Fertility preserving surgical approach to uterine arteriovenous malformation*

By Mirah D. Borja, MD; Sheryl Ann B. Dela Cruz, MD, DPOGS and
German D.C. Tan-Cardoso II, MD, FPOGS, FSGOP, FPSGE
Department of Obstetrics and Gynecology, East Avenue Medical Center

ABSTRACT

Arteriovenous malformations (AVM) are vascular disorders with a mixture of arterial, venous and small capillary-like channels with fistulous connections. Uterine arteriovenous malformations are rare cause of abnormal uterine bleeding with only a few reported cases. They may arise from pregnancy, miscarriage, previous cesarean section or other uterine surgery and gestational trophoblastic disease. Diagnosis can be made through angiography or doppler ultrasonography. Traditionally, uterine AVMs are treated with hysterectomy but with the advances in technology, minimally invasive conservative approaches such as radiologic arterial embolization or laparoscopic uterine artery ligation have become available.

We present a case of a 29-year-old, G2P1 (1011) who had a three- month history of heavy, intermittent vaginal bleeding from uterine arteriovenous malformation after a miscarriage. Laparoscopic bilateral uterine artery occlusion, offered a minimally invasive treatment with high symptomatic effectiveness.

Keywords: abnormal uterine bleeding, uterine artery ligation, uterine arteriovenous malformation

INTRODUCTION

Uterine Arteriovenous malformation is a rare cause of abnormal uterine bleeding. It is an abnormal connection between uterine arteries and vein lacking an intervening capillary network observable at histopathologic examination.[8]

The classical presentation of patients with uterine arteriovenous malformations is heavy and prolonged bleeding, usually with an intermittent and torrential pattern with no obvious cause.[6]

The current reference standard for diagnosing arteriovenous malformation is pelvic angiography. It aids confirmation and can be used concurrently to perform vessel embolization.[1] This, however, is invasive and relatively expensive, hence, it is reserved for mapping and therapeutic embolization procedures. Alternative diagnostic modalities are Doppler ultrasound, which is more readily available and inexpensive, computed tomography scan and magnetic resonance imaging.

Arteriovenous malformations are traditionally treated by medical management, uterine artery embolization or hysterectomy.[2] Uterine artery ligation via laparotomy or laparoscopy have also been used. Currently, the gold standard for treatment of women desiring future fertility is uterine artery embolization.

The objective of this paper is to enumerate the diagnostic and management options and explain the role of hysteroscopy and laparoscopy as new surgical approaches in the treatment of uterine arteriovenous malformation.

CASE REPORT

This is a case of DD, a 29 year-old, gravida 2 para 1 (1011), single, Roman Catholic, housewife, who came to our institution with a chief complaint of vaginal bleeding.

The patient's past medical history, family history and social history are non-contributory to her current condition. Her menarche was at age 10 years and subsequent menses were regularly occurring in monthly cycles, lasting for seven days amounting to two pads per day. Her last menstrual period was 10 weeks (May 28, 2018) prior to her first consult for vaginal bleeding. Her first pregnancy was in 2008, which was home delivered at term attended by a midwife.

Four months prior to admission, at 4 weeks amenorrhoeic, her pregnancy test was positive. No prenatal consult yet during this time.

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She was apparently well until 3 months PTA when she started to have vaginal bleeding amounting to one fully soaked pad per day. Upon consult at a private clinic, ultrasound revealed blighted ovum at 8 weeks gestation for which dilatation and curettage was recommended. She requested to be referred to a local hospital due to financial constraint.

On her consult in a district hospital, cervical ripening with medication was attempted for 14 days in preparation for curettage. Cervical ripening failed. Her vaginal bleeding, however, ceased. Two months prior to admission, her bleeding recurred with increased amount. Upon consult, her cervix was still unripe, so she was referred to our institution.

Upon consult in our institution one month PTA, she was stable but with persistent vaginal bleeding amounting to one to two moderately soaked pads per day. Her transvaginal sonogram showed normal sized retroverted uterus with dilated myometrial vessels demonstrating abundant and turbulent flow and thickened endometrium. Serum B HCG was 20 mIu/ml. Retained products of conception was considered (Figure 1). However, we suspected Uterine AV Malformation because of the localized area of increased vascularity within the myometrial vessels seen on color doppler. Hence, the initial plan of dilatation and curettage was deferred. A repeat ultrasound evaluation by a senior sonologist was planned.

