Rectal varices due to chronic inferior mesenteric vein thrombosis caused by external compression in a large hiatus hernia containing the pancreas

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ABSTRACT

A 79-year-old woman presented with a large fresh rectal bleed. Computed tomography revealed that she had a large type IV hiatus hernia, which contained the stomach and pancreas. Compression of the inferior mesenteric vein and splenic vein had led to thrombosis within these vessels and retrograde flow within the inferior mesenteric vein. This had led to the formation of portosystemic rectal varices. Ectopic varices occasionally form in the rectum, often in the context of liver cirrhosis. At the time of writing, ours is the first reported case of portosystemic rectal varices formulated in response to obstruction of vessels within a hiatus hernia.

KEYWORDS

Hiatus hernia – Rectal varices – Portal vein obstruction

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Case history

A 79-year-old woman presented to the emergency department following a large fresh rectal bleed. She had not had any abdominal pain, weight loss or change in bowel habit prior to this incident. She was stabilised and underwent blood transfusion prior to a period of active observation. Computed tomography (CT) of her abdomen and pelvis revealed a large type IV hiatus hernia containing the majority of the stomach as well as the body and tail of the pancreas (Fig 1). CT also identified thrombosis of the splenic vein and the distal portion of the inferior mesenteric vein (IMV) at the level of the portal confluence. Marked prominent venous collaterals were also noted to arise from the iliac veins and inferior vena cava, which had led to the formation of rectal varices (Figs 1 and 2).

We present an unusual case of rectal bleeding secondary to portosystemic rectal varices. Chronic compression from a large hiatus hernia containing the pancreas had led to thrombosis at the portal confluence of the IMV and splenic vein. This had subsequently caused retrograde flow within the IMV and the formation of portosystemic varices within the rectum. Following consideration of available treatment options, as well as the patient's age and comorbidities, the decision was made to manage her case conservatively. The patient was discharged with clinic follow-up and advice to seek medical attention in the event of further bleeding.

Discussion

Hiatus hernias are classified according to the position of the gastro-oesophageal junction and the extent of stomach herniation. Type I, or 'sliding', hiatus hernias occur when the gastro-oesophageal junction herniates into the mediastinum, accounting for over 95% of all cases.¹ Type II or 'rolling' defects are pure paraoesophageal hernias, where the fundus of the stomach is herniated in the presence of a normally positioned gastro-oesophageal junction. Type III hiatus hernias are combination of the aforementioned, with displacement of both the gastro-oesophageal junction and stomach. Type IV hiatus hernias are rare, accounting for only 5–7% of all paraoesophageal hernias. They comprise herniation of the stomach alongside other abdominal viscera, most commonly small bowel or omentum.

The presenting features of paraoesophageal hernias can vary from symptoms typical of gastro-oesophageal reflux disease to more subtle symptoms of postprandial chest fullness and shortness of breath, while some patients remain asymptomatic. Although there is not complete consensus on how best to manage paraoesophageal hernias, current guidance would advocate surgical repair in symptomatic cases. In asymptomatic patients, surgery may not always be indicated, with careful consideration of age and comorbidities advised.¹

It is exceedingly rare for the pancreas to herniate into a paraoesophageal hernia, with a limited number of cases

Figure 1 Coronal reformatted computed tomography images of the abdomen and pelvis, posterior to anterior. a) Incompletely imaged hiatus hernia with organoaxial volvulus (H); prominent venous collaterals arising from inferior vena cava, iliac veins and inferior mesenteric veins (V). b) Occluded distal segment of inferior mesenteric vein at level of portal confluence, likely due to chronic thrombosis (arrow). More proximal inferior mesenteric vein (IMV). Prominent venous collaterals arising from inferior vena cava, iliac veins and inferior mesenteric veins (V). Herniated pancreatic body within hiatus hernia (P). c) Absent segment of splenic vein at level of portal confluence (*) which extends to hiatus hernia (H), likely due to chronic thrombosis. Main portal vein (PV). Prominent venous collaterals arising from inferior vena cava, iliac veins, and inferior mesenteric veins (V)

Figure 2 Axial reformatted computed tomography of the upper pelvis (a) and upper/mid pelvis (b). a) Prominent venous collaterals arising from inferior vena cava and iliac veins (V). Proximal inferior mesenteric vein (IMV). b) Prominent venous collaterals arising from inferior mesenteric vein (V)

previously reported.² However, the unique aspect of this case was the formation of portosystemic rectal varices, secondary to compression of the splenic vein and IMV by a large hiatus hernia containing the pancreas. At the time of writing, we were unable to find a similar case in the available literature.

