Management of rectal varices in portal hypertension

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Abstract

Rectal varices are portosystemic collaterals that form as a complication of portal hypertension, their prevalence has been reported as high as 94% in patients with extrahepatic portal vein obstruction. The diagnosis is typically based on lower endoscopy (colonoscopy or sigmoidoscopy). However, endoscopic ultrasonography has been shown to be superior to endoscopy in diagnosing rectal varices. Color Doppler ultrasonography is a better method because it allows the calculation of the velocity of blood flow in the varices and can be used to predict the bleeding risk in the varices. Although rare, bleeding from rectal varices can be life threatening. The management of patients with rectal variceal bleeding is not well established. It is important to ensure hemodynamic stability with blood transfusion and to correct any coagulopathy prior to treating the bleeding varices. Endoscopic injection sclerotherapy has been reported to be more effective in the management of active bleeding from rectal varices with less rebleeding rate as compared to endoscopic band ligation. Transjugular intrahepatic portosystemic shunt alone or in combination with embolization is another method used successfully in control of bleeding. Balloon-occluded retrograde transvenous obliteration is an emerging procedure for management of gastric varices that has also been successfully used to treat bleeding rectal varices. Surgical procedures including suture ligation and porto-caval shunts are considered when other methods have failed.

Keywords: Rectal varices, Portal hypertension, Liver cirrhosis, Colonoscopy, Gastrointestinal bleeding

Core tip: Rectal varices complicate portal hypertension. Although rare, bleeding from rectal varices can be life threatening. There are no established guidelines for the treatment of rectal varices. In this article, the authors review endoscopic, radiological, and surgical techniques which have been suggested to be effective in the management of bleeding rectal varices.

INTRODUCTION
In 1954, Cabot et al[1] discussed the first reported case of bleeding rectal varices. Ganguly et al[2] defined rectal varices as dilated veins that originate more than 4 cm above the anal verge, clearly distinct from hemorrhoids, and not contiguous with the anal columns and/or pectinate line. The incidence of rectal varices in cirrhotic and non-cirrhotic patient varies in different reports and ranges between 38% and 94%[3,4]. Their prevalence in patients with cirrhosis is between 38% and 56%, whereas their prevalence in extrahepatic portal vein obstruction was reported between 63% and 94%[3,5]. Despite the high prevalence of rectal varices, clinically significant bleeding is rare and occurs in 0.5%-5% of patients[6].

While the management of esophageal varices has been well established[7], the optimal treatment of rectal varices remains to be determined. Endoscopic therapies, Transjugular Intrahepatic Portosystemic Shunt placement (TIPS), balloon-occluded retrograde transvenous obliteration (BRTO), and surgical management are some of the therapeutic options for management of rectal varices. The purpose of this article is to provide an updated review of current management of rectal varices.

**PATHOGENESIS**

Rectal varices are collaterals between the portal and systemic circulations that manifest as a dilation of the submucosal veins and constitute a pathway for portal venous flow between the superior rectal veins which branch from the inferior mesenteric system and the middle inferior rectal veins from the iliac system[8].

The normal hepatic venous pressure gradient (HVPG) ranges between 1 and 5 mmHg, becomes clinically significant when it reaches 10 mmHg and varices usually develop when the value of HVPG increases to at least 12 mmHg[9,10]. In the western hemisphere, sinusoidal portal hypertension secondary to liver cirrhosis is the most common cause of portal hypertension. There is a direct correlation between the progression of cirrhosis reflected by the Child Pugh or MELD scores and the degree of hyperdynamic circulation[11,12]. Hosking et al[13] studied 100 patients with cirrhosis and reported that the overall prevalence of rectal varices was 44%, this prevalence increased with the degree of portal hypertension. The authors described rectal varices in 19% of patients with cirrhosis without esophageal varices, 39% in patients with esophageal varices without history of bleeding, and 59% in patients with esophageal varices and history of bleeding. In this study, hemorrhoids occurred independently of the presence of rectal varices and 30% of patients had rectal varices and coexistent hemorrhoids[13].

There have been conflicting reports regarding the occurrence of rectal varices after obliteration of esophageal varices. However, a large study conducted in Japan by Watanabe et al[14] reported that 95% of patients with rectal varices had a history of esophageal varices and 87% of these patients had previously undergone endoscopic variceal obliteration for esophageal varices. The mechanism of rectal varices after treatment of esophageal or gastric varices is thought to be the result of obliteration of supplying vessels such as the left gastric, posterior gastric and short gastric veins leading to development of collateral vessels of the inferior mesenteric venous system and thus the formation of rectal varices. In this nationally representative study in Japan, the most frequent afferent vessel to the rectal varices was the inferior mesenteric vein, followed by the superior rectal vein and the efferent vessels included the internal iliac vein and the inferior rectal vein[14].

