

中文題目：以不規則血管變化且嚴重出血為表現的腸胃道基質瘤

英文題目：A gastrointestinal stromal tumor presenting with severe GI bleeding and disordered vasculature in small intestine

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Introduction:

Gastrointestinal stromal tumors (GISTs) are rare tumors, which represent approximately 1 to 2 percent of primary gastrointestinal cancers¹, but the most common mesenchymal neoplasms of the gastrointestinal tract. The overall incidence rate was between 10 and 15 cases per million population per year². GIST commonly affects older adults, with a median age of diagnosis between 65 and 69 years³⁻⁵. Upper GI bleeding is the most common clinical presentation of GISTs, caused by pressure necrosis and ulceration of the overlying mucosa. Other presentation of GISTs varies depending on the primary tumor location, including dysphagia, nausea, vomiting, abdominal pain, constipation, or obstructive jaundice.

Case presentation:

A 48-year-old man presented with epigastric pain, feeling of fullness, nausea and melena for 3 days. There were no other associated symptoms. Physical examination revealed soft, mildly distended abdomen. Initial screening tests showed a hemoglobin level of 9.2 g/dl.

He experienced an episode of massive melena on the first day of hospital admission, and his hemoglobin level had dropped to 6.1 g/d. Urgent upper endoscopy was performed, which reported some blood clots over second portion of duodenum, but early terminated due to patient's intolerance. Computed tomography (CT) angiography reported one exophytic tumor (5.3 cm) arising from proximal jejunum, with tumor related active bleeding.

Three attempts of transcatheter arterial embolization (TAE) were then performed for active bleeding. The first and the second attempts of TAE both reported no definite extravasation identified, therefore, a microcatheter was placed at a duodenal branch, partially supplying the tumor, and artificial spasm was given. During the third attempt of TAE, TAE with N-Butyl-2-cyanoacrylate (NBCA) was successfully injecting into the site of active extravasation, the focal segmental jejunal branches.

The abdomen was inspected with laparoscopy and a 6.5 × 4 × 2 cm tumor was seen arising from proximal jejunum, anti-mesentery site, without lymphadenopathy or adjacent organ invasion. The tumor was excised with segmental resection of about 12 cm of jejunum, followed by side-to-side anastomosis.

Histopathology of the tumor revealed 6.1 × 3.9 × 3.1 cm in size, with spindle cell feature. Mitotic rate was 1/per 5 square mm. The surgical margin was negative for tumor. Immunohistochemistry reported the cells were positive for CD117, DOG1. SMA and S100 were negative. These findings were

consistent with the diagnosis of gastrointestinal stromal tumor. The risk category was moderate.

The patient was discharged on day 10 post surgery in a stable condition. Adjuvant imatinib therapy was initiated 2 months after surgery. Follow-up CT of the abdomen reported no evidence of recurrence.

Discussion:

Primary GISTs are most frequently located in the stomach (55.6%), followed by small bowel (31.8%), colorectal (6.0%) and other location (6.2%). GISTs can present with a broad range of gastrointestinal symptoms. It varies according to their location or size. On average, 18.7% of cases are asymptomatic and found incidentally on CT scans, endoscopy, or surgical procedures. Common symptoms of GIST were abdominal pain, satiety, nausea, palpable abdominal mass, obstruction and gastrointestinal bleeding⁶.

Gastrointestinal bleeding is the most common symptom of GIST (54%)⁷. In addition, in cases of GIST-related emergencies, gastrointestinal bleeding was the most common (48.9%) cause of admission⁸. Few cases have reported the use of catheter embolization, surgical resection such as wedge resection or subtotal gastrectomy, to control the life-threatening bleeding from GISTs^{9,10}. A multi-center study identified independent risk factors for bleeding in GIST including tumor location in the jejunum, prolonged prothrombin time, presence of surface dimpling in CT images, and positivity of S100¹¹. A cohort study reported that GIST patients with age <60, male gender, tumors located in the small intestine, and tumors 5-10 cm in size had a higher risk of GI bleeding¹². Gastric GISTs tend to bleed when they are larger than 5 cm and are Ki-67 positive. Clinically, GI bleeding associated with jejunal GIST usually arises from mucosal break or ulceration after tumor development beneath the mucosa as tumors grow⁷.

