Uterine arteriovenous malformation in pregnancy: A case report*

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ABSTRACT

Background. Uterine arteriovenous malformation (AVM) is a web of arteries and veins lacking an intervening capillary network. Color flow Doppler is a popular method of diagnosis of uterine AVM. The definitive management is hysterectomy. However, for patients desirous of pregnancy, transarterial embolization is a safe and effective option. Although rare, uterine AVM can complicate pregnancy with torrential bleeding due to hormonal changes and significant remodeling of the myometrium.

Case. We report a case of a term pregnancyin a 33 year old with a uterine AVM and a previous transarterial embolization procedure who developed a uterine AVM during multi-agent chemotherapy for gestational trophoblastic disease. She consulted for prenatal checkup. Due to the risk of massive bleeding during labor, she underwent elective cesarean section at term and delivered a baby with good outcome.

Conclusion. This case suggests that uterine AVM in pregnancy can be managed conservatively with serial ultrasound monitoring and close follow up.

Keywords: uterine arteriovenous malformation (AVM), transarterial embolization, pregnancy, gestational trophoblastic neoplasia

INTRODUCTION

Uterine arteriovenous malformation (AVM) is a rare vascular disease characterized by a vascular tangle of arteries and veins without a communicating capillary network.[1] The vessels have thin muscular walls and are prone to rupture. Early diagnosis is important to prevent life-threatening hemorrhage. The definitive management of uterine AVM is hysterectomy. However, for patients who are still desirous of pregnancy, transarterial embolization poses as a viable alternative. Poor pregnancy outcome such as preterm deliveries and abortions are common after transarterial embolization. Although rare, uterine AVM can co-exist and complicate pregnancy. Remodeling of the myometrium and ensuing uterine contractions can cause rupture of the arteriovenous malformation resulting in torrential bleeding.

This report describes a rare case of successful term pregnancy after transarterial embolization for an asymptomatic uterine arteriovenous malformation as a sequela of gestational trophoblastic neoplasia. This paper emphasizes the importance of conservative management of uterine AVM particularly among patients who wish to preserve their fertility. Emphasis is also placed on proper prenatal counseling and evaluation to determine the appropriate route of delivery and ensure a successful pregnancy outcome.

CASE HISTORY

This is the case of a 33 year old Gravida 2 Para 0 (0010) who was admitted at our institution for elective cesarean section.

Six years prior to this admission, the patient underwent suction curettage for a complete hydatidiform mole. Two months after, the patient was diagnosed with Gestational Trophoblastic Neoplasia (GTN). After 12 cycles of Methotrexate chemotherapy there was note of plateauing βhCG values. The patient was asymptomatic and on internal examination, the cervix was 2x2 cm, firm smooth, the corpus was small, with no adnexal masses or tenderness. A pelvic ultrasound was done which showed an anechoic vascular cystic mass measuring 4.6 x 4.5 x 4.1 cm at the right posterolateral wall which on color flow mapping showed hypervascularity, suggestive of gestational trophoblastic neoplasia. The serum βhCG was 422 mIU/ml. She received multiple agent chemotherapy in the form of Etoposide, Methotrexate, Actinomycin, Cyclophosphamide, Oncovin (EMACO) with adequate response. Complete remission was achieved after 5 cycles of chemotherapy. Serial pelvic sonogram during treatment showed persistence of the vascular uterine mass despite

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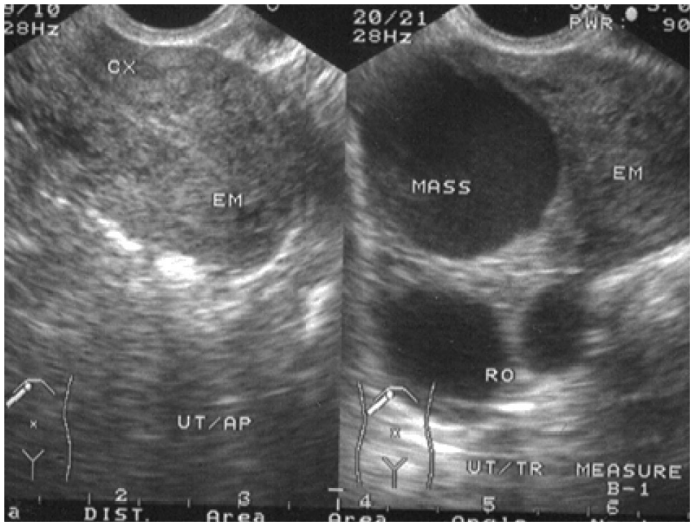