**DIAGNOSIS**

**Endoscopy**

Endoscopy is the main method for diagnosing rectal varices. They are visualized as blue tinted submucosal elevations located near the anus[15]. Rectal varices may be confused with internal...
hemorrhoids because of their location. However, hemorrhoids are not related to portal hypertension. Hemorrhoids result from a displacement of the anal cushions and hyperperfusion of the arteriovenous plexus vascular cushions without direct communication with any of the major branches of the portal venous system[16].

According to the general rules for recording endoscopic findings of esophago-gastric varices prepared by the Japanese Research Committee on Portal Hypertension, all codes for esophageal varices are used to describe ectopic varices including rectal varices[15]. Varices are classified into four groups according to their shapes and sizes. When there are no varices (F0), small and straight (F1), enlarged and tortuous (F2) and large and coil-shaped (F3). The color (C) of the varices is classified as either white (Cw) or blue (Cb). The dilated, small vessels or telangiectasia on the variceal surface is referred to as the red color sign (RC) which endoscopically indicates a high risk of bleeding. RC signs are graded as 0, 1, 2 or 3 according to their density and distribution. RC0 refers to no RC sign, RC1 to only a few RC signs, RC2 to several RC signs and RC3 to many RC signs. The bleeding signs as well as the mucosal findings can also be evaluated and described by endoscopy[15] (Table 1).

<table>
<thead>
<tr>
<th>Form (F)</th>
<th>F0: No varicose appearance</th>
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<tbody>
<tr>
<td></td>
<td>F1: Straight, small-caliber varices</td>
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<tr>
<td></td>
<td>F2: Moderately enlarged, beady varices</td>
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<tr>
<td></td>
<td>F3: Markedly enlarged, nodular or tumor-shaped varices</td>
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<tr>
<td>Color (C)</td>
<td>Cw: White varices</td>
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<tr>
<td></td>
<td>Cb: Blue varices</td>
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<tr>
<td></td>
<td>Cw-Th: Thrombosed white varices</td>
</tr>
<tr>
<td></td>
<td>Cb-Th: Thrombosed blue varices</td>
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<tr>
<td>Red color signs (RC)</td>
<td>RWM: Red wale markings</td>
</tr>
<tr>
<td></td>
<td>CRS: Cherry red spots</td>
</tr>
<tr>
<td></td>
<td>HCS: Hematocystic spots</td>
</tr>
<tr>
<td></td>
<td>RC(-): Absent</td>
</tr>
<tr>
<td></td>
<td>RC(+): Small in number and localized</td>
</tr>
<tr>
<td></td>
<td>RC(++): Intermediate between (+) and (+++)</td>
</tr>
<tr>
<td></td>
<td>RC(++)+: Large in number and circumferential</td>
</tr>
<tr>
<td></td>
<td>Te: Telangiectasia</td>
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<tr>
<td>Bleeding signs</td>
<td>Gushing bleeding</td>
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<td></td>
<td>Spurting bleeding</td>
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<tr>
<td></td>
<td>Oozing bleeding</td>
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<tr>
<td></td>
<td>Red plug</td>
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<td></td>
<td>White plug</td>
</tr>
<tr>
<td>Mucosal findings</td>
<td>E: Erosion</td>
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</table>
Endoscopic ultrasound

Conventional endoscopic ultrasound (EUS) reveals rectal varices as rounded, oval, or longitudinal echo free structures in the submucosa and also shows perirectal collateral veins outside the rectal wall. EUS can detect deep rectal varices in a large proportion of patients who do not have identified varices on routine endoscopy[17]. Dhiman et al[18] showed that EUS is better than endoscopy in detecting rectal varices (85% vs 45%) and in determining their number. Sato et al[19] demonstrated that intramural rectal varices, perirectal collateral veins, and the communicating veins between intramural rectal varices and perirectal collateral veins could be observed clearly with an ultrasonic microprobe. They also showed that the mean velocity of blood flow in rectal varices in the patients with rectal bleeding was significantly higher than in those cases experiencing no bleeding which indicates that the color doppler ultrasonography may be helpful in identifying high-risk group for rectal variceal rupture via the measurement of velocity[20]. Endoscopic color Doppler ultrasonography is better equipped than conventional EUS to evaluate the hemodynamics of varices, it can detect rectal varices through color flow images, calculate the velocity of blood flow in rectal varices for an effective and safe endoscopic variceal management[21].

