Unusual association of diseases/symptoms

Bleeding from peristomal varices: a complication of portal hypertension

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Summary

Peristomal varices are a recognised complication of stomas in the presence of portal hypertension. There has been a progression of treatment options described in the literature, including the transjugular intrahepatic portosystemic shunt (TIPS). The use of TIPS, a percutaneous procedure allowing connection between the portal and systemic circulations within the liver, is a well-recognised method of treating the complications of portal hypertension. This report presents a case of peristomal varices successfully treated with TIPS and subsequently reviews the literature relating to its management.

BACKGROUND

This case report serves as an aid to clinicians to highlight a relatively uncommon complication of stomas. Patients may suffer from recurrent haemorrhaging before a definitive treatment option is explored.

CASE PRESENTATION

A 66-year-old gentleman with an ileostomy located in the right iliac fossa for the preceding 5 years presented to the colorectal surgery outpatients' department with heavy bleeding from the mucocutaneous junction. The stoma-care specialist nurse had been using silver nitrate in an attempt to control the bleeding, but was of no benefit.

His previous medical history included ulcerative colitis, which was diagnosed 8 years previously, type 2 diabetes mellitus and hypertension. The patient's alcohol consumption was less than six units per week and had never been excessive.

The patient had initially undergone a total colectomy with ileorectal anastomosis for the ulcerative colitis because symptoms failed to respond to medical therapy. However, with the relapse of symptoms and the presence of residual disease in the rectal stump, the patient underwent refashioning of the ileostomy, with rectal excision, 3 years later.

The same year as the ileostomy, deranged liver function tests were observed and failed to improve despite the cessation of the therapeutic drug, mesalazine. Although primary sclerosing cholangitis was hypothesised as the cause, a liver biopsy demonstrated cirrhosis and moderate/severe steatohepatitis. The morphological features of primary sclerosing cholangitis were not evident and the biopsy was in keeping with non-alcoholic steatohepatitis.

Two years from the formation of the ileostomy, bleeding from the stoma was reported. Review in the outpatients department identified the patient as overweight (body mass index of 29 kg/m²), with a mid-line laparotomy scar and an ileostomy bag in the right iliac

fossa. A purple discolouration of the skin was evident around the stoma, but there was no organomegaly palpable and no caput medusae. The patient had not required any inpatient admissions, but nonetheless, the presence and persistence of symptoms warranted further investigation.

INVESTIGATIONS

Laboratory studies identified a thrombocytopenia ($90 \times 10^9/l$) and a prolonged prothrombin time of 17 s (international normalised ratio 1.4). Haemoglobin, white cell count and renal function tests were normal. The autoantibody screen, including anti-liver kidney microsomal type 1, antimitochondrial and antismooth muscle antibodies were negative. The viral hepatitis screen was also negative and the α -fetoprotein was normal.

Annual gastroscopies were performed once liver disease was identified to search for varices. The gastroscopies repeatedly identified oesophageal varices that were barely noticeable, but with an absent red sign and no active bleeding. There was also mild duodenitis with no evidence of active bleeding.

A CT of the abdomen and pelvis in arterial and portovenous phase was performed. The extent of abdominal varices including gastro-oesophageal, splenic and peristomal varices is demonstrated (figure 1a–c). The peristomal varices are extensive surrounding the stoma and correspond to the site of bleeding. Other features of portal hypertension such as splenomegaly are seen and complications such as ascites and portal vein thrombosis are excluded (figures 2 and 3).

TREATMENT

Upon discovery of the oesophageal varices, the patient was commenced on propranolol 20 mg twice daily. The patient underwent transjugular intrahepatic portosystemic shunt (TIPS) 4 months after presenting to the outpatients clinic.

Figure 1 (a) Sagittal reformatted images. Ileocolic varices surrounding the ileostomy site (white arrow). (b) Sagittal reformatted images. Varices surrounding the ileostomy site (white arrow). (c) Axial images. Varices surrounding the ileostomy site (white arrow).

OUTCOME AND FOLLOW-UP

A review, 1 month after TIPS, highlighted no evidence of significant deterioration in liver function and there have been no further reports of bleeding to date. At 10 months follow-up, there have been no reports of complications from our case and an ultrasound scan has demonstrated a patent TIPS with turbulent flow.

