HHS Public Access

Author manuscript

Am J Med Case Rep. Author manuscript; available in PMC 2019 October 24.

Published in final edited form as:

Am J Med Case Rep. 2019 ; 7(4): 62–66. doi:10.12691/ajmcr-7-4-3.

Duodenal Varix Rupture – A Rare Cause of Fatal Gastrointestinal Hemorrhage: A Case Report and Review of Literature

Sadat Iqbal[1], **Michelle Likhtshteyn**[1], **Daniel O'Brien**[1], **Samy I. McFarlane**[1,*], **Manuel Martinez**[1,2]

[1]Department of Internal Medicine, SUNY Downstate Medical Center, Brooklyn, NY, USA

[2]Department of Gastroenterology, Veterans Affairs Hospital, Brooklyn, NY, USA

Abstract

Duodenal varices are a rare complication of portal hypertension; with cirrhosis being the most common cause. Reports regarding the disease prognosis and natural history are limited. In addition to the diagnostic difficulty, ectopic duodenal varices pose a significant therapeutic challenge owing to the lack of specific management guidelines. Given the high risk of rupture, they can have devastating clinical outcomes. Rupture typically presents as a gastrointestinal hemorrhage and requires emergent interventions. We present a case of duodenal varix seen on upper endoscopy in a patient with portal hypertension and cirrhosis, together with review of the literature outlining the current understanding of this disease entity. We also highlight the pathogenetic mechanisms as well as the current diagnostic and therapeutic approaches for this potentially fatal disease.

Keywords

duodenal varices; gastrointestinal hemorrhage; cirrhosis; portal hypertension

1. Introduction

Duodenal varices are quite rare, representing only up to 17% of all ectopic varices. [1] The majority of cases are discovered incidentally and are not a common cause of variceal bleeding. [1] However, these type of variceal lesions have poor prognosis, particularly when associated with bleeding, with a mortality rate nearing 40%. [2] Duodenal varices are most common in patients with portal hypertension (PHT) of which 30% are known to be caused by liver cirrhosis; other causes of duodenal varices include idiopathic PHT or extra-hepatic obstruction of the portal vein. [3] The resistance of blood flow in the intra-hepatic portal vein with PHT leads to opening of embryonic channels resulting in variceal formation (either in the gastroesophageal tract or ectopic locations). [4] Given advances in endoscopic and imaging techniques, ectopic varices are being identified with increased frequency. However, due to the paucity of data from randomized controlled trials, guidelines for management of duodenal varices are quite limited. Currently avilable treatment modailtiies

*Corresponding author: Samy.mcfarlane@downstate.edu.

include endoscopic and radiologic intervention, as well as surgery in complicated cases. [5,6]

2. Report of the Case

A 55-year-old man presented to the Emergency Department with bright red blood per rectum ongoing for one day. He had several episodes of soft, brown stools mixed with blood after accidentally taking a higher than usual dose of lactulose. There was no report of abdominal pain, distention, nausea, vomiting, hematemesis, or rectal pain. Furthermore, there were no symptoms suggestive of hemodynamic compromise on physical exam. His past medical history was significant for coronary artery disease, prior polysubstance and alcohol abuse. The patient also had a history of hepatitis C-related cirrhosis with episodes of encephalopathy, ascites, esophageal varices, and portal and splenic vein thrombosis. The patient was not on anticoagulants or non-steroidal anti-inflammatory drugs at the time of presentation. He was receiving home hospice care and was able to independently perform most activities of daily living.

