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**Case Report**

# Pulmonary arterio-venous malformation: a catastrophe

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## ABSTRACT

Pulmonary Arteriovenous Malformations (PAVM) are abnormal fistulous connections between a pulmonary artery and a pulmonary vein that generate a right-to-left shunt by avoiding the normal pulmonary capillary bed. We report an unusual case of a young female patient who presented to the department of Obstetrics and Gynaecology with Bleeding per vagina diagnosed as Failed intrauterine pregnancy; Unilateral pedal edema and Breathlessness. Pulmonary Thromboembolism was suspected and Computed Tomography pulmonary angiogram (CTPA) was advised for, which showed abnormal communication between dilated left main pulmonary artery and dilated tortuous superior pulmonary vein. Multiple adjacent solid and ground glass nodules were also noticed which were suggestive of initial telangiectatic state of PAVMs. Most of PAVMs are related to Hereditary Hemorrhagic Telangiectasia, whereas only 10 to 20% are isolated sporadic cases. Pregnancy has been considered as a precipitant factor for PAVMs in most of the cases, patients and pregnant women affected by PAVMs are asymptomatic, but when the clinical manifestations occur, they are often related to the right-to-left shunting and may include dyspnoea, hypoxia, and pulmonary hypertension. Moreover, presence of one or multiple PAVMs during pregnancy is associated with an increased risk of severe complications such as rupture, haemothorax, and hypovolemic shock. Hence this case reports highlights the necessity for the radiologists to think in terms of PAVM as a differential diagnosis beyond the suspicion of Pulmonary thromboembolism to look for any abnormal arteria venous communication while reporting CTPA in pregnant women with breathlessness and foresee the catastrophic complications in an already known case of PAVM during pregnancy. Also, the radiologists should identify subtle solid or ground-glass nodules adjacent to large PAVMS which are the initial telangiectatic stage of PAVMs.

**Keywords:** AVM, Breathlessness, Hereditary haemorrhagic telangiectasia, Pulmonary arteriovenous malformation, Pregnancy, Rendu-Osler-Weber disease

## INTRODUCTION

Abnormal fistulous connections between a pulmonary artery and a pulmonary vein that circumvent the typical pulmonary capillary bed and cause a right-to-left shunt are known as pulmonary arteriovenous malformations (PAVMs).[1] A rare congenital disorder which results from abnormal development of pulmonary arteries and veins from a common plexus. Hereditary hemorrhagic telangiectasia (HHT) or Rendu-Osler-Weber disease has the most common association with PAVMs (80-90% cases), an autosomal dominant disorder which is characterised by multiple arteriovenous malformations of skin, mucous membranes and visceral organs like brain, lungs, spinal cord, liver and pancreas.[2]

## CASE REPORT

A 28-year-old female patient presented to the obstetrics and gynaecology out-patient department with bleeding per vaginum and acute breathlessness. On examination she was found to have unilateral right leg pedal oedema. She

did not give any history of other comorbidities like diabetes, hypertension, asthma, previous cardiac disorder or renal disorder. General physical examination of the patient revealed central cyanosis for the patient (room air, SpO2: 60%) and clubbing in both the upper and lower limbs. Blood work-up revealed elevated haemoglobin level, total count, and D-dimer levels. She was referred to the department of radio-diagnosis for plain chest radiograph, ultrasound abdomen, venous doppler of Bilateral lower limbs and computed tomography pulmonary angiography (CTPA). Plain chest radiograph revealed heterogenous patchy opacity in the left upper and mid zone with adjacent prominent serpiginous vessels which are extending towards hilum and prominent left pulmonary trunk (Figure 1).

