

## Case Report

# Pulmonary arteriovenous malformation: a case report

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## ABSTRACT

Pulmonary arteriovenous malformation (PAVM) is an uncommon pulmonary condition characterized by communications between the arterial and the pulmonary venous system. This disease is often related to hereditary hemorrhagic telangiectasia (HHT), also known as Osler-Weber-Rendu syndrome, and in some cases can be classified as idiopathic. It can be either congenital or acquired, some cases may be asymptomatic, while a less percentage may present symptoms like dyspnea, hemoptysis, hypoxemia, skin telangiectasias, epistaxis and may be associated with significant morbidity and mortality. PAVM should be suspected by clinical manifestations and chest X-ray altered imaging, and the diagnosis should be confirmed by a contrast-enhanced chest computed tomography (CT) scan and pulmonary angiography, being the gold standard test. The most effective therapy is embolization, but some patients may need alternative treatments, like surgical excision or lung transplantation, as an alternative reserved for severe cases where other interventions have been ineffective, or due to the lack of resources to perform the embolization procedure because of the socioeconomic reality of the country. We report the case of a patient with a 5-year history of hemoptysis, cough, dyspnea, and fatigue, with a possible apparent cause related to a surgical procedure performed in 2018. Initially, the differential diagnosis was aspergillosis, but after conducting laboratory tests and a contrast-enhanced CT scan, the diagnosis of arteriovenous malformation in the right lower lobe was made, which was successfully corrected surgically.

**Keywords:** Pulmonary arteriovenous malformation, Hemoptysis, Pulmonary angiography

## INTRODUCTION

Pulmonary arteriovenous malformations (PAVMs) are caused by abnormal communications between pulmonary arteries and pulmonary veins communicating directly without interposition of capillary bed.<sup>1</sup> Near 10% of the cases of PAVM are detected in childhood but the majority are detected around the 5th and 6th decades when symptoms appear.<sup>2</sup> There are factors such as puberty pregnancy and pulmonary artery hypertension that make PAVM increase in size over time making more likely the start of symptoms around the 4th and 6th decade.<sup>2</sup>

Pulmonary arteries supply most of the PAVM, less frequently by systemic arteries. The most common symptom is epistaxis followed by dyspnea, mainly seen in patients with large or multiple PAVM.

Hemoptysis is the third most common symptom, but it is the most common presenting complaint, massive hemoptysis may occur, but it is rarely fatal. Regarding to signs, superficial telangiectasias are usually the only physical finding in patients with PAVM.<sup>2</sup> There is evidence of a female predominance, with PAVMs observed approximately twice as frequently in females compared to males. Over 90% of PAVMs are unilateral

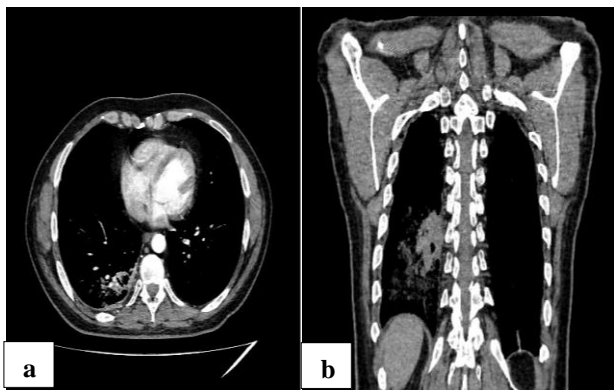
and solitary, with a marked predilection for the lower lung lobes.<sup>3</sup>

## CASE REPORT

A 50-year-old male patient with a clinical history beginning in 2018 presented with an episode of frank hemoptysis lasting one month. He was diagnosed with pleural effusion consistent with empyema and right-sided pleural thickening secondary to community-acquired pneumonia, without identification of a causative organism (BAAR negative). As a result, he underwent right thoracotomy, decortication, fistulectomy, and partial pleurectomy in the same year.

Two-months post-surgery, the patient experienced chest pain, cough, and self-limiting episodes of minor hemoptysis (approximately 30 ml over 72 hours). Similar episodes recurred in 2019, 2020, and 2022. During this period, he attended general medicine and pulmonology consultations.

