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CASE REPORT | LIVER

Ectopic Vaginal Varices With Hemorrhage After Hysterectomy

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ABSTRACT

Vaginal and uterine varices are well documented in pregnancy, although development of vaginal varices in patients with portal hypertension occurs in an exceptionally rare subset. Only 12 cases are reported in the literature; all but 3 of these cases involved patients with a history of hysterectomy, with 1 of the remaining 2 exhibiting partial obliteration of the uterine plexus due to radiation therapy for cervical cancer. We present a case of recurrent vaginal variceal bleeding in a patient with a history of hysterectomy, initially managed with vaginal tamponade and ultimately requiring definitive treatment with transjugular intrahepatic portosystemic shunt insertion.

INTRODUCTION

The development of varices as a consequence of shunting between the portal and systemic venous circulation is common in the setting of portal hypertension. Collateral venous flow is often found in the form of esophageal, gastric, rectal, and umbilical varices; however, ectopic varices may occur in less than 5% of cases.[1–3] Since the portal venous system provides outflow from the digestive organs, these ectopic sites are most often found in the small bowel or colon.[4] Acute bleeding from these ectopic sites can be fatal because many cases are unrecognized for an extended period. Bleeding from vaginal varices is among the rarest complications observed in patients with portal hypertension.[5] Twelve cases have been reported since 1967, and another case is detailed here.[6–16] Among reports of vaginal variceal bleeding, 9 occurred in the setting of portal hypertension after hysterectomy. In this report, we describe an episode of recurrent severe vaginal variceal hemorrhage in a patient with primary biliary cholangitis, portal hypertension, and a history of hysterectomy.

CASE REPORT

A 49-year-old woman presented to the emergency department with heavy vaginal bleeding. She had a history of decompensated cirrhosis secondary to primary biliary cholangitis and previously underwent hysterectomy for abnormal uterine bleeding 4 years before her presentation. Her course of cirrhosis was complicated by ascites, hepatic encephalopathy, and esophageal variceal hemorrhage requiring band ligation of esophageal varices. On initial presentation, she was found to have profuse vaginal bleeding with a decline in hemoglobin from 10.2 to 7.7 g/dL within her first 12 hours of hospitalization despite attempted tamponade with vaginal packing. Additional laboratory results included total bilirubin of 3.1 mg/dL, platelets of 68 $\times$ $10^3/\mu$L, international normalized ratio of prothrombin time 1.0, and Model for End-Stage Liver Disease-Na score of 12.

On further review, she had previously presented with vaginal hemorrhage attributed to vaginal varices on 2 occasions within the first 2 years after her hysterectomy. On the first presentation, she underwent percutaneous transhepatic portal venography with subselective catheterization and coil embolization of peripelvic collateral veins branching from the splenic hilum which supplied the

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vaginal varices. She experienced recurrent bleeding, requiring additional coil embolization of multiple peripelvic collateral veins followed by inferior mesenteric venography and coil embolization of small pelvic collateral veins arising from the superior rectal vein. Attempts were made at various times to initiate nonselective beta blocker prophylaxis from the time of her initial presentation with esophageal variceal hemorrhage; however, she did not tolerate therapy due to bradycardia.

Gynecologic evaluation with pelvic ultrasound at this time confirmed that the bleeding was again the result of vaginal variceal hemorrhage (Figure 1). Cross-sectional contrast-enhanced abdominal imaging with computed tomography angiography revealed extensive portal venous collateralization, but no portal vein thrombosis. She then underwent urgent transhepatic inferior mesenteric venography with sclerotherapy and embolization of the vaginal varices using a combination of 3% sodium tetradecyl sulfate foam and coil embolization of major feeding veins. This was immediately followed by transjugular intrahepatic portosystemic shunt (TIPS) insertion using an 8–10 mm × 7 cm × 2 cm Gore Viatorr stent graft with postdilation to 8 mm (Figure 2). A hepatic venous pressure gradient (HVPG) decreased from 19 mm Hg at baseline to 5 mm Hg after successful TIPS insertion. No recurrence of vaginal bleeding occurred after TIPS placement, and a follow-up pelvic and transvaginal ultrasound with Doppler evaluation demonstrated resolution of the vaginal varices (Table 1).

