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Massive Hemoptysis in Severe Idiopathic Pulmonary Hypertension- A Rare Presentation

Kanishka Salunkhe[1], Jolsana Augustine[2*], Vidhyapriya Sellamuthu[3], Jamalul Azizi Bin Abdul Rahaman[4], Fatimah Azmah Mohammad[5], Haizal Bin Haron Kamar[6] and Mohd Redzuan Bin Ismail[7]

[1]Kanishka Salunkhe, Fellow in Interventional Pulmonology, Department of Pulmonology, Sultan Idris Shah Serdang Hospital, Selangor Darul Ehsan, Malaysia

[2]Jolsana Augustine, Fellow in Interventional Pulmonology, Department of Pulmonology, Sultan Idris Shah Serdang Hospital, Selangor Darul Ehsan, Malaysia

[3]Vidhyapriya Sellamuthu, Fellow in Interventional Pulmonology, Department of Pulmonology, Sultan Idris Shah Serdang Hospital, Selangor Darul Ehsan, Malaysia

[4]Jamalul Azizi Bin Abdul Rahaman, Consultant Pulmonologist and Interventional Pulmonologist, Thomson Hospital, Kota Damansara, Selangor Darul Ehsan, Malaysia

[5]Fatimah Azmah Mohammad, Consultant Pulmonologist and Expert in Pulmonary Hypertension, Sultan Idris Shah Serdang Hospital, Selangor Darul Ehsan, Malaysia

[6]Haizal Bin Haron Kamar, Consultant cardiologist, Thomson Hospital, Kota Damansara, Selangor Darul Ehsan, Malaysia

[7]Mohd Redzuan Bin Ismail, Consultant interventional Radiologist, Thomson Hospital Kota Damansara, Selangor Darul Ehsan, Malaysia

ABSTRACT

Hemoptysis is not an uncommon presentation of a disease in a pulmonologist's usual practice. The significance of hemoptysis comes from the fact that even a minimal amount of intrabronchial bleeding can lead to life-threatening airway compromise and escalate the mortality risk. Infections such as tuberculosis and other bacterial or fungal parenchymal infectious causes are often cited as major culprits in hemoptysis cases. We present an unusual case of a young gentleman with no prior comorbidities who initially presented with nasal bleeding and over a span of 2 weeks experienced two major episodes of hemoptysis. He underwent multiple imaging studies and bronchoscopic evaluations but was eventually found to have elevated pulmonary pressures on echocardiogram and a pattern of right heart strain on electrocardiogram. These findings shed light on a rare presentation of idiopathic pulmonary hypertension being the cause of massive hemoptysis. This case was an eye-opener for many reasons including missing important cardiac examination findings during the clinical evaluation and the presence of a dilated pulmonary trunk in contrast CT images ultimately unravelling one of the rarest causes of massive hemoptysis.

***Corresponding author**

Jolsana Augustine DNB, Fellow in Interventional Pulmonology, Department of Pulmonology, Sultan Idris Shah Serdang Hospital Selangor Darul Ehsan, Malaysia.

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Introduction

Hemoptysis is not a rare presentation in pulmonary practice. Most cases of hemoptysis are mild and self-limiting. Severe or massive hemoptysis is defined as any amount of bleeding that can jeopardize the airway or lead to cardiovascular compromise. Life-threatening hemoptysis can occur with even 100 ml of blood rapidly clogging the airway resulting in airway compromise and carrying high mortality rates. The usual causes of hemoptysis range from acute lower respiratory infections, chronic obstructive pulmonary disease and malignancies to bleeding disorders, and diseases that cause structural damage to airways such as bronchiectasis. immunologic diseases like Goodpasture syndrome, systemic lupus erythematosus (SLE), granulomatosis with polyangiitis, microscopic polyarteritis and vascular anomalies such as aneurysms and arteriovenous malformations need to be ruled out [1]. The diagnostic workup and management of massive hemoptysis can often be challenging. In around 40% of cases, the etiology remains unknown even after detailed evaluation with CT scans and bronchoscopy making it an idiopathic or cryptogenic hemoptysis [2].

