Treatment of Refractory Gastrointestinal Bleeding in Patients With Portal Hypertension: A Case Series and Treatment Algorithm

J. Balogh[a,b,*], S. Gordon-Burroughs[a,b], P. Schwarz[e], J. Galati[f], R.A. McFadden[f], M. Cusick[c], M.J. Snyder[c], H.R. Bailey[c], M. Weiner[d], A. Wong[d], R.A. Ochoa[b], A. Saharia[a,b], A.O. Gaber[a,b], and R.M. Ghobrial[a,b]

[a]Sherrie and Alan Conover Center for Liver Disease and Transplantation; [b]Department of Surgery; [c]Department of Colon and Rectal Surgery, Department of Surgery; [d]Interventional Radiology; [e]Department of Gastroenterology; and [f]Department of Hepatology, Houston Methodist Hospital, Houston Texas

ABSTRACT

In patients with portal hypertension, ectopic varices can develop at any site along the gastrointestinal tract outside the classically described gastroesophageal location. Like esophageal variceal hemorrhage, bleeding from ectopic varices can be life-threatening. Diagnosis and treatment of ectopic varices can be challenging; to date, no effective treatment algorithm has been described. A systematic teamwork approach to diagnosing and treatment of ectopic varices is required to successfully manage hemorrhage from ectopic varices.

INTESTINAL portal hypertensive varices are rare but challenging sequelae of mesenteric portal hypertension (pHTN). pHTN is caused by increased intrahepatic or extrahepatic mesenteric venous pressure due to cirrhosis or mesenteric thrombosis or stenosis. Varices are classically described as portosystemic communications and most commonly occur in the distal esophagus and rectum. The presence of abnormal collateral vessels in the abdomen is the most sensitive (70%–83%) and specific sonographic sign for the diagnosis of pHTN [1]. Mesenteric blood flow can be obstructed from occlusion (eg, extrahepatic portal vein obstruction) or distortion (eg, hepatic cirrhosis), leading to the development of collateral pathways as blood attempts to bypass the occlusion, thereby allowing flow down a pressure gradient from high-pressure to low-pressure vessel or vascular bed [2]. It has been proposed that the severity of the pHTN is demonstrated by the number of collateral channels that develop. This is said to be due to the duration of pHTN and the differential gradient driving the flow between the portal and systemic circulation. Others hypothesize that the formation of portosystemic collateral circulation is due in part to angiogenesis secondary to elevated levels of vascular endothelial growth factor [2]. As a consequence of obstruction to the portal circulation, a network of distended fragile splanchnic varices develops or embryonic channels recanalize [3] and may rupture, potentially leading to exsanguination [4]. Portosystemic beds prone to variceal formation are known to be gastroesophageal, rectal, and caput medusa. Ectopic varices are large portosystemic venous collaterals occurring anywhere in the abdomen except the locations noted previously [4]. Ectopic intestinal varices may also be found at sites of prior resections, anastomoses, and ostomies [4–6].

The development of varices is an important prognostic marker in the progression of pHTN. Spontaneous acute variceal bleeding from nonoperated small intestine has been reported to occur in 5% to 6% of patients with pHTN [7]. A sentinel variceal bleed can occur without warning and is a major cause of mortality among patients with pHTN [6]. These varices are difficult to localize and control during massive hemorrhage. Norton et al reviewed 169 cases of bleeding ectopic varices and the commonest site was the small bowel; 17% were duodenal, 17% were jejunal or ileal, 14% were colonic, 8% were rectal, and 9% were peritoneal. Mortality from bleeding ectopic varices varies between 1% and 5% [4].

The management of bleeding ectopic varices in the small bowel is challenging and rare, thus the development of an algorithm to manage these patients successfully may help improve outcomes. Herein, we report 3 cases of massive bleeding from ectopic jejunal varices successfully managed with a combination of coil embolization and mesenteric vein sclerosant, and we propose a treatment algorithm for the care of such patients.

*Address correspondence to Julius Balogh, MH, MHA, Houston Methodist Hospital, 6550 Fannin St Smith Tower 1661, Houston, TX 77030. E-mail: jbalogh@houstonmethodist.org