DISCUSSION

Postmenopausal vaginal bleeding is relatively common in the general patient population and is most commonly attributed to benign causes, including endometrial atrophy or polyps;

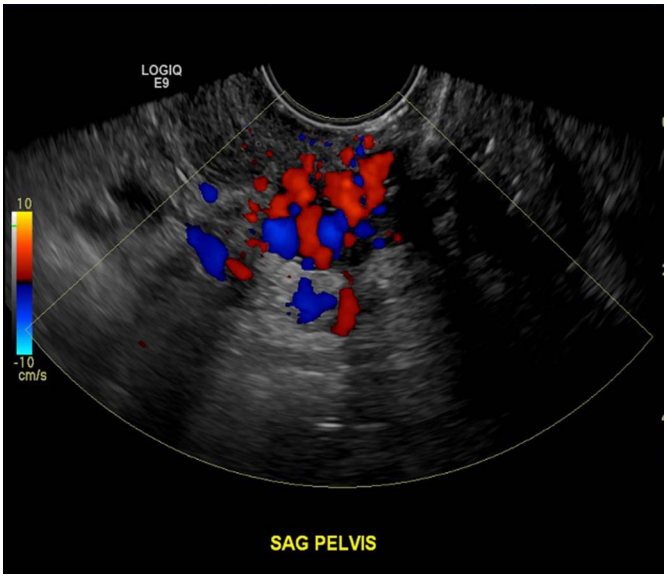

Figure 1. Pelvic ultrasound image demonstrating large varices at the vaginal cuff; the patient previously underwent hysterectomy.

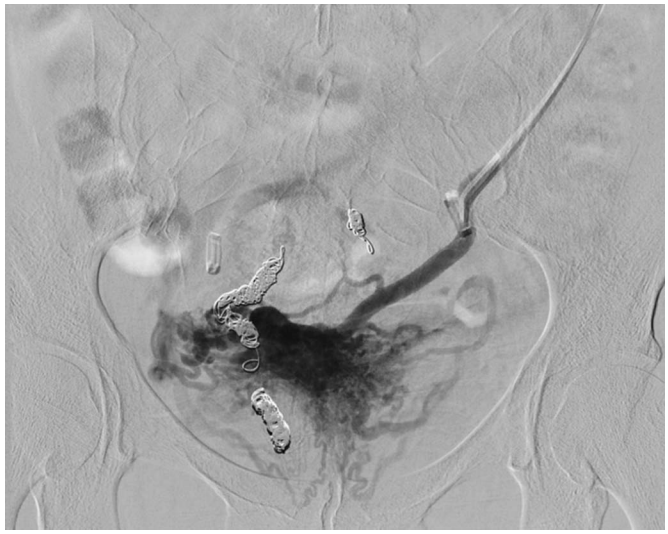

Figure 2. Venogram performed through transhepatic access identifying inferior mesenteric vein branch with large vaginal cuff varices. Note metallic coils from previous coil embolization of varices 2 years prior.

however, it can also occur in the setting of endometrial malignancy.[17] By contrast, vaginal bleeding in patients who have undergone a hysterectomy is rare but can result from various causes, such as postoperative uterine artery pseudoaneurysm, granulation tissue, infection, malignancy, or postmenopausal vaginal mucosal atrophy. Bleeding from vaginal varices in the setting of portal hypertension, as reported in this case, is among the rarest causes of vaginal bleeding.

Varices are a common complication of portal hypertension, most often occurring in the esophagus, stomach, and periumbilical vasculature.[4] However, varices can also develop at ectopic sites.[1–3] Under normal circumstances, the vast uterine plexus has the ability to dissipate the vascular pressure resulting from portal hypertension and prevent the development of vaginal varices.[4] When patients undergo a hysterectomy, the removal of the uterine plexus may decrease the capacitance of the venous system within the pelvis, leaving the vaginal veins vulnerable to the development of varices. The altered circulatory pathway through which vaginal varices may occur in this setting seems to originate from the formation of collateral vasculature between the superior hemorrhoidal plexus and the uterine and hypogastric veins, with subsequent venous congestion within the vaginal veins downstream.[5] Among cases occurring in patients who had not undergone hysterectomy, the vaginal bleeding was related to the collateral formation of ovarian veins that communicated with hepatofugal blood flow from the splenic vein in the setting of portal hypertension, compounded by an increased risk of bleeding associated with thrombocytopenia and elevated prothrombin time.[14]