On admission, his blood pressure was 105/76 mm Hg without orthostasis, heart rate 70 beats per minute, temperature 98.6○F, and respiratory rate 20 breaths per minute. His body mass index (BMI) was 23.9 (normal, 18.5–24.9). Physical examination revealed temporal wasting, no icterus, and dry mucous membranes. His abdomen was soft, non-tender and non-distended with normal bowel sounds. No scars or hernias were noted. On digital rectal examination, trace amount of red blood was observed, but rectal vault was devoid of stool content. His laboratory evaluations are consisent with anemia and relatively normal LFTs suggestive of advanced cirrhosis (Table 1). Upper endoscopy was planned to explore the cause of his bleeding. Overnight the patient sustained a large episode of bright blood per rectum, but remained hemodynamically stable with hemoglobin of 8.8 g/dL (normal, 13–18 g/dL). An esophagogastroduodenoscopy (EGD) was performed which was notable for small esophageal varices without stigmata of recent bleeding and mild portal hypertensive gastropathy in the gastric body and antrum. A 1.5 cm submucosal bulge was noted in the second portion of the duodenum with an apical red spot, suggestive of a duodenal varix (Figure 1). Of note, two years ago on an EGD a submucosal bulge was recognized in the same spot that was positive for the “pillow sign” (Figure 2).

Treatment of the duodenal varix was planned with endoscopic hemoclipping. As the hemoclip (Resolution 360™ Clip, Boston Scientific, Natick, MA) was deployed, the lesion started profusely bleeding and obscured endoscopic visibility preventing further intervention. The endoscope was then withdrawn from the patient. The patient was emergently intubated for airway protection; efforts to resuscitate the patient with fluids and blood transfusion were initiated. His computed tomography (CT) scan of the abdomen and pelvis performed six months ago was reviewed. It was notable for portal vein thrombosis, periduodenal, perisplenic varices, perigastric and periesophageal varices (Figure 2A & Figure 2B). After consulting surgery and interventional radiology (IR), the patient was transferred to the IR suite for an emergent angiogram. The arteriogram revealed a normal celiac axis, with normal caliber hepatic and splenic arteries and the gastroduodenal artery was markedly narrowed (Figure 3A). The angiogram also revealed evidence of portal vein

thrombus and duodenal and gastroesophageal varices (Figure 3B). Given the endoscopic and fluoroscopic images, the source of bleeding was consistent with a duodenal varix that eroded into the second portion of the duodenum presenting as a sentinel bleed. The gastroduodenal artery was then pre-emptively embolized with injection of gelfoam slurry and placement of a 3-mm diameter coil (Figure 3C).

There was no clinical evidence of further bleeding following the angiogram and embolization. However, the patient remained hemodynamically unstable and required pressors as well as ventilatory support for aspiration pneumonia. His underlying cirrhosis and presumed aspiration pneumonia portended a poor prognosis. The patient's clinical course was complicated by multi organ failure and he expired within 24 hours post admission.

3. Discussion

Duodenal varices were first visualized endoscopically in 1973. [1] Since then, only a handful of cases have been documented. As most information is gleaned from case reports, experience has been limited given the lack of randomized controlled trials (RCT) informing treatment guidelines. Fortunately, in addition to being less common than esophageal or gastric varices, bleeding risk is also relatively low. [2] However, the cause for concern is a very high mortality when bleeding does indeed occur. These entities are often found in patients with PHT secondary to cirrhosis; splenic vein thrombosis and occlusion of the portal vein are other notable causes. [2] Duodenal varices develop with collateral veins forming between the superior mesenteric vein/portal trunk and inferior vena cava. [3] Most often, duodenal varices form closer to the duodenal bulb, which is a component of the first part of the duodenum; the frequency of presentation decreases towards the distal portion of the duodenum. [3] In our patient, the lesion involved the second portion of the duodenum. Although few cases have been reported in Japan, duodenal varix in the second portion of the duodenum is a remarkably rare occurrence in the United States. [4] The mucosal color of small bowel varices may differ minimally from that of the surrounding mucosa. As in our case, on three prior endoscopic evaluations a 1.5 cm submucosal smooth lesion in the mid to distal second portion of the duodenum was described, it did not flatten when probed; it was taken to be consistent with a leomyoma, a GIST or potentially a duplication cyst (Figure 2A). On one endoscopy, the 1.5cm lesion appeared soft on probing and completely flattened intermittently during peristalsis, which was further suggestive of a duplication cyst (Figure 2B). Before and after images of the "pillow sign" were taken and what is apparent is that the overlying mucosa was the same color and consistency as the surrounding duodenal mucosa (Figure 2A,B,C).

