Case Report

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Uterine arteriovenous malformation: a case of challenges in diagnosis and management of a patient with a history of misdiagnosed hemorrhage

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Introduction: Uterine arteriovenous malformations (AVMs) are a rare cause of obstetrical hemorrhage. It can be congenital due to a defect during embryogenesis or acquired. Uterine AVMs can cause life threatening postpartum hemorrhage, and is most frequently misdiagnosed. This case highlights the diagnostic challenges posed by uterine arteriovenous malformation, a rare vascular anomaly that poses significant challenges in diagnosis and management.

Case presentation: This case report details the clinical presentation, diagnostic challenges, and treatment approach for a 39-year-old woman. In the absence of a medical history indicative of pre-existing ailments, the individual in question has undergone two emergency cesarean sections as documented in her surgical history, in addition to two previous dilation and curettage D&C operations. The patient presented with heavy vaginal bleeding 6 months after a cesarean section. The patient's clinical presentation, imaging findings, and intraoperative observations collectively substantiate the diagnosis of uterine AVMs.

Discussion: Women who have had uterine instrumentatio surgery, such as a cesarean section or dilatation and curettage (D&C) are more likely to develop acquired uterine AVMs. The absence of uterine artery embolism options compelled the use of alternative diagnostic methods, including contrast MRI, which successfully detected abnormal vascular lesions. The choice for hysterectomy was influenced by the patient's completion of childbearing and the presence of large vessels in proximity to critical regions.

Conclusion: This case emphasizes the significance of adapting treatment plans based on local resource constraints and the need for ongoing efforts to enhance diagnostic capabilities in undeserved regions.

Keywords: abdominal hysterectomy, postpartum bleeding, uterine, uterine arteriovenous malformation (AVM), vaginal bleeding

Introduction

Atypical vein and artery connections that avoid the capillary system are known as arteriovenous malformations, or AVMs. AVMs can be found anywhere in the body and they are classified as either central or peripheral if they are inside or outside the central nervous system. Although uterine AVMs are a rare peripheral location, the differential pressure gradient between the venous and arterial systems leads to intermittent yet excessive bleeding, which can make it a potentially fatal condition necessitating blood transfusions in up to 30% of cases[1–9]. The term 'arteriovenous aneurysm' or 'arteriovenous malformations' has been used since 1926, when Dubreil and Loubart reported the first case as a cricoid aneurism of the uterus[2,4,7,9]. The literature does not include much information regarding how frequently these lesions occur. Since 1926, less than 150 cases have been recorded in the literature; of them, only 73 cases were reported before 1997[2]. Clinical manifestations can differ from sporadic to severe vaginal bleeding. It is often detected in women who have unexplained vaginal bleeding at childbearing age, accounting for 1–2% of all genital and intraperitoneal hemorrhages[2,4,7]. Traditionally, uterine AVMs are classified as either acquired or congenital. The acquired are more

HIGHLIGHTS

- This case report highlights challenges that doctors may face in diagnosing uterine arteriovenous malformation (UAVM), in some areas suffering from the consequences of war, especially the shortage of medical equipment.
- Using ultrasound and Doppler color ultrasound in this case may indicate the effectiveness of these methods in directing and increasing suspicion of (UAVM).
- The unavailability of uterine artery embolism necessitated reliance on alternative diagnostic tools, such as contrast MRI, which proved effective in identifying abnormal vascular lesions, which may allow it to be an alternative confirmatory diagnostic method.

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common than congenital lesions. Women of reproductive age who have had uterine instrumentation surgery, such as a cesarean section or dilatation and curettage (D&C) are more likely to develop acquired uterine AVMs[1–9]. Pelvic discomfort, meno-metrorrhagia, and vaginal bleedings are common symptoms of both congenital and acquired AVMs. One possible outcome of arteriovenous shunting is pelvic varicocele. Additionally, dyspareunia and recurrent miscarriages can be indicative of uterine AVM because of increased vascularization that hinders embryonic implant. Incontinence, polyuria, and pollachiurya are frequently linked urinary symptoms[1–9]. Puberty, pregnancy, thrombosis, infection, biopsy, and injury can all lead to sudden worsening or flare-ups. Very high flow damage seldom results in high flow heart failure in young children[2]. The recommended first imaging modality for diagnosis is transvaginal ultrasonography with color flow and spectral analysis. Conventional angiography which is the gold standard for diagnosis, is mainly used for uterine artery embolization, especially in women who want to maintain their fertility[3,4]. Although it is invasive, hysteroscopy is another diagnostic tool[4]. Treatment options include selective embolization, laparoscopic coagulation of uterine arteries, hysterectomy, and hysteroscopic excision of the lesion. The conservative strategy involves medical therapy with gonadotropin-releasing hormone (GnRH) agonist, methylergonovine maleate, danazol, and NSAIDs, although it is linked to a high failure rate and, in certain cases, prolonged bleeding[4].

