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Rare Case of Vulval Arteriovenous Malformation

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Abstract

Vascular malformations are rarely located in the female lower genital tract. Vulvar arteriovenous malformation is rare and complex to manage because they are associated with varied presentation and their association with numerous complications like ulceration, bleeding, vulvar distortion etc. Presenting a very rare case of vulvar arteriovenous malformation in a 36 years young female patient who presented in emergency as post coital bleeding, had hemorrhagic hypovolemic shock, suspected on ultrasound, operated, needed intensive care management and histopathology of vulva clinched the diagnosis of such a rare case with rare presentation.

Keywords: Arteriovenous Malformation (AM), Venous Malformation (VM), Vulva Magnetic Resonance Imaging (MRI)

1. Introduction

AVM belongs to disorders known as vascular malformations. AVM belongs to the venous malformation which can occur anywhere in the body including Pelvis. AVM are the defects of circulatory system that generally arise during embryonic/fetal development or soon after the birth. Most common site for AVM is spinal cord and brain. AVM may be asymptomatic. Incidence is 1 in 100000 people per year and prevalence is 18/100000 people per year. Most AVM bleeds between 10-55 years. Diagnosis of AVM is by symptoms (Mass, increase in size of mass after trauma). Hemorrhagic, ultrasound and MRI are diagnostic. Pelvic AVM are mainly noted in uterus, vulva (Labia Majora, Mons Pubis, Labia Minora) and perineum. Pelvic AVM / Labia Majora AVM is very rare and can be asymptomatic or can cause abnormal and potentially threatened vaginal bleeding, Labia Majora AVM is very rare with less than 150 reported in the literature. Presenting here with a rare case of Labia Majora AVM who presented as post coital bleeding and necessitated intensive medical and surgical treatment.

2. Case Report

36-year-old para 4 patient came to the emergency department in late January 2025 at midnight with complaints of post coital bleeding since afternoon 4 PM, i.e. for 8 hours. Patient had sexual intercourse at 2 PM and later she noticed bleeding at 4 PM. She went to multiple facilities and was advised to have cold compression, admission for observation and she finally came to NMC Al Nahda hospital in the emergency at 00.17 hours. She was para 4, three normal delivery and last cesarean section. There was no history of forceful sexual intercourse, no history of physical abuse or trauma or foreign body insertion. On asking, the patient said that her bowel and bladder habits were normal, but she could not pass urine since 4 PM. Her menstrual history was normal. LMP was 15/01/2025. In past history, patient informed us that after her third delivery she had small swelling in the right side of labia and was not increasing in size. She had a history of fall inside the bus and hitting with some object at labial site 15 days back, but there was no bleeding per vagina, there was minimal pain on right side of vulval region since then. On examination, she was anxious, afebrile, well oriented in time and space. There were no marks of injury over our body. Per abdomen was soft no mass, scar of LSCS was noted. There was no tenderness in the abdomen. On the local exam examination swelling was noted on Mons Pubis with discoloration of skin. There was no tenderness. Right labial large swelling of 10/8 cm was seen, color was bluish red. It was extending to the left side and covering the left labia and skin over it was bluish red color. The skin over labia minora had split. The left side of labia was obsessed due to large swelling /

