

A case of vanishing lung syndrome

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CASE REPORT

A 37-year-old African-American male with past medical history of tobacco and marijuana abuse presented with sudden onset shortness of breath on exertion and chest pain. The chest pain was severe, right-sided, intermittent, and pleuritic. He reported diaphoresis and a 50-pound weight loss over the last year. He also noted a rapid decline in effort tolerance over the last year and being symptomatic on activities of daily living. He denied trauma, hemoptysis, and chronic lung diseases. The patient was a chronic cigarette smoker, averaging seven cigarettes per day for 22 years, and had a distant history of chronic marijuana smoking, averaging 1 ounce per day. On examination, the patient was in respiratory distress, tachypneic, tachycardic, with tenderness to palpation of the right chest. Chest X-Ray (Figure 1) showed right apicolateral pneumothorax with underlying severe lung parenchymal bullous disease. Computed tomography scan of chest with contrast (Figure 2 and Figure 3) confirmed moderate-sized right pneumothorax with bilateral giant emphysematous bulla, left greater than right, with rightward shift of mediastinum. While in the emergency room, the patient suddenly developed acute respiratory distress with distended neck veins, which was concerning for tension pneumothorax. Chest tube

was inserted into the right lateral chest, and there was return of air. Chest X-ray showed improvement in the right pneumothorax. A V/Q scan (Figure 4) showed 91% perfusion occurring in the right lung, 9% in the left lung, and no lung ventilation on the left. Alpha-1-antitrypsin testing was negative. The chest tube was removed after >96 hours without air leak, and the patient did not have recurrence of the pneumothorax. Cardiothoracic surgery was consulted and felt that the patient lacked lung reserve to successfully undergo bullectomy and recommended lung transplantation. The patient declined transplantation and was lost to follow-up.

DISCUSSION

Idiopathic giant bullous emphysema, also known as vanishing lung syndrome (VLS), is a rare condition. It is often asymptomatic but may present with progressive

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Received: 21 November 2017

Accepted: 05 December 2017

Published: 22 December 2017

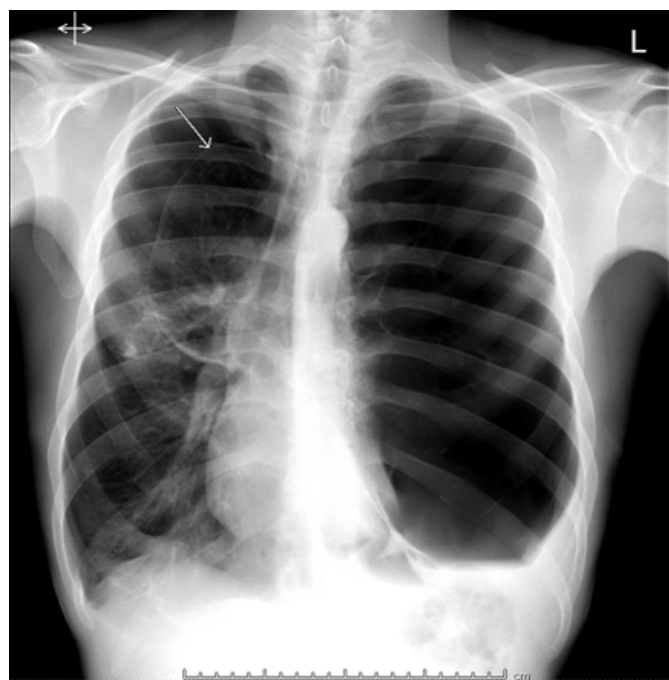


Figure 1: Chest X-Ray showing right pneumothorax and large bulla on the left pushing the mediastinum to the right.

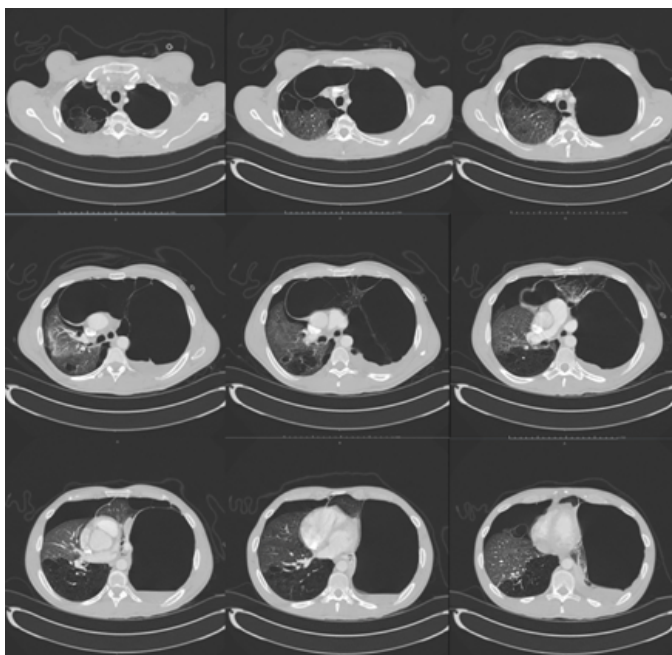


Figure 2: Axial chest computed tomography scan with intravenous contrast revealing multiple giant bullae and right pneumothorax.

