



Case report

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

Sanket Solanki^{*}, Suwendu 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

ARTICLE INFO

Keywords:

EHPVO
Ectopic varices
Splenectomy
Decompressive surgery

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

^{*} Corresponding author at: Sir Ganga Ram Hospital, New Delhi, Delhi 110060, India.

E-mail address: sanketsolanki54@gmail.com (S. Solanki).

<https://doi.org/10.1016/j.ijscr.2021.106299>

Received 28 June 2021; Received in revised form 6 August 2021; Accepted 7 August 2021

Available online 10 August 2021

2210-2612/© 2021 The Authors. Published by Elsevier Ltd on behalf of IJS Publishing Group Ltd. This is an open access article under the CC BY-NC-ND license

(<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

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 present 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]. Intraabdominal 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 jejuno-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 jejuno-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

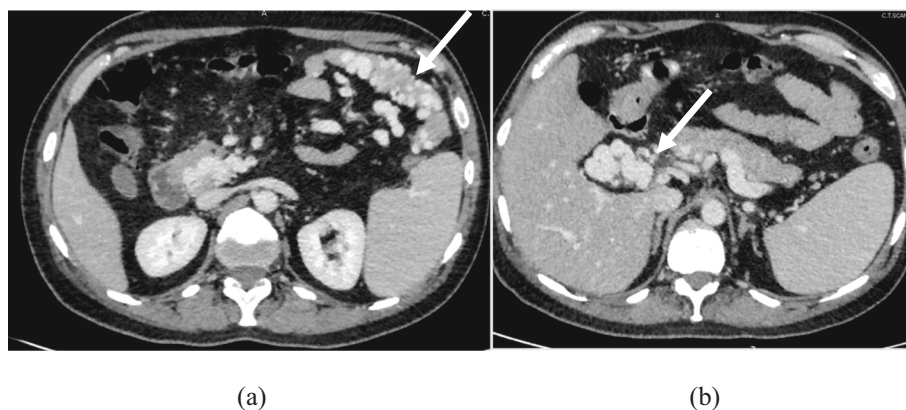


Fig. 1. Shows the transverse sections of contrast enhanced CT of the abdomen, with (a) showing the dilated jejunal varices (white arrow) and (b) showing the obstructed extrahepatic portal vein with cavernoma formation (white arrow).

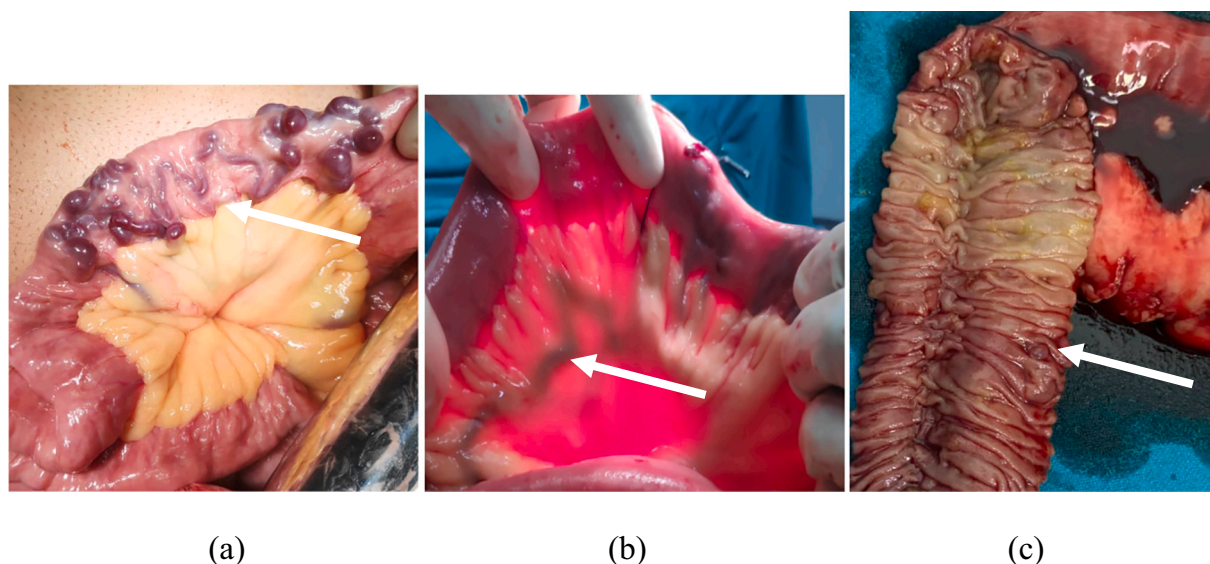


Fig. 2. (a) Shows intraoperative dilated jejunal varices on the jejunal segment (White arrow), (b) shows the trans-illuminated jejunal varices visible through the mesentery (White arrow) and (c) cut open specimen of the diseased segment of the jejunum that was resected, with visible opening of the feeder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

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 enterectomy 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 mmH₂O, 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.

Sources of funding

None.

Ethical approval

N/A.

Table 1

Literature review of ectopic varices with their etiology and management.

Author	Year	Etiology	Intervention
Joo et al. [11]	2000	Superior mesenteric vein thrombosis with liver cirrhosis	Segmental intestinal resection
Hiraoka et al. [15]	2001	Extrahepatic portal vein stenosis post pancreatoduodenectomy	Percutaneous transhepatic balloon dilatation and stent placement
Sato et al. [16]	2003	Extrahepatic 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 trans-splenic 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	Extrahepatic portal vein stenosis post pancreatoduodenectomy	Transhepatic portal vein stenting

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.

Guarantor

Sanket Solanki (Solanki S).

