OBSTETRICS

Ultrasound diagnosis and management of acquired uterine enhanced myometrial vascularity/arteriovenous malformations

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BACKGROUND: Arteriovenous malformation is a short circuit between an organ's arterial and venous circulation. Arteriovenous malformations are classified as congenital and acquired. In the uterus, they may appear after curettage, cesarean delivery, and myomectomy among others. Their clinical feature is usually vaginal bleeding, which may be severe, if curettage is performed in unrecognized cases. Sonographically on 2-dimensional grayscale ultrasound scanning, the pathologic evidence appears as irregular, anechoic, tortuous, tubular structures that show evidence of increased vascularity when color Doppler is applied. Most of the time they resolve spontaneously; however, if left untreated, they may require involved treatments such as uterine artery embolization or hysterectomy. In the past, uterine artery angiography was the gold standard for the diagnosis; however, ultrasound scanning has diagnosed successfully and helped in the clinical management. Recently, arteriovenous malformations have been referred to as enhanced myometrial vascularities.

OBJECTIVES: The purpose of this study was to evaluate the role of transvaginal ultrasound scanning in the diagnosis and treatment of acquired enhanced myometrial vascularity/arteriovenous malformations to outline the natural history of conservatively followed vs treated lesions.

METHODS: This was a retrospective study to assess the presentation, treatment, and clinical pictures of patients with uterine Enhanced myometrial vascularity/arteriovenous malformations that were diagnosed with transvaginal ultrasound scanning. We reviewed both (1) ultrasound data (images, measured dimensions, and Doppler blood flow that were defined by its peak systolic velocity and (2) clinical data (age, reproductive status, clinical presentation, inciting event or procedure, surgical history, clinical course, time intervals that included detection to resolution or detection to treatment, and treatment rendered). The diagnostic criteria were "subjective" with a rich vascular network in the myometrium with the use of color Doppler images and "objective" with a high peak systolic velocity of $\geq$20 cm/sec in the vascular web. Statistical analysis was performed and coded with statistical software where necessary.

RESULTS: Twenty-seven patients met the diagnostic criteria of uterine enhanced myometrial vascularity/arteriovenous malformation. Mean age was 31.8 years (range, 18-42 years). Clinical diagnoses of the patients included 10 incomplete abortions, 6 missed abortions, 5 spontaneous complete abortions, 5 cesarean scar pregnancies, and 1 molar pregnancy. Eighty-nine percent of patients had bleeding (n = 24/27), although 1 patient was febrile, and 2 patients were asymptomatic. Recent surgical procedures were performed in 55.5% patients (15/27) that included curettage (n = 10), cesarean deliveries (n = 5), or both (n = 1); 4 patients had a remote history of uterine surgery that included myomectomy. Treatment was varied and included expectant treatment alone in 48% of the patients with serial ultrasound scans and serum human chorionic gonadotropin until resolution (n = 13/27 patients), uterine artery embolization (29.6%; 8/27 patients), methotrexate administration (22.2%; 6/27 patients), hysterectomy (7.4%; 2/27 patients), and curettage (3.7%; 1/27 patients). Three patients required a blood transfusion. Of the 9 patients whose condition required embolization, the conditions of 7 patients resolved after the procedure although 1 patient's condition required operative hysteroscopy and 1 patient's condition required hysterectomy for intractable bleeding. Average peak systolic velocity after embolization in the 9 patients was 85.2 cm/sec (range, 35-170 cm/sec); the average peak systolic velocity of the 16 patients with spontaneous resolution was 58.5 cm/sec (range, 23-90 cm/sec).

CONCLUSIONS: Acquired enhanced myometrial vascularity/arteriovenous malformations occurred after unsuccessful pregnancies or treatment procedures that included uterine curettage, cesarean delivery, or cesarean scar pregnancy. Triage of patients for expectant treatment vs intervention with uterine artery embolization based on their clinical status, which was supplemented by objective measurements of blood velocity measurement in the arteriovenous malformation, appears to be a good predictor of outcome. Ultrasound evaluation of patients with early pregnancy failure and persistent bleeding should be considered for evaluation of a possible enhanced myometrial vascularity/arteriovenous malformation.

Key words: arteriovenous malformation, cesarean scar pregnancy, ultrasound, uterine artery embolization, uterus

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Enhanced myometrial vascularity/arteriovenous malformation (EMV/AVM) is a pathologic phenomenon described as a faulty "short circuit" of the blood stream between an organ's arterial and venous supply. The blood stream assumes an unusually high velocity, rendering the vessels into a vascular fistula. They were first described by Dubreil and Loubat[1] in 1926 as "aneurysme cirsoide de l'uterus." They are vascular lesions that may cause life-threatening gynecologic hemorrhage.[2-7] They can occur when the thin wall of the abnormal vessels are disrupted either naturally after menstruation or artificially after instrumentation[8-10] and have been reported in women across all ages.