It is important to understand the vascular supply of lungs. Identifying the source of bleeding is vital for management. Most often, the cause of a massive severe bleed is from the bronchial arteries as it is a high-pressure systemic circulation. Bleeding from the pulmonary arteries is noted in only 5% of cases [3]. We present an extremely rare case of massive hemoptysis in a young man with no comorbidities with massive hemoptysis and nasal bleeding as the sole symptoms.

Case Report

A 24-year-old man presented to a general practitioner's (GP) clinic complaining of a cough for 2 weeks and nosebleeds for the last 7 days. He was a reformed smoker and reported having occasional episodes of nosebleeds since his school days. He had no other known medical illnesses and had never experienced similar

issues or had any hospitalizations. He denied any breathlessness or limitations in his daily activities due to chest pain or fatigue. He had no seasonal cough or allergic symptoms. He smoked 4-5 cigarettes daily from the age of 15 until he stopped at 19. He denied any substance abuse or past history of bleeding from other sites such as coughing up blood or blood in stools. His family history was unknown as he was adopted in early childhood. The GP suspected pulmonary tuberculosis and referred him to our center for further investigation. In light of the nosebleeds, he underwent naso-laryngoscopy and the bleeding was arrested with nasal packing. Within a few hours, he experienced 3 episodes of hemoptysis (around 150 ml) and reported feeling 'something in the chest' and hence he was admitted to the ICU.

On general examination he appeared cachectic with a body mass index of 16.4kg/ m2. During clinical examination, he was afebrile, tachypneic with a respiratory rate 26 breaths per minute, tachycardic with a pulse rate of 118 bpm and normotensive with a blood pressure of 100/76 mm Hg. The rest of the physical examination was normal, except for chest auscultation which revealed bibasilar crepitations. His oxygen saturation on room air was 88%.

His full blood count showed hemoglobin 12.2 g/dL, white cell count 12640/mm3, platelets 164/mm3. His blood picture was microcytic and hyperchromic. His biochemical profile was normal and his renal, liver profile, and clotting parameters were with normal limits. His viral markers were negative.

He was evaluated with a chest x-ray, a computed tomography (CT) of the chest with contrast and bronchoscopy on an emergent basis to investigate the cause of massive hemoptysis. The chest X-ray showed right lower zone consolidation. (Figure1) A contrast CT chest showed the presence of consolidative changes in the posterior basal segment, associated with air bronchograms and a tree in bud pattern. (Figure 2a, 2b). Pulmonary tuberculosis (TB)was suspected to be the initial differential diagnosis, given a cachectic young patient with no significant prior medical history presenting with hemoptysis. The right lower lobe segmental consolidation in the X-ray and subsequent CT scan of the thorax strengthened the suspicion of an active pulmonary infection. Flexible bronchoscopic evaluation of the lower airways demonstrated organized clots in the right lower lobe bronchus (Figure 3) which were successfully removed with a cryoprobe (ERBECRYO single use cryoprobe 1.7mm) restoring luminal patency. No endobronchial lesions were observed (Figure 4). Bronchoalveolar lavage (BAL) samples were obtained for microbial evaluation particularly to rule out tuberculosis (GeneXpert and liquid culture) and for cytological evaluation. He was observed for another 5 days with no recurrence of bleeding and was subsequently discharged with an oral antibiotic and advice to follow up for BAL reports.

He presented to the emergency department 10 days later with another episode of massive hemoptysis and was readmitted to our HDU. His TB evaluation was negative and the BAL reports showed no evidence of infection. Given the recurrent massive hemoptysis, an emergency bronchial artery embolization was warranted for the bleeding vessel (Figures 5 and 6). He reported chest discomfort post-embolization, a bedside 12-lead ECG was performed showing sinus rhythm, right ventricular hypertrophy with strain, deep T wave inversion in leads V1 to V4 and ST depression in leads II, III and aVF (Figure 7). An urgent CT pulmonary angiography was conducted to rule out pulmonary embolism, which was ruled out but pulmonary angiography revealed a significantly dilated pulmonary trunk (Figure 8). A bedside echocardiogram performed subsequently showed a very dilated, hypokinetic right ventricle, a D-shaped interventricular septum, preserved left ventricular ejection fraction of 63.1%, tricuspid regurgitation with regurgitant velocity of 3.78 m/s, and an estimated PAP of 76 mmHg. Upon further probing, he admitted to feeling tired after routine daily activities which he attributed due to his malnourishment. He still denied any severe breathlessness that prevented him from performing his usual chores. Furthermore, a detailed cardiological examination following the echocardiogram report revealed loud P2 sounds and pansystolic murmur along the left sternal border which had been overlooked during the initial clinical evaluation. Detailed immunological work up was conducted to rule out any connective tissue disease, which returned negative. Genetic studies could not be performed due to limited access and financial constraints.

