

Treatment of ectopic varices with portal hypertension

Takahiro Sato

Takahiro Sato, Department of Gastroenterology, Sapporo Kosei General Hospital, Sapporo 060-0033, Japan

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Correspondence to: Takahiro Sato, MD, Department of Gastroenterology, Sapporo Kosei General Hospital, Kita 3 Higashi 8, Chuo-ku, Sapporo 060-0033, Japan. taka.sato@ja-hokkaidoukouseiren.or.jp
Telephone: +81-11-2615331
Fax: +81-11-2616040

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Abstract

Ectopic varices are unusual with portal hypertension and can involve any site along the digestive tract outside the gastroesophageal region. Hemorrhage from ectopic varices generally are massive and life threatening. Diagnosis of ectopic varices is difficult and subsequent treatment is also difficult; the optimal treatment has not been established. Recently, interventional radiology and endoscopic treatments have been carried out successfully for hemorrhage from ectopic varices.

Key words: Portal hypertension; Endoscopic injection sclerotherapy; Balloon-occluded retrograde transvenous obliteration; Ectopic varices; Endoscopic band ligation; Percutaneous transhepatic obliteration; Transjugular intrahepatic portosystemic shunts

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Core tip: Ectopic varices with portal hypertension are considered to be the cause of hemorrhage presenting as lower gastrointestinal bleeding. Recently, interventional radiology and endoscopic procedures have been performed successfully as a treatment option for ectopic varices.

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INTRODUCTION

Portal hypertension results from an increase in the resistance of blood flow in the intrahepatic portal vein and can cause the reopening of embryonic channels^[1]. Portal venous pressure is critical in the liver function of cirrhosis and esophageal varices are the most common complication of cirrhosis. Ectopic varices are portosystemic collaterals along the digestive tract outside the gastroesophageal region and are unusual^[2,3]. Endoscopic procedures such as endoscopic injection sclerotherapy (EIS) and endoscopic band ligation (EBL) have been carried out for treating esophageal varices^[4,5]. Balloon-occluded retrograde transvenous obliteration (B-RTO) is an angiographic technique for fundic varices of stomach^[6]. But, the optimal procedure has not been defined for hemorrhage from ectopic varices.

Here, we review the options in the treatment of

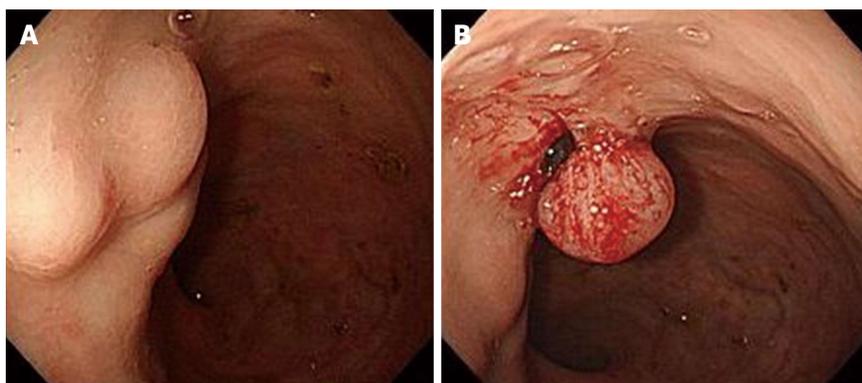


Figure 1 Endoscopy showed enlarged and tortuous, erosion positive rectal varices in a cirrhotic patient with anal bleeding (A) and endoscopic band ligation was performed successfully (B).

hemorrhaging ectopic varices.

ECTOPIC VARICES

The majority of portosystemic collaterals with portal hypertension are located in the esophagus and stomach and, especially, esophageal varices are the most common site. Ectopic variceal bleeding is massive and serious condition. Other sites of ectopic varices are the duodenum, small intestine, colon, rectum, and the peritoneum^[7]. A current survey of ectopic varices in Japan has been reported and the most frequent sites of ectopic varices are the rectum in 44.5%, followed by the duodenum in 32.9%^[8].

Rectal varices

Rectal varices are a consequence of portosystemic collaterals from the superior rectal veins to the middle inferior rectal veins. Endoscopy can detect the discrete dilated submucosal varices in the rectum and some investigators have reported that rectal varices occur at a high frequency^[9-11]. Hemorrhage from rectal varices occurs at a low frequency from 0.5% to 3.6%^[12-14].

Several medical procedures have been performed for controlling rectal variceal bleeding, however, a standard treatment has not been established.

Surgical procedures such as portosystemic shunting, ligation, and under-running suturing, also have been reported^[9]. Interventional radiologic techniques, including transjugular intrahepatic portosystemic shunts (TIPS), have been performed successfully for bleeding rectal varices^[15-17].

As an endoscopic treatment, Wang *et al*^[18] used EIS for rectal variceal bleeding. EBL has been performed as an effective procedure for rectal varices^[19-21] and Levine *et al*^[19] used EBL successfully for the rectal varices remaining after EIS. EBL is a safe and effective treatment for rectal varices. However, a retrospective study comparing EIS and EBL concluded that recurrence rate tended to be greater with EBL^[22] and stated that 5% ethanolamine oleate of the sclerosant should be carefully injected using fluoroscopy, avoiding injection

into the systemic circulation. Sato *et al*^[22] evaluated the efficacy of EIS for rectal varices and reported that EIS was a useful and safe treatment for rectal varices with regard to effectiveness and complication^[23].

