

## Ectopic gastrointestinal variceal bleeding with portal hypertension

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### Abstract

Massive gastrointestinal bleeding from gastrointestinal varices is one of the most serious complications in patients with portal hypertension. However, if no bleeding point can be detected by endoscopy in the predilection sites of gastrointestinal varices, such as the esophagus and stomach, ectopic gastrointestinal variceal bleeding should be considered as a differential diagnosis. Herein, we report a case of ectopic ileal variceal bleeding in a 57-year-old woman, which was successfully diagnosed by multi-detector row CT (MDCT) and angiography and treated by segmental ileum resection. To date, there have been no consensus for the treatment of ectopic ileal variceal bleeding. This review was designed to clarify the clinical characteristics of patients with ectopic

ileal variceal and discuss possible treatment strategies. From the PubMed database and our own database, we reviewed 21 consecutive cases of ileal variceal bleeding diagnosed from 1982 to 2017. MDCT and angiography is useful for the rapid examination and surgical resection of an affected lesion and is a safe and effective treatment strategy to avoid further bleeding.

**Key words:** Ectopic gastrointestinal bleeding; Ileal varix; Portal hypertension

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**Core tip:** Massive gastrointestinal bleeding from gastrointestinal varices is one of the most serious complications in patients with portal hypertension. If no bleeding point can be detected by endoscopy in the predilection sites of gastrointestinal varices, ectopic gastrointestinal variceal bleeding should be considered as a differential diagnosis. We report here a 57-year-old female case of ectopic ileal variceal bleeding, which were diagnosed by multi-detector row CT (MDCT) and its angiography and treated by segmental ileum resection. From the review results of previous reports, MDCT and its angiography is a rapid and useful examination. Moreover, surgical resection of responsible lesion is safe and effective treatment strategy to avoid further bleeding.

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## INTRODUCTION

Massive gastrointestinal bleeding from a gastrointestinal varix is one of the most serious complications in patients with portal hypertension. However, if the point of continuous bleeding in the predilection sites of a gastrointestinal varix, such as the esophagus and stomach, is not found and no further strategy for the accurate diagnosis and effective treatment of the bleeding point exists, the condition may become life threatening.

Lebrec *et al*<sup>[1]</sup> classified the gastrointestinal varices other than those of the esophagus and stomach as ectopic varices. Ectopic gastrointestinal varices were reported in the sites of the duodenum, small intestine, colon, rectum, peristomal, biliary, peritoneal, umbilical, and other locations. Ectopic gastrointestinal varices cause an unusual hemorrhage and account for 5% of all variceal bleeding. In particular, ectopic ileal variceal bleeding is the major type of ectopic gastrointestinal variceal bleeding<sup>[2]</sup>. Herein, we report a case of ectopic ileal variceal bleeding, which was diagnosed by MDCT and angiography and was surgically treated. Moreover,

we reviewed previous case reports regarding the clinical behaviors, diagnosis, and treatment strategies of ectopic ileal variceal bleeding, including our cases diagnosed between 1982 and 2017 from the PubMed database.

## CASE REPORT

A 57-year-old Asian woman with autoimmune portal hypertension due to polymyositis was admitted to our hospital with a 2-d history of hematochezia. She had a history of esophageal variceal rupture, which had been treated by endoscopy 3 years before. At admission, she had a blood pressure of 92/58 mmHg, heart rate of 85/min, respiratory rate of 16/min, and body temperature of 35.2 °C. Although she was pale and showed conjunctival pallor, and there was no jaundice, abdominal pain, or shifting dullness. Laboratory data were as follows: hemoglobin 7.3 g/dL, hematocrit 23.4%, platelets 112000/mm<sup>3</sup>, prothrombin time 98%, serum albumin 3.5 g/dL, total bilirubin 1.1 mg/dL, aspartate aminotransferase/alanine aminotransferase 35/51 IU/L. Hepatitis B surface antigen was positive and hepatitis C virus antibody was negative. There was no encephalopathy. Her Child-Pugh score was 6 (class A).