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CASE REPORTS

Patient 1

A 45-year-old man with alcoholic cirrhosis and a Model for End-stage Liver Disease (MELD) score of 16 was transferred to our facility for melenic stool and coffee-ground emesis. His past medical history was significant for diabetes mellitus type II, cholelithiasis, and rectal abscesses. Urgent esophagogastroduodenoscopy (EGD) was performed. This demonstrated 2 large (>5 mm) nonbleeding varices in the middle and lower third of the esophagus, mild portal gastropathy, fresh blood in the duodenum, with an additional varix in the third portion of the duodenum (>5 mm) with fibrin clot attached. The esophageal varices were banded and hemostasis achieved by the end of the procedure. Despite this, the rectal bleeding continued, and he was taken for an emergent transjugular intrahepatic portosystemic shunt (TIPS) procedure. Prior to graft placement, a catheter was advanced inferiorly through the portal vein into the superior mesenteric vein (SMV) and a large varix noted upon injection of contrast in the mid-SMV. Despite control of the large esophageal varices with concomitant portal pressure reduction, the patient continued to have significant blood loss via rectum and nasogastric tube, necessitating resuscitation with blood and blood products. The patient was taken back to the interventional radiology suite to investigate TIPS patency or persistent varix, at which time a 14-mm SMV varix with significant flow but no extravasation was demonstrated. The varix was embolized with coils and 3% Sotradecol/Lipiodol slurry solution with GelFoam. The patient's hematochezia and hematemesis briefly stopped postprocedure but then recurred with hematochezia. An EGD was repeated at bedside without any additional findings. Given the continued bleeding and lack of obvious source, the patient was sent for a triple-phase computerized tomography (CT) scan angiogram demonstrating splenomegaly (21 cm), multiple varices in the SMV, and increased diameter of the portal vein (26 mm), splenic vein (25 mm), and SMV (22mm) consistent with severe pHTN. Subsequent tagged red blood cell scan demonstrated massive amounts of tracer uptake in the left upper quadrant, consistent with active jejunal hemorrhage. When correlated with the CT scan, an additional varix was noted to be patent with contrast in the arterial phase. A balloon-occluded retrograde transvenous obliteration procedure was planned. Systemic venogram did not demonstrate varix. Additional venograms of the left renal vein and splenic vein did not demonstrate variceal flow. Reaccess of the portal system via the TIPS was accomplished by entry in the right internal jugular vein into the SMV and demonstrated residual varix arising from the distal aspect of the vein coursing medially and inferiorly with delayed images demonstrating opacification of the small bowel likely representing jejunum. This appeared to be the culprit source of hemorrhage. The varix was embolized using multiple coils and a slurry of GelFoam and contrast to sclerose the varix (Fig 1). Total product administration was 34,326 mL of packed red blood cells (PRBCs), fresh frozen plasma (FFP), cryoprecipitate, and platelets. Following the second embolization, the patient recovered without incident and was discharge home on day 4 postprocedure.

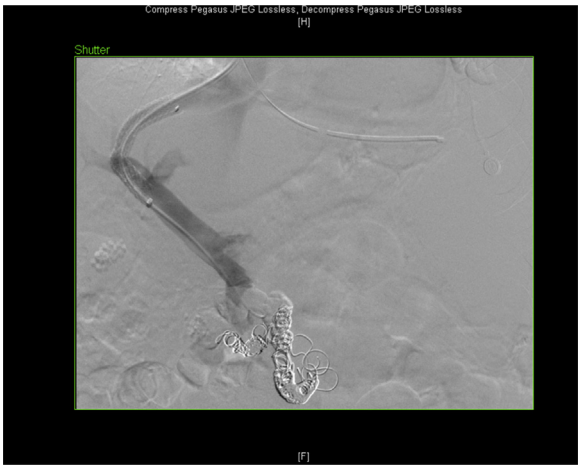

Fig 1. Patient 1, second variceal embolization.

Patient 2

A 64-year-old man was admitted with a history of passing melenic stool for 72 hours. His past medical history was significant for hypertension, diabetes mellitus type 2, hyperlipidemia, and colon cancer. His surgical history was significant for total abdominal colectomy with ileorectal anastomosis 5 years prior and negative for recurrence on surveillance. Social history was positive for 4 to 5 alcoholic beverages a day. His calculated MELD was 13. A triple-phase CT scan demonstrated gastroesophageal varices, splenomegaly, and infrarenal ectopic varix measuring 2.4 × 2.5 cm. Hepatic ultrasound demonstrated hepatopetal flow within the portal vein with suggestion of hepatic fibrosis or early cirrhosis and mild splenomegaly. The hematochezia continued and an aortogram was performed, which did not demonstrate extravasation of contrast from any jejunal branches. Due to persistent bleeding and clinical deterioration, he was taken to the operating room for exploratory laparotomy, lysis of adhesions, transabdominal balloon endoscopy, and small bowel resection with primary anastomosis. No obvious source of bleeding was identified during the procedure. The rectal bleeding continued, and therefore venous and atrial pressures were obtained using a transjugular access. The hepatic wedge pressure was 19 mm Hg, free hepatic pressure 15 mm Hg, right atrial pressure 16 mm Hg. TIPS was thought to not be indicated. Supportive treatment was continued and the bleeding ceased, but hematochezia recurred a few days later. Capsule endoscopy was performed and no site of bleeding was noted; however, melena was seen within the small intestine. Upper balloon-assisted enteroscopy was performed, which demonstrated a normal esophagus but congestive gastropathy with a normal duodenum. Following the procedure, his melena or hematochezia ceased and he was discharged home. The total amount of blood product administered was 11,534 mL of PRBCs, FFP, cryoprecipitate, and platelets. Eleven days postdischarge, he represented with recurrent gastrointestinal bleeding (melena and hematochezia). A transjugular liver biopsy was performed and showed stage 3 fibrosis, mild steatosis (macrovascular and focal microvesicular), and focal mild portal chronic inflammation with microfocal minimal interface hepatitis. Focal incomplete cirrhosis could not be entirely excluded at this time. Multiple transfusions were required to resuscitate the patient's continued bleeding. The patient was sent for TIPS with embolization. The pre-TIPS pressures were right atrial 8 mm Hg, portal 26 mm Hg, with a gradient pressure of 18 mm Hg. The post-TIPS and embolization pressures were right atrial 11 mm Hg, portal 17 mm Hg, with a gradient of 5 mm Hg. Access to the area for embolization was achieved via the SMV, which demonstrated multiple venous collaterals on venogram. The left lower quadrant was noted to have a large cluster of varices in the proximity of small bowel. Following the TIPS with embolization, the bleeding ceased