Figure 1. Pelvic ultrasound after 5 cycles of multi-agent chemotherapy which showed an irregular complex mass measuring 5.6 x 3.1 x 3.6 cm located at the right posterolateral wall, which on color flow mapping, showed turbulent vascular flow.

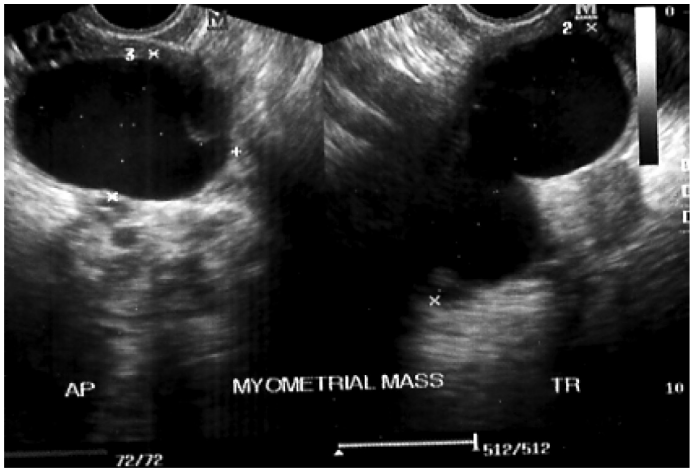

Figure 2. Pelvic ultrasound done one year after remission showed an increase in the size of the uterine arteriovenous malformation to 8.1 x 5.8 x 3.9 cm (from 5.6 x 3.1 x 3.6 cm). It extended to the right parametria and right infundibulopelvic vessels. Color flow mapping of the said mass showed hypervascularity and turbulent flow. The sonologic impression was myometrial mass consistent with arteriovenous malformation as a sequela of gestational trophoblastic neoplasia; potentially high risk for spontaneous rupture.

adequate response to chemotherapy. After the last course of chemotherapy, repeat sonography showedpersistence of the uterine mass at the right posterolateral wall, which on color flow mapping showed turbulent vascular flow (Figure 1). Because of the normal βhCG titers, the impression was Uterine Arteriovenous Malformation as a sequel of Gestational Trophoblastic Neoplasia. The patient remained in remission. Monthly pelvic ultrasound showed no increase in the size of the uterine AV malformation for 11 months.

One year from remission, during routine pelvic ultrasound, there was note of an increase in the size of the uterine arteriovenous malformation by almost 100% (Figure 2). It extended up to the right parametria and right infundibulopelvic vessels. Color flow mapping of the said mass showed hypervascularity and turbulent flow, potentially high risk for spontaneous rupture. Abdominal computed tomography scan with contrast was done which showed a vascular malformation, likely right uterine artery in origin, with aneurysmal dilatation of its draining vein (Figure 3). She underwent embolization of the right uterine artery and the other feeding branches from the anterior trunk of the right internal iliac artery using PVA particles, gelfoam and silk particles until total occlusion was observed (Figure 4) which decreased the size of the uterine AV malformation by approximately fifty percent. Upon discharge, the patient was advised regular ultrasound monitoring of the lesion. However, the patient was lost to follow-up.