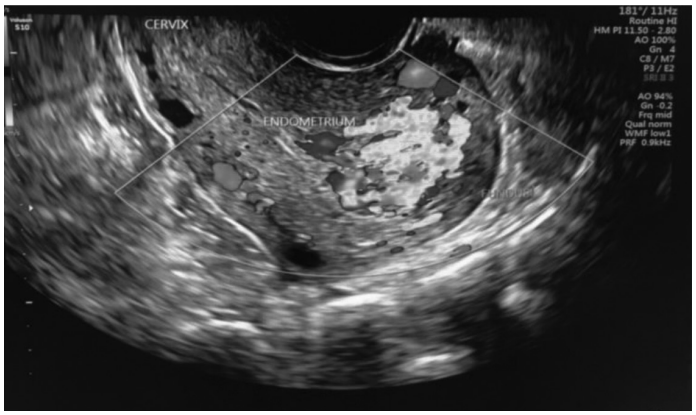


Figure 1. Transvaginal sonogram revealed normal sized retroverted uterus measuring 5.71 x 5.27 x 5.06 cm with cervix measuring 3.50 x 2.0 x 2.57 cm. The myometrial vessels were noted to be markedly dilated, which demonstrate abundant and turbulent flow. The endometrium is thickened at 2.52 centimeters with mixed echoes and abundant flow, to consider retained products of conception, blood and/or blood clots. Normal ovaries

Repeat ultrasound examination with doppler studies (2 weeks PTA delete) showed normal sized retroverted uterus, thickened endometrium, heterogenous with cystic spaces, midline echoes not defined. The endomyometrial junction at the posterior mid to upper segment is interrupted by dilated myometrial vessels with abundant and turbulent color on flow mapping, consider arteriovenous malformation. (Figure 2). Confirming the presence of uterine arteriovenous malformation, the patient was then advised diagnostic hysteroscopy and laparoscopic bilateral uterine artery coagulation.

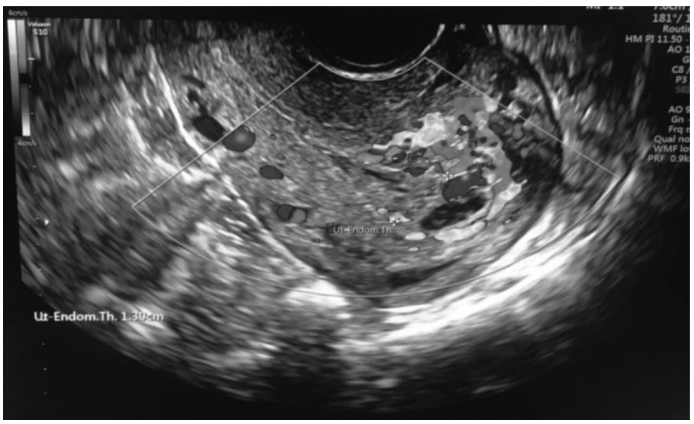


Figure 2. Repeat transvaginal sonogram revealed normal sized uterus, retroverted, measuring 5.23 x 5.31 x 5.53 cm with cervix measuring 3.99 x 2.29 x 2.77 cm. The endometrium is thickened at 1.3 cm, with cystic spaces, midline echoes not defined. The endomyometrial junction at the posterior mid to upper segment is interrupted by dilated myometrial vessels with abundant and turbulent color flow on mapping

She was finally admitted for the planned procedure in stable condition. CBC, coagulation studies and blood chemistry were normal. Her abdomen was flabby with normoactive bowel sounds, tympanitic on all quadrants, soft and nontender. On speculum examination, the cervix was pink, smooth, with bleeding per os. Upon internal examination, cervix was closed with no cervical motion tenderness; the uterus was not enlarged with no adnexal mass nor tenderness.

