Accepted Manuscript online: 2023-07-03 Article published online: 2023-12-14

Spontaneous Hemothorax by Pulmonary Arteriovenous Malformation during Pregnancy

Chuxiao Duan[1] Yunfei Mu[1] Yi Yang[1]

[1] Department of Thoracic Surgery, The Third People's Hospital of Chengdu, The Affiliated Hospital of Southwest Jiaotong University, Chengdu, People's Republic of China

Thorac Cardiovasc Surg Rep 2023;12:e63–e65.

Address for correspondence Yi Yang, MD, Department of Thoracic Surgery, The Third People's Hospital of Chengdu, The Affiliated Hospital of Southwest Jiaotong University, Chengdu 610031, People's Republic of China (e-mail: 191888@qq.com).

Abstract

Background: Pulmonary arteriovenous malformation (PAVM) is a rare vascular malformation that may cause hemothorax, especially during pregnancy.

Case Description: A 25-year-old woman presented sudden-onset left chest pain, dizziness, and dyspnea in the 27th week of gestation. Computed tomography angiography showed left pleural effusion with complete hemithorax opacification and an aneurysmal PAVM. She exhibited hemorrhagic shock and received emergency exploratory video-assisted thoracic surgery. A ruptured PAVM was identified and stopped by wedge resection in the upper lobe of the left lung. The patient's postoperative clinical course was uncomplicated. She subsequently delivered a healthy live baby vaginally at 41 weeks gestation.

Conclusion: PAVM should be considered in pregnant women with hemothorax. Emergency thoracoscopic surgery is the best treatment option.

Keywords
- pulmonary artery and vein
- pregnancy
- shock

Introduction

Pulmonary arteriovenous malformation (PAVM) is a rare vascular malformation resulting from direct communication between the pulmonary artery and vein without interposition of the capillary bed.[1] Most PAVMs are asymptomatic; they are usually found by accident and occasionally, via dyspnea due to a right-to-left shunt.[2] Symptomatic PAVM is usually related to hereditary hemorrhagic telangiectasia, which is an autosomal dominant vascular disorder that is also known as the Osler–Weber–Rendu syndrome.[3] The remaining causes are related to malignancy, trauma, hepatopulmonary syndrome, and heart surgery.[4]

Pregnancy is considered a risk factor for PAVM, mainly due to increased blood volume and cardiac output resulting in increased cardiac load, as well as relaxation of arterial smooth muscle due to increased levels of progesterone during pregnancy. Although the vast majority of pregnant women with PAVM are asymptomatic, when they present with symptoms such as hemoptysis, rupture, or hemothorax, the condition can be lethal.[5] Previous studies have reported the presence of PAVM during pregnancy due to complications, such as rupture, hemothorax, and hypovolemic shock.[6–9]

As early diagnosis and appropriate treatment are crucial for PAVMs, we herein present the case of a pregnant patient with life-threatening spontaneous tension hemothorax caused by a ruptured PAVM who was successfully treated with emergency thoracoscopic surgery.

Case Description

A 25-year-old previously healthy woman in the 27th week of gestation of her first pregnancy was rushed to our

received
February 2, 2023
accepted
June 15, 2023
accepted manuscript online
July 3, 2023

DOI https://doi.org/10.1055/a-2121-7350.
ISSN 2194-7635.

