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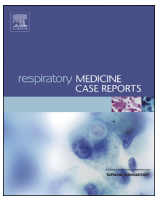

Case Report

Pulmonary artery pseudoaneurysm arising from primary lung neoplasm: A proposed mechanism

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ABSTRACT

Pulmonary artery pseudoaneurysms (PAPs) are rare and life-threatening occurrences. We present a 57-year-old male patient with squamous cell lung cancer, who presented with hemoptysis. Bronchoscopy did not reveal ongoing bleeding. Imaging showed a left lower lobe tumor, a cavitary lesion communicating with the bronchus, and a pulmonary artery pseudoaneurysm. Successful embolization of the originating segmental branch of the pulmonary artery was performed. The pathogenesis of PAPs associated with primary lung malignancies remains poorly understood. We propose a four-step mechanism involving primary tumor expansion, central cavitary necrosis, direct arterial invasion, inflammatory response, vessel wall damage, pseudoaneurysm formation, and subsequent filling of the former cavitary lesion. This case emphasizes the importance of considering PAPs in primary lung malignancies, particularly in male patients with squamous cell pathology. Understanding the proposed pathogenic mechanism could lead to early detection, prompt intervention, and improved outcomes.

1. Introduction

Pulmonary artery pseudoaneurysms (PAPs) are a rare but potentially life-threatening vascular condition characterized by an abnormal localized outpouching of the pulmonary artery. Unlike true aneurysms, PAPs affect only the external layers of the arterial wall, making them more prone to rupture due to the low resistance of the surrounding tissue [1]. The most common and alarming clinical presentations of PAPs is massive and life-threatening hemoptysis. The management of PAPs can be challenging, requiring a multidisciplinary approach that may involve surgical repair, endovascular techniques, or medical therapy.

The factors and mechanisms contributing to the development of PAPs in the context of primary lung malignancy are poorly understood, and there is a pressing need to explore these complexities. This case report and subsequent proposal for PAPs formation aim to address this gap in our understanding, offering insights into the pathogenesis of PAPs associated with primary lung malignancies, ultimately contributing to improved diagnostic and therapeutic strategies for this rare and challenging condition.

2. Case presentation

A 57-year-old man with a notable medical history of recent stage III squamous cell lung cancer affecting the left mainstem bronchus, presented to the Emergency Department with acute onset of hemoptysis and severe respiratory distress. The patient had

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previously completed chemoradiation therapy. He reported a significant volume of continuous, bright red blood, exceeding 50 cc, since awakening from a nap. He denied any prior history of hemoptysis, cough, fever, or chills preceding the sudden onset of his symptoms.

Upon initial evaluation, the patient exhibited signs of respiratory distress, with tachypnea (40 breaths per minute), sinus tachycardia at 144 beats per minute, and oxygen saturation as low as 79 % despite 100 % Fi02 via a non-rebreather mask. The physical examination revealed dried blood in the bilateral nares and oral cavity. The patient displayed increased work of breathing, severe subcostal and intercostal retractions, and diminished breath sounds throughout both lung bases.

Laboratory investigation indicated a baseline hemoglobin level of 10.9 g/dl, a normal platelet count and an INR of 1. Arterial blood gas analysis revealed respiratory acidosis, with a pH of 7.27, a pCO2 of 54, a pO2 of 94, and a bicarbonate level of 25. Although the patient was initially trialed on continuous positive pressure noninvasive ventilation, he was unable to tolerate the mask due to anxiety. During the patient's stay in the emergency room, he coughed an additional 40–60 cc of bright red blood, further exacerbating his critical condition.

