Pulmonary arteriovenous malformation unmasked in pregnancy: A case report

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Abstract

Pulmonary arteriovenous malformations are anomalous communications between arteries and veins of the pulmonary vasculature. Its incidence is rare. Pulmonary arteriovenous malformations can be asymptomatic or cause profound cardiovascular compromise and adverse neurological sequelae, as a result of right to left shunting of deoxygenated blood. Pregnancy and its physiological demands can unmask and exacerbate pulmonary arteriovenous malformations with attendant risks of life threatening complications and rarely, death. This case report describes a first presentation of pulmonary arteriovenous malformation in pregnancy and the tendency for misdiagnosis with pulmonary embolism. A multidisciplinary approach to management is pertinent considering the challenges involved in deciding the appropriate therapeutic management in pregnancy which has to be weighed against potential maternal and fetal risks.

Keywords

Maternal-fetal medicine, high-risk Pregnancy, Hypoxia Pulmonary arteriovenous malformation, transcatheter embolisation

Case presentation

A 28-year-old primigravida 18 weeks pregnant and previously fit and well, presented with a recent episode of fresh per vaginal bleeding. Physical examination showed no active bleeding and no evident cause for the event. From triage observations, it was noted that the patient's oxygen saturations were 92% on room air and when rechecked, lower still at 89%. The patient gave no history of respiratory disease or symptoms and was a non-smoker. A referral to the medical physicians was made and the patient was found to have a PaO₂ of 7.9 kPa on Arterial Blood Gas analysis (ABG). Examination revealed no stigmata of chronic disease, a normal chest X-ray (CXR) and ECG and only a pansystolic murmur thought to be secondary to pregnancy. Other blood investigations, including a liver function were unremarkable. The clinical suspicion was initially of a pulmonary embolism in pregnancy and the patient was commenced on treatment dose low-molecular-weight heparin (LMWH) prior to excluding the diagnosis by objective testing.

A half dose perfusion scan was requested with a finding of distinct perfusion deficit in the left lower lobe with activity outside of the lungs, in the kidney and thyroid suggestive of a right to left shunt. For elucidation, a Computed Tomography Pulmonary Angiography (CTPA) was performed and the presence of a left lower lobe pulmonary arteriovenous malformation (PAVM) was visualised. On further questioning, the patient reported suffering a brain abscess secondary to chicken pox aged three. Archived medical notes were reviewed and it was discovered that six years earlier, when pre-assessed for a dental procedure under general anaesthetic, the patient had low saturations of 93%. CXR at the time had been deemed normal, but on closer scrutiny a subtle opacity of the left lower lobe was apparent; recent images misjudged as normal were compared and the same opacity was found to be present. The LMWH was stopped and an urgent referral made to a tertiary cardiothoracics unit.

At 22 weeks, with conscious sedation, embolisation was performed by the insertion of femoral catheters to introduce four coils to occlude blood flow to the left lower lobe AVM (Figures 1 and 2). After the procedure, saturations immediately rose from the mid-80s to the mid-90s and the patient was discharged back to the care of obstetrics at her referring hospital. A multidisciplinary plan was made between cardiothoracics, obstetrics and anaesthetics regarding continuing antenatal care and intrapartum management. Saturations were monitored weekly and normal till delivery. The patient ultimately went into spontaneous labour at term and achieved a normal vaginal delivery with saturations maintained above 95% throughout. There was no

deterioration in the puerperium. A referral to a geneticist postnatal excluded an association of PAVM with hereditary haemorrhagic telangiectasia (HHT). Before a follow-up CT scan of the chest could be arranged to exclude persistence of PAVM or recanalisation, the patient conceived again and is currently in her third trimester with an uneventful pregnancy thus far.

Discussion

PAVM in pregnancy can cause serious and life-threatening complications with events such as haemothorax and heart failure being the most common complications reported. Recent reviews of case reports provide mortality rates of 0–15% for untreated PAVM.^{1,2} It can occur primarily or in association with HHT, an autosomal dominant genetic disorder characterised by the presence of multiple visceral arteriovenous malformations. The majority of morbidity and mortality from PAVM occurs during second and third trimesters of pregnancy, furthermore in the intrapartum period. Previously stable patients can dramatically deteriorate in this relatively short space of time.³

The diagnosis of PAVM is not uncommon, as a first presentation in pregnancy. This is mostly due to increased cardiovascular demand worsening right to left shunting, with increasing dyspnoea or low oxygen saturations. Deterioration is multifactorial and also affected by hormonal changes in pregnancy. Increases in blood volume and cardiac output also occur, by 40% and roughly 45%, respectively,⁴ and the resultant increase in pulmonary blood flow can lead to dilatation and eventual rupture of thin-walled PAVM. Exacerbating this is the fact that venous distensibility increases to 150% of normal just before the time of delivery.