Georg Thieme Verlag KG, Rüdigerstraße 14, 70469 Stuttgart, Germany

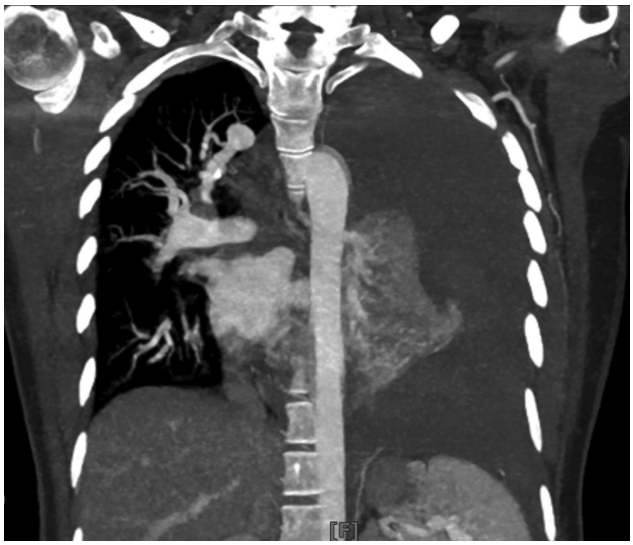

Fig. 1 CT showing an aneurysmal pulmonary arteriovenous malformation located in the upper lobe of the right lung and a massive pleural effusion on the left side.

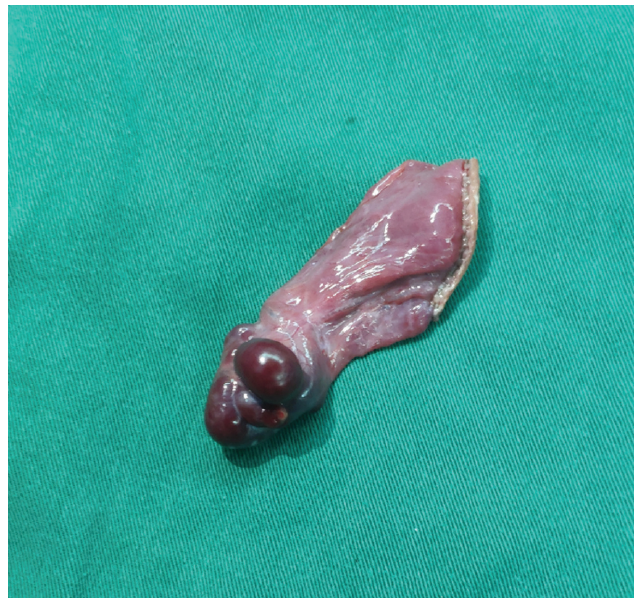

Fig. 3 Lung wedge resection involving the ruptured pulmonary arteriovenous malformation.

emergency department with the chief complaint of sudden-onset left chest pain, dizziness, and dyspnea without trauma. Physical examination revealed decreased breath sounds on the left side, dull percussion notes, and decreased vocal tactile fremitus but no evidence of cyanosis. Obstetric ultrasound revealed a single live intrauterine fetus. Computed tomography angiography (CTA) showed left pleural effusion that caused complete hemithorax opacification and an aneurysmal PAVM with a feeding branch of the upper right pulmonary artery and a dilated draining vein (►**Fig. 1**). Systolic blood pressure decreased to 70 mm Hg despite continuous intravenous infusion. Initial laboratory test results revealed normal platelets, a normal coagulation panel, and hemoglobin of 7.8 g/dL. Thoracentesis revealed blood collection in the left chest, and the patient was diagnosed with hemothorax with persistent bleeding. After multidisciplinary discussions with anesthesiologists and obstetricians, we decided to treat the primary cause first and extend the gestational age to the extent possible. We decided to perform emergency video-assisted thoracic surgery (VATS) because the patient's condition was considered life-threatening. After removal of the retained thrombus and blood inside the pleural space, measuring ~3,000 mL, a ruptured PAVM was identified in the upper lobe of the left lung (►**Fig. 2**). Wedge resection was performed using an endostapler (►**Fig. 3**). No other obvious lesions were found in the lung parenchyma or thoracic wall. As a result, the bleeding was successfully stopped, and the patient's vital signs recovered. Red blood cells (800 mL) and fresh-frozen plasma (800 mL) were transfused during the surgery. Histological examination of the resected lung specimen confirmed a diagnosis of PAVM (►**Fig. 4**). The postoperative clinical