We performed an emergent upper gastrointestinal endoscopy, which showed a mild esophageal varix without bleeding. However, lower gastrointestinal endoscopy revealed a large blood clot at the ileocecum, but there was no active bleeding lesion during the endoscopy. MDCT showed no definitive liver cirrhosis, but dilation of the hepatic portal vein and umbilical vein and splenomegaly and portosystemic collaterals indicated portal hypertension. In addition, enhanced MDCT and MDCT and angiography revealed the presence of an ileal varix, which showed no active bleeding into the abdominal cavity. In particular, the ileal varix had a portosystemic shunt *via* the superior mesenteric vein into the right ovarian vein.

She was treated conservatively for 2 d with a blood transfusion. On the 3<sup>rd</sup> day after admission, she had massive hematochezia. We performed a second MDCT and angiography and diagnosed the patient as hematochezia due to massive ileal varix bleeding because there was a massive coagula at the distal ileal lumen of the ileal varix. We performed emergent segmental ileal resection, which included the ileal varix, *via* a small laparotomy (Figure 1). The varix was located at the 20-cm proximal portion of the ileocecal valve. Her postoperative condition was uneventful. She had no further bleeding and was discharged on the 8th day after surgery.

## DISCUSSION

Portal hypertensive enteropathy is present in 5%-11% of patients with portal hypertension and often gives rise to gastrointestinal varices in the esophagus and stomach, which cause active bleeding<sup>[3]</sup>. Gastrointestinal varices other than those of the esophagogastric area

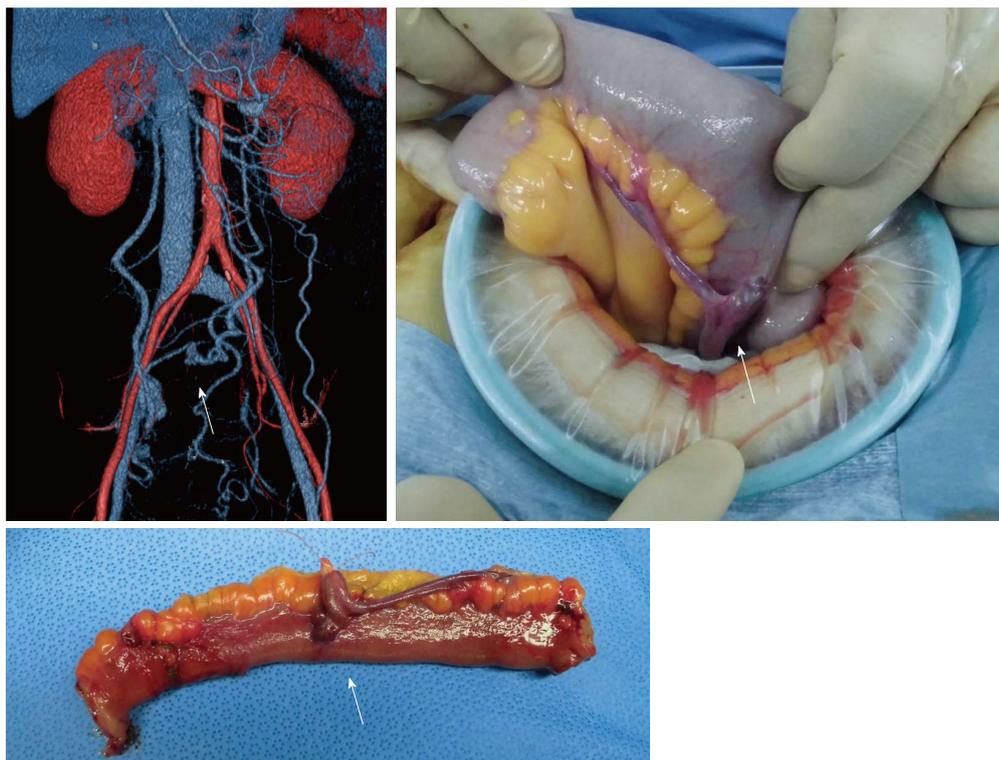


Figure 1 Ileal varices (arrow) were detected using multi-detector row CT and angiography and were resected by laparotomy.

