Transplant International ISSN 0934-0874

CASE REPORT

Vaginal bleeding complicating portal hypertension: a particular entity – Report of two cases and review of the literature

Giuseppe Orlando,[1] Pierre Goffette,[2] Andre Geubel[3] and Jan Lerut[4]

1 Liver Transplantation, Tor Vergata University, Rome, Italy
2 Department of Radiology (Prof. Baudouin Maldague)
3 Department of Hepatogastroenterology (Prof Yves Horsmans)
4 Unit Liver Transplantation and General Surgery, Cliniques Universitaires Saint-Luc, Université catholique de Louvain - UCL, Brussels, Belgium

Keywords
liver transplantation, portal hypertension, transjugular portosystemic stent shunt, vaginal haemorrhage, vaginal varices.

Correspondence
Prof. Jan P. Lerut MD, PhD, FACS, Unit Liver Transplantation and General Surgery, Cliniques Universitaires St-Luc – STARZL Room, Université catholique de Louvain – UCL, 10 Avenue Hippocrates, B – 1200 Brussels, Belgium. Tel.: +32 2 7645306; fax: +32 2 7649039; e-mail: lerut@chir.ucl.ac.be

Received: 27 June 2005
Revision requested: 21 July 2005
Accepted: 17 September 2005

doi:10.1111/j.1432-2277.2005.00232.x

Summary

Vaginal haemorrhage arising from varices is an exceptional complication of portal hypertension. Two cases successfully treated by transjugular intrahepatic portosystemic stent shunt are presented here. Both these patients previously had a total hysterectomy as did three out of four cases reported in the literature. Clinicians should be aware of 'vaginal bleeding' in the context of both hysterectomy and chronic liver disease in order to adopt the correct diagnostic and therapeutic approach. Definitive treatment can be obtained only if the underlying portal hypertension is adequately taken into consideration.

Introduction

The most frequent complications of portal hypertension (PHT) are ascites, hydrothorax and bleeding. The bleeding usually arises from varices in the gastrointestinal tract, and sometimes at the site of a surgical anastomosis. Bleeding from vaginal varices is extremely rare, as exemplified by the fact that only four cases have been reported [1–4]. We describe two further cases, which were successfully treated using transjugular intrahepatic portosystemic stent shunt (TIPSS) and liver transplantation (LT).

Material

Case 1

A 48-year-old female with cryptogenic cirrhosis, complicated by PHT and insulin-independent diabetes mellitus presented with menorrhagia. She was treated with lynestrenol (Orgametril®; Organon, Oss, The Netherlands). She went on to develop metrorrhagia and a total radical hysterectomy (TRH) was performed in March 2001. Thirteen months later, she presented with a vaginal bleed that originated from vaginal dome varices. After failure of tamponade, surgical haemostasis under general anaesthesia became necessary. At that time her Hgb was 10.5 g/dl, platelet count was 64 000/mm^3 and INR was normal. Three months later, she was re-hospitalized for metrorrhagia that was mainly occurring after sexual intercourse. As gynaecological examination showed only mild bleeding, she was treated conservatively. Hgb was 12.2 g/dl, platelet count 63 000/mm^3 and INR was normal. Two months later, tamponade and surgical haemostasis were required again. She was finally referred to our unit for evaluation of her underlying liver disease and considera-

tion for LT. As hepatic function was still satisfactory (Child-Pugh A), she was judged suitable for a TIPSS placement. This procedure was carried out in November 2002 using CO_2-venography. The portal venous system was patent and a large re-permeabilized umbilical collateral ending in a homolateral hypogastric vein was visualized (Fig. 1a and b). The mesenteric veins, catheterized through this collateral, drained a huge pelvic variceal venous network (Fig. 1c). The TIPSS included both an uncovered Palmaz and a 12-mm Smart® (Cordis Corporation, Miami, FL, USA) stent in order to adequately bridge the distance between portal and hepatic veins. At the end of the procedure, venography showed a well functioning shunt as well as shrinkage of the pelvic varices. The umbilical vein could still be visualized. Portosystemic venous pressure gradient fell from 16 to 7 mmHg. Post-TIPSS course was uneventful. Four months later, hysteroscopy showed that the vaginal varices had disappeared. The portosystemic pressure gradient had fallen to 5 mmHg. The paraumbilical vein was still patent but was occluded by coils and gel foam. She is now in regular follow-up at our outpatient clinic and her general condition and liver function are good. The TIPSS functions perfectly and there has been no recurrent vaginal haemorrhage (VH) after a follow-up of 34 months.

