Ectopic variceal bleeding from chronic superior mesenteric vein thrombosis after hemorrhagic pancreatitis

Kathryn Flynn 1 | Kevin Chung 2 | Thomas Brooke 1 | Jonathan Keung 3

1 Internal Medicine, Walter Reed National Military Medical Center, Bethesda, Maryland, USA
2 Pulmonary and Critical Care Medicine, Walter Reed National Military Medical Center, Bethesda, Maryland, USA
3 Interventional Radiology, Walter Reed National Military Medical Center, Bethesda, Maryland, USA

Correspondence
Kathryn Flynn, Internal Medicine, Walter Reed National Military Medical Center, Bethesda, MD, USA.
Email: k.flynn8820@gmail.com

Abstract
Varices are a common cause of gastrointestinal (GI) bleed. When ectopic, there is often a delay in diagnosis as it is difficult to localize these varices. Ectopic small bowel varices usually arise from portal hypertension, which commonly develops in the setting of cirrhosis. This case presents a much rarer cause of bleeding ectopic varices with portal hypertension secondary to chronic superior mesenteric vein (SMV) thrombosis that developed after an episode of hemorrhagic pancreatitis. An 81-year-old man with a past medical history of a recent GI bleeds secondary to an arteriovenous malformation presented to the hospital with continued melena after a recent admission at another hospital for the same symptom. Upper endoscopy and colonoscopy showed no evidence of active bleeding. Subsequently computed tomography angiography (CTA) showed bleeding from collaterals in the third part of the duodenum, consistent with ectopic varices. The CTA also showed SMV thrombosis. The patient underwent an ultrasound-guided transhepatic venogram with coiling and sclerosant embolization of SMV varices and distal SMV balloon angioplasty. Capsule endoscopy after showed no evidence of further bleeding. The patient was discharged 72 h after the intervention with stabilized hemoglobin and resolved melena. Ectopic varices should be on the differential diagnosis for patients presenting with a GI bleed that remains nonlocalized after endoscopy and colonoscopy. EGD or colonoscopy is the first-line intervention for the treatment of bleeding ectopic varices. If unreachable by these means, percutaneous coil embolization is an alternative way to stabilize the patient. As no general management guidelines exist, treatment of bleeding ectopic varices should continue to be case-dependent and involve a multidisciplinary team.

KEYWORDS
angioplasty, bleed, bleeding, colonoscopy, critical care, duodenal, duodenum, ectopic, ectopic varices, embolization, endoscopy, gastroenterology, gastrointestinal bleed, GI bleed, hemorrhagic pancreatitis, interventional radiology, pancreatitis, percutaneous coil embolization, portal hypertension, superior mesenteric vein thrombosis, varix
1 | INTRODUCTION

Varices are a common cause of gastrointestinal (GI) bleeds and are either gastroesophageal or ectopic. Ectopic varices, by definition, are large portosystemic venous collaterals located anywhere outside the gastroesophageal region. Ectopic varices account for only 5% of all variceal bleeds. Due to their infrequency and location outside the gastroesophageal region, ectopic variceal bleeds pose a diagnostic dilemma as these sources often take longer to localize in a patient presenting with a GI bleed. Ectopic varices have a mortality rate of up to 40% during a bleeding episode and a four-fold increased risk of bleeding when compared to gastroesophageal varices.1,2

Ectopic varices most commonly develop in the setting of portal hypertension from pre-existing liver disease, such as cirrhosis. It is important to investigate other causes when a patient does not exhibit cirrhotic findings in the setting of portal hypertension. One cause of portal hypertension is superior mesenteric vein (SMV) thrombosis, and its most common presenting symptom is bleeding from esophageal or gastric varices.3 The clinical presentation of bleeding ectopic varices from SMV thrombosis after hemorrhagic pancreatitis is rare to find in the literature. There is a 1–2% reported incidence of portosplenomesenteric thrombosis after severe acute pancreatitis.4 To our knowledge, only two cases of bleeding small bowel varices after pancreatitis exist.5 Here, we present a case of bleeding duodenal varices caused by portal hypertension from SMV thrombosis secondary to hemorrhagic pancreatitis.

