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CASE REPORT

Uterine Arteriovenous Malformation: A Near-Missed Fatal Misdiagnosis?

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ABSTRACT

Less than 100 cases of uterine arteriovenous malformations (AVM) have been documented. The true incidence remains unknown with 30% complicated by hypovolemic shock. Women who experience unexplained vaginal bleeding may consider this diagnosis. We chose to emphasize this case because, while being a rare disease, it could cause mortality if misdiagnosed. In this case, a 23-year-old lady, para 1+2 with a history of several miscarriages and curettage, complained of per vaginal bleeding more than 2 months after her suction and curettage on September 2018. Her second visit was managed as having retained the product of conception and another curettage was conducted. Despite this, the patient still has worsening vaginal bleeding which leads to her third visit. A bedside ultrasound was conducted and suspected uterine AVM. This was then further confirmed with contrast-enhanced computed tomography of the pelvis and pelvis angiography. Five Micronester coils and 10% Histoacryl adhesive were used to successfully embolize the uterine artery. This case report served as a valuable lesson on the importance of having a proper diagnosis and prompt treatment to avoid fatal misdiagnosis which could have disastrous consequences.

INTRODUCTION

Dubreil and Loubat published the first description of uterine arteriovenous malformations (AVM) in 1926 (Evans et al., 2017). In women who are not pregnant, AVMs are rare. Although less than 100 occurrences were reported, 30% of those people required blood transfusions due to hypovolemic shock (Polat et al., 2002; Manolitsas et al., 1994; Hasegawa et al., 2012). Therefore, women who experience unexplained vaginal bleeding may need to consider this diagnosis. We chose to emphasize this case because, while being a rare disease, it could cause mortality if misdiagnosed.

AVMs of the uterus can result in deadly vaginal bleeding due to intervillous space enlargement deep into the myometrium. This enables direct passage without the involvement of capillary vessels from the arterial system to the venous system (Evans et al., 2017; Farias et al., 2014). Uterine AVMs can be acquired or congenital. An anomaly in the embryologic development of early vascular structures gives rise to congenital uterine AVMs (Kwon & Kim, 2002). The causes of acquired uterine AVMs include pelvic trauma, surgery (including cesarean section and curettage), malignancy, and infection. Multiple feeding arteries, a central nidus of vessels with characteristics of both arteries and veins and numerous big draining veins are the structural features that distinguish structurally congenital uterine AVMs. Contrarily, acquired uterine AVMs are made up of one or more arteriovenous fistulas (AVFs) between the myometrial venous plexus and intramural arterial branches. Due to the similarity of their radiologic findings, congenital uterine AVMs and acquired uterine AVFs must be distinguished by the patient's medical history (Lalitha et al., 2015; Timmerman et al., 2000). We highlight a 23-year-old lady, post suction and curettage complicated with uterine AVMs which was successfully embolized and we discussed our management strategy for managing this fatal condition.

CASE REPORT

A 23-year-old lady, para 1+1 with a history of missed miscarriage in 2014 (required dilatation and curettage) and emergency caesarean section for breech in labour (2015), was admitted for a second miscarriage in September 2018. Suction and curettage were done with an estimated blood loss of 1.8 litres. However, two months later, the patient still complained of per vaginal (PV) spotting. The patient denied having any fever or abdominal pain. On bedside ultrasound examination, noted a thickened endometrial wall and elevated beta-HCG level. It then proceeded with second suction and curettage as it was initially presumed there were retained products of conception (POC) which was complicated with an estimated blood loss of 1.3 litres. After the procedure, the patient still complained of having per vaginal bleed (quarterly soaked 3 pads/day) and seek gynaecology consultation again when it worsened to fully soaked 2 pads/day.

