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Pelvic arteriovenous malformation involving the uterine and internal iliac vessels with concomitant true aneurysm of uterine artery diagnosed through color Doppler ultrasound

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Abstract:

BACKGROUND: Pelvic arteriovenous malformations (AVMs) are rare but carries life-threatening consequences.

CASE REPORT: A 47-year-old multipara who had previously undergone four cesarean sections came for re-evaluation of a malignant ovarian new growth. She was asymptomatic. Repeat ultrasound revealed normal ovaries, and a cystic structure at the left adnexa with abundant mixing of colors, turbulent flow and pulsative waveforms on spectral Doppler. It arises from serpentine tubular structures from the uterine isthmus. Uterine artery aneurysm was considered. Magnetic resonance angiography confirmed the findings of aneurysm and pelvic arteriovenous malformation. The patient underwent a hysterectomy with ligation and excision of aneurysm. Histopathologic findings showed pelvic AVM and a true aneurysm of the uterine artery.

CONCLUSION: Ultrasound with color Doppler is a low-cost and readily available tool for gynecologists for the diagnosis and management of pelvic AVM.

Keywords:

Doppler ultrasound, pelvic arteriovenous malformation, uterine artery aneurysm

Introduction

The vascular supply of the uterus mostly originates from the uterine artery that branches from the internal iliac artery.[1] Pelvic vascular malformation in this location is rare.[2] Clinical symptoms may be absent and can be an incidental finding during a routine ultrasound. It carries life-threatening consequences; hence, clinicians and sonographers should have a high index of suspicion when dealing with these vascular lesions. We present a case of a pelvic arteriovenous malformation (AVM) and its color Doppler ultrasound features.

Case Report

A 47-year-old Gravida 4 Para 4, who had a cesarean section for all her term deliveries, was referred due to an ultrasound finding of ovarian new growth with malignant features. One week before consult, during her annual physical examination, a transvaginal ultrasound was done, which revealed a cystic mass at the right adnexa [Figure 1a] with abundant color on flow mapping [Figure 1b]. The patient was referred for gynecologic Doppler studies. She denies weight changes, abdominal discomfort, changes in bowel or bladder habits, or changes in her menstrual cycle

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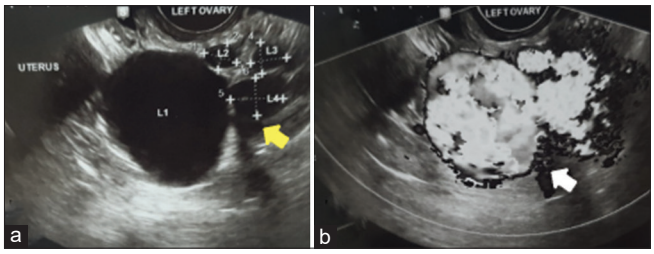

Figure 1: Noninstitutional ultrasound: Initial transvaginal ultrasound. (a) Grayscale image showing multiple cystic structures at the left adnexa, yellow arrow. (b) Color Doppler showed abundant color