2 | CASE PRESENTATION

An 81-year-old man with a past medical history of a recent GI bleed presented to the hospital after 2 days of melena and left-sided and mid-epigastric abdominal pain associated with fatigue and dizziness with ambulation. The patient had been discharged a week prior from a different intensive care unit (ICU) after a prolonged admission for a GI bleed. Colonoscopy during that admission showed descending colon arteriovenous malformation that was treated with argon plasma coagulation (APC) and ulcerated mucosa in the descending colon that was treated with epinephrine and clips. Esophagogastroduodenoscopy (EGD) showed evidence of Barrett’s esophagus but no active bleeding. Both computed tomography angiography (CTA) and magnetic resonance imaging of the abdomen showed no evidence of active GI bleeding, but it was significant for chronic occlusion of the SMV and splenic vein with significant intramural/periduodenal venous collaterals. The patient was diagnosed with a lower GI bleed in the descending colon. The patient presented to this hospital shortly after discharge due to the continuation of melena and symptomatic anemia. Symptoms included feeling clammy, weak, and nauseated with dyspnea on exertion without chest pain. The patient reported continued black, tarry stools without pain during bowel movements.

The patient was normotensive and without tachycardia. On physical examination, the patient was well-developed and in no apparent distress, but there was some mild tenderness to palpation on the mid-epigastrum and left lower quadrant. Hemoglobin was 8.1 g/dl on admission, down from a baseline of 14 g/dl 2 months prior. Liver function tests and coagulation studies (PT, PTT, and INR) were within normal limits. The hemoglobin dropped to 6.9 g/dl overnight, so the patient was admitted to the medical ICU with symptomatic anemia in the setting of an ongoing GI bleed.

EGD and colonoscopy both showed no active source of bleeding or targetable lesion. The patient received six units of RBCs while in the ICU. The hemoglobin stabilized with no source for an active bleed, so the patient was transferred to the general floor.

In the setting of known chronic SMV thrombosis, hematology/oncology performed a comprehensive hypercoagulable workup. The workup was completely negative and included JAK2 V617F mutation, lupus anticoagulant, direct Russell viper venom time, cardiopin antibodies, beta-2 glycoprotein antibodies, Factor V Leiden, Factor II, and protein C levels.

The patient was transferred back to the ICU 2 days later for symptomatic anemia with another hemoglobin drop to 5.8 g/dl. The patient received three more units of RBCs and had a CTA (Figure 1) that showed bleeding varices around the third part of the duodenum, which prompted
the consultation of interventional radiology (IR) and general surgery. IR performed a successful ultrasound-guided transhepatic venogram with coiling and sclerosant embolization of varices around the third part of the duodenum as seen in Figures 2 and 3. Balloon angioplasty of distal SMV stenosis was also performed. After the procedure, Gastroenterology performed capsule endoscopy that showed no active bleeding. Subsequent CT imaging in Figure 4 showed the resolution of the variceal network. The patient received a total of 12 units of RBCs during this admission. Fibroscan confirmed that the patient did not have cirrhosis. The patient’s melena resolved, and his hemoglobin remained stable for 72 h, at which point he was deemed safe to be discharged home.

At follow-up 1 year later, the patient has not had any recurrent bleeding episodes. He had a repeat EGD and colonoscopy without evidence of any bleeding.

3 | DISCUSSION

Patients with ectopic varices often have underlying portal hypertension in the setting of cirrhosis. SMV thrombosis is a known, but uncommon cause of portal hypertension and represented 3% of reported cases in a study evaluating the etiology of portal hypertension in adults.6 The increasing hydrostatic pressures from the backflow behind the thrombus can lead to bleeding varices, which most often form in the esophagus or stomach. Varices developing from SMV thrombosis can go undetected for long periods of time due to the vague, intermittent abdominal pain that may be the only symptom until a patient presents with bleeding collaterals. If EGD and colonoscopy cannot localize a bleeding source, the diagnosis becomes more difficult as the bleeding may be coming from ectopic varices.3

Ectopic varices, by nature of their location, are harder for clinicians to locate when a patient presents with a GI bleed, as they can be unreachable by EGD and colonoscopy. The patient had undergone two prior endoscopies, but the endoscopies evaluated up to the second part of the duodenum and were just shy of the area of the bleeding varix. An interdisciplinary team was critical to identifying and treating the lesion, as the lesion was not identified until IR was consulted and found it on a personal read of the imaging. The prolonged hospital course and delayed diagnosis reinforce the relevant and challenging nature of ectopic varices.

