CASE REPORT

Ectopic varices: a potential cause of gastrointestinal bleeding in patients with portal hypertension

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SUMMARY

A newly diagnosed 53-year-old woman with cirrhosis has repeated gastrointestinal bleeding with resulting symptomatic anaemia. She underwent routine diagnostic endoscopic evaluation without localisation of the aetiology of her bleed. Ultimately, she was found to have ectopic varices in the small bowel as a result of underlying high portal pressures. She underwent transjugular intrahepatic portosystemic shunt for portal system decompression with resolution in her bleeding.

1 month prior. The remainder of her physical examination was notable for abdominal distension and ascites which had started a few months prior to her presentation in clinic. Given concern for haemodynamic instability and acute blood loss, the patient was sent to the emergency department for triage and possible admission. Initial labs were notable for Hgb level of 5.8 mg/dL and the rectal examination was consistent with active bleeding. She was subsequently admitted to the inpatient hepatology service for further management.

BACKGROUND

Gastrointestinal (GI) bleeding is a common and often life-threatening cause of symptomatic anaemia in the general population. Patients with cirrhosis and portal hypertension represent a more complicated clinical challenge as their concomitant coagulopathy may predispose them to large volume variceal bleeds as well as infection. Recent estimates of mortality associated with variceal bleeding are as high as 30% in patients with Child Class C disease at 6 weeks.¹ This is a case of a 53-year-old woman with alcohol cirrhosis who presented with recurrent obscure GI bleeding. The lesion was not discovered on extensive previous evaluation including endoscopy and provocative angiogram. Further evaluation with capsule endoscopy and CT angiography raised concern for an ectopic variceal bleed in the small intestine in the setting of decompensated cirrhosis and portal hypertension. She ultimately underwent transjugular intrahepatic portosystemic shunt (TIPS) placement and mesenteric variceal embolisation with resolution of her bleeding and improvement of her overall volume status.

CASE PRESENTATION

A 53-year-old woman with newly diagnosed alcohol cirrhosis presented to gastroenterology clinic to establish care after multiple recent inpatient admissions for GI bleeding and symptomatic anaemia at another facility. Approximately 1 month prior, the patient was admitted to an outside hospital with haematochezia and found to have an initial haemoglobin (Hgb) level of 3.1 mg/dL. She was subsequently discharged home in stable condition after the bleeding stopped and she was appropriately volume resuscitated.

On initial evaluation in clinic, the patient was tachycardic with a heart rate of about 130 beats/min. The patient was also having ongoing haematochezia and melaena that began shortly after discharge

INVESTIGATIONS

Prior to admission, the patient had undergone extensive evaluation to determine the aetiology of her GI bleeding: two upper endoscopies, two colonoscopies with blood in the caecum each without active extravasation and two tagged red blood cell (RBC) scans which showed evidence of bleeding in the right lower quadrant. She then underwent two separate mesenteric angiographies, including one provocative study with tPA, that were both unrevealing.

DIFFERENTIAL DIAGNOSIS

The causes of GI bleeding are varied and are of special concern in patients who also have cirrhosis as these patients also have a concomitant coagulopathy. The initial concern in patients with cirrhosis is often oesophageal variceal bleeding, but the patient had no evidence of oesophageal varices on endoscopy. Further considerations included diverticular bleed, peptic ulcer disease, portal hypertensive gastropathy or angiodysplasia.

TREATMENT

The patient was volume resuscitated on admission and received 2 units of packed red blood cells as well as intravenous fluids. Her Hgb improved slightly from 5.8 to 6.6 mg/dL, which was not an appropriate response and was concerning for ongoing blood loss. She then received another single unit of packed red blood cells and capsule endoscopy was pursued for further evaluation of a presumed small bowel bleed (figures 1 and 2). The capsule study was positive for active bleeding noted at approximately 3 hours into the study. Passage into the duodenum was confirmed after 6 min.