are rare and are classified as ectopic gastrointestinal varices. Ectopic gastrointestinal varices occur at sites such as the duodenum, small intestine, colon, rectum, peristomal, biliary, peritoneal, umbilical, and other locations. Various related factors of an ectopic gastrointestinal varix such as portal hypertension due to cirrhosis, portal vein thrombosis, a history of abdominal surgery, chronic intraperitoneal inflammation, and hematochezia have been reported<sup>[4,5]</sup>.

An ileal varix is the major type of ectopic gastrointestinal varix. In a review 169 cases of ectopic gastrointestinal variceal bleeding, 17% was the highest rate of bleeding among all sites and was derived from jejunal and ileal varices<sup>[2]</sup>. Ileal varices are associated with a history of abdominal surgery and adhesions<sup>[6]</sup>. Presumably, abdominal surgery and intraperitoneal inflammation may cause adhesion of the intestinal tract. Then, collateral vessels within the adhesion may give rise to ectopic intestinal varices, particularly, in the jejunum and ileum<sup>[7]</sup>. Ectopic ileal varices most commonly flow into systemic circulation through the gonadal veins and less commonly through branches of the internal iliac veins<sup>[7]</sup>. In our case, there were various compatible features such as autoimmune portal hypertension and previous surgeries for appendicitis and hematochezia. Moreover, a portosystemic shunt, which flowed from the superior mesenteric vein into the right ovarian vein, was detected.

From the PubMed database including our own, we reviewed 21 consecutive cases of ileal variceal bleeding diagnosed from 1982 to 2017. The clinical features of

21 patients are shown in Table 1. Patients with ileal variceal bleeding consisted of 5 male and 16 female patients with a median age of 57 years (range 33-80 years). From the medical history, 71.4% (15/21) of patients were associated with portal hypertension due to liver cirrhosis. Previous abdominal surgery was noted in 57.1% (12/21) of patients. Regarding the diagnosis, 61.9% (13/21) of patients were diagnosed by SMA angiography. Capsule endoscopy was used in two cases. However, recent cases were mainly diagnosed by MDCT or MDCT and angiography and treated by surgical resection with no further bleeding. Surgical resection was performed in 76.1% (16/21) of all patients. Some recent patients underwent interventional radiology (IVR) treatment methods such as transjugular intrahepatic portosystemic shunt (TIPS)<sup>[8-10]</sup> and balloon-occluded retrograde transvenous obliteration (BRTO)<sup>[11,12]</sup>.

