

## Case Report

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# Pulmonary arteriovenous malformation causing hemothorax in a pregnant woman without Osler-Weber-Rendu syndrome

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**Abstract:** Pulmonary arteriovenous malformations (PAVMs), although most commonly congenital, are usually detected later in life. Case report: We present a case of a 19-year-old woman with no previous history of AVM or telangiectasia, who presented dyspnea and hypoxia by massive left hemothorax in the 34th week of gestation. After emergent cesarean delivery, a chest computed tomography (CT) with i.v. contrast showed a likely 3 cm area of active contrast in left lower lung. Chest tube placement revealed about 2 liters of blood. The patient was subsequently found to have pulmonary AVM. A successful embolisation of AVM followed by lung atipic resection involving AVM and decortication for lung re-expansion were the treatments provided. Conclusions: Women with known pulmonary AVM should be maximally treated prior to becoming pregnant, and the physician should be alert to complications of pulmonary AVM during pregnancy.

**Keywords:** Pulmonary arteriovenous malformations, Osler-Weber-Rendu syndrome, pregnant woman

## 1 Introduction

A pulmonary arteriovenous malformation is an abnormal connection between a pulmonary artery and vein that bypasses the alveolocapillary complex. Most cases tend to be simple AVMs (single feeding artery) although up to 20% of cases can have complex (two or more) feeding vessels. There is a recognized female predilection. They can be multiple in around one-third of cases. The impact of this malformation is right-to-left shunting and, subsequently, a varying degree of cyanosis. The lesion is the product of abnormal development of the primitive pulmonary vasculature at the level of the terminal capillary loops. Although there may be only one large artery and one large draining vein, often there are multiple feeder and draining vessels. These lesions may be single, multiple, bilateral, or diffuse. A somewhat complex classification scheme was devised based on the number and relative location of these fistulas within the lung parenchyma. There is an association with Osler-Weber-Rendu syndrome (hereditary hemorrhagic telangiectasia), a syndrome that is also associated with arteriovenous malformations elsewhere. Pulmonary arteriovenous malformations may develop in the presence of liver failure as a presumably acquired phenomenon. In addition PAVMs have been found in hepatic cirrhosis, (as part of the hepatopulmonary syndrome), schistosomiasis, mitral stenosis, trauma, previous cardiac surgery (e.g. Glenn and Fontan procedures for cyanotic congenital heart disease), actinomycosis: thoracic actinomycosis infection, Fanconi syndrome, metastatic thyroid carcinoma, and tuberculosis (Rasmussen aneurysm). Despite most patients being asymptomatic, the connection between the venous and arterial system can lead to dyspnea (due to right-to-left shunting), as well as embolic events (due to paradoxical emboli). Although it is assumed that the vascular defects are present at birth, they are seldom manifested clinically until adult life when the vessels have been subjected to pressure over several decades. Clinically a murmur or bruit may be audible over the lesion (especially if peripheral). Other clinical

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symptoms include easy fatigability and hemoptysis. The patient may appear cyanotic on physical examination. Other complications seen in these patients as a result of the fistula include paradoxical embolus with cerebral injury and brain abscess. The diagnosis is usually made by pulmonary angiography. At the time of diagnosis, therapeutic embolization is possible and is the treatment of choice. Surgical therapy for these malformations has largely been supplanted by embolization therapy. Surgical resection would invariably require removal of otherwise normal lung in addition to that involved in the lesion, but in many cases resection provides long-standing relief of symptoms and clinical manifestations. The concern with embolization therapy is the possibility of recurrence due to recanalization, which may occur in 25% of the cases. The arteriovenous malformations may be too diffuse to effectively treat with either embolization or surgical resection. In rare cases, lung transplantation may be the only feasible treatment possible.