Bedside ultrasound (Figure 1) was then repeated by the gynaecology team which noted there was a hypoechoic lesion at the left side of the uterine wall in the transabdominal scan and multiple short tubular-like hypoechoic structures seen at the uterine wall in the transvaginal scan. There was no residual product of conception (POC) detected. The constellation of these findings of a hypoechoic lesion surrounded by multiple tubular-like structures raised the suspicion of uterine AVMs. However, the overall findings and raised beta-HCG level also raised the suspicion of possible gestational trophoblastic disease. Hence, a computed tomography (CT) of the pelvis was done to assess further the hypoechoic lesion at the left uterine wall.

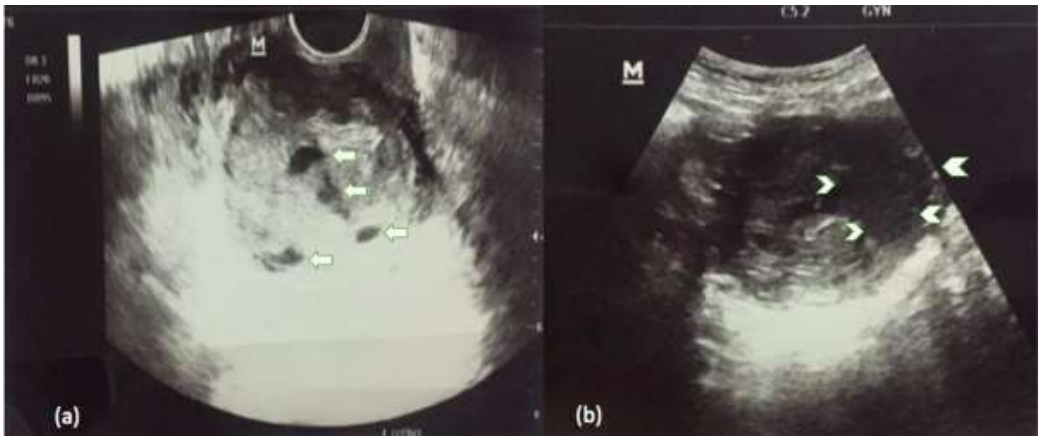

Figure 1 (a) Transvaginal scan shows multiple tubular hypoechoic structures at the uterine wall (white arrows). (b) Transabdominal scan shows a hypoechoic lesion noted at the left side of the uterine wall which raised the suspicion of uterine mass or if correlating with the tubular-like structure in (a), this raised the suspicion of a possible uterine AVM nidus (arrowheads)A contrast-enhanced CT of the pelvis (Figure 2) showed tortuous dilated vessels noted at the parametrium around the uterus with some seen traversing the hypodense lesion. There is a hypodense lesion arising from the left anterolateral wall of the uterus measuring 5.7 × 8.6 × 5.4 cm (AP × W × CC). No calcifications were noted within. Both ovaries are normal. The tortuous dilated vessels raised the suspicion of uterine arteriovenous malformation while the hypodense lesion may represent a uterine fibroid.

