



**Cardiopulmonary Imaging** Case Report

## Pulmonary Artery Pseudoaneurysm due to Mucormycosis: Case Report and Literature Review

Roberto Mempin<sup>1</sup>, Nader Kamangar<sup>1</sup>

<sup>1</sup>Division of Pulmonary and Critical Care Medicine, Olive View-UCLA Medical Center, David Geffen School of Medicine at UCLA, Los Angeles, California, United States.



**\*Corresponding author:**

Nader Kamangar,  
Division of Pulmonary and  
Critical Care Medicine,  
Olive View-UCLA Medical  
Center, David Geffen School  
of Medicine at UCLA,  
Los Angeles, California,  
United States.

[kamangar@ucla.edu](mailto:kamangar@ucla.edu)

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### ABSTRACT

Pulmonary artery pseudoaneurysm (PAP) is a rare vascular phenomenon with a high mortality rate, as these entities can enlarge, rupture, and lead to asphyxiation. Pulmonary mucormycosis (PM), an underdiagnosed but an increasingly seen entity in the era of chemotherapy and immunosuppression, is a known cause of PAP, and should be suspected in immunosuppressed patients with hemoptysis. We present a case of PAP due to PM in a patient with recently diagnosed diffuse large B-cell lymphoma of the liver who underwent chemotherapy and developed acute cavitory lung disease and hemoptysis. His diagnosis was delayed due to the withholding of iodinated contrast with computer tomography (CT) imaging in the setting of renal failure. He then underwent embolization of his PAP with resolution of his hemoptysis. PAP is an uncommon cause of hemoptysis that can be diagnosed with CT pulmonary angiography, and mucormycosis is a known but rare cause of PAP in patients with malignancy receiving immunosuppression.

**Keywords:** Pulmonary artery pseudoaneurysm, Pulmonary mucormycosis, Cavitory lung disease, Hemoptysis

### INTRODUCTION

Pulmonary artery pseudoaneurysm (PAP) is seen in up to 11% of patients presenting with hemoptysis and who underwent bronchial or pulmonary angiography.<sup>[1]</sup> They can be congenital or acquired, with the most common etiologies of acquired PAP being infection, chronic inflammation seen in vasculitis, malignancy, and traumatic injury.<sup>[2]</sup> PAPs ultimately arise from inflammation of the vessel wall, either due to pneumonia, septic emboli, or direct invasion by an organism or tumor, leading to eventual thinning of the vessel wall and creation of the pseudoaneurysm. Pulmonary mucormycosis (PM) is a disease that is similarly uncommon, but is a known cause of PAP, especially in patients with underlying malignancy receiving immunosuppression. PAP due to PM is an extremely rare clinical entity that requires a high clinical suspicion and necessitates computer tomography (CT) pulmonary angiography and bronchoscopy for diagnosis. We herein report a case of a patient with recently diagnosed diffuse large B-cell lymphoma (DLBCL) of the liver who presented with failure to thrive and developed hemoptysis due to PM shortly after chemotherapy. He was ultimately found to have a PAP due to his underlying infection and underwent embolization which resolved his hemoptysis.

### CASE HISTORY

A 65-year-old man a history of autoimmune hepatitis, hypertension, hyperlipidemia, and benign prostatic hypertrophy was admitted to our intensive care unit (ICU) after presenting with

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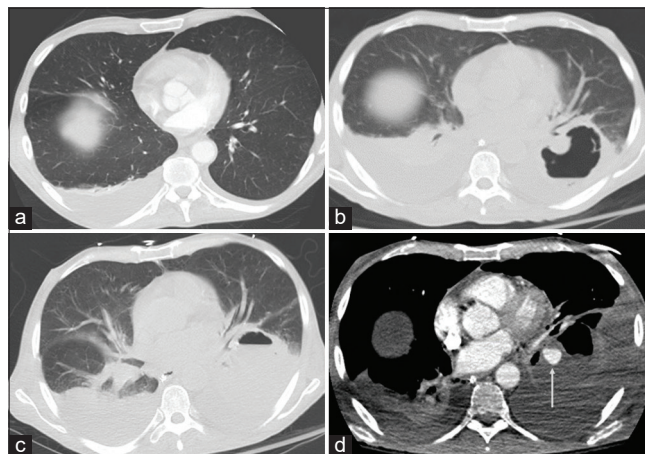
severe symptomatic hypotonic hyponatremia 1 month after being diagnosed with DLBCL of his liver. His medications included furosemide, allopurinol, prazosin, folic acid, ursodiol, pantoprazole, and prednisolone. On resolution of his hyponatremia, he was transferred to the oncology service and was started on therapy for his DLBCL with cyclophosphamide, rituximab, and dexamethasone. His hospital course was complicated by tumor lysis syndrome with associated renal failure requiring hemodialysis, as well as volume overload resulting in respiratory failure necessitating intubation and mechanical ventilation. He was later extubated and transferred back to the oncology service.

