OPEN ACCESS

Case of postpartum uterine artery pseudoaneurysm associated with von Willebrand disease

Jesse Wayson,[1] Jennifer Tomlinson Allen ,[1] Shahar Laks,[1] Michael Allen[2]

[1]ObGyn, Augusta University, Augusta, Georgia, USA
[2]Emergency Medicine, Augusta University, Augusta, Georgia, USA

Correspondence to
Dr Jennifer Tomlinson Allen; jenallen@augusta.edu

JW and JTA are joint first authors.

Accepted 11 December 2022

SUMMARY

A woman in early 20s with type 1 von Willebrand disease (vWD) presented to the emergency department with abdominal pain and vaginal bleeding at 5 weeks post partum following primary caesarean section. Imaging revealed a uterine artery pseudoaneurysm (UAP), which is a rare condition that can cause postpartum haemorrhage. Caesarean birth and vWD are two risk factors for pseudoaneurysm. Swift postpartum recognition of a pseudoaneurysm is essential to prevent a potentially life-threatening outcome. Successful diagnosis and treatment of the patient's symptoms required interdisciplinary teamwork between obstetricians, interventional radiologists and haematologists. Uterine artery embolisation (UAE) was performed and complete resolution of the pseudoaneurysm was noted 6 weeks after the procedure. Haemorrhage was averted due to early detection of UAP prior to its rupture, and future fertility was preserved. The patient successfully conceived a second child 1 year after UAE and delivered via caesarean birth without haemorrhage or recurrence of UAP.

BACKGROUND

Von Willebrand disease (vWD) is the most common inherited bleeding disorder, affecting an estimated 1.3% of American women.[1 2] It may be caused by either a qualitative or quantitative deficiency of von Willebrand factor (vWF), a critical protein for platelet adhesion and protection against degradation of clotting factors.[1] The disease is divided into three main types. Type 1 vWD is the most common of these, and accounts for approximately 80% of cases. In type 1 vWD, levels of vWF can be either minimally to moderately decreased, whereas type 3 vWD is characterised by severely low or undetectable levels of vWF.[1] Type 2 vWD involves a qualitative deficiency of vWF, which is characterised by normal to near normal levels of vWF, but functional deficiency of the protein itself.[1]

Pregnant women with vWD are at a significantly higher risk of postpartum haemorrhage (PPH) than pregnant women at baseline.[3] In the setting of concurrent uterine artery pseudoaneurysm (UAP), an obstetric case can quickly become fatal. A pseudoaneurysm is a blood-filled cavity communicating with the arterial lumen due to a deficiency of one or more layers of an arterial wall.[4] Trauma to the uterine artery during perforation (such as during surgery) leads to a defect in the arterial wall, through which arterial blood escapes and diffuses through adjacent tissue, causing a collection of blood to form. When this collection is in continuity with the uterine artery which supplies a continuous flow of blood, a pseudoaneurysm forms.[5] The absence of a three-layered arterial wall lining in the pseudoaneurysm differentiates it from a true aneurysm.[6] The prevalence of UAP is difficult to determine due to its rarity; however, the prevalence is cited as 2–6 per 1000 deliveries when asymptomatic patients are included.[7 8] Spontaneous evolution of a pseudoaneurysm can lead to rupture. UAP can occur after caesarean delivery, especially when there is arterial injury during the procedure.[9] Pseudoaneurysm rupture is a rare cause of PPH, which can occur up to 3 months after caesarean delivery.[6] Common misdiagnoses include hypermenorrhoea, retained products of conception or endometritis.

While the risk of increased bleeding in pregnant patients with type 1 vWD has been well documented, and the risk of PPH in patients with pseudoaneurysm is well established in the literature, delayed PPH resulting from UAP in patients with vWD is only rarely reported, and there are no standardised recommendations to guide management in these situations. Notably, the patient in that case was found to have vWD after she presented with a PPH.[9]

We present a case of a delayed PPH due to a UAP in the setting of vWD. We also discuss detailed strategies for preventing peripartum haemorrhage and PPH in patients with vWD, as well as the diagnosis and treatment of UAP in a postpartum patient.

