

CASE REPORT

Arteriovenous Malformation Causing Ileocecal Variceal Bleeding in Liver Cirrhosis: Case Report and Review of the Literature

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Varices that occur at sites other than the esophago-gastric area are termed ectopic varices. An ileal varix is a very rare cause of lower gastrointestinal bleeding. Although ileal varices are generally associated with prior intra-abdominal surgery and adhesions, an arteriovenous malformation (AVM) in the ileocecal area can cause ileal varices and bleeding in patients with portal hypertension who have not received previous intra-abdominal surgery, which is due to an intestinal or colonic AVM dilating the collateral veins and further aggravating portal hypertension. Surgical treatment should be considered in patients with massive ectopic variceal bleeding. We report a case of massive ileocecal variceal bleeding associated with an AVM that occurred in a patient with alcoholic liver cirrhosis. (*Gut and Liver* 2008;2:54-59)

Key Words: Ileocecal varix; Arteriovenous malformation; Portal hypertension

INTRODUCTION

Esophagogastric variceal bleeding is a frequent complication of the portal hypertension. In the setting of the portal hypertension, portosystemic collaterals can occur anywhere along the gastrointestinal tract. Ectopic variceal bleeding, which is defined as bleeding from outside the esophagogastric lesion, is an uncommon life-threatening complication of portal hypertension.¹ Bleeding from ileal varices is far less frequent and the reported cases seem to be associated with prior intra-abdominal surgery and adhesions.²⁻⁵ However, considering that intestinal and co-

lonic arteriovenous malformation (AVM) could cause dilated collateral veins and aggravate portal hypertension,^{6,9} AVM in the ileocecal area can cause ileal varices and bleeding in patients with the portal hypertension without previous intra-abdominal surgery. We report a case of massive bleeding from the ileocecal varices associated with AVM in a patient with alcoholic liver cirrhosis.

CASE REPORT

A 50-year-old man visited an emergency room due to massive hematochezia. He had histories of longstanding alcohol consumption and liver cirrhosis for 3 years. He had no history of operation. He passed large amount of bright red bloody stools on admission. On physical examination, ascites and hepatomegaly were prominently present. His blood pressure was 130/80 mmHg and pulse rate was 69/min. Laboratory findings were as follows: hemoglobin of 6.2g/dL; hematocrit of 19%; platelets of $2.43 \times 10^5/\text{mm}^3$; prothrombin time of 42.8%; serum albumin of 2.7 g/dL; total bilirubin of 4.5 mg/dL; aspartate aminotransferase/alanine aminotransferase of 563/149 IU/L. Hepatitis B surface antigen and hepatitis C virus antibody were negative. He was classified as having liver cirrhosis Child-Pugh class C at this point. Esophagogastroduodenoscopic examination revealed large non-bleeding esophageal varices and portal hypertensive gastropathy. The Colonoscopic examination showed severe bowel wall edema and a few diverticula in the ascending colon without an active bleeding site. One day later, he developed a massive hematochezia accompanied

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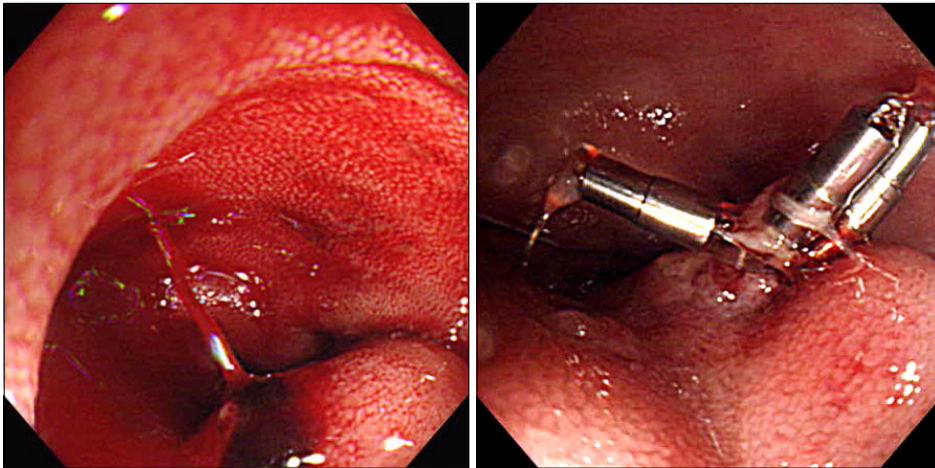