TREATMENT

Medical management

The management of bleeding rectal varices essentially includes prompt resuscitation and correction of coagulopathy. The intravascular volume repletion is done with crystalloids and packed red blood cells. The Asian Pacific Association for the Study of the Liver recommends to maintain systolic blood pressure between 90-100 mmhg, and the heart rate below 100 beats/min[22]. The goal of blood transfusion is a hemoglobin level approximately 8 g/dL (hematocrit of 24)[7]. A short course of prophylactic antibiotic therapy should be administered to improve survival and decrease the risk of spontaneous bacterial peritonitis in all patients presenting with cirrhosis and gastrointestinal bleed including rectal bleeding[23]. There are no randomized control trials to recommend the use of vasoactive drugs such as vasopressin, terlipressin or octreotide in bleeding rectal varices. However, these drugs have a proven benefit in the management of bleeding gastro-oesophageal varices and hence can be considered for use in bleeding rectal varices[24].

Endoscopic management

Endoscopic injection sclerotherapy: Endoscopic injection sclerotherapy (EIS) was first reported to be useful for treatment of rectal bleeding in 1985[25]. Later on, other case reports of successful EIS for treatment of bleeding rectal varices were published[26-28]. Sato et al[29] performed EIS using 5% ethanolamine oleate with iopamidol, which was injected intermittently under fluoroscopy in 32 patients. The patients were successfully treated without serious complications. The authors suggested the necessity to evaluate the hemodynamics of the rectal varices before EIS to avoid severe complications such as pulmonary embolism. They also recommended injecting the sclerosant slowly under fluoroscopy. The recurrence rate in this series was 24% over the 1-year follow-up period.

Endoscopic band ligation: Endoscopic band ligation (EBL) has been well studied and its efficiency in treating bleeding esophageal varices is well known[30,31]. EBL has also been used in treatment of gastric varices. However, its efficacy in this regard is equivocal[32,33]. In 1996, Kojima et al[34] used EBL in the management of bleeding rectal varices. Subsequently, Uno et al[35] reported a successful
use of EBL to treat bleeding rectal varices after failure of sclerotherapy in a child with extrahepatic portal hypertension. Long term follow up of 46 mo after successful use of EBL in treatment of bleeding and obliteration of rectal varices as the initial therapy in an adult patient was reported by Firoozi et al [36] EBL is a safe and effective therapy for rectal varices, however the risk of recurrence is high [34,37]. Sato et al[38] compared EIS to EBL in the management of rectal varices. EIS appeared to be superior to EBL with regard to effectiveness. The recurrence rate was less with EIS 33.3% vs 55.6% with EBL. No complications were noted with EIS, however one patient who received EBL developed bleeding ulcer[38].

**Cyanoacrylate injection:** Cyanoacrylate glue is an accepted therapeutic method for gastric varices, although its use is off-label in the United States[39]. It was first described by Soehendra et al[40]. This glue preparations work by immediate polymerization upon contact with blood, causing vascular obstruction and is eventually extruded into the gastric lumen, typically about 1 mo after injection [41,42]. Weilert et al[43] reported a case of rectal varix managed successfully with EUS-guided cyanoacrylate injection and embolization coils. The use of coils is believed to provide a scaffold to retain glue within the varix, thereby minimizing the risk of embolization and allowing for a decreased volume of glue injection for variceal obliteration[43]. Color Doppler-EUS has been used to diagnose submucosal endoscopically invident rectal varices bleeding and to manage it by histoacryl glue injection[44]. The most serious adverse events of glue injection therapy is systemic embolization and sepsis which has been reported secondary to embolized glue acting as a septic focus[45]. Embolization into the arterial circulation (via a patent foramen ovale or arteriovenous pulmonary shunt) can result in stroke and multiorgan infarction[45].