Duodenal varices are often identified endoscopically and can be found in duodenal portions that are relatively difficult to reach using an upper endoscope. In cases with high suspicion that cannot be identified endoscopically, other diagnostic modalities might be necessary; including CT scan, angiography or even surgical exploration. [4] In cases with obscure gastrointestinal bleeding or patients with chronic liver disease, small bowel capsule endoscopy has also been shown to be helpful. [5] Eisen *et al.*[6] tested the efficacy of PillCam ESO™ capsule endoscopy in 32 patients with liver cirrhosis for the detection of

esophageal varices and portal hypertensive gastropathy. They reported a concordance of 96.9% and 90.6% for upper endoscopy and capsule endoscopy respectively in the diagnosis of varices and portal hypertensive gastropathy. McCarthy *et al.* [7] performed a systematic review and meta-analysis that demonstrated a diagnostic accuracy of wireless capsule endoscopy (CE) in the diagnosis of esophageal varices was 90% (95% CI, 0.88–0.93). They concluded that although Wireless CE is well tolerated, current sensitivity is insufficient to replace an index EGD screening for varices. Tang, S *et* al reported a case series of diagnosing small-bowel varices by wireless CE. Over a 12-month period, small bowel varices were found in 4 of 46 patients (8.7%) who underwent CE for GI bleeding. Their conclusion was that CE is an invaluable tool for diagnosing ectopic varices. [5]

With regards to initial management using pharmacologic therapy, the Baveno VI consensus recommends starting vasoconstrictors, such as terlipressin, somatostatin, or octreotide and antibiotics within 24 hours of suspected variceal bleeding. [8] However, medical therapy has limited efficacy in cases of actively bleeding ectopic varices. [7] In terms of definitive therapy, surgical, endoscopic or interventional radiology procedures are used. While surgical methods including variceal suture ligation or resection were previously preferred, given the high rate of re-bleeding and mortality endoscopic and radiologic methods have recently come to the forefront of therapy. [7] Currently surgery is reserved as a treatment of last resort.

Keeping in mind that endoscopic treatment of duodenal varices is often operator dependent, endoscopic variceal ligation (EVL) and endoscopic variceal obturation (EVO) using cyanoacrylate have been shown to be successful in achieving primary hemostasis and having less adverse events compared to surgery. In a case series of 8 patients by Park *et al.*, hemostasis was achieved with various endoscopic methods alone without requiring surgical or radiologic treatment. [7] Five patients achieved hemostasis with cyanoacrylate injections, 2 with endoscopic variceal ligation and one with hemoclipping; a technique that was employed in our patient. [7] Only one of the five cases managed with cyanoacrylate injection experienced a re-bleed thus further proving the authors' view that cyanoacrylate is the safest and effective method for hemostasis. EVL and EVO both have few adverse events, including treatment-induced ulceration, perforation, and stricture. However, EVO may also cause embolic events or bleeding due to cast extrusion from the injector needle site and portal biliopathy. [7] Placement of stents to control esophageal variceal bleeding have been reported, yet to date there are no reports of utilizing this as an option for ectopic varices of the duodenum.

Interventional radiology approaches include percutaneous transhepatic obliteration (PTO), transileocolic vein obliteration (TIO), balloon-occluded retrograde transvenous obliteration (B-RTO) or transjugular intrahepatic portosystemic shunt (TIPS). [3] While TIPS is highly efficacious, in up to 67% of the time in a published case series of 28 patients [8], increased risks of encephalopathy and procedure-related morbidity must be taken into account. TIPS can be used as first-line therapy or as second-line treatment if endoscopy fails. Its use as first-line therapy has been observed more frequently for patients unfit for surgery or as a bridge to transplantation. [8] B-RTO is effective for patients with bleeding at a lower pressure, patients with hepatic encephalopathy and when the portal vein is not patent. BRTO

with or without transiliocolic obliteration of vein has been shown to be effective for up to 3 years without complication or recurrence in bleeding in patients who failed endoscopic therapy or are not surgical candidates. [8] B-RTO however is not widely available, (reports showing its success mainly originated in Japan) and lack of technical expertise among interventionalists, especially given the complexities in its use. [8]

