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RARE VASCULAR DISEASES OF THE FEMALE REPRODUCTIVE TRACT: A LITERATURE REVIEW ON THE ETIOLOGY, DIAGNOSIS, AND TREATMENT OF UTERINE ARTERIOVENOUS MALFORMATIONS AND VULVAR ANEURYSMS

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ABSTRACT

Vascular disorders of the female reproductive tract, including uterine arteriovenous malformations (AVMs) and aneurysms or pseudoaneurysms of the uterine and pudendal arteries, represent extremely rare but potentially life-threatening conditions in gynecology. Due to their low incidence and nonspecific clinical presentation, these entities pose significant diagnostic and therapeutic challenges. Although uncommon, they may result in severe hemorrhage, particularly in the postpartum period or following intrauterine procedures.

The aim of this narrative review is to summarize current evidence regarding the etiology, clinical manifestations, diagnostic imaging modalities, and treatment strategies for rare vascular abnormalities of the female reproductive system. A literature search was conducted using PubMed, Scopus, and Web of Science databases, including articles published between 2000 and 2025.

Available data indicate that color Doppler ultrasonography is the primary diagnostic tool, while magnetic resonance imaging and angiography play a crucial role in confirmation and treatment planning. Selective endovascular embolization has emerged as the gold-standard therapy, providing high efficacy in hemorrhage control while preserving uterine integrity and reproductive potential in most patients. Despite favorable outcomes, standardized diagnostic and therapeutic algorithms are lacking. Further multicenter studies and case registries are required to optimize management strategies for these rare but clinically significant conditions.

KEYWORDS

Uterine Arteriovenous Malformation, Uterine Artery Pseudoaneurysm, Vulvar Aneurysm, Endovascular Embolization, Abnormal Uterine Bleeding

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Introduction

Rare vascular diseases of the female reproductive tract, including uterine arteriovenous malformations (AVMs) and aneurysms or pseudoaneurysms of the uterine and pudendal arteries, constitute a significant diagnostic and therapeutic challenge in gynecology [2,5,14]. Although infrequent, these conditions may lead to severe complications such as recurrent or massive genital bleeding, anemia, and hemorrhagic shock, particularly in the postpartum period or after gynecological procedures [2,4,5,18]. Their nonspecific clinical presentation often results in delayed or incorrect diagnosis [8].

Uterine AVMs are characterized by abnormal direct connections between arteries and veins without an intervening capillary network. They may be congenital or, more commonly, acquired following pregnancy-related events, uterine curettage, cesarean section, hysteroscopic interventions, or electrosurgical procedures [2,4,6]. Aneurysms and pseudoaneurysms of the uterine and pudendal arteries usually arise from vascular wall injury, leading to focal dilation or extravascular blood collection surrounded by soft tissues [5–7,11]. These pathologies may mimic neoplastic lesions of the uterus, further complicating differential diagnosis [8].

Advances in imaging techniques, particularly Doppler ultrasonography, computed tomography, and magnetic resonance imaging, have significantly improved the detection of uterine and vulvar vascular abnormalities [2,3,14]. Concurrently, progress in interventional radiology has transformed therapeutic approaches. Selective or superselective endovascular embolization is now considered the treatment of choice, enabling effective hemorrhage control while preserving the uterus and reproductive potential [1,9,12,16]. Surgical management, including hysterectomy, is currently reserved for emergency situations or failure of minimally invasive therapy [2,14].

Methodology

A narrative literature review was conducted to identify relevant publications concerning rare vascular diseases of the female reproductive tract. The PubMed, Scopus, and Web of Science databases were searched for articles published between 2000 and 2025. Search terms included combinations of: *uterine arteriovenous malformation*, *uterine artery pseudoaneurysm*, *vulvar aneurysm*, *embolization*, and *postpartum hemorrhage*. Original articles, case reports, case series, and review papers published in English were included. As this study was based solely on published literature, ethics committee approval was not required.

Results

Etiology

The etiology of uterine and vulvar vascular abnormalities is complex and involves both congenital and acquired mechanisms. The vast majority of reported cases represent acquired lesions, frequently associated with iatrogenic trauma or pregnancy-related events [2,14].