Case presentation

A 39-year-old gravida 7, para 5, presented to our institution with a chief complaint of persistent and profuse vaginal bleeding, amounting to the use of 6–7 sanitary pads daily, occurring 6 months post an uneventful elective cesarean section. In the absence of a medical history indicative of pre-existing ailments, the individual in question has undergone two emergency cesarean sections as documented in her surgical history because of poor fetal heart rate monitoring result, in addition to two previous dilation and curettage D&C operations. The first was done as a result of the persistence of fetal fragments after birth, while the second, conducted recently, serves as a diagnostic and therapeutic measure for persistent bleeding. However, the diagnostic results were negative, and the woman's bleeding did not improve. She presented to us in a critical clinical condition.

Pregnancy was ruled out through a negative pregnancy test.

Despite prior interventions with ergot compounds, combined oral contraceptive pills (COCPs), and Norethisterone, the patient experienced limited improvement in her bleeding symptoms. Hemodynamically, she remained stable, with a recorded hemoglobin level of 10.5, subsequently decreasing to 10.1 within 1 week. All other hematological parameters were within normal limits.

Initial transvaginal sonography revealed a uterus of normal size with an ordinary endometrial shape and thickness. However, hypoechoic areas within the uterine muscular layer adjacent to the internal cervical OS were noted. Subsequent Doppler color ultrasound examination focused on these areas, revealing marked hypervascularity and the presence of a substantial vascular mesh structure, prompting suspicion of a uterine arteriovenous malformation (AVM) or fistula (Fig. 1).

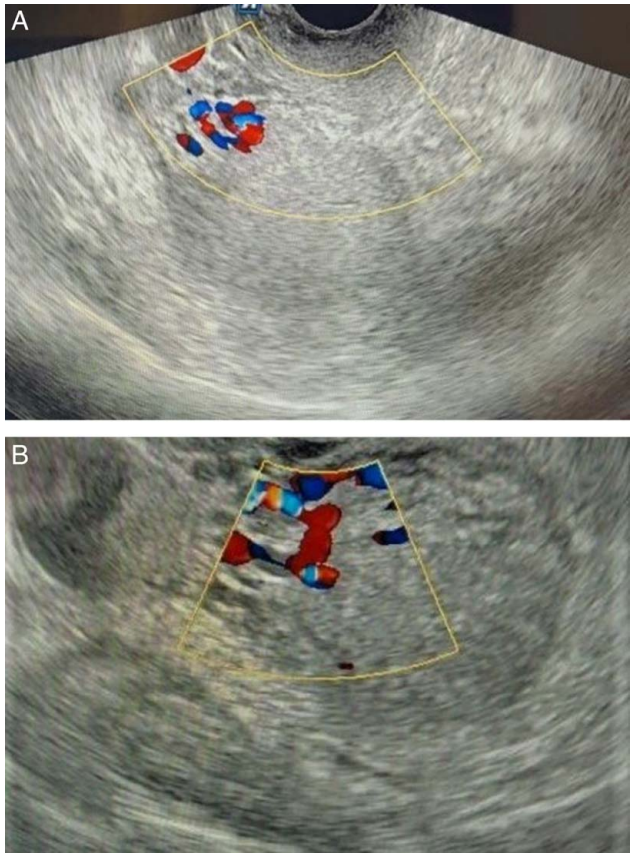

Figure 1. (A) Doppler Ultrasound image showing abnormal multidirectional vascularity near uterine isthmus. (B) close up Doppler image showing more hypoechoic (black) gaps, which show hypervascularity on color Doppler.