Two weeks prior to this admission, at 34 weeks age of gestation, the patient sought prenatal consult for the first time at our outpatient clinic. Pelvic ultrasound showed a

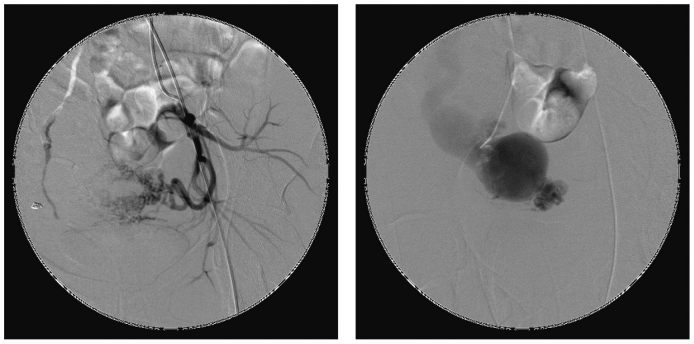

Figure 3. Abdominal computed tomography scan with contrast done one year after remission which showed a vascular malformation, likely right uterine artery in origin, with aneurysmal dilatation of its draining vein.

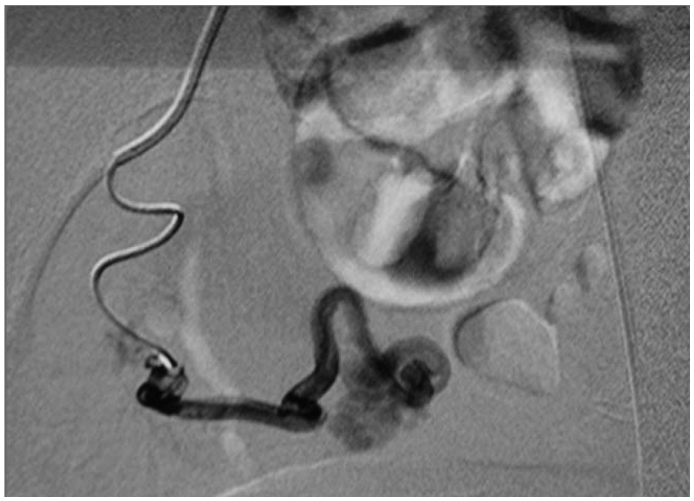

Figure 4. Embolization of the uterine artery using PVA particles (355-500 u), gelfoam and silk particles.

single live intrauterine pregnancy, in cephalic presentation with good cardiac and somatic activities. There was note of a vascular dilatation at the right lateral myometrium at the level of the lower uterine segment measuring 4.6 x 4.5 x 4.1 cm which on color flow mapping showed abundant vascularity with mosaic pattern, consistent with an arteriovenous malformation (Figure 5). The patient was counseled regarding risks of bleeding for the mother and the baby during delivery. Because of the risk of rupture and bleeding of the arteriovenous malformation during labor and delivery, the team arrived at a consensus to do elective cesarean section at 38 weeks age of gestation with possible bilateral internal iliac artery ligation.

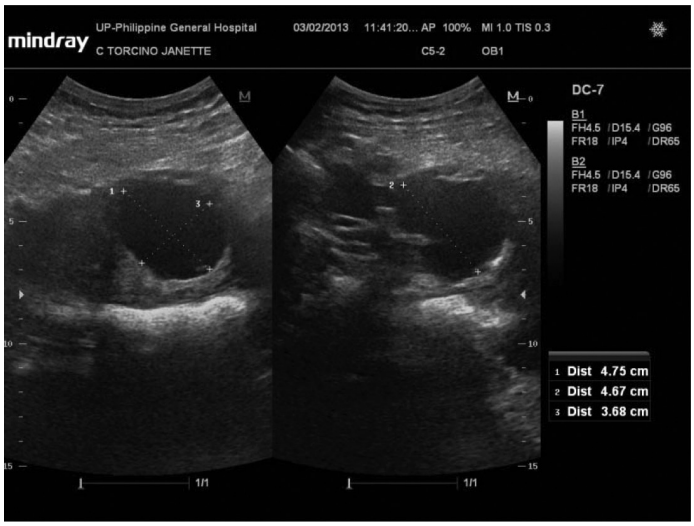

Figure 5. Pelvic ultrasound at 37 weeks age of gestation showed a vascular dilatation at the right lateral myometrium at the level of the lower uterine segment measuring 4.6 x 4.5 x 4.1 cm which on color flow mapping showed abundant vascularity with mosaic pattern. This was consistent with an arteriovenous malformation.