Case 2

A 51-year-old woman underwent TRH for a huge fibroma. Six years later, she presented with multiple gastrointestinal bleeds. These originated from ruptured oesophageal varices that had developed in the context of alcoholic cirrhosis. After failure of medical and endoscopic treatment a TIPSS was performed in June 1994. Eight years later, she developed liver insufficiency and grade III encephalopathy. It was decided to calibrate the TIPSS. The low portosystemic gradient of 3 mmHg and the absence of intrahepatic portal perfusion were consistent with an excessively large shunt, so a vascular reducing device $14 \times 4 \times 40$ mm, 70 cm/12 F, Sinus-Rep® (Belgium Medical, Ettlingen, Germany) was inserted. The shunt diameter was reduced from 13 to 7 mm and the portosystemic gradient rose to 9 mmHg. Her postoperative course was characterized by severe ascites and chronic vaginal blood loss, resulting in a fall in Hgb from 10 to 8.3 g/dl. These findings were explained by a TIPSS occlusion (confirmed on venography) that had

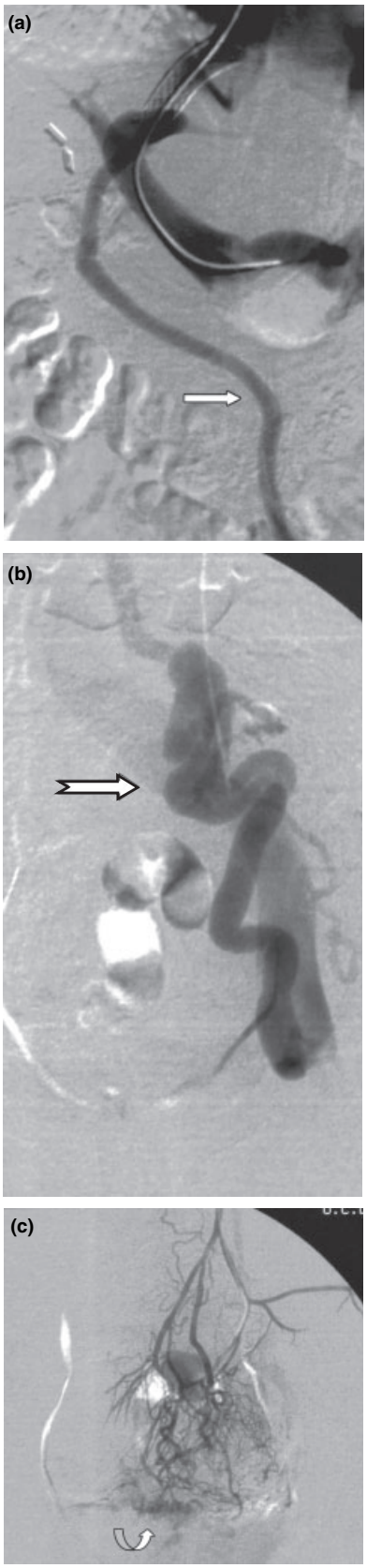

Figure 1 (a) Venography shows a patent portal venous system, a recanalized umbilical vein arising from the left portal vein. (⟹) which was (b) connected with a large hypogastric vein (⟹) and (c) hypertrophic pelvic venous network and vaginal varices (↺).

caused the development of pelvic varices originating from the inferior mesenteric vein (IMV). The TIPSS was easily repermeabilized and a Corinthian stent® (Cordis Corporation, Roden, The Netherlands) was placed within the reducing device, expanding the TIPSS diameter to 10 mm. The procedure was completed using selective intravascular thrombolysis in order to treat the associated thrombosis of the extrahepatic portal vein. The postoperative course was uneventful and the VH ceased. She underwent a successful transplant 12 months later (June 2003) for her progressive liver dysfunction. She is doing very well, 26 months post-LT, without any gynaecological problems.

Discussion

Most bleeding that occurs in the context of portal hypertension originates from oesophageal or gastric varices. Occasionally some patients develop varices in other areas. These ectopic varices may have unusual clinical presentations such as haemoperitoneum, haemophilia and haematuria. Previous surgical interventions favour the formation of these varices. The most frequent localizations of ectopic varices are stomal (27%) duodenal (18%), jejunal or ileal (18%), colonic (15%) peritoneal (10%) and rectal (9%) [5]. We have reported the cases of two cirrhotic patients, who had both undergone hysterectomy, and in whom portal hypertension resulted in the development of vaginal varices responsible for recurrent episodes of bleeding. Both were successfully treated using TIPSS.

Vaginal varices develop rarely in cirrhotic patients, if so they may cause acute and/or chronic bleeding. Reports in the literature are scarce (Table 1). The first case of VH from varices complicating PHT was reported in 1967 by Kreek in a 40-year-old alcoholic patient who had undergone previous TRH to control a postabortion haemorrhage [1]. During a 24-month period, she presented with three bleeds which were treated with urgent vaginal packing followed by partial vaginectomy. The second case was successfully treated with tamponade, portocaval shunt and interruption of the umbilical vein. She had also undergone previous TRH for recurrent severe menorrhagia. She died 3 months later from alcohol intoxication [2]. A third case concerned a 50-year-old woman with decompensated alcoholic liver disease. She died of uncontrollable bleeding from vaginal varices which developed 3 years after TRH for endometrial cancer [3]. The fourth case concerned a 69-year-old primary biliary cirrhosis patient who was successfully treated using retrograde transvenous obliteration of vaginal varices with a balloon occlusion technique. She had not had previous abdominal surgery [4].