Uterine AVMs consist of abnormal arteriovenous connections bypassing the capillary bed. Congenital forms result from aberrant angiogenesis during embryonic development and are rarely reported. Acquired AVMs are far more common and typically develop following childbirth, miscarriage, uterine curettage, cesarean section, hysteroscopic procedures, or electrosurgical interventions such as loop electrosurgical excision procedures (LEEP) [2,4,6,18]. Vascular injury, reparative angiogenesis, and hormonal influences are believed to play key roles in their pathogenesis [2].

Aneurysms and pseudoaneurysms of the uterine and pudendal arteries are exceedingly rare and almost exclusively acquired. Pseudoaneurysms result from disruption of the arterial wall with subsequent blood extravasation into surrounding tissues, most often after obstetric trauma or gynecological procedures [5–7,11]. Clinical symptoms may be delayed, contributing to diagnostic difficulties. Rare associations with coexisting conditions such as deep infiltrating endometriosis have also been described [7].

Pudendal and ovarian artery aneurysms typically present as postpartum complications, related to increased blood flow, hemodynamic changes, and the effects of steroid hormones on vascular wall integrity [13,15]. Increased recognition of these rare entities is likely related to the growing use of advanced imaging and interventional techniques in gynecological practice [2,14].

Clinical Manifestations

The most common clinical presentation of uterine and vulvar vascular abnormalities is abnormal genital bleeding. This may be recurrent, profuse, or refractory to conservative treatment [2,4,18]. In postpartum patients or following gynecological procedures, these lesions can cause massive hemorrhage, leading to severe anemia or hemorrhagic shock [5,11,18].

In some cases, AVMs and pseudoaneurysms remain asymptomatic and are detected accidentally during imaging performed for abnormal uterine bleeding [8,18]. Less frequently, pelvic pain, local discomfort, or pressure symptoms may occur, particularly in patients with larger vulvar aneurysms or hematomas [13,17]. Importantly, these vascular lesions may clinically and radiologically mimic uterine tumors or fibroids, complicating diagnostic evaluation [8].

Diagnostic Evaluation

Diagnosis of rare vascular diseases of the female reproductive tract remains challenging due to their rarity and nonspecific clinical features. Transvaginal ultrasonography with color Doppler assessment is the primary diagnostic modality. Typical findings in uterine AVMs include multiple anechoic spaces within the myometrium with turbulent, high-velocity, low-resistance blood flow, producing a characteristic “mosaic” pattern [3,20]. This technique allows differentiation from fibroids, retained products of conception, or malignancy [3,14]. In pseudoaneurysms, the classic “yin–yang” flow pattern may be observed [5,6,11].

Contrast-enhanced computed tomography and magnetic resonance imaging serve as complementary tools, particularly in equivocal cases or when extensive disease is suspected. These modalities facilitate assessment of lesion extent and relationships with adjacent structures, and assist in excluding neoplastic processes [2,8]. Magnetic resonance imaging is especially valuable in women of reproductive age due to the absence of ionizing radiation [2,14].

Selective angiography remains the diagnostic gold standard, enabling precise identification of pathological vessels and simultaneous endovascular treatment [1,2,9]. Given its invasive nature, angiography is generally reserved for patients requiring intervention or presenting with acute hemorrhage [4]. Accurate diagnosis is critical, as invasive uterine procedures performed without excluding vascular pathology may precipitate catastrophic bleeding [2,18].

Table 1. Etiology and Clinical Presentation of Rare Vascular Diseases of the Female Reproductive Tract

Pathology	Predominant Etiology	Typical Clinical Presentation	Key References
Uterine AVM	Acquired (postpartum, curettage, cesarean section, hysteroscopy, LEEP)	Abnormal uterine bleeding, postpartum hemorrhage	[2,4,6,18]
Uterine artery pseudoaneurysm	Vascular injury after delivery or procedures	Secondary postpartum hemorrhage	[5,6,11]
Vulvar/pudendal artery pseudoaneurysm	Obstetric trauma, surgical injury	Vulvar pain, swelling, hematoma, rupture	[13,17]
Congenital uterine AVM	Developmental vascular anomaly (rare)	Recurrent or incidental bleeding	[14]

Treatment: Endovascular Embolization

Endovascular embolization is currently considered the first-line treatment for most uterine and vulvar vascular abnormalities, including uterine AVMs and aneurysms or pseudoaneurysms of the uterine and pudendal arteries [1,2,14]. This minimally invasive approach enables effective hemorrhage control while preserving the uterus, which is particularly important for women desiring future fertility [1].