Radiological studies revealed a ground-glass opacity pattern in the pulmonary parenchyma with bilateral bronchovascular enhancement, pleural thickening of the right lower lung, and parenchymal scarring in the right lower lobe. No additional treatment was administered due to spontaneous symptom resolution.



**Figure 1: Chest CT scan (mediastinal window), (a) axial section; and (b) coronal section. Right paracardiac vascular nodular image with significant enhancement with contrast medium.**

In 2023, the patient presented to the emergency department with significant hemoptysis (300 ml within 24 hours), accompanied by sharp right dorsal chest pain, cough, and severe fatigue. A chest X-ray revealed infiltrates, right pleural thickening, ipsilateral diaphragmatic elevation, a fibrotic band in the right hilum, and signs of diffuse alveolar hemorrhage. He was hospitalized for 21 days and managed with tranexamic acid and analgesics. Attempts to perform pulmonary arteriography and embolization were unsuccessful due to resource limitations in our country, prompting referral to a tertiary care hospital for evaluation by thoracic surgery.

At the tertiary center, serum galactomannan testing was performed to rule out aspergillosis, yielding negative results. Subsequently, a contrast-enhanced computed tomography (CT) in Figures 1a and b, revealed an arteriovenous malformation (AVM) in the right lower lobe. Due to the persistence of symptoms and the risk of recurrent hemorrhage, surgical intervention was determined to be the definitive treatment.

## DISCUSSION

PAVMs are abnormal communications between the pulmonary arterial and venous systems. Most patients have multiple PAVMs, of variable size and generally located in the lower lung lobes, which become clinically apparent from the second decade of life, perhaps due to their slow growth with age, hormonal influence, and trauma.

They usually have a rounded, oval or lobulated morphology, and a caliber that ranges from a few millimeters to several centimeters. These are classified according to their angiographic appearance as simple or complex PAVMs.<sup>4</sup>

The most frequent forms of PAVMs (up to 80% of the total) are simple, which have only one afferent artery and one efferent vein, while complex PAVMs have one or more afferent arteries and a highly developed and complex venous drainage system. The venous bed is usually dilated with a fusiform or aneurysmal morphology and has a system of vascular channels interposed between the arteries and veins. Paradoxical embolism can be observed in this type of PAVM.

Diagnosis is based on clinical suspicion and is then confirmed with imaging such as CT, MRI or digital angiography, which would help plan treatment. More than 95% of patients have radiological changes and the most frequent is the presence of a well-defined, non-calcified peripheral nodule, attached to hilar structures by blood vessels. Occasionally, they may calcify and may grow with age. A sensitivity and specificity of 92% and 83%, respectively, have been reported. In the initial X-ray of the case presented a nodule was not observed but a very well-defined mass without calcifications.<sup>5</sup>

A contrast computed tomography was performed, showing the presence of a cluster in the right lower lobe, associated with ground glass changes consistent with an arteriovenous malformation of the right lower lobe, leading to a surgical resolution. Pre-surgical evaluation was requested by internal medicine, where no surgical contraindication was found; evaluating him as follows: Intermediate surgical risk, LEE index 0, complications of 0.4%, moderate risk of deep vein thrombosis with indication of antithrombotic measures.<sup>6</sup>

Thoracotomy + right lower lobectomy + pulmonary raffia + right unilateral total lung lavage + placement of right chest tubes 28 and 32 FR was performed with a clean-

contaminated wound. An indurated lesion was found at the level of the right lower lobe in posterior segments measuring almost 4cm in diameter and not dependent on a communication with aortic vessels.

There were no surgical complications. Following her right lower lobectomy, her oxygen saturation declined 88% symptoms ceased and discharged well. In future medical evaluations, there is no evidence of respiratory distress, reaching saturation levels above 90%. The last follow-up chest X-ray performed 7 months after the surgery shows: no pleural effusion, expanded lung fields, elevation of the right hemidiaphragm, and a centered mediastinum. The patient today, after one and a half years' post-surgery, leads a normal life without complications.

## CONCLUSION

PAVMs are a rare entity, usually asymptomatic, which must be taken into account in the differential diagnosis in the presence of a pulmonary mass. Scanning angiography is a reliable diagnostic method that allows adequate characterization of the components of the malformation and appropriate surgical planning.

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