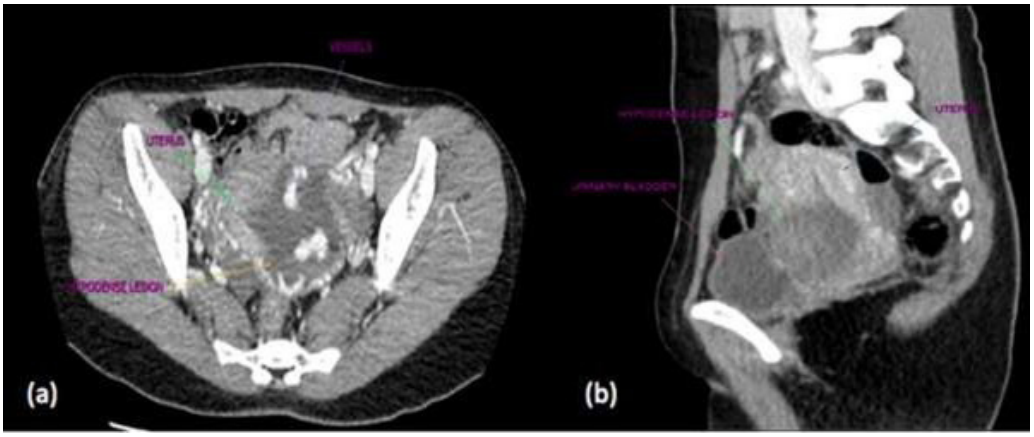

Figure 2 Contrast-enhanced CT of the pelvis at (a) axial view showing a hypodense lesion with tortuous dilated vessels seen traversing within. Dilated vessels are also seen at the parametrium around the uterus. The hypodense lesion pushed the uterus towards the right side. At sagittal view (b), the hypodense lesion was seen with its epicentre within the anterior uterine wall protruding into the uterine cavity

The decision to proceed directly with a diagnostic pelvic angiogram instead of a CT angiogram was made after the patient began to show worrying vital signs and haemoglobin level after the CT Pelvis. This was then followed by therapeutic embolization in the same setting after confirming the arteriovenous malformation in the pelvic angiogram. Pre-embolization (Figure 3a) noted arteriovenous malformation supplied by three feeding arterial branches from the left ascending uterine artery and drained into the nidus and uterine venous plexus. Post embolization (Figure 3b) with 5 Micronester coils and 10% Histoacryl glue noted successful occlusion of the left ascending uterine artery AVM. No AVM was seen at the right uterine artery.

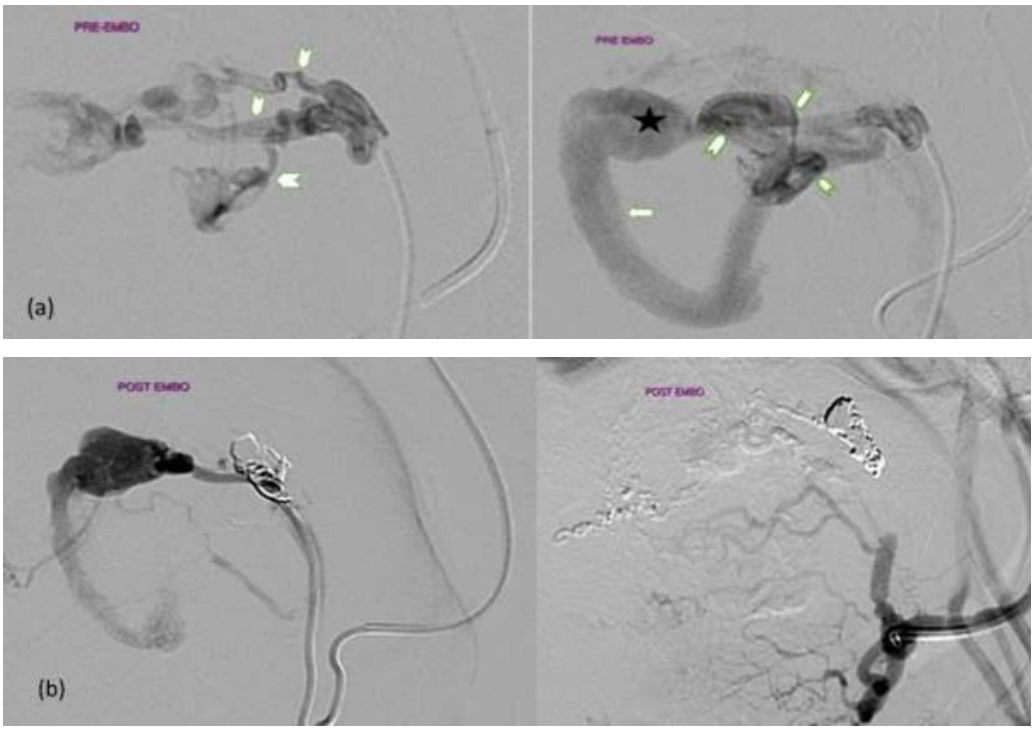

Figure 3 Pre-embolization images (a) showing three feeding arterial vessels (arrowheads) from the left ascending uterine artery draining into the nidus (star-shaped) and the venous plexus (arrow). Post- embolization images (b) showing a successful occlusion of the left ascending uterine artery AVM with 5 Micronester coils and 10% Histoacryl Glue. No AVMs were seen at the right uterine artery

