

Pan-Colonic Varices and Idiopathic Portal Hypertension

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Abstract

Varices of the lower GI tract, although rare, are a known cause of hematochezia. They are usually found in a segmental distribution and are often associated with cirrhosis, portal hypertension, or portal vein obstruction. We present the case of a 43-year-old male with no personal or family history of liver disease, who experienced recurrent rectal bleeding over a 27-year period. Colonoscopy revealed varices from the rectum to the cecum confirmed with endoscopic ultrasound, while esogastroduodenoscopy, small bowel series, and CT were all normal. Portal hypertension was present without an identifiable cause.

Key words

Varices - adult - angiography - colon - sigmoid - gastrointestinal hemorrhage - hematochezia - venous pressure - varices

Case report

A 43 year-old man was admitted for further evaluation of recurrent lower gastrointestinal bleeding. He reported that his first episode of hematochezia occurred at the age of 16. The bleeding episodes were always associated with defecation and were painless. He reported a predictable pattern of blood-coated stools for 2-3 days that progressed to passage of larger amounts of blood for 2-3 days, followed by resumption of bloodless bowel movements. Over the years, he had noticed that the bleeding episodes would occur 1-2 days after consuming beans and rice. He denied abdominal pain, fever, anorectal manipulation or passing mucus with bleeding episodes. Bleeding occurred about three times per year until 9 months prior to this admission

when the frequency of hematochezia increased to two episodes per day. On admission he was hemodynamically stable, and did not report fever, chills, abdominal pain, weight loss, or change in his bowel habits.

His past medical history was significant for hypothyroidism after ablation therapy for Graves' disease, and he was maintained on 75 mcg of Synthroid. He reported drinking 1-2 beers on the weekends and smoking 1 pack of cigarettes per day. He recalled developing an erythematous rash in the past with penicillin administration. He was born and educated in Honduras and came to the United States at the age of 20. His mother has Type II diabetes, otherwise there is no family history of significant illnesses. He appeared as a well-developed overweight male in no acute distress with stable vitals. His physical examination was significant for exophthalmos, truncal obesity, a non-tender abdomen with active bowel sounds, no dermatologic lesions, and gross blood on digital rectal exam. There were no stigmata of liver disease.

The hemoglobin was 9.3 g/dL and mean corpuscular volume was 68 fL (82-92 fL). Biochemical tests of liver function, electrolyte analysis, and coagulation profile were all normal. There was no serologic evidence of chronic viral hepatitis. Colonoscopy revealed several columns of dilated tortuous bluish vessels extending from the rectum to the cecum (Fig.1). The varices were largest proximal to the rectum (Fig.2), and smaller in the rectum. Several areas had evidence of superficial erythematous venules suggestive of red wale markings (Fig.3). No active bleeding site was found and no endoscopic therapy was done. Given these findings, an esogastroduodenoscopy was performed which showed no evidence of esophageal varices, gastric varices, duodenal varices, or portal hypertensive gastropathy. A small bowel series and contrast abdominal CT were normal; specifically, there was no splenomegaly, evidence of cirrhosis, thrombosis, and no collateral veins. A liver biopsy showed no evidence of cirrhosis or abnormal fibrosis (Fig.4). A portogram revealed normal flow, and superior mesenteric artery and inferior mesenteric artery angiography revealed normal arterial and venous phases. There was no evidence of portal venous obstruction or obstruction of the superior

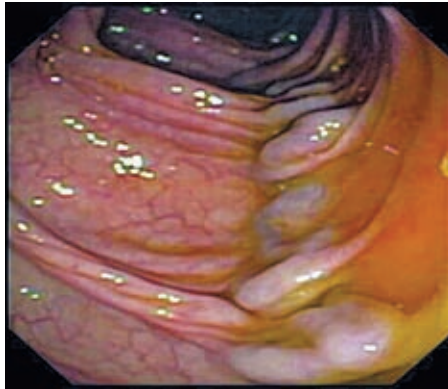


Fig.1 Continuous varix column in sigmoid colon.

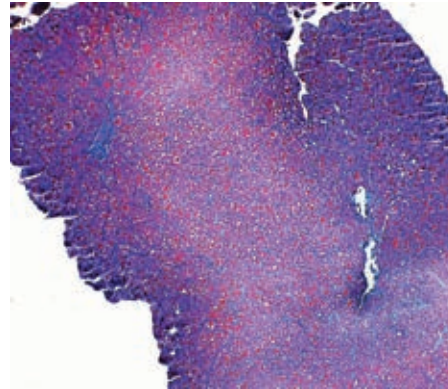


Fig.4 Liver biopsy - no evidence of fibrosis.



Fig.2 Varix protruding into lumen in descending colon.

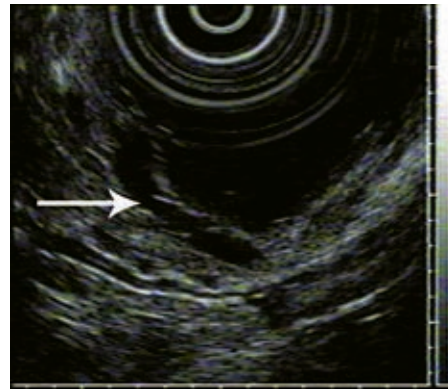


Fig.5 EUS demonstrating varix.



