

Bleeding Ectopic Varices: An Uncommon Cause of Hemorrhagic Ascites in Cirrhotic Patients

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Abstract

Bleeding ectopic varices are a very rare cause of hemorrhagic ascites in cirrhotic patients. We presented a case of a 52-year-old cirrhotic man with a history of hepatitis C for 20 years, with complaints of abdominal distension and discomfort for 1 day. Paracentesis showed frank hemorrhagic ascites. There was no evidence of hepatocellular carcinoma or spleen rupture on the abdominal CT scan. In absence of these causes, the ectopic variceal bleed was suspected to be the most likely cause. Although endoscopic intervention with sclerotherapy and band ligation, trans jugular intrahepatic portosystemic shunting, endovascular embolization, transvenous balloon occlusion, and percutaneous transhepatic embolization had all been employed in the management of ectopic varices with success. But these had failed to identify bleeding origin in some cases and surgery being the only option remaining.

Introduction

Ascites is one of the most common complications of cirrhosis. Decompensated ascites develops in about 57 % of cirrhotic patients after 10 years of diagnosis [1]. Patients having cirrhosis fail to maintain normal arterial pressure and systemic vascular resistance. This leads to the development of hyperdynamic circulation with raised cardiac output. In response, kidneys retain sodium and water. All these factors lead to the development of ascites. Ascites also marks poor prognosis in cirrhotic patients. The study has shown 15% cirrhotic patients presenting with ascites die within 1 year and 44 % die within 5 years [2].

However, hemorrhagic ascites is a very rare complication in cirrhotic patients. It happens in less than 5 % of cirrhotic patients [3]. Management of hemorrhagic ascites is very challenging as it is very hard to determine etiology. Prognosis is very poor for

cirrhotic patients having hemorrhagic ascites. Mortality associated with hemorrhagic ascites is very high about 87% at one month and 72 % at one year [4]. Hemorrhagic ascites in cirrhotic patients may be caused by hepatocellular carcinoma, traumatic rupture of the spleen, iatrogenic puncture of vessels during paracentesis, and bleeding ectopic varices.

Ectopic varices are dilated portosystemic collaterals located at any point other than the esophagogastric region. In cirrhotic patients, ectopic varices dilate and bleed due to portal hypertension. Ectopic varices can develop at various locations like duodenum, jejunum, ileum, colon, rectum, peritoneum, retroperitoneum, abdominal wall, vagina, ovary, etc. Hemorrhagic ascites usually develops from bleeding peritoneal varices

Case Presentation

A 52-year-old man came to the emergency with a history of rapid abdominal distention and discomfort for 1 day. The patient was a known case of Hepatitis C for 20 years and was diagnosed with cirrhosis 3 years ago. Patient-reported dull abdominal pain that was diffuse. His oral temperature was 99°F (37.2 °C), heart rate of 102 beats per minute, blood pressure 90/60 mm Hg, respiratory rate 20 per minute and oxygen saturation of 99 % on 2 L/min oxygen by nasal cannula. On physical examination, there was mild scleral icterus, hand tremors, mild abdominal tenderness, and

shifting dullness to percussion without rebound tenderness. Paracentesis showed grossly hemorrhagic ascitic fluid. Serum hematocrit was

25.5 %, hemoglobin 6.7 g/dl, INR 1.8, platelet count $90 \times 10^3/\mu\text{L}$, total serum bilirubin 4.5 mg/dl, serum potassium 2.6 mEq/L. ECG showed sinus tachycardia. An abdominal computed tomography (CT) scan showed a small liver with diffuse nodules, an enlarged spleen, mild portal vein distention, and massive dense cities. He was started on intense fluid resuscitation with potassium and

vitamin K supplementation, transfused with 6 units of packed red blood cells and 6 units of fresh frozen plasma (FFP). His condition stabilized during his stay at an emergency and was transferred to the intensive care unit (ICU). Angiography was done which showed variceal oozing from the jejunal and ileal region.

Discussion

Hemorrhagic ascites is an uncommon finding in cirrhotic patients. It has been shown that it mostly results from structural lesion rather than low platelet count or coagulopathy in cirrhotic patients. Rupture of the spleen in cirrhotic patients having splenomegaly from minor or blunt trauma can cause hemoperitoneum but spontaneous hemorrhagic ascites is the most common etiology (up to 72 %) [4]. Rupture of hepatocellular carcinoma also contributes a major portion of hemorrhagic ascites cases in patients with cirrhosis ranging 13 % to 24 % [4]. Iatrogenic hemorrhagic ascites contributes to only about 11 % to 17 % of cases [4]. Iatrogenic ascites usually results from accidental puncture of the peritoneal or epigastric artery. But it is a rare cause of hemorrhagic ascites if hematocrit of the ascitic fluid is more than 5 % [5]. Repeating the paracentesis in such cases can be helpful [6].