Given ongoing hypoxemic hypoxic respiratory failure, the patient was intubated with a standard 8.0 endotracheal tube and oxygen saturations improved to 97 % on 100 % FiO2 that was quickly decreased to 30 %.Frank blood was visible from the endotracheal tube and an emergent bronchoscopy was performed to visualize the site of bleeding. Bronchoscopy showed blood originating from the left lung and the posterior wall of the distal left mainstem bronchus was necrotic with evidence of a cavitary lesion in the superior segment of the left lower lobe extending into the lung parenchyma through which a large vessel could be seen (Fig. 1A). No active bleeding was further visualized, and a 9-French endobronchial blocker was advanced into the proximal left mainstem bronchus under direct visualization for contingency inflation in the setting of a recurrent bleed. Following endobronchial blocker placement, no further hemoptysis recurred, and hemoglobin levels nadired at 7.4 g/dl.

Subsequent CT with arterial and venous phase contrast revealed a mass encasing the distal left mainstem bronchus and a cavitary lesion communicating with the left lower lobe bronchus. Within the cavitary lesion, a contrast-enhancing segmental branch lesion of the left lower lobe pulmonary artery could be identified without evidence of contrast extravasation, consistent with a pulmonary artery pseudoaneurysm (Fig. 1B).

Patient underwent left main pulmonary artery angiography with interventional radiology demonstrating a segmental left lower lobe pulmonary artery pseudoaneurysm without extravasation (Fig. 1C). A 5-French catheter was used to select the contributing inferior and posterior pulmonary artery branches, at which point a high flow microcatheter was advanced and the segmental branch from which the pseudoaneurysm arose was embolized using platinum coils (Ruby, Penumbra, Alameda, United States). A persistent communication to the pseudoaneurysm was seen and a mesh plug (Amplatzer, Abbott, Chicago, United States) was deployed to embolize the remaining proximal branch. Repeat arteriography demonstrated no further filling of the pseudoaneurysm and preservation of the nearby segmental artery (Fig. 1D).

Unfortunately, one month after discharge the patient suffered a cardiac arrest in the setting of recurrent massive hemoptysis and died after a brief hospitalization.

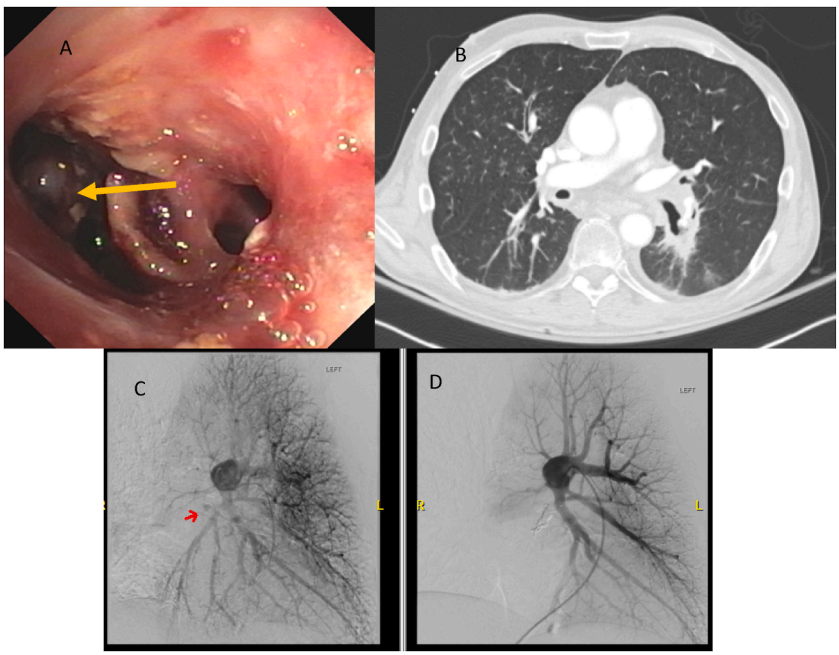

Fig. 1. (A) Bronchoscopic image of cavitary lesion in left lower lobe superior segment, visible pulmonary artery pseudoaneurysm (arrow). **(B)** Contrast enhancing pulmonary artery pseudoaneurysm within cavitary lesion seen on CT chest angiogram with IV contrast. **(C)** Segmental left lower lobe pseudoaneurysm without extravasation **(D)** Successful embolization of segmental lower lobe branch.