DISCUSSION

Peristomal varices are an unusual, but recognised complication of patients with ileostomies,¹ colostomies² and ileal conduits^{3–5} with coexisting portal hypertension. Primary sclerosing cholangitis in the presence of ulcerative colitis has been identified as the most frequent aetiology for patients with an ileostomy and portal hypertension.²

The varices develop at the vulnerable peristomal mucocutaneous junction, between the high-pressure portal system and the low-pressure systemic venous system.^{1 6 7} The period between gastrointestinal (GI) surgery and the development of stomal varices is dependant upon the severity of the underlying liver disease, but has been reported to range from 19 months to 24.5 years.⁸ Furthermore, time to subsequent bleeding has been reported to range from 1.5 to 29 years,⁹ each carrying an estimated mortality risk of 3%–4% per episode.¹⁰ This is in contrast to gastro-oesophageal variceal bleeding which has an associated

30%–40% mortality risk.⁷ However, it is deemed inevitable that all patients with peristomal varices develop major bleeding from the peristomal site.¹¹

Despite not being considered as life-threatening as gastro-oesophageal varices, deaths from peristomal variceal exsanguinations have been reported,¹² but the majority of deaths are secondary to liver failure or massive upper GI tract bleeding.⁷ The average survival time from initial haemorrhage from peristomal varices in conservatively treated patients is thought to be approximately 4 years.⁷ The significant morbidity during this period, such as the risk of recurrent bleeds and the development of chronic anaemia warrants appropriate early management of peristomal varices.

In 1968, Resnik *et al*¹³ were the first to describe peristomal varices in the literature when three patients suffered from stomal haemorrhaging after developing ileostomy varices. Since then, various management options have been described in the literature with varying outcomes. These have included, stoma revision,¹ injection sclerotherapy,¹⁴ liver transplantation,¹¹ embolisation,¹⁰ portosystemic shunt surgery^{1 16} and TIPS.^{17–20}

The varices are susceptible to bleeding from minor local trauma including general stoma care. Non-invasive treatment methods such as pressure dressings,¹¹ manually applied pressure¹⁶ and suture ligation^{7 8 12 21} have been

Figure 2 Axial image of upper abdomen with intravenous contrast. Splenomegaly and fatty infiltration to the liver.

Figure 3 Axial images of upper abdomen with intravenous contrast. Fatty infiltration of the liver and gastro-oesophageal varices (white arrow).

successful in controlling an acute bleed, but provide only a temporary resolution. Re-bleeding has been estimated at 74% with conservative measures,⁷ which commonly requires hospitalisation and blood transfusion. A preventive measure would be in accordance to the recommendations of performing primary intestinal anastomosis to avoid a stoma.²² However, in this case, a primary anastomosis was the preferred primary treatment option but due to a relapse of disease was not a viable long-term solution.

Stoma revision with devascularisation requires a laparotomy, is associated with the development of new varices and shown to not prevent recurrence of peristomal variceal bleeding.²³

Injection sclerotherapy is effective in controlling the acute bleed and permits more definitive procedures to be performed on an elective basis.⁸ However, the varices need to be visible to the operator and complications have included skin necrosis and mucosal ulceration.¹⁴

Liver transplantation has been demonstrated to provide long-term benefit with regression of peristomal varices within 2 weeks of liver grafting.¹¹ However, some patients may be unsuitable candidates for surgery and the worldwide lack of donors requires alternative therapeutic options to be considered.

Addressing the underlying portal hypertension can be achieved using either a surgical portosystemic shunt or TIPS, both of which would reduce the risk of re-bleeding by reducing the portosystemic pressure gradient.

Reports of surgical shunt procedures have been successful and are more effective in preventing recurrence of haemorrhage than either sclerotherapy or stomal revision.^{1 12 21 23} However, it is associated with high mortality (5%–15%) and morbidity,²³ and should be avoided in potential candidates for liver transplantation.

The goal of TIPS placement is to divert portal blood flow into the hepatic vein, so as to reduce the pressure gradient between portal and systemic circulations. It has been shown to be an effective therapy for portal hypertension when conservative medical management has failed.

TIPS was first described in the management of peristomal varices in 1994¹⁷ and has, since then, been successful in resolving bleeding in numerous cases.^{18 19} Documented procedure-related complications, included immediate shunt thrombosis and intraperitoneal bleeding. Therefore, Shibata *et al* recommended routine surveillance of shunts by duplex ultrasonography every 3 months to identify an occlusion. TIPS is a relatively safe procedure and can be performed in those patients unsuitable for surgery. When weighing the risks versus benefits, the presence of gastro-oesophageal varices would positively influence its use.

This case presented a management challenge due to its uncommon encounter. The definitive therapy for patients with hepatic failure is liver transplantation, but TIPS has proved to be a valuable bridge to transplantation. We recommend TIPS in those patients who fail to respond to conservative measures.

Learning points

- Our recommendations include; local pressure, transfusion of blood products, shunting for those unresponsive to conservative treatment or those with coexisting oesophageal varices and liver transplantation.

Competing interests None.

Patient consent Obtained.

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Please cite this article as follows (you will need to access the article online to obtain the date of publication).

Khan F, Vessal S, Gordon-Williams R. Bleeding from peristomal varices: a complication of portal hypertension. *BMJ Case Reports* 2011; 10.1136/bcr.08.2011.4598, date of publication

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