CASE REPORT

Uterine Arteriovenous Malformation as a Cause of Secondary Postpartum Hemorrhage: A Case Report

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Abstract

Background: Uterine arteriovenous malformation (AVM) is a very rare cause of secondary postpartum hemorrhage (PPH) which might present along with retained products of conception (RPOC). Despite being a rare entity, it is a life-threatening condition and hence high degree of suspicion is required for prompt diagnosis and appropriate treatment as certain procedures, such as instrumental evacuation, commonly performed for RPOC could be devastating. With the recent advancements, color Doppler ultrasonography has become a preferred noninvasive method for diagnosing AVM though angiography remains the gold standard for diagnosis.

Case description: This case report describes a 36-year-old multiparous woman who presented 3 weeks after delivery by lower segment cesarean section (LSCS) with secondary PPH. Transabdominal ultrasound (TAS) revealed an involuting uterus with an ill-defined hypoechoic area within the myometrium adjacent to the endometrium with significant intrinsic vascularity raising the suspicion of placenta accreta. Magnetic resonance imaging (MRI) with contrast was performed for confirmation which showed an AVM with coexistent RPOC. Since the patient did not give consent for uterine artery embolization which was offered to her during counseling, hysterectomy was carried out. Histopathological diagnosis of uterine AVM was conclusive.

Conclusion: Arteriovenous malformation, although a rare entity, should be ruled out in a patient presenting with hemorrhage after delivery/miscarriage even if the imaging shows the presence of RPOC by the diagnostic modalities available so that appropriate treatment can be instituted.

Keywords: Arteriovenous malformation, Color Doppler, Hysterectomy, Secondary postpartum hemorrhage, Uterine artery embolization.

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Introduction

With less than 100 cases reported so far, uterine arteriovenous malformation (AVM) is a rare entity which can be congenital or acquired.[1,2] Failure in the embryological vascular differentiation leading to multiple vascular channels results in the formation of a congenital AVM.[3] However, most cases are acquired and result from previous uterine surgery or curettage, infection, gestational trophoblastic disease, exposure to diethylstilbestrol, malignancy of endometrium, or cervix.[4–8]

Arteriovenous malformations are extremely variable in size and location, leading to a spectrum of clinical presentations. They have been reported not only in head, neck and extremities but also in bowel, lung, spleen, stomach, pancreas, bladder, uterus and vagina. Congenital uterine AVM tends to venture into the surrounding tissues and constitutes a difficult therapeutic challenge. The bleeding caused by these malformations is more often episodic, torrential, and can lead to significant anemia or even shock warranting hospital admission.

Historically, the diagnosis of uterine AVM was made at laparotomy or upon histopathological examination of the uterine specimen following hysterectomy. Subsequently, with advances in technology, angiography became the gold standard modality for diagnosing this condition.[9] Recent reports suggest that angiography should be reserved for cases in which surgical intervention or therapeutic embolization of the lesion is planned and that color Doppler ultrasonography is the preferred method of diagnosing AVM.

Although it is not a commonly encountered condition, we have to keep in mind this rare entity while evaluating a case of secondary postpartum hemorrhage (PPH). This will avoid unnecessary surgical intervention and enable us to provide other conservative treatment options for those women desirous of retaining the uterus for reproductive or menstrual function.

In this case report, we share our experience of managing a patient who presented to us with secondary PPH. Although the initial diagnosis was retained products of conception (RPOC), magnetic resonance imaging (MRI) showed uterine AVM which was later confirmed by histopathological examination. Since the woman did not agree for conservative management, she underwent hysterectomy denying uterine artery embolization.

Case Description

A 36-year-old multiparous woman presented to our hospital with history of two bouts of heavy vaginal bleeding interspersed by periods of minimal bleeding for 1 week. She had undergone emergency lower segment cesarean section (LSCS) in view of previous LSCS with breech presentation 21 days ago. She was

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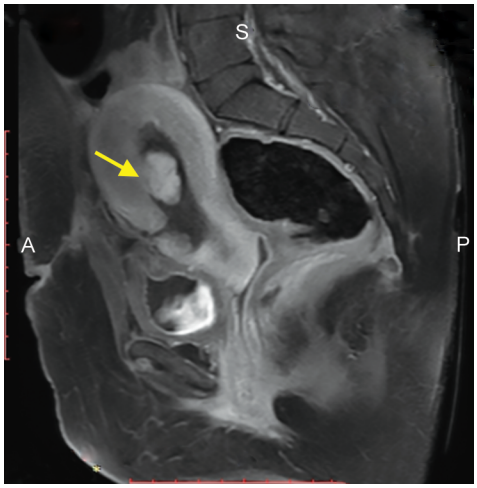