CASE PRESENTATION

A woman in her early 20s, gravida one para one, with a history of type 1 vWD, underwent a primary caesarean delivery for arrest of descent and cephalopelvic disproportion, complicated by acute chorioamnionitis. The patient was followed by haematology throughout her pregnancy and had no history of antepartum bleeding. At the time of admission for induction of labour, she received Humate-P (antihaemophilic factor-vWF (coagulation factors)) prior to delivery and Amicar (antifibrinolytic agent) following delivery for bleeding prevention. The caesarean delivery itself was uncomplicated, with an estimated blood loss of 850 mL. The preoperative haemoglobin (Hb) level was 112 g/L, and Hb level at discharge on postoperative day 2 was 95 g/L. This was considered an appropriate decline on the basis of the estimated blood loss during her surgery. No other haemostatic agents were required during her postpartum hospital stay.

The patient presented to the obstetric emergency room on postoperative day 35 with heavy vaginal bleeding, nausea, diarrhoea, and both periumbilical

To cite: Wayson J, Allen JT, Laks S, *et al*. *BMJ Case Rep* 2022;**15**:e253804. doi:10.1136/bcr-2022-253804

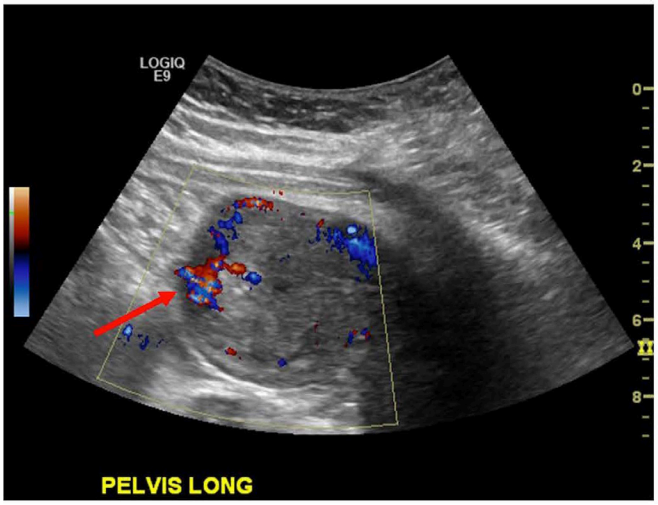

Figure 1 Transvaginal pelvic ultrasound showing 1.8×1.6×1.8 cm focal hyperechoic area within the endometrial region at the fundus of the uterus, with prominent vessels in the area.

and right lower quadrant abdominal pain. She was afebrile and vital signs were within normal limits. Abdominal examination revealed a skin incision without erythema or warmth and there was mild tenderness to palpation along the incision and in bilateral lower quadrants. She had normoactive bowel sounds and the uterine fundus was palpated 3 cm below the umbilicus and moderately tender. There was no evidence of guarding or rebound. Pelvic examination was significant for bright red blood in the vaginal vault. Her Hb level on admission was 101 g/L and white cell count was $79x10^9$/L. A urine pregnancy test was negative. Based on these findings, there was a low suspicion for the more common causes of delayed PPH, such as endometritis, retained products of conception, or gestational trophoblastic disease or neoplasia.

INVESTIGATIONS

A pelvic ultrasound revealed a 1.8 cm hyperechoic area at the level of the uterine fundus, and colour Doppler showed prominent vessels around the superior, anterior and posterior aspect of the hyperechoic area, suggestive of arterial malformation (figure 1). An emergent CT scan of the abdomen and pelvis was performed with intravenous contrast, which revealed a hypervascular structure within the fundus of the endometrial cavity measuring 1.6×1.0×1.4 cm. This structure was seen to be in communication with fundal branches of the uterine artery and was highly concerning for pseudoaneurysm (figure 2). There was no CT evidence for acute appendicitis or other pathology. Emergent MRI confirmed the diagnosis of pseudoaneurysm (figure 3).

TREATMENT

Interventional radiology performed a catheterisation of the left internal iliac artery with angiogram, followed by gelfoam embolisation of the left uterine artery (figure 4). The patient tolerated the procedure well and was administered intramuscular Depo-Provera for contraception prior to discharge.

OUTCOME AND FOLLOW-UP

CT angiogram was performed 6 weeks after uterine artery embolisation (UAE) was performed, with complete resolution of the pseudoaneurysm noted. Fertility was preserved, and the patient conceived again approximately 12 months later. She had an uncomplicated pregnancy and delivered by repeat caesarean section at term.

