Ectopic varices in portal hypertension

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ABSTRACT

Varices most commonly occur and bleed in the gastro-esophageal region but ectopic varices can arise at extra-gastro-esophageal locations in the gastrointestinal tract like duodenum, jejunum, ileum, colon, rectum, biliary tree and at the site of a surgical ostomy. These varices pose diagnostic as well as therapeutic challenges during endoscopic procedures. Ectopic varices can also occur outside the gastrointestinal tract, giving rise to diagnostic difficulties on imaging and unusual hemorrhage. Although ectopic varices bleed less commonly than gastro-esophageal varices, they are difficult to diagnose and problematic to treat. The absence of stigmata of recent esophageal or gastric variceal bleeding and certain clues in the patient’s history and clinical presentation should raise the clinician’s suspicion of an extra-gastro-esophageal site of variceal bleeding. Patients with extrahepatic causes of portal hypertension, cirrhotic patients with a prior history of gastrointestinal surgery and patients who present with profound bleeding but without hematemesis should in particular be evaluated further if an obvious site of gastro-esophageal variceal bleeding is not observed at UGI endoscopy. Accurate preoperative diagnosis is often difficult in patients bleeding from ectopic varices from extra-gastro-esophageal sites, rebleeding is certain even in those patients who respond to medical treatment, and the optimal surgical decision making protocol has not yet been evolved due to the low prevalence of these ectopic varices. Nevertheless, an appreciation and awareness of these unusual causes of gastrointestinal bleeding, allied with prompt and appropriate diagnosis can lead to successful endoscopic, radiological or surgical management of ectopic variceal bleeding.

Key words: Cirrhosis, ectopic varices, gastrointestinal bleeding, portal hypertension

How to cite this article:

DUODENAL VARICES

Duodenal varices occur in about 0.4% in all patients with portal hypertension and account for one third of bleeding episodes from ectopic varices. Early detection is important, as duodenal varices are a potential source of massive hemorrhage. At upper gastrointestinal endoscopy, an uninitiated observer may misinterpret bleeding from duodenal varices as that from duodenal ulcer. These should be considered in all patients with duodenal tumoral lesions and suspected portal hypertension. In this context, duodenal biopsy can be dangerous and should be avoided. A diminution in the volume of the duodenal varices with inspiratory movements may help in the differential diagnosis during endoscopy.
The duodenal bulb is the most common site of duodenal varices, the second portion of the duodenum appears to be the next most common site but duodenal varices in the other portions are rare. [4] Hashizume et al. studied these varices angiographically and histopathologically; and found that the duodenal varix consisted of a single vessel with afferent and efferent vessels, forming a portosystemic shunt in the retroperitoneum. The varix traversed the duodenum and was present in the submucosal layer of the posterior wall; while the afferent vessel was the superior or inferior pancreaticoduodenal vein originating in the portal vein trunk or superior mesenteric vein, and the efferent vein drained into the inferior vena cava. [2] They have also been reported at the site of previous duodenal operations and the resultant adhesions and after endoscopic sclerotherapy. [5,6] Duodenal varices are more common in patients having extrahepatic portal vein obstruction and in those with thrombosed portosystemic shunts. [7,8]

Apart from endoscopy, hypotonic duodenography, ultrasonography, computed tomography, venous phase of superior mesenteric angiography, and percutaneous transhepatic portography have been used to diagnose duodenal varices. [4]

Medical therapies, including vasopressin and octreotide may have limited success in controlling active duodenal variceal bleeding. [9] Endoscopic sclerotherapy or endoscopic variceal ligations are the main treatment modalities. [10,11] Embolization and transjugular intrahepatic portosystemic shunt are the therapeutic alternatives, if endoscopic sclerotherapy or variceal ligation fails to control the bleeding. [12,13]

When conservative measures cannot control the hemorrhage, emergency laparotomy may be indicated. Duodenal varix suture ligation or resection results in a high rate of rebleeding. [14] End-to-side portacaval shunt may be effective. [9] An arteriovenous fistula requires resection of the paramural varix and surgical occlusion. [14] In view of the difficulty during the duodenal mobilization and the precarious condition of patient, it is not surprising that the operative mortality is high.