or flow. She denies any known comorbidity. She is a nonsmoker and denies illicit drug use. Ultrasound re-evaluation showed normal ovaries, and a thin-walled, anechoic, cystic structure lateral and inferior to the left side of the uterus measuring 4.06 cm × 3.58 cm × 4.18 cm with a volume of 31.80 mL [Figure 2a]. It appears to be arising from a serpentine tubular structure coming from the uterine isthmus, suggestive of a dilated blood vessel [Figure 2b]. On color Doppler, the cystic structure had abundant mixing of colors [Figure 2c], turbulent flow [Figure 2d], and pulsative waveforms on spectral Doppler [Figure 2e], with peak systolic velocity (PSV) of 145.92 cm/s and resistance index of 0.45. Three-dimensional (3D) ultrasound confirmed the location of the cystic mass [Figure 2f]. Considering the location, grayscale, and color Doppler findings, uterine artery aneurysm was considered. Magnetic resonance angiography with contrast confirmed the ultrasound findings. It showed a dilated, tortuous anterior branches of the left internal iliac artery and vein in the left side of the uterine cervix and lower uterine segment [Figure 3a], particularly the uterine vessels, with apparent fistulous connection between artery and vein associated with a saccular aneurysmal component measuring approximately 3.4 cm × 4.3 cm × 3.5 cm [Figure 3b]. The patient was advised regarding possible complications, such as life-threatening rupture of the aneurysm. After informed consent, she opted for surgery. On mobilization of the uterus, an unruptured cystic structure was seen at the lateral aspect of the uterus measuring 4.0 x 3.8 x 2.8 cm, with a thrill upon palpation, confirming the diagnosis of an aneurysm. Distal to this were tortuous vessels [Figure 4]. The thoracovascular surgeon performed the ligation of distal vessels and excision of the aneursym. Owing to the location and vascular connections of the aneurysm to the uterus, which can increase the risk of bleeding if dissected, the uterus and cervix were removed with subsequent bilateral adnexectomy. The patient tolerated the procedure well, with a total blood loss of 400 mL. On gross examination of the specimen, the aneurysm has a unilocular cavity covered with red–brown granular material [Figure 5a and b]. Histopathologic examination of the specimen confirmed a vascular malformation compatible

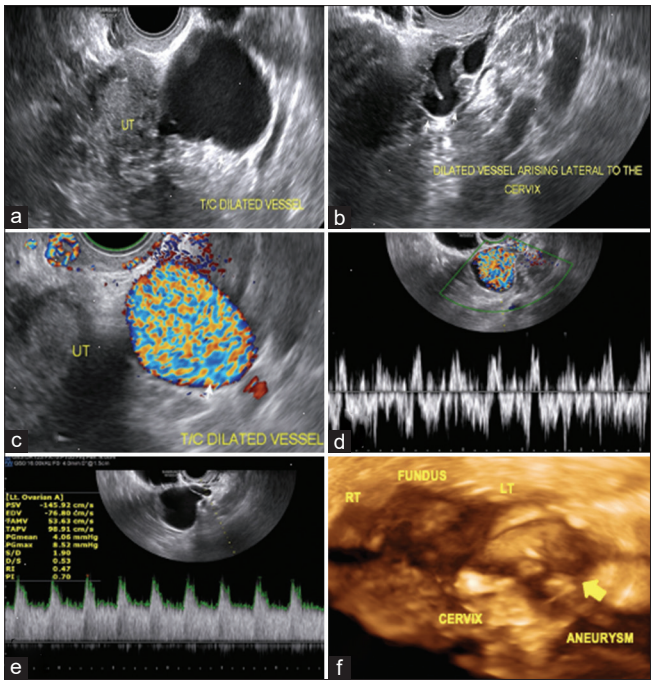

Figure 2: Gynecologic Doppler studies: (a) Grayscale image showing cystic structure lateral and inferior to the left side of the uterus measuring 4.06 cm × 3.58 cm × 4.18 cm (volume 31.80 mL). (b) Serpentigious structure adjacent the cystic structure is a blood vessel arising from the uterine isthmus suggestive of the uterine artery. (c) Color flow mapping of the cystic structure showing abundant and mixed colors. (d) Color Doppler of the cystic structure showed bidirectional turbulent flow. (e) Color Doppler of the adjacent serpentigious structure showed pulsative waveforms. (f) 3D image of the uterus with adjacent cystic structure (yellow arrow). All findings are suggestive of uterine artery aneurysm. PSV: Peak systolic velocity, UT: Uterus, RT: Right adnexa, LT: Left adnexa

with AVM [Figure 6a] involving the internal iliac and uterine vessels. The wall of the aneurysm contains three layers of an arterial wall [Figure 6b], hence the diagnosis of a true aneurysm. The patient had an unremarkable postoperative course and was subsequently sent home.

