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CASE REPORT

Pulmonary arteriovenous malformations in pregnancy – a case report and review of literature

William Hayes, MD[1], Emily O'Brien, MD[1], Benjamin Hammelman, MD[2], James M. O'Brien, MD[3]

[1]Penn State College of Medicine, Department of Obstetrics & Gynecology
[2]Penn State College of Medicine, Division Interventional Radiology
[3]Penn State College of Medicine, Division of Maternal Fetal Medicine

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ABSTRACT

Pulmonary arteriovenous malformations are potentially life-threatening abnormalities that can be exacerbated by pregnancy physiology. Despite prenatal optimization, patients with hereditary hemorrhagic telangiectasia and associated vascular abnormalities continue to experience significant maternal morbidity. Currently, there is a paucity of clinical guidance for managing obstetric patients affected by pulmonary arteriovenous malformations. This case exemplifies the potential to minimize maternal-fetal morbidity and mortality by managing pregnancies affected by pulmonary arteriovenous malformations with a proactive and multidisciplinary approach. Developing an emergency action plan eases decision-making in critical circumstances, allowing for timely intervention. Diagnostic testing for hereditary hemorrhagic telangiectasia and/or pulmonary arteriovenous malformations should be pursued in all at-risk individuals to afford optimization of outcomes. Although this case describes an overall positive outcome, additional research is still needed to help improve maternal and fetal outcomes.

Introduction

Arteriovenous malformations (AVM) are vascular anomalies due to aberrant communications between arterial and venous systems, most commonly affecting the lungs, brain, and liver[1]. When looking specifically at pulmonary arteriovenous malformations (PAVM), approximately 3 in every 100,000 people of the general population are affected. Upwards of 80% of these cases are associated with an inherited disorder like hereditary hemorrhagic telangiectasia (HHT)[2-3]. HHT is an autosomal dominant condition that has a much higher prevalence, affecting up to 1 in every 5,000 people[4]. Approximately 50% of patients are asymptomatic from this disease process, particularly if PAVMs are less than 2cm in diameter, but these are still identified by the third decade of life for most people[2,5].

Despite the rarity and varied clinical presentation of PAVM, patients of reproductive age require guidance regarding the effect pregnancy may have on their disease process. Significant elevations in estrogen, vascular growth factors, cardiac output and progesterone induced vasodilation may result in PAVM growth and subsequent pulmonary hemorrhage, cardiopulmonary collapse, and maternal morbidity and mortality. Existing literature describes an increased risk of poor maternal and potentially fetal outcomes in pregnancies affected by PAVM hemorrhage, but there is still a paucity of clinical guidance regarding the optimal management of obstetric patients affected by PAVMs[6].

We aim to discuss a case in which maternal mortality was avoided in a patient with PAVM. Through multidisciplinary collaboration and patient education, the morbidity associated with this disorder for be minimized, and maternal, fetal, and neonatal outcomes optimized. Founded on the physiologic changes of pregnancy, we justify a framework for multidisciplinary obstetric care that may improve preconception, antenatal, and postpartum management of patients affected by HHT and similar disease processes driven by arteriovenous malformations.

Case

The patient is a 31-year-old gravida 4, para 2 who presented for consultation at 30 weeks gestation. Her medical history was significant for previous embolization of more than 20 PAVM in the setting of autosomal dominant HHT, which was diagnosed via genetic testing at age 15. Her first two pregnancies were complicated by ruptured PAVM in the postpartum period that required vascular coil placement. In her second pregnancy, she also experienced ruptured PAVM during her antenatal course around 29 weeks gestation, and subsequently had a successful vaginal birth after cesarean at term.

She presented to the high-risk obstetrics clinic in the third trimester of pregnancy for coordination of care and planned delivery at a tertiary medical center with an interventional radiology team. At the time of consultation and transfer of obstetrical care, she was without recent imaging and had yet to establish care with a multidisciplinary team. We recommended urgent chest and brain imaging, maternal echocardiogram, and an assisted second stage of labor if imaging identified untreated PAVM that could not be managed prior to delivery. The patient then met with our obstetric anesthesia team and neonatal intensive care unit. Once known to all members of the multidisciplinary team that would be part of her delivery, an emergency action plan was developed.

This plan emphasized that if the patient presented with concerning symptoms, immediate chest imaging should be obtained and the interventional radiology, surgical intensive care unit, neonatal intensive care unit, and obstetric anesthesia teams should be promptly notified. If cesarean delivery was required, this should occur in an operating room with proximity to the surgical intensive care unit (ICU).