On admission, the patient was ambulatory with stable vital signs, normal systemic physical examination findings, and good fetal heart tones. The admitting impression was Pregnancy Uterine, 38 weeks age of gestation, cephalic not in labor, status post suction curettage for a Complete Hydatidiform mole (2006); Gestational Trophoblastic Neoplasia III:8, in remission; status post Methotrexate XII (2006-2007); status post EMACO V (May 2007 – September 2007); Uterine AV malformation; status post uterine artery embolization (2008); Gravida 2 Para 0 (0010).

Intraoperatively, there were dilated and tortuous vessels at the right lower uterine segment which seemed to arise from the right uterine artery with communication to the infracolicomentum and the right pelvic sidewall (Figure 6). A classical uterine incision was done to avoid the vascular mass. The patient delivered a term baby girl, 2700 grams, with an APGAR score of 9 remaining 9. The

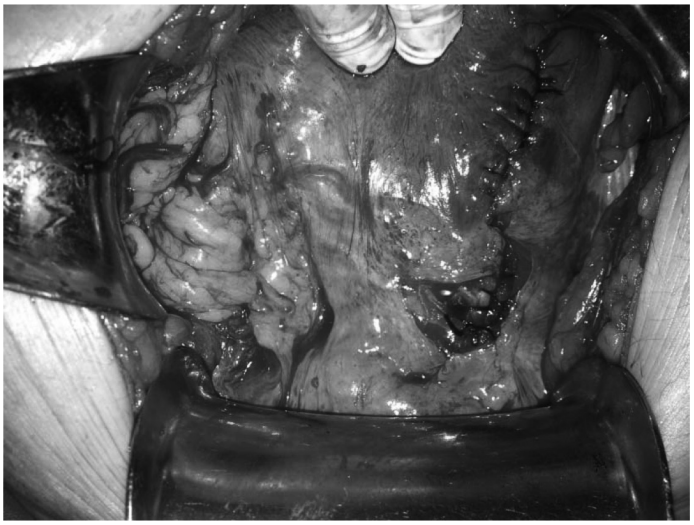

Figure 6. Intraoperative findings showed dilated and tortuous vessels at the right lower uterine segment which seems to arise from the right uterine artery with communication to the infracolicomentum and the right pelvic sidewall.

total blood loss was 700 cc. She had an unremarkable postoperative course and was discharged on her 4th postoperative day.

Serum ßhCG determination done at six weeks and six months postpartum were both normal at <0.100mIU/ml. Pelvic ultrasound done six months postpartum still revealed the uterine AVM measuring 5.3 x 4.1 x 3.7 cm. There were no episodes of abnormal uterine bleeding. She is on regular follow-up at the Section of Trophoblastic Diseases with B-HCG determination and pelvic ultrasound every 6 months.

DISCUSSION

Uterine arteriovenous malformation (AVM) is a rare vascular disease that was first described by Dubreil and Loubat in 1926.[1] Uterine AVM is a tangle of varied size vessels with arterial and venous histologic characteristics but without evidence of an intervening capillary network.[2] It can be either congenital or acquired. Congenital uterine AVM result from failure in embryological vascular differentiation. Acquired uterine AVMs, on the other hand, are mainly due to previous uterine trauma or surgery, endometrial or cervical carcinoma, maternal diethylstilbestrol (DES) exposure, or from gestational trophoblastic disease.[3]

Uterine AVM developing after gestational trophoblastic neoplasiaoccur in around 10-15% of patients.[2,5,6] Arteriovenous communications in malignant trophoblastic diseases develop as a result of destruction of the uterine vasculature by trophoblastic proliferation and invasion of the endometrium, myometrium and spiral arteries, leading to the development of abundant small

vessels that penetrate the invading trophoblast coupled with a prominent arteriovenous shunt.[13]