Discussion

Pelvic AVMs are rare, all the more if with a concomitant true aneurysm.[2,3] There are only few case reports and most are associated with pseudoaneurysms. AVM is undifferentiated high-flow vascular malformations that resulted from multiple abnormal communications between the arterial and venous system without an intervening capillary network.[2,4] Arteriovenous communications in pelvic AVM can vary widely in size, number, and location.[2] It may arise from any of the arteries in the pelvis and the main supplying arteries are usually the gonadal (testicular or ovarian) arteries, branches of the internal iliac arteries, hemorrhoidal branches of the inferior mesenteric artery, and midline sacral artery.[5] Pelvic AVM may extend to the uterus but is distinguished from uterine AVMs because the latter are arteriovenous communications that are located entirely within the uterus.[2]

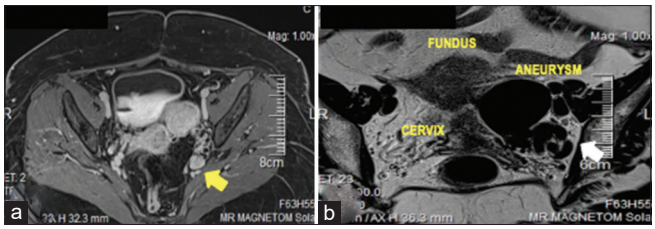

Figure 3: Magnetic resonance image/magnetic resonance angiogram images: (a) Dilated tortuous anterior branches of the left internal iliac artery and vein in the left side of the uterine cervix and lower uterine segment. (b) Saccular aneurysm component

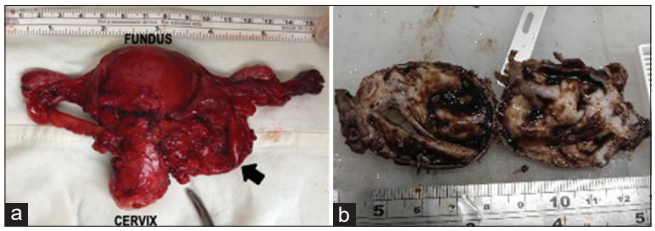

Figure 5: Gross examination: (a) Uterus with cystic structure at the left parametria. (b) Cut section of the cystic structure showing a unilocular cavity with red–brown granular material

AVM of the pelvis can be congenital or acquired. Congenital AVM results of abnormal endothelial cell proliferation and delayed vascular remodeling during angiogenesis,[6] whereas acquired AVM is secondary to infection, traumatic deliveries, cesarean sections, and other interventions.[7] Congenital AVM may remain asymptomatic up into adulthood.[8] However, it may progress to symptomatic AVM after stimulation by trauma, hormonal change during puberty or with pregnancy,[8] or a consequence of thrombosis or infection.[9] It is difficult to determine whether the pelvic AVM in this case is acquired or congenital, but what is certain is that her prior cesarean section placed her at risk for its development.

The AVM in this case is an incidental finding, and the patient denies any related symptoms. Most of the reported clinical presentations in symptomatic pelvic AVMs are abnormal vaginal bleeding ranging from frequent spotting to catastrophic hemorrhage, pelvic pain or pressure, signs of increased venous pressure such as vulvar varices, and in cases of high-flow left-to-right shunts, high-output cardiac failure.[2,7,10,11] Pelvic AVM can lead to life-threatening hemorrhage and hence timely diagnosis is important.

Diagnostic tools such as computed tomography (CT) angiogram and magnetic resonance angiogram (MRA) have been extensively used in AVM because it offer better tissue contrast and delineate the surrounding pelvic organ involvement.[10] MRA findings include high-flow serpentine and enlarged feeding arteries and draining veins, which appear as large flow voids on