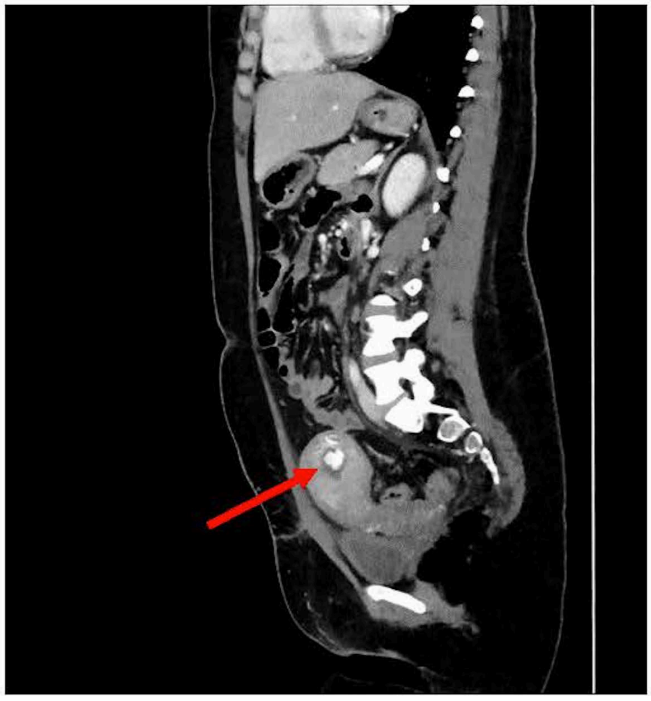

Figure 2 Sagittal CT of the abdomen and pelvis showing 1.6×1.0×1.4 cm hypervascular structure within the fundus of the endometrial cavity that appears to connect with the fundal branches of the uterine artery, consistent with pseudoaneurysm.

DISCUSSION

PPH is the most common cause of maternal death worldwide,[9] and prompt recognition, diagnosis and treatment are necessary to prevent poor outcomes. Primary PPH occurs within 24 hours of delivery and is caused by uterine atony in 80% of cases.[9] Secondary PPH occurs from 24 hours to 6 weeks after delivery, and is usually due to retained products of conception, infection, delayed involution of the placental bed or bleeding disorders.[9] Appropriate workup should be performed to rule out these common aetiologies. In a stable patient, imaging should be conducted prior to diagnostic dilation and curettage, as this operation could cause significant harm to a patient with

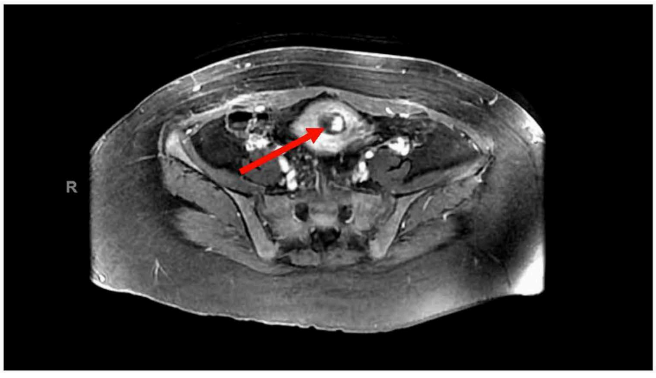

Figure 3 Axial MRI with contrast of pelvis showing 1.4×1.5 cm enhancing structure involving the uterine fundus consistent with uterine artery pseudoaneurysm.