Fig. 1: Magnetic resonance imaging of pelvis with contrast showing sagittal section of uterus with soft tissue mass adherent to anterior uterine wall suggestive of retained products of conception with coexistent arteriovenous malformation

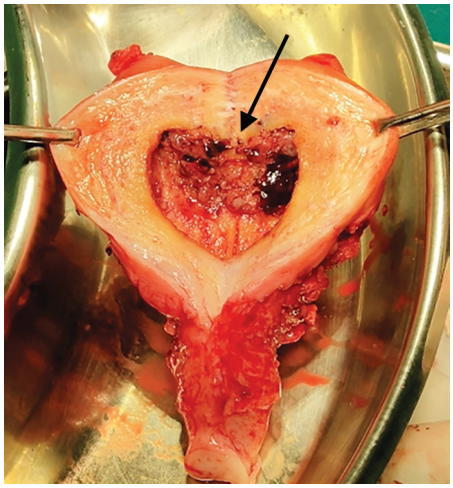

Fig. 2: Gross specimen of the uterus with posterior wall cut open showing adherent fleshy mass (2 × 2 cm) on the anterior wall near the fundus

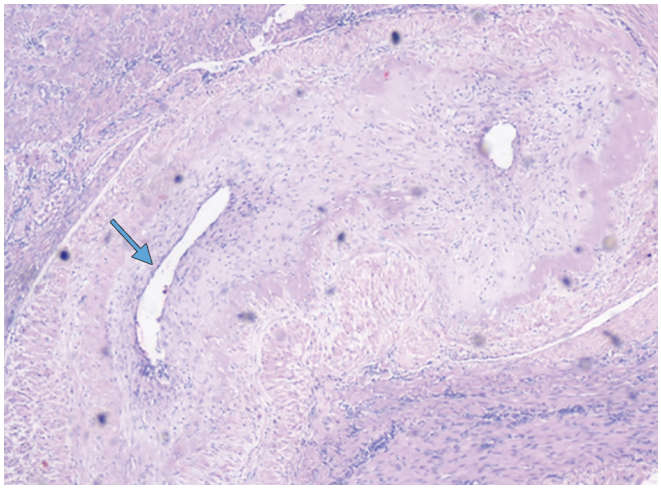

Fig. 3: Thick walled, hyperplastic vessels showing degenerative changes within the tunica media; with narrowed vessel lumen, seen within the myometrium

discharged on postoperative day 6 in a stable condition. Her obstetric history was uneventful. She had an abortion prior to her last pregnancy but there was no history of any curettage being performed. On admission, she was hemodynamically stable and hemoglobin (Hb) level was 12.6 g/dL. Moderate amount of active bleeding through the os was noted on per speculum examination and on bimanual examination, uterus was corresponding to 10 weeks size. To rule out gestational trophoblastic disease, serum beta-human chorionic gonadotropin (hCG) was performed and it was 66.4 mIU/mL. The patient experienced another bout of bleeding per vaginum after admission, following which her Hb dropped to 10 g/dL.

Suspecting RPOC, transabdominal ultrasound (TAS) was performed which showed an involuting uterus of 11.5 × 5.6 × 6.4 cm with endometrial thickness of 20 mm with an ill-defined hypoechoic area measuring 17 × 13 mm within the myometrium adjacent to the endometrium with significant intrinsic vascularity raising a suspicion of placenta accreta. Hence, MRI pelvis with contrast was performed which showed mildly bulky uterus with soft tissue mass adherent to anterior uterine wall with restricted diffusion and homogeneous enhancement, with a large feeder vessel traversing the myometrium and extending to the uterocervical junction, suggestive of RPOC with coexistent AVM (Fig. 1).

Patient was counseled regarding the various treatment options and was offered uterine artery embolization. Due to personal reasons, patient opted for hysterectomy and hence, definitive procedure was carried out. No complications were encountered. When the uterus specimen was cut open from the posterior surface, a fleshy mass of 2 × 2 cm was noted adherent to the anterior wall of uterine cavity near the fundus which could not be separated (Fig. 2). Histopathology report was suggestive of uterine AVM alone without any e/o trophoblast tissue (Fig. 3). Patient was then discharged on postoperative day 6.