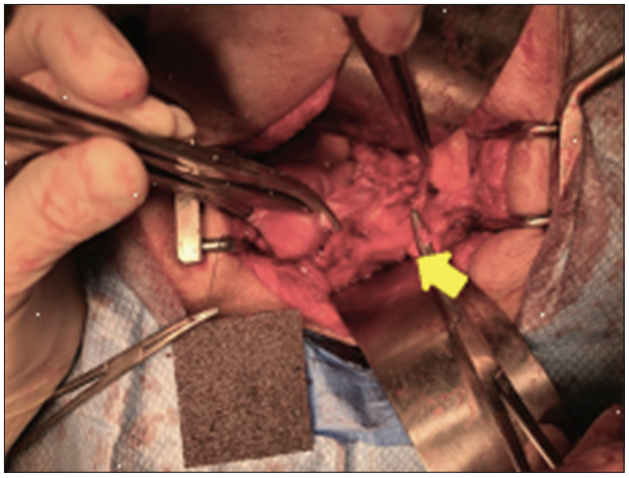

Figure 4: Intraoperative findings: Located at the left parametria is a dilated structure measuring 4.0 cm × 3.8 cm × 0.8 cm with bruit upon palpation (yellow arrow)

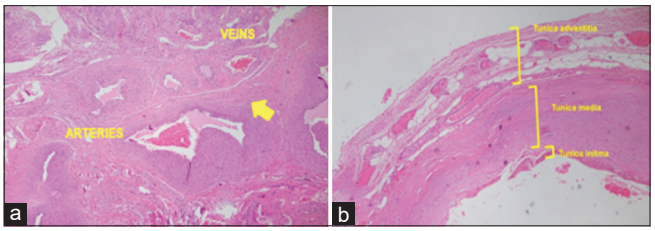

Figure 6: Histopathologic examination (scanner view), 4x magnification. (a) Pelvic arteriovenous malformation showing mixture of thick-walled vessels (large arteries, veins, and small vessels, yellow arrow) separated by dense fibrous connective tissue and fibroadipose tissues. (b) Wall of the aneurysm exhibiting arterial layers: tunica externa, tunica media, and tunica intima

spin-echo images or high-signal-intensity structures on gradient-echo images, with the absence of a well-defined mass.[12] MRA is preferred over CT angiogram to avoid ionizing radiation, especially in child-bearing age.[10] Digital subtraction angiography is the gold standard in the diagnosis of AVM[2] but is an invasive procedure and is reserved and performed together with endovascular treatment.

Pelvic ultrasonography can be offered as an inexpensive screening and diagnostic method for AVM. Grayscale sonographic findings can be nonspecific[2] and operator dependent. Owing to the rarity of the condition, a sonographer can possibly miss the diagnosis using only grayscale sonography. Ultrasonographic evaluation of pelvic masses should always employ color Doppler with flow velocity measurements to increase sensitivity, specificity, positive predictive value, and negative predictive value in establishing the diagnosis.[13] Once a vascular malformation is determined, Doppler interrogation further differentiates between venous and arterial flow, the latter having pulsative waveform. Doppler can also categorize vascular malformation into slow-flow and fast-flow lesions, thus aiding in

differential diagnosis.[14,15] High-flow malformations include both AVM and arteriovenous fistulas, whereas slow-flow malformation is venous and lymphatic malformations.[14] Venous malformations of the uterus and ovaries are typically associated with insufficiency of the ovarian vein and may coexist with pelvic congestion syndrome.[15] In this patient, color Doppler showed abundant color, turbulent flow and pulsative waveforms on spectral Doppler therefore ruling out venous or lymphatic malformation.

Pelvic AVM in color Doppler shows the presence of enlarged uterine and/or pelvic vessels exhibiting intense signal, aliasing, and apparent flow reversal indicative of turbulent high-velocity flow.[2,12] Spectral Doppler can also demonstrate elevated flow velocities within low-resistance vessels, indicating the presence of an arteriovenous communication.[2,3] The latter exhibits a resistance index between 0.25 and 0.55.[16] The present case showed a resistance index of 0.45, increasing the probability of an AVM.

