

CONGENITAL UTERINE ARTERIOVENOUS MALFORMATION

Syed Mehdi Raza Rizvi, Anjum Mehdi, Shama Aslam, Amir Hayat Mahais, Hassan Bukhari

Department of Diagnostic Imaging and Radiology, Allied Hospital, PMC, Faisalabad, Pakistan.

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ABSTRACT

Uterine Arterio-venous malformation (AVM) is a rare but potentially life-threatening source of bleeding. A high index of suspicion and accurate diagnosis of the condition in a timely manner are essential because instrumentation that is often used for other sources of uterine bleeding can lead to massive haemorrhage. Although angiography remains the gold standard for diagnosis, ultrasound (US) and magnetic resonance imaging (MRI) are the modalities of choice for the evaluation of a suspected AVM. Up till now in literature all cases of acquired AVM are reported with a very few cases of congenital AVM.

Keywords: AVM(Arterio-venous malformation), MR Angiogram, Colour Doppler.

Case Report

An 18 year old girl, unmarried, having no history of conception presented with complaints of episodic profuse vaginal bleeding. She was having no history of previous surgery. Trans-abdominal ultrasound of that girl revealed bulky uterus and multiple tortuous tubular and serpiginous anechoic spaces throughout the myometrium more marked in the uterine fundus and anterior wall. (Fig. 1)



Figure 1: Gray scale pelvic ultrasound showing multiple anechoic spaces throughout the uterine myometrium, most prominent in the fundus.

Colour Doppler US scan showed intense flow with colour aliasing and apparent flow reversals (Fig. 2).

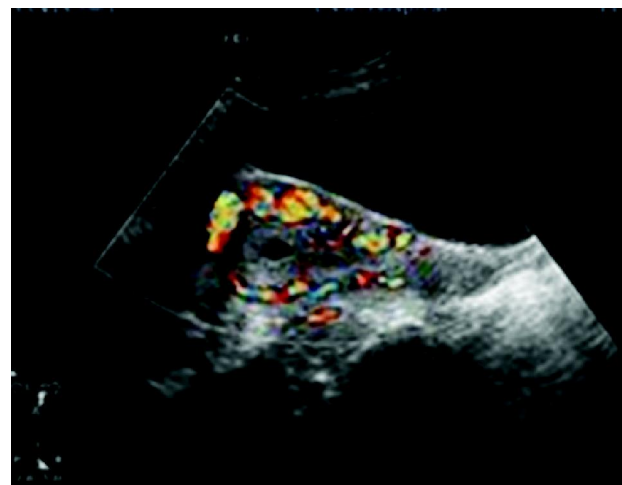


Figure 2: Colour Doppler US scan showing intense colour fill in within the anechoic spaces. There is evidence of colour aliasing and apparent flow reversals.

Spectral Doppler revealed pulsatile high velocity venous waveforms with differentiation between arterial and venous waveform difficult as signals are perceived simultaneously above and below the baseline because of proximity of the vessels (Fig. 3).

Correspondence : Dr. Amir Hayat Mahais
Department of Diagnostic
Imaging and Radiology,
Allied Hospital, PMC,
Faisalabad, Pakistan.
Email: dramirhayat@gmail.com

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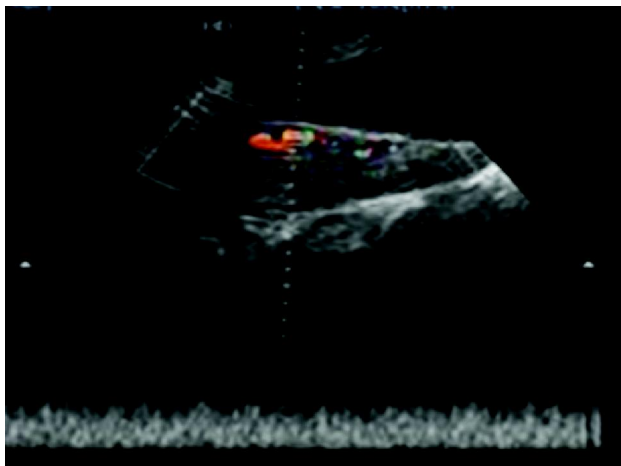


Figure 3: Spectral colour Doppler showing, pulsatile high velocity venous waveforms.

