

# Congenital Uterine Arteriovenous Malformation: A Case Report

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**Abstract** Uterine arteriovenous malformation (UAVM) is an infrequent vascular condition although it can be fatal and threaten the patient's life. UAVM can be categorized as congenital or acquired. The majority of cases usually present with heavy intractable abnormal uterine bleeding in the childbearing age and/or recurrent miscarriages that's why the diagnosis of UAVM requires a high index of clinical suspicion. The exact incidence of UAVM remains to be elucidated but recent years have seen an increase in reported cases particularly after pregnancy, miscarriage and uterine surgical procedures such as Dilation and curettage (D&C). The Initial assessment of UAVM usually begins with transvaginal ultrasound (TUS) whereas angiography remains the modality of choice to confirm the diagnosis. Despite several medical agents have been employed in the treatment of the condition, In the past years, Embolization has become widely used as a first option in the management of UAVM because of its highly satisfying outcomes and more importantly avoidance of invasive procedures use with subsequent complications, especially hysterectomy which is reserved as a last resort. We herein report a young female with a confirmed congenital UAVM managed successfully via bilateral embolization of the uterine artery without the need for hysterectomy.

**Keywords:** Uterine arteriovenous malformations (UAVMs), vascular anomalies, vascular malformation, angiography, uterine artery embolization

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## 1. Introduction

Uterine arteriovenous malformations (UAVMs) are extremely uncommon conditions that may be responsible for vaginal hemorrhage. They have great significance in gynecological clinical practice due to their potential of causing life-threatening hemorrhage. In addition to the usual physiologic alterations occurring during gestation, there are several precipitating elements closely linked to this condition. Approximately all uterine patho-physiological processes are involved in its development [1].

Recent studies have shown that mutations of the RASA-1 gene on 5q13-22 cause a variety of congenital vascular anomalies including UAVMs [2,3,4]. UAVMs were firstly identified in 1926 by Dubreuil and Loubat [5].

UAVMs are abnormal connections of uterine blood vessels that frequently appear during the reproductive period [6].

Approximately 100 cases of uterine AVM have been documented in the literature, so the precise prevalence of uterine AVM remains unclear [7]. In spite of this, reports are on the increase due to the increasing accessibility of diagnostic methods particularly imaging ones [8]. US-

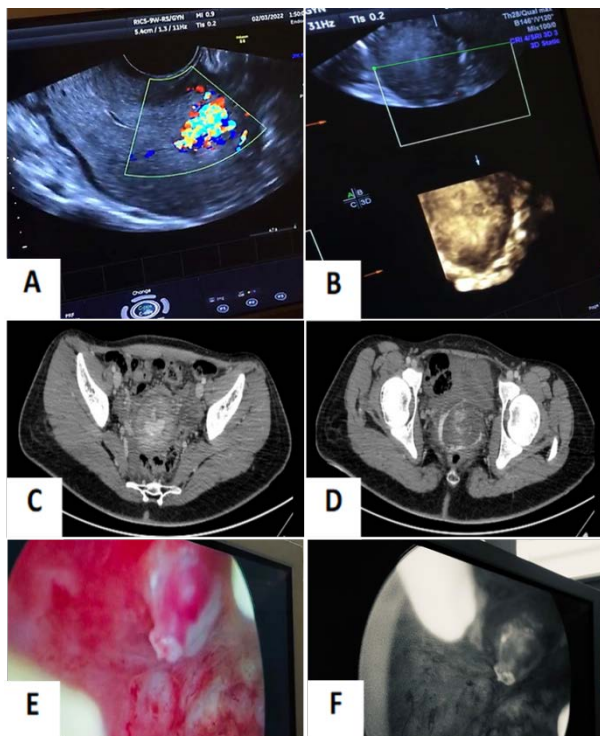
Doppler may be used as an initial diagnostic modality while an angiogram may be required for confirmation of the diagnosis [9,10]. The most common type is acquired such as following uterine surgical procedures or pregnancy; however, there are congenital types as well [11,12,13].