On hospital day 18, on completion of a chest CT, the patient suffered PEA arrest, secondary to central airway obstruction caused by blood clots, requiring emergent intubation. He achieved return of spontaneous circulation within 3 min and did not require vasopressor support.

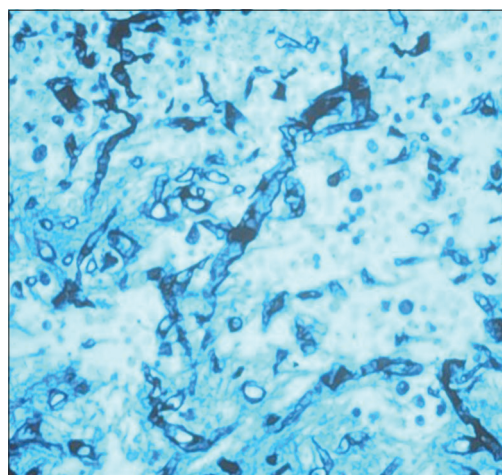
The patient was intubated and appeared cachectic. His vital signs were as follows: Temperature, 38.2°C; pulse, 72/min; blood pressure 107/61 mmHg; respiratory rate 21; and oxygen saturation 97% while on FiO<sub>2</sub> 50%. The patient's pupils were equally round and reactive, but he was nonresponsive to verbal or physical stimuli off sedation. Dried blood was present in the oropharynx and bloody secretions were suctioned from the endotracheal tube. He had significant anasarca, but otherwise cardiovascular, pulmonary, and abdominal examinations were unremarkable.

Notable laboratory data on ICU transfer were as follows: WBC 3.8 10<sup>3</sup>/uL, hemoglobin 9.5 g/dL, BUN 56 mg/dL, and creatinine 1.69 mg/dL, which were all stable compared to prior. CT of the abdomen and pelvis performed during hospital day 17 to evaluate for small bowel obstruction incidentally revealed a thick-walled cavitory lesion in the left lower lobe, which was not present on chest CT obtained at the time of admission [Figure 1, Panels a and b]. Subsequent non-contrast-enhanced chest CT performed before the patient's cardiac arrest revealed a new air fluid level within the cavitory lesion [Figure 1, Panel c]. Bronchoscopy was notable for extensive blood clots admixed with thick mucus originating from the left lower lobe bronchus. CT pulmonary angiography demonstrated a 1.9 × 1.5 cm saccular contrast-enhancing lesion in the left lower lobe posterior basal segment, medial to the previously noted left lower lobe cavitory lesion [Figure 1, Panel d]. Cytologic findings from the bronchoalveolar lavage were consistent with *Mucor* and the patient was diagnosed with PAP secondary to PM [Figure 2].

He was treated with isavuconazonium for his underlying PM and underwent successful embolization of the PAP [Figure 3]. Isavuconazonium was chosen over Amphotericin B due to the patient's renal failure. No further hemothysis was noted.