Fig. 1. Colonoscopic view of ileocecal variceal bleeding performed for a Dieulafoy lesion due to spurting bleeding at the terminal ileum. Hemoclips are applied.

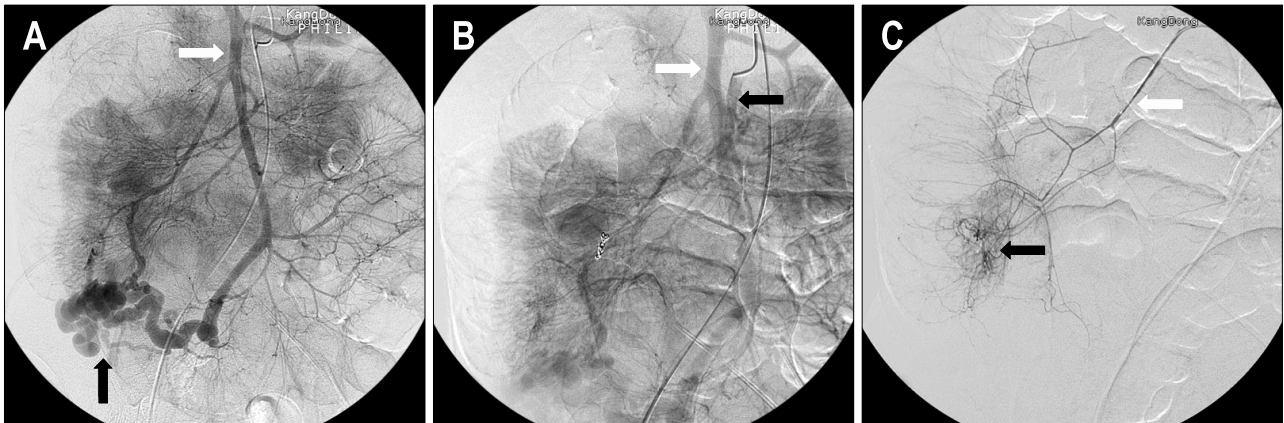


Fig. 2. (A) Selective angiography of the superior mesenteric artery (SMA) showing dilated tortuous veins (black arrow) in the delayed phase. White arrow: superior mesenteric vein (SMV). (B) Early filling of the dilated draining veins. Black arrow: SMA, white arrow: SMV. (C) Suspicious contrast extravasation (black arrow) in distal portion of the right colic branch, for which embolization is performed. White arrow: right colic artery.

by hypotension. Active resuscitation was commenced and emergent colonoscopy was performed again. The colonoscopy revealed a Dieulafoy lesion with active bleeding at the terminal ileum. Three hemoclips were applied to the bleeding site successfully (Fig. 1). However, on the next day there was a massive and recurrent bleeding despite the clipping, necessitating transfusion of 11 units of packed red cells. A superior mesenteric angiography was undertaken to exclude an arterial bleeding during the episode of acute bleeding. In delayed phase dilated and tortuous mesenteric varices were observed at the ileocecal area (Fig. 2A), and early filling of the dilated draining veins was observed in the ileocecal region (Fig. 2B). AVM with ileocecal varices was suspected. We performed coil embolization due to suspicious contrast extravasation observed in the distal portion of the right colic branch during the arterial phase (Fig. 2C). Nevertheless, massive

