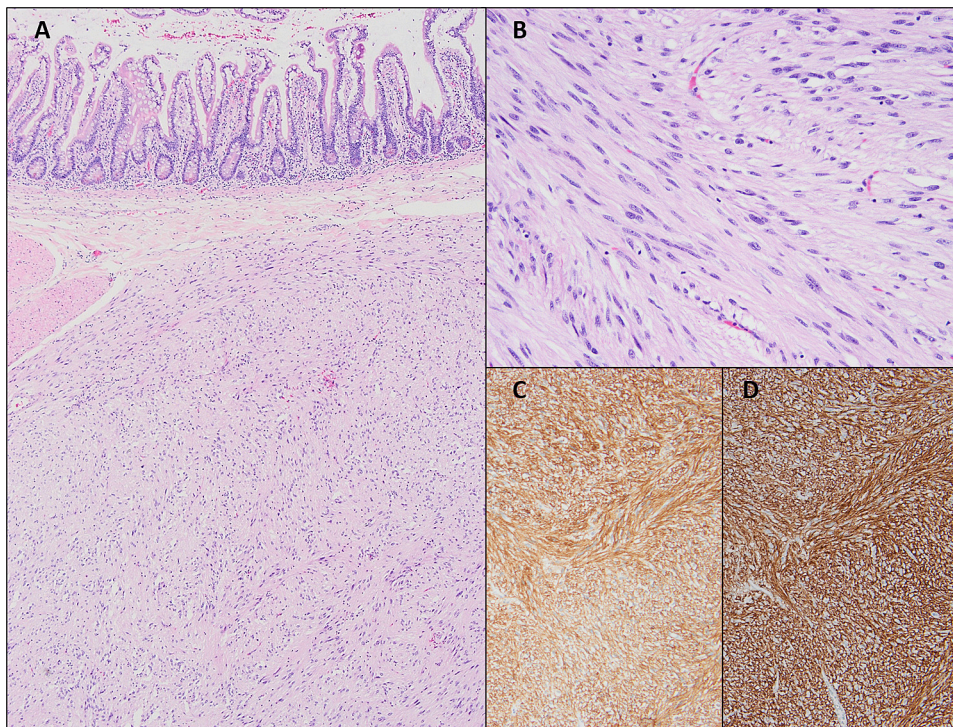


Fig. 5. Histopathology results. (A) Submucosal spindle cell proliferation with diffuse sheets or vague storiform arrangements of bland tumor cells. (B) At higher magnification, there is no nuclear pleomorphism, mitosis or necrosis. (C) KIT (CD117) immunohistochemistry shows intense and diffuse positivity (cytoplasmic and membrane-associated). (D) Immunohistochemistry for DOG1 (ANO1) is also a sensitive and specific marker and may be helpful in KIT-negative GISTs.

rates, and deaths, did not differ between two groups [10]. Visceral arteriography can detect bleeding in the upper GI tract at rates as low as 0.5 ml/min and it also has the ability to define in greater detail the vascular anatomy which can help to find the origin of the lesions [11] such as the case that has been exposed in this review. Recently, a small series of cases has been published showing the usefulness of CT for the management of acute massive over bleeding due to jejunal GIST [12].

Pathologically, CD117 expression is the GIST main feature (only 5 % of GISTs are CD117-negative). The mitotic count and the mutational analysis have predictive value for sensitivity to molecular-targeted therapy (PDGFRA D842V mutations are associated to lack of sensitivity to imatinib) [13].

Standard treatment of localized GISTs is complete surgical excision of the lesion, with no dissection of clinically negative lymph nodes. Pre-operative imatinib should be considered for those tumors where R0 surgery with no expected major sequelae is not feasible [9].

After surgery, risk assessment should be taken into account. A widely used risk classification was proposed by the Armed Forces Institute of Pathology, which incorporates the primary mitotic count, tumor size and tumor site and the presence or absence of tumor capsule rupture [14,15].

However, recent research demonstrates GI bleeding is of great significance to the prognosis of GIST [16] due to increases the risk of recurrence and affects the overall survival rate in a retrospective analysis [17]. Authors suggest that the causes of GI bleeding in patients with GIST are tumor rupture and tumor invasion of mucosal or submucosal blood vessels, thus favoring tumor dissemination, tumor recurrence and worse overall survival. Additionally, in a recent meta-analysis, authors found that mitotic index >5 per 50 HPF can significantly increase the risk of GI bleeding in GIST [18]. Moreover, patients with GI bleeding usually need emergency care, including transfusion support requirements, urgent endoscopic techniques and even emergency surgery, that also may contribute to worse prognosis.

4. Conclusion

Jejunal GIST is a very rare cause of small intestinal bleeding. Moreover, it could be part of a difficult differential diagnosis in women with abdominopelvic mass. In patients with GI bleeding with negative upper and lower endoscopies, a small intestinal source should be suspected, and CT may be useful in the diagnosis, especially in emergency situations. In addition, it also can show the precise location of bleeding, and guide subsequent management. Early diagnosis of bleeding GIST is of great importance because it has a worse prognosis and lower overall survival.

Consent statement

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Provenance and peer review

Not commissioned, externally peer reviewed.

Ethical approval

This case report was approved by the Hospital del Mar Clinical Research and Ethics Committee in 2023 (reference number: 2023/10881).

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Author contribution

Maria González-Vivó: Conceptualization, Writing- Original draft preparation. **Ander Zugazaga:** Visualization, Writing- Reviewing and Editing. **Josep Maria Dedeu Cusco:** Visualization, Writing- Reviewing and Editing. **Dolores Naranjo-Hans:** Visualization, Writing- Reviewing and Editing. **Anna Casajoana:** Supervision, Writing- Reviewing and Editing. **Laura Carot:** Conceptualization, Writing- Original draft preparation.

Guarantor

Maria Gonzalez-Vivo
Laura Carot

Research registration number

This case report was not registered in any place because it is not “First in Man”.

Conflict of interest statement

None.

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