

Uterine Arteriovenous Malformation: A rare cause of massive primary postpartum haemorrhage

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Introduction

Arteriovenous malformations (AVM) occur when the normal architecture of the capillary bed is bypassed; forming an abnormal connection between the arterial and venous system ⁽¹⁾. The venous system, which is normally a low-pressure system, is subjected to high pressure arterial blood flow ⁽¹⁾. The vessels involved in this abnormal connection are significantly weaker making them prone to rupture and bleeding ⁽¹⁾. AVMs can be categorized as congenital or acquired, with congenital being considered the most common form of most AVMs ⁽²⁾. Congenital AVMs may occur as part of the following autosomal dominantly inherited syndromes; Hereditary Haemorrhagic Telangiectasia, Capillary malformation- arteriovenous malformation and Parkes Weber Syndrome⁽³⁾. AVMs can form anywhere in the tissues or organs but are most often identified in the CNS ⁽⁴⁾. Uterine AVMs are predominantly seen in pre-menopausal women and typically present varying degrees of abnormal vaginal bleeding. Large uterine AVMs may present with a pulsatile mass. Congenital uterine AVMs tend to have multiple feeding arterioles, a central nidus and multiple large draining veins⁽⁵⁾ whereas acquired uterine AVMs tend to feature single arterioles connecting with a single veins- sometimes termed AV fistulae ⁽⁶⁾. In contrast to the majority of AVMs , acquired uterine AVMs are more common and may occur in the postpartum period, after spontaneous miscarriage, medical termination of pregnancy or following uterine surgery (STOP, D+C or C-section)⁽⁷⁾

Case Report: Background

- PO.
- 33 years old.
- History of severe peanut allergy and mild asthma
- BMI 20
- IVF pregnancy for male factor infertility
- 1x UTI antenatally, otherwise uncomplicated pregnancy
- Induction of labour due to reduced growth velocity from 40th to 10th centile at 38 week growth scan

Case Report: Outcome

- After 24 hours the patient was extubated with no apparent neurological deficit. The Bakri balloon was removed after 48 hours, and no further bleeding occurred.
- Total blood products required were 10x RBC, 1x platelets 2x cryoprecipitate, 5x FFP.
- The patient was discharged on day 6 but was readmitted on day 10 with raised blood pressure and proteinuria which required treatment with antihypertensives.
- MRI scan 6 weeks postpartum demonstrated a uterine AVM being fed from the right uterine artery.



Discussion

The incidence of uterine AVM largely unknown because symptomatic AVMs may respond to medical management and therefore many cases are not investigated, additionally, the findings on grey scale ultrasound are nonspecific meaning the diagnosis is often missed⁽⁸⁾. One study of 959 women diagnosed uterine AVM in 0.73% on colour doppler US⁽⁹⁾. But O'Brien et al quoted an incidence as high as 4.5% in a study of 464 patients with abnormal uterine bleeding⁽⁷⁾. On colour doppler US uterine AVMs appear as a mass of vessels with multidirectional high-velocity flow which produce a colour mosaic pattern⁽¹⁰⁾ and spectral doppler analysis shows arteriovenous shunting with high-velocity but low-resistance flow⁽¹¹⁾. However, these features may also be present in multiple other conditions including retained trophoblastic tissue, gestational trophoblastic disease, placental polyps and vascular neoplasm which presents a diagnostic challenge as is potentially dangerous as inappropriate D+C has been shown to exacerbate bleeding from AVMs⁽¹²⁾. CT angiography is still considered the gold standard investigation but as can be seen in our case, the CT findings can still be misinterpreted; especially in the acute setting ⁽¹³⁾. For our patient, diagnosis was discovered on postpartum MRI. The findings on T1 and T2 weighted images include multiple serpentine flow-related signal voids in the uterine wall, endometrial cavity, and parametrium⁽¹⁴⁾. For patients with mild symptoms, medical management with agents including progestogens, the COCP, danazol and GNRH analogues are effective as evidenced by a meta-analysis of 121 women which demonstrated a success rate of 88% ⁽¹⁵⁾. Case reports of uterine AVMs presenting with acute life-threatening haemorrhage describe using balloon catheterization and subsequent uterine artery embolisation or emergency hysterectomy to control bleeding ⁽¹⁶⁾. Timmerman et al. discovered that on US, uterine AVMs with a peak systolic velocity value of 0.83 m/s carried a greater chance of requiring embolisation and values below 0.39 m/s never required embolisation ⁽¹¹⁾. In a study of 19 patients bilateral uterine artery embolization radiographically and clinically resolved the abnormality in 17 of the cases and all these women returned to a normal menstrual cycle ⁽¹⁷⁾. Other studies have quoted success rates of 92% and 79% although in the minority repeat embolisations were required^(5, 18). A recent literature review concluded that embolisation did not interfere with women's ability to conceive and carry a pregnancy to term as 27 women who were treated achieved 34 pregnancies and 31 live births ⁽¹⁹⁾. In current literature there are only 12 reported cases of uterine AVM resulting in PPH. The blood loss was reported between 1000ml and 3000ml and 8 out of the 12 cases required blood transfusion. Ours is the only reported case where bleeding was catastrophic enough to result in cardiac arrest. Interestingly, two of the reported patients had undergone IVF, as had our own case which raises the possibility of a link between the two ⁽²⁰⁾. Evidence is very limited with regards to the management of these women in subsequent pregnancies, but it has been anecdotally recommended that women should undergo embolization with follow up US doppler and MRI prior to the next pregnancy, and that serial growth scans should be performed due to the feared risk of intrauterine growth restriction⁽⁸⁾.

References

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Case Report: Timeline of Events