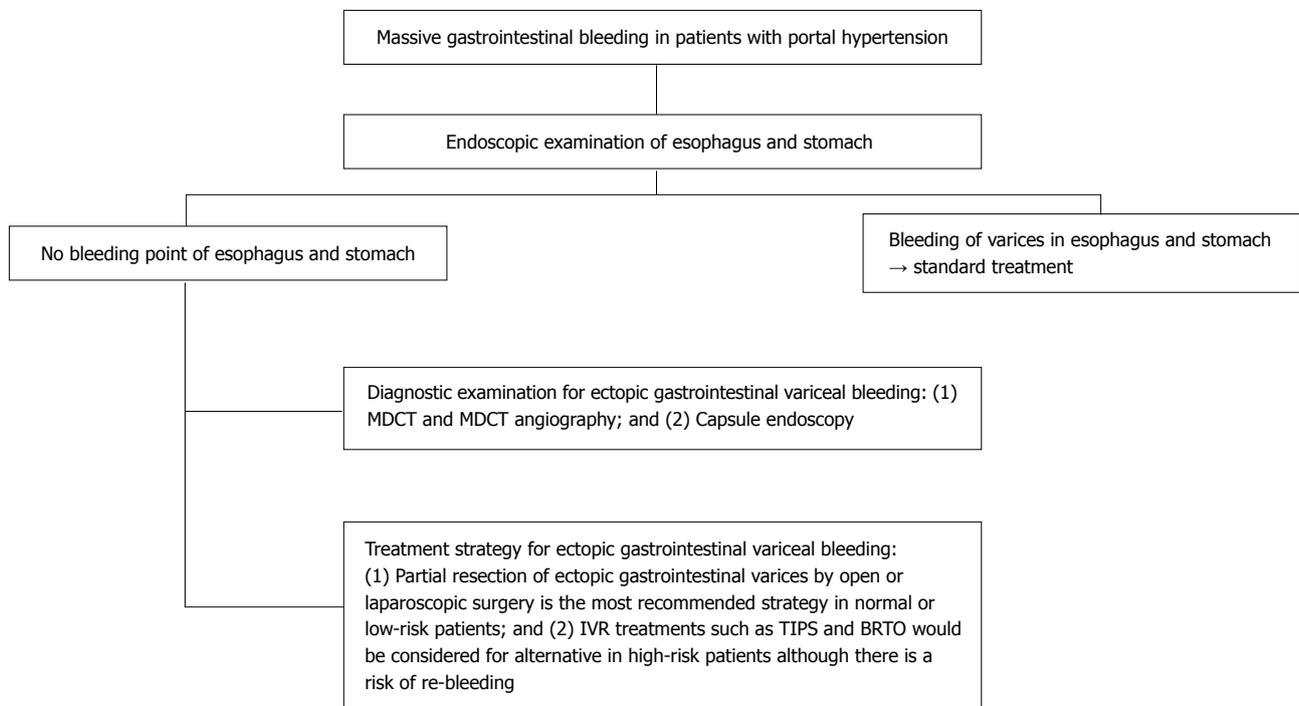
There were no patients with re-bleeding in previous reports of ileal variceal bleeding. However, re-bleeding rates of 23%-39% have been reported in TIPS and 5%-16.6% in BRTO in all reports of ectopic gastrointestinal variceal bleeding<sup>[13-16]</sup>. Although non-invasive treatment such as IVR may be desirable for ectopic gastrointestinal variceal bleeding in high-risk patients with co-morbidities, surgical resection of an affected intestine is currently a safe and effective treatment strategy to avoid further re-bleeding. Moreover, laparoscopic surgical resection of an affected intestine could be possible effective strategy as a minimally invasive procedure (Figure 2).

Ectopic gastrointestinal varices bleeding, especially ileal variceal bleeding, in patients with portal hypertension

**Table 1 Summary of the reported ileal variceal bleeding**

Case	Year	Age	Sex	Past history	Previous abdominal surgery	Diagnosis	Treatment	Outcome	
1	1982	Falchuk	52	F	liver cirrhosis	Cholecystectomy	SMA angiography	Partial enterectomy	Dead
2	1984	Shimada	49	M	liver cirrhosis	Ruputured esophageal varix	SMA angiography	Partial enterectomy	Alive
3	1986	Hojhus	80	F	Periappendicular abscess	(-)	SMA angiography	Partial enterectomy	Dead
4	1986	Arst	56	F	Liver cirrhosis	(-)	Laparotomy	Ileocelectomy	Dead
5	1990	Lewis	72	F	Liver cirrhosis	Hysterectomy	SMA angiography	Ileocelectomy	Alive
6	1994	Kurihara	43	M	(-)	(-)	SMA angiography	Partial enterectomy	Alive
7	1997	Ahn	54	M	Liver cirrhosis	(-)	SMA angiography	Ileocelectomy	Dead
8	1999	Ohtani	66	F	Liver cirrhosis	Ectopic pregnancy	SMA angiography	Partial enterectomy	Alive
9	2001	Kobayashi	62	F	Hepatocellular carcinoma	Hysterectomy	SMA angiography	Ligation of ileocecal and ovarian vein	Alive
10	2006	Ueda	72	F	Liver cirrhosis	Abdominal aortic aneurysm	MDCT	Partial enterectomy	Alive
11	2007	Lopez	56	F	Liver cirrhosis	Pelvic surgery	SMA angiography	TIPS	Alive
12	2007	Mashimo	33	F	Liver cirrhosis	Endometriosis	SMA angiography	Partial enterectomy	Alive
13	2009	Suzuki	74	F	Liver cirrhosis	Acute appendicitis	MDCT	Partial enterectomy	Alive
14	2009	Traina	58	F	Liver cirrhosis	(-)	ES	Sclerotherapy + TIPS	Alive
15	2009	Sato	55	M	Liver cirrhosis	Laparotomy for colonic tumor	Retrograde transvenous venography	BRTO	Alive
16	2010	Konishi	54	F	(-)	(-)	CE	Partial enterectomy	Alive
17	2011	Ambiru	62	F	Liver cirrhosis	Ectopic pregnancy	MDCT	Partial enterectomy	Alive
18	2011	Castagna	70	M	Liver cirrhosis	(-)	CE	TIPS	Alive
19	2013	Vamaddevan	48	F	Liver cirrhosis	(-)	MDCT	TIPS	Alive
20	2015	Garcia	74	F	Venous thromboembolism	(-)	MDCT	Partial enterectomy	Alive
21	2017	Our case	57	F	Portal hypertension	Acute appendicitis	MDCT	Partial enterectomy	Alive