Portosystemic varices form in response to backpressure from the tributaries of the portal vein. When blood flow is

obstructed due to distortion of the portal vasculature, as in liver cirrhosis, or due to external compression of local vessels, as in extra-hepatic portal vein obstruction (EHPVO), collateral pathways open up allowing blood to bypass. So-called portosystemic collateral veins are formed when collapsed embryonic channels reopen or when the direction of flow within existing adult vessels is reversed, allowing blood to flow down a gradient away from areas of

high pressure.⁵ Backpressure transmitted via portosystemic collateral veins can then lead to the engorgement of vessels within visceral walls and the formation of varices.

This phenomenon is most commonly seen in the context of portal hypertension due to liver cirrhosis, where oesophageal varices are detected in 50% of patients and gastric in 5–33%.⁴ However, ectopic varices may form in various locations across the gastrointestinal tract including the duodenum, small bowel, large bowel and rectum. Ectopic varices can be the product of portal hypertension or alternatively formulate in response to EHPVO.

Rectal varices can formulate as a result of cirrhosis or portal vein obstruction, as mentioned previously. However, rare aetiological mechanisms include mesenteric vein thrombosis and splenic vein thrombosis, as seen in our case. Thrombosis of these vessels leads to a reversal of normal hepatopetal flow, with blood instead moving from the inferior mesenteric vein into the superior rectal vein. From here, blood flows into the extrinsic rectal venous plexus, which lies outside the rectum, below the level of the peritoneal reflection. As backpressure builds, blood flows via perforating vessels, through the muscularis propria and into the intrinsic rectal venous plexus. The intrinsic rectal venous plexus is subdivided into a superior and inferior group of veins. The superior group lies in the rectal submucosa and it is engorgement of these vessels that leads to the formation of rectal varices. The inferior group of intrinsic rectal venous plexus vessels lies in the anal subcutaneous tissue, with backpressure here leading to the formation of external haemorrhoids.⁵ From the site of rectal varices, blood is able to drain into the inferior and middle rectal veins, which are both tributaries of the internal iliac vein. Thus, rectal varices represent portosystemic collaterals, allowing blood to flow

between the inferior mesenteric and iliac systems in the event of obstruction.

Although rare, anorectal variceal bleeds may be heavy, with life-threatening potential. In the event of massive haemorrhage, haemodynamic resuscitation and prophylactic antibiotic therapy prior to urgent endoscopic assessment is advised. Pneumatic tamponade with a Foley catheter or Sengstaken–Blakemore tube may be employed as a temporising measure. Endoscopic injection sclerotherapy should be considered as the first choice of treatment option, although band ligation and variceal obturation with either glue or thrombin are viable alternatives depending on operator expertise. In the event of refractory bleeding, consideration may be given to alternative therapies such as transjugular intrahepatic portosystemic shunt.⁵ Fortunately, in the reported case the bleeding resolved rapidly, without the need for operative management. The patient was discharged following blood transfusion, intravenous fluid therapy and a period of active observation.

References

1. Kohn GP, Price RR, Demeester SR *et al.* *Guidelines for the Management of Hiatal Hernia*. Los Angeles, CA: Society of American Gastrointestinal and Endoscopic Surgeons; 2013.
2. Coughlin M, Fanous M, Velanovich V. Herniated pancreatic body within a paraoesophageal hernia. *World J Gastrointest Surg* 2011; **3**(2): 29–30.
3. Sharma M, Rambeshbabu CS. Collateral pathways in portal hypertension. *J Clin Exp Hepatol* 2012; **2**: 338–352.
4. Arora A, Rajesh S, Meenakshi YS *et al.* Spectrum of hepatofugal collateral pathways in portal hypertension, an illustrated radiological review. *Insights Imaging* 2015; **6**(5): 559–572.
5. Robertson M, Thompson AI, Hayes PC. The management of bleeding from anorectal varices. *Curr Hepatol Rep* 2017; **16**(4): 406–415.