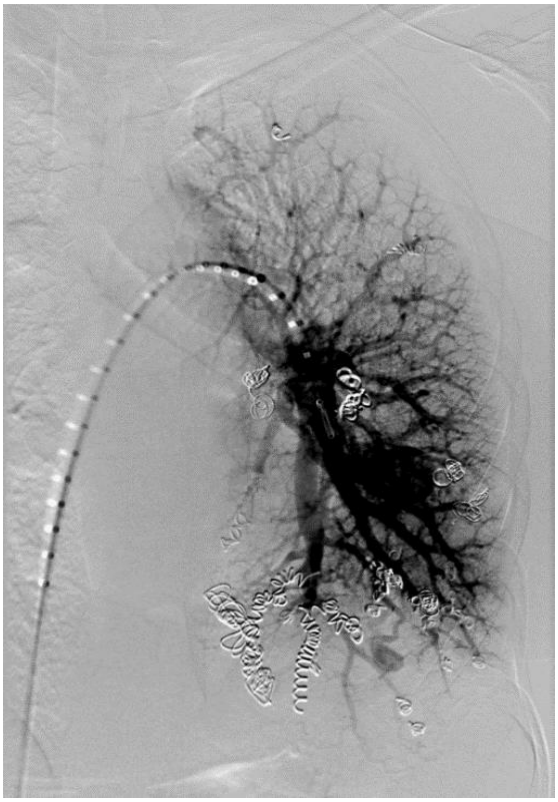

Figure 1: Interventional radiology coil placement into left sided AVMs.

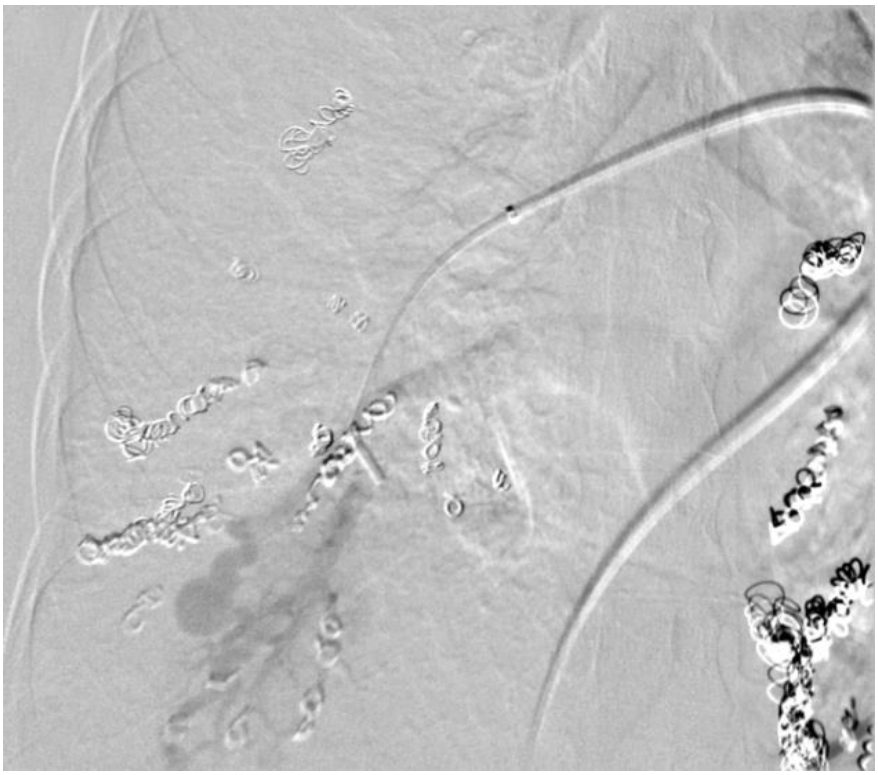

Figure 2: Interventional radiology coil placement into right sided AVMs.

The recommended imaging studies were significant for numerous bilateral PAVM and she underwent placement of 10 total vascular coils antenatally. Unfortunately, the largest PAVM measured 32mm and was located within the left lingula, where a feeding vessel could not be identified. The next largest PAVM measured 17 mm (located in the right lower lobe) and 23 mm (located in the right upper lobe), both of which were successfully treated.