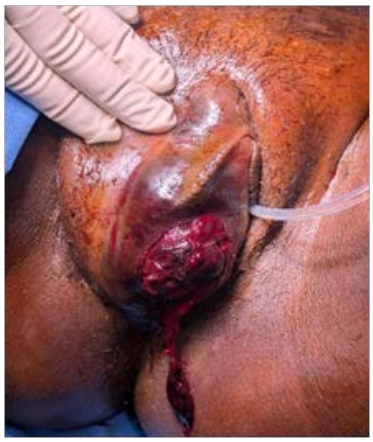
hematoma. There was no tenderness. There was no active bleeding. The Gentle Per vaginal and per rectal examination was done. Her bladder was drained with foleys catheter and 800 ML of urine drained. There with clinical diagnosis of large vulval hematoma right side secondary to? Post Coital trauma? Vulval varicosities? Vascular malformation. Patient and her husband / relatives were counseled for admission, investigations, and management. Patient investigations were carried out. CBC, CRP and USG abdomen, and Pelvis. CBC hemoglobin 9.9, hematocrit 31, platelet adequate. USG abdomen shows large oval vascular hemorrhagic lesion 6.3/5/6 cm in subcutaneous plane of right vulva with internal - vessels with Doppler signal of low systolic flow suggestive of vulval hemangioma or Venous malformation of vulva left side. MRI was suggested. Patient was started on IV fluid and investigations were sent and management plan was discussed with relatives - admission, examination under anesthesia, evacuation of hematoma. Patient had sudden fainting attacks and giddiness and bout of bleeding from labia and hypotension and was shifted to Operation Theater from the emergency department. Under general anesthesia cleaning and draping was done. Large right side vulval hematoma of size9/9/7 was noted extending all over Labia Majora and Minora and extending into mons pubis. Left side vulva was obscured by large hematomata. Hematoma was explored, evacuation of hematoma was done, active bleeding vessels, where ligated and complete hemostasis was achieved, that space was obliterated with 2.0vicryl. In the skin over labia was— bluish discoloration presenting edges were freshened and sent for histopathological examination. On per speculum examination cervix was healthy, anterior, posterior and lateral vaginal fornices were normal. There was no evidence of any tear or injury. Urethra and clitoris were normal. Wound was packed and fully cut. Foley catheter was continued.

2.1. Ultrasound (USG) Description of AV Malformation – Right Vulva: Grayscale (B-Mode)

• **Findings:** Ill-defined, heterogeneous, soft tissue lesion in the right vulvar region. Multiple anechoic or hypoechoic tubular structures within the lesion. No distinct mass or solid component.

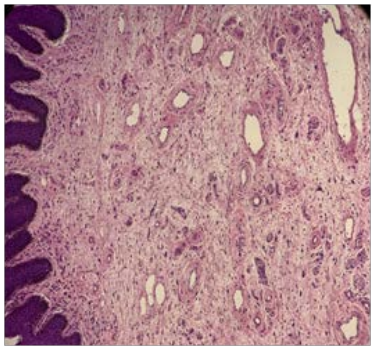
• **Color Doppler Findings:** Markedly increased vascularity with tortuous, serpiginous vessels., Multidirectional turbulent flow (aliasing may be present), Low-resistance, high-velocity flow pattern, Arterialization of venous flow may be observed.

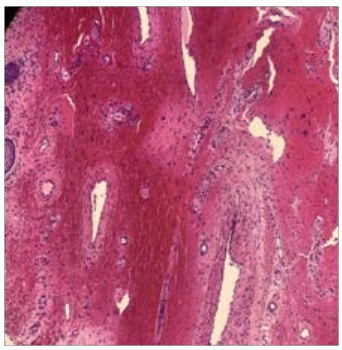
• **Spectral Doppler:** High peak systolic velocities. Low resistive index (RI), consistent with arteriovenous shunting. Findings are suggestive of a high-flow vascular malformation, consistent with an arteriovenous malformation (AVM) involving the right vulvar region. Further evaluation with MRI/MRA or CT angiography is recommended for anatomical delineation and treatment planning.



Preoperative presentation

Post operative





Squamous mucosa with subepithelial proliferation of blood vessel and nerve bundles. Blood vessels with areas of haemorrhage

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3. Discussion

3.1. Epidemiology

AVMs of the external genitalia are extremely rare, with only isolated case reports and small series published. Most reported cases occur in reproductive-age women, though they may present at any age. There is no definitive data on incidence or prevalence due to the rarity and under-reporting.