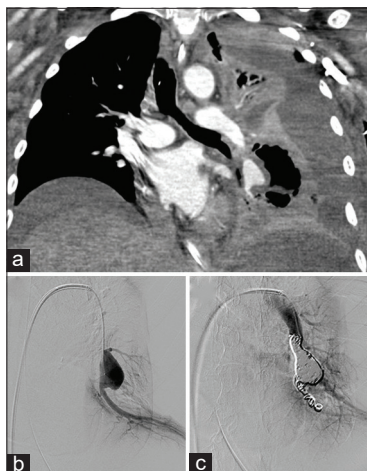


**Figure 1:** A 65-year-old man with diffuse large B-cell lymphoma of the liver who presented with failure to thrive and received inpatient chemotherapy. (a) Chest computer tomography (CT) lung window (1.5 mm thickness axial reconstructed image) from hospital day 1 showing unremarkable lung parenchyma; (b) CT abdomen lower lung window (3 mm thickness axial reconstructed image) from hospital day 17 demonstrating a 7.3 cm thick-walled cavitory lesion in the left lower lobe; (c) chest CT lung window (1.5 mm thickness axial reconstructed image) from hospital day 18 showing an air-fluid level within the thick-walled left lower lobe cavitory lesion; (d) CT pulmonary angiogram (1.5 mm thickness axial reconstructed image) from hospital day 24 with arrow showing a 1.9 × 1.5 cm saccular contrast-enhancing lesion in the left lower lobe posterior basal segment.



**Figure 2:** A 65-year-old man with diffuse large B-cell lymphoma of the liver who presented with failure to thrive and received inpatient chemotherapy. Bronchoalveolar lavage specimen demonstrating broad thin-walled non-septate hyphae.

The patient unfortunately suffered irreversible anoxic brain injury from his cardiac arrest and remained nonresponsive to physical or verbal stimuli. His renal failure worsened and given the lack of neurological recovery the patient's family elected to forgo hemodialysis or cardiopulmonary



**Figure 3:** A 65-year-old man with diffuse large B-cell lymphoma of the liver who presented with failure to thrive and received inpatient chemotherapy. (a) Computed tomography pulmonary angiogram (3 mm thickness coronal reconstructed image) showing the left inferior pulmonary artery pseudoaneurysm; (b) digital subtraction angiogram from a selectively catheterized left inferior pulmonary artery demonstrating the pseudoaneurysm; (c) post embolization digital subtraction angiogram demonstrating no flow to the inferior pulmonary artery pseudoaneurysm.

resuscitation. The patient subsequently passed away 1 month after his initial cardiac arrest.

## DISCUSSION

Acquired PAP is a rare cause of hemoptysis with a wide array of etiologies. PAP can be caused by complications from pulmonary artery catheters, chest tube insertion, and biopsies, as well as penetrating thoracic trauma (i.e., stabbing). Malignancies such as bronchogenic squamous cell carcinoma, primary sarcoma, and metastatic sarcoma have been reported to be associated with PAP as well. Chronic inflammation due to Behcet's disease and Takayasu arteritis has also resulted in PAP. However, infection is the most commonly identified cause of PAP, accounting for up to 33% of cases.<sup>[2]</sup> Infectious causes include septic emboli from tricuspid endocarditis, tuberculosis, syphilis, pyogenic bacteria, and various fungi. Historically, before the era of efficacious antibiotics, tuberculosis and syphilis were the most common infections associated with formation of PAP. An aneurysm arising from the pulmonary artery adjacent to or within a tuberculous cavity, also known as a Rasmussen's aneurysm, is reported in 5% of patients with tubercular cavities on autopsy.<sup>[3]</sup> More commonly seen infectious etiologies causing PAP today include pyogenic bacteria such as *S. aureus*, *S. pyogenes*, *Klebsiella*, and *Actinomyces* and fungi such as *Mucor*, *Aspergillus*, and *Candida*.

PAP ultimately arises because of inflammation of the vessel wall. In cases attributable to a tuberculous cavity or pyogenic

pneumonia, chronic inflammation leads to gradual weakening of the adjacent pulmonary arterial wall. The granulation tissue replaces the adventitia and media of the pulmonary arterial wall, which, in turn, is replaced by fibrin, leading to thinning, and pseudoaneurysm formation.<sup>[4]</sup> Successive endovascular seeding of the pulmonary artery lumen by septic emboli from tricuspid endocarditis can lead to pseudoaneurysm formation as well. The destruction of the vessel wall starts in the lumen and progresses to the outer wall. Some organisms such as *Mucor* can cause pseudoaneurysms by direct invasion of the vessel wall. PAP associated with malignancy is thought to be due to a similar mechanism through direct tumoral invasion and erosion of the vessel wall. The pseudoaneurysms tend to be solitary and found in the segmental or subsegmental branches. The presence of multiple pseudoaneurysms is commonly associated with endocarditis or metastatic disease to the lung.<sup>[2]</sup>