On diagnostic hysteroscopy, the uterine cavity was well visualized despite mild bleeding. The endometrium was thin with prominent tortuous vascularities on the wall. There was an intracavitary spongy mass with prominent engorged and tortuous vascularities at its base at the postero-fundal portion. (Figure 3)

After confirming the presence of AVM, laparoscopy then was performed. The uterine corpus was of normal size and retroverted. Adnexa and the rest of the pelvoabdominal organs were grossly normal. Laparoscopic bilateral uterine artery coagulation with bipolar forceps was performed. The patient was placed in modified dorsal lithotomy position using a CO2 as distention medium; transumbilical open Hasson technique for the primary port and two accessory port for hand instruments. After inspection and identification of the great vessels and ureter, the peritoneum was opened by creating a linear incision at the posterior broad ligament along the

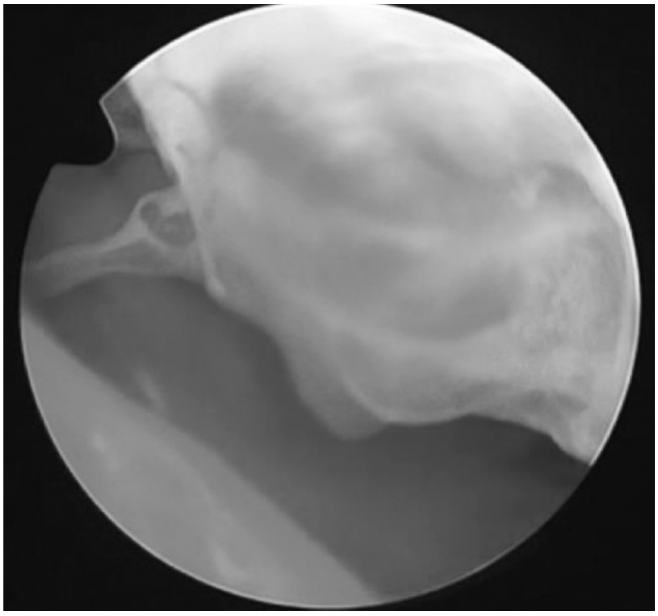


Figure 3. On diagnostic hysteroscopy, the uterine cavity was well visualized despite mild bleeding. The endometrium was thin with prominent tortuous vascularities on the wall. There was an intracavitary spongy mass with prominent engorged and tortuous vascularities at the base attached postero-fundal portion

infundibulopelvic ligament, displacing it medially. The paravesical and obturator spaces were developed by blunt dissection. The ureters were traced until it crossed under the uterine arteries. The uterine arteries were isolated then triply coagulated using bipolar device. (Figure 4A). The same was done on the contralateral side (Figure 4B).

An immediate repeat hysteroscopic visualization of the uterine cavity was executed showing significant decrease in bleeding as well as regression of the initially seen tortuous vessels on the uterine wall. The engorged prominent vascularity at the base of the intra-cavitary mass was noted to have constricted. (Figure 5)

The patient's postoperative course was unremarkable, and she was discharged on her 2nd postop day in stable condition and improved. Her vaginal bleeding completely stopped.

On the first post-operative week, a repeat transvaginal sonogram showed regression of the abnormal myometrial vascularities, normal-size retroverted uterus, thickened heterogenous endometrium with irregular cystic spaces, interrupted endomyometrial junction at the posterofundal area containing mixed echoes, probably representing blood or blood clots, and myometrial vessels at the posterofundal area appeared dilated with moderate color on flow mapping.

2 months post-op, on her day 8 of menses, a repeat transvaginal sonogram revealed normal size retroverted uterus with thickened endometrium at 1.01cm. There was no area of increased vascularity in the endometrium nor myometrium (Figure 6). The patient reported resumption of normal monthly menstrual cycle.