The most striking characteristic of AVM in this patient is the presence of an aneurysm. Aneurysm manifests in grayscale ultrasound as a pulsating anechoic structure with abundant color from on Doppler. It can be further described as fusiform or saccular based on its shape.[3] The aneurysm, in this case, is saccular and represents localized extrusions from the main vessel. 3D ultrasound was also used in this case to further evaluate the location of the aneurysm. This provides a clear depiction of the vascular lesion in relation to its surrounding structures.[17] Owing to its location, uterine artery aneurysm was considered. Aneurysms of the uterine artery are rare, and within this location, mostly are pseudoaneurysms.[18] A true aneurysm can be distinguished from pseudoaneurysm through color Doppler. True aneurysm manifests with an excessive color filling of the parent artery, whereaspseudoaneurysm exhibits a to-and-fro pattern when blood flows into the sac during systole and away during diastole.[3] The latter was not seen in this patient. It was later confirmed in the histopathology that it was a true aneurysm, having all layers of the arterial wall.

Ultrasonography does not only aid in diagnosis, but it can also guide management. PSV can predict the natural history of AVM.[19] PSV of 40 cm/s distinguishes an AVM with a high probability of regression without intervention and can be expectantly managed with serial sonographic follow-up and stable hemoglobin.[19] PSV between 40 and 60 cm/s can be managed with close follow-up in patients without excessive bleeding, as it may regress or persist.[19] PSV of more than 60 cm/s identifies a high-velocity AVM with a low likelihood of spontaneous regression and a high risk of significant bleeding; therefore, it should be managed expeditiously.[10] The PSV in this case was 145.92 cm/s. Additionally, the concomitant uterine artery aneurysm measures 4 cm in widest diameter. There is a high incidence of spontaneous rupture occurring in uterine artery aneurysm with a diameter of more than 1 cm.[20] With all of this imaging data, surgical intervention was chosen in this case rather than conservative care.

In managing these pelvic and uterine AVM, the literature suggests that transcatheter embolization has high rates of success.[2] A systematic review of 40 studies that included 54 patients with acquired AVM who underwent transcatheter embolization found a primary success rate of 61% and a secondary success rate of 91%.[10] However, AVM may persist or recur if one or more arteriovenous communications remain patent.[2]

Hysterectomy is the definitive surgical therapy for pelvic AVM and may be performed either first-line or after the failure of embolotherapy.[2] The aneurysm in this case was seen at the lateral aspect of the uterus. Attempting dissection of pelvic AVM from adjacent pelvic structures is technically difficult and may result in increased blood loss. Since the patient has also no desire for pregnancy, hysterectomy was performed after ligation of the distal blood supply with excision of the aneurysm. Complications after hysterectomy include infection (13%), venous thromboembolism (1%–12%), genitourinary system injuries (0.75% to 1.5%), and rarely, gastrointestinal tract injuries, bleeding complications, neuropathy, and vaginal cuff dehiscence.[2]

Similar to any other form of disease, management and treatment strategy depend on correct diagnosis. Ultrasound with color Doppler is a low-cost tool for the diagnosis of pelvic AVM, which can direct preoperative planning and guide the treatment options which, subsequently optimizing management and improving the outcome of this life-threatening condition.

Summary

This article presented the case of a 47-year-old multipara, asymptomatic, with incidental finding of pelvic AVM with true saccular aneurysm of the uterine artery diagnosed through color Doppler ultrasound. Color Doppler ultrasound is a readily available tool for gynecologists in the diagnosis of pelvic vascular malformation. In addition, it is also valuable in the decision-making process, specifically in the timing of intervention and in what appropriate treatment should be instituted. As pelvic AVMs are rare but are life-threatening conditions, a high index of suspicion and application of color Doppler should always be imperative during ultrasound evaluation of pelvic masses.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Authorship contributions

Ma. Cresilda Paz Sabularce, MD, FPOGS, FPSMFM, FPSUOG - Involved in conceptualization, over-all writing, review and editing.

Gumersinda Cruz-Javier, MD, FPOGS, FPSMFM, FPSUOG - Involved in supervision and mentorship.

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Conflicts of interest

There are no conflicts of interest.

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