Discussion

Arteriovenous malformations can be defined as vascular structural anomalies involving abnormal communication between arteries and veins, eventually bypassing the capillary system. However, there is no standard definition of a true AVM. Literature search did not give us much information on the frequency of these lesions. Since uterine bleeding due to AVM subsides on treatment with drugs in a few cases, further investigations may not be warranted and AVM may be missed. Therefore, it is difficult to determine the incidence of this condition. O'Brien et al. gave a rough incidence of uterine AVM as 4.5%.[10] Yazawa et al. noted a lower incidence on ultrasound of 0.6%.[11]

With the increasing use of ultrasound and Doppler for detection, uterine AVMs may be over diagnosed. Therefore, it has been proposed that lesions detected with hypervascular and turbulent flow be designated as uterine vascular malformations.[12] The term uterine AVM should be restricted to those lesions which on angiography demonstrate a hypervascular mass with early filling or following hysterectomy on histopathological examination of the uterus.[13]

The pathogenesis of acquired uterine AVMs can be traced back to the previous pregnancy. Hence, they are more common in women in their reproductive years, although they could occur in any age group. In women who present with abnormal excessive uterine bleeding a few months after a miscarriage or termination of pregnancy, uterine AVM has to be suspected when the pregnancy

test is negative.[14] Infection, inflammation, RPOC, gestational trophoblastic disease, and gynecologic malignancies could also be risk factors for AVMs.[15]

Congenital AVMs, which are not as common as acquired AVMs, have been thought to be due to failure of embryological differentiation where the vessels may invade the adjacent structures.[3] In contrast, acquired malformations may arise when the venous sinuses get incorporated in the scars within the myometrium following the necrosis of chorionic villi.[14] Acquired AVMs lack a characteristic nidus and can be supplied by one or both uterine arteries.[10]

In a patient with uterine AVM, if surgical treatment is resorted to consider the diagnosis to be RPOC or trophoblastic disease, there could be catastrophic vaginal bleeding which could result in emergency hysterectomy and eventual increase in morbidity and mortality. Therefore, every attempt has to be made to diagnose uterine AVMs using available facilities.[16]

Uterine AVMs were initially diagnosed when histopathological examination of uterus was performed in women who underwent hysterectomy for excessive vaginal bleeding.[17] Recently, ultrasound has been used for screening and early diagnosis of this condition and color and spectral Doppler are the tools which help us determine appropriate treatment plan. Doppler in uterine AVM shows low impedance in uterine artery.[18] Differential diagnoses include subinvolution of the placental bed and adenomyosis which appear similar to AVMs on ultrasound.[19] Due to limitations of ultrasound, MRI is recommended as it provides a better tissue contrast and helps delineate the adjacent pelvic organ involvement. Digital subtraction pelvic angiography not only gives the accurate diagnosis but also identifies the main feeding vessels which helps in embolization. Therefore, it is considered the gold standard for diagnosis of uterine AVMs.[13]

Treatment is decided after taking into consideration the patient's age, degree of bleeding, hemodynamic stability, and desire for future fertility. If the patient is not hemodynamically stable, resuscitation is the first line of management. Once the patient is hemodynamically stable, further treatment is determined by the desire for future fertility and ability for close follow-up. Young women desirous of pregnancy may be administered medical management and followed-up to know the response. There are case reports of successful medical management of uterine AVM. An 18-year-old woman with acquired uterine AVM was given a course of combined oral contraceptive pill. There was complete cessation of bleeding 6 days after starting the pill. On day 37, computed tomography (CT) scan was performed and no abnormal vasculature was seen.[20] Various other medications which have been considered in the treatment are danazol, progesterone, implanon, and methotrexate.

Among the minimally invasive surgical techniques, transcatheter embolization (TCE) has proven to be an effective option for patients desirous of future fertility. The embolic agents used include polyvinyl alcohol, histoacryl (glue), detachable balloons, microspheres, and hemostatic gelatin. Some cases may require repeat embolization for additional treatment. This is not considered as due to failure of initial embolization. Success rate of embolization is 61% after one embolization and 91% after repeat embolization.[21]

Other minimally invasive surgical techniques have been tried for treatment of uterine AVMs when embolization has failed. These include laparoscopic occlusion of the internal iliac arteries using non-resorbable clips[22] and unilateral or bilateral laparoscopic bipolar coagulation of uterine arteries.[23] When the conservative measures fail or if the patient is not willing to come for follow-up, hysterectomy should be considered with 100% success rate.

For asymptomatic AVM, there are no definitive guidelines as to whether they require treatment or need follow-up and the modalities for the same. Furthermore, there is no robust data comparing different modalities of treatment described above for symptomatic uterine AVM. More studies are needed to compare the short- and long-term outcome of different conservative modalities of treatment of uterine AVMs.

This case report highlights that the cause of secondary PPH could be AVM. Although it is a rare entity, the treating clinicians should be aware of it. It also highlights that, an understanding of the pathophysiology of uterine AVM and its prompt identification followed by appropriate treatment in symptomatic patients is of utmost importance because inadvertent curettage can result in massive hemorrhage and unplanned hysterectomy with significant morbidity.

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