Case report

Isolated ectopic jejunal varices in a patient with extrahepatic portal vein obstruction – A case report

Sanket Solanki *, Suvendu Sekhar Jena, Sri Aurobindo Prasad Das, Amitabh Yadav, Naimish N. Mehta, Samiran Nundy

Department of Surgical Gastroenterology and Liver Transplantation, Sir Ganga Ram Hospital, New Delhi, Delhi 110060, India

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ABSTRACT

Introduction and importance: Among the various causes for lower gastrointestinal bleeding, ectopic varices constitute a small chunk. Though rare, these can pose a diagnostic challenge with recurrent bleed leading to multiple admission and blood transfusions.

Case presentation: A 41-year-old male presented to our department with multiple episodes of melena. On further evaluation with CT angiography, a diagnosis of extrahepatic portal vein obstruction with moderate splenomegaly and ectopic jejunal varix was made. He underwent splenectomy with resection of involved jejunal segment with side to side anastomosis.

Clinical discussion: The diagnosis of ectopic varices remains elusive in a large number of cases in view of the varied etiology. Various newer endoscopic and imaging modalities can play a diagnostic as well as therapeutic role but this also further complicates the management as there is a lack of substantial guidelines directing the treatment protocol. As a result, we have to resort to a case by case approach for the optimal management in these cases.

Conclusion: The main modality of management for bleeding ectopic varices is percutaneous or endoscopic. Surgery is reserved for refractory cases, with decompressive shunts combined with segmental resection of involved intestine being at the forefront of surgical options.

1. Introduction

Ectopic varices represent an unusual cause of lower GI bleeding and are defined as the porto-systemic collaterals that are present anywhere in the abdomen, except in the gastro-oesophageal region. They are usually rare and account for about 5% of all variceal bleeds. The most common site of ectopic varices is duodenum (17%), followed by jejunum and ileum (17%), colon (14%), peritoneum (9%), rectum (8%) and rarely other sites like the ovary, vagina or retroperitoneum [1]. Most common etiology for these varices is portal hypertension with others being congenital anomalies, previous abdominal operations, vascular thrombosis and certain familial syndromes. The triad of portal hypertension, prior history of abdominal surgery and haematochezia without haematemesis can characterize the source to be intestinal varices. Various diagnostic modalities have been applied, including upper and lower GI endoscopies, computed tomography (CT) scans, angiographies, and recently capsule endoscopy. The management includes a multi-disciplinary stepwise approach including endoscopic or interventional radiological procedures, and surgery. Here we report a rare case of isolated ectopic jejunal varix in a young patient with portal hypertension secondary to extrahepatic portal vein obstruction (EHPVO).

2. Case report

A 41-year-old male, without any comorbidity, presented with complaints of diffuse abdominal pain, which had gradually increased over the period of a week. It was associated with dark stools with increased frequency (up to 4 times a week) and non-bilious vomiting. There was no history of fever, jaundice, abdominal distension, weight loss, constipation or any bleeding disorders in the family. He had a similar episode in the past for which he had undergone upper GI (UGI) endoscopy, which didn’t reveal any oesophageal varices. On clinical examination, the patient was haemodynamically stable with mild pallor and palpable splenomegaly. On laboratory investigations, his haemoglobin...
was 6.5 g/dL with negative viral markers and normal coagulation studies. Repeat UGI endoscopy revealed no varices. CT angiography (Fig. 1) showed a normal liver echotexture with chronic occlusion of the superior mesenteric and portal veins with multiple collaterals in the peripancreatic region, hepatoduodenal ligament and the porta, with ectopic varices in the distal jejunal loops. Capsule endoscopy showed varices in the proximal small bowel with altered blood presentation distally. A diagnosis of EHPVO with lower GI bleed from the ectopic jejunal varices was made and he was planned for a splenectomy with proximal splenorenal shunt. Intra-operatively, the liver was normal with moderate splenomegaly with multiple peri-portal collaterals. Jejunal varices were present about 20 cm from the duodeno-jejunal junction spreading along a length of 20 cm (Fig. 2). In view of the unhealthy and friable splenic vein which was not suitable for shunting, splenectomy along with segmental resection of the jejunum bearing varices and ligation of the feeding vessel, followed by a side to side jejunal anastomosis in 2 layers was done. The post-operative course was uneventful and he was discharged on post-operative day 9. He is doing well in subsequent follow up without any further bleeding episodes.