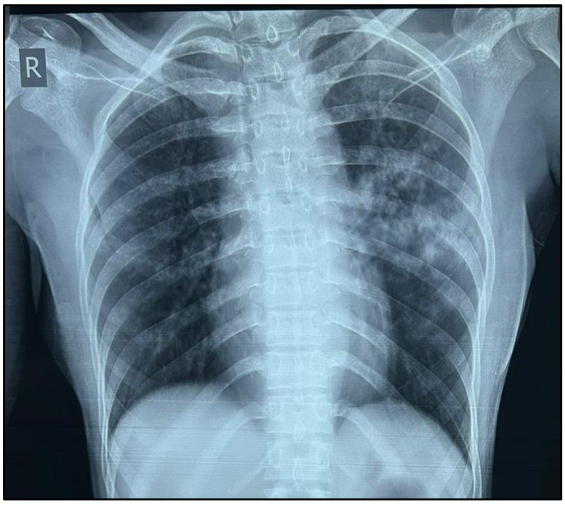


**Figure 1: Plain radiograph of chest reveals heterogenous patchy opacity in the left upper and midzone with adjacent prominent serpiginous vessels which are extending towards hilum and prominent left pulmonary trunk.**

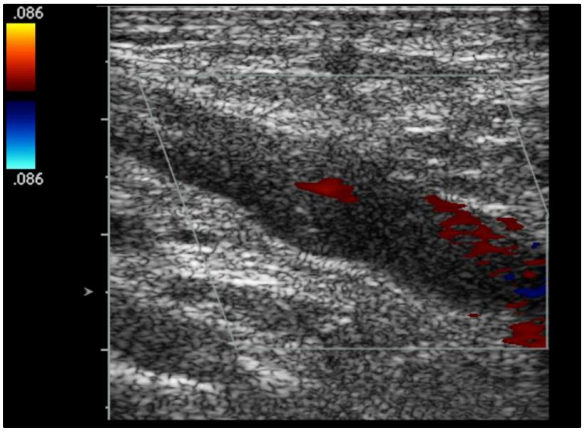


**Figure 2: Venous doppler of right lower limb reveals hypoechoic thrombus within the right popliteal vein with no internal flow; No obvious compressibility - suggestive of acute deep vein thrombosis.**

Ultrasonography of abdomen – pelvis revealed early pregnancy failure of 7 weeks gestation. The patient also gave a history of previous three failed pregnancies, all between 7th to 10th gestational weeks. Venous doppler of the right lower limb revealed acute deep vein thrombosis involving right common femoral vein, superficial femoral vein, popliteal vein, and posterior tibial vein (Figure 2).

CTPA with maximum intensity projection (MIP) images revealed abnormally dilated left pulmonary artery, dilated and tortuous superior pulmonary vein and its branches. An abnormal communication between the dilated left pulmonary artery and superior pulmonary vein (Figure 3 and 4).

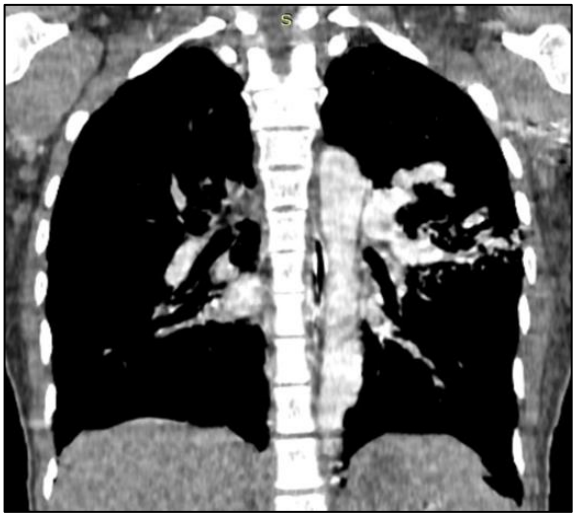

**Figure 3: CTPA coronal section of thorax reveals: Dilated and tortuous course of the superior pulmonary vein and its branches.**

