

Chest Pain in a Middle-Aged Smoker with Heart Failure and Missing Lung

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Background

Bullous lung disease refers to radiolucencies measuring greater than 1cm in diameter. Bullae formation is most commonly seen in smokers with COPD or alpha-1-antitrypsin deficiency, but has also been associated with HIV, injection drug use, collagen vascular disease like Ehlers Danlos or Marfan syndrome, and rarely with autoimmune disease like Sjogrens's syndrome or sarcoidosis and vasculitis like polyangitis with granulomatosis. Typically, patients are asymptomatic, but can suffer from dyspnea with or without exertion, cough, or rarely, hemoptysis. Complications can result from infection, pneumothorax, and a possible association with bronchogenic carcinoma (1-3).

Case Presentation

A 45-year-old man with a history of pulmonary embolism on therapeutic anticoagulation, heart failure with reduced ejection fraction of 9% with an automated intracardiac defibrillator in place, and active tobacco and cocaine use disorder who presented with chest pain. The pain was sharp, left-sided, lasting one hour, with radiation to the right chest and back and started two hours after snorting cocaine. Vital signs on admission were unremarkable with the exception of a respiratory rate of 20 breaths per minute. Laboratory findings were unremarkable, which included an unchanged electrocardiogram from prior. A chest x-ray showed the absence of pulmonary vascular markings in the right lung apex (Figure 1A). A chest computerized tomography angiogram was obtained to evaluate for aortic dissection, which was negative. However, the computerized tomography of the chest showed the right lung findings in Figure 1B. What is the diagnosis for Figure 1B?

Discussion

These right lung findings are consistent with vanishing lung syndrome, which is defined as the presence of giant bullae in one or both lungs, occupying at least one third of the hemithorax and compressing surrounding normal lung parenchyma (1). The disease usually affects young male smokers with a history of cannabis use or alpha-1 antitrypsin deficiency, and it is frequently unilateral (2). The condition is progressive, with continuous enlargement of the bullae that results in further compression of the adjacent lung parenchyma and worsening dyspnea. Its management includes medical comorbidity optimization and surgical interventions such as bullectomy or endobronchial valve insertion. Indications for bullectomy include severe dyspnea, pneumothorax, pain, infection, and/or hemoptysis (3). Pulmonary function testing is required for pre-operative evaluation. Although bullectomy is the treatment of choice, indications for surgery should be assessed individually. Severe comorbidities such as pulmonary hypertension or end-stage heart failure may preclude any surgical intervention.

Conclusion

Initially, this patient was admitted to rule out an acute aortic dissection, which was negative. A review of the patient's previous imaging findings revealed progressive expansion of the right apical bullae over one year (Figures 1C and 1D). The patient did not undergo surgical evaluation for bullectomy due to his end-stage heart failure and decision to continue smoking. Ultimately, this patient was found to have vanishing lung syndrome, with the goal being optimization of medical management for improvement of quality of life and symptomatic relief.

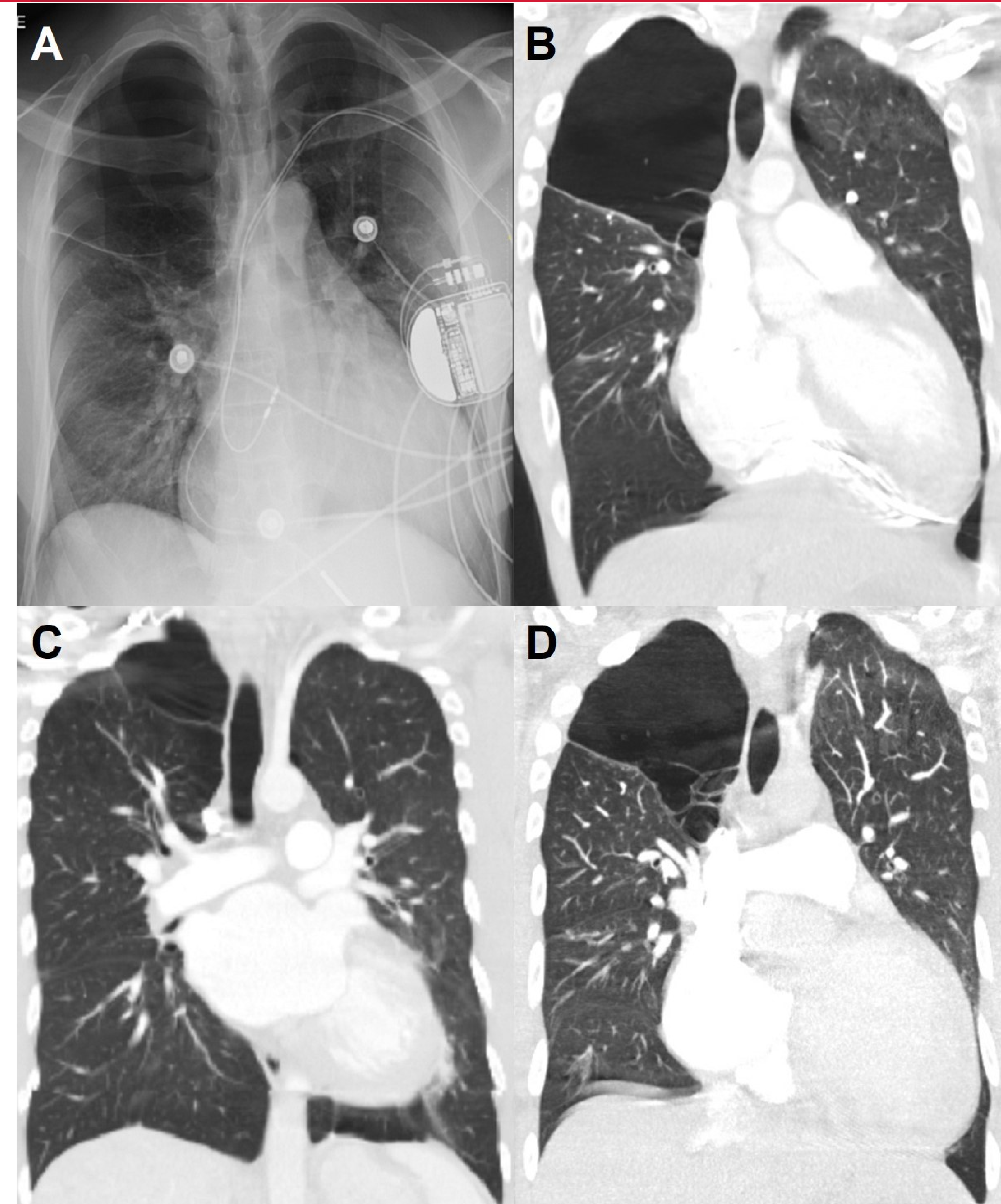


Figure. **A)** Chest x-ray on admission showing absence of pulmonary vascular markings in the apex of the right lung. **B)** Right giant bullae on presentation, occupying more than one-third of the right hemithorax. **C)** Right apical bullae 12 months before presentation. **D)** Right apical bullae 4 months before presentation.

References:

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