Massive hemoptysis in a patient with Eisenmenger syndrome, polysplenia, and transverse liver

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October 10, 2024

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Abstract

Introduction : Hemoptysis is defined as blood-streaked sputum from the lower parts of the respiratory tract. Hemoptysis, even in small amounts, is a frightening and an alarm sign for possible underlying conditions such as infections, pulmonary diseases, neoplastic conditions, cardiovascular alterations, vasculitis, traumatic events, hematological derangements, and iatrogenic or drug-induced events. The initial step in the evaluation of hemoptysis is to determine the source of bleeding.

Case presentation : Herein, we report an unusual case of massive hemoptysis in a young patient with polysplenia and Pulmonary Artery Hypertension (PAH) in the setting of Eisenmenger syndrome. Chest radiography was suggestive of multiple lung opacities bilaterally. Chest Computed Tomography (CT) revealed

a non-heterogeneous mass-like lesion measuring 4 × 5.2 × 5.6 cm in the superior segment of the inferior lobe of the left lung, concerning for accessory spleen. The patient underwent Video bronchoscopy, which showed tracheomalacia and active bleeding in the left main bronchus. The bleeding was controlled by Argon Plasma Coagulation (APC) technique. Broncho-alveolar lavage (BAL) was negative for acid-fast bacilli on staining and on culture. After stabilization, the patient was discharged home on medical management for PAH. On two-week follow-up, imaging revealed resolution of the pulmonary mass-like lesion.

Conclusion : Our report highlights the importance of bronchoscopy in determining the bleeding source in patients with hemoptysis and managing it via the APC technique.

Keywords: Hemoptysis; Pulmonary Hypertension; Eisenmenger syndrome; Polysplenia; Transverse liver

INTRODUCTION

Hemoptysis is defined as blood-streaked sputum from the lower parts of the respiratory tract. The initial step in the evaluation of hemoptysis is to determine the source of bleeding. Pseudo hemoptysis, which refers to blood expectoration via a source other than the bronchial or pulmonary system, should be ruled out by history and physical examination (1).

Hemoptysis, even in small amounts, is a frightening and alarm sign. The clinical spectrum ranges from minor blood-stained sputum to major bleeding causing respiratory failure and hemodynamic instability. Underlying causes may vary from benign self-limiting conditions to severe and potentially life-threatening conditions. In terms of severity, hemoptysis is considered scant when presents with bleeding <5 mL, mild when <20 mL, and moderate when >20 ml, while massive hemoptysis is defined as bleeding amount of 100 mL/24 hours or more (2).

Hemoptysis may derive from multiple underlying conditions, such as infections, pulmonary diseases, neoplastic conditions, cardiovascular alterations, vasculitis, traumatic events, hematological derangements, and iatrogenic or drug-induced events (Table 1) (3).

In adults, acute respiratory tract infections (e.g., bronchitis, pneumonia), bronchiectasis, asthma, chronic obstructive pulmonary disease, and malignancy are the most common etiologies. Tuberculosis (TB) is a major cause of hemoptysis in endemic regions and in developing countries (4). However, in industrialized areas, bronchial carcinoma and bronchiectasis are more common culprits.

Pulmonary artery hypertension (PAH) is a serious condition causing progressive obstruction and obliteration of the pulmonary vascular bed. PAH is a rare cause of hemoptysis, which is responsible for 0.2- 4% of the cases. Although, hemoptysis is a relatively more common finding in patients with Eisenmenger syndrome (1).

In the past, depending on the severity and etiology of hemoptysis, several management strategies were recommended including supportive care, surgical resection, and lung transplant. Currently, the more commonly used strategy is bronchial artery embolization (BAE). In this technique, a particulate material is injected into angiographically identified abnormal bronchial arteries, helping in hemostasis. BAE is usually well tolerated; however recurrent bleeding is commonly associated with the procedure (6).

Herein, we report a case of hemoptysis in a patient with Eisenmenger syndrome and polysplenia. We aim to portray the importance of bronchoscopy as both a diagnostic and therapeutic tool, which helped us to identify the source of bleeding and manage it via the BAE technique.

CASE DESCRIPTION

History

A 34-year-old female with past medical history of Eisenmenger syndrome in the setting of Ventricular Septal Defect (VSD), who presented with 10 days' history of cough, hemoptysis (about 300 ml/day), pleuritic chest pain, and exertional dyspnea. She did not report any fever, gum bleeding, or epistaxis.

Examination

On physical examination the patient was tachypneic, tachycardic, and hypoxic with oxygen saturation of 85% in room air. Chest auscultation revealed left upper lobe crackles and holosystolic murmur in the lower sternal border. Also, she was found to have clubbing in her hands bilaterally.