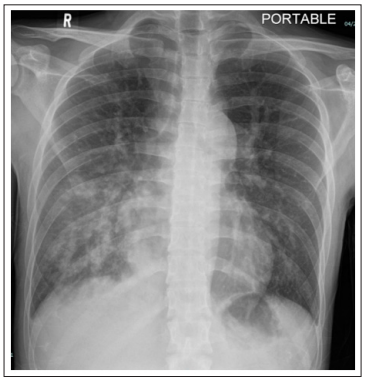

Figure 1: Initial Chest X-Ray Showing a Right Lower Zone Hazy Opacity

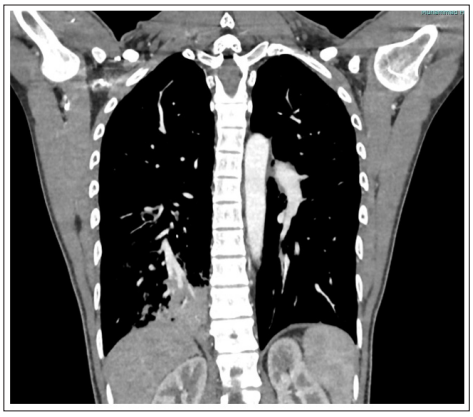

Figure 2a: Initial CT scan (Coronal Image) of the Chest Showing a Consolidation in the Right Lower Lobe

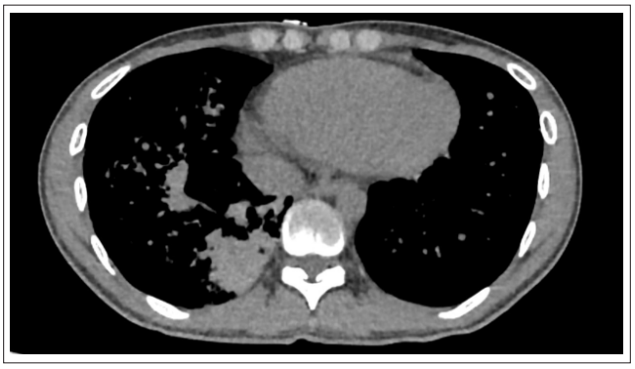

Figure 2b: Initial CT scan (Axial Image) of the Chest Showing a Consolidation in the Right Lower Lobe

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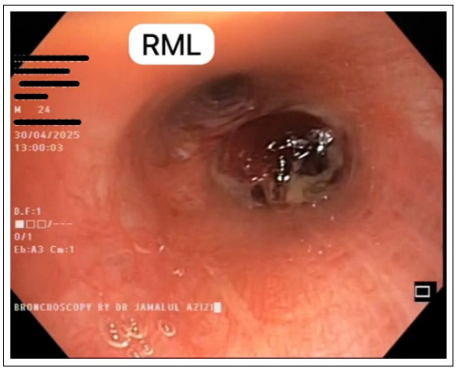

Figure 3: Bronchoscopy on presentation. Right Lower Lobe Bronchus is Completely Occluded by a Blood Clot