Fig.3 Red wale marks in ascending colon.

mesenteric vein or inferior mesenteric vein. The hepatic wedge pressure was measured at 24mm Hg (IVC=9mm Hg). EUS confirmed that the visible bluish structures were in fact submucosal vessels (Fig.5).

The patient required 4 units of packed red blood cells during hospitalization. Propranolol was started at 10 mg tid and titrated to 40mg tid over three weeks. A response in heart rate was noted from baseline 90's to 60's. Repeat sigmoidoscopy demonstrated a marked decrease in the size of the varices. However, the patient decreased the dose of propranolol due to perceived effects on his libido. He continues to have monthly episodes of hematochezia, and

a persistent microcytic anemia, despite iron therapy, but has not required transfusions. The patient has repeatedly declined colonic resection.

Discussion

Colonic varices are an uncommon cause of hematochezia. There are approximately 80 cases of lower gastrointestinal bleeding from colonic varices reported in the literature. However, the vast majority of these cases are related to portal hypertension, either from cirrhosis, or portal vein obstruction. Embryologically derived anastomoses between the portal and systemic systems exist in the esophagus, terminal ileum, colon, retroperitoneum, and anterior abdominal wall (1). In general the coronary-azygous anastomosis is the most developed, and favors the development of esophageal varices in the setting of portal hypertension. It has been hypothesized that colonic varices in patients with portal hypertension arise in patients in whom the colonic anastomosis is highly developed. Colonic varices not associated with portal hypertension may be due to mesenteric venous or splenic vein obstruction. This may be due to thrombosis, tumor invasion, extrinsic compression, acute or chronic pancreatitis, mesenteric adhesions, or congenital anatomic variations. Twenty-two of the reported cases did not have a specific etiology and were classified as idiopathic or primary.

The incidence of non-idiopathic colonic varices is equal in males and females and the mean age at diagnosis is 50 years (2). By comparison, idiopathic colonic varices occur more commonly in males than in females, and the mean age at diagnosis is 41.3. While the majority of non-idiopathic colonic varices have a segmental distribution, half of the reported idiopathic cases are pan-colonic (3). A familial tendency for the idiopathic cases is suggested by several case reports demonstrating patients having a family member with the same condition (4). Approximately one third of the reported cases of idiopathic colonic varices appear to have a familial component.

In certain cases of idiopathic colonic varices, distinct venous abnormalities may be identified. For example, an annular aneurysm of the ileoceocolic artery which "encircled and compressed the SMV" was identified in three related individuals described by Hardy in 1967 (5). Colonic varices have been described in Klippel-Trenaunay-Weber (K-T-W) syndrome, which is a rare disease characterized by the triad of cutaneous hemangiomas, varicose veins, and bony and soft tissue hemihypertrophy that can cause unilateral limb lengthening. There are two cases of colonic varices (which are also termed "cavernous colonic hemangiomas") in association with K-T-W syndrome (6). However, in most cases, the etiology will not be identified, even after laparotomy and colonic resection. In our patient, mesenteric angiography, portography, and venography did not demonstrate any identifiable venous abnormality. Our evaluation did not reveal a cause for the patient's non-cirrhotic portal hypertension, although it resulted in colonic varices.

In some instances, video endoscopy has allowed for rapid diagnosis. At endoscopy, colonic varices appear as tortuous, bluish structures that may protrude into the lumen (7). Careful examination may reveal superficial venules (red wale markings) on the varices (Fig 3). Although endoscopy can provide useful information, it is not the most reliable modality for diagnosing varices. Overinflation during colonoscopy may hide the varices by collapsing them, and active bleeding may also obscure visualization. Colonic varices may be misinterpreted on barium enemas as polyps, cancer, air bubbles, or fecal material (8). The most reliable modality for diagnosing colonic varices is selective mesenteric angiography, where the varices are often identified during the venous phase (9).

The management of bleeding colonic varices beyond the initial resuscitation period has not been standardized. Treatment is dictated by the underlying etiology as well as the distribution of the varices and has included somatostatin infusion (10), arterial vasopressin infusion (11), catheter embolization (12), cyanoacrylate infusion (13), argon plasma coagulation (14), and vascular stenting (15). Portal decompression is appropriate in patients with portal hypertension. In patients with varices limited to the rectum, variceal banding (16) or sclerotherapy (17-21) have been used. There are no reports of these endoscopic therapies being applied to more proximal locations in the colon.

Variceal banding or sclerotherapy may not be safe to perform in the colon proximal to the rectum due to the risk of perforation from transmural injury. Colonic resection has been reserved for younger patients with hemodynamically significant bleeding. Conservative management, which may consist of observation only or periodic transfusions, is preferable for the elderly and those with less significant bleeding.

Conclusion

The discovery of colonic varices should prompt a thorough evaluation to exclude liver or cardiac disease, as well as vascular abnormalities. Based on our experience and a review of the medical literature, we recommend conservative medical management initially with a Beta Blocker for those presenting with bleeding varices in the setting of elevated portal pressures. Careful patient follow-up should be instituted for those who are incidentally noted to have varices but have not bled. In general, patients with idiopathic colonic varices are likely to have a better prognosis than those whose varices arise secondary to portal hypertension (22).

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