Hitting the Target: An Uncommon Case of Obscure-Overt Gastrointestinal Bleeding Caused by a Jejunal Gastrointestinal Stromal Tumor

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Abstract

Significance

GIST are mesenchymal tumors of the GItract, representing less than 1% of GI neoplasms. GIST arising from the jejunum is extremely rare and accounts for 0.1%-0.3% of all GI tumors. Symptoms are non-specific and relate to the tumor location and size. Large tumors have wide range of manifestations including nausea, vomiting, abdominal pain and bleeding. GI bleeding from GISTs are mostly obscure in origin and has always been a diagnostic and therapeutic challenge. We report an uncommon case of obscure-overt GI bleeding from a jejunal GIST. We aim to highlight the advantage of push enteroscopy and CT angiography (CTA) in the approach to patients presenting with obscure-overt GI bleeding.

Clinical Presentation

A 72-year-old female presented with sudden episode of massive hematochezia associated with lightheadedness and anemia.

Management

Esophagogastroduodenoscopy and colonoscopy was unremarkable. Push antegrade enteroscopy using a pediatric colonoscope showed an ulcer with a spurting blood vessel in the proximal jejunum. Endoscopic clips were deployed resulting to complete hemostasis. CTA showed a heterogeneously enhancing jejunal mass with prominent vessels. Embolization using polyvinyl alcohol embolic particles was performed. Post-embolization contrast showed complete revascularization of the tumor. She underwent exploratory laparotomy with segmental jejunal resection. Final histopathology diagnosis is CD117 positive, DOG1 positive, spindle cell-type GIST.

Recommendation

Jejunal GIST presenting as obscure-overt gastrointestinal bleeding is extremely rare. Normal findings from an upper and lower endoscopy in a patient presenting with GI bleeding should raise a suspicion of small intestinal bleed. When a vascular lesion is suspected, abdominal CTA should be performed. This case highlights the importance of a stepwise and multidisciplinary approach in such cases.

INTRODUCTION

Gastrointestinal stromal tumors (GIST) are mesenchymal tumors of the gastrointestinal (GI) tract, representing less than 1% of GI neoplasms. The stomach is the most commonly involved site, followed by the small intestines, colon, rectum and esophagus. GIST arising from the jejunum is extremely rare and accounts for 0.1% to 0.3% of all GI tumors.[1,2] Symptoms are non-specific and relate to the location and size of the tumor.[3] Small GISTs that are less than 2 cm are usually asymptomatic. However, tumors larger than 4 cm have a wide range of manifestations including nausea, vomiting from intestinal obstruction, abdominal pain and bleeding from pressure necrosis and mucosal ulceration.[2,4] GI bleeding from GISTs are mostly obscure in origin, which has always been a diagnostic and therapeutic challenge to gastroenterologists.(5) We report an uncommon case of obscure-overt GI bleeding from a jejunal GIST, leading to anemia requiring blood transfusion. The objective of this case report is to highlight the advantage of push enteroscopy and CT angiography (CTA) in the approach to patients presenting with obscure-overt GI bleeding.

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CASE REPORT

A 72-year-old female with hypertension presented with sudden episode of hematochezia associated with lightheadedness. There was no melena, hematemesis, abdominal pain or anal pain. Further history revealed previous colonoscopy findings of colonic diverticulosis. She denied any family history of GI malignancy. On assessment, she had stable vital signs with unremarkable physical examination. Focused examination revealed a flabby, soft and non-tender abdomen with no palpable mass. Digital rectal exam showed maroon colored stools. Complete blood count (CBC) was within normal limits with an initial hemoglobin of 12.8 g/dL. After an episode of massive hematochezia, repeat hemoglobin was 10.7 g/dL, she was then advise to undergo endoscopy.

Colonoscopy revealed descending to sigmoid colon diverticulosis and grade 1 internal hemorrhoids; however, no sign of bleeding was identified. Esophagogastroduodenoscopy up to the distal portion of the duodenum was also unremarkable. Two episodes of hematochezia were reported few hours after endoscopy. Vital signs remained stable with no other associated symptom. Repeat CBC showed a 9.3 g/dL hemoglobin; hence, a unit of packed red blood cells was transfused. Repeat colonoscopy with terminal ileoscopy revealed black stools with maroon blood clots from the visualized terminal ileum to proximal transverse colon. Several diverticular openings were filled with black stools and clots but with no evidence of active bleeding. Two small, superficial clean-based ulcers without bleeding were seen in which endoscopic clips were applied. Push antegrade enteroscopy was performed using a pediatric colonoscope. An ulcer with an adherent clot was seen at the proximal jejunum (Fig. 1a). The clot was dislodged after continuous flushing which revealed a spurting and pulsating blood vessel (Fig. 1b). Seven endoscopic clips were deployed resulting to complete hemostasis (Fig. 1c). She was referred to interventional radiology service for abdominal CTA and possible embolization.

