



Bleeding Jejunal Varices in a Patient with Extensive Porto-Mesenteric Thrombosis: Clinical Conundrum

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Abstract

Ectopic varices are rare cause of obscure GI bleeding and hence should be always suspected in patients with history of portal hypertension who present with GI bleeding and have negative upper and lower GI endoscopies. We reviewed a case of a 58-year-old patient who presented with melena and symptomatic anemia and was a known case of factor V Leiden heterozygous mutation on long term anticoagulation. Unexpectedly his extensive workup showed; bleeding jejunal varices as the source of his anemia. Patient failed multiple trials of interventional radiology embolization and was managed laparoscopically with small bowel segmental resection and re-anastomosis. Establishing the diagnosis is challenging and management should be through a multidisciplinary approach. We conclude that surgical removal of the involved small bowel is a feasible and safe option in similar patients resistant to other modalities of treatment.

Keywords: Jejunal ectopic varices; Surgical resection; Upper GI bleeding; Anemia

Introduction

Small bowel ectopic varices can develop due to multiple causes such as portal hypertension, abdominal surgical procedures, congenital anomalies in venous outflow, and abdominal vascular thrombosis. Given all these etiologies; abdominal vascular thrombosis is among the rarest reasons to cause ectopic varices. Small bowel varices have been reported few times in literature to be seldomly caused by Portomesenteric Vein Thrombosis (PMVT) that can also lead to portal hypertension [1]. Ectopic varices are uncommon cause of lower Gastrointestinal (GI) bleeding, account for approximately 5% of all variceal bleeding [1]. Most of the cases are segmental and secondary to portal hypertension or portal venous obstruction. However, some isolated cases were reported in the absence of portal hypertension or venous obstruction [2]. Intestinal ectopic varices commonly present with melena, hematochezia, or intraperitoneal bleeding.

Case Presentation

A 58-year-old man presented to the emergency department with 3-day history of dizziness, melena, and passage of large amount of fresh blood per rectum. This was the third hospitalization due to melena. He was taking oral warfarin and had IVC filter in place due to recurrent Deep Venous Thrombosis (DVT) and development of Pulmonary Embolism (PE). His past medical history was significant for extensive portomesenteric thrombosis involving superior mesenteric, splenic, and main portal veins which was diagnosed 2 years ago. Extensive workup revealed heterozygous mutation of factor V Leiden, while protein C, protein S, and antithrombin III levels were all within normal range. Autoimmune workup, hepatitis workup and HIV serology were all negative and he has no similar family history. He is a known to have proximal jejunal varices diagnosed by capsule endoscopy after three episodes of lower GI bleeding in the past; first one was in 2016, was managed conservatively without need for intervention and second episode was in 2018 which was managed by percutaneous trans-hepatic embolization of the varices, and similarly in 2019 and he underwent a second trial of coiling of the jejunal varices. On clinical examination he was hemodynamically stable and physical examination revealed marked pallor and normal abdominal examination (Figure 1). Digital rectal examination showed no evidence of melena at the time of examination. Laboratory tests revealed a hemoglobin level of 5.5 g/dl, platelet count 200,000/ul, INR 1.5. Urgent Esophagogastroduodenoscopy (EGD) revealed normal esophagus and stomach. Subsequently, push Upper GI enteroscopy was attempted in the same setting revealing multiple varices in the proximal jejunum with red wale signs involving short segment of the jejunum. Computed Tomography (CT)

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Received Date: 06 May 2022

Accepted Date: 01 Jun 2022

Published Date: 06 Jun 2022

Citation:

AL Zoubi M, Toffaha A, Elmatbouly M, Suliman A, Al-Bahrani A. Bleeding Jejunal Varices in a Patient with Extensive Porto-Mesenteric Thrombosis: Clinical Conundrum. Clin Surg. 2022; 7: 3518.