## 2 Case report

Our patient was a 19-year-old previously healthy female at the 34th week of gestation presented to our institute with severe dyspnea, hypoxia and left-sided pleuritic chest pain. Her past medical history presented several short hospitalizations in another hospital for chest pain. During the previous hospitalizations laboratory investigations showed anemia and increased flogistic markers. The discharge diagnosis was: rheumatic disease in pregnant women, but no chest x-ray was performed. Subsequently, after last ospedalization in peripheral hospital, the worsening of her clinical conditions resulted in the transfer to the Gynecology Unit of our structure, where, she finally underwent chest X-ray that showed a massive left pleural effusion (Figure 1) but no history of epistaxis or family history of Osler-Weber-Rendu syndrome was noted. An exploratory thoracentesis showed liquid blood [17-20] without a history of chest trauma. A physical exam was significant for decreased breath sounds on the left side, dull percussion note, and decreased vocal tactile fremitus but no evidence of cyanosis. Vital signs were systolic blood pressure of 90 mm Hg, diastolic blood pressure of 50 mm Hg, pulse rate 120/min, afebrile, and respiratory rate of 30/min. Initial labs revealed normal platelets, normal coagulation panel, and hemoglobin of 8.5 gm/dL. Critical care panel showed pH of 7.4, pCO<sub>2</sub> of 40 mm hg, pAO<sub>2</sub> 60 mm hg, and saturation of 88%. Since the clinical condition was stable and pregnancy almost completed, we decided

to perform emergent cesarean delivery to avoid the risk of the CT-chest with i.v. contrast and of emergent surgery procedure in order to avoid fetal complications [12,13]. A chest tube was placed in operating room and about two liters of frank blood was removed with the chest tube placement which also resulted in normalization of the blood pressure and improved oxygenation on the monitor. CT-chest with i.v. contrast showed a likely 3 cm area of active contrast along with compressive atelectasis of the left lower lobe and pleural effusion (Figure 2). She also underwent a fibrobronchoscopy showing no pathological signs [14,15]. Pulmonary angiography confirmed an AVM of a basilar left pulmonary artery supplying aneurysmal AVM and dilated draining vein. Embolisation of the culprit vessel was performed by placement of a non-adhesive liquid embolic agent (Onyx 34<sup>®</sup>). (Figure 3). Repeat chest X-ray showed no re-expansion of left lung due to protract hemothorax. For this reason, decortication for lung re-expansion and lung wedge resection involving AVM were performed (Figures 4-6). Patient had an uncomplicated course subsequently and was discharged home after 7 days. Evaluation for AVM with a head MRI was negative. Examination or genetic testing for hereditary hemorrhagic telangiectasia (HHT) was not performed during this hospitalization but a genetic examination performed a few months afterwards showed no association.

**Ethical approval:** The research related to human use has been complied with all the relevant national regulations, institutional policies and in accordance the tenets of the Helsinki Declaration, and has been approved by the authors' institutional review board or equivalent committee.

**Informed consent:** Informed consent has been obtained from all individuals included in this study.

## 3 Discussion

Pulmonary AVMs are an abnormal connection between pulmonary artery and a vein, bypassing the pulmonary alveolar-capillary complex. This leads to the abnormal development of the primary vascular system at the level of the terminal capillary loops that provide a direct capillary-free communication between the pulmonary and systemic circulation which causes right-to-left shunt, hypoxia, with a variable degree of cyanosis and increases the risk of paradoxical emboli including cerebral abscess [1, 2]. Although there may be only one large artery and one large draining vein, often there are multiple feeder and

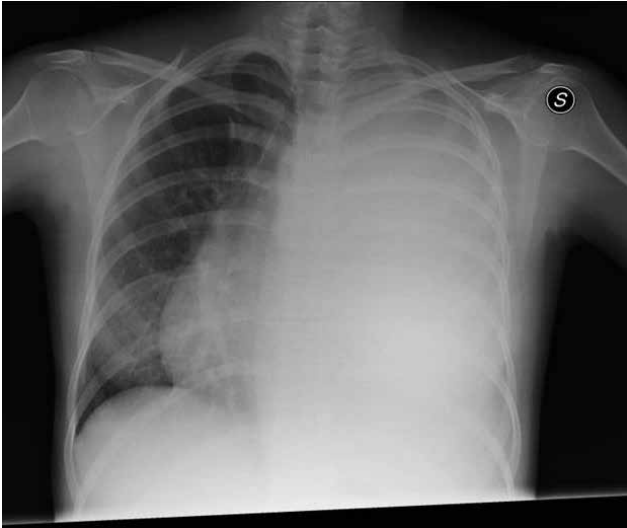


Figure 1: Chest X-ray showing left hemithorax opacification.