Diagnosis

The most common clinical presentation is heavy and prolonged menstrual bleeding.[3] The increased propensity to bleed may be initiated by hormonal changes such as pregnancy, menstruation or estrogen and progestin therapy. Bleeding usually occurs when the abnormal vessels are exposed due to sloughing of the endometrium during menstruation or iatrogenically during endometrial curettage.[7] Clinical examination are often unremarkable but may sometimes reveal a pulsating uterine mass with an audible bruit.[1]

The gold standard for diagnosis is angiography[3] but the use of transvaginal ultrasound with color flow Doppler is a minimally invasive, accessible and cheaper alternative. One characteristic of uterine AVM on gray scale ultrasonography is subtle myometrialinhomogeneity with many distinct, small anechoic spaces in the thickened myometrium or endometrium, which are focally or asymmetrically distributed. Color flow Doppler will show a tangle of tortuous vessels that are multidirectional, high-velocity and low resistance, and are focally or asymmetrically distributed. There is a mosaic pattern of color with apparent flow reversal of juxtaposed reds and blues with different flow directions.[7] Color flow Doppler combined with history and physical examination can be sufficient to diagnose cases of uterine AVM.

Uterine AVM and GTN have similar ultrasonologic features henceserum βhCG determination is warranted. For this patient, there was note of a persistent vascular uterine mass despite normal serum βhCG values, hence the diagnosis of uterine AVM was entertained and subsequently confirmed by computed tomography angiography.

Conservative Management of Uterine Arteriovenous Malformation

The definitive treatment of uterine AVM involves hysterectomy for patients who are not desirous of future pregnancy or those presenting with severe hemorrhage.[3] In recent years, arterial catheterization and embolization of the uterine artery has been successfully employed in the management of AVMs. It is now considered the primary therapeutic option when preservation of reproductive capacity is desired.[3] Transarterial embolization is a safe and effective conservative treatment for arteriovenous malformations.[2,5,8] Compared to hysterectomy, it is associated with lower morbidity, shorter hospital stay and uterine preservation.[10] In a study by Ghai et al in 2003, the success rate of transarterial embolization was 93% where in bleeding was controlled in 14 out of 15 patients.[3] McGrath et al reported the success of embolization of uterine AVM in patients with gestational trophoblastic tumor at Charing Cross Hospital from 2000-2009.[2] Embolization achieved control of hemorrhage in 18 out of 19 patients. Risks involved in the procedure include uterine ischemia and necrosis. A major disadvantage with transarterial embolization is the high cost of the procedure as well as the need for 2 to 3 sessions before complete success is achieved.

For the case presented, no intervention was considered immediately after the diagnosis of uterine AVM since she was asymptomatic. Intervention was deemed necessary when there was note of a rapid increase in the size of the AVM. Bilateral internal iliac artery ligation ortransarterial embolization of the uterine artery was offered. The risks, complications, and success rates were thoroughly discussed with the patient and her relatives. She opted for the latter option. Embolization of the right uterine artery and the other feeding branches from the anterior trunk of the right internal iliac artery was done using PVA particles, gelfoam and silk particles until total occlusion was observed. This decreased the size of the uterine AVM by 50%. She was then advised regarding the need for another embolization.

Pregnancy and Fertility Outcomes following Transarterial Embolization

Literature is sparse on the occurrence and outcome of pregnancy following arterial embolization of a uterine AVM. There is decreased vascularity of areas treated with arterial embolization causing compromised placental development and fetal growth. Poor pregnancy outcome such as preterm deliveries and abortions are common.[9]Chia et al (2003) reported a case of a successful term pregnancy with good outcome following embolization of a uterine AVM. She presented with menometrorrhagia and a 2.6 x 2.0 x 2.3 cm cystic mass on the lower right uterine wall which on Doppler sonography showed intracavitary blood flow with arterial and venous waveforms. Embolization was done successfully using a mixture of histoacryl and lipiodol. She conceived spontaneously 9 months after the procedure. Garner et al (2002) reported another case of successful term pregnancy after selective arterial embolization of a symptomatic uterine AVM in the setting of Gestational trophoblastic tumor.[5] In the series of patients who underwent embolization at Charing Cross Hospital, nine out of 18 patients who had a successful embolization were able to conceive and deliver healthy infants.[2]