**JEJUNAL AND ILEAL VARICES**

A triad of portal hypertension (generally due to liver cirrhosis), history of abdominal surgery, and hematemesis without hematemesis characterizes small intestinal varices. [16] Bleeding from varices may present with vesical varices and gross hematuria if an intestinal segment is used for an augmentation cystoplasty. [17]

A history of abdominal surgery appears to predispose the development of ectopic varices (portosystemic communication) in adhesions. Possible physiological origins of this entity were studied in Edward’s demonstration of network of fine communication between the parietal surface of the viscera and the posterior abdominal wall, arising in the embryo due to the juxtaposition of the developing systemic and visceral venous plexus. [18] Formation of collaterals, de novo, is unlikely if the anatomy is undisturbed. In some cases no cause can be found. Histological examination demonstrates a massive varicose vein and several dilated veins in the submucosa. [19]

Although rare, bleeding from small bowel varices is associated with a high mortality as accurate preoperative diagnosis is often difficult. Detection of these varices has been a challenging task and several invasive diagnostic techniques such as enteroclysis, TC-99m RBC studies, venous phase of mesenteric arteriography, enteroscopy, color flow Doppler ultrasound and magnetic resonance angiography have been used for this purpose. [16,20-23] Intraoperative Sonde enteroscopy is safe and effective, providing complete visualization of the small-bowel mucosa without enterotomy while avoiding the trauma that can be caused by push endoscopy. It is the diagnostic assessment of choice. [24] Medical therapy, including vasopressin infusion via the superior mesenteric artery, is often useful in controlling acute variceal bleeding. [25] Percutaneous transhepatic embolization and transcatheter intrahepatic portosystemic shunt are the therapeutic alternatives. [26,27] Surgical treatment consists of lysis of adhesions and bowel resection combined with portosystemic shunt, under the presumption that the portal pressure in these patients has been partially decompressed through these spontaneous shunts and may increase significantly after their surgical division. [28] Patients with excellent hepatic reserve survive and have no further gastrointestinal bleeding. [29]

**COLONIC VARICES**

Colonic variceal bleeding is a rarity and is most commonly due to portal hypertension, with local mesenteric vein obstruction constituting a rare cause. The true prevalence of colonic varices is not known, but Feldman et al. found an incidence of 0.07% in autopsy material. [30] Esophageal varices were present in approximately half of the group with colonic varices. [13] Bleeding has been reported to occur in 2.5% of patients attending sclerotherapy sessions for esophageal varices. [32] In patients with portal hypertension the coronary azygous system was the primary portosystemic channel in at least half of the cases, but in a quarter of cases it was the inferior mesenteric-internal iliac system. [31] Possible etiologies of this condition may be esophageal transection and devascularization and extensive thrombosis of the portal vein resulting in obliteration of the coronary-azygous anastomotic system. In
such a situation, other potential sites of portal-systemic anastomoses, such as that in the colon, may open, leading to development of colonic varices.\[33\] Idiopathic/primary, familial, secondary to splenic vein thrombosis and adhesion-related colonic varices without portal hypertension have also been reported.

Varices of the colon are usually segmental, involving predominantly (66%) the distribution of inferior mesenteric vein and less frequently (26%) the distribution of superior mesenteric area, and never confined to transverse colon.\[31\] Diffuse variceal involvement of the colon is uncommon and implies an unknown cause.

In case of colonic varices the differential diagnosis should include portal hypertension with chronic liver disease, portal vein thrombosis, vascular anomalies or postoperative complications. If this entity is not considered, a rectal or colonic biopsy may lead to brisk and dangerous bleeding. Apparent similarity of radiological and endoscopic appearance of varices to polyps, misdiagnosis and inappropriate biopsy remain the potential pitfalls. Colonoscopist visualizes these varices as serpiginous to nodular, often bluish submucous lesions. They are often missed on colonoscopy due to collapse of varices during periods of hypotension or because of increase in the intraluminal pressure due to air insufflation during the endoscopic examination.\[31,34\] Sensitivity of colonoscopy is greatly reduced during periods of active bleeding and in the absence of good bowel preparation.

In cases where the cause of lower GI bleeding is not clear, even after colonoscopy; venous phase of mesenteric angiogram and scintigraphic studies may be useful.\[31,35\] If doubt persists, intraoperative colonoscopy may be useful to pinpoint the problem.\[36\] Conservative therapy consists of vasopressin and somatostatin analogue, which may be useful in the control of bleeding.\[37\] Sclerotherapy using a colonoscope and transjugular intrahepatic portosystemic shunt are other therapeutic alternatives.\[58\]

The choice of surgical therapy in portal hypertension is portal decompression and not colonic resection; as colectomy is associated with significantly greater mortality due to risk of infection and considerable technical difficulty of this surgery in the presence of portal hypertension.\[31,34\]

### ANORECTAL VARICES

Anorectal varices are a rare cause of rectal bleeding and are often erroneously diagnosed as bleeding hemorrhoids. Although rare, rectum is the most common site of lower gastrointestinal varices.\[39\] Rectal varices occur due to high pressure in the inferior mesenteric venous system in patients with portal hypertension. Bleeding from them is uncommon, and often mild and self-limiting, but rarely it can be fatal. It is equally important to be aware of the presence of rectal varices in case rectal biopsy is needed in patients with portal hypertension.