Table 1. Literature review of vaginal bleeding in the presence of portal hypertension.

Authors	Year	Age	Previous TRH	Delay between TRH and VH onset	Aetiology of liver disease	Failure of local treatment	Need of transfusions	Treatment	Outcome
Kreek (1)	1967	40	Y	8 years	Alcoholic	Y	Y	Packing followed by elective partial vaginectomy	Recovery at 18 months
Enksson (2)	1982	42	Y	2 years	Alcoholic	Y	Y	Tamponade followed by elective portocaval shunt	Dead at 3-month alcohol intoxication
Marzotko (3)	1996	50	Y	3 years	Alcoholic	Y	Y	Emergency laparotomy with intra-abdominal and transvaginal tamponade	Perioperative dead
Hoshida (4)	1999	69	N	–	PBC	Y	N	Transvaginal ligation and tamponade followed by balloon-occluded retrograde obliteration	Complete recovery at 4 months
UCL	2004	49	Y	1 year, 1 month	Cryptogenic	Y	N	Tamponade and surgical haemostasis, followed by TIPSS	Complete recovery at 34 months
UCL	2004	66	Y	15 years	Alcoholic	Y	N	TIPSS revision followed by LT	Complete recovery at 24 months

TRH, total radical hysterectomy; VH, vaginal haemorrhage; PBC, primary biliary cirrhosis; LT, liver transplantation; TIPSS, transjugular intrahepatic portosystemic stent shunt.

Increased resistance to portal flow is the initiating factor in the development of PHT in liver cirrhosis. The portal system is partially decompressed through the development of collaterals, which mostly arise at the oesogastric junction and the periumbilical, retroperitoneal and perianal areas. Collaterals can also develop in the ovarian and pelvic regions. Haemorrhage, occurring as a result of both venous dilatation and increased endovascular pressure, most frequently originates from oesogastric varices [6]. Vaginal varices are very uncommon, they are probably due to the development of spontaneous venous shunts involving inferior mesenteric, ovarian, common iliac and pelvic veins. One should consider vaginal variceal bleeding especially in cirrhotic patients who have undergone previous hysterectomy. Indeed, both our patients, as well as three of the four reported cases, had undergone previous gynaecological surgery. The lack of a uterine vascular network may be responsible for the formation of vaginal varices by shunting venous blood directly to the vaginal venous system, causing massive congestion with risk of burst.

Gastrointestinal bleeding occurring in the context of PHT can be controlled effectively by combining pharmacological and/or endoscopical treatments. As different forms of PHT-derived haemorrhage have the same pathophysiological mechanism, the rationale to control VH should be to obtain local control by using similar strategies of haemostasis (tamponade, packing, sclerotherapy, ligature and surgical suture) and to pharmacologically reduce the splanchnic venous pressure. In case of the failure of such measures, TIPSS and finally, LT should be considered [7].

This report illustrates that VH complicating liver cirrhosis may be responsible for life threatening bleeding. TIPSS, and eventually LT, should be proposed in a timely manner to solve this difficult therapeutic problem. The first of our cases represents the first occurrence in which VH was the indication for TIPSS placement in the absence of any other bleeding source.

References

1. Kreek MJ, Raziano JV, Hardy RE, Jeffries GH. Portal hypertension with bleeding vaginal varices. *Ann Intern Med* 1967; **66**: 756.
2. Eriksson LS, Hårdstedt C, Law DH, Thulin L. Massive haemorrhage from vaginal varicose veins in patient with liver cirrhosis. *Lancet* 1982; **1**: 1180.
3. Marzotko E, Pfeiffer R, Nenning H, Kohler U. Vaginal hemorrhage after hysterectomy as a complication of alcohol-induced liver cirrhosis. *Zentralbl Gynakol* 1996; **118**: 417.
4. Hoshida Y, Saitoh S, Murashima N *et al.* Vaginal variceal haemorrhage in a patient with primary biliary cirrhosis: a case successfully treated by balloon-occluded retrograde transvenous obliteration. *Am J Gastroent* 1999; **94**: 3081.
5. Bosch J, Burroughs AK. Clinical signs and treatment of digestive bleeding in cirrhotic patients. In: Benhamou JP, Birger J, McIntyre N, Rizetto M, Rodes J, eds. *Hepatologie Clinique*, 2nd edn. Medecine-Science Flammarion, 2002: 680–691.
6. Stanley AJ, Hayes PC. Portal hypertension and variceal haemorrhage. *Lancet* 1997; **350**: 1235.
7. Azoulay D, Castaing D, Majno P *et al.* Salvage transjugular intra-hepatic portosystemic shunt for uncontrolled variceal bleeding in patients with decompensated cirrhosis. *J Hepatol* 2001; **35**: 590.