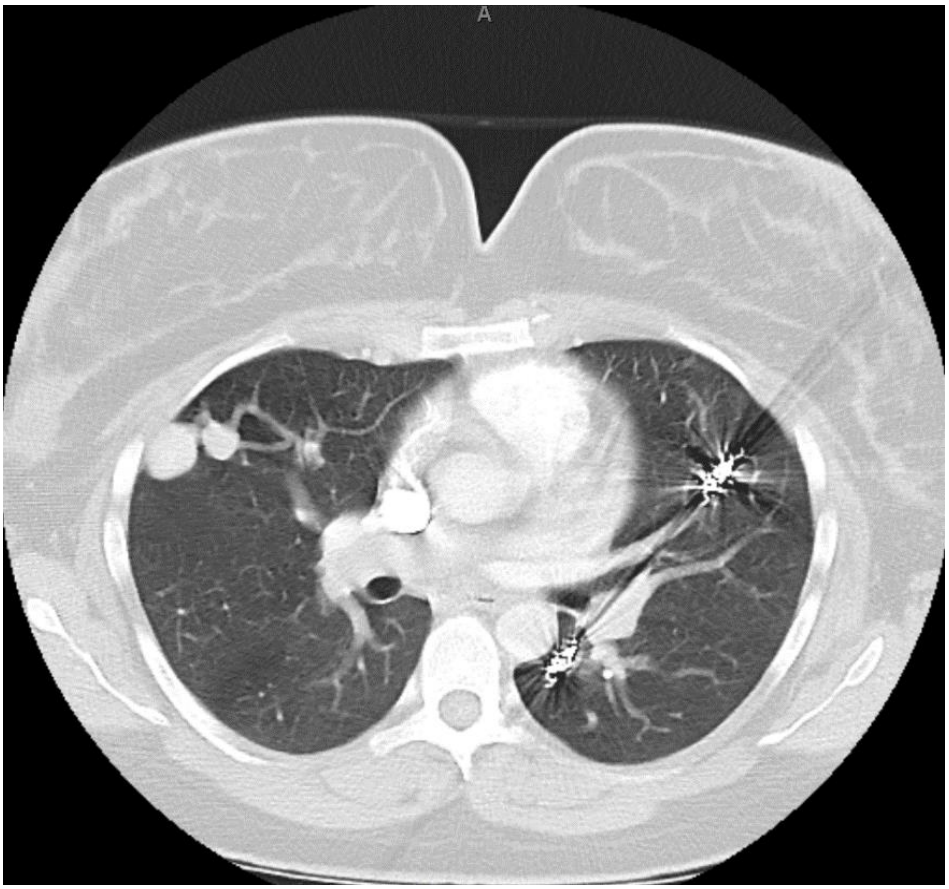

Figure 3: Computed tomography scan of chest demonstrating coil placement during third trimester of pregnancy.

At 38 weeks 4 days estimated gestational age, the patient presented to the hospital with complete cervical dilation and fetal malpresentation. A repeat C-section was performed followed by direct admission to the surgical ICU for recovery. She was downgraded to the postpartum floor later that evening after an uneventful and brief surgical ICU course.

On postpartum day 1, the patient experienced an episode of acute onset hemoptysis, oxygen desaturation, tachycardia, and tachypnea. Prompt laboratory and imaging studies were obtained and pulmonology, anesthesia, and high-risk obstetrics were consulted. The patient immediately returned to the ICU where she was intubated due to persistent hypoxia. Repeat chest imaging was consistent with pulmonary hemorrhage. This was managed with urgent PAVM embolization, at which time a total of 4 vascular coils were placed in a right lower lobe PAVM that measured 17 mm. Notably, this was not one of the larger PAVMs targeted during antenatal embolization. The patient was subsequently extubated on postpartum day 3 and returned to the postpartum floor on postpartum day 4. The remainder of her hospital course was unremarkable and she was discharged home on postpartum day 5 in stable condition with appropriate follow up.

