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Case Report

Uterine Arteriovenous Malformation, Grey- Scale and Colour Doppler findings: A Case Report and Review

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Abstract: ***Introduction:*** Uterine arteriovenous malformations can either be congenital or acquired and are rare vascular disorders seen in women of child bearing age, with varied presentation. The clinical features range from being asymptomatic to life threatening haemorrhage. Imaging, particularly grey scale and color Doppler ultrasonography play a major role in the prompt diagnosis of this condition as the findings are useful when differentiating between the types as well as in planning the management of these patients. ***Objectives:*** To make a report of a rare case of arteriovenous malformation of the uterus in a young female patient with a review of relevant literature as well as to highlight the importance of Colour Doppler ultrasonography in the evaluation of uterine lesions. ***Case Presentation:*** The case is of a 36year old female patient with a 2 years history of amenorrhoea following multiple sessions of dilatation and curettage. She also had chemotherapy due to choriocarcinoma. Pregnancy test was negative. Pelvic ultrasound scan done showed multiple cystic tubular spaces that were hypervascular on colour Doppler interrogation. High velocities and low RI values were recorded. ***Conclusion:*** Uterine arteriovenous malformations (AVMs) are important rare vascular disorders of the uterine vessels that need to be recognized early on imaging for appropriate management to be instituted.

Keywords: Uterine arteriovenous malformations, Colour Doppler ultrasonography, case report.

INTRODUCTION

Uterine arteriovenous malformation (AVM) is a rare vascular condition and can either be congenital or acquired [1]. Uterine arteriovenous malformations are rare in non- pregnant women and were first described in 1926 by Dubriel and Loubat. Since then, several terms have been used to describe these lesions, including carvenous haemangioma, cirsoid aneurysm, racemose aneurysm, pulsatile angioma and arteriovenous fistula [2,3].

Acquired uterine arteriovenous malformations are abnormal communications between the intramural branches of the uterine artery and the myometrial venous plexus deep inside the myometrium and endometrium. They may obtain blood supply by either one or both uterine arteries [1]. The most common vascular disease affecting the uterine arteries are arteriovenous malformations (arteriovenous fistulas), true aneurysms and pseudoaneurysms [2].

The congenital arteriovenous malformation is very rare and it usually results from abnormal development of the embryonic primitive vascular structures which then determine multiple abnormal communications between arteries and veins [4].

The acquired type is usually more encountered in clinical practice and is associated with conditions such as gestational trophoblastic disease (GTD), multiple gestations, miscarriages, previous surgeries and instrumentation (dilatation and curettage), caesarean section, pelvic trauma, cervical or endometrial carcinoma as well as infections and exposure to diethylstilbesterol [4-6].

Clinical presentation of this entity varies with the major presenting feature being uterine bleeding

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which maybe life –threatening in young women [6]. Bleeding usually occurs when the endothelium of the abnormal vessels is disrupted as during instrumentation or due to endometrial desquamation during menstruation. Chronic blood loss associated with AVM may also cause abdominal pain, dyspareunia and anaemia [6,7]. The association of the clinical history with imaging findings is quite useful when differentiating between the congenital or acquired types of uterine AVM.

Several imaging modalities can be employed in the imaging of these lesions. Ultrasonography (both gray scale and colour Doppler), contrast enhanced computed tomography, angiography and in recent times the use of Magnetic resonance imaging (MRI) have been very valuable [7,8]. Colour Doppler ultrasound is the modality of choice in the diagnosis of uterine AVM [7]. The grey scale features may be non- specific with a wide range of manifestations including areas of subtle or diffuse myometrial inhomogeneity, heterogenous ill - defined mass with multiple hypoechoeic cystic or tubular structures of varying sizes [6-8]. Doppler USS typically shows the lesions to be hypervascular with low resistance waveforms and high velocity flow pattern [9].

As already mentioned, these lesions can also be evaluated with contrast enhanced computed tomography and Magnetic Resonance Imaging as well as by angiography which is the preferred method in patients who may undergo embolization [3-5].

CASE PRESENTATION

We report a case of a 36 years old P_1^{+2}(1 alive) lady with a history of treated gestational trophoblastic disease (choriocarcinoma). She had dilatation and curettage as well as complete course of chemotherapy 2 years prior. She presented on an outpatient basis to the gynaecology unit with 2 years history of amenorrhoea following the last dilatation and curettage she did before the commencement of the chemotherapy. Since completing her drugs, she has had normal beta-hcg levels and serial pregnancy tests done have been negative. There were no complaints of cyclical monthly abdominal pains or discomfort to suggest hydrometrocolpus, no history of abnormal vaginal discharge or irregular bleeding. She was also not desirous of any more children. Her last confinement was 5 years prior. Physical examination and review of the body systems were essentially normal. She was also haemodynamically stable.