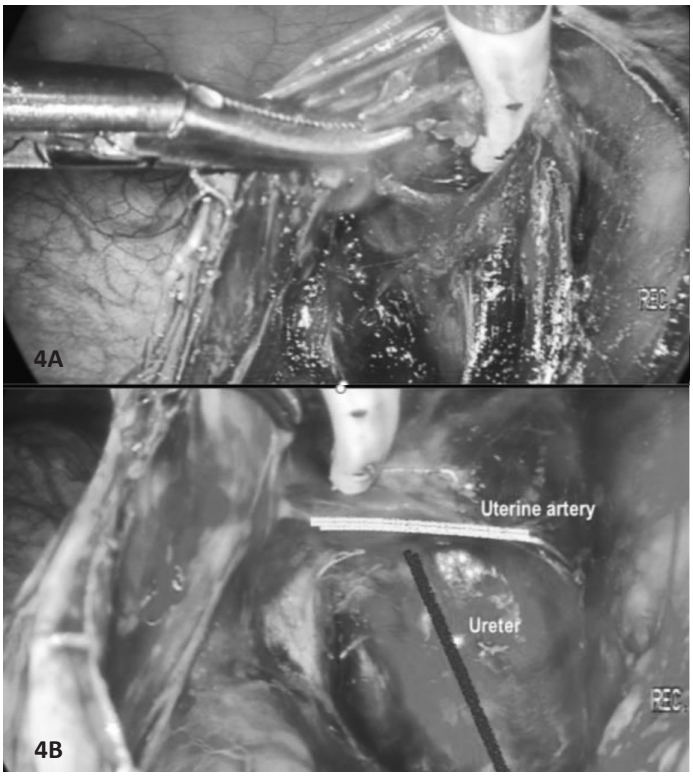
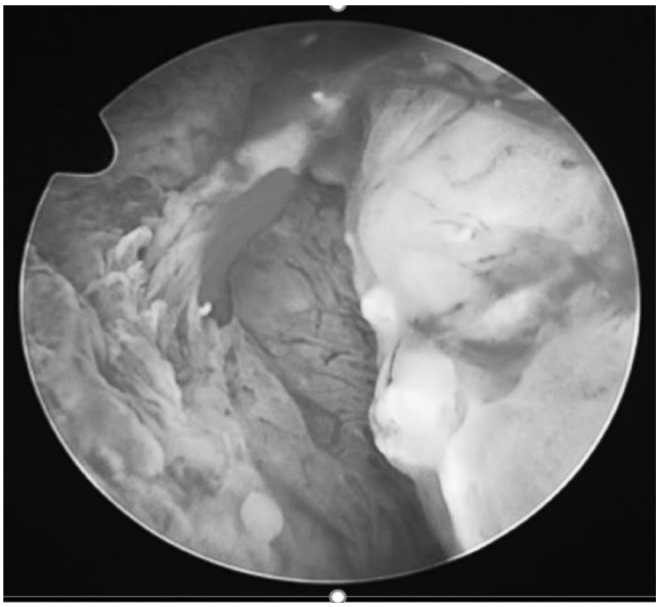


Figure 4 (A&B). After inspection and identification of the great vessels and ureter, the peritoneum was opened by creating a linear incision at the posterior broad ligament along the infundibulopelvic ligament, displacing it medially. The paravesical and obturator spaces were developed by blunt dissection. The ureters were traced until it crossed under the uterine arteries. The uterine arteries were isolated then triplycoagulated using bipolar device



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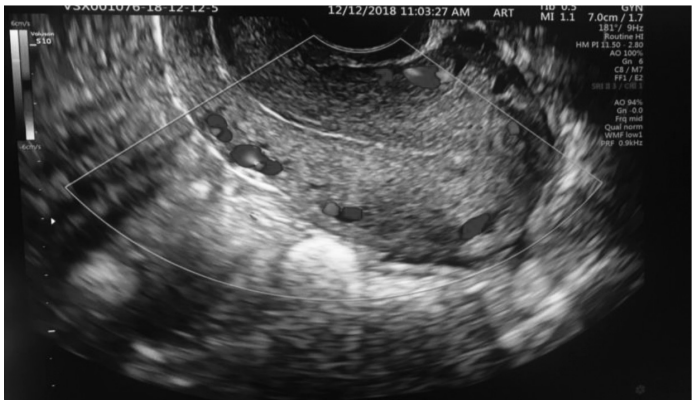

Figure 6. 2 months post-operatively, on her day 8 of menses, a repeat transvaginal sonogram revealed normal sized retroverted uterus with thickened endometrium at 1.01cm. There was no area of increased vascularity in the endometrium nor myometrium.