Though chronic SMV thrombosis was identified in earlier imaging studies, this diagnosis does not immediately point clinicians towards suspecting bleeding duodenal varices because SMV thrombosis-induced varices are often found in the esophagus or stomach. Chronic SMV thrombosis is often a sequela of prothrombotic states like coagulopathies or malignancy, local foci of irritation or inflammation like pancreatitis or splenectomy, or a combination of these. Any patient with SMV thrombosis should undergo a full hypercoagulable workup, and their history should be thoroughly examined for any precipitating events.3 The most plausible cause of this patient’s chronic SMV thrombosis was from an episode of hemorrhagic pancreatitis years ago, which led to a prothrombotic state of thrombosis formation and allowed for the gradual development of venous collaterals. A retrospective study from the University of Pittsburgh showed that 22 of 162 patients (14%) with acute pancreatitis developed portosplenomesenteric venous thrombosis. Necrotizing pancreatitis was associated with all but one patient with thrombus formation, and 27% of these patients developed varices. All varices were localized to the gastroesophageal region, and no patients had any gastrointestinal bleeding complications.7 While portosplenomesenteric venous thrombosis is a known complication of necrotizing pancreatitis, the majority of patients do not have long-term sequelae of variceal bleeding or ectopic variceal formation.4

Due to the infrequency in which bleeding ectopic varices are identified, data regarding management are scarce and often come from case reports and retrospective series.3 In general, treatment requires a multidisciplinary approach and is tailored to each patient. After resuscitation and stabilization, the preferred initial intervention is endoscopic with endoscopic glue injection being the first choice for duodenal varices.5 For bleeding ectopic varices unreachable by EGD or colonoscopy, percutaneous coil embolization is regarded as a safe short-term approach to stabilize the patient.5,10 The ultimate
treatment goal is to relieve the elevated pressures, which is why patients treated solely with embolization often have episodes of re-bleeding and require subsequent intervention.\textsuperscript{1,11} Re-opening the SMV in this patient was allowed for restoration of anatomical flow to relieve the pressure source that initially led to the development of these varices. In unstable patients with no access to IR and with varices not amenable to endoscopic intervention, there must be a discussion among GI and general surgery to decide on salvage therapy options or transfer to another facility. Duodenal varices pose unique anatomical challenges that are case-specific, which is why ongoing communication with multiple specialties is paramount.\textsuperscript{11} More research is needed to better define prophylactic and therapeutic data when it comes to treating bleeding ectopic varices. One area of continued debate revolves around β-blocker secondary prophylaxis against duodenal variceal re-bleeding, and more studies will need to be conducted to understand whether and who this treatment would most benefit.\textsuperscript{1} Chronic SMV thrombosis is often treated with long-term anticoagulation, but large studies have not evaluated the risks and benefits of anticoagulation in the setting of bleeding varices.\textsuperscript{3} As no general management guidelines exist, treatment of bleeding ectopic varices should continue to be case-dependent and involve a multidisciplinary team.

**CONCLUSIONS**

In any patient with portal hypertension and GI bleeding, the presence of ectopic varices must be considered whether both EGD and colonoscopy fail to localize a bleeding source. This patient’s remote history of hemorrhagic pancreatitis allowed for a hypercoagulable condition in which chronic SMV thrombosis developed, thus propagating undiagnosed portal hypertension until presenting with ectopic variceal bleeding. EGD or colonoscopy are the first-line interventions for the treatment of bleeding ectopic varices. If unreachable by these means, percutaneous coil embolization is an alternative way to stabilize the patient. As no general management guidelines exist, treatment of bleeding ectopic varices should continue to be case-dependent and involve a multidisciplinary team.

**CONFLICT OF INTEREST**

No competing interests to declare.

**AUTHOR CONTRIBUTIONS**

Kathryn Flynn led the writing, editing, revising and followed up the patient; Thomas Brooke and Kevin Chung contributed to the clinical presentation of the patient and helped with revisions; Jonathan Keung contributed to the imaging findings and interpretations.

**ETHICAL APPROVAL**

The authors declare that they have no conflicts of interest, and no funding was received for this case report. No patient-identifying data have been released in the article. Patient has consented for this to be published. No further acknowledgements or ethical statements were to be made.
CONSENT
Written informed consent was obtained from the patient to publish this report in accordance with the journal’s patient consent policy.

ORCID
Kathryn Flynn  🅰️ https://orcid.org/0000-0002-1026-8778

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