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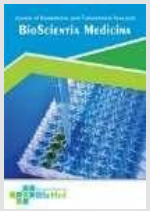

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# A Deadly Spontaneus Hemopneumothorax: A Case Report

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**A B S T R A C T**

**Background:** Spontaneous hemopneumothorax (SHP) is a subtype of hemopneumothorax in which there is an accumulation of blood and air in the pleural space without trauma or other definitive cause. Spontaneous hemopneumothorax is a rare disease. **Case presentation:** A man, aged 32 years, came with a complaint of sudden shortness of breath due to a right hemopneumothorax patient who was put on a chest tube insertion. It looked like 1500 cc of blood came out accompanied by an air component. Next, the chest tube is connected to the water-sealed drainage. The patient found a decrease in Hb from 15 mg/dl to 12 mg/dl. In the next nine hours, the patient's Hb became 10 mg/dl. As a result of this condition, the patient underwent an emergency thoracotomy to evaluate the source of bleeding, where the patient's source bleeding was found to be an arterial vein malformation (AVM) at the apex of the lung attached to the chest wall. Anatomical and pathological examination showed confirmed arterial vein malformation. **Conclusion:** Spontaneous hemopneumothorax, in this case, is a rare case caused by arterial-vein malformation. The right decision in patient management will determine the outcome and outcome of the treatment given.

## 1. Introduction

Spontaneous hemopneumothorax (SHP) is a subtype of hemopneumothorax in which there is an accumulation of blood and air in the pleural space in the absence of trauma or other definitive cause. Spontaneous hemopneumothorax is a rare disease, with the risk of developing SHP around 0.5-11.6% in patients with initially spontaneous pneumothorax (SP), and this disease can be life-threatening.[1,2] Bleeding most commonly results from the rupture of a vascularized bulla or tearing of the adhesion between the visceral and parietal pleura. This bleeding may worsen due to hemodynamic instability resulting from hypovolemic shock. In this case, we report where SHP with consequent Artery Vein Malformation (AVM) of the lung.[3]

Pulmonary arterial vein malformation (PAVM) is defined as the presence of an inappropriate direct vascular connection. Hemothorax in PAVM is a rare clinical entity. Spontaneous hemothorax, in this case, due to ruptured subpleural pulmonary arteriovenous malformation (PAVM) is a potentially life-threatening event that requires urgent interventional therapy.[4]

## 2. Case Presentation

A male aged 32 years presented with complaints of sudden shortness of breath. This complaint has been

experienced since one day ago and is accompanied by right chest pain. This patient was then taken to the emergency department. Examination of vital signs showed an unstable hemodynamic state, blood pressure 92/52 mmHg, heart rate 138 x/minute, RR 32 x/minute, and $SpO_2$ of the patient was 99%. The patient was given $O_2$ via a nasal cannula at 2L/min. Chest x-ray imaging showed a right hydropneumothorax.

The management of this patient is by inserting a chest tube. When a chest tube was inserted, 1500 cc of blood was seen, accompanied by an air component. Next, the chest tube is connected to the water-sealed drainage. After the chest tube was placed, the patient's hemodynamics began to stabilize with a MAP > 70 mmHg and a decrease in heart rate to 110 x/min.

On observation after chest tube insertion, continuous bleeding was found until the total bleeding was 2500 cc, so serial hemoglobin checks were performed. The patient found a decrease in Hb from 15 mg/dl to 12 mg/dl. In the next nine hours, the patient's Hb became 10 mg/dl. As a result of this situation, the patient underwent an emergency thoracotomy to evaluate the source of the bleeding. In intraoperatively, the patient's source of bleeding was found from arterial vein malformation (AVM) at the apex of the lung attached to the chest wall. In patients with wedge resection AVM, the tissue obtained was examined for anatomical pathology with confirmed AVM results. The AVM attaches to the blood vessel in a clip so that bleeding can be stopped. The patient went home after 3 days postoperatively in good condition.

## 3. Discussion

SHP was defined as the accumulation of more than 400 ml of blood in the pleural space associated with a primary spontaneous pneumothorax in the absence of trauma. Pneumothorax caused by hemothorax is the result of impaired adhesion of blood vessels located between the visceral and parietal pleural surfaces.[4,5] Cases of SHP are rare cases. SHP is a potentially life-threatening condition. SHP is hemodynamically unstable. This hemodynamically unstable state is a determining factor for the success of treatment. Where this is a frequent occurrence in males causing bleeding may be due to added strength in training and the reported male-to-female ratio in SHP of 25:1.[6]

The mechanism of bleeding in SHP is due to the tearing of the parietal and visceral pleural vascular adhesions or to tearing of congenital aberrant vessels, but cases of these aberrant vessels have only been reported in a few studies. Pathological studies showed fibrosis with positive alcian bulla deposits in the media and intima.[7,8] Chest radiography is the most useful diagnostic tool for finding bleeding. These patients usually show radiological evidence suggesting a hydropneumothorax.[8]

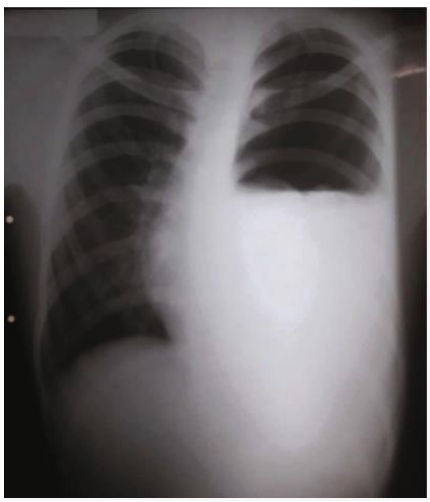

Figure 1. Hydropneumothorax.