If a cirrhotic patient presents with hemorrhagic ascites with no evidence of hepatocellular carcinoma lesion, no findings of traumatic spleen rupture and iatrogenic cause ruled out then rupture of an intraperitoneal ectopic varix, although a rare complication (only up to 5 % of all variceal bleeds) [7] of portal hypertension in cirrhotic patients, is most likely. Ectopic varices can be present in different locations. In a study by I D Norton et al. [8] of 169 cases, bleeding ectopic varices were in the peristomal area (26 %), duodenum (17 %), jejunum or ileum (17 %), colon (14 %), rectum (8 %), and peritoneum (9 %). Ectopic varices can also be rarely found in other locations like the ovary and vagina. Presenting complaints of bleeding ectopic varices depend on the location of the varices like hematemesis and hematochezia in small intestinal varices, fresh blood per rectum in rectal varices, and bleeding from the edge of the stoma in peristomal varices. Rupture of peritoneal ectopic varices leads to hemoperitoneum, and patients usually present with rapidly developing abdominal distention. Abdominal pain or discomfort can also be present. Massive bleeding can also lead to hypotension and patient can complain of lightheadedness or syncope. Hypovolemia due to bleeding can also lead to shock. Physical examination may show stigmata of cirrhosis like gynecomastia, spider angioma, palmar erythema, scleral icterus, asterixis, shifting dullness, etc. Patients with rupture of peritoneal varices can also have ecchymoses of flanks and periumbilical area (Grey Turner's and Cullen's sign) [5]. Cirrhotic patients with hemorrhagic ascites usually has higher rates of complications like hepatic encephalopathy, spontaneous

Embolization was attempted to stop bleeding from these sites. Despite all these measures patient had liver failure, raised intra-abdominal pressure, and worsening hepatic encephalopathy after the procedure and the patient expired 5 days after presentation.

bacterial peritonitis, acute kidney injury and mostly require management in ICU [4]. Hemorrhagic ascites is also shown to increase mortality in cirrhotic patients as compared to other similar patients without hemorrhagic ascites. A study by N H Urrunaga et al. [4] has shown that patients with hemorrhagic ascites had very high mortality 87 % at one month, 72 % at 1 year, and 61 % at 3 years.

Initial management of hemorrhagic ascites with unstable hemodynamics is aggressive fluid resuscitation. Two or more large-bore intravenous lines should be developed. Resuscitation with crystalloids and colloids should be started while waiting for blood grouping and matching. Vitals monitoring should be continued. Vasoactive drugs have been very beneficial in the management of esophageal varices but their role in ectopic varices is undetermined [9]. After fluid resuscitation, upper gastrointestinal endoscopy is considered first-line intervention. If it fails to find a bleeding site, colonoscopy is the next step in management. If endoscopy shows bleeding varices, then management with either sclerotherapy or band ligation should be done [10]. A study by T Sato et al. [11] has shown band ligation and sclerotherapy have similar clinical efficacy. Endoscopic management usually detects intestinal, colonic, and rectal varices but it has low efficacy for the detection of peritoneal varices. Diagnosis and management of ectopic peritoneal varices are very challenging as most of the interventions fail to show the origin of bleeding. Trans jugular intrahepatic portosystemic shunting (TIPS) is the best next step after the failure of endoscopic intervention. A study by Kochar N et al. [12] has shown that initial hemostasis was developed in 66.67% cases treated by TIPS presenting with acute ectopic variceal bleed [11]. However, the chances of rebleed remain high when treated with TIPS alone. TIPS also predispose to the development of hepatic encephalopathy. Endovascular embolization has also been shown promising results. Vengali M et al. [13] has shown that TIPS combined with embolization has reduced the chances of rebleed. Transvenous balloon occlusion [14] and percutaneous transhepatic embolization [15] have also been tried with success in the treatment of bleeding varices. Surgery is the last resort after all treatment options failure.

Laparotomy has both diagnostic and therapeutic roles. But it is not recommended as most patients with cirrhosis do not qualify for

surgery and mortality is also very high. The proposed mechanism is that blood in the peritoneum has a tamponade effect which prevents further bleeding. Laparotomy removes this tamponade effect which leads to heavy bleeding and rapid exsanguination of

Conclusion

Hemorrhagic ascites is an uncommon complication of cirrhosis. It may be due to traumatic spleen rupture, hepatocellular carcinoma lesion rupture, iatrogenic or ectopic peritoneal varices. If there is no hepatocellular lesion on imaging, no evidence of traumatic

the patient [5]. However, even after all these interventions bleeding ectopic varices cannot be found in some cases. Management of such cases remains a dilemma until this date and mortality are very high.

spleen rupture and iatrogenic cause ruled out by repeating paracentesis then bleeding ectopic varices are most likely. Diagnoses and management of ectopic varices is very challenging as most interventions may fail to identify a bleeding ectopic varix.

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