Uterine arteriovenous malformation – Possible association to uterine fibroids?

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SUMMARY
A 32-year-old, gravida 2 para 0+1, was managed in Selayang Hospital, Selangor for uterine fibroids in pregnancy and placenta previa major. The lady went into preterm labour at 33 weeks, requiring emergency Caesarean section. Intraoperatively, we found a thinned-out bulge between the intramural uterine fibroids at the posterior uterine wall, which then perforated and was repaired. Persistent bleeding post operatively led to relaparotomy and hysterectomy. Histology of the uterus reported arteriovenous malformation (AVM). We postulate the possibility of these lesions co-existing with uterine fibroids. Screening for uterine AVMs in patients with fibroids may lead to early detection with option of embolization; deferring the need for hysterectomy.

INTRODUCTION
Uterine arteriovenous malformations (AVMs), although considered rare, have an important place in gynaecological practice due to risk of massive bleeding that is potentially life-threatening. There are numerous risk factors associated with this condition; encompassing nearly the entirety of uterine pathophysiology, as well as the normal physiologic changes of pregnancy. As such, a high index of suspicion is required when faced with persistent or heavy uterine bleeding, after common causes have been ruled out. This case report highlights our encounter with a patient with multiple uterine fibroids, whose bleeding during Caesarean section was only later confirmed to be due to a uterine AVM. We hope to raise awareness of the possibility of these lesion co-existing with uterine fibroids when seen in women whose uterine bleeding remains uncontrolled despite routine measures.

CASE REPORT
We describe here a case of a 32 year old, gravida 2 para 0+1 who was followed up in Selayang Hospital, Selangor throughout her antenatal period for multiple uterine fibroids and placenta previa major. She had a complete miscarriage in the past, managed conservatively and not requiring curettage. She denied having any uterine surgeries done before.

She went into preterm labour at 33 weeks of gestation and required emergency Caesarean section. Intraoperatively she was reported to have a highly vascularised lower segment with a distorted uterine cavity. There were multiple intramural fibroids at the posterior wall of the uterus; largest of which measured 7x6cm, located at the left cornua. Due to persistent bleeding, the uterus was exteriorised for optimal view. Interestingly, she was found to have a thinned-out bulge measuring 5x6cm, located right in between the fibroids previously mentioned at the posterior wall of the uterus, away from the placental implantation site. Gentle palpation over this lesion caused it to perforate; a defect of 1x2cm in measurement. Multiple haemostatic sutures had to be applied to secure the bleeding. Post operatively, she was transferred to the ICU for close monitoring.

She unfortunately continued to bleed (evidenced by persistent bleeding from the abdominal drain) and required relaparotomy the same night. Intraoperatively, she was found to be oozing from the friable area at the thinned-out bulge of the posterior uterine wall stated earlier. Hysterectomy was done; however, she had by then developed disseminated intravascular coagulopathy (DIC). Abdominal packs were placed into the pelvic cavity; these were only removed 3 days later after her coagulation profile had improved. Her condition improved gradually and she was discharged home well on Day 15 post surgery. She remained well during her follow-up at 6 weeks post delivery.

Histology of the uterus reported the posterior wall to have groups of dilated and ectatic vessels with variable muscular wall thickness. There were an admixture of malformed vessels displaying muscular disruption with incomplete muscular wall and intimal protrusion; causing abrupt changes in thickness of medial and elastic layers of vessels and abnormal vascular dilatation. Elsewhere, some vessels were dilated and thin-walled where the uterine stroma form part of the vessel wall. These findings were in keeping with arteriovenous malformation. These were found to be in close proximity with two well-circumscribed intramural lesions composed of interlacing bundles of smooth muscle cells in sweeping fascicles. These smooth muscle cells are of bland-looking spindle cells having cigar shape nucleus and abundant bright eosinophilic cytoplasm with marked stromal hyalinization – suggestive of leiomyomata.