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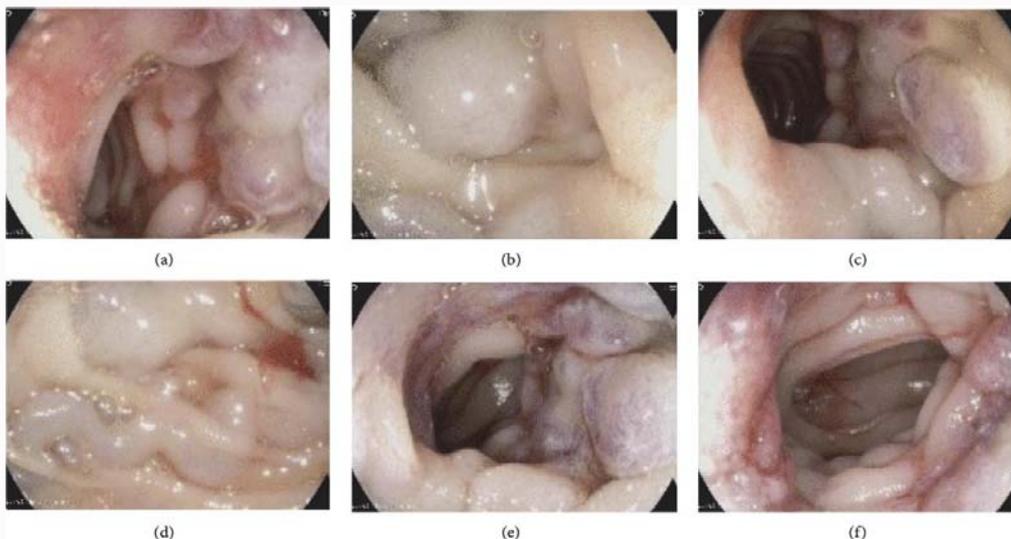


Figure 1: images of push enteroscopy showing multiple dilated jejunal varices with evidence of recent bleeding.



Figure 2: Radiological imaging for the patient pre trials of embolization of the varices. A: Abdomen CT scan with contrast shows distended portal vein with filling defect (white arrow point to the thrombus); B: Cross-section CT shows distended splenic vein with filling defect (white arrow); C: Coronal section of abdominal CT showing distended portal confluence with filling defect occluding the lumen (white arrow).

angiography of the abdomen showed no evidence of active intra-abdominal bleeding. CT abdomen with contrast was sought which demonstrated total occlusion of superior mesenteric and splenic veins with well-established collateral venous circulation (Figures 2A-2C). In 2019, patient had selective angiogram with embolization of the jejunal varices using metal coils (Figures 3A-3D). He was started on IV PPI infusion and later he received IV terlipressin along with ceftriaxone 1 g/day for 5 days after the enteroscopy findings. A Multidisciplinary Team (MDT) meeting was arranged between foregut surgeon, interventional radiologist, and gastroenterologist. The agreement was to surgically resect the affected small bowel segment for definitive therapy as the patient continued to have lower GI bleeding and this was the fourth hospitalization with melena.

Laparoscopic findings of short segment of proximal jejunum covered with sub serosal varices fed by large veins from the mesenteric border (Figures 4A-4D). Laparoscopic segmental small bowel resection with side-side jejunum-jejunostomy stapled anastomosis was done. The surgery and postoperative period were uneventful; he was discharged four days after surgery. Patient remained symptom free and did not develop any further episodes of melena up to one year follow up.

Discussion

Ectopic varices are the least common culprit causing variceal bleeding and one of the least common sites to be affected is the jejunum. Cirrhosis leading to portal hypertension is the most