Based on the ultrasound findings a diagnosis of uterine Arterio-venous malformation was considered. The patient was taken for a MR examination Axial T1 spin echo MR image showed a bulky uterine fundus with multiple tortuous flow related signal voids. Axial and coronal T2W MR Image revealed multiple flow voids in the entire myometrium with disruption of the junctional zone. The endometrium was normal (Fig. 4 to 6).

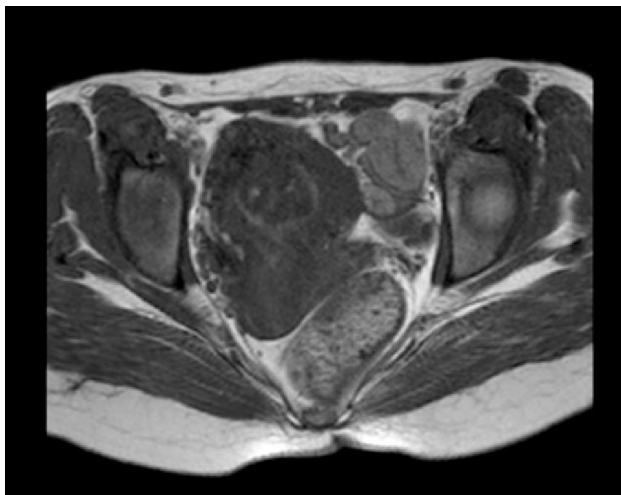


Figure 4: Axial T1W spin echo MR image showing a bulky uterine fundus with multiple tortuous flow related signal voids within the myometrium.

MR angiogram revealed feeder arteries from the bilateral internal iliac arteries and early visualization of the pelvic veins with opacification of the right common iliac vein and then the IVC (Fig. 7).

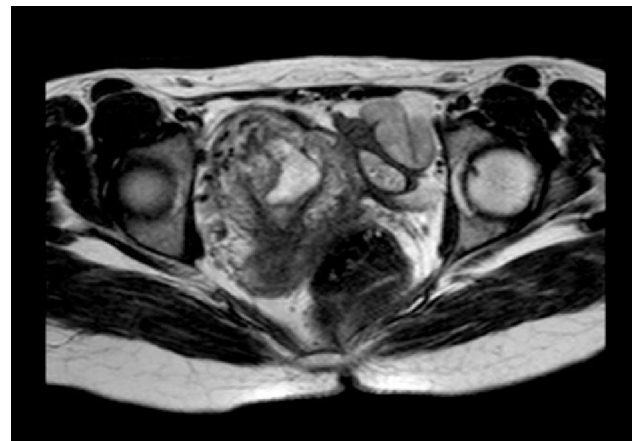


Figure 5: T2-weighted magnetic resonance image (MRI) of the pelvis shows diffuse enlargement of the uterus with abnormal T2 hyperintense thickening of the myometrium and multiple flow voids in the myometrium with disruption of the junctional anatomy.

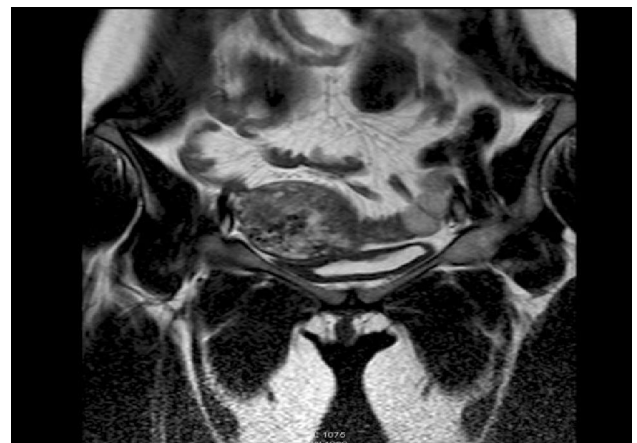


Figure 6: T2-weighted magnetic resonance image (MRI) of the pelvis shows diffuse enlargement of the uterus with multiple flow voids in the myometrium with disruption of the junctional anatomy.