Indications and Technique

Indications for embolization include symptomatic uterine AVMs presenting with heavy or recurrent bleeding, postpartum hemorrhage, and anemia unresponsive to conservative management [2,4,18]. Embolization is also the preferred treatment for uterine and vulvar artery pseudoaneurysms, particularly following obstetric or gynecological procedures [5–7,11,13]. The procedure is performed percutaneously, usually via femoral artery access, following selective angiography to identify and embolize the pathological vessels [2,9,12].

Embolic Materials

Various embolic agents are used, including polyvinyl alcohol particles, metallic coils, tissue adhesives, absolute alcohol, and resorbable materials such as gelatin sponge [1,9,14]. Selection of embolic material depends on lesion characteristics, vascular anatomy, and the patient's reproductive plans. Recent studies suggest that temporary embolic agents may better preserve uterine perfusion and reproductive function [16].

Efficacy, Safety, and Fertility Outcomes

Systematic reviews and case series report high success rates of embolization in controlling hemorrhage associated with uterine AVMs, with most patients requiring only a single procedure [1,4,12]. Treatment of pseudoaneurysms is similarly effective, often resulting in immediate symptom resolution [5,6,11,13]. Although embolization is generally safe, rare complications such as uterine necrosis or recurrent bleeding have been reported, particularly following bilateral embolization or use of permanent agents [10].

Preservation of fertility is a major advantage of embolization. Numerous cases of successful pregnancies following treatment have been described in the literature [1,2,12]. Nonetheless, careful patient selection and close obstetric monitoring in subsequent pregnancies are recommended due to limited long-term data [1,10].

Table 2. Diagnostic Modalities and Treatment Outcomes of rare vascular diseases of the female reproductive tract

Modality / Treatment	Role	Key Findings	References
Color Doppler ultrasound	First-line diagnosis	High-velocity, low-resistance turbulent flow	[3,20]
MRI / CT	Adjunct imaging	Lesion extent, differentiation from tumors	[2,8,14]
Selective angiography	Gold standard	Definitive diagnosis and treatment planning	[1,9]
Endovascular embolization	First-line therapy	~85–90% success, fertility preservation	[1,4,12,16]
Surgical treatment	Second-line / emergency	Reserved for failed embolization	[2,14]

Discussion

Rare vascular diseases of the female reproductive tract, including uterine AVMs and uterine or vulvar artery aneurysms, are uncommon but can cause life-threatening bleeding. Diagnosis is often delayed due to nonspecific symptoms and similarity to more common gynecological conditions.

Imaging advances have improved detection. Color Doppler ultrasound is the primary screening tool, while MRI aids in differentiating vascular anomalies from tumors. Selective angiography remains the gold standard for both diagnosis and interventional planning.

Endovascular embolization has emerged as the preferred treatment, offering high success rates, rapid hemorrhage control, and fertility preservation. Complications are rare but underscore the need for individualized planning.

Despite these advances, evidence is limited to case reports and small series. Long-term reproductive outcomes and standardized protocols remain poorly defined. Multidisciplinary collaboration and further prospective studies are essential to optimize management and establish evidence-based guidelines for these rare but potentially serious conditions.

Conclusions

1. Rare vascular diseases of the female reproductive tract should be considered in the differential diagnosis of abnormal uterine bleeding, particularly in postpartum patients and following gynecological procedures.

2. Doppler ultrasonography is the primary diagnostic modality, while selective angiography remains the gold standard for definitive diagnosis and treatment.

3. Endovascular embolization is the treatment of choice for uterine AVMs and uterine or vulvar artery aneurysms and pseudoaneurysms, offering high efficacy and fertility preservation.

4. Surgical intervention should be reserved for emergency cases or failure of minimally invasive therapy.

Further prospective studies and case registries are essential to develop standardized diagnostic and therapeutic algorithms.

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