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# Surgery for pulmonary arteriovenous malformations complicating pregnancy: An effective treatment strategy

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## Abstract

Pulmonary arteriovenous malformation (PAVM) is a vascular anomaly of the lung. These lesions which are usually congenital can lead to life threatening complications later in life. It can occur primarily or in association with hereditary haemorrhagic telangiectasia. They are very rare during pregnancy but can cause life threatening complications. We present the case report of a young woman with acute onset of a large hemothorax during the third trimester of pregnancy. Multiple PAVM were noted in the lower lobe of the lung. There was intrauterine death and foetus was evacuated. She underwent thoracotomy and segmental resection of multiple lesions. PAVM should be considered in young women with acute onset of a spontaneous hemothorax and should be managed emergently. Treatment should follow a multidisciplinary approach and complications of PAVMs can be managed effectively with surgery.

**Keywords:** Hemothorax; Pregnancy; Pulmonary arteriovenous malformations

## Introduction

Pulmonary arteriovenous malformations (PAVM) are abnormal direct communications between pulmonary arteries and veins. Incidence of PAVM is about 1:50,000 cases. It occurs twice as often in women as in men. About 13 – 55% of patients are asymptomatic. Symptoms and complications of PAVM manifest in women mostly during pregnancy at the second and third trimesters. This is due to the physiological demands associated with pregnancy. PAVM in pregnancy has a high mortality rate of 0 – 15%. Most of these patients present with chest pain and dyspnoea and are misdiagnosed and treated as pulmonary embolism. Rupture of PAVM should be the first suspicion in a pregnant patient with sudden spontaneous hemothorax.

## Case Report

A 27 years old primigravida with 32 weeks of gestation presented to the emergency department with sudden onset breathing difficulty, profuse sweating and left chest pain. She also gave history of nose bleed at 16 weeks of gestation. She was evaluated elsewhere initially where chest roentgenoogram revealed homogenous opacification of the left lung fields completely. Pleural tapping was done and 400ml of blood was evacuated. On examination she was found to be diaphoretic and severely tachypnic. Her heart rate was 156/minute and blood pressure was 100/70mmHg. Her oxygen saturation was 97% with 5L of oxygen given by mask. At our centre she was admitted in the intensive care unit. Her haemoglobin was 11.1g/dL, total count was 43,000 cells /cumm, platelet count was 4,06,000 cells/cumm. Renal and liver function tests were normal. Coagulation profile was within normal limits. Blood and pleural fluid cultures did not grow any organism. She was treated with non-invasive ventilation. She received intravenous piperacillin and tazobactum 4.5g intravenously every six hours and also analgesics. Bed side ultrasound of the abdomen to assess the foetus revealed foetus corresponding to 33 weeks of gestation and intrauterine foetal death. Chest roentgenogram anteroposterior (Fig. 1) view revealed homogenous opacifications of the upper and lower zones of the left lung with significant collapse of the lung. Bedside ultrasound of the left chest revealed a well-defined hyperechoic lesion measuring approximately 11 X 9 X 7.5cm (400cc) which was suggestive of a hematoma. After initial stabilization she underwent intercostal drainage procedure for the left hemothorax. During the procedure one litre of blood was evacuated from the left pleural cavity and clots were also noted. She continued to have severe bleeding into the pleural cavity. Computed tomography of the chest with intravenous contrast (Fig. 2) revealed two large arteriovenous fistula from the left pulmonary circulation in the superior and anterior basal segments of the lung with large venous sacs and prominent draining veins. They were noted along the pleural surface. The largest venous sac measured 3.2 X 1.7 X 1cm. On the second day of admission she underwent transcatheter embolization (Fig. 3 & 4) of the pulmonary arteriovenous fistula using Amplatzer 4 vascular plug after elective intubation. Subsequently she underwent lower segment caesarean section for evacuation of the dead foetus. Post embolization chest roentgenogram anteroposterior view revealed homogenous opacification of the lower zone of the left lung. On the third day of admission she underwent left posterolateral thoracotomy and non-segmental excision of the PAVMs under general anaesthesia. Intraoperatively two litres of clots were found in the left pleural cavity (Fig. 5). In the lingular lobe a ruptured PAVM with active bleeding was noted (Fig. 6). In the lower lobe medial aspect two pulsatile PAVMs were noted largest measuring 2 X 3cm (Fig. 7). All the clots

were evacuated. Wedge resection of the portions of lung with PAVMs was done using 75mm linear stapler (Ethicon) (Fig. 8). Hemostasis was achieved. Thoracotomy was closed in layers with two pleural drains. She was extubated on the second postoperative day (POD). Post op chest roentgenogram anteroposterior view revealed a fully expanded left lung with clear lung fields and ICDs in position (Fig. 9). Patient was shifted to the ward on the third postoperative day. ICDs were removed on the 4$^{th}$ POD. She was discharged from the hospital on the 8$^{th}$ POD. Histopathological examination of the resected segments revealed lung parenchyma with few vascular spaces of varying thickness with congestion and focal luminal narrowing. The vessel wall was hyalinized and thickened in some areas. A few thin walled vessels were seen around the airways. There was haemorrhage in the lung parenchyma. In the follow-up patient was doing well with no further bleeding episodes.