CASE DISCUSSION

Emmanuel, Luschka, and Virchow first described arteriovenous malformations in the mid-1800s.

The reported cases of uterine AVM are few. A study done by O. Brien et al. identified uterine arteriovenous malformations in 21 women out of 464 with abnormal uterine bleeding using color doppler sonography and spectral doppler analysis, resulting to an incidence of 4.5%.[4] Contrary to this, Yazawa, et.al, performed a prospective study of 959 women and reported a lower incidence of 0.6% of uterine vascular malformation.

Locally, the first reported case was published by Dr. John Eymard Sy in 2003. It was a case of a 27 year-old, gravida 4 para 3 (2112) who presented with a four–month history of menorrhagia with symptomatic anemia. Uterine AVM was diagnosed by ultrasound with Doppler studies and further confirmed by histopathologic result. The patient was managed with total abdominal hysterectomy.

Uterine arteriovenous malformation may be congenital or acquired. Congenital AVM are thought to result from failure of embryological vascular differentiation leading to multiple vascular connections.[2] The mesenchyme forms large flexiform structures that differentiate into mature vessels with the primitive components disappearing or they may develop partially with primitive vascular communications persisting.[2] Congenital AVMs, therefore, are focal areas of inadequate vascular development. The lesions tend to have multiple feeding arteries, draining vein and intervening nidus; hence, it grows by recruitment of collateral vascular channels.[2]

In contrast, acquired AVMs are usually large vessels that arise from prior dilatation and curettage, cesarean section, previous uterine surgery, pelvic trauma, gestational trophoblastic disease and uterine and cervical cancers.

I would suspect that our index patient had spontaneous abortion 2 months PTA which she did not notice. It is also possible that the product of conception ended in spontaneous resorption. After that event, there developed connection of arterial and venous vessels within the myometrium during involution of the chorio-placental villi following cessation of pregnancy. As explained in the literature, in cases of spontaneous abortion, it is postulated that these malformations may arise when venous sinuses become incorporated in scars within the myometrium after necrosis of the chorionic villi.

Uterine arteriovenous malformations are more commonly diagnosed in the postpartum period or a few months after a spontaneous miscarriage or termination of pregnancy.[6]

The classical presentation of patients with uterine arteriovenous malformations is abnormal vaginal bleeding, ranging from frequent spotting to catastrophic hemorrhage, usually with an intermittent and torrential pattern which can be severe enough to cause anemia and shock.[1] Therefore, awareness of the condition is essential to be able to facilitate effective management and prevent complications or even death from catastrophic bleeding.

In the past, when imaging modalities were not available, diagnosis of arteriovenous malformation was made only after hysterectomy and histopathologic examination since the diagnosis of uterine arteriovenous malformation cannot be made by clinical presentation and assessment alone.

In the present time, many modalities are available that can be used to diagnose AVM. Digital subtraction angiography (DSA) remains the current gold standard for diagnosis of uterine AVM. This invasive procedure provides an unmatched level of detail in identifying and localizing arteriovenous communications.[4] The defining feature of AVMs is brisk early filling of numerous enlarged veins emerging from a nidus, or nest of abnormal vessels after the arterial contast injection.[4]

Pelvic sonography with color doppler is often the initial screening study performed in the setting of suspected uterine AVM It provides a non-invasive method of diagnosis of arteriovenous malformation by displaying the vascular flow in hypoechoic areas within the myometrium.[4] Color doppler evaluation documents the presence of hypervascular tangles of tortuous vessels with a color mosaic of irregular turbulent flow.4 The arteriovenous communications show multidirectional mixing of arterial and venous waveforms, low resistance high velocity flow within the

lesion is highly diagnostic of uterine AVM.[6]

Additionally, uterine arteriovenous malformation may also be detected using hysteroscopy, computerized tomography, and magnetic resonance imaging.[4]

What made us to suspect the presence of uterine AVM in our patient was the finding on TVS with color doppler studies. The patient's transvaginal ultrasound with color doppler studies showed normal sized retroverted uterus, thickened endometrium with endomyometrial junction at the posterior mid to upper segment interrupted by dilated myometrial vessels with abundant and turbulent color on flow mapping which is highly suggestive of arteriovenous malformation (Figure 2).