DISCUSSION

The proposed incidence of 4.5% occurred at the mean age of 30 years old with 96% premenopausal and 4% postmenopausal. The most typical presenting symptom is menorrhagia. Some authors assert that the uterus may have enlarged with noticeable pulsation during bimanual examination. A vascular "steal" pattern with considerable blood shunting from the arterial to the venous circulation will appear in serious cases. This circumstance might lead to serious circulatory disturbance and cardiac insufficiency which may complicate large AVM (Evans et al., 2017; Farias et al., 2014). However, instead of menorrhagia, our patient just complained of prolonged vaginal bleeding. Her stomach and vulva/vaginal examinations were unremarkable as well. Our patient suffered massive blood loss in two separate suction and curettage events before she was confirmed to have uterine AVM. This is considered to be dangerous as the massive blood loss might turn into a catastrophic event of hypovolaemic shock or death secondary to uterine AVM (Evans et al., 2017; Polat et al., 2002).

Ultrasound is usually performed as the initial imaging assessment as it is easily accessible and non-invasive. As noted in our patient, the greyscale ultrasound shows subtle multi-foci of hypoechogenicity scattered within the thickened myometrium. Colour Doppler ultrasonography should show serpiginous/ tubular anechoic structures within the myometrium with rapid and turbulent blood flow in a mosaic pattern. Spectral analysis of the arterial vessels within the lesion might show high-velocity flow with a low resistive index (0.51 – 0.65) (Evans et al., 2017; Polat et al., 2002). However, unfortunately, no Doppler examination was done for our patient by the primary team.

Along with doing an ultrasound as a preliminary examination, pelvic magnetic resonance imaging and CT angiography

are crucial for supporting the confirmation of the uterine AVM diagnosis (Evans et al., 2017). Contrast-enhanced examinations will demonstrate the enhancement of the collection of serpentine vessels which will enhance as intensely as normal vessels in the arterial phase. These findings will be accompanied by evidence of early venous return which is signified by opacification of venous structures on the arterial phase images (Evans et al., 2017). Even though our patient did not have a CTA examination, the dilated and tortuous vessels traversing the hypodense lesion and within the bilateral parametrium regions seen in the contrast-enhanced CT of the pelvis, increased the degree of suspicion of an arteriovenous malformation. This suspicion was then further confirmed with a diagnostic pelvic angiogram and proceeded with therapeutic embolization in the same setting.

The recommended gold standard is an angiography, especially in patients who might eventually undergo embolization. Since 1986, surgical intervention for uterine AVMs has been replaced with embolotherapy, which is now widely accepted. The preferred course of treatment for patients with significant uterine bleeding or in whom fertility is not the primary concern is hysterectomy, particularly in post-menopausal women due to the 17% recurrence rate (Polat et al., 2002; Hasegawa et al., 2012).

It is still unclear whether the pregnancy is safe after uterine AVMs have been successfully embolized. Pregnancies following effective embolotherapy are uncommon. This is due to the decreased placental vascularization that occurs after embolotherapy, which causes an unfavourable pregnancy outcome (Evans et al., 2017). However, some studies showed there is a 27% success rate for pregnancies after post-uterine artery embolization (UAE). According to their findings, after embolization, sufficient collateral supply can form to support a full-term pregnancy. A retrospective analysis reveals that during 10 years, five unremarkable intrauterine pregnancies were successfully carried to term in 4 of 15 patients who underwent pelvic arterial embolization for traumatic uterine AVMs. They concluded that percutaneous embolotherapy is a secure and efficient treatment for traumatic uterine AVMs while maintaining the chance of a subsequent pregnancy (Evans et al., 2017, Lalitha et al., 2015; Timmerman et al., 2000).

