Obscure gastrointestinal bleeding in a patient with congenital hepatic fibrosis and Crohn’s disease

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CASE REPORT

A 42-years-old man with congenital hepatic fibrosis, portal vein thrombosis, and autosomal dominant polycystic kidney disease, presented with melena in the last week. The patient has Crohn’s disease and had undergone right hemicolecction 7 years ago because of an enterovesical fistula. His medication included warfarin and azathioprine. Laboratory data revealed: Hemoglobin 10.9 g/dl, white blood cell count 1.49x10³/L, platelet count 70.x10⁹/L and international normalized ratio 2.4. Upper endoscopy revealed small esophageal varices without bleeding stigmata and no bleeding lesions were found at colonoscopy. For persistent melena and daily red blood cells transfusion requirements, an abdominal angio-CT was performed which suggested a small bowel (SB) ectopic varix (EcV) near the surgical abdominal scar (Fig. 1). Retrograde single-balloon enteroscopy showed blood in the colon and ileum; however, the EcV was impossible to reach because an ulcerated ileal stenosis 5 cm from the ileocolic anastomosis prevented the progression of the overtube. To confirm the radiological findings, a capsule endoscopy was performed. It revealed a blood in the distal SB and a varix in the ileum (Fig. 2). A transjugular intrahepatic portosystemic shunt (TIPS) was indicated without recurrence of gastrointestinal bleeding.

DISCUSSION

EcV are portosystemic collaterals in locations other than the gastroesophageal region, accounting for up to 1-5% of all variceal bleeding (1). Given their sporadic nature, literature is limited to case reports, small case series and reviews (2,3). EcV develop secondary to portal hypertension, abdominal surgical procedures and vascular thrombosis. Congenital hepatic fibrosis is a rare cause of portal hypertension. Jejuno-ileal varices are the result of collateral circulation through postoperative adhesions between the SB and the abdominal wall in portal hypertension. Early diagnosis and treatment is required because bleeding can be massive with high mortality rates (1). Treatment depends on the location of varices, patient’s condition and local available expertise (4). Endoscopic procedures are relatively faster and easier than surgery or radiologic intervention and include band ligation, sclerotherapy or cyanoacrylate injection (5). However, TIPS offers a highly effective modality for bleeding control when the varix is not reachable with endoscopy.

REFERENCES