lower gastrointestinal bleeding continued. Since the patient remained in a hemodynamically unstable state, an emergency exploratory laparotomy was performed although the fact that he was Child-Pugh class C liver cirrhosis. At the operation, a nodular cirrhotic liver and massive ascites were found. There was a dilated and tortuous vein with a diameter of about 0.5 cm-1 cm, on the surface of the ileocecal wall. A bleeding point was found at the 5 cm proximal portion of the ileocecal valve. Segmental resection of the ileum and colon were performed. The specimen consisted of 15 cm of the terminal ileum and 5 cm of the colon. Histologic sections showed a focal increase in the number of irregularly enlarged submucosal vessels (Fig. 3). Microscopic examination demonstrated several dilated angiodysplastic vessels with ruptures and hematoma formation in the submucosa (Fig. 4A) and showed the vascular element, which con-



Fig. 3. Gross specimen shows a dilated submucosal vein in the resected ileum.

sisted of both thick and thin walled vessels (Fig. 4B). A plume of fibrin over a thick artery-like vessel was observed in the elastic stain (Fig. 4C). These findings were consistent with AVM. Although the patient's vital signs were stable and there was no further bleeding right after the surgical resection of the ileocecal varices, his condition deteriorated. The patient died of hepatic failure on the 13th day after the operation.

DISCUSSION

To our knowledge, this is the first case which illustrated that massive ileal variceal bleeding can be caused by AVM at the ileocecal area in a patient with alcoholic liver cirrhosis. Varices can occur in the distal gut, including small intestine and colon, though variceal bleeding is usually secondary to collateral venous pathways in the esophagus or stomach. Varices that occur at the site other than the esophagogastric area are described as ectopic varices and account for less than 5% of all varix-related bleedings.¹⁰ They are typically associated with portal hypertension. As the flow and pressure become increasingly high, portosystemic anastomotic veins may become ectatic and varicose in regions of potential portosystemic anastomoses.¹¹ Although the most common site of ectopic varices is duodenum, varices involving the small intestine are commonly found at anastomotic sites and in adhesions after abdominal surgery.²⁻⁵ Collaterals within adhesions are possible causes of ectopic varices, particularly, in jejunum and ileum. Thus, previous abdominal surgery in patients with portal hypertension may be a predisposing factor in the development of ileal varices. Ueda *et al.*⁵ reviewed 57 reports that described small intestinal varices in PubMed dating from 1951 to 2003. Previous abdominal