**Interventional radiology**

**TIPS:** TIPS is a minimally invasive and effective method used for management of rectal varices during active bleeding. It can serve both as a bridge to transplantation and as the definitive therapy in patients who are not good candidates for surgery[24]. TIPS was first used in 1993 by Katz et al[46] in a patient with repeated bleeding from anorectal varices (ARV) with marked decompression of the varices 24 h after placement of the TIPS. The patient had no recurrent bleeding after 6 mo of follow up. Several case reports and small case series of bleeding ARV successfully managed with TIPS have been described in the literature[47-55]. Kochar et al[56] reported in 2008 the largest series of patients (n = 28) with bleeding ectopic varices, 12 of them were rectal varices treated by TIPS placement. Hemostasis was effectively achieved in 67% of the patients. This was achieved solely with TIPS without concomitant embolization in 21 of the 22 (95%) patients and in three of the five (60%) patients who had TIPS and concomitant variceal embolization. Rebleeding from ectopic varices occurred in five (21%) patients. In two (40%) patients, the rebleeding was secondary to shunt dysfunction and responded to revision of the shunt. However, rebleeding occurred in three patients despite a functioning shunt with low portal pressure gradients[56].

**Embolization:** Embolization is a procedure performed by interventional radiologist to occlude the feeding vein to the rectal varices. It can either be performed alone or in combination with band ligation or TIPS[53,57]. When used alone, embolization results in high 1 year rebleeding rates[44]. The combination of TIPS and embolization has been described as efficient in the prevention of recurrent bleeding from esophagogastric varices[58]. After embolization, the communication of the portal vein and the rectal veins remains partially interrupted even after shunt stenosis. Hence, the increase of the pressure in the portal vein is not directly transmitted into the rectal plexus[53]. Ahn et al[59] reported recently a case of recurrent bleeding after successful TIPS treated with variceal embolization. Various embolization materials are used, including coils, gelfoam, thrombin, collagen, autologous blood clot and ethanol[60,61].
BRTO: Developed by Kanagawa et al[62] in the early 1990s, the BRTO procedure is an endovascular technique that causes occlusion of outflow portosystemic shunt, such as a gastrorenal shunt, using an occlusion balloon followed by the endovascular injection of a sclerosing agent directly into the gastrovariceal system[57,62]. For the past two decades, this procedure has become common practice in Asia for the management of gastric varices. It is now becoming more popular in the United States. It has been shown to be effective in controlling gastric variceal bleeding with low rebleeding rates. BRTO has many advantages over TIPS. It is less invasive and can be performed on patients with poor hepatic reserve and those with encephalopathy[63]. Anan et al[64] reported a case of successful treatment of colonic varices by means of BRTO in a patient with hepatic encephalopathy leading to resolution of the encephalopathy and worsening of preexisting esophageal varices. This reflects postprocedural increased portal hypertension. A more recent article reported the success of BRTO as an additional therapy to surgical suture in controlling bleeding rectal varices with 1.26 cm feeding vessel. However, the patient died 6 mo later from liver failure[65].

Surgical management

Surgery has been used for treatment of rectal varices mainly when endoscopic management has failed. Surgical methods include simple suture ligation, inferior mesenteric vein occlusion and porto-caval shunt surgery. The later has been shown to be effective in controlling life threatening bleeding. However, the majority of patients presenting with bleeding rectal varices have a poor general condition and are not good candidate for these major surgical procedures[66]. The mortality in these patients is high and is mainly secondary to liver failure. Bittinger et al[66] reported 80% mortality within 2 mo despite adequate local treatment of the rectal varices.

Direct suture ligation is a technically challenging option and often not successful. However, the stapled approach seems to be a suitable alternative. Stapled procedure for the control of bleeding varices was first reported in 2002 by Botterill et al[67]. The authors reported a circumferential stapling device was used to successfully control bleeding ano-rectal varices after failure of injection sclerotherapy and band ligation. In 2005, another case report also demonstrated that stapled procedure may be an effective means of bleeding control[68]. A case series of nine patients was published by Kaul et al[69] with successful control of bleeding following a circumferential stapled procedure. Four of the nine patients were previously treated with endoscopic therapy (three with banding and one with injection sclerotherapy). No further rebleeding was noted during the follow up period of 4 to 24 mo.

CONCLUSION

Bleeding rectal varices can be a life threatening condition in patients with portal hypertension and should be considered in the differential diagnosis of these patients when they present with lower gastrointestinal bleeding.

The management of rectal varices is multidisciplinary and involves gastroenterologists, interventional radiologists and surgeons. There are no established guidelines to define the appropriate management strategies for rectal varices. Published studies consist mainly of case reports and series. This article provides a review of the literature summarizing the different therapeutic options to manage rectal varices.

Footnotes

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