Due to the length of time between passage into the duodenum and the first sighting of active bleeding in the small bowel, there was concern that the bleed could not be reached with single balloon

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Figure 1 Oedematous and congested small bowel mucosa.

enteroscopy. The patient was sent for CT angiography. CT angiography did not show any intraluminal extravasation but was concerning for prominent mesenteric varices in the right lower quadrant (consistent with previous tagged RBC scans) and associated mesocaval shunting (figure 3). Given ongoing concern for portal hypertension causing an ectopic variceal bleed, the patient underwent TIPS for decompression followed by embolisation of a right lower quadrant mesocaval varix. The initial portal venous pressure pre-TIPS was noted to be 31 mm Hg. After TIPS, portal venous pressure was successfully reduced to 20 mm Hg with a portal pressure gradient (PPG) of 15 mm Hg (RA pressure 5 mm Hg). During TIPS placement, the mesocaval varices were embolised given high flow and large size noted on SMV angiogram during the procedure (figure 4). Additionally, there was

Figure 2 Oedematous and congested colonic mucosa.

Figure 3 Prominent mesenteric varices—right lower quadrant prominent mesocaval shunt (blue arrow) from the distal branch of the superior mesenteric vein into the right gonadal vein (red arrow).

concern that the mildly elevated PPG (15 mm Hg) after TIPS (goal PPG 12 mm Hg) increased her risk for re-bleeding.

OUTCOME AND FOLLOW-UP

The patient did well following the TIPS and embolisation procedures without any further GI bleeding. She was able to return to many of her daily activities and was no longer short of breath on exertion. She did have some issues with optimisation of her volume status following the TIPS and was seen in clinic for follow-up and management of an outpatient diuretic regimen.

DISCUSSION

Bleeding from the small bowel is a relatively uncommon cause of GI bleeding but is responsible for the majority of recurrent bleeding cases in patients without an obvious aetiology discovered on initial evaluation with upper and lower endoscopies.² General evaluation of presumed small bowel bleeding includes capsule endoscopy, CT or MRI enterography, radionuclide scanning and

Figure 4 Coil embolisation of the mesocaval shunt.

angiography. Studies using capsule endoscopy to evaluate for small bowel pathology in patients with compensated cirrhosis estimated the prevalence of small bowel varices to be 8.1%³ and 7%.⁴ However, the likelihood of small bowel varices increased with the presence of larger oesophageal varices, ascites, portal gastropathy or colonopathy, and increasing Child-Pugh classification.^{3,4} Interestingly, a few cases of small bowel variceal bleeding without EVs^{5,6} and with portal-systemic pressure gradients as low as 15 mm Hg⁷ have been reported.

Currently, there are no routine screening recommendations for ectopic varices (varices outside of the gastro-oesophageal region) in cirrhosis as it affects a rather small subset of patients. Ectopic varices are responsible for 2.5%–5% of all GI bleeding with an associated mortality of up to 40%.⁷ There is no suggested management of ectopic variceal GI bleeding, but efforts have been made to better classify the haemodynamics of ectopic varices when determining appropriate treatment options. Some studies have shown that re-bleed rates following a TIPS are 20%–40%. Other endovascular management options, including balloon-retrograde transvenous obliteration, have similar re-bleeding risk when compared with TIPS.^{8,9} For small bowel varices accessible by endoscopy (push enteroscopy, double-balloon enteroscopy and colonoscopy), other therapeutic interventions to achieve haemostasis have shown success including variceal banding,¹⁰ sclerotherapy and thrombin injection.¹¹ In these cases, EUS may be helpful in the characterisation of lesions and assessment of sclerosant effect if sclerotherapy is used.¹²

Learning points

- ▶ Although relatively rare, bleeding ectopic varices are a common cause of gastrointestinal bleeding in patients with portal hypertension and no sites of active bleeding identified on upper and lower endoscopy.
- ▶ Small bowel varices are often seen in patients with other sequelae of portal hypertension (oesophageal varices, ascites and so on) but can be seen in patients without these findings.
- ▶ Capsule endoscopy, radionuclide scanning and angiography can be used to evaluate for small bowel varices.
- ▶ The approach to management of bleeding small bowel varices should be multidisciplinary and include input from providers trained in advanced endoscopy and interventional radiology. Transjugular intrahepatic portosystemic shunt and balloon-retrograde transvenous obliteration are treatment options for bleeding small bowel varices not amenable to endoscopic intervention.

This case highlights the prevalence of small bowel varices in patients with portal hypertension and the importance of considering small bowel varices as a possible cause of GI bleeding in patients with an otherwise negative evaluation. Given the multiple treatment options that can be used in ectopic bleeding, we recommend a multidisciplinary approach to treatment that should include advanced endoscopists as well as interventional radiologists.

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