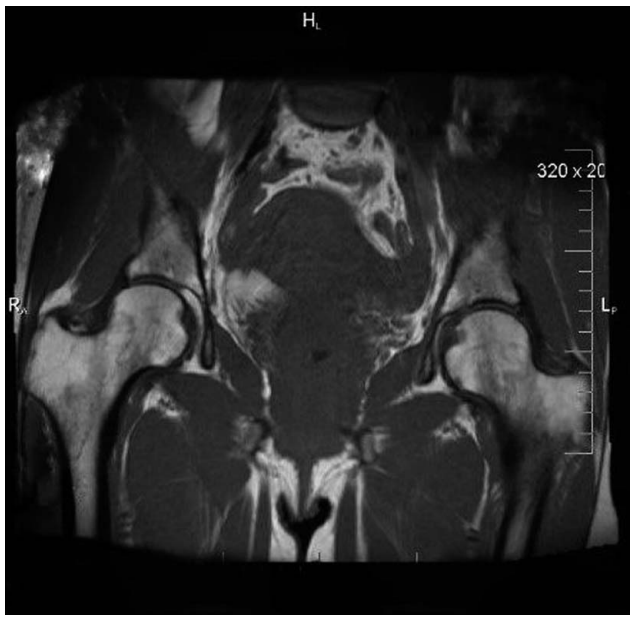

Figure 2. Pelvic MRI in coronal view coronal view demonstrating serpiginous vascular structure in bulk of the uterus with some even presenting increased contrast at t1.

While an angiogram traditionally serves as the gold standard diagnostic tool for such conditions. However, this technology is not yet available in Syria, which led to the scheduling of a contrast-enhanced MRI study.

The contrast MRI elucidated abnormal vascular lesions infiltrating the uterus, primarily localized near the internal cervical OS, with extensions reaching the posterior uterine wall (Fig. 2), (Fig. 3). In the absence of the option for uterine artery embolism within the country and considering the patient's completion of childbearing, marked by bilateral tubal ligation during the last cesarean section, an informed decision was made to proceed with an open abdominal hysterectomy without complications.

During the surgical intervention, the presence of large vessels in proximity to the prior cesarean scar and the uterine artery around the posterior uterine wall was noted (Fig. 4). Histopathological examination confirmed the diagnosis, revealing multiple vascular malformations infiltrating the uterine muscular tissue (Fig. 5).

The patient's clinical presentation, imaging findings, and intraoperative observations collectively substantiate the diagnosis of uterine AVMs, necessitating the implementation of an open abdominal hysterectomy as the definitive therapeutic measure in light of resource limitations and the patient's reproductive history.

Discussion

Uterine AVMs is an infrequent condition that can be fatal due to postpartum hemorrhage, which can vary in severity. Uterine AVMs are characterized by abnormal vascular communication between the uterine arterial and venous systems and are typically classified as congenital or acquired[3,5]. In a research investigation involving nearly a thousand patients, all women were evaluated for AMV after delivery or abortion, and only six cases were identified, resulting in an incidence rate of 1 in 1000. Due to the scarcity of cases, it remains challenging to accurately determine the true incidence and prevalence of AVMs. AVMs account for ~1–2% of all cases of genital and intraperitoneal hemorrhages[2,6,8]. Congenital cases of uterine AVM are

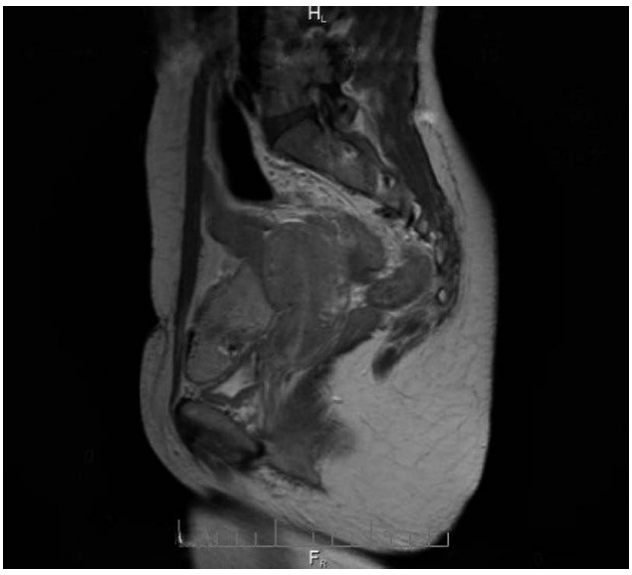

Figure 3. Sagittal MRI view showing contrast taking serpentine vessels on the posterior uterine wall.