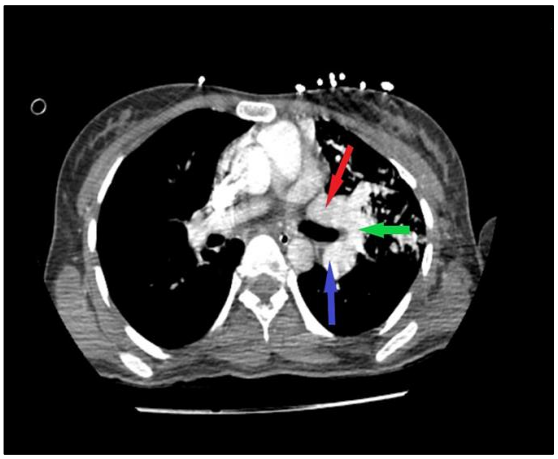

**Figure 4: CTPA axial section of thorax reveals: The left pulmonary artery (red arrow), superior pulmonary vein (blue arrow) and a communication (green arrow) between the two.**

Lung window revealed multiple solid and ground glass nodules in anterior and lingular segments of left upper

lobe- Likely telangiectatic stage of PAVMs (Figure 5A & 5B).

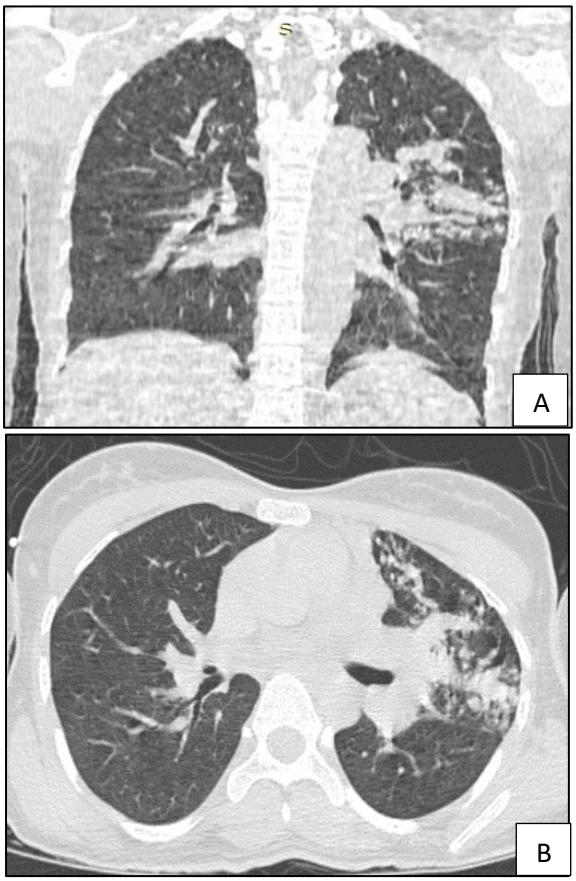


**Figure 5: (A) Coronal section of HRCT Thorax reveals: multiple solid and ground glass nodules in anterior and lingular segments of left upper lobe- likely telangiectatic stage of PAVMs. (B) Axial section of HRCT Thorax reveals: multiple solid and groundglass nodules in anterior and lingular segments of left upper lobe- likely telangiectatic stage of PAVMs.**

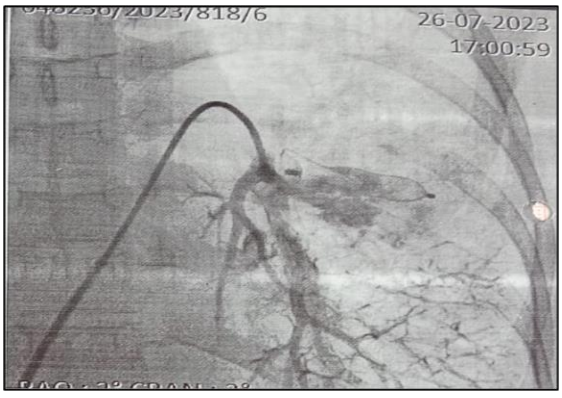


**Figure 6: Pre-closure Angiogram reveals: The contrast injection revealed the two dominant feeding arteries with multiple small ones, the nidus and draining veins.**

No evidence of hemothorax. No obvious rupture of PAVM. No evidence of thromboembolism. The Patient was referred to higher centre for further management following diagnosis and was treated surgically by Pulmonary AV Fistula Device Closure with a 18/20 Patent ductus arteriosus (PDA) Device. The pre- and post-surgical images have been attached for better understanding (Figure 6 and 7). Patient is symptomatically better.