DISCUSSION
AVMs are rare vascular anomalies that are potentially life-threatening. There are difficulties in obtaining accurate incidence rate due to the rarity in reported cases; to date, there are fewer than 150 cases reported in the literature. A prospective study of 959 patients after abortions or delivery found sonographically evident uterine AVMs in 5.2% of women after dilatation and curettage, and in 0.22% of...
women after delivery; however only 0.1% deemed clinically significant. It is estimated from that study that the incidence of true uterine AVMs is <0.1% of women who have had abortion or delivered. Increased use of imaging modalities is likely to lead to identification of these lesions more frequently.

Arteriovenous malformations result from abnormal direct communication between an artery and a vein without an intervening capillary bed, causing diversion of blood flow into the venous system following the pressure gradient, and resulting in increased intraluminal pressure, high-velocity flow, and marked vascular enlargement of the venous system. These lesions rarely occur in the uterus.

Mainly acquired, uterine AVMs are typically associated with damage to uterine tissue; development of abnormal vascular connections between arteries and veins may occur during the healing process. They usually follow a history of previous uterine trauma – e.g. curettage procedures, caesarean section, or pelvic surgery. Acquired arteriovenous communications are analogous to fistula formation, but more complex and numerous, likely due to the rich vascularity and dense network of anastomoses within the uterus. With increased surgical gynaecological procedures in current practice, the prevalence of uterine AVMs will likely increase.

Outside the context of pregnancy, acquired AVMs are associated with uterine infections, inflammation, endometriosis, gynaecological malignancies, and exposure to diethylstilbestrol (DES). The pathophysiology behind non-surgical-related cases of uterine AVMs (such as in the case described above) remain unclear.

Uterine fibroids have occasionally been described in cases of uterine AVMs. AVMs are histologically reflected by abrupt changes in the thickness of the medial and elastin layer of the vessels. During the course of fibroid development, the abnormal increase in muscle growth is accompanied by dysregulation of its normal vascular pattern. From available studies, there appear to be alterations in the distribution of vessels in the myometrium around the fibroid as well. The gradient of vessel size seen in non-fibroid myometrium, is lost in the myometrium around the fibroid. It is proposed that the loss of the vessel size gradient could lead to vascular pressure changes and predispose to the formation of uterine AVMs.

Given the rarity of the condition and its nonspecific ultrasound findings, the diagnosis of uterine AVM is often challenging. Traditionally, and as with our case, AVM was diagnosed by laparotomy and histopathologic examination of the uterus after a hysterectomy. With advancing technology, colour and spectral Doppler ultrasound is now widely used to detect this condition; revealing multiple enlarged uterine vessels, intense signalling, and apparent multidirectional flow indicative of turbulent high-velocity flow within low-resistance vessels. CT, MRI, CTA, and MRA can be used to better delineate the AVM. Digital subtraction angiography (DSA) however remains the gold standard for diagnosing AVMs, though rarely performed due to its invasive nature.

Endovascular management with transcatheter embolization of uterine artery is the mainstay of therapy for uterine AVMs as it is effective, can be done without general anaesthesia, and maintains fertility. Cases refractory to endovascular interventions can be treated definitively with hysterectomy.

Many uterine AVMs spontaneously resolve without requiring treatment. Growing evidence suggests that conservative management with flow monitoring and imaging may be appropriate for patients who are asymptomatic with lower-flow lesions. There have also been case reports describing the use of gonadotropin-releasing hormone (GnRH) agonists, oral contraceptives, as well as methylergonovine maleate to treat uterine AVMs in stable patients.

Described in this case report was a woman in her early 30s, now with only one child. While screening for uterine AVMs (i.e. by means of ultrasound Doppler scan) in all patients with uterine fibroid in pregnancy may not be practical, it would have led to the early detection of this lesion with the option of embolization; hence deferring the need for a hysterectomy. Although rare, there are case reports available on successful uterine artery embolization during an ongoing pregnancy, not resulting in any acute complication to the mother or the developing foetus.

CONCLUSION

Uterine AVMs may present with torrential uterine bleeding, potentially leading to life-threatening haemorrhage. As such, they should be considered as part of the differential diagnoses when faced with unexplained heavy uterine bleeding, so as to avoid any unnecessary intervention which may significantly worsen the bleeding or delay treatment. As far as screening goes, awareness of this condition, recognition of its imaging features, as well as a high index of suspicion is essential in maximizing the chances of a favourable outcome, especially in the presence of risk factors.

REFERENCES