The most frequent adverse reaction that results from myometrium temporary ischemia is pelvic discomfort. The pain might occur right away and can last anywhere between 12 and 24 hours. As a result, it is typical for patients to need an overnight stay with proper pain management following the treatment. Post-embolization syndrome, which lasts a few hours to a few days and includes fever, discomfort, malaise, and nausea, is another common morbidity in the UAE. The syndrome frequently develops following the embolization of any solid organ and is assumed to be an immune-mediated reaction. Approximately 50% of patients are said to experience it, but controllable with analgesics, antipyretics, and anti-inflammatories (Memtsa & Homer, 2012).

A more severe complication is uterine infection, and patients may experience rapid onset severe pain, vaginal discharge, and/or bleeding. Since infection can sometimes result in systemic sepsis and necessitate hysterectomy, it must be aggressively treated. Rarely, sepsis-related deaths have also been reported. Fortunately, it is a rather uncommon complication that only occurs in less than 1% of cases (Memtsa & Homer, 2012). Pulmonary embolism is an uncommon but potentially fatal consequence of UAE that is hypothesized to be caused by a transient hypercoagulable state that is similar to but less severe than that seen during surgical procedures. Before undergoing UAE, it is wise to conduct a thromboprophylaxis risk assessment because the incidence is thought to be 1 in 400 (Memtsa & Homer, 2012). The ovaries are most likely to be affected by off-target organ embolization. Although the vaginal artery typically develops

as a distinct branch of the internal iliac artery, it occasionally may share a common trunk with the uterine artery or form anastomoses within the broad ligament. In such cases, the vagina is susceptible to embolization-induced ischaemia, which can cause sexual dysfunction and/or dyspareunia (Memtsa & Homer, 2012). Despite everything stated above, multicentre trials consistently demonstrate that the rate of major adverse events following UAE is significantly lower than that of more conventional surgical interventions like myomectomy and hysterectomy (Memtsa & Homer, 2012).

Although most of the complications start to occur within the first month post-procedure; long-term monitoring is also required to identify any developing complications and determine whether or not the therapy was successful. In large clinical trials, the standard follow-up is 3, 6, 12, 21, or 24 months, and then annually after that. In one of the studies done, the patients were followed up with phone calls for 6 months, at which time patients returned to the department for follow-up imaging. The most accurate way to document interval change is through imaging modality continuity. If the patient had ultrasound imaging done before the procedure, the same method of re-assessment should be used for post-procedure imaging follow-up. However, till now there are currently no clear recommendations for long-term follow-up (Carrillo, 2008).

CONCLUSION

In conclusion, uterine AVMs are still uncommon since they are only identified in symptomatic individuals who are pregnant or have recently experienced a miscarriage. The rarity of this disease and the scarcity of relevant literature have caused a lack of awareness among medical professionals, which eventually may lead to a fatal misdiagnosis. This case may have been considered a near–missed situation when the patient was presumed to have retained the product of conception during her second visit for persistent per-vaginal spotting. Fortunately, her third visit with worsening symptoms triggered a high clinical suspicion of this disease and extensive imaging studies were conducted. The patient was then successfully treated before more catastrophic complication occurs. Therefore, from this case report, we have gained a valuable lesson on the importance of having a proper diagnosis and prompt treatment to avoid fatal misdiagnosis which could have disastrous consequences.

CONFLICT OF INTEREST

The authors declare that they have no competing interests in publishing this case report.

CONSENTS

Written informed consent was obtained from the patient to publish the case. A copy of the written consent is available for review by the Chief Editor.

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