3. Discussion

Lower GI bleeding primarily affects people of 65 years of age or more, with an annual incidence in the third decade being 1/100,000 and reaching 200/100,000 by the ninth decade [2].

The etiology of overt lower GI bleeding can usually be diagnosed with colonoscopy and traditional imaging, but the causes of obscure bleeding (with normal upper and lower GI endoscopy) can be difficult. Ectopic varices are an unusual cause of such bleeding and account for 5% of cases. Duodenal varices account for most ectopic varices which bleed, but jejunoileal varices are the most common in occurrence as shown by Saad et al. [3] Liver cirrhosis is the most common cause of ectopic varices, followed by EHPVO, idiopathic portal hypertension and causes like liver metastasis, biliary stricture and biliary atresia among others [4]. Intrabdominal adhesions from previous abdominal surgeries also predispose to collaterals at unusual places leading to ectopic varices. Extra hepatic portal vein thrombosis is a common cause of ectopic varices as was seen in our case. In our case, the patient presented with lower GI bleed with negative endoscopic study and was found to have EHPVO with ectopic jejunal varices on CT angiography and capsule study.

The clinical presentation varies according to the site of the varices, with the most common presentation being anaemia. They may also present with melaena as was seen in our case. The presence of ectopic varices should also be strongly considered in patients with known liver disease or with stigmata of portal hypertension, who have obscure blood loss where upper and lower gastrointestinal (GI) endoscopic studies fail to identify the site of bleed.

Various diagnostic modalities have been utilized for these patients. The duodenal and rectal varices are usually identified with upper and lower GI endoscopies respectively. But the main diagnostic dilemma is with jeuno-ileal varices. In the setting of an acute or subacute haemorrhage, CT angiography [5] is the preferred choice which can promptly locate the source of bleeding. MR angiography can also enhance the unusual sites of portosystemic collaterals but has a diminishing role in the acute or subacute setting. Double balloon enteroscopy or push enteroscopy is useful in selected cases with high suspicion of small intestinal bleeding, where it can play a diagnostic as well as a therapeutic role. Capsule endoscopy [6] can aid the diagnosis in obscure cases but has a limited role in the acute setting. Also, Technetium Tc-99m red blood cell scintigraphy has been explored for slow bleeding but its role for massive bleeds is diminishing. Conventional angiography has been at the forefront for its dual role as a diagnostic and therapeutic modality and also being the least invasive of the options.

The most challenging aspect is the management of these ectopic varices due to the paucity of guidelines for the management due to rarity of the disease. The management varies with each case based on the site of haemorrhage, haemodynamic stability, local resources, and underlying cause. The majority of the ectopic varices are amenable to endoscopic treatment with band ligation and injection sclerotherapy. Among the interventional radiological procedures, direct percutaneous access to the varices has proven to be the optimal approach. Transjugular Intrahepatic Porto-Systemic Shunt (TIPSS) can be used for oesophageal varices, but it is often found that selective decompression of ectopic varices does not occur at the same pressure as that of oesophageal varices [7]. This is owing to the fact that ectopic varices decompress via pathways other than the left gastric-azygous vein. This reiterates the need for definitive embolization at the times of TIPSS creation. A novel retrograde approach to shunt occlusion is balloon occluded retrograde transvenous obliteration (BRTO) which has been successfully used in cases of bleeding gastric varices, but the results with ectopic jeuno-ileal varices are not as promising [8]. Percutaneous embolization using the transhepatic method has been one of the most widely used methods with success rates reaching up to 80% [9]. However, it is associated with a high rebleeding rate. Also, a transjugular approach can be applied where decompression is not required. In cases with extrahepatic PV obstruction, correction of stenoses or thromboses with stents may improve the outcomes.

The role of surgery in the modern-day management of bleeding ectopic varices is limited due to the morbidity and mortality that accompanies the liver disease in these patients. It is often the salvage option offered after exhaustion of endoscopic and interventional radiological options. Surgical management aims to decrease the portal pressure by the creation of a porto-systemic (which can be selective or

Fig. 1. Shows the transverse sections of contrast enhanced CT of the abdomen, with (a) showing the dilated jejunal varices (white arrow) and (b) showing the obstructed extrahepatic portal vein with cavernoma formation (white arrow).
non-selective) or porto-portal shunt, with or without an associated splenectomy [10]. This can be accompanied by regional specific devascularization depending on the presence of varices.