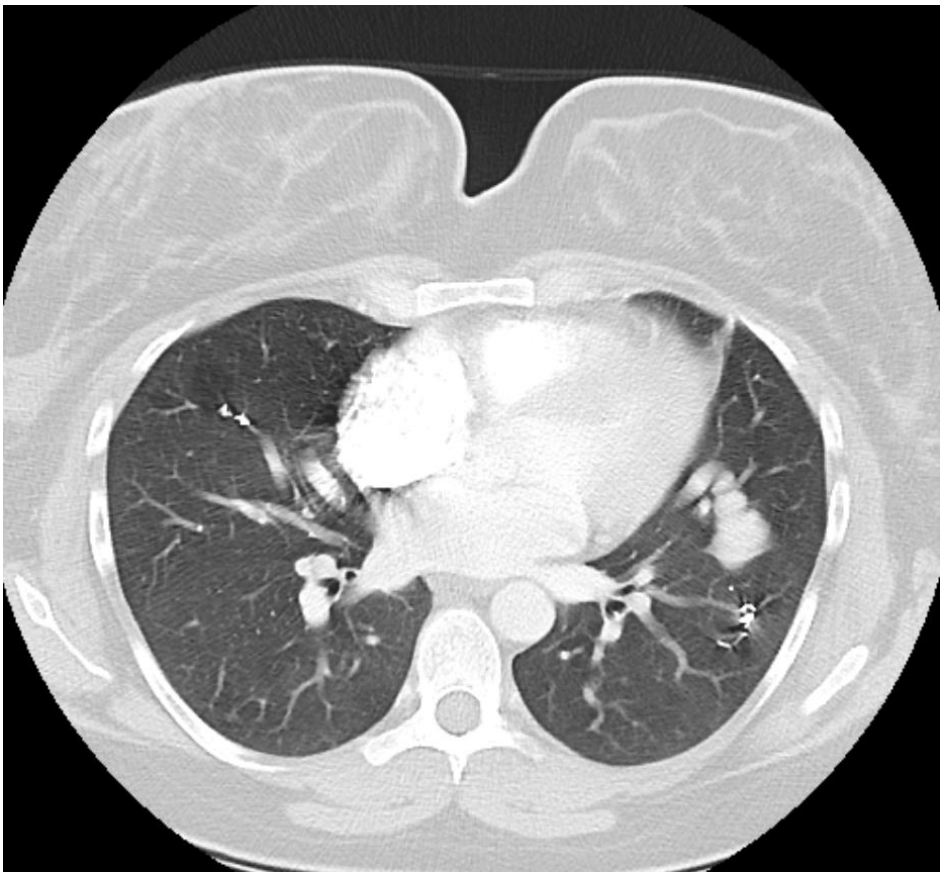

Figure 4: Computed tomography scan following coil placement following acute PAVM rupture in the postpartum period.

Discussion

We have presented a unique case of a gravida with known HHT and PAVM who was able to successfully carry a pregnancy to term with the assistance of multidisciplinary care and intervention. Despite the patient experiencing a pulmonary hemorrhage, the neonatal outcome was positive and preparation prevented additional maternal morbidity and mortality.

For both pregnant and non-pregnant patients, the natural history of PAVM includes an increase in size over time. Treatment is generally recommended in all patients with a PAVM greater than 3 mm in diameter and can occur via surgical resection or embolization of feeding vessels with coils[3,7-8]. Our team was afforded the knowledge that all of this patient's PAVMs could not be treated antenatally, and were able to further counsel and educate the patient that unrecognized and/or untreated PAVM unfortunately have an increased rate of mortality[9]. This highlights the importance of both preconception and antenatal screening in order to best improve outcomes.

Regardless of this patient's family history and personal diagnosis of HHT at age 15, she opted to not complete genetic testing for her children. She had been counseled by the obstetric and neonatal teams regarding the autosomal dominant inheritance pattern of HHT, and knew that each of her children had a 50% chance of being affected. She was counseled that genetic testing would not only allow for optimization of her children's lifelong health, but also could play a role in their future obstetrical management. For example, knowing a neonate will be affected by vasculopathy might be an absolute contraindication to a vacuum-assisted delivery. Interestingly, this patient's first pregnancy was a cesarean delivery after a failed vacuum at an outside institution.

As seen in our case, preventative embolization improves outcomes but does not completely eliminate the risk of PAVM rupture and sequela.

Based on available data, the risk of PAVM rupture and subsequent hemothorax is approximately 1-2% in pregnancy and the immediate postpartum period[9-10]. This is significantly higher than the annual 0.16% PAVM rupture risk in the non-pregnant state, and approaches the 2.7% lifetime risk of PAVM rupture[11]. This drastic elevation in risk can be attributed to the numerous physiologic cardiovascular and respiratory changes of pregnancy and labor.

Some of the most notable physiologic changes to consider are the following: blood volume increase of up to 45%, increase in arterial and venous compliance, increase in cardiac output by approximately 30-50% (first stage of labor is associated with a 12% to 31% increase in cardiac output, while the second stage of labor is associated with an even greater increase in cardiac output of up to 49%), transient increase in intrapartum blood pressure, and autotransfusion of approximately 500mL of uteroplacental blood and rapid mobilization of extravascular fluid in the immediate postpartum period[12-15]. Finally, the vascular resistance within PAVM is lower than in the normal artery-capillary-vein circulatory system, consequentially resulting in blood preferentially flowing through the low resistance PAVM[16]. This can be potentiated by the increased circulating blood volume that is a normal physiologic response in pregnancy. Understanding this physiology is important in anticipating possible obstetric outcomes and informed our decision for initial recovery in the surgical ICU, as well as the plans for antepartum imaging and preventative treatment as indicated.

Conclusion

Our case exemplifies the potential to minimize maternal-fetal morbidity and mortality by managing pregnancies affected by PAVM with a proactive and multidisciplinary approach. The development of an emergency action plan eased decision making and allowed for timely intervention. Diagnostic testing for HHT and/or PAVMs should be pursued in all at-risk individuals to afford optimization of outcomes.

Conflict of Interest:

None

Acknowledgements:

None

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