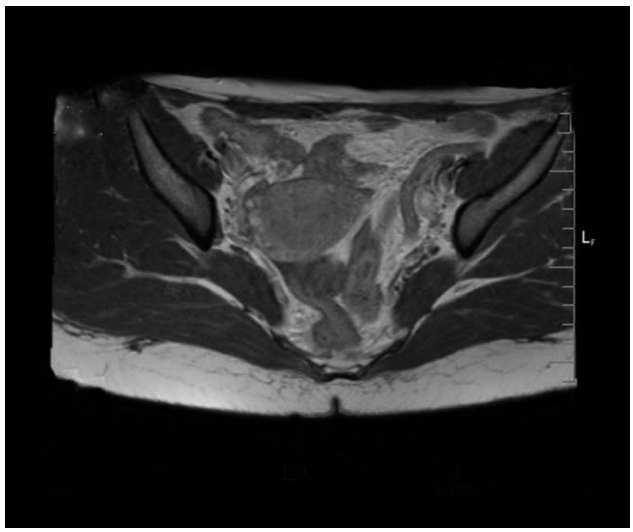

Figure 4. Uterus during surgery demonstrating high vascularity around the isthmus.

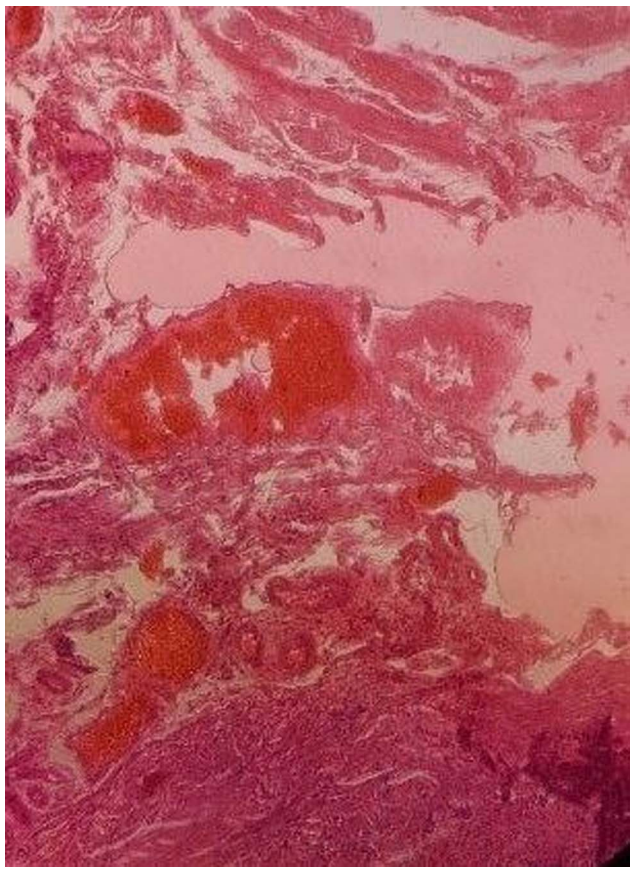

Figure 5. Histopathological examination shows multiple vascular lumens within the uterine wall near the cervical OS.

uncommon and thought to be caused by a failure of differentiation during fetal angiogenesis. while acquired cases are typically associated with gestation, uterine surgical procedures, or neoplastic diseases[3,9]. A review of 54 women with acquired uterine AVM revealed that 96% of them had a history of interventions such as dilatation, myomectomy, curettage, cesarean section, or intrauterine device placement. Nonsurgical causes, including infections, trophoblastic diseases, and uterine malignancies, have also been reported[3,7,9]. Based on information from the medical literature, it appears that in cases where the uterine vascular plexus is disrupted during an invasive procedure and surgical packing is used for hemostasis, it is crucial to ensure proper approximation of the newly formed artery and venous margins. This is important because if larger vessels are nearby without an intervening capillary plexus during the healing process, there is a risk of developing a fistulous connection when blood flow is restored. Therefore, it is essential to take all necessary precautions to prevent this complication[9]. In the patient's surgical history, there are two cesarean sections, in addition to two previous dilation and curettage D&C operations, so most probably that the cause of AVMs is acquired.