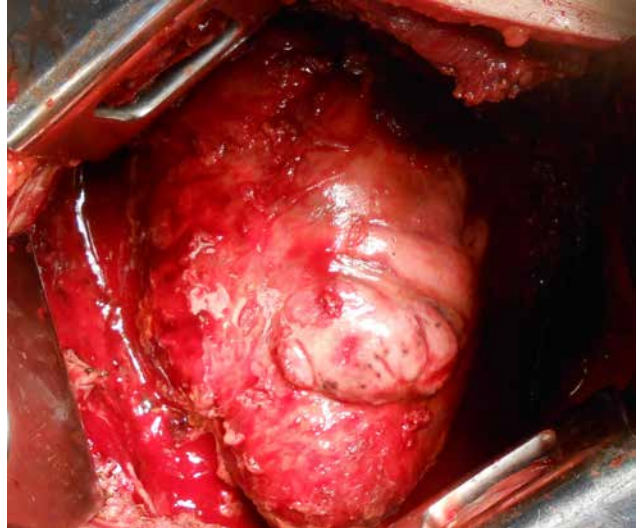


Figure 4: Unexpandable lung showing after thoracotomy



Figure 2: Computed tomography of the chest with I.V. contrast showing left-sided pleural effusion and a likely 3 cm area of active contrast.

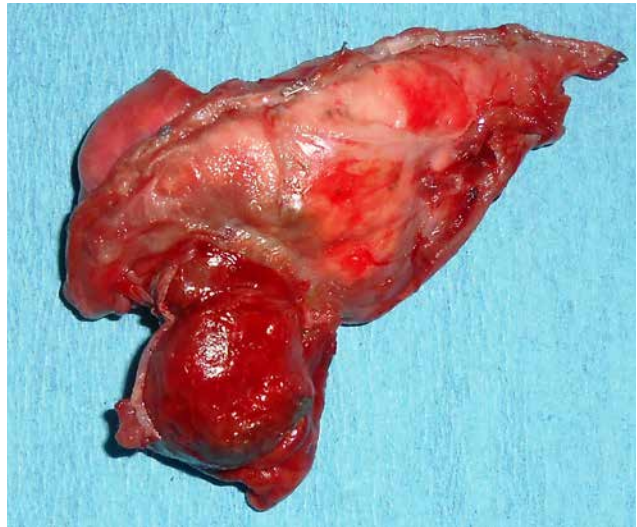


Figure 5: Lung wedge resection involving AVM

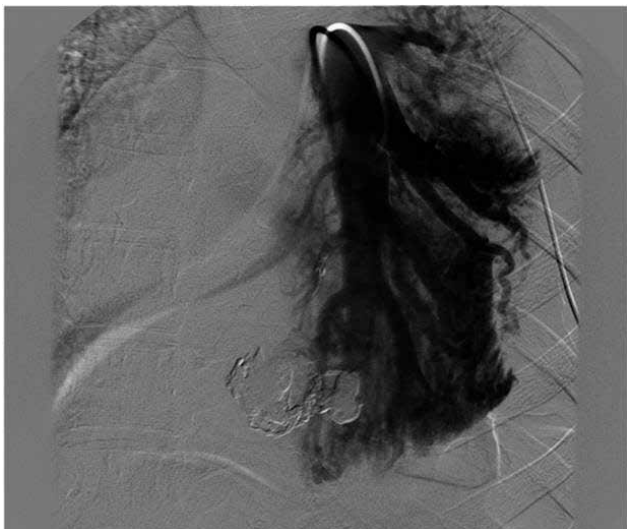


Figure 3: Interventional radiologist-guided embolisation of the left pulmonary artery which was the culprit vessel.