Injection therapy with a sclerosing agent is increasingly becoming first-line modality despite considerable associated recurrence risks. Furthermore, repeat injection therapy can increase the risk of perforation due to the thin structure of duodenal wall. [9] EVL is lower on the list likely due to concerns of ligation when the varix is large, difficulty in obtaining good views, and accessibility to collateral vessels with severe bleeding. [9] Hemoclipping on the other hand, which was selected for our case has the advantage of occluding the blood vessel by applying direct mechanical pressure and therefore is useful in the coagulopathic patients. [9-11] Furthermore, the effectiveness of hemoclipping has been shown to be similar to band ligation, however no head to head studies exist to compare between these interventions.

In this patient, hemoclipping was unsuccessful and therefore the patient underwent emergent angiogram followed by embolization with gelfoam slurry, and 3 mm coil placement. While our patient was successfully embolized, due to his hemodynamic instability and multi organ failure, he was transferred to the intensive care unit where he shortly expired. Although the present case resulted in a poor outcome, it is important to keep in mind that no further bleeding was noted after embolization and the cause of death in our case was likely due to advanced stage of liver failure as well as failure in other organ systems.

4. Conclusions

We believe our case report should raise awareness that duodenal varix should be included in the differential diagnosis of upper gastrointestinal hemorrhage, especially when the patient is known to have PHT/cirrhosis. Prompt endoscopic recognition of this lesion is fundamental to a successful treatment strategy. This case also suggests that direct referral to IR or surgery might be a prudent strategy given the high risk of complications if endoscopic intervention is attempted.

References

[1]. Bhagani S, Winters C, Moreea S. Duodenal variceal bleed: an unusual cause of upper gastrointestinal bleed and a difficult diagnosis to make. BMJ Case Rep. 2017; 2017.

[2]. Park SB, Lee SH, Kim JH, et al. Successful treatment of duodenal variceal bleeding by endoscopic clipping. Clin Endosc. 2013; 46(4): 403–6. [PubMed: 23964340]

[3]. Mora-Soler A, Velasco-Guardado A, Acosta-Materan R, et al. Endoscopic treatment of duodenal varices with cyanoacrylate. Rev Esp Enferm Dig. 2013;105(10): 629–32. [PubMed: 24641462]

[4]. Sato T Treatment of ectopic varices with portal hypertension. World J Hepatol. 2015;7(12): 1601–5. [PubMed: 26140080]

[5]. Tang SJ, Zanati S, Dubcenco E, et al. Diagnosis of small-bowel varices by capsule endoscopy. Gastrointest Endosc. 2004; 60(1): 129–35. [PubMed: 15229446]

[6]. Eisen Glenn M. et al. “The accuracy of PillCam ESO capsule endoscopy versus conventional upper endoscopy for the diagnosis of esophageal varices: a prospective three-center pilot study.” Endoscopy 38 1 (2006): 31–5. [PubMed: 16429352]

[7]. McCarth T,, Afinogenova Y Basile N, Use of Wireless Capsule Endoscopy for the Diagnosis and Grading of Esophageal Varieces in Patients with Portal Hypertension: A Systematic Review and Meta-analysis. J Clin Gastroenterolo. 2017 2: 51(2) 174–182.