In SHP patients, it is necessary to ask about the patient's age and history, clinical evaluation, radiological examination, hemoglobin examination, aspiration of air and blood, and respiratory fluid results are very useful to distinguish between SHP and effusion mixed with blood.[9,10] The therapeutic policy is decided individually based on the patient's condition and the clinical presentation of the patient's disease. The clinical picture depends on the amount of blood loss and air leakage. Initial treatment goals include resuscitation, hemostasis, and lung reexpansion.[11,12] Initial treatment is fluid resuscitation and pleural drainage. Homologous blood transfusions may also be required if the patient's hemoglobin is low.[13,14]

Indications for thoracotomy are hypovolemic shock due to continuous bleeding (100 ml/hour), persistent air leakage, impaired lung expansion, patchy pleurisy, or recurrent pneumothorax. In the absence of such cases, the patient is treated conservatively with a tube thoracostomy.[15] Intraoperative management, where the source of bleeding is identified, adhesion cauterized or ligated. Subsequently, pleural abrasion was performed, and drainage was performed on the patient. Video-assisted thoracoscopic surgery (VATS) is the best treatment for thoracopulmonary problems, especially for spontaneous pneumothorax.[16,17] This therapy is an alternative method that reduces trauma, causes less postoperative pain, and results in shorter hospital stays. VATS was used after initial resuscitation. According to previous studies, thoracotomy is safer in patients with shock in whom major bleeding occurs.[18] Tube thoracostomy and thoracentesis alone are effective. While there are also things that must be resolved, such as bleeding adhesion ligation, pleural cavity irrigation, and pleural abrasion through a posterolateral or lateral thoracotomy, depending on the cause of the patient's SHP.[19]

Pulmonary arterial vein malformation (PAVM) is a rare condition. PAVM is defined as there is an inappropriate direct vascular connection, called the nidus, between the pulmonary artery and vein, which passes from the base of the pulmonary capillaries. This is what causes right-to-left shunts.[20] PAVM requires prompt diagnosis and correct management when clinically suspected, although the frequency is low: Serious clinical conditions such as cerebrovascular stroke or transient ischemic attack, abscess, embolization, hemoptysis, and hemothorax are described as a result of thrombosis, distal embolization or rupture of the PAVMs.[21] This imaging examination serves as a PAVM decision-making process; CXR, CT, and MR provide an important element for the treatment and careful monitoring of diagnosed PAVM.[22]

The clinical presentation of PAVMs is highly variable:[23] Some patients are completely asymptomatic, and others present with unexplained or life-threatening respiratory symptoms such as dyspnea and hypoxemia at rest or during activity. Platypnea (orthostatic dyspnea) and orthodeoxia (orthostatic hypoxemia) are hallmarks of PAVM. PAVM may also manifest as a clinical emergency with hemoptysis and/or hemothorax.[24]

The chest x-ray appearance of the PAVM is chameleonic (Figure 2). Arteries and veins usually point in different directions: Arteries originate at the hilum of the lung and reach the fistula, whereas drainage veins arise from the PAVM to reach the left atrium. If visible, the feeding vessel appears as a tubular opacity connecting the suspected fistula to the mediastinum, and the veins are usually larger than the arteries.[25]

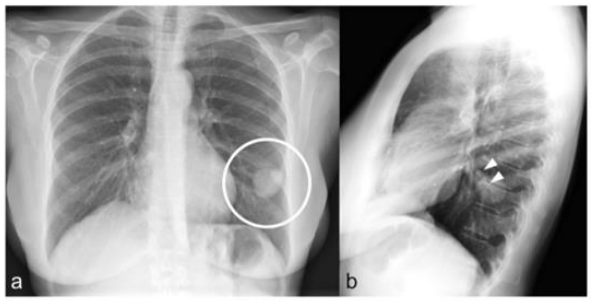


Figure 2. Chest x-ray radiograph shows two well-defined homogeneous masses with a lobed (circle) shape in the left subpleural parenchymal of the lung (a) confirmed on a lateral radiograph (b) which better also shows serpiginous opacities directed from lesion to the hilum, this figure refers to the blood vessels (arrows) .