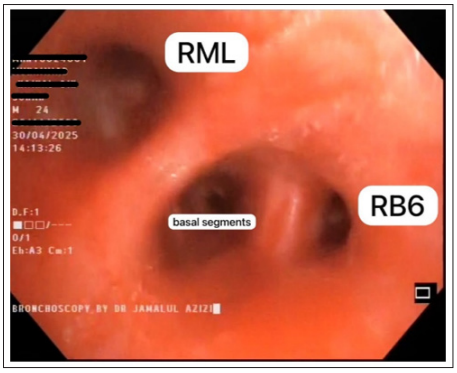

Figure 4: Post Cryoextraction of the Clot, Right Lower Lobe Bronchus is Patent

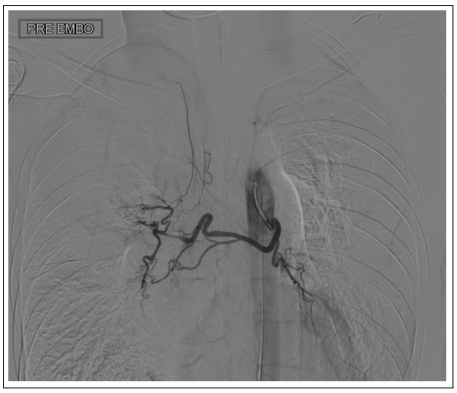

Figure 5: Pre-Bronchial Artery Embolisation (BAE): Common Bronchial Trunk Arteriogram Demonstrating Prominent and Tortuous Right Bronchial Artery

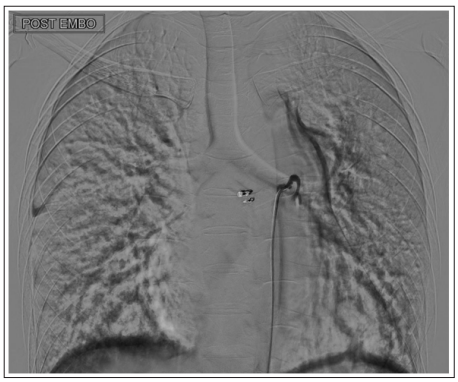

Figure 6: Post BAE Angiogram Showing Complete Cessation of Flow into the Right Bronchial Artery

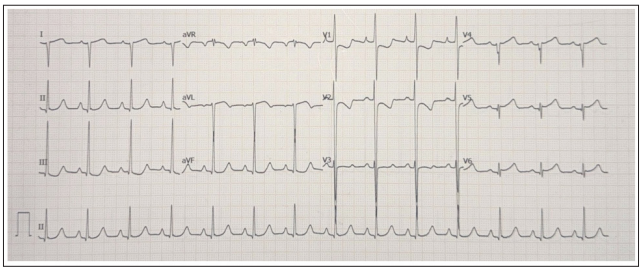

Figure 7: 12-Lead ECG After Chest Pain Following BAE

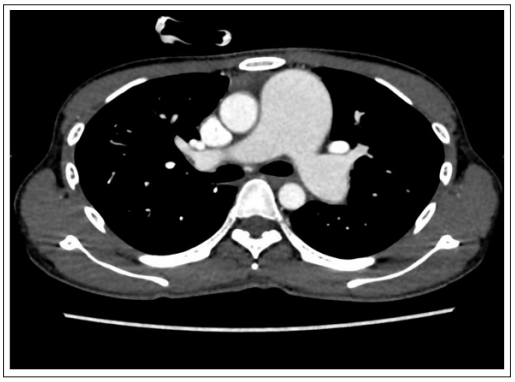

Figure 8: CT Pulmonary Angiogram Image Showing a Very Dilated Pulmonary Artery Trunk. Aortopulmonary Ratio >1:1

After 3 days the patient recovered well. A right heart catheterization (RHC) study was performed confirming the presence of Pulmonary Artery Hypertension (PAH) with pressures in the right ventricle 110/16 mmHg and in the main pulmonary artery of 129/62 mmHg without any intracardiac shunts. This case was an eye opener because this patient had never reported dyspnea or limitation in his routine physical activities aside from cachexia and bleeding manifestations. He has been referred to a center of excellence in pulmonary hypertension, Sultan Idris Shah Serdang Hospital for further work up and management. The patient is currently prescribed sildenafil 25 mg every 8 hours orally, whilst awaiting further evaluation for lung transplantation by the multidisciplinary team. He is under close follow up in the pulmonary hypertension clinic.