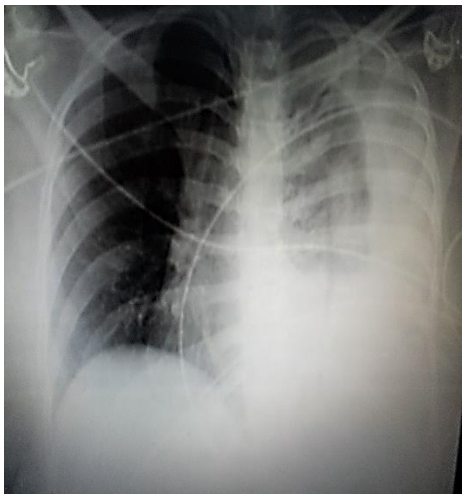

**Fig. 1: Chest X-ray anteroposterior view shows large left hemothorax and collapsed lung**

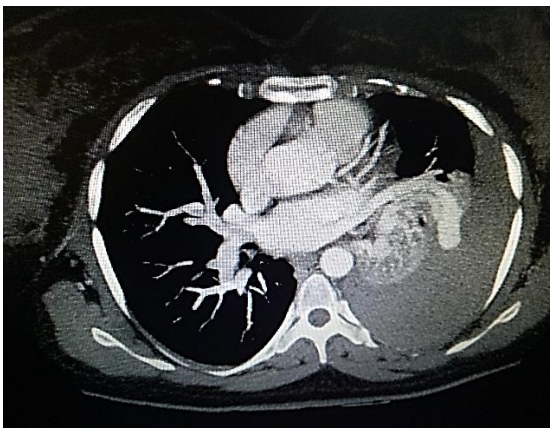

**Fig. 2: Computed tomography of the chest shows large venous sacs and prominent draining veins in the superior and anterior basal segments of the left lung**