CT scan is the gold standard among non-invasive diagnostic tools for PAVM (Figures 3 & 4). PAVM picture: solid nodules are found in the periphery, round or polylobate, with well-defined borders.[26] Complex PAVMs are also recognized on CT scans for the presence of more than one arterial supply originating from multiple segmental arterial branches. PAVM telangiectasias present as ground-glass areas, with a solid nodular component within them, with multiple characteristic PAVMs usually occurring simultaneously in both lungs.[27]

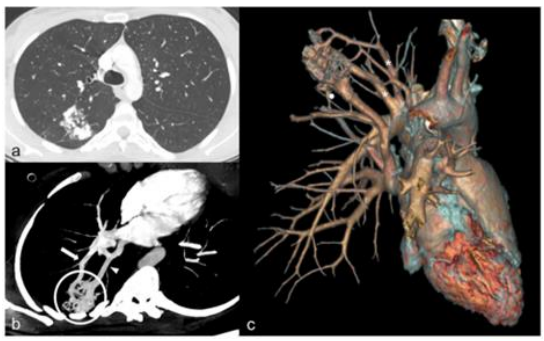


Figure 3. Pathological vasculature in the right upper lobe (a). The maximum intensity projection image confirms the presence of a macroscopic PAVM characterized by the presence of a nidus (circle) that receives the arterial feeder (arrow) from the apical segmental pulmonary artery and drains into the large venous collector (arrow) (b) Reconstruction of volume rendering well illustrates the complex nature of the lesion. Which shows the feeder arteries arising from the apical (asterisk) and posterior (dot) segmental arteries of the right upper lobe.

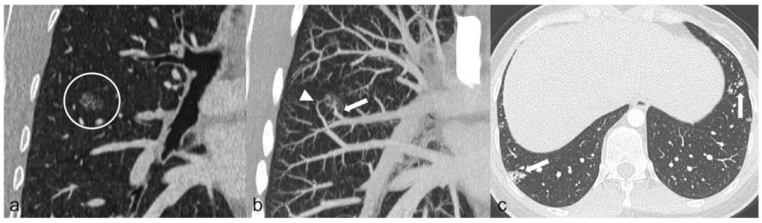


Figure 4. (a) Coronal CT-scan image showing the presence of a 23 mm (circle) nodule with well-defined margins and a “ground-glass” appearance. (b) Maximum intensity projection clearly shows the artery feeding (arrows) and veins (arrows) of the PAVM. (c) The presence of multiple simple PAVMs (arrows) in the lower lobes strengthens the presumptive diagnosis.

The PAVM classification currently used was proposed by White (Figure 5). PAVMs are classified as simple and complex based on their angioarchitecture.[24,28] The two types differ in the number of segmental afferent arteries: one segmental afferent artery for the simple type and two or more segmental arteries for the complex type. Two specific subtypes of complex PAVM are diffuse and telangiectasia:[29] The diffuse subtype is characterized by the involvement of a large part of the parenchyma, sometimes even the entire lung, by a network of deformed blood vessels; PAVM telangiectasias involve only a portion of the lung lobes, and the vascular tissue is usually microscopic. PAVMs are also classified according to the size of the feeding arteries: If the feeding arteries have a diameter of 3 mm or greater, the PAVM is classified as macroscopic or microscopic.

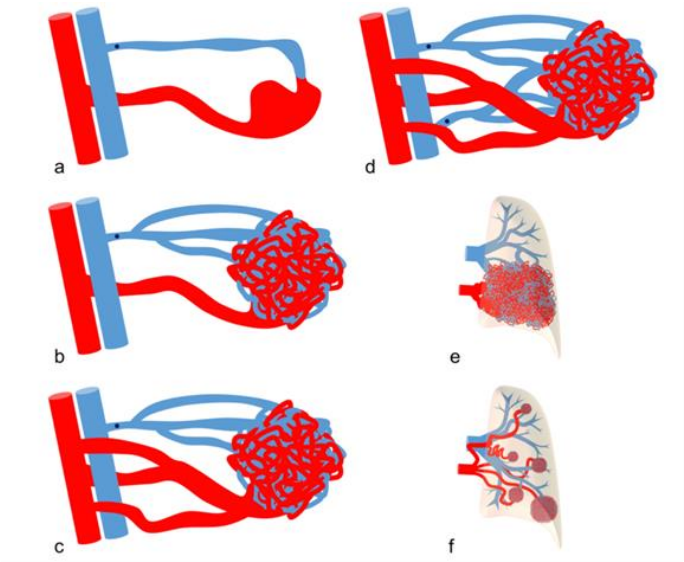


Figure 5. Schematic drawing of the PAVM classification. Simple PAVM is characterized by the presence of a single segmental feeding artery; The angio architecture may consist of a direct connection between a segmental artery and a vein, usually an aneurysm (a) or several sub-segmental arteries joining in a network of malformed vessels that drain into one (b) or several (c) veins. The complex PAVM receives its blood supply from two or more segmental arteries (d); the diffuse subtype showed extensive parenchymal involvement with defective vessel damage of the same size (e) that in the telangiectasia subtype, it was too small to be seen on imaging (f).

## 4. Conclusion

Spontaneous hemopneumothorax, in this case, is a rare case of arterial-vein malformation. The right decision in patient management will determine the outcome and outcome of the treatment given.

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