Patient presented to the ultrasound suite for a pelvic ultrasound evaluation. On examination, there was an enlarged and bulky uterus measuring about 12cmx9.0cmx7.6cm (CCXTRXAP) with heterogenous myometrial echotexture and the presence of multiple anaechoiec tubular structures, more in the fundal region but also involving the lower uterus and the cervix to a lesser extent. The parametrial areas also had these anaechoiec spaces within it. On colour doppler interrogation, the lesions were seen to be vascular structures, demonstrating intense myometrial hypervascularisation and turbulent flow and showing a mosaic pattern. Spectral analysis of the arterial components within the lesion showed high velocity flow with a low resistivity index (RI). Similar pattern was seen in the venous flow on spectral evaluation. An impression of uterine AVM was made with the risk factors being previous instrumentation and history of GTD. Patient was then referred back to the gynaecology team to discuss the possible management options, since she was asymptomatic. She was placed on conservative management and is presently stable.

Images obtained are as shown in figs 1-4 below.

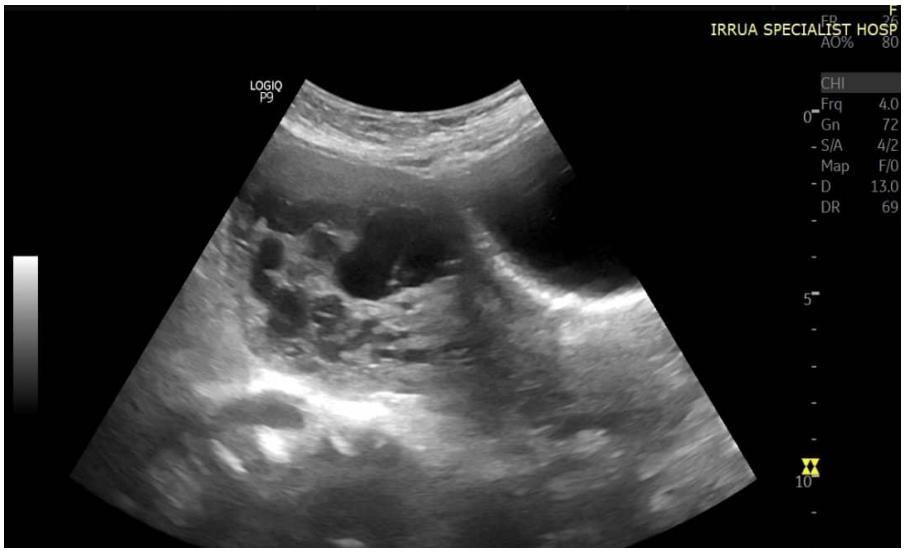

Fig 1: Grey-scale longitundinal view of the uterus containing multiple cystic spaces within it

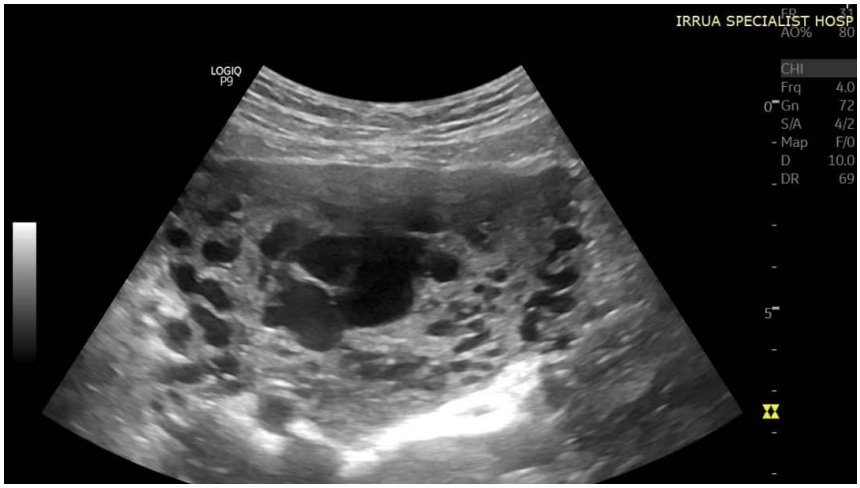

Fig 2: Grey-scale transverse view of the uterus containing multiple cystic spaces within it. The parametrial areas are shown to also contain these cystic areas