Differential diagnoses of uterine AVM are retained products of concepcion and GTD. Although it is relatively easy to detect abnormal blood flow in the myometrium by using color Doppler ultrasonography, retained products of conception can sometimes be difficult to differentiate using ultrasonographic imaging alone.

To differentiate uterine AVM from these two possibilities, serum beta HCG was measured. Her serum beta HCG concentrations taken 2 weeks apart were only 20 and 22 miu/ml which rules out gestational trophoblastic disease.

To emphasize the importance of correct diagnosis, a misdiagnosis of the patient may have led to performing a dilatation and curettage which would eventually lead to a torrential and fatal vaginal bleeding. A high index of suspicion coupled with good ultrasound interpretation and measurement of beta HCG would lead to a correct diagnosis.

Individualizing the treatment for patients with uterine arteriovenous malformation involves consideration of the patient's hemodynamic stability, amount of bleeding and desire for future fertility.[1]

In the past, hysterectomy was the mainstay of treatment for uterine arteriovenous malformation but with advances in technology, conservative management has become available.

For asymptomatic and stable patients, medical management of uterine arteriovenous malformation may be considered. There have been reports of effective treatment of uterine AVM using GnRHa, methylergometrine and danazol, all of which reduce blood flow to the uterus.[4]

In the recent years, transarterial embolization of uterine artery was considered the primary therapeutic option for management of uterine AV malformation due to its potential to preserve fertility.[2] Various studies have shown that uterine artery embolization (UAE) has high clinical success rates from 93% to 95%.[4]

Uterine artery embolization has been reported to have complications in approximately 10% of cases which include pelvic pain, low grade temperature, and infection. It also carries a risk of recurrence and has also been suggested that embolization is responsible for decreased uterine contractions in the treated areas.[10] A Cochrane review from 2014 demonstrated that uterine artery embolization can result in endometrium ischemia and provokes a "postembolization syndrome", such as massive necrosis and infarction of the uterus, pelvic pain, and uterine artery rupture.

Poor pregnancy outcomes were also related to UAE such as abortions and preterm deliveries. A study by Vilos, et al. questioned the efficacy of this procedure, hence, concluding that uterine artery embolization may not be as effective at managing AVM as previously thought and should be questioned as an initial therapy in symptomatic women of reproductive age and desiring fertility preservation. [10]

Presently, there is limited literature available regarding the utilization of laparoscopic bilateral uterine artery ligation in managing patients diagnosed with uterine arteriovenous malformation. Owing to the high magnification view with laparoscopy, it can provide safe dissection and identification of the uterine vessels for definitive ligation. Compared to laparotomy, fertility preservation is optimized with minimal tissue handling in laporoscopy lessening the risks of adhesion formation.

Since our patient is young and desirous of having more children, conservative management is appropriate. Our patient has been bleeding continuously making medical management not an option. Transarterial embolization is not available in our hospital. The best approach we can offer our patient was laparoscopic uterine artery ligation.

Post-operative surveillance six weeks after the procedure showed regressions of the initially described vascularites with thin endometrium and resumption of regular menses.

This patient was fortunate to have averted the consequences of the initially proposed dilatation and curettage procedure in a primary hospital that could have resulted to catastrophic intraoperative hemorrhage, hysterectomy or worst death from intractable bleeding.

There were 2 other cases that were managed laparoscopically in our institution. One underwent total laparoscopic hysterectomy. The other case published by Matundan et. al., was that of a 30 y.o. with acquired AVM after a cesarean section that was successfully managed by laparoscopic bilateral uterine artery ligation. This patient already became pregnant and delivered via repeat cesarean section.

SUMMARY

Many lessons have been learned from this case. First, Uterine AVM must be suspected in a patient suffering from unexplained vaginal bleeding. Second, even with unavailability of modern technology diagnostic modalities like angiography, CT scan and MRI, diagnosis of uterine AVM can be made with good ultrasonographic interpretation. Third, missing the diagnosis of uterine AVM could have led to catastrophic curettage. Finally, laparoscopic uterine artery ligation is a feasible, safe, and economical procedure, which offers a fertility preserving alternative treatment modality for uterine AVM. ■

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