Discussion

In cases of hemoptysis, a basic hematologic work up and clotting parameters and chest radiography are routinely performed. CT Chest with contrast or CT angiography with contrast is highly sensitive in identifying the source of bleeding [3]. In cases of recurrent hemoptysis where bleeding recurs after initially successful cessation, bronchial artery embolization is suggested as per the The American College of Radiology Appropriateness Criteria [4]. Bronchoscopy is indicated for both diagnostic and therapeutic reasons being particularly useful in identifying the bleeding site, evacuating clots and employing several interventional techniques in arresting intrabronchial active bleeding such as instillation of cold saline, balloon tamponade, bronchial blockers, argon plasma coagulation or electrocautery. Bronchoscopy is also beneficial in identifying intrabronchial tumors or inflammatory conditions and obtaining samples for microbiological and cytological evaluation [4].

Pulmonary Arterial Hypertension is a heterogenous and rare disease, defined by a mean Pulmonary Arterial Pressure (mPAP) ≥20 mmHg as measured by right heart catheterization (RHC), with normal pulmonary artery wedge pressure ≤15 mmHg and elevated

pulmonary vascular resistance ≥3 Wood units [5]. Hemoptysis is a rare but well documented complication in PAH. Spontaneous bleeding manifestations such as dental bleeds, epistaxis and menstrual bleeding can occur in PAH associated with congenital heart disease, although they are usually mild and self-limiting. The pathophysiology of hemoptysis in cases of PAH is complex and multifactorial. Changes in pulmonary microvascularity, obliterative remodelling of the pulmonary vascular bed along with endothelial damage, vasoconstriction and thrombosis have been implicated [6]. Neoangiogenesis in response to the inflammatory process might induce the development of collaterals and aneurysms between the high pressure systemic bronchial circulation and low-pressure pulmonary circulation potentially leading to significant bleeding [6].

A case report published by Zylkowska et al described a 27-year-old woman suffering from recurrent hemoptysis while already on treatment for Idiopathic PAH (IPAH); she died despite multiple BAE [7]. In another study involving a total of 4 patients diagnosed with PAH on prostacyclin analogues, BAE was performed multiple times to halt hemoptysis [8].

Tio et al studied the factors leading to hemoptysis in idiopathic and hereditary PAH in 129 patients; showing that hemoptysis was more frequent in subjects with bronchial arterial hypertrophy, and associated with faster disease progression [9].

In our patient, the initial presentation was misinterpreted as pseudo hemoptysis secondary to nasal bleeding at the local health centre. The first CT chest with contrast performed at our centre showed a dilated pulmonary trunk which was initially missed. Furthermore, clinical examination overlooked key findings such as loud second heart sounds and a pansystolic murmur. Performing an echocardiogram at this initial stage could have identified severe pulmonary hypertension and prompting detailed cardiological work up at the first instance. Therapeutic bronchoscopy was crucial in resolving airway obstruction by removing intrabronchial clots. Due diligence is essential in every case of hemoptysis as it can prove life threatening at any point. Patients with IPAH experiencing recurrent hemoptysis should be evaluated for lung transplantation as early as possible.

Conclusion

In conclusion, hemoptysis in PAH is rare. Hemoptysis in IPAH can be effectively managed by a multidisciplinary, tailored decision-making approach in well-equipped centers. 2 D Echocardiogram should be conducted in all cases of unexplained hemoptysis especially when it is severe. A low threshold for performing CT pulmonary angiogram with contrast studies in such patients must be maintained. BAE combined with bronchoscopic management remains the safest approach to managing hemoptysis in an acute setting, facilitating the time to lung transplant as the preferred treatment option. Although BAE can be attempted multiple times alongside bronchoscopic evaluation to halt hemoptysis, further definitive treatment modalities are necessary to modify the disease and improve the quality of life.

Acknowledgements in Managing the Case

Dr Elang Kumaran a/l Krishnan
Consultant ENT, Head & Neck surgeon, Thomson Hospital Kota Damansara, Selangor Darul Ehsan, Malaysia

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