The reported incidence of rectal varices ranges from 40 to 89.3%.\[39,45\] No correlation has been found between the presence of anorectal varices and the Child's grade of cirrhosis, intrahepatic V/s extrahepatic causes of portal venous obstruction, the grade of esophageal varices, the presence of gastric varices, portal hypertensive gastropathy, or whether or not patients received sclerotherapy.\[39,40,43-45\]

Identifying the source of lower gastrointestinal hemorrhage in patients with chronic liver disease and portal hypertension can be challenging but the differential diagnosis between hemorrhoids and anorectal varices has been elucidated in many studies.\[40,46\] It has also been documented that the prevalence of hemorrhoids is not increased in patients with portal hypertension and their presence is unrelated to the degree of portal hypertension.\[40,44\] A careful examination is essential to prevent misdiagnosis and inappropriate and inadvertent treatment like surgical excision of varices in mistake for hemorrhoids, with disastrous results.\[40\] Anorectoscopy is the initial investigation of choice. Rectal endoscopic ultrasonography, transvaginal sonography and magnetic resonance imaging are useful in detecting the presence and number of rectal varices.\[47-49\]

The principal emergency treatment is endoscopic sclerotherapy or endoscopic ligation, failing which surgical ligation should be performed.\[50-52\] Before the advent of transjugular intrahepatic portosystemic shunt (current choice of treatment), a portosystemic shunt, preferably between the inferior mesenteric vein and the vena cava or renal vein, was the treatment of choice.\[53\] Transjugular embolization of the inferior mesenteric vein is an alternative to TIPS, where TIPS is not feasible.\[54,55\]

### STOMAL VARICES

Variceal bleeding from enterostomy is an unusual complication of portal hypertension and represents a cause of recurrent or intractable gastrointestinal bleeding. Presence of caput medusae/varices developing around a stoma may herald the presence of mild to moderate portal hypertension before other signs of hepatic decompensation are evident. Once variceal communications have been formed between the portal venous system of the gut and subcutaneous systemic circulation, heavy bleeding from dilated venous plexus may occur spontaneously or from microtrauma. In a review,
the average interval found was 48 months for ileostomies, 38 months for ileal conduits and 23 months for patients with a colostomy.\(^{[56]}\)

Proper diagnosis requires careful inspection of the muco-cutaneous region of the stoma for venous bleeding sites and endoscopy examination of the stoma to rule out the presence of recurrent bowel disease or other lesions like arteriovenous malformations, polyp or Crohn’s disease.\(^{[57]}\)

The emergent treatment of bleeding of the colostomy must combine several methods, quite often consecutively: local compression, ligation, and sclerotherapy.\(^{[58]}\) Palliative local measures, like suture ligature or sclerotherapy, however, remain the treatment of choice in the high-risk, cirrhotic patient who is unlikely to survive a major operation and may increase the interval between bleeding episodes and decrease the severity of bleeding.\(^{[57,58]}\) The hemorrhage can be managed temporarily in most patients with local measures. Once bleeding is controlled, the treatment must be primarily medical (hygienic and dietary habits, b-adrenergic blocking agents), but complementary surgery is invariably necessary because of recurrence of bleeding.

There is no consensus on which of the various surgical options is best, but by and large, the type of further surgical treatment is determined by the severity of the underlying liver disease and the patient’s life expectancy.\(^{[56]}\) Mucocutaneous disconnection (MCD) is simple, quick, repeatable and associated with a lower morbidity and intraoperative blood loss than stomal relocation. In the select group of patients that cannot be managed conservatively, MCD is favored and relocation considered only if MCD is technically impossible i.e. improperly placed stoma, symptomatic peristomal hernias and those with poor appliance fit.\(^{[60]}\) It should be kept in mind that repeated use of local operative procedures leads to the formation of scar tissue and causes problems in the care of the stoma. Although stomal manipulation is the most commonly performed procedure, portosystemic shunting has the lowest incidence of both rebleeding and need for additional procedures and provides the longest mean postoperative survival and is the choice in patients who are good surgical candidates.\(^{[56]}\) In particular, the absence of postoperative encephalopathy in the ileostomy group may be attributed to the absence of colon, the major source of bacteria generated nitrogenous products. Transjugular intrahepatic portosystemic shunt and stomal varices embolization are effective alternatives in case of recurrent bleeding of stomal varices.\(^{[61]}\)

The overall prognosis mainly depends on the function of the liver, the deterioration of which is accelerated by the successive hemorrhagic accidents. Particular attention should be paid to stoma care and the prevention of trauma from appliances.