Endoscopy is often the first-line diagnostic and therapeutic management in patient with acute GI bleeding, particularly if there is a suspected upper GI source. However, for patients with failed upper or lower endoscopy and patients with a source of bleeding visualized on an imaging modality, catheter angiography is considered the first-line imaging and treatment option. Embolization can be performed when active extravasation of contrast material into the bowel lumen is visualized¹³.

Our patient presented with massive bleeding from jejunal GIST, accompanied with severe anemia requiring blood transfusion. Upper endoscopy was unsuccessful and the source of bleeding was not identified. Angiography and embolization were thus considered to be performed early to detect the bleeding site and treat bleeding promptly.

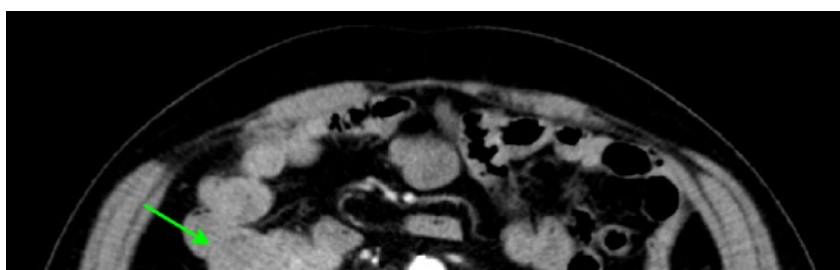
The gold standard of treatment for localized primary GISTs is radical surgical resection through laparoscopy. A retrospective review concluded that complete surgical resection with negative margins is the curative treatment for GISTs. However, if there is metastatic disease then imatinib should be recommended as the targeted therapy in conjunction with surgery¹⁴.

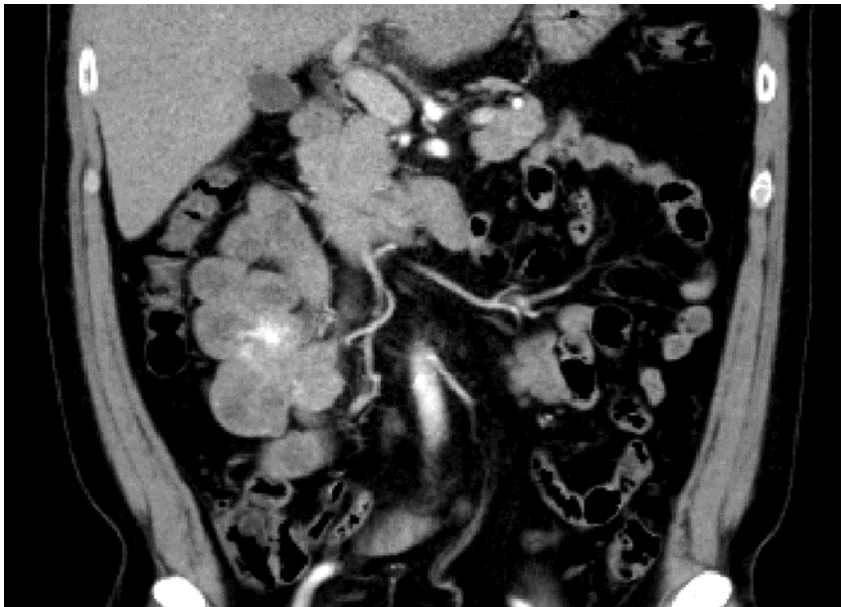
Diagnosis of bleeding jejunal GISTs is often challenging to diagnose and treatment, particularly in patient with massive bleeding and hemodynamic instability, due to inaccessibility of endoscopy.

Angiography is considered to be a single-step diagnostic and therapeutic tool for managing bleeding. Surgical resection is the main treatment for both management of bleeding and to provide oncologically clear resection margins.

Conclusion:

GIST is an uncommon tumor in small intestine. It may present with many different symptoms, but bleeding is one of the most life-threatening presentation. In order to disclose accurate bleeding source, combined endoscopy and CT angiography are mandatory. Angiography embolization for the treatment of severe and emergent bleeding of small bowel GIST is crucial in the first step. The mainstay treatment is still surgical resection.





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