3.2. Etiology and Pathogenesis

Congenital AVMs result from aberrant embryonic development of the vascular system. Acquired AVMs may occur due to trauma, surgery (e.g., episiotomy or vulvar surgery), or obstetric procedures. Hormonal influences, especially during pregnancy, may exacerbate existing AVMs.

3.3. Clinical Presentation

Painless or painful vulvar swelling, Palpable pulsatile mass, Intermittent or persistent bleeding, Vulvar varicosities or discoloration, May mimic Bartholin's cyst, varicocele, or hemangioma.

3.4. Diagnostic Modalities

Doppler ultrasound is the first-line modality, showing high-flow vascular lesions with turbulent flow. MRI/MRA provides detailed views of soft tissue and vascular anatomy. CT angiography is particularly useful in preoperative planning. Digital subtraction angiography (DSA) is considered the gold standard for diagnosis and planning endovascular treatment.

3.5. Differential Diagnoses

The differential diagnoses for AVMs in the external genitalia include vulvar varicosities, hemangioma, Bartholin's cyst or abscess, lymphangioma, malignancies, and pelvic congestion syndrome. AVM belongs to the age of disorders known as vascular malformation (VM). Vascular malformation occurs anywhere in the body including female pelvis. They are best categorized according to the biological classification proposed by Milliken and Glowacki in 1982. It is according to their flow characteristic (fast/slow) and their vascular channel component. (Capillary, vein lymphatic, AV combined). They can involve any organ and tissue plane and occur in focal and different form. Vascular malformation (VM) is commonly symptomatic VM. Typically, there anomalies are caused by germline or somatic mutation in TIE2 gene which is involved in signaling between endothelial and mesenchymal cells during vasculogenesis and angiogenesis. They (VM) affect all tissue layers and can be focal multifocal or differ. AVM belongs to a group of disorders called VM. Most common site for AVM is spinal cord and brain.

AVM are defects of circulatory system that generally arise during embryonic or factor development or soon after birth. They mainly consist of abnormal blood vessels. There are three major groups of AVM.

- Truncal - Seen in hand, neck, VPP and corral, limb and pelvis.
- Diffuse- Common in lower limb.
- Local- Seen in any organ.

Congenital VM affects approximately 0.3 to 0.5% of population. There are benign lesions arising from developmental abnormalities of vessels during interactive growth and differ from vascular tumors like hemangioma. AVM are present at birth, but they are often not detected until symptoms appear later in life. Incidence of AVM in 1 in 100000 people per year and overall prevalence is 18/100000 people per year. AVM lesions may be located superficially with only minimal AV shorting or more deeply with significant film AV shutting. Some people are born with nidus and as the years pass by It tends to enlarge as the pressure of arterial vessel cannot be handled by vein that drain out of it. Most AVM bleeds between the age 10-55 years (Hemorrhagic 3-4%) after the age of 55 chance of bleeding is significantly low. The AVM typically evolves overtime. Changing from asymptomatic lesions with minimal lesions to large one and this evolution is often stimulated by trauma or hormonal change (Puberty/Pregnancy). It can be diagnosed by its clinical and imaging pictures, on physical examination, skin and bluish discoloration, bruit thrill. USG and Color Doppler are useful to detect, and MRI is diagnostic.

- Ultrasound findings indicate Arteriovenous Malformation (AVM).
- Magnetic Resonance Imaging findings indicate Arteriovenous Malformation (AVM).

Pure vascular malformation of vessels is rare VM of uterus and ovaries are typically associated with ovarian vein insufficiency. Pelvic AVM can be noted in Uterus, Vulva (Labia majora, Mons pubis, Labia minora and Perineum). Pelvic AVM and uterine AVM are rare and cause abnormal and potentially threatened vagina. Pelvic AVM are benign, rare and are associated with severe pain, vaginal or rectal bleeding, hematuria and typically followed by trauma. AVM of labia majora, very rare less than 150 cases reported in the lifespan. Natural history can either be quiescent (never present) or expansion (Soft tissue mass, Dark bone). Destruction- often precipitated by trauma so present as ulcer, bleeding. Common differential diagnosis is hemangioma, ulcer, growth (Benign / Malignant) Intrauterine AVM typically arise after pregnancy or after uterine trauma. Among asymptomatic women it is diagnosis by USG.

