BMJ Case Reports

## Complete the template below

Please type your report directly into this template. Read the tips and reminders in each section as you type. Use formal US or UK English and scientific terminology.

### TITLE OF CASE

Embolisation of an acquired uterine arteriovenous malformation.

### SUMMARY

A G5P2+2 female in her 30s presented to hospital with PV bleeding, approximately two weeks post electric vacuum aspiration (EVA) for retained products of conception. Ultrasound and MRI demonstrated a large vascular myometrial lesion, suggestive of a uterine arteriovenous malformation (UAVM). She underwent digital subtraction angiography (DSA) with interventional radiology, and simultaneous uterine artery embolisation (UAE). She represented two weeks later with recurrent PV bleeding and anaemia. She underwent repeat DSA, demonstrating persistent UAVM, and a repeat embolisation was performed. Symptoms resolved following the second embolisation, and a repeat MRI performed twelve weeks later demonstrated complete resolution of the UAVM.

Although UAVMs are rare lesions, they can cause significant haemorrhage and morbidity. The presence of UAVM should particularly be considered after uterine intervention such as EVA or caesarean section.

UAE is a safe and effective therapy, which preserves fertility.

### BACKGROUND

UAVM is an uncommon vascular lesion, typically presenting with vaginal bleeding, which can be life-threatening. They are relatively rare lesions, with less than 200 cases published in the literature.

Given the risks of significant morbidity and mortality, prompt diagnosis and management is essential. The diagnosis can be made based on a combination of clinical, biochemical, and radiological findings.

Close communication between the obstetric and interventional radiology teams can provide rapid diagnosis and effective treatment.

### CASE PRESENTATION

A G5P2 +2 (caesarean section x 2, early medical termination of pregnancy x2) woman in her thirties attended our unit requesting termination of pregnancy. The patient had recently moved to our jurisdiction and had limited English. Translation services were used throughout consultations with the patient. Her past medical and surgical history was unremarkable, and she had a normal BMI. A transvaginal ultrasound revealed a viable intrauterine normally sited pregnancy of 6 weeks' gestation. Medical termination of pregnancy (MTOP) was performed as per guidelines with Mifepristone 200mg PO stat followed by misoprostol 800mg buccal after at home 36 hours later. Six days later the woman was reviewed again in clinic as her bleeding had been minimal in the period since the administration of medications. A repeat transvaginal ultrasound showed the pregnancy still in situ however foetal cardiac

activity was not present. The woman was counselled about options and elected to proceed with electric vacuum aspiration under general anaesthesia. The bleeding was noted to be heavier than usual following removal of pregnancy tissue, however haemostasis was achieved following bimanual compression with a total estimated blood loss of 450ml. There were no clinical concerns for molar pregnancy on examination of the pregnancy tissue. An etonogestrel subdermal contraceptive implant (Implanon NXT®) was inserted for long-acting reversible contraception at the woman's request and she was discharged home.

Eleven days later the woman presented to the emergency department with heavy vaginal bleeding, suprapubic pain and a history of a watery vaginal discharge. On examination suprapubic and uterine tenderness were noted. Haemoglobin was 8.7g/dL and serum β-hCG was 28,411 IU/L. An ultrasound revealed a thin endometrial thickness of 5.6mm implying no retain products of conception were present. The anterior myometrium appeared heterogenous with prominent vascularity. The woman was admitted and commenced on intravenous co-amoxiclav 1.2g eight-hourly with a working diagnosis of endometritis. She was observed as an inpatient for four nights and received 48 hours of intravenous antibiotics prior to switching to oral. She remained well throughout this period with light vaginal spotting only and was discharged home when serial β-hCG monitoring showed a downward trend.

Two days later she re-presented to the emergency department again complaining of heavy vaginal bleeding. She was haemodynamically stable with a haemoglobin of 6.6 g/dL so was admitted to the high dependency unit (HDU) and four units of red cell concentrate (RCC) were transfused.

## INVESTIGATIONS *If relevant*

Multiphasic gadolinium-enhanced MRI of the pelvis was performed (figure 1). This demonstrated a mass in the uterine body measuring 5.2 x 6.1 x 8.2 cm. The mass was isointense on T1-weighted sequences and heterogeneously hyperintense on T2-weighted sequences. Arterial phase imaging showed enhancement of several serpiginous structures within the mass, thought to represent arteriovenous shunting. A non-enhancing component was present centrally consistent with haemorrhage or necrosis. In addition, the pelvic veins were dilated.

The patient proceeded to have further investigation with digital subtraction angiography (DSA) in the interventional radiology suite (figure 2). This demonstrated a large enhancing vascular lesion with an irregular tangled network of arteries, veins and capillaries, and early venous filling of the left ovarian vein. Appearances were consistent with a uterine arteriovenous malformation (UAVM).