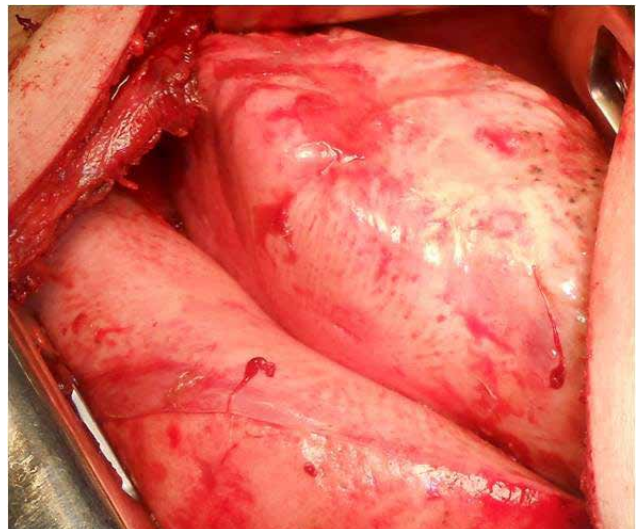


Figure 6: Expanded lung after decortication XXX



draining vessels. These lesions may be single, multiple, bilateral, or diffuse. These AVMs increase in size during pregnancy [3]. The differential diagnosis was for: abnormal systemic vessels, highly vascular parenchymal mass, other congenital or acquired pulmonary arterial or venous lesions (e.g. pulmonary varix), retroperitoneal varices [11,16,21-32].

There is an association (93.6% of cases) with Osler-Weber-Rendu syndrome (hereditary hemorrhagic telangiectasia), a syndrome that is also associated with arteriovenous malformations elsewhere. Pulmonary arteriovenous malformations may develop in the presence of liver failure as a presumably acquired phenomenon [4, 5]. Criteria for hereditary hemorrhagic telangiectasia include spontaneous recurrent nosebleeds, mucocutaneous telangiectasia, visceral involvement [5]. It is important to evaluate all patients with PAVM for signs of HHT. This involves meticulous exam for telangiectatic lesions by looking in the oral cavity, the lips, and nasal mucosa using rigid rhinoscopy. The skin of the face and fingertips and look in the conjunctive of the eye should be included.

Screening for pulmonary AVM includes chest radiograph [23-25, 33-37], arterial blood gas on oxygen, and echocardiogram to evaluate for pulmonary shunting [6]. Screening for cerebral AVM includes MRI and MR angiography (MRA) of the brain although cerebral angiography is most sensitive. Computed tomography of the abdomen and pelvis is recommended for screening for AVM in the abdomen [7].

PAVMs were historically treated with surgical resection. As endovascular techniques developed, embolization became the mainstay of treatment. At the time of diagnosis, therapeutic embolization is possible and is the treatment of choice. Indications for embolotherapy include the following: progressive enlargement of the lesions, paradoxical embolization, symptomatic hypoxic, feeding vessels of 3 mm or larger. The technique of coil embolotherapy involves the localization of the pulmonary arteriovenous malformation by means of angiography, followed by selective catheterization of the feeding artery. A steel coil is advanced through the catheter and placed distal to any branch of the vessel. Sometimes, more than 1 coil is required to completely occlude the vessel. Multiple pulmonary arteriovenous malformations can be embolized in a single session.

Surgical therapy for these malformations has largely been supplanted by embolization therapy. Surgical resection would invariably require removal of otherwise normal lung in addition to that involved in the lesion, but in many cases resection provides long-standing relief of symptoms and clinical manifestations [8, 9]. Pregnancies should

be managed with close liaison between obstetricians, pulmonologists, and interventional radiologists, using appropriate “high-risk” obstetric management strategies. To reduce risk of brain abscess in cases of undiagnosed pulmonary AVM, antibiotic prophylaxis is recommended prior to dental procedures. Embolisation remains the treatment of choice as demonstrated by multiple studies.

## 4 Conclusions

Several cases of pulmonary AVM have been described in the literature with a large proportion of cases associated with hereditary telangiectasia. Our case presenting AVM causing hemothorax in a pregnant woman without Osler-Weber-Rendu with no previously significant medical history is not reported in literature. These AVM expand during pregnancy because of increases in blood volume, cardiac output, and venous distensibility [3]. In this occasion, the combined treatment of embolization with Onyx LES, a non-adhesive liquid embolic agent device utilized for the pre-surgical embolization of arteriovenous malformations (AVM's), and subsequent lung wedge resection [10] involving AVM and decortication for lung re-expansion were performed. Women with known pulmonary AVM should be maximally treated prior to becoming pregnant, and the physician should be alert to complications of pulmonary AVM during pregnancy.

**Conflict of interests:** The Authors have no conflict of interest or any financial support

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