CTA with 3D reconstruction showed a heterogeneously enhancing mass measuring 4 x 3.9 x 4.4 cm with prominent vessels adjacent to the jejunum (Fig. 1d). Multiple linear densities medial to the mass representing endoscopic clips were noted. Superior mesenteric arterio-portography showed intense neovascularity and tumor staining at this region supplied by the first jejunal branch of the superior mesenteric artery (Fig. 2a). Embolization using polyvinyl alcohol embolic particles (355-500 microns) was performed by superselective catheterization of the described jejunal artery. Post-embolization contrast showed complete devascularization of the tumor (Fig. 2b). She underwent exploratory laparotomy which revealed an 8 x 5 cm well-circumscribed, violaceous mass in the jejunum (Fig. 2c). Segmental jejunal resection and stapled anastomosis were performed. Histopathology (Fig. 2d-f) showed CD117 positive and DOG1 positive GIST of spindle cell type involving the jejunum, with a mitotic rate of 0-1/5 mm^2. She was discharged 3 days after surgery without recurrence of bleeding.

DISCUSSION

GISTs, despite considered the most common mesenchymal tumor of the GI tract, rarely occurs in the jejunum. It originates from the interstitial cells of Cajal which are the pacemaker cells of the gut, coordinating GI motility and peristalsis.[6] Compared with large GISTs, small tumors (<2 cm) are commonly asymptomatic and incidentally found on endoscopy or imaging. When symptomatic, patients would present with abdominal pain/discomfort, vomiting, bloatedness or hemorrhage. GI bleeding, which is present in 50% of cases are obscure in origin due to its location, making it difficult to be identified by routine endoscopy. The main mechanisms leading to GI bleeding are ulceration of the overlying mucosa and pressure necrosis.[7]

The patient presented with episodes of hematochezia without hemodynamic compromise. Her initial endoscopy did not reveal any actively bleeding lesion hence, was inconclusive. It is estimated that 5% of patients with overt GI bleeding will have an initial non diagnostic endoscopy findings. Persistent visible evidence of bleeding anywhere from the GI tract after a negative diagnostic evaluation is defined as obscure-overt GI bleeding. As seen with this case, majority of patients with obscure-overt GI bleeding present with either melena or hematochezia.[8] These patients are likely to possess a more serious lesion with a higher morbidity and mortality.[9]

Small bowel pathologies account for the majority of the etiologies of obscure-overt GI bleeding. The most common of which are vascular lesions. Other potential etiologies include small bowel tumors, inflammatory lesions and less commonly, extraluminal sources.[10,11] Therefore, a high index of suspicion for these etiologies is warranted to prevent delays in the definitive diagnosis and management. In our case, push antegrade enteroscopy was able to reach the jejunum where the spurting blood vessel was noted.

According to Gralnek and colleagues, push enteroscopy may be performed directly in situations where in small bowel lesions are suspected. It would be beneficial since it permits direct visualization of the small intestines extensively into the jejunum and/or ileum. Simultaneously, tissue samples for histopathologic diagnosis can be obtained and more importantly, therapeutic endoscopic measures can be carried out. A dedicated videoenteroscope, pediatric or standard adult colonoscope can be used. Reported diagnostic yields in the evaluation of GI bleeding have ranged from 13% to 38% using a pediatric colonoscope, which increases with greater depth of scope insertion.[11] With the endoscopic findings suggestive of a vascular lesion, performing a CT angiography is the next best diagnostic imaging of choice to identify site of bleeding.[5] Angiography can identify non-bleeding lesions with the demonstration of typical vascular features seen in vascular ectasias (i.e. slow-filling vein, vascular tuft in the arterial phase, and early-filling vein) and tumors. Simultaneously, interventional radiologists are able to administer embolization therapy if an amenable lesion is detected.[11]

In these rare occasions that GISTs present as obscure-overt GI bleeding, an early multidisciplinary management is of utmost importance. Collaborating with interventional radiology and surgery services will be the cornerstone in the approach to these patients, and should be considered in all cases of massive GI bleeding. A case report by Saad et al. described a 46-year- old male with jejunal GIST who presented with melena, dizziness, palpitations and anemia requiring multiple blood transfusions.[12] Esophagogastro-duodenoscopy was inconclusive, whereas colonoscopy showed fresh blood throughout the colon. CTA revealed a 5.2 x 4.2 cm heterogeneously mass in the proximal jejunum with persistent enhancement through the delayed phase. The patient underwent laparoscopic segmental small bowel resection, after failure of radiologic embolization.[7]

This case report underscores the importance of a systematic approach in managing patients with GI bleeding. Inconclusive findings from an upper and lower endoscopy in cases of overt GI bleeding should increase the suspicion of a small intestinal pathology. In the absence of a dedicated enteroscope, a pediatric or an adult colonoscope may be used for antegrade enteroscopy. In addition, this case highlights the utility and effectiveness of therapeutic endoscopy in the initial management of patients with massive small intestinal GI bleeding prior to definitive surgical resection.