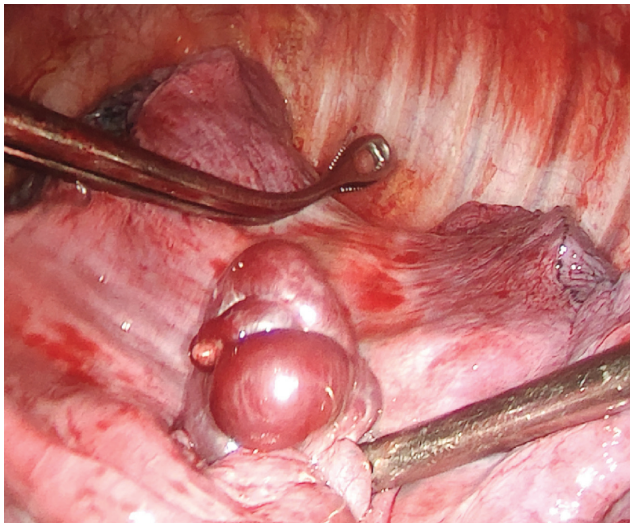

Fig. 2 A ruptured pulmonary arteriovenous malformation in the upper lobe of the left lung.

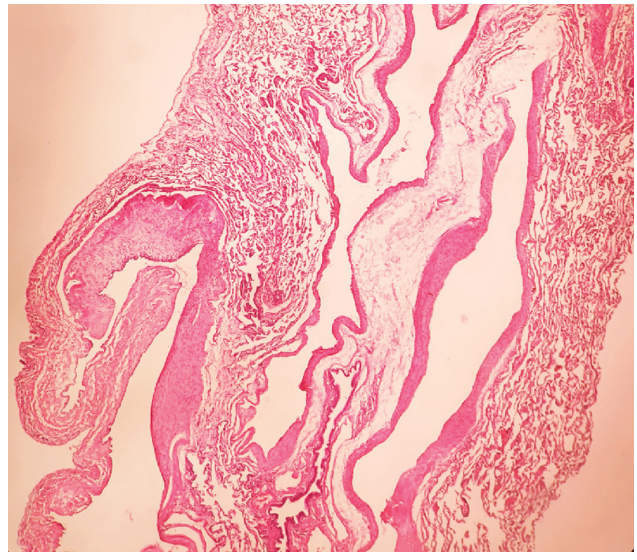

Fig. 4 The histological examination of the resected lung specimen. The histological preparation is hematoxylin-eosin staining (×40).

course was uncomplicated, and the patient was discharged on the sixth postoperative day. At the time of publication, the patient had vaginally delivered a live baby. She and the baby are currently healthy.

Discussion

Our case report discussed a pregnant patient affected by a ruptured PAVM that caused a massive hemothorax compressing the lung, which was successfully treated with emergency thoracoscopic surgery. Moreover, the report summarized the diagnosis and appropriate treatment for PAVMs in pregnant women to avoid life-threatening complications and reduce maternal mortality from the disease.

The prevalence of hemorrhage associated with PAVM is extremely low; however, the risk would relatively increase during pregnancy,[10] which is related to the increase in cardiac work and blood volume, as well as the effect of the estrogen-progesterone imbalance on vessels.[2] Once a PAVM ruptures, bleeding into the pleural cavity results in hemothorax and may lead to progressive dyspnea, pleuritic pain, hypoxia, and hypovolemic shock.

CTA is useful for the diagnosis of PAVM rupture, but it is not recommended in pregnancy because of fetal radiation exposure. However, the fetus is exposed to maximum radiation of 0.66 mGy when chest CT is performed, whereas a dose of $\geq$1 Gy is lethal to the fetus. Therefore, chest CT is safe and can be performed as needed.[7]

Two therapeutic options are recommended for treating patients with PAVM: transcatheter arterial embolization (TAE) and surgical resection (ligature, wedge resection, segmentectomy, lobectomy, and pneumonectomy). Surgical resection of the PAVM is recommended for all patients who can receive general anesthesia, especially for patients with neurological complications, newborns, or central localization of the PAVM.[11] In cases of massive hemothorax, surgery is the only choice.[11] TAE is also an effective treatment and should be performed at the time of diagnosis or when the following criteria are satisfied: progressive enlargement of a detected PAVM, paradoxical embolization, symptoms of hypoxemia, and feeding vessels of 3 mm or larger.[6]