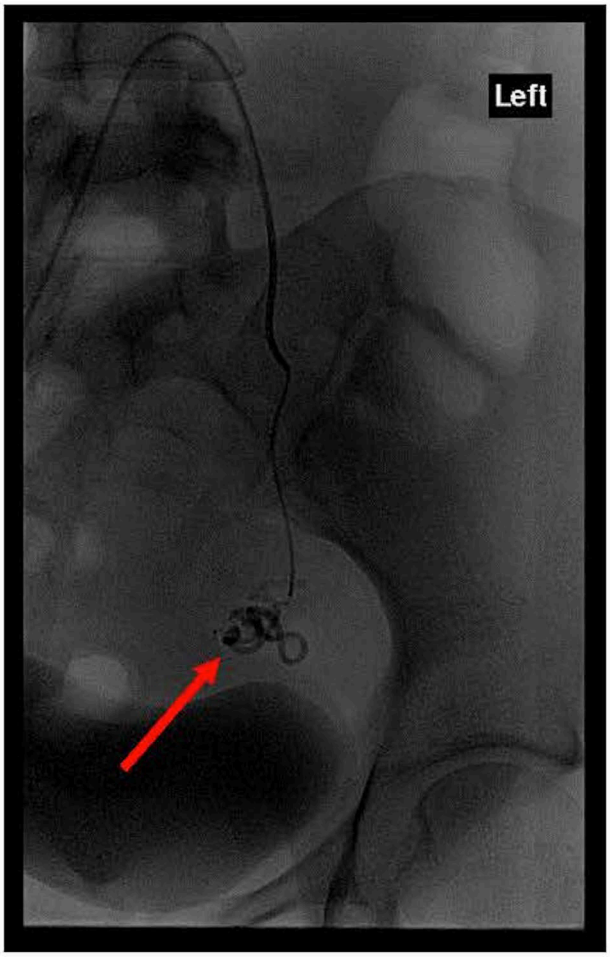

Figure 4 Uterine angiography demonstrating a pseudoaneurysm of the distal left uterine artery fundal branch, with successful gelfoam embolisation of the pseudoaneurysm.

a uterine vascular malformation, and potentially lead to catastrophic bleeding.

Bleeding due to a UAP is a rare cause of delayed PPH.[9] The most common presenting symptoms of UAP are abnormal vaginal bleeding and pelvic pain. Caesarean delivery is the greatest risk factor for having a UAP. Spontaneous haemostasis can occur, reinforcing a misdiagnosis, and there can be a delay of up to 4 weeks before recurrence of bleeding.[4 5]

Our patient developed a UAP in the setting of type 1 vWD. In a case report of a PPH due to UAP in a patient with vWD, the patient endured significant blood loss, likely because the diagnosis of vWD was not known prior to delivery, and delayed evaluation of the patient's postpartum bleeding led to rupture of the pseudoaneurysm. In our case, a prenatal diagnosis of vWD facilitated preventive measures to avert heavy bleeding. Her pre-existing diagnosis of vWD also led to a heightened awareness of her increased risk of delayed PPH, which in turn prompted early imaging at the time of her presentation to the emergency room. This allowed for early detection of her pseudoaneurysm and likely prevented its rupture, which is associated with significant blood loss when it occurs. The finding of a pseudoaneurysm in a patient without a history of bleeding disorders should prompt an evaluation for possible bleeding disorders.[10]

Pregnant women with type 1 vWD should be co-managed with a haematologist throughout pregnancy. Measurement of factor VIII and vWF levels is recommended and treatment should be administered when the factor VIII level is less than 25% of its normal range and the vWF level is below 50 IU/dL.[11] However, providers may also prophylactically treat in the late stages of labour and 3–4 days post partum if there is concern for bleeding despite normal levels of vWF and factor VIII. The treatment most commonly offered is intranasal desmopressin, a synthetic vasopressin that increases levels of vWF and factor VIII.[11] vWF/factor VIII concentrates have also been developed and are approved for use as an alternative therapy. Our patient received Humate-P (antihaemophilic factor-vWF) 50 units/kg intravenous loading dose with maintenance dose 20–30 units/kg intravenously at delivery, with repeat dosing every 12 hours for 72 hours post partum. She received Amicar 8000 mg every 6 hours orally for 48 hours. Additionally, the patient was prescribed Humate as needed to treat any acute onset of heavy bleeding in the outpatient setting.

Non-operative vaginal delivery is the preferred mode of delivery for patients with vWD; however, caesarean delivery should still be performed for usual obstetric indications.[11] Our patient had a primary caesarean delivery for arrest of descent in the setting of cephalopelvic disproportion.

For the detection of a pseudoaneurysm, B-mode imaging usually shows an anechoic lesion in the wall of the lower uterus, which may be pulsatile. Duplex Doppler ultrasound shows a mosaic flow pattern due to the arterial flow swirling in different directions and at different velocities.[4 5] Sonographic features may disappear during active bleeding due to arterial pressure deflation and spastic communication, especially when the pseudoaneurysm is small in diameter. Contrast-enhanced CT or MRI may be used to confirm the diagnosis as its sensitivity to detect arterial lesions is comparable with angiography.[4] Angiography remains the gold-standard method for the diagnosis of UAP, and may facilitate definitive treatment.[4] Traditional management of UAP includes bilateral internal iliac or uterine artery ligation, and, when other treatments fail, hysterectomy. Transcatheter selective arterial embolisation has recently emerged as a safe and highly effective treatment due to its high success rate (>90%), low complication rate and its ability to preserve future fertility.[8]