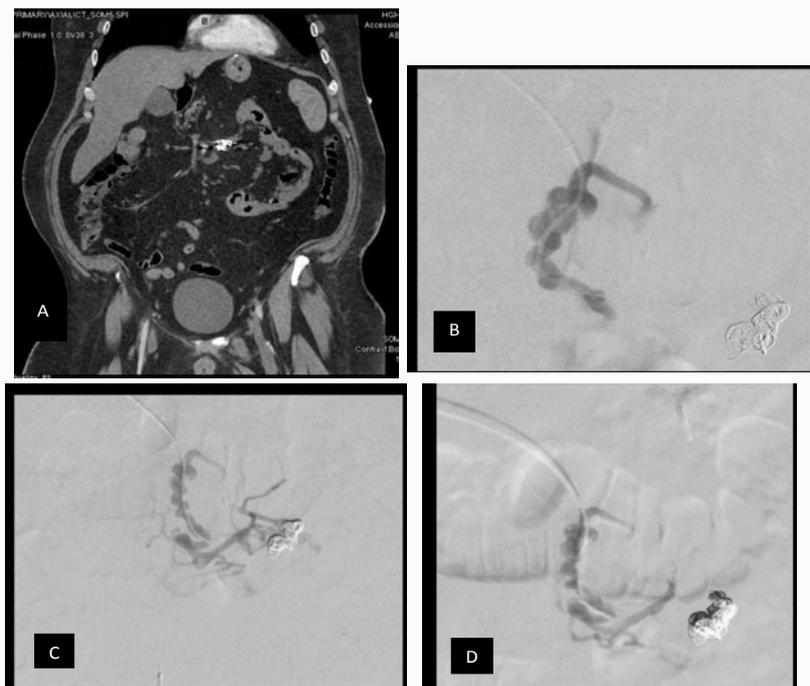


Figure 3: Radiological images post three trials of embolization and coiling of the varices.
 A: Coronal CT scans showing coils in the jejunal varices (arrow in A).
 B-D: interventional radiology showing varices (arrow in B) and coil in the jejunal varices (arrow in C).



Figure 4: Intra-operative images showing isolated segment of the jejunum with ectopic engorged varices.
 A-D: intra-operative images showing isolated jejunal varices (white arrow).
 D: post resection and anastomosis of healthy jejunum

common etiology for ectopic varices; whereas extra hepatic portal hypertension is a very rare reason for ectopic varices [1]. Most cases of ectopic varices are segmental (involving a segment of small bowel) and secondary to portal hypertension or portal venous obstruction, however, some isolated cases were reported in the absence these conditions [2-13]. Presence of ectopic varices quadruple the risk of bleeding when compared with esophageal varices [3] and can have a mortality rate as high as 40% [4]. Etiology of jejunal variceal formation is not completely understood. Common predisposing risk factors are hepatic cirrhosis with portal hypertension, hepatocellular carcinoma and/or pancreatitis [6]. A familial aggregation of small

intestinal varices in the absence of portal hypertension has been described [7]. Intra-abdominal adhesions from previous abdominal surgery, arteriovenous fistula secondary to trauma and nodular lymphoid hyperplasia have been also reported [8-20]. Jejunal varices carry a high mortality rate if not properly and timely managed [21]. Chronic portomesenteric thrombosis leads to development of venous collaterals which can bleed leading to melena and sometimes passage of large amount of fresh blood per rectum like our patients in this case. This is the second reported case of ectopic varices with mesenteric vein thrombosis attributed to heterozygous factor V Leiden mutation [22]. Our patient was kept on lifelong anticoagulation due to extensive

Table 1: Summarizes the different modalities available for management of ectopic varices.

Therapeutic options	Description	Pros	Cons
Endoscopic Therapy	Band ligation, sclerotherapy, injections	Accessible, available	High rebleeding rate
TIPS	Trans-jugular intra hepatic portal systemic shunting. Shunt procedure with concomitant embolization for better success and decrease rate of rebleeding	High success in selected patients, can be used as primary modality or salvage therapy	Requires interventional radiology, not suitable in occluded varices Risk of hepatic encephalopathy
BRTO	Balloon occluded retrograde transvenous obliteration. Shunt closure procedure	High success rate in gastric varices. Need more studies for isolated ectopic varices	Requires expert interventional radiology. Not feasible in occluded varices
Surgical Resection	Surgical resection of the involved bowel with varices	Definitive therapy Low risk of recurrence	Risk associated with major intestinal resection if long segment is involved