AVMs are classified broadly as either congenital or acquired.[10] Acquired uterine AVMs are usually traumatic and result from previous uterine surgery including diagnostic or therapeutic curettage,[11,12] cesarean delivery, or myomectomy.[13] Endometrial carcinoma, cervical carcinoma, and gestational trophoblastic disease have also been implicated as causes of acquired uterine AVMs.[9] Lately, causative connection was recognized between cesarean scar pregnancy (CSP) and acquired AVM, as it was noted in 8 of 751 published cases of CSP[14] and in 5 additional case reports since 2010.[4-7,15] Because acquired uterine AVMs are considered very rare, their true incidence is unknown. Until 2005 there have been <100 cases reported in the literature, with just 73 cases reported before 1997.[16] Available data come from only small case series or single case reports.

The previously used term *AVM* is now subject to revision by those who contributed to the understanding of this disease during the last years.[17-20] The authors refer to the rich vessel network in cases of retained placental tissue, the term *SEVM* (as a descriptive term) can be applied to all acquired uterine AVMs, regardless of the presence or absence of products of conception. Therefore, it is justified to refer to all the aforementioned rich acquired vascular structures of the uterus defined by color Doppler interrogation as *EVM/AVM*. This new term will be used in this article in the appropriate places.

The clinical symptoms of uterine EMV/AVMs can appear gradually or suddenly, with patients most commonly presenting with heavy or irregular vaginal bleeding after a miscarriage, uterine surgery, or treated or untreated CSP. The natural history is variable; some cases slowly revert to normal circulation, and the condition disappears over a period of weeks to months, although some persist without regression, which puts the patient at higher risk of hemorrhage. EMV/AVMs represent 12% of all pelvic and intraperitoneal hemorrhages[21,22]; in 30% of cases, a blood transfusion is necessary.[3] With significant bleeding, treatment is of the essence, often in the form of uterine artery embolization (UAE). Importantly, curettage for patients with heavy vaginal bleeding because of an AVM may exacerbate the bleeding and may be life-threatening when the diagnosis of EMV/AVM has not been made before the intervention.[18]

In the past, the diagnosis of EMV/AVM was made via laparotomy. Later, angiography became the "gold standard" diagnostic method.[3] Although angiography is still used in the treatment of uterine EMV/AVM during a UAE, less morbid diagnostic methods have emerged. Saline solution infusion sonohysterography may be useful as an aid to detect an AVM.[23] Tal et al[24] described the use of color Doppler in diagnosing postabortal placental remnants. Most recently, transvaginal ultrasound scanning (TVUS) has emerged as an efficient, simple, and accessible diagnostic modality to detect and follow the vascular pattern of the EMV/AVM with the use of blood velocity blood flow indices.[18,25,26] Sugiyama et al[26] reported 1 case in which TVUS was used by measuring the resistance index of the blood flow. Capmas et al[27] used 3-dimensional ultrasound scans to diagnose EMV/AVM; however, the blood flow velocity was not reported. Lee et al[28] reported using TVUS in patients with EMV/AVM, measuring Doppler blood flow indices. Singh et al[29] used mainly magnetic resonance imaging; when they used TUVS, no quantification was reported. Tullius et al[30] reported using TVUS in 1 case.

The aim of this study was to review the value of TVUS in the diagnosis and treatment of suspected uterine EMV/AVMs to outline the disease's natural history. In addition to including the well-established uterine procedures that predispose to acquired EMV/AVM, this case series directs attention to an emerging link between CSPs and EMV/AVM. We also emphasize that, through early and proper identification of patients with EMV/AVM, we may be able to avoid potentially morbid treatments that include transfusion, curettage, UAE, or ultimately, hysterectomy.

Materials and Methods

This study is a retrospective review of medical records and ultrasound images from a single ultrasound unit to assess the presentation, management, treatment, and clinical outcomes of patients with uterine EMV/AVMs. All patients who presented to the New York University Langone Medical Center OB/GYN Ultrasound Unit from January 1, 2011 to August 31, 2014, and were diagnosed with pregnancy-related uterine EMV/AVM on 2-dimensional TVUS were eligible for inclusion in the study. As per our ultrasound scanning protocol, all patients who undergo gynecologic scans are first examined with the grayscale mode followed by color/power Doppler interrogation. There were no restrictions on age or racial/ethnic origins for inclusion. Exclusion criteria included patients with viable pregnancies at the time of an EMV/AVM diagnosis. The study was approved by the New York University School of Medicine Institutional Review Board.

Criteria for the sonographic diagnosis were (1) unusual, tubular, tortuous, anechoic structures seen by 2-dimensional grayscale ultrasound imaging on sagittal and/or transverse section of the uterus, which subjectively reveal an unusually rich vascularity with tortuous-appearing blood vessels that are concentrated in a small area of myometrium adjacent to the uterine cavity, with or without clearly visible products of conception (POC) that are detected by grayscale ultrasound imaging followed by color or power Doppler imaging (Figure 1, A and B) and (2) objectively a demonstration of high-velocity blood flow within the vascular "web" with a peak systolic velocity (PSV) of ≥20 cm/sec (Figure 1, C). Because measurement of blood velocity is angle dependent and the direction of blood flow within a vascular "web" is not necessarily parallel with the angle of insonation, it is expected that measurements within the web will differ from 1 another. To determine the highest blood flow velocity, we used a very narrow sampling window (2 mm) and measured at least 5-10 different sites