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Figure 1. Pre embolisation.

Figure 2. Post embolisation.

As in this case, the presentation of PAVM in pregnancy is commonly mistaken for pulmonary embolism, a far more common cause of respiratory distress and reduced breath sounds.⁵ Furthermore, careful examination and analysis of past medical history will usually reveal the true diagnosis. Our patient later gave a history of brain abscess and previous hypoxia which if obtained at initial presentation would have at least alerted clinicians to the likelihood of an underlying chronic medical condition. This will have precluded the preliminary diagnosis of pulmonary embolism and the need for therapeutic LMWH.

In PAVM, CXR appearances are largely non-specific and as such if there is strong clinical suspicion of malformation, more definitive imaging is required to make the diagnosis. There is a place for CT and Magnetic Resonance Imaging (MRI), useful in visualising the location and size of such lesions, but pulmonary angiography remains the gold standard and in stable women.

Treatment is by transcatheter embolisation (TCE). Surgical resection is still recommended in the setting of haemodynamic instability or the severely compromised. Though previously a third option, conservative management is no longer advised in symptomatic pregnant women as it poses a significant risk of development of complications. TCE in the non-pregnant patient is well described in the literature, although its use in pregnancy is still surrounded by controversy regarding exposure of the fetus to radiation. Shielding the abdomen and pelvis with lead is of little benefit as, unlike with X-ray, there is internal scatter of the radiation exposed to the thorax.¹ This risk is reduced by keeping fluoroscopy time and the number of diagnostic exposures pre procedure to a minimum. In this case as presentation was in the second trimester in a stable but symptomatic patient, the balance of risks favoured embolisation with overall fetal radiation exposure well within the 500 mrad threshold associated with any increased risk of birth defects or fetal loss. In a particular series, the fetal radiation dose ranged from 50 to 220 mrad, less than half of the maximum recommended in pregnancy.^{1,6}

A multidisciplinary approach is needed in the care of women with PAVM in pregnancy. The considerations include the risks of further deterioration with advancing gestation with potential lethal consequences in the event of PAVM rupture. Furthermore, the choice of therapeutic intervention is crucial as the option of surgical resection under general anaesthesia is not without attendant maternal morbidity risks in a haemodynamically stable patient.

In the event of worsening or recurrence of PAVM after treatment, management needs to be focussed on maintaining cardiac and respiratory function, particularly as labour is a time of exertion and possible worsening of hypoxia. The haemodynamic effect on the vasculature is also more pronounced during this period. Although our patient remained asymptomatic and stable following treatment, an intrapartum anaesthetist plan was sought to assess the safety of general anaesthetic versus regional anaesthesia should this become necessary. The latter because there may be coexisting epidural AVM in women with PAVM and so to determine this, CT/MRI of the lumbar spine is recommended in the early third trimester. An epidural insertion, if performed needs to be titrated to no further than T10 dermatome, so as to avoid significant reduction in systemic vascular resistance and worsening of right to left shunt.⁷ This, and any other plans, needs explicit documentation in the notes so that all attendants at the birth, midwives and doctors alike, are aware of what is safe and more importantly what is contraindicated.

PAVM is not a contraindication to vaginal delivery; however, the consideration in the event of worsening or recurrence is to minimise haemodynamic stresses and systemic hypertension.⁷ The advice is to shorten the second stage where possible. Saturations need close monitoring (to assess clinical stability) during the intrapartum period. Cardiothoracic input does not stop at embolisation, as contingency planning is needed if there is worsening or recurrence of PAVM after treatment, hence the need for postpartum review. Long-term follow-up has identified subsequent complications such as persistence of PAVM, recanalisation, growth of accessory vessels and ischaemic strokes.^{8,9} There is a reported small risk of recanalisation and recurrence and PAVM growth in a subsequent.⁶

Our patient conceived soon after delivery with no evidence of recurrence of PAVM. She had an unremarkable pregnancy and delivery before submission of this report. There are not many cases in the literature of a second pregnancy after embolotherapy in a previous pregnancy.

Conclusion

This case emphasises the consideration of PAVMs as a differential diagnosis of hypoxia in pregnancy and the importance of treatment in pregnancy to reduce risk of adverse complications. It highlights the need for a multidisciplinary approach in informing the choice of definitive treatment, guided by maternal and fetal safety considerations.

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