## DIFFERENTIAL DIAGNOSIS *If relevant*

The DSA imaging appearances, with arteriovenous shunting, capillary tangles and early venous filling, were consistent with UAVM. In addition, the recent history of medical termination of pregnancy with electrical vacuum aspiration is a described risk factor for acquired UAVM.

Initial ultrasound appearances were potentially compatible with a molar pregnancy, however the down trending serum β-hCG made this a less likely diagnosis.

## TREATMENT *If relevant*

Following DSA, a decision was made to perform uterine artery embolisation (UAE) to treat the AVM. Both uterine arteries were selectively catheterised, and temporary embolic material (Gelfoam®) administered. The post embolisation angiogram (figure 3) demonstrated a satisfactory and significant devascularisation of the AVM.

Two weeks later the woman re-presented to the emergency room with heavy vaginal bleeding. She was febrile, hypotensive and tachycardic. Her haemoglobin was 10g/dL on admission and the speculum examination revealed minimal vaginal bleeding. She was treated for sepsis with the devascularised uterus and endometrium the presumed source. She was commenced on broad spectrum antibiotics and transferred to the high dependency unit. Blood cultures were positive for multidrug resistant E. Coli and following consultation with clinical microbiology, was administered meropenem 1g every 8 hours.

The patient was further investigated with a CT mesenteric angiogram which showed a recurrence of the AVM with both bilateral ovarian and uterine artery supply, and an accessory artery arising from the left internal iliac artery.

The woman wished to retain her uterus for potential future pregnancy but was counselled extensively regarding the risk of requiring hysterectomy should she develop life-threatening haemorrhage or infection. She continued to improve clinically and following seven days of intravenous antibiotics and 3 negative blood cultures, a repeat DSA and uterine artery embolisation was performed, utilising Gelfoam® and coils (figure 4).

## OUTCOME AND FOLLOW-UP

The patient's symptoms resolved following the embolisation, with no further menorrhagia or anaemia. The patient was observed as an inpatient for a further six days and discharged home well. Weekly β-hCG monitoring was performed for 10 weeks and normalised to non-pregnancy levels.
Subsequent MR imaging of the pelvis performed twelve weeks post repeat embolisation demonstrated complete resolution of the AVM (figure 5).

## DISCUSSION *Include a very brief review of similar published cases*

UAVM is an uncommon vascular lesion, typically presenting with vaginal bleeding, which can be life-threatening. AVM pathophysiology involves an abnormal communication between uterine arteries and veins, often via an abnormal cluster of capillary vessels (figure 6).

AVMs are described as either acquired or congenital. Congenital AVMs are thought to be due to failure of normal embryological uterine vascular development. Acquired AVMs most commonly occur due to trauma to the myometrium, for example during dilatation and curettage (D&C), caesarean section or direct trauma to the uterus. Uterine malignancy (such as gestational trophoblastic disease) or infection can also cause a UAVM.

They are relatively rare lesions, and most of the literature consists of case reports – less than 200 case reports have been published (2, 3). Overall, there exists a paucity of quality research - Peitsidis et al (3) performed a systematic review, identifying six observational studies involving a total of 103 patients, and graded the quality using the Scottish Intercollegiate Guidelines Network (SIGN). Only one graded as 2+, indicating moderate probablity that demonstrated relationships were causal; the rest demonstrated significant risks of non-causal relationships.

Given the often dramatic presentation with post partum haemorrhage or vaginal bleeding, prompt diagnosis and management is crucial. Ultrasound, CT and MR angiography are useful, but DSA is considered the gold standard for diagnosis and permits simultaneous treatment with embolisation (3, 4).

Owing to the rarity of the condition and a lack of high quality evidence, established guidelines for management of UAVM are sparse (5). UAxE and hysterectomy are well described treatments, and the choice of management will vary depending on local skills and expertise – however UAE has the important benefit of preserving fertility (6, 7). Hoang et al describe an approach to investigation and treatment, and further outline interventional radiology strategies depending on the Yakes classification of the aneurysm (8). Medical treatment has also been described in a minority of case reports, and can be achieved using contraceptives, hormonal therapy or methylergotramine maleate.

## LEARNING POINTS/TAKE HOME MESSAGES *3-5 bullet points*

- Uterine AVM is a rare vascular lesion that can present with significant vaginal bleeding.
- Most uterine AVMs are acquired, rather than congenital, and can occur following obstetric procedures such as D&C, caesarean section, and also direct uterine trauma, malignancy and infection.
- Ultrasound, CT, and MR angiography are useful in establishing the diagnosis, but DSA is the gold standard, and allows for simultaneous treatment in the form of uterine artery embolisation (UAE).
- Treatment with either UAE or hysterectomy is highly effective, but UAE has several advantages including preservation of fertility and the absence of a need for general anaesthetic.