PMVT. High index of suspicion is needed to diagnosis such difficult cases of ectopic intestinal varices. Many diagnostic modalities can be used to assess small bowel lesions but push enteroscopy is more sensitive and allows for interventional opportunities. However, push enteroscopy does not come without risks and it calls for a trained gastroenterologist to perform it [17]. There are no clinical trials to guide the management of ectopic varices; evidence about management is limited to a few case reports and retrospective case series. Some of the different modalities that have been used include TIPS (Transjugular Intrahepatic Portosystemic Shunts) and BRTO (Balloon-occluded Retrograde Transvenous Obliteration). Our gastroenterology group has studied this patient back in 2018 and published the medical management for ectopic varices [23]. We are updating the literature with the surgical management for such patients. There is an ongoing debate over the best management of such varices either by decompression with (TIPS), transvenous obliteration or through surgical bowel resection. Unfortunately, the number of ectopic varices ‘reports in the literature is small and no definite conclusion can be made as to which is the ideal way to manage these varices [5]. Endoscopic management of jejunal varices including endoscopic band ligation and injection sclerotherapy had been reported [18], but its role is limited and carries high rates of recurrent variceal bleeding [19]. Embolization, via intervention radiological techniques, using steel coils, thrombin, gel foam, collagen or autologous blood clot, either alone or in combination with band ligation or TIPS, are an effective short-term therapy for bleeding ectopic varices [9]. The rate of successful bleeding control has been reported to reach 94%; unfortunately, embolization does not decompress the portal venous system, resulting in high 1-year re-bleeding rates reaching up to 8% annually [10]. If endoscopic modalities or interventional embolization fail to control bleeding from ectopic varices, creating a TIPS or proceeding with surgery could be optimal option depending on the availability of expertise, liver function and the cause of portal hypertension. Management of ectopic varices is the most challenging aspect of such cases. There are no clinical trials to guide the management of ectopic varices. Evidence for management is limited to a few case reports and retrospective case series. Some of the different modalities that have been used include TIPS (Transjugular Intrahepatic Portosystemic Shunts) and BRTO (Balloon-occluded Retrograde Transvenous Obliteration). TIPS offer a highly effective modality to control bleeding, the long-term survival of patients is mainly dependent on their liver function. The high efficacy of TIPS has to be balanced against the potential for increased encephalopathy and the procedure-related morbidity. TIPS together with variceal embolization has the advantage of being effective, minimally invasive, can be performed in one session, does

not preclude subsequent liver transplantation, and therefore may be used during the acute situation both as a bridge to transplantation and as the definitive therapy in patients unfit for surgery [1]. Direct surgery or local de-vascularization of the varices is a useful minimally invasive procedure that does not take much time, does not involve resection of long segments of the small intestine and can be done even if the portal vein is not patent or if the patient has Child B or C cirrhosis [11,12]. Taking into consideration the previously mentioned points about TIPS; as this patient had normal liver function and his portal hypertension was due to his chronic medical predisposition; decision was taken to explore his abdomen and perform resection as there was an isolated short segment involved of the small bowel. Table 1 summarizes the different modalities available for management of ectopic varices. Surgical resection of the affected bowel segment showed low recurrence rates in few reported cases and proven to be a viable option for difficult cases with recurrent hospitalizations due to melena [13]. In some reports; surgery was considered the main treatment option of bleeding jejunal varices especially in isolated cases [14-16]. However, high risk and morbidity of surgery should be taken into consideration in cirrhotic patients [4,17].

Conclusion

Ectopic varices are rare; however, they carry a difficult diagnostic and therapeutic dilemma, so they should be managed in a multidisciplinary team with patient centered approach. Surgical resection of the involved bowel segment remains a feasible and safe approach particularly in presence of an isolated short segment of small bowel and other minimally invasive approaches failed to stop the bleeding.

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