A hysterectomy is becoming a less common treatment option due to the development and increasing accessibility of embolization, which has become more convenient and avoids more invasive methods [14]. In this case report, we introduce a 25-year-old woman as a first case of congenital UAVM in Palestine.

## 2. Clinical Report

Our patient is a multiparous 25-year-old Palestinian woman who is gravida two, para two, with a history of two vaginal deliveries. Her first pregnancy resulted in the delivery of a live fetus at 32 weeks gestation. Meanwhile, the second pregnancy was complicated by antepartum hemorrhage. It ultimately terminated at 28 weeks gestation due to intrauterine fetal demise on November 21<sup>st</sup>, 2021, followed by postpartum hemorrhage managed appropriately. The patient underwent dilation and curettage (D&C) on January 9<sup>th</sup>, 2022 due to retained products of conception,

after which the patient began suffering from recurrent episodes of abnormal uterine bleeding that necessitated hospitalization and a blood transfusion of 4 units of packed red blood cells. The patient transferred to us from a peripheral hospital and was afterwards admitted on February 3<sup>rd</sup>, 2022 for the assessment of an abnormal uterine bleeding. The patient experienced the first episode of abnormal uterine bleeding at age of 20. initial laboratory findings on admission are as follows; hemoglobin (Hb) was 10.26 g/dl, hematocrit (HCT) was 30.8%, red blood cells (RBC) were  $3.48 \times 10^6/\mu\text{l}$ , platelet count (PLT) was  $94.9 \times 10^3/\mu\text{l}$ . The patient was suspected to have very vascular lesion on USS query retained piece of placenta (Figure 1A, Figure 1B), then she underwent urgent hysteroscopy which showed obvious pulsating artery and abnormal vascular communication (Figure 1E, Figure 1F, Figure 1B), intraoperative consultation carried out with consultant radiologist who asked for urgent computed tomography angiography (CTA) (Figure 1C, Figure 1D), and was thus sent to the radiological department to do computed tomography angiography (CTA). It revealed active contrast extravasation in the uterine cavity which increased in Porto-venous and delayed phases consistent with Uterine arterio-venous malformation (UAVM) shown to be mainly supplied by the right uterine artery. In addition, it was associated with multiple dilated peri-uterine blood vessels and veins. Lastly, a mild amount of pelvic free fluid consistent with the density of blood was also noticed (Figure 1C, Figure 1D).



**Figure 1.** (1A, 1B): Ultrasound demonstrates heterogenous myometrial echotexture due to the presence of multiple serpiginous anechoic structures within the myometrium and the color doppler shows turbulent flow within these structures. (1C, 1D): computed tomography angiography reveals active contrast extravasation in the uterine cavity. (1E, 1F): hysteroscopy shows an abnormal vascular lesion

Bilateral uterine artery embolization (UAE) with gelfoam was successfully performed while the patient still under general anesthesia all the procedures carried out

under general anesthesia (hysteroscopy, CTA and UAE) after taking consent from her family ( husband and her mother).

### 3. Discussion

Uterine arteriovenous malformations (AVMs) are rare lesions that may present with life-threatening hemorrhage if not diagnosed or treated promptly and properly. Although true incidence of such lesions still unclear, some reports suggest that to be less than 4.5%, mainly occurring in women of reproductive age [12].

An arteriovenous malformation (AVM) is an abnormal connection between an artery and a vein which bypasses the capillary system. Uterine AVMs may be congenital or acquired. Congenital AVMs are due to the arrest of normal vascular development resulting in the failure of primitive vessels to differentiate into arteries or veins [13,15].

Furthermore, congenital AVMs are supplied by multiple arteries that may be extrauterine and flow into a central tangle of vessels which drain into large veins. In contrast, acquired AVMs are supplied by single or bilateral uterine arteries and connect to the myometrial venous plexus [16].