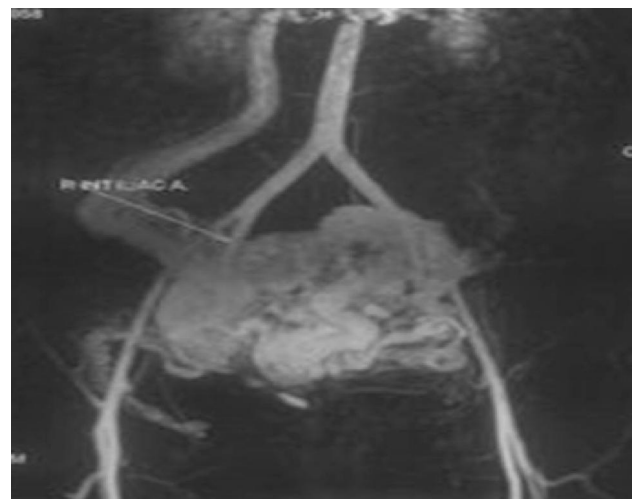


Figure 7: MR angiogram image showing the arteriovenous malformation with feeders from the internal iliac artery and early opacification of the right common iliac vein and the IVC.

Disucssion

AVMs of the uterus are a rare cause of uterine bleeding, with only over 100 cases reported since 1926.¹ They consist of a tangle of vessels with abnormal communication between arteries and veins and lack evidence of an intervening capillary network.

AVMs can either be congenital or acquired. Congenital uterine AVMs are thought to develop secondary to faulty arrest in the angiogenic process.^{1,2,3} Acquired uterine AVMs are thought to be related to uterine trauma including curettage or cesarean delivery, retained products of conception, choriocarcinoma, endometrial or cervical carcinoma, gestational trophoblastic disease, and diethylstilbestrol exposure.^{1,2,3} Regardless of the etiology, uterine AVMs are a potential source of significant morbidity and, rarely, mortality. The true incidence is unknown, but the majority of cases are found in women of reproductive age. Most commonly, they present with menorrhagia or menometrorrhagia requiring blood transfusions in 30% of reported cases.³

The differentiation between a uterine AVM and other causes of uterine bleeding is necessary because instrumentation can lead to massive haemorrhage. The precise diagnosis of AVM is of vital importance. Traditionally, diagnosis was made after hysterectomy and histopathologic examination. Currently, angiography is the gold standard for diagnosis.

Gray-scale US features of uterine AVMs include multiple anechoic structures with a serpentine contour within the myometrium.⁴ However, these features are often confusing and nonspecific.^{3,4}

Doppler ultrasonography is also a good noninvasive technique, and uterine AVMs appear as nonspecific heterogeneous or anechoic spaces in the myometrium on gray scale ultrasound and show increased vascularity on Doppler.⁵ On spectral flow Doppler, the systolic and diastolic velocities are 4 to 6 times higher than observed in normal myometrial vessels. Resistance index measurements are low, and mixing of arterial and venous waveforms is seen.⁷ Doppler examination should be done prior to D&C, which should be avoided in these women because it is likely to worsen the bleeding.


Digital subtraction angiography is an invasive method of confirming the diagnosis of AVM and treating it by embolization. MRI provides accurate definition of

uterine AVMs and effectively delineates invasion of adjacent organs. Characteristic features include a bulky uterus with a focal mass, disruption of the junctional zones, multiple serpiginous flow-related signal voids within the lesion, and prominent parametrial vessels.^{2,3} Gadolinium-enhanced MRI demonstrates hyper vascular arterial dominant flow. On angiography, the affected common and internal iliac arteries appear thicker and more circuitous than those on the normal side. AVMs appear as a complex tangle of vessels supplied by enlarged feeding arteries and show early venous drainage during the arterial phase.⁸ Computed tomography and magnetic resonance imaging may be used to determine the size, extent, vascularity, and involvement of adjacent organs.^{6,8}

Prior to embolotherapy, hysterectomy or uni/bilateral uterine artery ligation were the therapies of choice. Since the first reported case of trans-catheter uterine artery embolization for a uterine AV fistula in 1982, embolotherapy has become a well-recognized alternative to surgical intervention for uterine AVMs, with the major advantage of maintaining childbearing capacity.

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