CE: Capsule endoscopy; MDCT: Multi-detector raw computed tomography; ES: Enteroscopy; TIPS: Transjugular intrahepatic portosystemic shunt; BRTO: Balloon occluded retrograde transvenous obliteration.



**Figure 2 The management algorithm for massive gastrointestinal bleeding in patients with portal hypertension.** MDCT: Multi-detector raw computed tomography; TIPS: Transjugular intrahepatic portosystemic shunt; BRTO: Baloon occluded retrograde transvenous obliteration.

might be considered as a differential diagnosis if upper or lower endoscopy cannot detect a bleeding point such as in the esophagus or stomach. MDCT or MDCT

angiography is useful for the rapid examination and surgical resection of an affected ileum and is a safe and effective treatment strategy to avoid further bleeding.

## ARTICLE HIGHLIGHTS

### Case characteristics

A 57-year-old Asian woman with autoimmune portal hypertension due to polymyositis was admitted to our hospital with a 2-d history of hematochezia. She had a history of esophageal variceal rupture, which had been treated by endoscopy 3 years before.

### Clinical diagnosis

On the 3<sup>rd</sup> day after admission, she had massive hematochezia. The authors performed a second multi-detector row CT (MDCT) and angiography and diagnosed as massive ileal varix bleeding because there was a massive coagula at the distal ileal lumen of the ileal varix.

### Differential diagnosis

There was no differential diagnosis because upper and lower endoscopic examinations could not detect the responsible lesion.

### Laboratory diagnosis

Laboratory diagnosis was a severe anemia with hemoglobin 7.3 g/dL and hematocrit 23.4% because other data showed no apparent disorder.

### Imaging diagnosis

Imaging diagnosis by MDCT and its angiography was massive ileal varix bleeding because there was a massive coagula at the distal ileal lumen of the ileal varix.

### Pathological diagnosis

Pathological diagnosis was the leal varix.

### Treatment

The authors performed emergent segmental ileal resection, which included the ileal varix, via a small laparotomy. The varix was located at the 20-cm proximal portion of the ileocecal valve.

### Related reports

Jejunal varices as a cause of massive gastrointestinal bleeding. *Am J Gastroenterol* 1992; **87**: 514-517.

### Term explanation

The authors used common terms, which were used in previous reports.

### Experiences and lessons

Ectopic gastrointestinal variceal bleeding might be considered as a differential diagnosis if upper or lower endoscopy could not detect bleeding point. From the review results of previous reports including our case, MDCT and its angiography is a rapid and useful examination. Moreover, surgical resection of responsible lesion is safe and effective treatment strategy to avoid further bleeding.

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