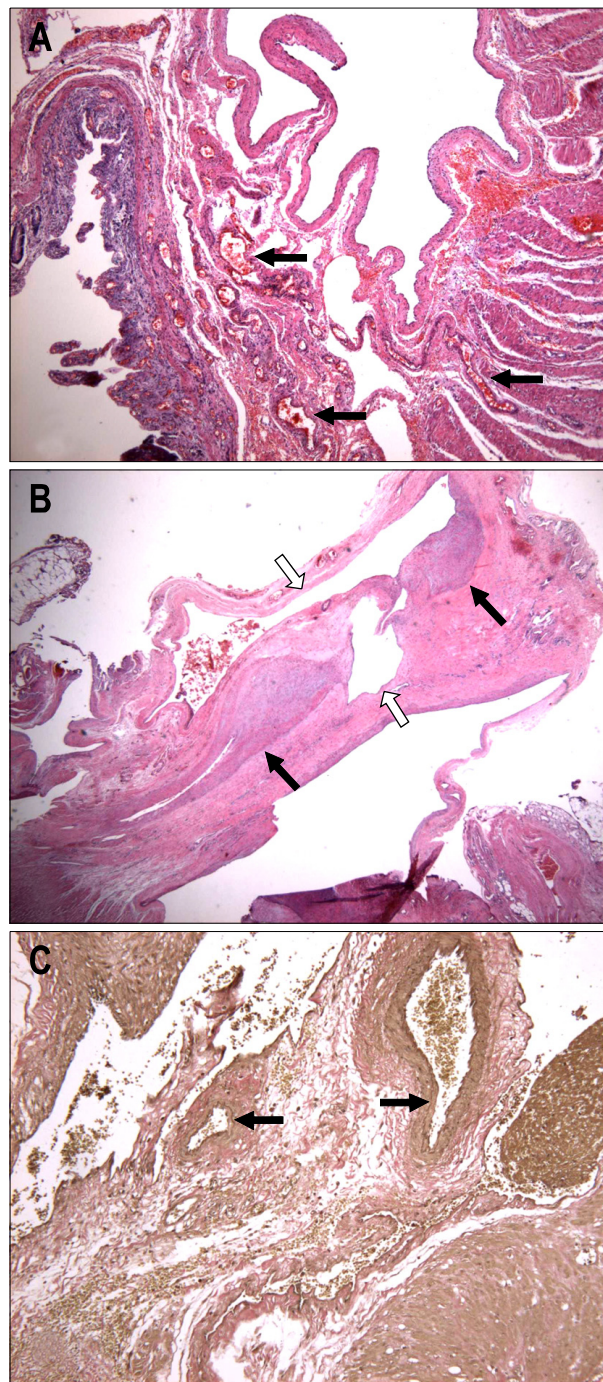


Fig. 4. (A) Irregularly enlarged angiodyplastic vessels with rupture and hematoma formation in the submucosa. (B) Thick- and thin-walled vascular channels (black and white arrows, respectively) in the submucosa (hematoxylin & eosin stain; ×10). (C) Internal elastic fibers (black arrow) in thick artery-like vessels (elastin stain; ×100).

surgery was noted in 48 cases (84%).

In our case, the patient had ileal varices without a history of abdominal surgery. However, mesenteric angiography showed AVM at the ileocecal area. Interestingly,

some studies noted that mesenteric AVM would cause portal hypertension and gastrointestinal variceal bleedings.^{6,7} Therefore, we hypothesized that the early and rapid drainage of blood through ileocecal AVM would develop dilated collateral vein at the ileocecal area and that accentuating the portal hypertension resulted in bleeding.

The term, AVM has been used to describe a wide variety of vascular lesions of the intestine, which include telangiectasia, angiodysplasia, vascular dysplasia and vascular ectasia.¹² Moore *et al.*¹³ classified intestinal AVM as follows: Type 1 is a solitary localized lesion within the right colon usually in older patients. Type 2 is a larger, endoscopically visible, and probably congenital lesion

common in small intestine. Type 3 is a punctate angio-mas causing gastrointestinal hemorrhage. Considering that the feature of AVM of our case was single, well localized and an endoscopically non-visible malformation within the right colon in an older patient, the AVM in this patient could be classified as an intestinal AVM in the Type I category. Colonoscopy has occasionally been of value in the diagnosis of colonic AVMs. Visible telangiectatic lesions and multiple areas of mucosal erosion have been most frequently described.¹⁴ However, in cases of emergency, early diagnoses were often difficult because the bleeding sites may not be revealed by endoscopic examinations due to limited and poor field of vision.