This paper has been reported in line with the SCARE 2020 criteria [20].

Provenance and peer review

Not commissioned, externally peer reviewed.

Ethical approval

None.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Registration of research studies

NA.

Declaration of competing interest

None.

References

- [1] A. Helmy, K. Al Kahtani, Fa.dda.M. Al, Updates in the pathogenesis, diagnosis and management of ectopic varices, *Hepatol. Int.* 2 (2008) 322–334.
- [2] L.L. Strate, Lower GI bleeding: epidemiology and diagnosis, *Gastroenterol. Clin. N. Am.* 34 (2005) 643–664. Dec.
- [3] W.E. Saad, A. Lippert, N.E. Saad, S. Caldwell, Ectopic varices: anatomical classification, hemodynamic classification, and hemodynamic-based management, *Tech. Vasc. Interv. Radiol.* 16 (2013) 108–125.
- [4] N. Watanabe, A. Toyonaga, S. Kojima, S. Takashimizu, K. Oho, S. Kokubu, K. Nakamura, A. Hasumi, N. Murashima, T. Tajiri, Current status of ectopic varices in Japan: results of a survey by the Japan Society for Portal Hypertension, *Hepatol. Res.* 40 (2010) 763–776.
- [5] D. Weishaupt, T. Pfammatter, P.R. Hilfiker, U. Wolfensberger, B. Marincek, Detecting bleeding duodenal varices with multislice helical CT, *AJR Am. J. Roentgenol.* 178 (2002) 399–401. Feb.
- [6] G.D. De Palma, M. Rega, S. Masone, F. Persico, S. Siciliano, F. Patrone, L. Matantuono, G. Persico, Mucosal abnormalities of the small bowel in patients with cirrhosis and portal hypertension: a capsule endoscopy study, *Gastrointest. Endosc.* 62 (2005) 529–534.
- [7] Z.J. Haskal, M. Scott, R.A. Rubin, C. Cope, Intestinal varices: treatment with the transjugular intrahepatic portosystemic shunt, *Radiology* 191 (1994) 183–187.
- [8] N. Hashimoto, T. Akahoshi, D. Yoshida, N. Kinjo, K. Konishi, H. Uehara, Y. Nagao, H. Kawanaka, M. Tomikawa, Y. Maehara, The efficacy of balloon-occluded retrograde transvenous obliteration on small intestinal variceal bleeding, *Surgery* 148 (2010) 145–150.
- [9] S.M. Koo, S.W. Jeong, J.Y. Jang, T.H. Lee, S.R. Jeon, H.G. Kim, J.O. Kim, Y.J. Kim, Jejunal variceal bleeding successfully treated with percutaneous coil embolization, *J. Korean Med. Sci.* 27 (2012) 321.
- [10] E. Mansoor, A. Singh, G. Nizialek, H.M. Veloso, J. Katz, G.S. Cooper, G. Isenberg, Massive gastrointestinal bleeding due to isolated jejunal varices in a patient with extrahepatic portal hypertension: a case report, *Am. J. Gastroenterol.* 111 (2016) 1209–1211.
- [11] Y.E. Joo, H.S. Kim, S.K. Choi, J.S. Rew, H.R. Kim, S.J. Kim, Massive gastrointestinal bleeding from jejunal varices, *J. Gastroenterol.* 35 (2000) 775–778.
- [12] A. Deshpande, P. Sampat, R. Bhargavan, M. Sharma, Bleeding isolated jejunal varices without portal hypertension, *ANZ J. Surg.* 78 (2008) 814–815.
- [13] M. Kastanakis, D. Anyfantakis, N. Katsougris, E. Bobolakis, Massive gastrointestinal bleeding due to isolated jejunal varices in a patient without portal hypertension, *Int. J. Surg. Case Rep.* 4 (2013) 439–441.
- [14] S. Matsubara, K. Ouchi, S. Matsuno, Portal venous pressure following splenectomy in patients with portal hypertension of differing etiology, *Eur. Surg. Res.* 24 (1992) 372–377.
- [15] K. Hiraoka, S. Kondo, Y. Ambo, S. Hirano, M. Omi, S. Okushiba, H. Katoh, Portal venous dilatation and stenting for bleeding jejunal varices: report of two cases, *Surg. Today* 31 (11) (2001) 1008–1011.
- [16] T. Sato, O. Yasui, T. Kurokawa, M. Hashimoto, Y. Asanuma, K. Koyama, Jejunal varix with extrahepatic portal obstruction treated by embolization using interventional radiology: report of a case, *Surg. Today* 33 (2003) 131–134.
- [17] J.Y. Lee, S.Y. Song, J. Kim, B.H. Koh, Y. Kim, W.K. Jeong, M.Y. Kim, Percutaneous transsplenic embolization of jejunal varices in a patient with liver cirrhosis: a case report, *Abdom. Imaging* 38 (2013) 52–55.
- [18] D.R. Kohli, M.F. Levy, G.B. Smallfield, Laparotomy-assisted endoscopic injection of jejunal varices for overt small bowel bleeding, *ACG Case Rep. J.* 21 (4) (2017), e79. Jun.
- [19] C.J. Heiberger, T.I. Mehta, D. Yim, Jejunal varices: an unconsidered cause of recurrent gastrointestinal haemorrhage, *BMJ Case Rep.* 7 (12) (2019), e228680. Mar.
- [20] for the SCARE Group, R.A. Agha, T. Franchi, C. Sohrabi, G. Mathew, The SCARE 2020 guideline: updating consensus Surgical CASE REport (SCARE) guidelines, *Int. J. Surg.* 84 (2020) 226–230.