The postulated mechanism by which the pregnancy is carried to term is the substantial collateral circulation of the uterus which prevents infarction despite total occlusion of the uterine artery during arterial embolization. It is also

thought that post-embolization arterial recanalization occurs to provide enough blood supply to the uterus to maintain the growing uterus to term.

For our patient, since the pregnancy occurred 5 years after the embolization, it is speculated that the uterus already developed sufficient blood supply to accommodate the pregnancy either through collateral circulation or recanalization of the occluded vessels.

Uterine Arteriovenous Malformation in Pregnancy

Currently, only a handful of cases describing the presence of a uterine AVM during pregnancy have been reported in literature. As such its clinical course during pregnancy as well as the optimal timing and route of delivery is still unknown. Garner and associates reported a case of uterine rupture with massive hemorrhage at 38 weeks age of gestation managed by cesarean section with subsequent oversewing of the AVM.[4] Another case was reported by Kelly in 2001 where the patient underwent preterm labor and delivered vaginally at 34 weeks with no complications.[6] Castro-Aragon et al in 2004 reported a case of a 29 year old primigravid who had an incidental finding of a 3x5 cm AVM during routine first trimester ultrasound.[4] The patient underwent monthly ultrasound which showed no significant change in the size of the uterine AVM. She delivered a live baby girl via elective low transverse cesarean section at 39 weeks. For our patient, her first prenatal checkup at our institution was already at 34 weeks age of gestation. Ultrasound with Doppler studies showed an arteriovenous malformationat the level of the lower uterine segment. Hormonal changes in pregnancy as early as the first trimester can trigger sudden hemorrhage of the AVM. When patients go into labor, significant remodeling of the myometrium with thinning of lower uterine segment may result to vulnerability of the vascular mass. Subsequent uterine contractions resulting in increased intrauterine pressure can cause rupture of the arteriovenous malformation. Due to the potential risk of bleeding and rupture of the uterine AVM when the patient goes into labor, the team arrived at a consensus to do abdominal delivery at 38 weeks age of gestation. She delivered by primary classical cesarean section to a term baby with good outcome. The case presented is different from the three reported cases because our patient was asymptomatic at presentation, the uterine AVM was a sequela of gestation trophoblastic neoplasia, and she underwent a previous transarterial embolization for the uterine AVM.

SUMMARY

In summary, reported is a case of a term pregnancy in a patient with an acquired uterine arteriovenous malformation with a previous transarterial embolization procedure. Uterine arteriovenous malformation is a rare vascular anomaly. Its early detection is vital as it may present with life-threatening hemorrhage. Pelvic ultrasound with Color flow Doppler is a popular non-invasive method of diagnosis uterine AVM. Doppler studies will show a tangle of tortuous vessels with a mosaic color pattern. Acquired uterine AVM can develop from gestational trophoblastic neoplasia, particularly those undergoing multi-agent chemotherapy, because of faulty angiogenesis. Transarterial embolization is a safe and effective treatment option that has the advantage of preserving the uterus. A successful pregnancy after transarterial embolization is possible through sufficient collateral circulation and recanalization of occluded vessels. Although rare, uterine arteriovenous malformation can occur with pregnancy. Hormonal changes as early as the first trimester can trigger sudden hemorrhage of the AVM. Also, as labor ensues, significant remodeling of the myometrium may further push the vascular mass to bleed. This case suggests that uterine AVM in pregnancy can be managed conservatively with serial ultrasound monitoring, close follow up and thorough counselling of the patient. Due to the limited number of cases reported in literature, the appropriate timing and route of delivery still needs to be determined.

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