## REFERENCES


*1. Yakes AM, Continenza AJ, Yakes WF. The Yakes AVM Classification System: Cracking the Code for Curative AVM Endovascular Treatment. In: Khanna AK, Tiwary SK, editors. Vascular Malformations. Singapore: Springer Singapore; 2021. p. 275-94.*

*2. Salmeri N, Papale M, Montresor C, Candiani M, Garavaglia E. Uterine arteriovenous malformation (UAVM) as a rare cause of postpartum hemorrhage (PPH): a literature review. Archives of Gynecology and Obstetrics. 2022;306(6):1873-84.*

*3. Peitsidis P, Manolakos E, Tsekoura V, Kreienberg R, Schwentner L. Uterine arteriovenous malformations induced after diagnostic curettage: a systematic review. Arch Gynecol Obstet. 2011;284(5):1137-51.*

*4. Davies R, Parker M, Basu A, Alleemudder D. The clinical applications of interventional radiological techniques in obstetrics and gynaecology. The Obstetrician & Gynaecologist. 2023;25(2):131-9.*

*5. Matsumoto MM, Caridi TM. Uterine Vascular Anomalies: Management and Treatment Overview. Semin Intervent Radiol. 2023;40(4):342-8.*

*6. Ahmadi F, Moukhah S. A case report of Ggeneralized uterine arteriovenous malformation after molar pregnancy in an infertile woman. Int J Reprod Biomed. 2018;16(2):119-22.*

*7. Eling R, Kent A, Robertson M. Pregnancy after uterine arteriovenous malformation-case series and literature review. Australas J Ultrasound Med. 2012;15(3):87-96.*

*8. Hoang VT, Van HAT, Trinh CT, Pham NTT, Huynh C, Ha TN, et al. Uterine Arteriovenous Malformation: A Pictorial Review of Diagnosis and Management. J Endovasc Ther. 2021;28(5):659-75.*


## FIGURE/VIDEO CAPTIONS

**Figure 1.**
MRI pelvis showing a large vascular mass within the uterus, serpiginous enhancing vasculature (*black arrow)* and dilated pelvic veins *(black arrowhead)*

**Figure 2.**

Digital subtraction angiography (DSA). Images *a-c* demonstrate the large rounded vascular mass, with a nidus of irregular blood vessels. Bilateral uterine arterial supply is present. Image *d* demonstrates the early venous filling of the left ovarian vein *(black arrow).*

**Figure 3.** Pre and post embolisation DSA of the pelvis, demonstrating significant devascularisation of the pelvic mass. The uterine arteries remain patent *(black arrows).*

**Figure 4. (a)** DSA demonstrating recurrence of the AVM. Large ovarian arteries bilaterally, with 'corkscrew' appearance (black arrows). **(b)** Angiogram post embolisation with Gelfoam and coils (black arrows). Residual AVM arterial supply from accessory branch of the left internal iliac artery (black arrowhead). **(c)** Final angiogram. Both uterine arteries and the accessory left internal iliac artery branch have been coiled.

**Figure 5. (a)** Sagittal MRI pelvis at initial diagnosis, showing large heterogenous mass in the uterus, obliterating the endometrial stripe and subsequently demonstrated to be an AVM**. (b)** Post embolisation sagittal MRI pelvis. The AVM has entirely involuted, and the uterus is normal in appearance, with a normal endometrial stripe. A Caesarean section scar is evident in the uterine body (*).

**Figure 6.** Original illustration of the pathophysiology of arteriovenous malformations, as described by Yakes et al (1).
**Type I** – AV fistula. Direct communication between an artery and vein.
**Type IIA** - Blood flows via an abnormal nidus/tangle of abnormal smaller vessels.
**Type IIB** - Like a type IIA, but with an aneurysmal outflow vein.
**Type IIIA** - The abnormal tangle of vessels is located within a single outflow vein.
**Type IIIB** - Like a type IIIA, except with multiple outflow veins.
**Type IV** - The malformation infiltrates the adjacent non-vascular tissue. Most common in congenital AVMs.

## PATIENT'S PERSPECTIVE

## INTELLECTUAL PROPERTY RIGHTS ASSIGNMENT OR LICENCE STATEMENT

I, **Aidan Coffey** the Author has the right to grant and does grant on behalf of all authors, an exclusive licence and/or a non-exclusive licence for contributions from authors who are: i) UK Crown employees; ii) where BMJ has agreed a CC-BY licence shall apply, and/or iii) in accordance with the relevant stated licence terms for US Federal Government Employees acting in the course of the their employment, on a worldwide basis to the BMJ Publishing Group Ltd ("BMJ") and its licensees, to permit this Work (as defined in the below licence), if accepted, to be published in BMJ Case Reports and any other BMJ products and to exploit all rights, as set out in our licence author licence.

**Date: 15/06/2024**

**PLEASE SAVE YOUR TEMPLATE WITH THE FOLLOWING FORMAT:**
Submitting author's last name and date of submission, e.g. Smith_April_2022.doc