Successful Treatment of a Case of Ectopic Jejunal Varices with Portal Venous Stenting

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Patient: Female, 77
Final Diagnosis: Ectopic variceal bleeding of the jejunum
Symptoms: Abdominal pain • hematemesis • melena
Medication: —
Clinical Procedure: Portal vein stenting
Specialty: Gastroenterology and Hepatology

Objective: Unusual clinical course
Background: Management of patients with ectopic variceal bleeding can be challenging and requires a multidisciplinary approach. Ectopic bleeding from jejunal varices at the anastomotic site of choledochojejunostomy is rare and difficult to treat.

Case Report: We report a case of ectopic jejunal variceal bleeding secondary to portal vein stenosis at the anastomotic site of a prior choledochojejunostomy for pancreatic cancer. The patient was successfully treated with portal vein metallic stent placement that remained patent for 7 months.

Conclusions: The management of ectopic variceal bleeding remains difficult and controversial. Portal vein stenting has been reported as a successful method of treatment that is minimally invasive especially in patients who had prior major surgeries.

MeSH Keywords: Gastrointestinal Hemorrhage • Jejunal Diseases • Portal Vein

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**Background**

Massive gastrointestinal (GI) hemorrhage from GI variceal bleeding is the most serious and life-threatening complication of portal hypertension. Typical locations for variceal hemorrhage include areas of physiologic portosystemic shunts with the esophagus and stomach as the most common site for bleeding. Variceal bleeding outside the gastroesophageal junctions is considered ectopic bleeding and can involve any locations along the GI tract. Portal hypertension secondary to liver cirrhosis is the classic underlying etiology in most patients. Other causes of portal hypertension include portal vein thrombosis and stenosis secondary to inflammatory changes or underlying malignancies. Management of patients with ectopic variceal bleeding can be challenging and requires a multidisciplinary approach. Ectopic bleeding from jejunal varices at the anastomotic site of choledochojejunosotomy is rare and difficult to treat.

**Case Report**

We describe a 77-year-old female with a past medical history significant for pancreatic adenocarcinoma that was diagnosed 3 years prior to presentation status post Whipple procedure with portal vein reconstruction and adjuvant chemotherapy with gemcitabine and capecitabine. The patient complained of melena and hematochezia that had been going on for the last week prior to her presentation. She noted associated chronic abdominal pain, mainly epigastric, and intermittent with no relation to exertion or oral intake. The pain was associated with nausea and abdominal distention but no vomiting, hematemesis or aspiration episodes. Patient denied any previous history of upper GI bleeding with no recent esophagogastroduodenoscopy (EGD).

Physical examination revealed a pale emaciated female in moderate distress with a systolic blood pressure of around 70–80 mmHg. On examination, abdominal distention with diffuse tenderness to palpation was noted. Bedside ultrasound showed minimal ascites but no rebound tenderness or signs of acute abdomen. Initial laboratory investigations were significant for a hemoglobin level of 6.6 mg/dL and elevated blood urea nitrogen in the 60–70 mg/dL range. Liver enzymes and bilirubin were consistent with a cholestatic picture without evidence of severe synthetic dysfunction.

The patient was managed initially with supportive therapy and fluid resuscitation. She received 2 units of packed red blood cells, in addition to intravenous proton pump inhibitors and octreotide infusion. EGD showed patent choledochojejunosotomy with hemorrhagic appearance. Medium sized dilated blood vessels at the anastomosis area were seen (Figures 1, 2). Attempted ablation by coagulation grasper was unsuccessful and resulted in brisk bleeding. Complete hemostasis was achieved by epinephrine injection. Computed tomography (CT) scan of the abdomen was consistent with marked narrowing of the portal vein reconstruction with apparent collateralization around the choledochojejunosotomy site (Figures 3, 4). Liver and spleen were unremarkable without evidence of hepatic cirrhosis. A multidisciplinary meeting was held to discuss the case with interventional radiology, surgery, and gastroenterology. Transjugular intrahepatic portosystemic shunt (TIPS) was considered initially but the patient did not receive the procedure due to lack of hepatic sinusoidal gradient. The portal vein stenosis was treated with endovascular metal stent placement via a percutaneous approach.
transhepatic balloon dilatation resulting in a significant reduction in prestenotic portal pressure and relief of the jejunal varices. The stent remained patent with no further evidence of bleeding for 7 months without concurrent anticoagulation therapy as the risks were estimated to outweigh the benefits at that time. The patient had another attack of ectopic variceal bleeding secondary to stent thrombosis in the setting of pancreatic cancer recurrence. Goals of care were discussed with the patient and her family who elected to proceed with comfort care measures without invasive procedures.