In the management of patients presenting with hemorrhage from vaginal varices, it is critically important to establish

Table 1. Summary of reported cases of vaginal variceal hemorrhage

Cases	Reference	Age	Liver diagnosis	Past history	Management/treatment
1	Kreek et al[8]	40	Alcoholic cirrhosis	Hysterectomy	Laparotomy with ligation of the vaginal plexus to portal system collateral veins
2	Eriksson et al[9]	42	Alcoholic cirrhosis	Hysterectomy	End-to-side portocaval shunt
3	Marzotko et al[10]	50	Alcoholic cirrhosis	Hysterectomy	Vaginal tamponade, laparotomy without successful treatment
4	Hoshida et al[11]	52	Primary biliary cholangitis with cirrhosis	Radiation for uterocervical cancer resulting in partial uterine plexus obliteration, no hysterectomy	Transvaginal ligation, BRTO
5	Orlando et al[12]	48	Cryptogenic cirrhosis	Hysterectomy	TIPS
6	Orlando et al[12]	51	Alcoholic cirrhosis	Hysterectomy	TIPS, liver transplantation
7	MacHugh et al[13]	58	NAFLD cirrhosis	Hysterectomy	Emergent suture, liver transplantation
8	Nagata et al[14]	47	Hepatitis C cirrhosis	No hysterectomy	TIPS followed by partial splenic artery embolization
9	Garg et al[15]	36	Hepatitis C cirrhosis	Hysterectomy	TIPS
10	Glick et al[6]	35	NAFLD cirrhosis	No hysterectomy	TIPS
11	Chan et al[11]	60	NAFLD cirrhosis	Hysterectomy	Liver transplantation, splenectomy, IMV ligation
12	Sun et al[7]	55	No liver disease or portal hypertension; congenital AV fistula in the lower extremity	No hysterectomy	AV fistula occlusion

AV, arteriovenous; BRTO, balloon-occluded retrograde transvenous obliteration; IMV, inferior mesenteric vein; NAFLD, nonalcoholic fatty liver disease; TIPS, transjugular intrahepatic portosystemic shunt.

control in the acute setting and proceed with definitive therapy. In our case, initial efforts to obliterate the vaginal varices using coil embolization were inadequate without achieving reduction in her HVPG with TIPS placement. Immediate management of acute bleeding from vaginal varices includes packing the vaginal canal with gauze and achieving tamponade. Although hemostasis can be achieved with measures such as coil embolization, the risk of bleeding recurrence may persist in the presence of ongoing portal hypertension, particularly with elevated HVPG, as in our case. Similarly, several reported cases of bleeding from vaginal varices noted unsuccessful attempts with medical management (beta blockers and octreotide) and local tamponade until definitive treatment with reduction of portal hypertension could be achieved through TIPS placement or liver transplantation.[6,16] Definitive therapy led to the lasting cessation of vaginal varices and absence of subsequent bleeding in cases that included follow-up evaluations.[11–14]

A clear understanding of the vascular network contributing to the development of vaginal varices is required because recurrent hemorrhage has been reported after liver transplantation in cases where the altered vasculature leading to hemorrhage was not resolved by transplantation. In 1 report, a patient with portal vein thrombosis presented with bleeding from vaginal varices arising from collateralization with the inferior mesenteric vein. Bleeding persisted even after liver transplantation likely because of residual venous occlusion and left-sided portal hypertension that had not been decompressed with liver transplantation and interpositional graft to the superior mesenteric vein , ultimately requiring splenectomy and ligation of the inferior mesenteric vein. Attention to the specific portal venous vasculature of each case is essential to determine which treatment will be most effective. Our case emphasizes the need for definitive treatment and reduction of portal hypertension in cases of bleeding ectopic vaginal varices and highlights the need to consider this diagnosis in the evaluation of women with a history of cirrhosis, portal hypertension, and hysterectomy who present with severe recurrent vaginal bleeding.

DISCLOSURES

Author contributions: JK Smith, M. Gautam, and H. Elsiesy are responsible for the conceptualization of the manuscript. JK Smith, PG Wortley, and SA Gonzalez are responsible for writing of the original draft. All authors participated in reviewing and editing the manuscript. All authors approve the final, submitted draft. SA Gonzalez is the article guarantor.

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