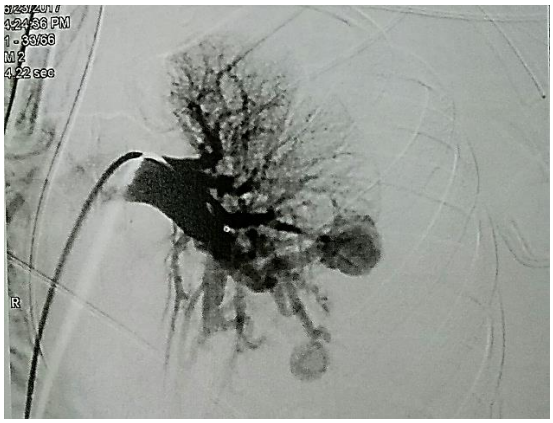

**Fig. 3: Left pulmonary artery angiogram shows two separate arteriovenous fistulas in the left lung**

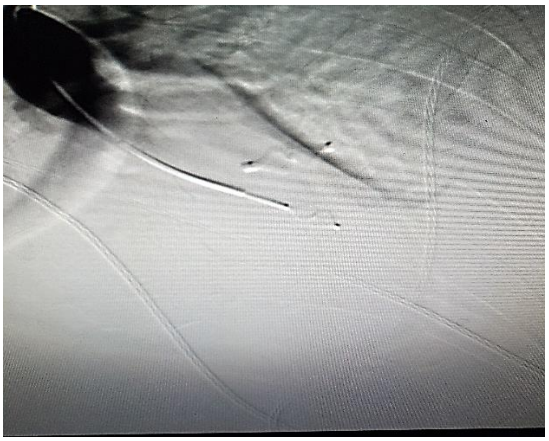

**Fig. 4: Vascular plug embolization of the arteriovenous malformations**

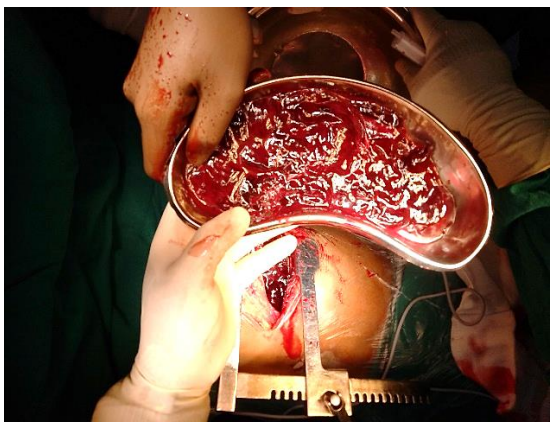

**Fig. 5: Intraoperative image of large amounts of retained clots evacuated from the pleural cavity**

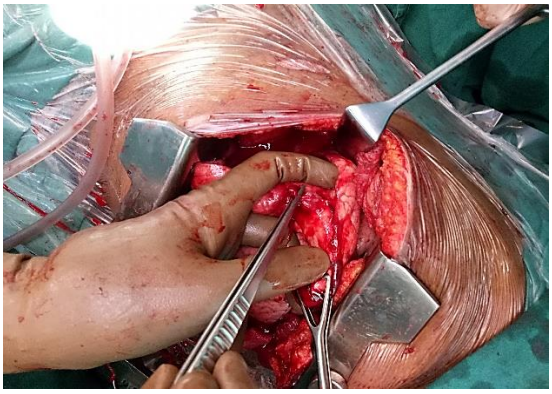

**Fig. 6: Surgical image shows ruptured PAVM with active bleeding**

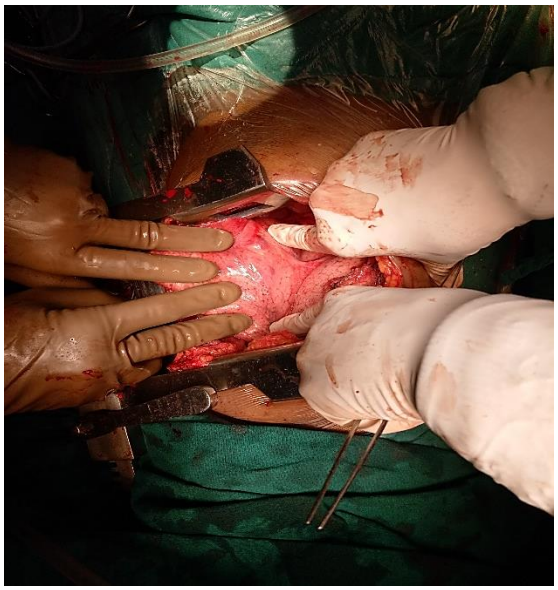

**Fig. 7: Surgical image of pulsating PAVM**

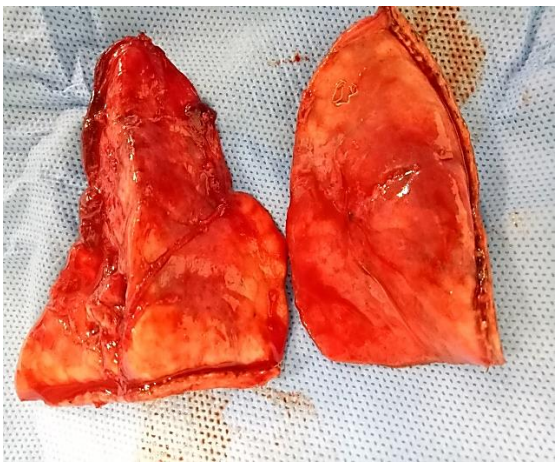

**Fig. 8: Resected lung tissue along with the PAVMs**

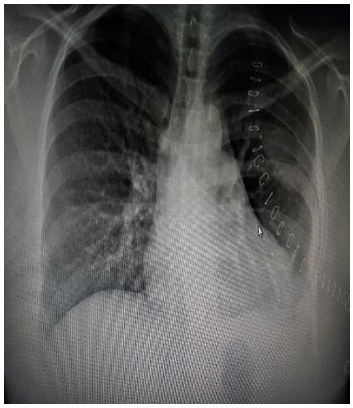

**Fig. 9: Post operative Chest X-ray shows clear left lung fields and a well expanded lung**

## Discussion

Most PAVM are asymptomatic and are unrecognized. Some are incidentally diagnosed while screening for other diseases. Most of it is diagnosed only after complications related to PAVMs occur. Life threatening complications are rupture, paradoxical cerebral embolism and high output congestive cardiac failure. Physiological factors which normally occur during pregnancy such as increased cardiac output, vascular distensibility and progesterone form a favourable environment for the growth of PAVMs. The PAVMs increase in size and the vessel wall becomes thin with the ensuing distension risking rupture anytime [1,2]. Approximately 70% of PAVMs are associated with hereditary haemorrhagic telangiectasia. 15–35 % of patients with HHT have PAVMs. 53-70% PAVMS are found in the lower lobes. 70% of patients have unilateral disease and 36% have multiple lesions of which 50% have bilateral disease. PAVMs can be microscopic but typical lesions are 1 – 5cm in size. The vascular channels are thin walled with a single layer of endothelium. The connective tissue stroma is scant and is disconnected from the normal lung.