In our case, the patient presented with rapid hemorrhagic shock without an obvious cause. After consulting with the obstetrician and the radiologist, we immediately performed enhanced CT. A PAVM was found in the upper lobe of the right lung although the lesion of the left side was indistinct. Considering the possibility of hemothorax caused by rupture of the PAVM, we performed an emergency VATS first and prepared thoracotomy if necessary, through which we clearly identified the ruptured and bleeding PAVM and successfully achieved hemostasis by removing the PAVM via wedge resection of the lung. Collaboration with anesthesiologists and obstetricians before, during, and after surgery is crucial for saving both the mother and fetus.

Previous studies have reported that hemothorax caused by rupture of PAVMs during pregnancy usually occurs in the third trimester.[7,12] Therefore, they all chose to perform a cesarean section to terminate the pregnancy first and then perform thoracoscopic surgery. However, in our case, the patient was in the second trimester, and we chose to perform emergency thoracoscopic surgery immediately to determine the cause and treat the disease, rather than directly terminate the pregnancy. At the time of publication, the mother and baby were healthy. We have fully informed the mother that the lung condition should be fully assessed as soon as possible after the end of lactation, and interventional therapy should be performed if necessary.

Several key factors are involved in the success of a patient's outcome. Timely and rapid thoracoscopic surgery was performed to determine the cause and to remove the lesions. Second, multidisciplinary cooperation, including obstetricians, anesthesiologists, and other departments, contributed to quick and accurate judgment and action. Finally, systematic treatment and nursing were provided during the perioperative period.

Conflict of Interest
None declared.

References

1 Shovlin CL. Pulmonary arteriovenous malformations. Am J Respir Crit Care Med 2014;190(11):1217–1228
2 Esplin MS, Varner MW. Progression of pulmonary arteriovenous malformation during pregnancy: case report and review of the literature. Obstet Gynecol Surv 1997;52(04):248–253
3 Kühnel T, Wirsching K, Wohlgemuth W, Chavan A, Evert K, Vielsmeier V. Hereditary hemorrhagic telangiectasia. Otolaryngol Clin North Am 2018;51(01):237–254
4 Giordano P, Nigro A, Lenato GM, et al. Screening for children from families with Rendu-Osler-Weber disease: from geneticist to clinician. J Thromb Haemost 2006;4(06):1237–1245
5 Meek ME, Meek JC, Beheshti MV. Management of pulmonary arteriovenous malformations. Semin Intervent Radiol 2011;28 (01):24–31
6 Di Crescenzo V, Napolitano F, Vatrella A, Zeppa P, Laperuta P. Pulmonary arterovenous malformation causing hemothorax in a pregnant woman without Osler-Weber-Rendu syndrome. Open Med (Wars) 2015;10(01):549–554
7 Naito J, Nakajima T, Morimoto J, et al. Emergency surgery for hemothorax due to a ruptured pulmonary arteriovenous malformation. Gen Thorac Cardiovasc Surg 2020;68(12):1528–1531
8 Di Guardo F, Lo Presti V, Costanzo G, et al. Pulmonary arteriovenous malformations (PAVMs) and pregnancy: a rare case of hemothorax and review of the literature. Case Rep Obstet Gynecol 2019;2019:8165791
9 Funaki B. Embolization of pulmonary arteriovenous malformations. Semin Intervent Radiol 2007;24(03):350–355
10 Shovlin CL, Winstock AR, Peters AM, Jackson JE, Hughes JM. Medical complications of pregnancy in hereditary haemorrhagic telangiectasia. QJM 1995;88(12):879–887
11 Kuhajda I, Milosevic M, Ilincic D, et al. Pulmonary arteriovenous malformation-etiology, clinical four case presentations and review of the literature. Ann Transl Med 2015;3(12):171
12 Wang HC, Zhang JH, How CH. Emergency video-assisted thoracic surgery for ruptured pulmonary arteriovenous malformation-related hemothorax in a pregnant woman: a case report. J Med Case Reports 2018;12(01):75