They may be caused by infectious disease, gestational trophoblastic disease, retained products of conception, malignancy, and exposure to diethylstilbestrol [17]. It is also associated with uterine trauma such as uterine curettage [18]. It is postulated that the shunting of blood from the capillary plexus towards the venous system from hemostatic control during surgery such as, packing and clamping, gives rise to the lesion. In addition, the healing process after such procedures may result in an abnormal connection between an artery and vein [19].

We suspect that our patient's uterine AVM was congenital in nature since she had not undergone any procedures prior to her presentation. It is assumed that the uterine curettage injured, rather than resulted in the formation of the AVM since the patient had presented with abnormal uterine bleeding prior to undergoing the procedure.

Uterine AVMs most commonly present with abnormal uterine bleeding. Hence, patients may be anemic or hypotensive at presentation, such as our patient. Menses as well as, dilation and curettage procedures cause sloughing of the endometrium which exposes the abnormal vessels within the AVM, precipitating hemorrhage [20]. Therefore, dilation and curettage are contraindicated when uterine AVMs are suspected. Patients may also present with lower abdominal pain, dyspareunia, urinary frequency, and incontinence. Also, systemic hypotension may occur due to pooling of blood in the lesion [18].

Historically, uterine AVMs were diagnosed by pathologic examination of uterine tissue after hysterectomy [13,18]. However, a less invasive approach is utilized in recent times. Ultrasound remains the best initial test to aid in the diagnosis of abnormal uterine bleeding [21]. On greyscale ultrasound, uterine AVMs are visualized as anechoic spaces in the myometrium, an intramural mass, or a bulky cervix. Thus, it can be confused with gestational trophoblastic disease or retained products of conception, but They can be distinguished from one another by obtaining serum B-hCG levels [22,23].

Color Doppler ultrasound of uterine AVMs demonstrates flow, Spectral analysis exhibits high velocity flow, as well as, a low resistive index [24]. The gold standard for diagnosis, CT angiography, which displays hypertrophy of the uterine arteries and early drainage into hypertrophied veins [25].

In some and rare cases, such as ours, hysteroscopy may allow direct visualization of the lesion. while on the contrary, the majority of cases have a normal hysteroscopic outcome [26,27,28].

This is not always plausible however, since the field of visualization may be obscured by blood [29,30].

Treatment of uterine AVMs is dependent on the patient age, presentation severity, future fertility desire, and the lesion size. Asymptomatic patients may receive conservative management with medications or serial testing with ultrasound. Patients with recurrent bleeding, anemia, or hemodynamic instability who desire future fertility should be treated with uterine artery embolization [31,32]. Arterial access is gained through the femoral artery under local anesthesia. In the past, there were concerns of post-procedural infertility since uterine vascularity is decreased, thus affecting placentation and fetal growth. However, there have been multiple case reports reporting successful pregnancies following uterine artery embolization [33].

Patients may receive medical treatment while awaiting uterine artery embolization. This includes estrogens that result in endometrial proliferation, thus covering the pathologic vessels in the myometrium. Another agent is methylergonovine maleate which reduces the blood flow within the AVM and results in its collapse [14,30].

GnRH agonists may also be used, suppressing estrogen production. This decreases stimulation of estrogen receptors on endothelium, decreasing its proliferation [34].

Surgical management includes hysterectomy for those who do not desire future fertility. In addition, uterine artery ligation, or hysteroscopic or laparoscopic coagulation of the AVM may be performed [35].

## 4. Conclusion

Although rare, it is important to include uterine AVMs in the differential diagnoses of abnormal uterine bleeding to avoid undergoing dangerous procedures such as dilation and curettage that may have detrimental effects on the patient. Furthermore, early diagnosis of congenital AVMs is essential since the lesion will grow further in pregnancy, resulting in complications, such as abortion and life-threatening hemorrhage.

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