Discussion

Massive GI hemorrhage from GI variceal bleeding is the most serious and life-threatening complication of portal hypertension. Portal hypertension results from increased pressure within the splanchnic circulation secondary to the resistance of venous return to the liver. The most common cause of portal hypertension is liver cirrhosis. Other causes include obstruction of portal circulation from underlying venous blood clots and underlying malignancies. Typical locations for variceal bleeding involve areas of physiologic portosystemic shunts with the gastroesophageal junction as the most commonly reported site. GI varices outside the esophagus and stomach are considered atypical or ectopic and can involve any location along the GI tract. Ectopic variceal bleeding represents an unusual and rare source for GI hemorrhage that is reported in up to 5% of variceal bleeding cases. Common sites for ectopic variceal bleeding are the duodenum and areas of previous surgery. The peritoneum, rectum, and ovaries are less commonly reported.

Atypical GI bleeding can be classified as either overt or occult bleeding depending on the presence or absence of symptoms, respectively. Clinical features for atypical variceal bleeding are similar to those of typical varices. Patients classically present with profuse hematochezia and melena. Hematemesis is less commonly reported but can occur with duodenal varices. Other reported symptoms include occult GI bleeding, iron deficiency anemia, and hypovolemic shock. High degree of suspicion is needed in patients presenting with the aforementioned symptoms in the setting of negative upper and lower endoscopy.

Evaluation of atypical varices involves upper GI endoscopy to assess for common sites of bleeding at esophagus and stomach, in addition to exclusion of other causes of GI hemorrhage. Initial assessment by EGD and colonoscopy should be repeated if needed in an attempt to identify the source of bleeding. When the initial diagnostic workup is negative, the small bowel should be further investigated for a source of bleeding. Diagnostic tools to evaluate the small intestine are limited and include capsule endoscopy and double balloon enteroscopy (DBE). Prior studies showed that capsule endoscopy to be superior to other methods in the ability to detect abnormalities of the small bowel. Capsule endoscopy is relatively non-invasive; however, it is a diagnostic technique and does not allow for therapeutic interventions. Computed tomography (CT) scan with angiography can be utilized to evaluate overt GI bleeding especially in patients with suspected lower GI bleeding. Other modalities include red blood cell scintigraphy, magnetic resonance angiography, endoscopic ultrasound (EUS), laparoscopy or laparotomy.
Management of ectopic varices can be challenging due to the diversity of locations and various clinical presentations. Treatment should always consider the risks and benefits of available therapeutic interventions. A multidisciplinary approach guided by the available local expertise should always be considered. Surgical approach involves resection or direct ligation of the involved segment [10,11]. Percutaneous embolization has been reported as a successful form of treatment in some patients [12–14]. Transjugular intrahepatic portosystemic shunts (TIPS) can successfully treat ectopic variceal bleeding in the setting of portal hypertension in up to 90% of patients, especially in case of recurrent bleeding [3,15,16]. Another endovascular technique is balloon-occluded retrograde transvenous obliteration (BROTO) that utilizes a specially shaped balloon catheter with a sclerosing agent [17–22]. Hekmat et al. reported a successful obliteration of jejunal varices by the use of cyanoacrylate through DBE [23].

Portal venous stenting has been reported as a viable therapeutic intervention for the management of jejunal varices secondary to portal vein compression [24–26]. Portal vein stent placement reduces portal hypertension and related symptoms in 84–85% of patients with malignant portal stenosis. The mean patent period is variable and ranges from 4 to 21 months [27–29]. Yamakado et al. noted higher rates of stent occlusion in patients with splenic vein involvement, obstruction of the portal venous system, severe hepatic dysfunction, and pancreatic cancer [27].

Our patient had ectopic jejunal variceal bleeding at the surgical anastomosis site of a prior choledochojejunostomy. Further evaluation was consistent with underlying portal hypertension secondary to narrowing of the portal vein and associated varices of the small bowel. She was successfully treated with portal vein stenting to reduce the elevated pressure within the portal system as a palliative measure. Her stent remained patent for 7 months and was later occluded by stent thrombosis. Saeki et al. described similar cases that were successfully managed by percutaneous embolization and mesocaval shunting [14]. Portal vein stenting remains a viable form of treatment with a variable stent patency period depending on the type of stent used and the underlying etiology.

Conclusions

Management of ectopic variceal bleeding can be challenging as it is difficult to diagnose such cases while treatment remains controversial. Patients with jejunal variceal bleeding after a choledochojejunostomy are not healthy enough to undergo surgical treatment most of the time. Non-invasive interventions should be considered as a form of palliative treatment in those patients.

Conflicts of interest

None.

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