Decompressive shunts are the best option to prevent rebleeding, with the distal splenorenal shunt being a commonly performed procedure for cases with ectopic varices where portal hypertension is well documented. Also, intraoperative endoscopy can help in selective enter-ectomy for bleeding jejuno-ileal varices [11,12,13]. The usual procedure is resection of the involved segment along with ligation of the feeding vessels accompanied by a decompressive procedure, as was planned in our case, but the friable and unhealthy splenic vein precluded the decompressive procedure and a splenectomy was done as splenectomy causes a decrease in the portal venous pressure of about 280 mmHg, with a pressure reduction of about 29% in patients with EHPVO as compared to 18% in patients with liver cirrhosis [14]. The decrease in portal pressure in patients with EHPVO following splenectomy was significantly higher than either those with liver cirrhosis (p < 0.025) or Idiopathic Portal Hypertension (p < 0.005). An extensive literature search revealed a few such cases of isolated jejunal varices, with varying etiology, which had been treated with a multitude of options (Table 1).

Thus, the lack of standard guidelines for the ideal management of ectopic varices has led to a case by case approach with often endoscopic and interventional radiological procedures being foremost. Regardless of the method used, complete obliteration with embolization of the target varix should be sought, as focal embolization of a feeder will eventually lead to rebleeding with development of newer collaterals. As seen in our case, a splenectomy with concomitant resection of involved intestinal segment with ligation of feeding vessels is also a viable treatment option.

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Table 1

| Author            | Year | Etiology                                      | Intervention                                      |
|-------------------|------|-----------------------------------------------|--------------------------------------------------|
| Joo et al. [11]   | 2000 | Superior mesenteric vein thrombosis with liver cirrhosis | Segmental intestinal resection                  |
| Hiraoka et al. [15]| 2001 | Extrarectal portal vein stenosis post pancreato-duodenectomy | Percutaneous transhepatic balloon dilatation and stent placement |
| Sato et al. [16]  | 2003 | Extrarectal portal vein thrombosis post radical cholecystectomy for carcinoma gall bladder | 1. Segmental jejunal resection 2. Intraoperative ethanol embolization of the varices |
| Deshpande et al. [12]| 2008 | Isolated jejunal varices without portal hypertension | Jejunal resection                                |
| Koo et al. [9]    | 2012 | Decompensated liver cirrhosis                 | Percutaneous transhepatic coil embolization       |
| Lee et al. [17]   | 2013 | PV thrombosis with liver cirrhosis             | Percutaneous transsplenic embolization            |
| Kastanakis M et al. [13] | 2013 | Idiopathic jejunal varices without portal hypertension | Segmental jejunal resection                      |
| Mansoor et al. [10]| 2016 | EHPVO                                         | Side to side portocaval shunt                     |
| Kohli et al. [18] | 2017 | Portal vein thrombosis post deceased donor liver transplantation | Intraoperative enteroscopy with glue injection   |
| Heiberger et al. [19]| 2019 | Extrarectal portal vein stenosis post pancreato-duodenectomy | Transhepatic portal vein stenting                |

Fig. 2. (a) Shows intraoperative dilated jejunal varices on the jejunal segment (White arrow), (b) shows the trans-illuminated jejunal varices visible through the mesentery (White arrow) and (c) cut open specimen of the diseased segment of the jejunum that was resected, with visible opening of the feeder vessels into the jejunum (white arrow).

Author contribution

Sanket Solanki – Writing original draft, methodology, Formal analysis.
Suvendu Sekhar Jena – Writing original draft, methodology, formal analysis.
Aurobindo Prasad Das – Writing (Reviewing and editing), methodology, Supervision.
Amitabh Yadav – Supervision, Conceptualization, Resources.
Naimish Mehta – Supervision, Conceptualization, Resources.
Samiran Nundy – Writing (Reviewing and editing), Supervision, Conceptualization, Resources.

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Consent
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NA.

Declaration of competing interest
None.

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