METHODS (DIFFERENTIAL DIAGNOSIS, INVESTIGATIONS, AND TREATMENT)

Laboratory findings were significant for microcytic anemia (Hemoglobin 10.6 gm/dl), elevated erythrocyte sedimentation rate (ESR) at 23 mm/hr., and negative sputum for both Acid Fast Bacilli (AFB) stain and geneXpert. Chest radiograph revealed non-homogenous opacities in both lungs (figure 1). Computed tomography (CT) Chest showed a non-heterogeneous mass like lesion measuring $4 \times 5.2 \times 5.6$ cm in superior segment of inferior lobe of the left lung (figure 2), in addition to a transversus liver and multiple spleens were noted (Figure 3). Echocardiography was remarkable for mild Pulmonary valve insufficiency, Pulmonary artery pressure (PAP) of 80 mmHg, abnormal septal motion, large membranous VSD, severe right ventricular (RV) enlargement, and left ventricular Ejection fraction (LVEF) of 55%.

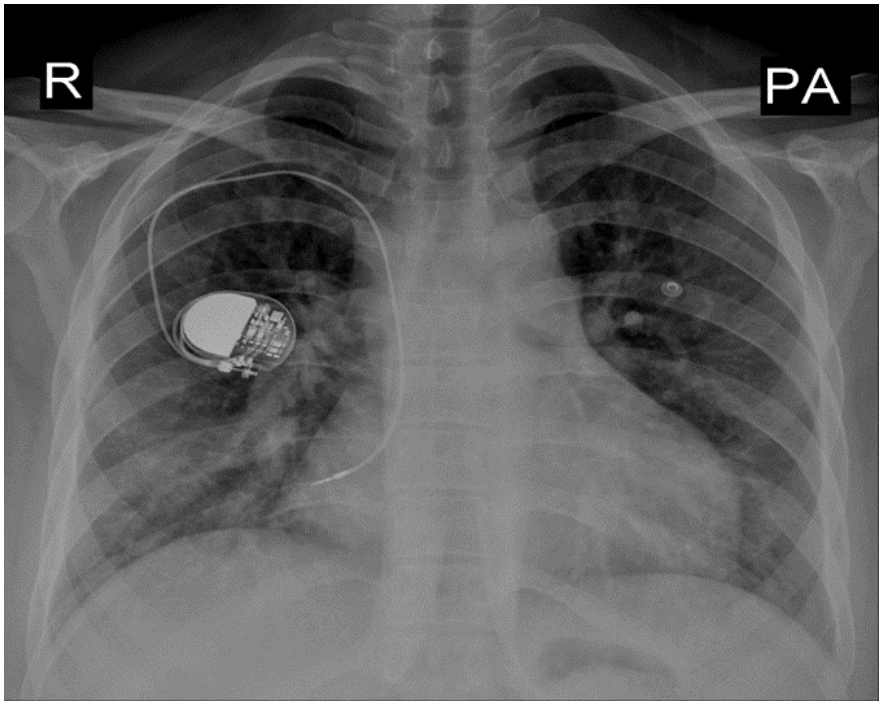
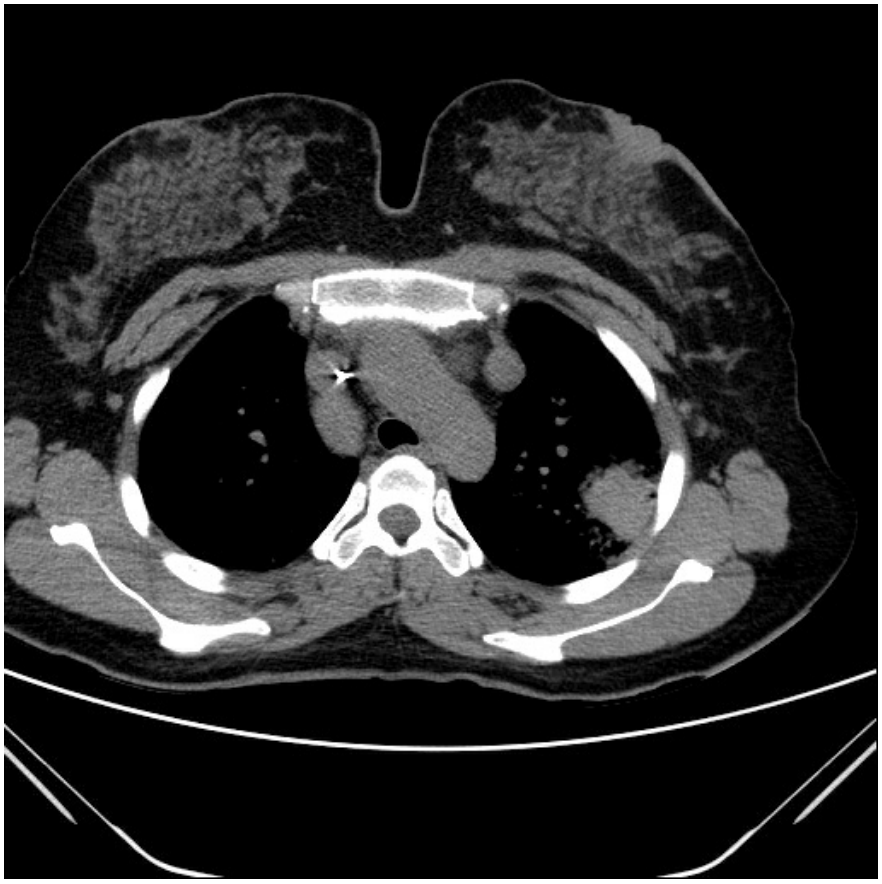