3.6. Histopathological Features of AVM

Gross examination revealed an irregular soft tissue mass, reddish-purple in color, with a spongy to firm consistency and areas of hemorrhage. The size of the mass varies depending on the extent, but no encapsulation is seen. Microscopic findings showed abnormal, dilated, and tortuous vascular channels interspersed in fibrous connective tissue. The vessels demonstrated direct connections between arteries and veins without an intervening capillary network. Arterial components included thick-walled vessels with prominent muscular layers, while venous components were characterized by thin-walled, dilated channels with

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irregular outlines. The endothelium was typically intact, with no evidence of malignancy or atypia. Focal areas showed thrombosis, hemorrhage, or fibrosis, depending on chronicity. The surrounding stroma exhibited congestion, edema, and inflammatory infiltrates, especially in symptomatic lesions.

4. Management Strategies

Conservative management is recommended for asymptomatic or minimally symptomatic lesions. Endovascular embolization is preferred for symptomatic lesions and may be curative or adjunctive before surgery. Surgical resection is considered for accessible, localized AVMs or when embolization fails. A multidisciplinary approach involving gynecology, interventional radiology, and vascular surgery is often necessary.

4.1. Pregnancy Considerations

AVMs can increase in size and become symptomatic due to hormonal and hemodynamic changes. There is a risk of rupture and hemorrhage during labor or delivery. Close monitoring with imaging during pregnancy is essential. The mode of delivery should be individualized; a cesarean section may be considered in cases of large or symptomatic AVMs.

4.2. Treatment of AVM in Complex Multidisciplinary Team Approach.

Treatment goal is to cure, manage pain and manage bleeding. MRI contrast is useful in detection among asymptomatic women diagnosed by USG the AVM resolves potentially in high % cases so conservative management t is recommended. Women presenting with bleeding, may needs ligation of bleeding vessel. Selective inter arterial embolization, resection and reconstruction with tissue flaps: AVM managed by embolization or sclerotherapy.

4.3. Prognosis

Recurrence is possible, especially if the AVM is not completely resected or embolized. Early diagnosis and appropriate treatment yields favorable outcomes. Long-term follow-up is recommended due to the potential for recurrence or complications. Most patients living with AVM can live a long life. Some individuals live with an AVM without it being diagnosed. 2-4 % risk of hemorrhage is present. If AVM ruptured survival rate is 90% [1-9].

5. Conclusion

Arteriovenous malformations (AVMs) in the female lower genital tract are rarely reported. Specifically, AVMs of the labia majora have fewer than 150 reported cases in a lifetime. The presentation of AVMs can range from asymptomatic to small masses that may become symptomatic, increasing in size with pain and bleeding, usually following trauma or during and after pregnancy. The occurrence of vulvar AVMs during pregnancy is rarely reported and remains largely unknown. Pelvic AVMs are rare and pose significant diagnostic and therapeutic challenges. They can affect individuals of any age, and symptoms can vary. The most frequent symptoms include palpable mass and pain with bleeding. Vulvar malformations are congenital lesions that may enlarge and become symptomatic at a young age, with more symptoms noted before the age of 55. Vulvar AVMs should be differentiated from vulvar varicosities, which are typically associated with pregnancy. Rapid enlargement of AVMs can occur following trauma, thrombosis, and physiological hormonal changes. Symptomatic AVMs of the external genitals are rarely reported. Main objective is to increase awareness regarding rare case of Vulval AVM necessitating surgical excision, ICU admission and blood transfusion.

Acknowledgment

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