[8]. de Franchis Roberto, on behalf of the Baveno VI Faculty, Expanding consensus in portal hypertension Report of the Vaveno VI Consensus Workshop: Stratifying risk and individualizing care for portal hypertesion Journal of Hepatology, 2015 vol. 63 743–752. [PubMed: 26047908]

[9]. Park SW, Cho E, Jun CH, et al. Upper gastrointestinal ectopic variceal bleeding treated with various endoscopic modalities: Case reports and literature review. Medicine (Baltimore). 2017; 96(1): e5860. [PubMed: 28072750]

[10]. Helmy A, Al Kahtani K, Al Fadda M. Updates in the pathogenesis, diagnosis and management of ectopic varices. Hepatol Int. 2008; 2(3): 322–34. [PubMed: 19669261]

[11]. Gunjan D, Sharma V, Rana SS, Bhasin DK. Small bowel bleeding: a comprehensive review. Gastroenterol Rep (Oxf). 2014; 2(4): 262–75. [PubMed: 24874805]

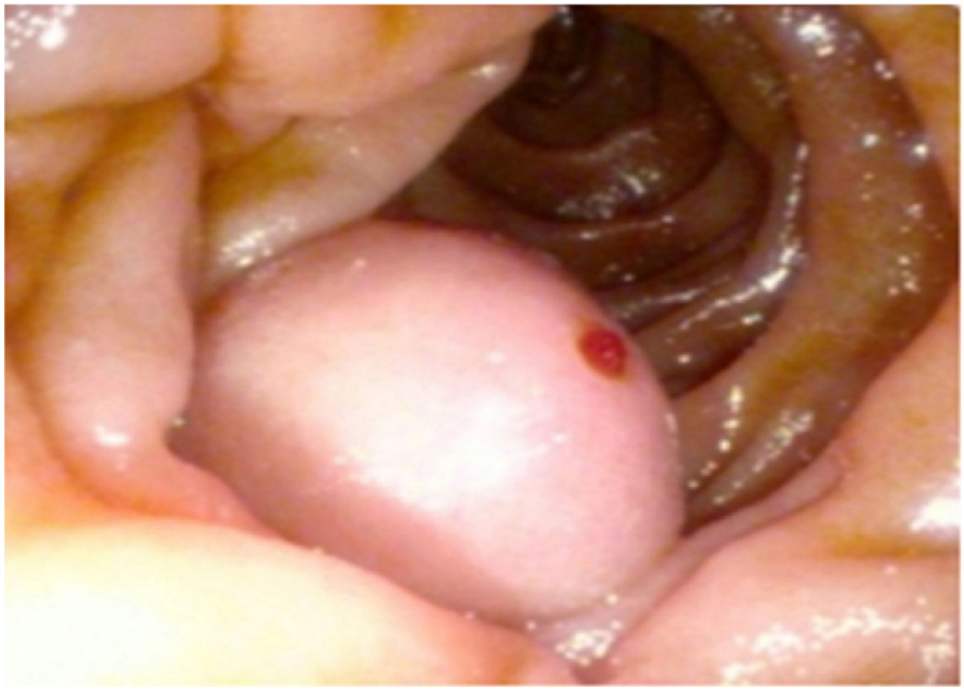

Figure 1.
From endoscopy on presentation described in case report. A submucosal bulge in the second portion of the duodenum with stigmata of bleeding at the apex noted by the apical red spot. As mentioned in the case report, a hemoclip was deployed and the lesion began actively bleeding and prevented further endoscopic visualization