Case presentation

EBL for rectal varices: Endoscopic finding of liver cirrhosis with anal bleeding revealed enlarged and tortuous, erosion positive rectal varices (Figure 1A), and endoscopic band ligation was performed successfully (Figure 1B).

EIS for rectal varices: EIS was performed using a flexible gastrointestinal endoscope, with a transparent hood attached to the tip, and the injection needle was placed into the varices (Figure 2A). Fluoroscopic observation with infusion of 5% ethanolamine oleate with iopamidol was made to determine the extent of the varices (Figure 2B).

Duodenal varices

Hemorrhage from duodenal varices is low frequent, however, it is often serious condition^[24]. The hemodynamics of duodenal varices involved the development of collateral veins between the portal vein trunk or superior mesenteric vein and the inferior vena cava^[25]. Hemorrhage from duodenal varices is generally massive and fatal^[25,26]. The diagnosis of duodenal varices is done ordinary by endoscopic examination, however, it is often difficult to observe hemorrhaging duodenal varices. The common site of duodenal varices is the duodenal bulb^[26], ranked next the second portion of the duodenum^[27]. In the United States and Europe, varices of the duodenal bulb occur most frequently because of extrahepatic portal obstruction, causing portal hypertension. On the other hand, in Japan, common site of duodenal varices is the second portion of the duodenum. In addition, hemorrhage from duodenal varices in the distal third portion is very rare^[28,29].

Recently, interventional radiology and endoscopic treatments have been performed successfully for duodenal varices. EBL is useful for obtaining hemostasis^[30,31],

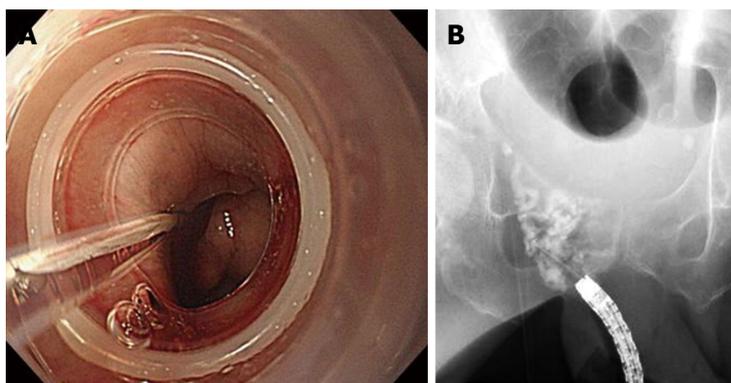


Figure 2 Endoscopic injection sclerotherapy was performed using a transparent hood attached to the tip and injection needle placed into the varices (A) and fluoroscopic observation with infusion of 5% ethanolamine oleate with iopamidol was made to determine the extent of the varices (B).



Figure 3 Endoscopy revealed tortuous duodenal varices in the anterior wall of the second portion (A) and N-butyl-2-cyanoacrylate was injected with contrast medium into the duodenal varices on fluoroscopic observation (B).

however, re-bleeding of varices is a weak point with EBL. EIS has been performed successfully for the treatment of duodenal variceal bleeding^[32,33] but there were serious problems of re-bleeding of the varices^[34,35]. N-butyl-2-cyanoacrylate is a tissue glue monomer; upon contact with blood, it immediately polymerizes and solidifies, resulting in hemostasis of variceal bleeding. Endoscopic therapy using N-butyl-2-cyanoacrylate is also very useful for massive duodenal variceal bleeding^[29,30] because of the high blood velocity and blood flow. Interventional radiologic techniques such as TIPS, B-RTO, and percutaneous transhepatic obliteration (PTO) are options for hemorrhaging duodenal varices and successful treatments have been reported, including TIPS^[26] and B-RTO^[35-37]. B-RTO is able to obliterate the afferent and efferent veins and may be considered as a treatment option for duodenal varices. PTO also has been used successfully in some cases^[38,39].

Case presentation

Endoscopic treatment using cyanoacrylate for duodenal varices: Endoscopy of liver cirrhosis with massive melena revealed tortuous duodenal varices in the anterior wall of the second portion (Figure 3A). N-butyl-2-cyanoacrylate was injected with contrast medium into the duodenal varices on fluoroscopic

observation (Figure 3B).

Small intestinal varices

Bleeding from jejunal and ileal varices may be massive and serious and it is difficult to achieve early diagnosis. Most cases are detected following intra-abdominal surgery. The development of collateral circulation *via* the post-operative adhesions is a risk factor of small intestinal varices in patients with portal hypertension.

Many literatures of hemorrhaging jejunal^[40-45] and ileal varices^[46-54] have been described. Bleeding small intestinal varices are as follows: portal hypertension, hematochezia without hematemesis, and previous abdominal surgery^[55]. Several treatments are available for jejunal varices, such as surgery^[40], portal venous stenting^[42,44,45] and percutaneous embolization^[40,43], and surgery has been used successfully for bleeding ileal varices^[50,51,56,57]. In ileal variceal patients with a serious condition, angiographic techniques including TIPS have been used successfully as a non-surgical approach^[3,52,54]. B-RTO is also practical for treating ileal varices^[58,59] and it also may be used for patients in a serious condition.

CONCLUSION

Ectopic varices in patients with portal hypertension are

considered to be the cause of hemorrhage presenting with lower gastrointestinal bleeding, and recently, their frequency has been increasing.

The diagnosis of ectopic varices and subsequent treatment are difficult. Recently, interventional radiology and endoscopic treatments have been performed successfully for ectopic varices. More investigations are necessary in a large number of patients.

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