Our patient presented with an acute massive hemothorax during her third trimester. She gave history of nose bleed during her fourth month of gestation which was not further evaluated. She could have been a patient of HHT but we had not evaluated her for the same. Ultrasound of the thorax revealed a hyperechoic mass which was due to clots formed within the pleural cavity due to persistent slow bleeding from the ruptured PAVM. Although hemothorax has adverse effects but in the setting of a ruptured PAVM which is a low pressure system it helps to cause tamponade and prevents exsanguination of the patient [3,4]. So emergent intercostal drainage in a pregnant patient who has a spontaneous hemothorax should be planned, only after careful scrutiny of the condition of the patient. Such patients will need emergent definitive intervention in the first place such as transcatheter embolization or surgery. Transcatheter embolization is an effective procedure but carries the risk of radiation exposure to

the foetus in pregnant patients. These patients need effective shielding of the abdomen against radiation but still one is not sure about the hazards of radiation exposure through thorax into the internal organs. In our patient as there was intrauterine demise of the foetus transcatheter embolization did not pose any risks. Other complications include cerebral abscess and recanalization of the PAVMs.

Surgery under general anaesthesia though a safe and effective procedure is not without attendant risks for the foetus. Although embolization causes shrinkage and involution of the PAVM the clots that have already formed within the pleural cavity causes collapse and consolidation of the lung. Clots don't drain thorough the ICD. If significant clots are retained within the pleural cavity it requires thoracotomy or a video assisted thorocoscopy procedure for removal of clots so that the collapsed lung attains adequate expansion and function. In our patient though embolization was done and an ICD was present she had lung collapse due to retained clots and continuing bleeding. During surgery it was noted that the ruptured PAVM was actively bleeding and the other PAVM was pulsating. So, nonanatomical resection of the lung was performed.

Even after embolization of all accessible PAVMs the contrast echocardiogram remains abnormal in 80% of patients. In a study by Lee et al. on embolization of large PAVMs majority were cured by first embolotherapy but 18% required second or third procedure and 12% had recurrence [5]. Bicakcioglu et al. did a retrospective study on 41 patients with PAVM [6]. In their study surgery was preferred in patients where treatment of the embolism was unsuccessful or not possible, in patients with symptomatic and complicated PAVM and/or when a differential diagnosis of PAVM was not possible.

## Conclusions

Management of PAVM during pregnancy needs a multidisciplinary approach with careful discussion and planning done in the best interest on safety of the mother and the foetus. Surgery is the mainstay of treatment of PAVM with complications as resection of the lesions can be done completely in majority of cases and the pleural cavity can be evacuated effectively for better lung expansion and preservation of its function. Surgery avoids the dangers of radiation to the foetus. Transcatheter embolization should be undertaken for patients with poor surgical risk and those with extensive or multiple lesions where effective surgical cure is not possible.

**Conflicts of interest:** None declared

**Acknowledgements:** None

## References

1. Majumder B, Dasgupta S, Chakraborty S, Ghosh S, Chakraborty S, Tandel V, Sudeep KN. Dyspnoea and Cyanosis in Pregnancy: An Extremely Rare Cause. Journal of Cardiovascular Disease Research. 2016;7(4):149-51.
2. Sood N, Sood N, Dhawan V, Pulmonary arteriovenous malformation (avm) causing tension hemothorax in a pregnant woman requiring emergent cesarean delivery. Pulmonary Medicine, 2011;865195.
3. Anin SR, Ogunnoiki W, Sabharwal T, Harrison-Phipps K. Pulmonary arteriovenous malformation unmasked in pregnancy: A case report. Obstetric Medicine. 2013;6(4):179-81.
4. Esplin MS, Varner MW. Progression of pulmonary arteriovenous malformation during pregnancy: case report and review of the literature. Obstet Gynecol Surv. 1997;52(4):248-53.
5. Lee DW, White RI, Egglin TK, et al. embolotherapy of large arteriovenous malformations: Long term results. Ann Thorac Surg 1997:64:930-40.
6. Bicakcioglu P, Gulhan SSE, Sayilir E, Erturk H, Agackiran Y, et al. Surgical treatment of pulmonary arteriovenous malformations. Turk J Med Sci. 2017;47:161-66.