Absent inferior mesenteric vein as a cause of lower gastrointestinal bleeding: The first reported case in the literature

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A B S T R A C T
To date, absent inferior mesenteric vein (IMV) has not been reported in the literature as a cause of or being associated with lower gastrointestinal (GI) bleeding. We describe a case of 13 year-old girl who presented with hematochezia and was subsequently found to have widespread colonic varices involving the ascending, transverse and proximal descending colon. The upper GI tract, small bowel, and rectum were not involved. Delayed venous phase of mesenteric angiography revealed an absent inferior mesenteric vein. The patient underwent laparoscopic extended right hemicolectomy with ileosigmoid anastomosis. No bleeding from recurrent varices occurred during a 1-year period of follow up. We conclude that extended right hemocelecytomy is a potential curative surgical option in patients presenting with lower GI bleeding from colonic varices caused by absent IMV. Upper GI and small intestinal involvement should be excluded to prevent future bleeding from missed varices these sites.

1. Background

Lower gastrointestinal (GI) bleeding can be caused by many etiologies in pediatric patients, a fact that makes the diagnosis challenging in this age group. Colonic varices have been described as a causative etiology in many case reports [1–9], none of these reports described the congenital absence of inferior mesenteric vein (IMV) as an underlying cause of these varices. We describe the first case of lower GI bleeding associated with colonic varices secondary to a congenital absence of the inferior mesenteric vein.

2. Case report

A 13 year-old female with no significant past medical history presented to her local emergency department with a five-day history of intermittent rectal bleeding, urgency, and tenesmus which was thought to be gastroenteritis on initial evaluation. She was admitted to our facility after she passed a loose bloody bowel movement followed by a syncopal attack. Her hemoglobin level was 5.9 g/dl (10 g/dl prior to admission). Her initial physical exam was remarkable for minimal left lower quadrant pain. Meckel’s diverticulum scan was negative. Esophagogastroduodenoscopy excluded the Upper GI as a potential cause of her bleeding. Subsequent flexible sigmoidoscopy revealed thick, sticky, tenacious, melanotic stool throughout the rectum and sigmoid colon with some fresh blood under the melanotic stool. Computed tomography (CT) scan of the abdomen revealed moderate thickening and edema of the wall of the ascending colon with multiple small prominent vessels in the region of the hepatic flexure. Abdominal ultrasonography with Doppler study excluded portal venous flow obstruction and hypertension.

Repeat colonoscopy after bowel preparation identified multiple varices in the ascending colon, hepatic flexure, transverse colon, and splenic flexure [Figs. 1–4], with stigmata of recent hemorrhage [Figs. 1–4], with stigmata of recent hemorrhage. Capsule endoscopy excluded the presence upper GI tract and small bowel varices.

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Laparoscopic extended right hemicolectomy with ileosigmoid anastomosis was successfully performed to remove the involved bowel segments. The postoperative course was complicated by pre-renal azotemia which resolved with IV hydration and intra-abdominal abscess that was drained percutaneously. Of note, during the operation we actively looked for the presence of the IMV to exclude the possibility of a present but thrombosed vein. The pathologist also looked for an ectatic or thrombosed vein or remnant and found none.

Six months after the operation the patient had an episode of rectal bleeding that was found to be due to ileosigmoid anastomotic ulcer which responded to Sucralfate therapy. No colonic varices were identified at the time of colonoscopy.

During the 1 year period of follow up, the patient did not have recurrent bleeding attributed to recurrent varices.

3. Discussion

Absent inferior mesenteric vein has never been reported in the literature as a cause of or being associated with lower gastrointestinal bleeding or inferior mesenteric venous hypertension. In this case, the patient had uneventful 13 years of her life without any GI bleeding. The ascending colon, transverse colon and left colon was draining via a single marginal vein that drained into the superior mesenteric drain. We postulate that either venous hypertension in the IMV collaterals led to the development of widespread colonic varices or a congenitally absent IMV may be associated with abnormal varices throughout the ascending, transverse and left colon. The ascending and proximal transverse colon—theoretically—should not be involved in this process as the venous drainage of this segment is via the superior mesenteric vein (SMV) which was both present and patent in this patient. IMV collaterals were partially draining via the SMV which could explain—along with IMV hypertension—the involvement of the ascending and proximal transverse colon with the varices process. To our surprise, the distal most descending colon and the sigmoid colon were completely free of any varices. However, the venous drainage from the hindgut could drain either proximally (toward the hepatic and ascending colon) to drain via the SMV tributaries, or distally, may be via the middle and inferior rectal veins and subsequently to the systemic venous system through the internal iliac veins. To our knowledge, we are not aware of any study that could provide us with such information.
The arterial phase of the mesenteric angiogram was completely normal. The absent IMV was only identified in the late venous phase of the study. We advocate obtaining a venous phase in patients with unexplained colonic varices to look for an underlying venous vascular malformation [10].

The presence of a vascular malformation in the colon could be associated with other vascular malformations throughout the GI tract. Therefore, exclusion of the Upper GI and small bowel involvement is important before considering any surgical treatment to avoid missing a concomitant presence of varices which can be a source of future bleeding.

The absence of data in this topic and the tempting option of expectant management were sources of decision making difficulties in this patient. We opted to proceed with resection of the involved colonic segments as expectant management will leave the real risk of life-threatening bleeding. Our patient presented with a herald bleed and syncope with a potential source of bleeding. This can put the patient at a higher risk of recurrent bleeding which could be fatal. Since the sigmoid colon and rectum were free of the disease process, we elected not to remove them. Total proctocolectomy would be an aggressive procedure for obvious reasons. The operation performed cannot guarantee cure and recurrence of varices and bleeding in the remaining bowel. During the period of follow up, the patient sustained no bleeding attributed to recurrent varices. Surveillance proctosigmoidoscopies might be considered to exclude the presence of recurrent varices.

Laparoscopic extended right hemicolectomy was successfully performed. During the period of one year follow up the patient did not experience any episode of bleeding attributable to recurrent colonic varices. She had one episode of bleeding from an anastomotic ulcer which responded to sucralfate treatment and lower endoscopy confirmed no varices.

4. Conclusion

Vascular malformations including absent inferior mesenteric vein should be considered in children presenting with lower GI bleeding due to colonic varices. Selective mesenteric angiogram is the gold standard diagnostic study for vascular anomalies and delayed venous phase is important in identifying venous anomalies. Upper GI tract and small intestinal involvement should be excluded by performing EGD and capsule endoscopy-if age allows. We propose colectomy of the affected area as potential treatment when bleeding occurs with an absent inferior mesenteric vein and colonic varices.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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