A typical patient with uterine AVM is a multiparous woman in her 30s who experiences symptoms such as menometrorrhagia, vaginal bleeding, and pelvic pain. Both acquired and congenital AVMs can present with these symptoms[7].

Diagnosing uterine AVMs requires two initial steps, imaging and laboratory tests. Imaging includes ultrasound (US), MRI, computed tomography (CT), and angiography, which is the gold standard[7]. Beginning with transvaginal US, hypoechoic areas are usually detected which in our case were found near the internal cervical OS. Hypervascularity with big vascular mesh structure was visible when Doppler color US was applied. The unavailability of angiography in Syrian hospitals posed a serious challenge for doctors in order to reach the definitive diagnosis, therefore contrast MRI was used and unusual vascular lesions were found, some of which extended up the posterior uterine wall and infiltrated the uterus close to the internal cervical OS. These findings led to diagnosis of the Uterine AV malformations despite the absence of Angiography.

Management of Uterine AVM varies from a patient to another depending on their age, level of bleeding and hemodynamic status, with putting the patient's desire for future fertility into consideration. Therefore, in patients seeking pregnancy, hysterectomy, which is done traditionally–should be avoided and other treatment alternatives should be considered, such as medical treatment with oral and intramuscular methylergonovine, oral contraceptive pills (OCPs), methotrexate, or misoprostol[10]. This is called conservative treatment and can be used in small lesion with peak systolic value <60 cm/s[7]. In addition to embolisation treatment, particularly in acute cases in order to stop blood loss and stabilize the hemodynamic status[10]. Unfortunately, this technique is not available in Syria, which prompted us to exclude this method and resort to an open abdominal hysterectomy after discussing the plan with our 39-year-old patient, who has already given birth and completed childbearing after performing a bilateral tubal ligation during her most recent C-section.

The most common complications of hysterectomy can be categorized as infectious, venous thromboembolic, genitourinary (GU), and gastrointestinal (GI) tract injury, bleeding, nerve injury, and vaginal cuff dehiscence. Injury to the GI tract after hysterectomy is the less common, with a range of 0.1–1%. Bleeding complications after hysterectomy also are rare[11]. The work has been reported in line with the SCARE (Surgical CAse REport) 2023 criteria[12].

Conclusion

This case highlights the diagnostic challenges posed by uterine arteriovenous malformation, a rare vascular anomaly that poses significant challenges in diagnosis and management.

The unavailability of uterine artery embolism necessitated reliance on alternative diagnostic tools, such as contrast MRI, which proved effective in identifying abnormal vascular lesions. The decision for an open abdominal hysterectomy was guided by the patient's completed childbearing and large vessels near-critical regions. This case emphasizes the significance of adapting treatment plans based on local resource constraints and the need for ongoing efforts to enhance diagnostic capabilities in undeserved regions. In line with our findings in medical literature, it is imperative to address such issues, as preserving a woman's reproductive health is a crucial matter. Overcoming it is not an easy task. A woman's life and her ability to conceive are fundamental aspects.

Ethical approval

Our institution does not require ethical approval for reporting individual cases or case series.

Consent

Written informed consent was obtained from the patient's parents/legal guardian for publication and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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Author contribution

F.N., G.K., and F.A.: contributed to drafting, reviewing, editing, and approved the final manuscript; L.R.: contributed to drafting, editing, and approved the final manuscript; N.G.: contributed to drafting, reviewing, editing, corresponding, and approved the final manuscript; H.A.: contributed to reviewing, supervising, and approved the final manuscript; M.H.: contributed to drafting, editing, and approved the final manuscript; S.L.: contributed to drafting, editing, and approved the final manuscript. All authors read and approved the final manuscript.

Conflicts of interest disclosures

None.

Guarantor

All authors have read and approved the manuscript, on behalf of all the contributors I will act and guarantor and will correspond with the journal from this point onward.

Data availability statement

Data sharing is not applicable to this article.

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