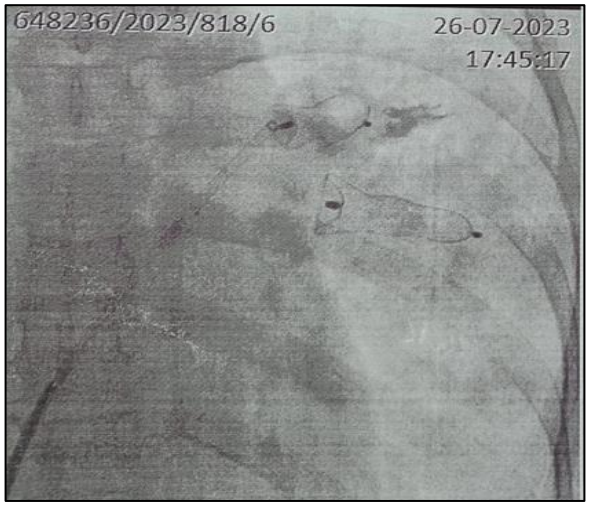


**Figure 7: Post-closure Angiogram reveals: The absence of contrast follows into the nidus through the feeding arteries.**

## DISCUSSION

Pulmonary arteriovenous malformations (PAVMs) are abnormal, direct communications between the pulmonary artery branches and pulmonary veins, but without pulmonary capillaries between them. They are the leading anomalies of the pulmonary vessels, usually congenital and without malignant potential. PAVM can be Simple (80% of cases have a single feeding segmental artery leading to a single draining pulmonary vein); Complex (20% of cases have ≥2 feeding arteries or draining veins); Diffuse: rare, it is a combination of simple and complex variants. Telangiectatic: these are typically seen in children more common in patients with telangiectasia. Based on how the lung and pulmonary vasculature develop throughout embryonic development, PAVMs are divided into five types.[7,8] (Table 1).

The most prominent pathophysiological feature in PAVM is an elevated proportion of right-to-left shunt from the pulmonary artery to the pulmonary vein. Dyspnea on excretion is the most prevalent symptom, occurring in 31-67% of cases. The degree of this symptom is influenced by the degree of hypoxemia and right-to-left shunt. Radiologically, the condition is an important differential diagnosis of the pulmonary nodule. In our case, PAVMs are detected on multimodality chest imaging: Roentography, contrast-enhanced computed tomography (CECT) and computed tomography pulmonary

angiography. On chest radiography, typically, PAVMs appear as well-defined homogeneous soft tissue lesions and are associated with dilated pulmonary vessels.[1,2] CECT is the modality of choice for the characterization of PAVMs.

**Table 1: Classification of PAVM based on the embryonic development of lung and pulmonary vasculature.**

| Group | Subgroup | Imaging features |
|---|---|---|
| **I** | | Multiple small arteriovenous fistulas without aneurysm |
| **II** | | Large arteriovenous aneurysm |
| **III** | A | Large arteriovenous aneurysm (central) |
| | B | Large arteriovenous aneurysm with anomalous venous drainage |
| | C | Multiple small arteriovenous fistulas with anomalous venous drainage |
| **IV** | A | Large venous aneurysm with systemic artery communication |
| | B | Large venous aneurysm without fistula |
| **V** | | Anomalous venous drainage with fistulas |