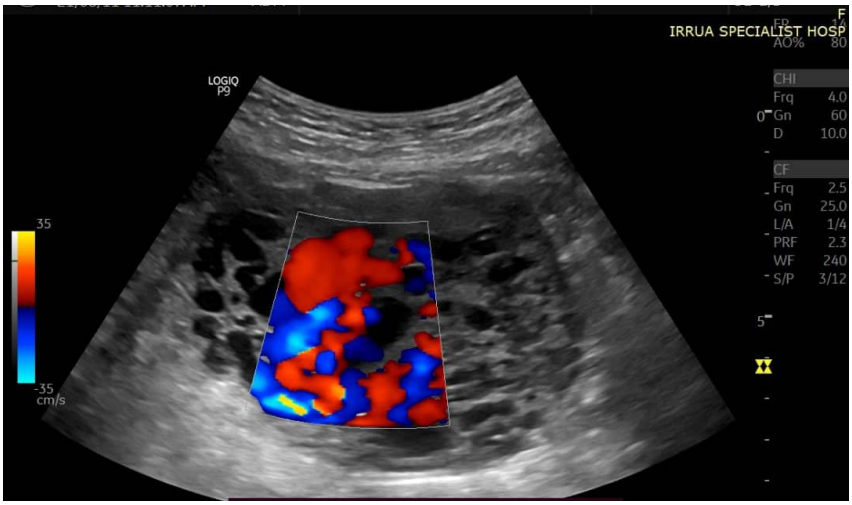

Fig 3: On colour doppler interrogation, the lesions are seen as vascular structures, demonstrating intense myometrial hypervascularisation and turbulent flow with some areas showing a mosaic pattern

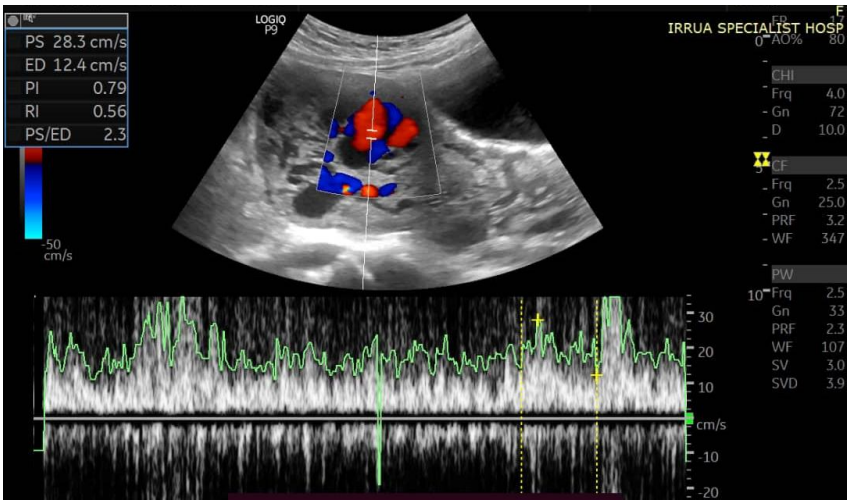

Fig 4: Spectral analysis of the arterial components within the lesion showed high velocity flow with a low resistivity index (RI)

DISCUSSION

Arteriovenous malformations are part of the vascular abnormalities affecting the uterine vasculature. Uterine AVMs are rare with just about only a hundred cases described in literature [1]. It is a condition where there is dilatation of the inter-villous spaces deep inside

the myometrium allowing a direct flow from the arterial system towards the venous system without the participation of the capillary vessels [1-3].

Uterine AVMs could be congenital or acquired. The index case is an acquired type as she has up to 2 of the risk factors associated with this type. The causes of acquired uterine AVMs include gestational trophoblastic disease (GTD), previous uterine instrumentation and surgeries, infections and mitotic cervical and endometrial conditions [4-6]. Also, AVMs persist in 10-15% of cases of GTD in remissions after chemotherapy [3]. Massive vaginal bleeding is said to be the main clinical presentation in these patients. These have been the case in most of the reports recorded in the literature. The index patient however had no history of bleeding and had amenorrhoea for 2 years.

Doppler ultrasonography is the imaging modality of choice in diagnosing the condition [3-5]. The findings being diffuse uterine enlargement, with multiple cystic tubular spaces as well as focal or asymmetric endometria thickening. These lesions also demonstrate arteriovenous shunts with low resistance and high velocity flow [4,6,8-9]. These findings were recorded in the index patient. Furthermore the spectral analysis can predict the degree of the vascular lesion arterialization and assist in treatment planning [9].

Conditions that can present with similar sonographic findings include Gestational trophoblastic diseases, hypervascular lesions like retained conception products and abnormal placentation [1,8].

Management of uterine AVMs can either be conservative or through endovascular and surgical methods [1,7,8]. Conservative medical treatment of uterine AVMs using Gonadotropin releasing hormone (GnRH) agonists is mainly reserved for stable patients [8]. Transarterial endovascular treatment is most commonly used to treat the congenital type particularly when they are multiple [7,8,10]. The surgical option involves doing a hysterectomy for these patients following a failed embolization or if the patient is no more desirous of children [10]. Index patient is presently on conservative management.

CONCLUSION

Uterine AVMs are uncommon lesions with the patients either being completely asymptomatic or presenting with life threatening vaginal bleeding. The diagnosis should be considered in patients of childbearing age with history of instrumentation or other risk factors like GTD or malignant cervical and endometrial conditions. Colour Doppler USS is an excellent non-invasive and widely available diagnostic tool that is used in the diagnosis of this condition, which can be life threatening in some cases.

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