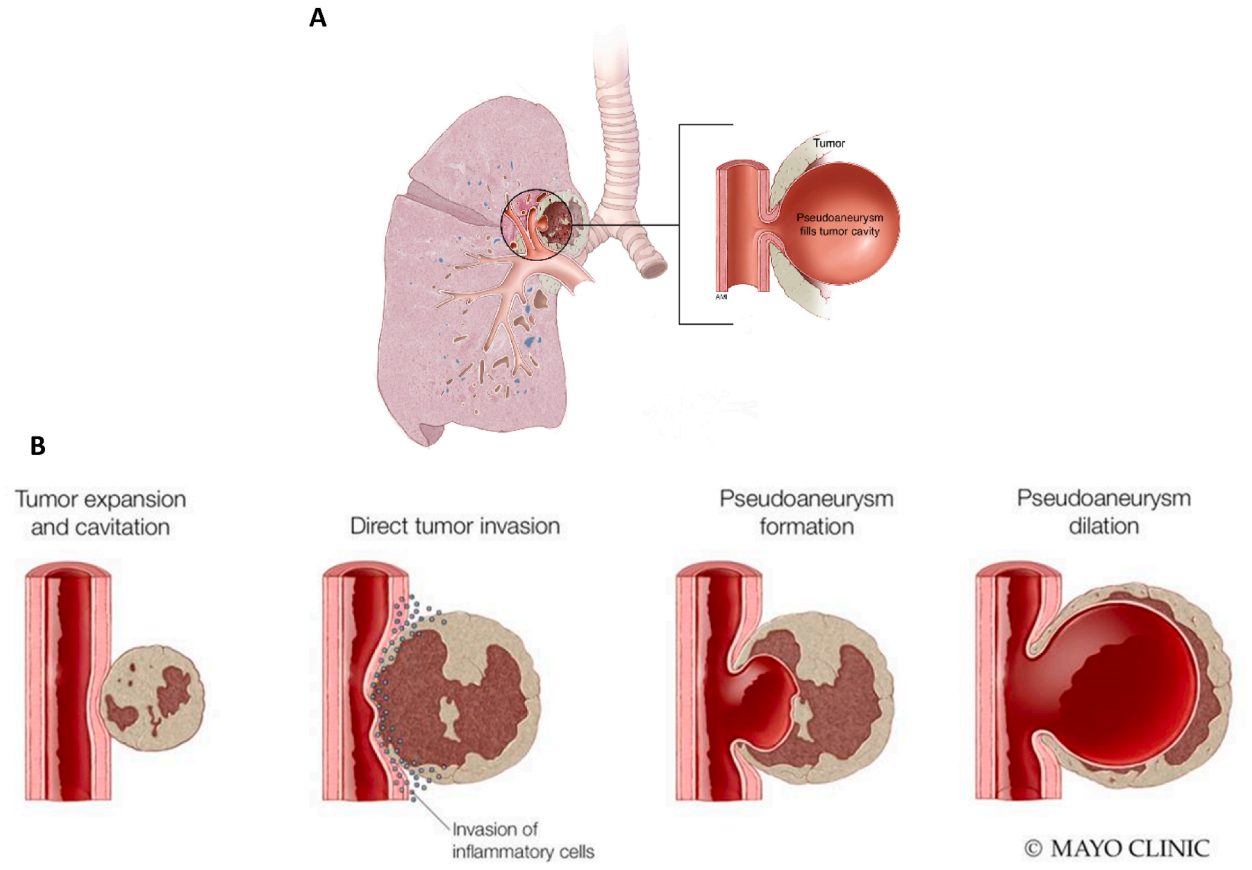

Fig. 2. **(A)** Typical presentation of PAP in setting of primary lung neoplasm. **(B)** Proposed mechanism of PAP formation in the setting of a primary lung malignancy.