A homogenous, well-circumscribed, high density, non-calcified enhancing nodule or serpiginous mass lesion connected with blood vessels, showing aneurysmal connection along with the feeding artery and draining vein related to PAVMs.[4] Three components have to be identified on a CT for establishing a diagnosis of PAVM: a feeding artery, a nidus and a draining vein. Ground glass opacities may also be noted between the feeding artery and draining vein, signifying microscopic telangiectasia.[1,6] 3D spiral CT is useful in the pre-therapeutic evaluation of the number and orientation of the feeding arteries. CT pulmonary angiography is the gold standard modality required when further intervention is planned. Multiplanar reformatted images confirmed the diagnosis of PAVMs by establishing the vascular communication with the lesion, with the origin and termination of PAVMs. Pulmonary angiography is the gold standard for diagnosing PAVMs; it is utilized to identify the vascular architecture of individual PAVMs as well as to diagnose PAVMs.[3] Transthoracic Contrast Echocardiography (TTCE), a simple, rapid and minimally invasive examination which is an initial PAVM screening tool with a high sensitivity.[2] Studies have shown that CT is the best non-invasive modality for the evaluation of pulmonary AVMs. Surgery and transcatheter embolotherapy is recommended for treatment of symptomatic patients or those AVMs having a diameter of more than 3mm. Transcatheter embolotherapy with stainless steel coils or detachable balloons is most performed by interventionists.[10]

Pregnancy also has effects of the progesterone on arteries and the rise in cardiac work and blood volume, pregnancy has been identified as a precipitant factor for PAVMs. Pregnancy can lead to changes in the thoracic cage's anatomy. Furthermore, alterations in maternal hormones might result in changes to the mucosa of the airways, including hypersecretion, edema, and hyperemia. Conversely, dyspnea in expectant mothers should always be closely monitored due to the numerous associated disorders, including the rarest ones like paroxysmal airway disease. Concerning the consequences of PAVMs, they can lead to secondary to septic emboli, brain, spinal, or systemic abscesses, and paradoxical septic or nonseptic emboli. While detection and treatment of PAVM should be recommended for asymptomatic pregnant women, this approach is not universally accepted, it has been linked to a significant increase in morbidity and mortality and a high risk of life-threatening complications with one or more untreated cases during pregnancy.[5] The most common complications in pregnancy are recanalization and and rupture of PAVMs. The most common complications associated with PAVMs are stroke, cerebral abscess, rupture, hemoptysis, hemothorax and pulmonary arterial hypertension. The most common vascular PAVM mimics to be ruled out during radiological examination are: fibrosing mediastinitis and venovenous collateals, arterial collaterals, pulmonary artery pseudoaneurysms, hepatopulmonary vessels, meandering pulmonary vein and pulmonary vein varix.[1] To conclude, a practical diagnostic algorithm would be a thorough clinical examination and a chest radiograph and any abnormality to be followed up by a contrast enhanced CT. A contrast echocardiography for establishing a right-to-left shunt and 100% oxygen method to estimate the shunt fraction. Pulmonary angiography is recommended to examine the architecture of the PAVM if any intervention is planned.[3] Transcatheter embolotherapy (TCE) is the treatment of choice in PAVMs.

## CONCLUSION

Pulmonary AVMs are a rare clinical entity. It is usually associated with Hereditary Hemorrhagic Telangiectasia / Osler Weber Rendu syndrome. Although mostly these are congenital malformations, they become overtly symptomatic and PAVM can reveal itself during pregnancy due to physiological changes. Dyspnea during pregnancy should not be attributed alone to Physiological changes of pregnancy or pulmonary thromboembolism alone. Additionally, using the modified early obstetric warning score (MEOWS) charts could guide staff in monitoring a pregnant woman and early recognition of signs and symptoms that are not within the range of normal physiological changes associated with pregnancy. PAVMs are abnormal vascular structures that most often connect a

pulmonary artery to a pulmonary vein, thus bypassing the normal pulmonary capillary bed and resulting in an intrapulmonary right-to-left shunt. The spectrum of PAVMs extends from microscopic lesions causing profound hypoxemia with ground-glass appearance on CT which represent telangiectatic stage of PAVMs to nodules/nidus with feeding arteries and veins. Sometimes rarely Abnormal vascular communication exists between main branch of pulmonary artery and vein. Following a proper imaging algorithm in pregnant women helps us to diagnose PAVMs and save the patient from the catastrophic outcomes.

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