Fig. 1. Chest X-ray PA view revealed non homogenous opacity in both lungs.

To identify the source of bleeding, Video-bronchoscope was performed, which revealed tracheomalacia and active bleeding in the left main bronchus, which was controlled by APC (figure 4). Bronchoalveolar lavage (BAL) was sent for cytology, anaerobic culture, Ziehl-Neelsen stain, and GeneXpert to detect Mycobacterium TB, all of which were unremarkable.



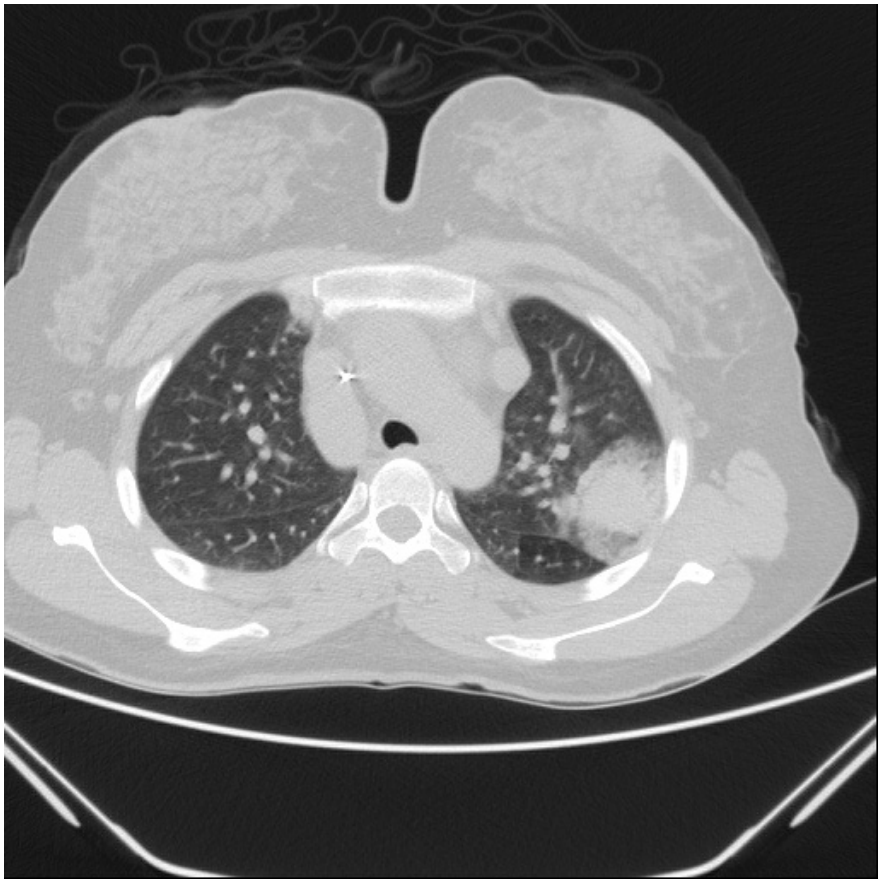


Fig. 2. Chest CT showed non heterogeneous mass like lesion measuring 4 × 5.2 × 5.6cm in superior segment of inferior lobe of left lung.