Major complications following UAE are rare. About 8.5% short-term and 1.25% serious complication rates have been reported to date.[12] Postprocedural imaging is routinely performed to evaluate effectiveness of treatment and to identify potential post-UAE complications. For our patient, a surveillance CT angiogram was performed 6 weeks after UAE, with complete

Learning points

- Uterine artery pseudoaneurysm is a rare complication of pregnancy or surgery, particularly caesarean delivery, and usually presents with abnormal vaginal bleeding and abdominal or pelvic pain.
- Patients with von Willebrand disease (vWD) are more prone to developing uterine artery pseudoaneurysm and may experience catastrophic bleeding with pseudoaneurysm rupture. Optimising medical management of vWD, consulting with haematology and early detection of pseudoaneurysm with imaging such as ultrasonography with Doppler, CT or MRI are critical to avoid a misdiagnosis or delay in care.
- Uterine artery embolisation is the treatment of choice when a pseudoaneurysm is identified. Surveillance angiogram is important to ensure resolution.

resolution of the pseudoaneurysm noted. The patient conceived again approximately 12 months later.

This case illustrates a scenario in which appropriate preventative measures were taken before delivery, during labour and in the postpartum period in a patient with known type 1 vWD. Postpartum vaginal bleeding or pain in a patient with known vWD should raise concern for pseudoaneurysm in the differential diagnosis. Appropriate workup for early detection and treatment with UAE is necessary to avert severe haemorrhage. This patient recovered well and was discharged home for outpatient surveillance the day after UAE was performed.

Twitter Jesse Wayson @WaysonMD

Acknowledgements We fully acknowledge the contribution of our patient to this work and appreciate her consent to share her medical journey.

Contributors All four authors listed contributed to this case report in a significant way. JW and JTA managed the patient's care and wrote the bulk of the case report, while MA and medical student SL edited the work and verified references. The senior author, JTA, accepts full responsibility for the content of the finished work, had access to the patient's data and controlled the decision to publish.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent for publication Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

ORCID iD

Jennifer Tomlinson Allen http://orcid.org/0000-0001-6172-5898

REFERENCES

1 Castaman G, James PD. Pregnancy and delivery in women with von Willebrand disease. *Eur J Haematol* 2019;103:73–9.
2 James AH. Von Willebrand disease. *Obstet Gynecol Surv* 2006;61:136–45.
3 Sladič M, Verdenik I, Smrkolj Špela. The effect of von Willebrand disease on pregnancy, delivery, and postpartum period: a retrospective observational study. *Medicina* 2022;58. doi:10.3390/medicina58060774. [Epub ahead of print: 07 Jun 2022].
4 Bouchet P, Chabrot P, Fontarensky M, *et al*. Pitfalls in diagnosis of uterine artery pseudoaneurysm after cesarean section. *Ultrasound Obstet Gynecol* 2012;40:482–3.
5 Isono W, Tsutsumi R, Wada-Hiraike O, *et al*. Uterine artery pseudoaneurysm after cesarean section: case report and literature review. *J Minim Invasive Gynecol* 2010;17:687–91.
6 Kuwata T, Matsubara S, Kaneko Y, *et al*. Asymptomatic uterine artery pseudoaneurysm after cesarean section. *J Obstet Gynaecol Res* 2010;36:405–10.
7 Parr K, Hadimohd A, Browning A, *et al*. Diagnosing and treating postpartum uterine artery pseudoaneurysm. *Proc* 2018;31:56–8.
8 Dohan A, Soyer P, Subhani A, *et al*. Postpartum hemorrhage resulting from pelvic pseudoaneurysm: a retrospective analysis of 588 consecutive cases treated by arterial embolization. *Cardiovasc Intervent Radiol* 2013;36:1247–55.
9 Shimizu M, Kondoh E, Ueda M, *et al*. Secondary postpartum hemorrhage due to uterine artery pseudoaneurysm rupture in von Willebrand disease. *J Obstet Gynaecol Res* 2011;37:1887–90.
10 Kwon JH, Kim GS. Obstetric iatrogenic arterial injuries of the uterus: diagnosis with us and treatment with transcatheter arterial embolization. *Radiographics* 2002;22:35–46.
11 Demers C, Derzko C, David M, *et al*. Gynaecological and obstetric management of women with inherited bleeding disorders. *J Obstet Gynaecol Can* 2005;27:707–32.
12 Spies JB, Spector A, Roth AR, *et al*. Complications after uterine artery embolization for leiomyomas. *Obstet Gynecol* 2002;100:873–80.