Table 1. Reported Cases of Ileal Variceal Bleeding

	Age/Sex	Underlying disease	Site of varices	Methods of diagnosis	Treatment	Outcome	Previous abdominal surgery	Reference no.
1	72/F	Alcoholic liver cirrhosis	Ileum	SMA angiography	Ileocelectomy	No further bleeding	Hysterectomy	1
2	66/F	HCV positive-liver cirrhosis	Ileum	SMA angiography	Segmental resection of the ileum	No further bleeding	Laparotomy for ectopic pregnancy	4
3	72/F	HCV positive-liver cirrhosis	Ileum	SMA angiography	Enterectomy	No further bleeding	(-)	5
4	43/M	Idiopathic	Ileum	SMA angiography	Segmental resection of the ileum	No further bleeding	(-)	7
5	52/F	HBV positive-liver cirrhosis	Ileum	SMA angiography	Segmental resection of the ileum	Died	Cholecystectomy	19
6	80/F	Periappendicular abscess	Ileum	SMA angiography	Segmental resection of the ileum	Died	(-)	20
7	56/F	Alcoholic liver cirrhosis	Ileum	Laparotomy	Ileocelectomy	Died	(-)	21
8	49/M	Alcohol liver cirrhosis	Ileum	SMA angiography	Segmental resection of the ileum	No further bleeding	Operation for rupture of esophageal varix	22
9	62/M	Deficiency of ileocecal vein	Ileocecum	SMA angiography	Rt. hemicolectomy	No further bleeding	Laparotomy for abdominal aortic aneurysm	23
10	54/M	Alcoholic liver cirrhosis	Ileum	SMA angiography	Ileocelectomy	Died	(-)	24
11	33/F	Alcoholic liver cirrhosis	Ileum	SMA angiography	Subtotal resection of the ileum	No further bleeding	Abdominal operation	25
12	56/F	Alcoholic liver cirrhosis	Ileum	SMA angiography	TIPS	No further bleeding	Pelvic surgery	26
13	62/F	Hepatocellular carcinoma	Ileum	SMA angiography	Ligation of ileocecal vein, and ovarian vein	No further bleeding	Hysterectomy	27
14	64/M	Idiopathic	Ileum~rectosigmoid colon	Colonoscopy	Ileocelectomy	No further bleeding	(-)	28
15	50/M	Alcoholic liver cirrhosis	Ileocecum	SMA angiography	Segmental resection of the ileum	Died	(-)	Current case

Ileocecal varices are often missed since air insufflations during the endoscopic examination raises the intraluminal pressure resulting in collapse of the varices.¹ We also could not recognize the varices or AVM during the colonoscopy in this case. The most accurate method in diagnosing ectopic varices and AVMs is selective mesenteric angiography. In ectopic varices the delayed phase of the angiography shows large dilated tortuous veins. Angiographic hallmarks of an AVM of the bowel include early venous filling with a dilated draining vein, a vascular tuft, and a slowly emptying vein.^{14,15} In the patient with an angiographically demonstrable AVM, preoperative arterial catheter placement and dye injection are the most efficient method for localizing these lesions, and it also provides a reliable means of limiting the extent of resection.¹⁴ Therefore, mesenteric angiography should be considered during the earlier stage when working-up cases involving suspected ectopic varix.^{15,16}

The histologic features of AVM are submucosal vascular malformation, the presence of thin walled ectatic venous channels, and thick walled fibromuscular vessels with internal elastic fibers connected with a thin walled vein,¹⁷ which are consistent with the pathologic results of our case.

The available therapies for variceal bleeding include conservative medical therapy; surgical options such as bowel resection and portosystemic shunt; and percutaneous treatment with either embolization or transjugular intrahepatic portosystemic shunt (TIPS).^{11,18} Coil embolization is a safe and technically easy treatment for ectopic variceal bleeding with effective, immediate results. However, recurrent bleeding is frequent and re-intervention is often required.¹⁸ Thus, surgery should be undertaken as soon as the diagnosis is made, since bleeding from ectopic varices is most often profuse and recurrent. Surgical management such as segmental resection or shunt operation seems to be successful for controlling the excessive bleeding from ectopic varices.²

In a review of the literatures in PubMed from 1982 to 2007, a total of 14 cases was reported as ileal variceal bleeding.^{1,4,5,7,19-28} Among these 14 cases of ileal varix, 12 cases were diagnosed with SMA angiography.^{1,4,5,7,19,20,22-27} All of these cases, bar one had operations performed.^{1,4,5,7,19,20,22-28} 8 cases of them had a history of previous abdominal surgery.^{1,4,19,22,23,25-27} There were no cases that involved AVMs. We illustrated a review of the 14 ileal variceal bleeding cases (Table 1).

In summary, we report a patient with alcoholic liver cirrhosis who had massive bleeding from ileocecal varices, which was associated with AVM. In patients with portal hypertension, ectopic variceal bleeding should be consid-

ered as another potential source of massive gastrointestinal hemorrhage.

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