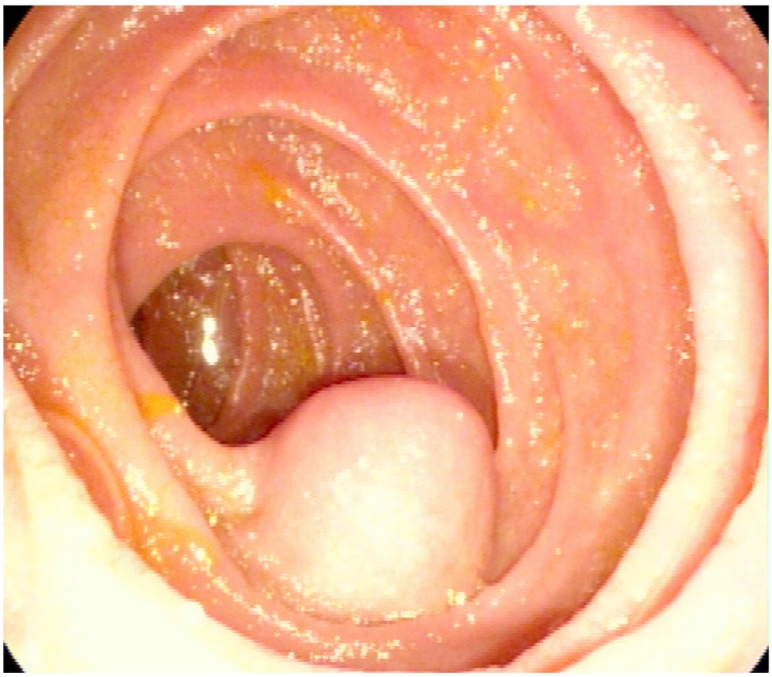

Figure 2A.
A 1.5cm round lesions with smooth border noted in the second portion of the duodenum

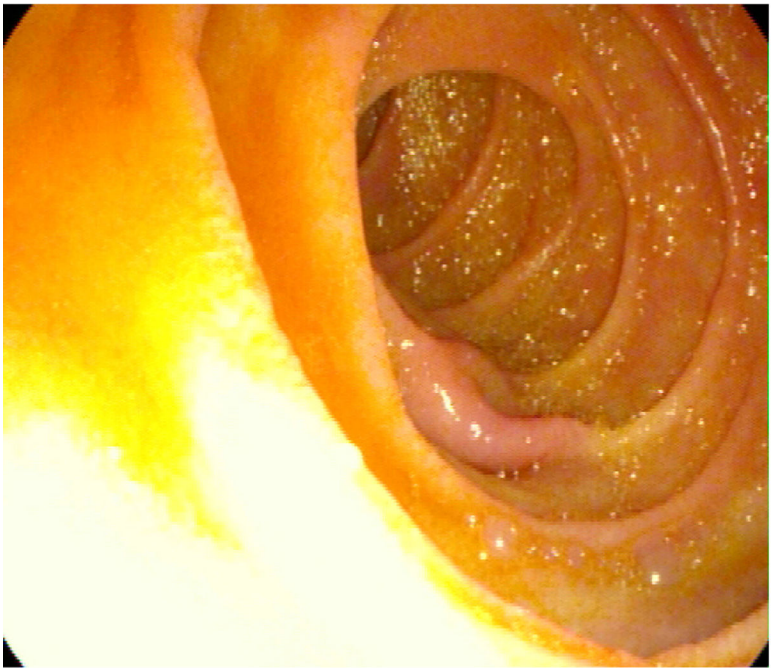

Figure 2B.
A 1.5cm lesion seen in the second portion of the duodenum; the mass appeared soft on probing and completely flattened during peristalsis

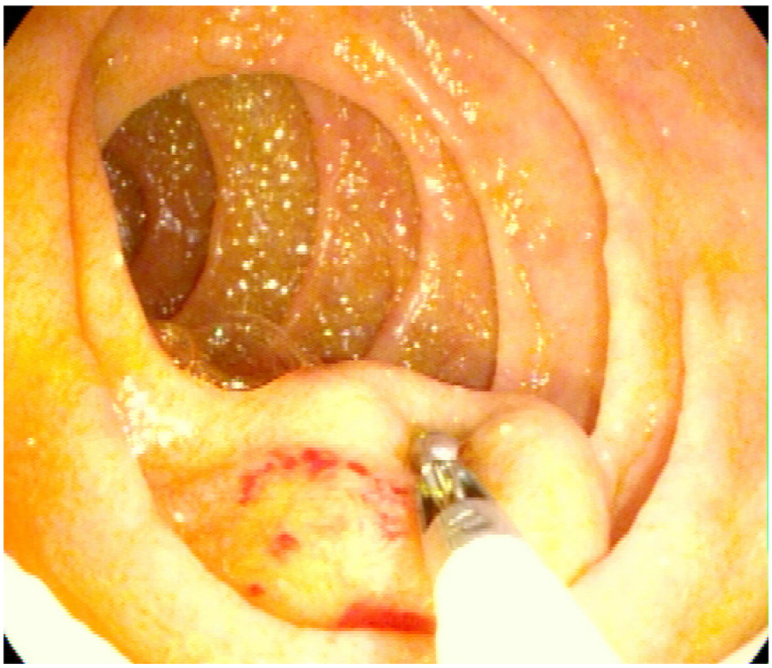

Figure 2C.
Duodenal submucosal bulge in second portion of duodenum that was positive for a “pillow sign”