3. Discussion

Pulmonary artery pseudoaneurysms (PAPs) are a rare source of life-threatening hemoptysis and airway compromise. As opposed to a true aneurysm, a pseudoaneurysm has a higher risk of rupture as it involves distortion of two layers of the vascular wall instead of all three leading to a focal dilatation of the pulmonary artery segment. Rupture of the weakened vascular vessel carries a mortality rate upwards of 50 % leading to intrapulmonary hemorrhage and subsequent asphyxiation [1,2].

Historically, PAPs are known complications of infections, pulmonary hypertension, collagen and vascular disorders, pulmonary embolism, trauma, and iatrogenic catheter misplacement [1,3,4]. In a retrospective review of 24 PAPs identified at a large academic medical center, PAPs were most frequently associated with infections (33 %). Trauma was identified as the second most common cause (17 %), both from penetrating injuries and iatrogenicity. PAPs associated with neoplasm represented the third most common unique category (13 %) while pulmonary embolism, traction bronchiectasis and pulmonary fibrosis, and PAPs of idiopathic origin accounted for the remaining cases [5].

PAPs associated with primary lung neoplasms remain rare, and the pathogenesis, risk factors, and presentation of PAPs in this setting are poorly understood. However, among the few cases described in literature, there is a clear association between the male gender and formation of PAPs in primary lung malignancy. Squamous cell carcinoma with central cavitation of the primary tumor complicated by arterial invasion is the most frequently described primary lung malignancy associated with PAP formation [6]. PAP presentation is typically 4–6 months after initial diagnosis of malignancy with massive hemoptysis as its primary symptom [4,6–8].

The pathogenesis of PAPs associated with infection has been previously characterized, the formation of PAPs in primary lung cancers is poorly understood. In PAPs associated with tuberculosis and with infectious pneumonia, contiguous spread of the pathogen along vasculature leads to tissue destruction from the outer vessel wall towards the lumen. This damage then predisposes the involved vessel wall to pseudoaneurysm formation [1–3,5]. PAP pathogenesis in the setting of primary lung malignancy has been hypothesized to occur by a similar mechanism. Direct tumor invasion of the vessel wall and subsequent erosion of the vascular wall seems to be a prerequisite to PAP development [1,6].

Based on prior PAPs reported in the literature as well as our own case, we propose the following mechanism for PAP formation in the setting of primary lung neoplasm: (1) primary tumor expansion with central cavitary necrosis (2) direct arterial invasion with resultant inflammatory response and damage to the vessel wall (3) altered vascular flow and increased intraluminal pressure (4) pseudoaneurysm formation (5) pseudoaneurysmal dilatation, often with filling of the former cavitary lesion (Fig. 2).

PAPs may cause life threatening bleeding via massive hemoptysis which can occur with the initial vascular injury or with pseudoaneurysmal dilatation. On presentation, the management priority remains maintenance of airway patency and hemodynamic resuscitation. PAPs are an unusual cause of massive hemoptysis, but if identified, the standard of care is minimally invasive intravascular coiling and embolization of the pseudoaneurysm or feeder vessels, as described in our case [5]. Other techniques such as aneurysmectomy, lobectomy and pneumonectomy are less commonly performed and may lead to increased mortality related to secondary infections [6,9].

4. Conclusions

PAPs due to primary lung malignancy resulting in massive hemoptysis are uncommon but are a high-risk clinical scenario with poor outcomes and a high mortality rate. We propose a possible mechanism for the pathophysiology of PAPs in primary lung malignancy similar to described PAP development in the setting of infection with prerequisite direct vascular invasion and pseudoaneurysmal dilation accompanying cavitary necrosis of the tumor mass. While uncommon, described cases often present as an early complication of aggressive, cavitary lung neoplasms, typically squamous cell carcinoma in male patients.

Key points

- Pulmonary artery pseudoaneurysms have low incidence but high mortality.
- The formation of PAPs in the setting of primary lung neoplasm is likely due to direct tumor invasion of the vessel and an increased inflammatory response leading to altered vascular flow and subsequent pseudoaneurysmal dilatation.
- A multidisciplinary approach in management with interventional pulmonology and interventional radiology is necessary.

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Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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