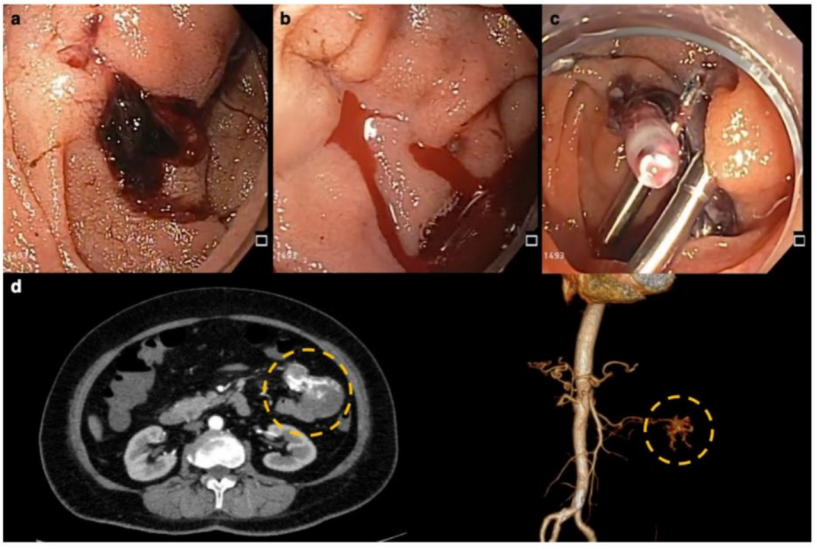

Figure 1. Push enteroscopy revealed an (a) ulcer with adherent clot at the proximal jejunum, (b) oozing visible vessel post-flushing and (c) partial hemostasis after application of endoscopic clips. (d) CT Angiogram with 3D reconstruction showed a heterogeneously enhancing mass with prominent vessels and multiple linear densities representing endoscopic clips.

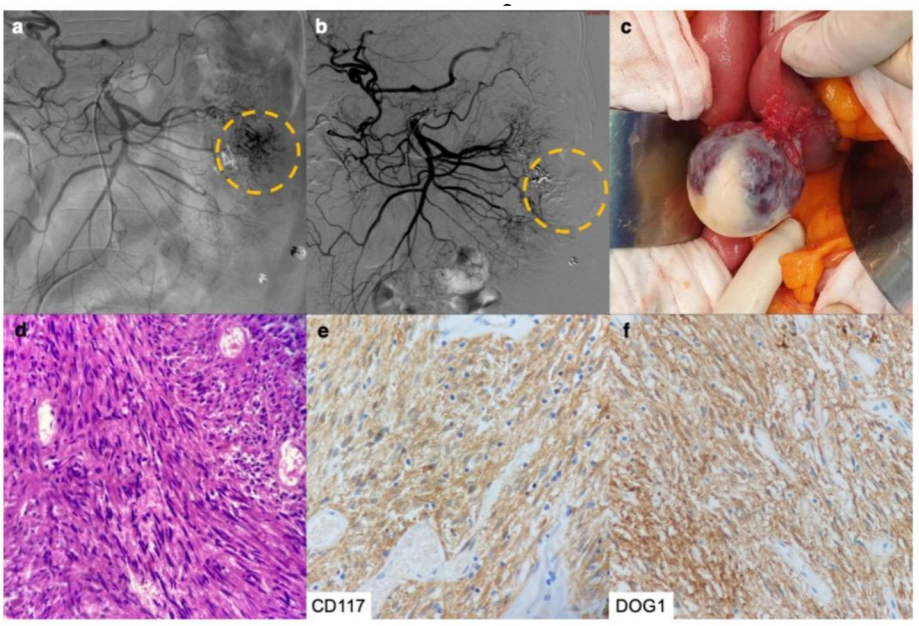

Figure 2 (a) Superior mesenteric arterio-portography showed intense neovascularity and tumor staining at the jejunal region supplied by the first jejunal branch of the superior mesenteric artery. (b) Complete devascularization of the tumor post-embolization. (c) Fungating mass in the antimesenteric side of the jejunum during exploratory laparotomy. (d) Histopathology showed cytologically bland spindle cells arranged in packets or whorls with normochromatic nuclei. Immunohistochemistry revealed (e) CD117 positive and (f) DOG-1 positive staining.

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