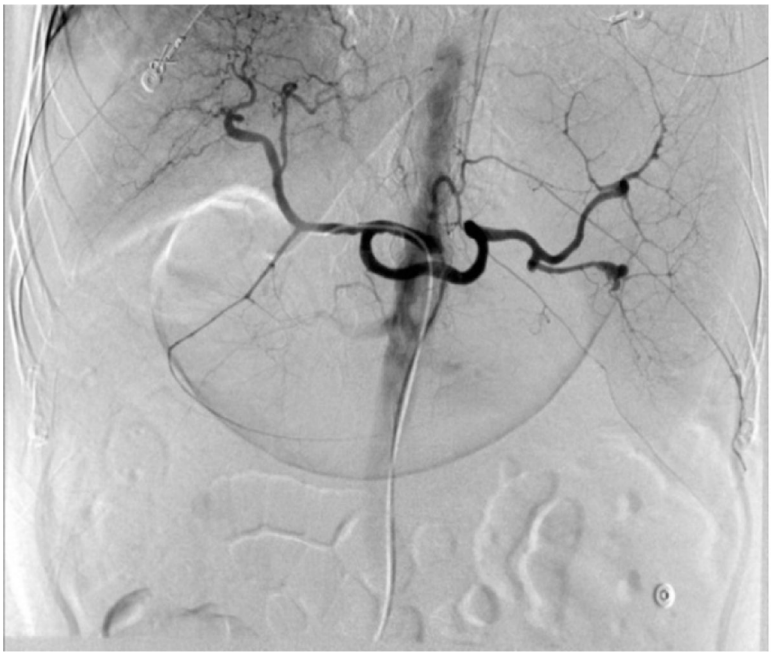

Figure 3A.

Arteriogram revealed a normal celiac axis, with normal caliber hepatic and splenic arteries and the gastroduodenal artery was markedly narrowed