### BILIARY VARICES

Gallbladder varices are often seen in portal hypertension, more often in extra hepatic portal vein obstruction patients.\(^{[62,63]}\) Gallbladder varices do not correlate with size of esophageal varices, number of sessions of sclerotherapy, presence or absence of gastric varices, portal gastropathy, Child Pugh grade or splenorenal shunt placement.\(^{[63]}\) These collaterals cause some gallbladder stasis but do not impede gallbladder function and hence seem unlikely to contribute to gallstone formation.\(^{[62]}\) Their clinical significance is their propensity to bleed during biliary surgery; thus, the operating surgeon should be aware of them. The color flow Doppler is the gold standard procedure for the diagnosis, although angiography, computerized tomography and magnetic resonance have also been reported.\(^{[64]}\)

Bile duct varices are seen more frequently in left hepatic duct, possibly due to the joining of umbilical vein to the left branch of portal vein adjacent to the left hepatic vein.\(^{[65]}\) The resultant filling defect in the ERCP has to be differentiated from sclerosing cholangitis and malignancy.\(^{[65,66]}\) Due to their propensity to bleed, balloon dilatation is probably best avoided in these patients and placement of pigtail biliary endoprostheses is preferred over straight stents with side flaps.\(^{[67]}\) Usually biliary varices are found incidentally during imaging, but their presence calls for a search for portal vein thrombosis. Rarely they can give rise to obstructive jaundice or haemobilia.

### INTRAPERITONEAL HEMORRHAGE FROM ECTOPICT VARICES

Intraperitoneal hemorrhage from ectopic varices is a rare occurrence. In cirrhotic patients, sudden onset of abdominal pain in combination with hypotension and falling hematocrit in the absence of external blood loss should result in ultrasonography of the abdomen. The main differential diagnosis is acute pancreatitis. Any free fluid present should be aspirated and when blood is encountered the patient must be operated upon immediately.\(^{[68]}\) Spontaneous hemorrhage from anterior abdominal wall varices has also been documented into the rectus abdominus muscle and peritoneal cavity.\(^{[69]}\) Exploratory laparotomy and suture ligation of the bleeding varix seems to give the greatest likelihood of survival. Angiography with special attention to the venous phase may demonstrate the varices, in addition, vasopressin infusion in the superior mesenteric artery can be tried, which may permit stabilizing the patient before surgery.\(^{[70]}\) Patient’s remaining liver function and the ability to withstand surgery determine
the ultimate prognosis.

**CUTANEOUS VARICEAL BLEEDING**

In portal hypertension, three types of cutaneous portosystemic collaterals may develop the ‘classical’ Caput Medusae, enterostomal varices and scar or adhesion-related abdominal collaterals.[71] Very few cases have been documented of a varicose umbilical vein with external hemorrhage significant enough to cause hemodynamic instability.[72,73] Coagulopathy and hemorrhagic shock, ending in a fatality may complicate the clinical course.[72] Local measures (direct pressure, suture ligation and sclerotherapy) and medical therapy should be applied early in the resuscitation of the patient. Once stable, definitive treatment has to be instituted otherwise rebleeding is a certainty. Transjugular intrahepatic portosystemic shunt, umbilical vein embolization and mesocaval shunt surgery have all shown good results, with stoppage of bleeding and disappearance of cutaneous varices.[72,74,75]

**MISCELLANEOUS**

Upper esophageal varices occur infrequently and may rarely cause massive upper gastrointestinal hemorrhage.[76] This case serves to stress the importance of a through examination of the cervical portion of the esophagus during routine endoscopy. Varices of the gastric antrum are seen in a small proportion of patients and are distributed equally amongst the etiologies of portal hypertension.[77] They rarely bleed and may be ignored during sclerotherapy of esophageal varices, however, if required, sclerotherapy is the treatment of choice.[78,79] Rarely, idiopathic varices have been reported throughout the gastrointestinal tract.[80] Significant varices can occur outside the gastrointestinal tract and have been described in kidney, lungs, tracheobronchial tree, mediastinum and vagina; giving rise to unusual hemorrhage as well as diagnostic difficulties on imaging.[81-86]

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ANNOUNCEMENT

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