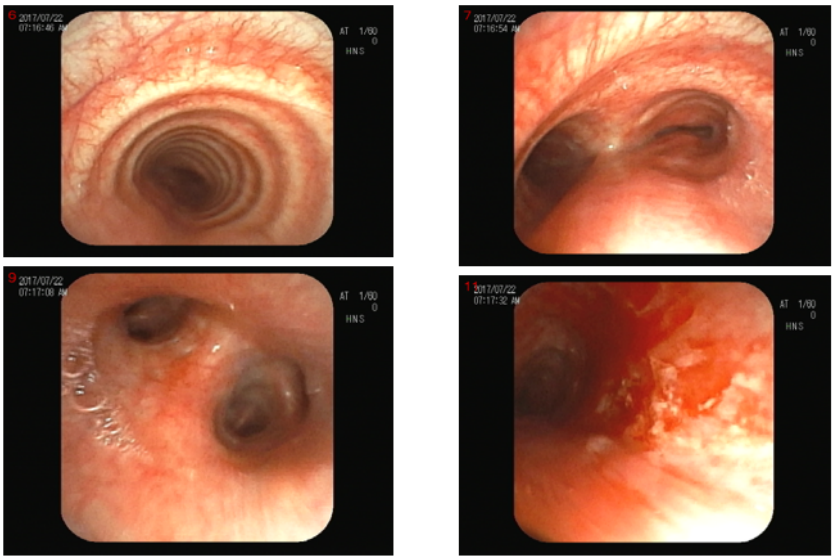


Fig. 3. Bronchoscopic findings revealed hemorrhagic lesion in left upper segments of inferior lobe.

CONCLUSION AND RESULTS

After stabilization, the patient was discharged on maintenance medications to control PAH e.g., diuretics, and was advised to follow up regularly with her pulmonologist. On a two-week follow-up, the patient remained asymptomatic. Lung examination was clear bilaterally and repeat CXR was consistent with the resolution of the lung opacities.

Discussion

Hemoptysis is a serious complication of PAH that is rarely reported in end-stage patients. The incidence of hemoptysis in PAH patients remains uncertain (7). In 75% of the patients, CT chest shows an evidence of bronchial artery hypertrophy. The number of dilated bronchial arteries correlates with the severity of PAH. The presence of hypertrophied bronchial artery increases the risk of hemoptysis in PAH patients (8).

Diagnosis of pulmonary hypertension can be established by Right Heart Catheterization (RHC) and revealing elevated mean pulmonary arterial pressure (mean PAP) [?]25 mmHg at rest. Recommended pharmacologic treatment for PAH includes vasodilators, prostanoids, nitric oxide, phosphodiesterase inhibitors, endothelin receptor antagonists, and anti-coagulants treatment (9). Bronchial artery embolization is suggested as an immediate emergency procedure in patients with PAH and severe hemoptysis or as an elective intervention in patients with frequent episodes of mild-to-moderate hemoptysis. Anticoagulant therapy in patients with PAH and hemoptysis should be postponed. Interventional Bronchoscopy and APC are useful tools to control the bleeding. APC is an electrosurgical technique similar to laser or electrocautery, which is used during bronchoscopy procedures to ablate malignant airway tumors, control hemoptysis, remove granulation tissue from stents or anastomoses, and treat a variety of benign disorders (10).

Our case is the first report of hemoptysis in a patient with PAH and multiple spleens. Nugraha et al in 2021 reported a 25-year-old woman with past history of PAH that presented with hemoptysis (400ml) and shortness of breath. Her examination revealed jugular venous dilatation, right ventricular heaving, accentuated 2nd heart sound, a grade 3/6 pansystolic murmur with best heard at the left lower sternal border. Further workups showed elevated PAP at 98 mmHg. A non-reactive O2 test, observation of hemoptysis suspected to be associated with pulmonary hypertension, suspected Hospital Acquired Pneumonia (HAP), and hypokalemia. The treatments included sildenafil, digoxin, furosemide, iloprost nebulizer, Aspar K, ceftazidime and ciprofloxacin (9).

Our patient had an atypical presentation of hemoptysis and pleuritic chest pain, and it was challenging to pinpoint the cause bleeding, especially in the setting of concomitant polysplenia. Hemoptysis is usually an end-stage presentation of PAH and has a poor prognosis. Given the severity of this condition, prompt and aggressive management is crucial to achieve the best outcome. Take away point from our report is to highlight the importance of bronchoscopy in determining the bleeding source in patients with hemoptysis and to manage the bleeding in a timely manner via the APC technique.

AUTHOR CONTRIBUTIONS

MS, AN managed the patient, MS, EP and BD drafted the paper, SB, AN and MS finalized the paper. All authors read and approved final version of the paper.

ACKNOWLEDGMENTS

The authors declare that there is no conflict of interest regarding the publication of this paper.

FUNDING INFORMATION

None.

CONFLICT OF INTEREST STATEMENT

None.

CONSENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

DATA AVAILABILITY STATEMENT

The data are not publicly available due to privacy or ethical restrictions.