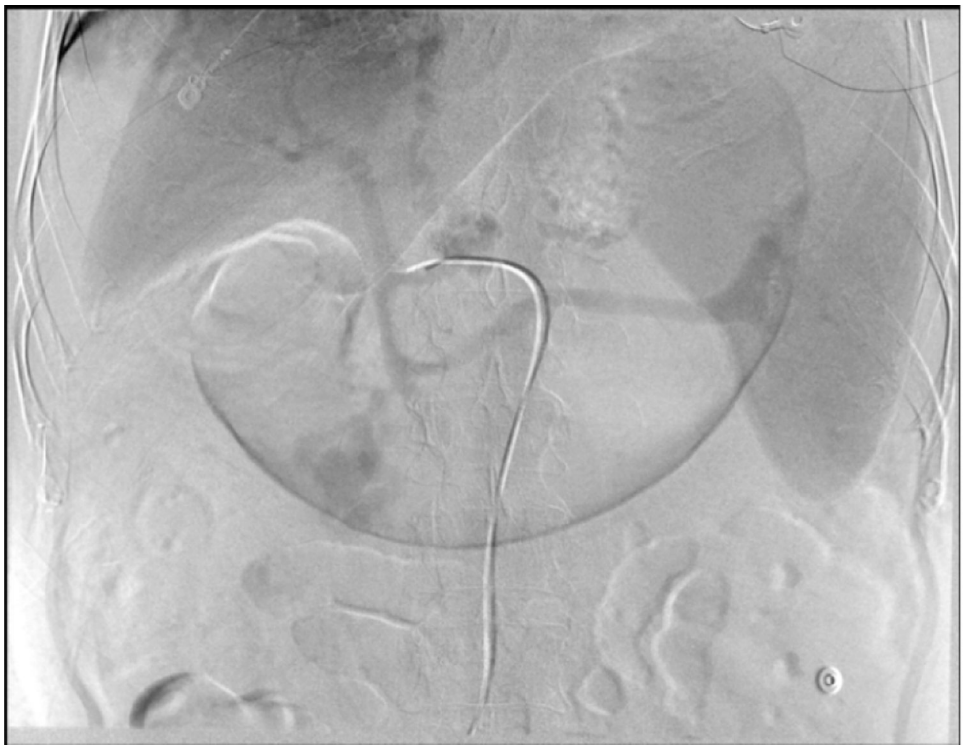

Figure 3B.
The angiogram also revealed evidence of portal vein thrombus, duodenal and gastroesophageal varices

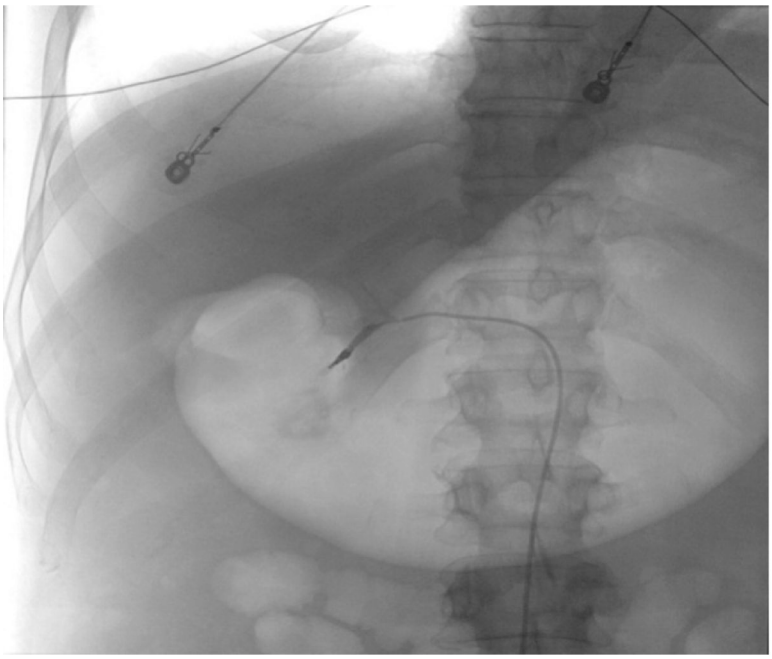

Figure 3C.
The gastroduodenal artery was pre-emptively embolized with injection of gelfoam slurry and placement of a 3 mm diameter coil

Table 1.

Initial laboratory investigations of the patient with respective reference ranges

Laboratory	Specimen	Patient	Reference range
White cell count	Serum	3.6	4.5–11.0/uL
Hemoglobin	Serum	8.8	13–18 g/dL
Hematocrit	Serum	25.80	40%−52%
Total proteins	Serum	2.8	6–8 mg/dL
Albumin	Serum	1.2	3.8–5.1 g/dL
Total bilirubin	Serum	0.7	0.1–1.2 mg/dL
ALP	Serum	36	44–147 IU/L
AST	Serum	18	5–40 U/L
ALT	Serum	41	7–56 U/L
Glucose	Serum	246	65–115 mg/dL
Blood urea	Serum	20	6–22 mg/dL
Creatinine	Serum	0.7	0.4–1.2 mg/dL
Sodium	Serum	142	135–145 mmol/L
Potassium	Serum	5.7	3.5–5.0 mmol/L
Chloride	Serum	120	100–110 mmol/L
Calcium	Serum	5.9	24–32 mmol/L
Magnesium	Serum	1.9	1.46–2.68 mg/dL
INR	Serum	1.41	0.8–1.2
PT	Serum	16.6	11–14 s
PTT	Serum	29.4	25–35 s

Abbreviations: ALP, alkaline phosphatase; AST, aspartate transaminase; ALT, alanine aminotransferase; INR, international normalized ratio; PT, prothrombin time; PTT, partial thromboplastin time