PM is the second most common form of Mucormycosis after the rhino-orbital-cerebral form and is typically only seen in immunocompromised patients. In fact, 80% of patients diagnosed with PM have an underlying malignancy, with the majority (71.4%) having a non-solid malignancy.<sup>[5]</sup> The genera in the order *Mucorales* are ubiquitous in nature and are usually found on decaying vegetation and in the soil.<sup>[6]</sup> Humans inhale the spores into the bronchioles and alveoli, leading to rapid progression of pneumonia. This can result in non-specific symptoms including fever, dyspnea, and cough, but vascular invasion of the organism can lead to hemoptysis. Clinical diagnosis is difficult, as definitive diagnosis often requires demonstration of the organism in tissue which often cannot be easily obtained; <50% of patients are diagnosed pre-mortem.<sup>[7]</sup> Bronchoalveolar lavage cytology can show characteristic broad non-septate hyphae with irregular branching, but the lack of septations leads to fragility of the specimen, further decreasing the diagnostic yield of bronchoscopy.<sup>[6]</sup> Common CT findings include pleural effusion (77%), nodules with a peripheral predominance (60%), consolidation (54.3%), ground glass (51.4%), and cavitation (37.1%).<sup>[5]</sup> The often cited reverse halo sign is actually seldomly seen (5.7%).<sup>[5]</sup> Ultimately, however, the overall mortality for PM remains 80% despite aggressive treatment.<sup>[5]</sup>

CT pulmonary angiography is not only the gold standard to establish a diagnosis of PAP but also assists in determining candidacy for potential endovascular intervention. Unfortunately, up to 46% of PAPs were overlooked by radiologists on the initial CT scans in one study, all of which were contrast-enhanced studies.<sup>[2]</sup> A clinical history of hemoptysis did not significantly alter the PAP detection rate on the initial CT study, further increasing the difficulty in making this diagnosis.<sup>[2]</sup> PAPs are frequently missed on CT studies without intravenous contrast enhancement as they may appear as endobronchial lesions or lung masses. Despite

contrast-enhancement, the diagnosis can be missed when radiologists focus on axial imaging alone, as sagittal and coronal multiplanar reformations are often very helpful in establishing the diagnosis.<sup>[2]</sup> Endovascular treatment modalities include direct coil embolization, stent placement, and embolization of the feeding vessel. Direct intra-saccular embolization with coils has the advantage of preserving pulmonary arteries distal to the PAP, preserving lung function, though carries an increased risk of PAP rupture.<sup>[8]</sup> Embolization of the vessels proximal to the PAP can be performed if the risk of rupture is high or intra-saccular embolization is not feasible. However, proximal embolization may be ineffective if a bronchopulmonary shunt is present and perfuses the aneurysm sac; it is thusly important to rule out such a shunt with a bronchial arteriogram.<sup>[8]</sup> Stent-assisted coil embolization is another option that allows for the preservation of flow distal to the aneurysm sac, but is best used for wide-necked aneurysms.<sup>[8]</sup> In one retrospective study, 12 patients underwent stenting or embolization of their PAP, and only one had immediate recurrent hemoptysis.<sup>[2]</sup> The risks of endovascular treatment of PAPs are no different from the risks of other endovascular embolization procedures throughout the body: Contrast-induced neuropathy, non-target embolization, arterial dissection, arterial thrombosis, and end-organ infarction. Operative repair is rarely required for PAP. Surgical interventions are typically reserved for cases of free pleural hemorrhage, unremitting hemoptysis, and infections likely to be refractory to medical therapy. Potential surgical interventions can include lobectomy, pneumonectomy, aneurysmectomy, and pulmonary artery arteriorrhaphy. Case reports describing management of PAP due to PM typically involve amphotericin B and surgical resection with varying results.<sup>[9-11]</sup>

## CONCLUSION

PAP and PM are individually uncommon clinical entities, but PAP due to underlying PM is a rare condition with a limited description in the literature. A high index of suspicion for both the clinician and radiologist is required and should be suspected in patients with underlying malignancy receiving immunosuppression whom develop hemoptysis. PAP can be successfully treated with embolization, but surgical resection should be considered in refractory infection.

## Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

## Conflicts of interest

There are no conflicts of interest.

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