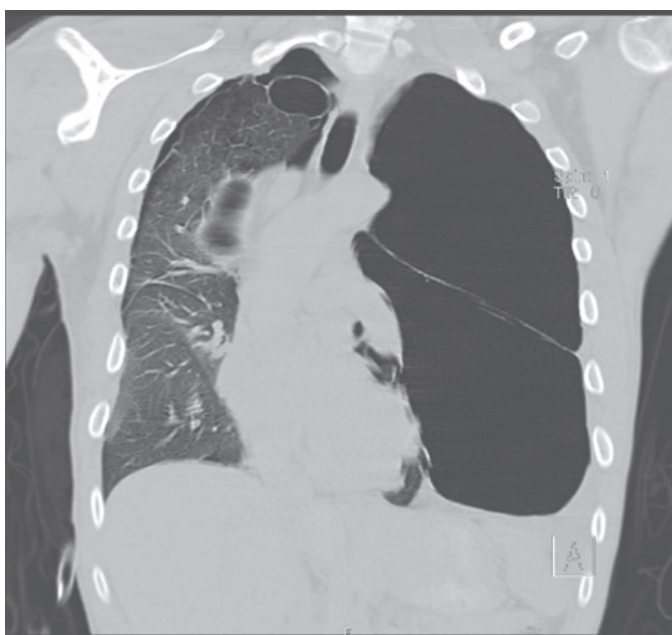


Figure 3: Coronal chest computed tomography scan with intravenous contrast revealing multiple giant bullae and right pneumothorax.

dyspnea and hypoxia. This condition usually occurs in young, thin, male smokers [1]. Risk factors include smoking, marijuana abuse, and alpha-1-antitrypsin deficiency [2–4]. Marijuana smokers have asymmetrical bullous disease with pathological changes happening approximately 20 years earlier than in tobacco smokers

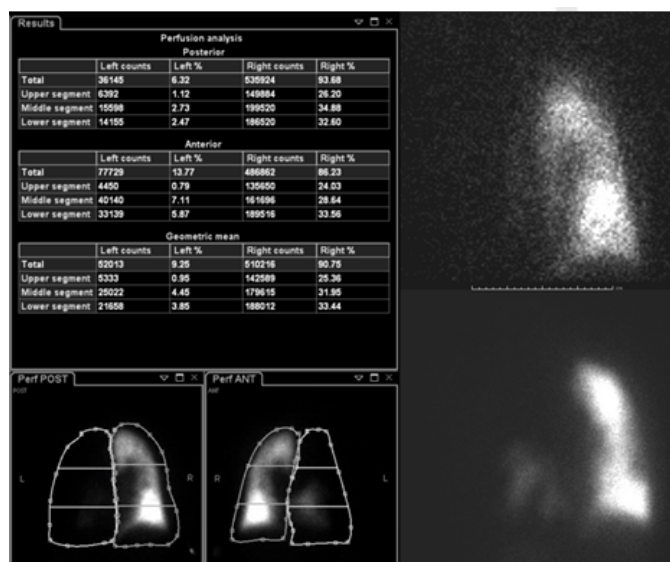


Figure 4: Ventilation perfusion scan showing major ventilation on right compared to left lung.

[2, 3]. The radiographic criteria proposed in 1987 [5] include giant bullae in one or both upper lobes occupying at least one third of the hemithorax and compressing surrounding normal parenchyma. One of the major complications of VLS is spontaneous pneumothorax, which presents as chest pain with acute deterioration in respiratory function [6]. Vanishing lung syndrome bullae can also mimic pneumothorax, and it is difficult to distinguish between the two clinically and with chest radiography. The distinction can usually be made on CT scan of chest [7]. Lung-volume-reduction surgery (or bullectomy) is considered for selected patients with VLS after assessment of exercise capacity, pulmonary function testing, and smoking cessation [1]. Bullectomy leads to improvements in pulmonary function for up to five years [8].

CONCLUSION

Vanishing lung syndrome (or idiopathic giant bullous emphysema) is a rare condition that may present with progressive dyspnea and hypoxia in young smokers. Imaging reveals giant bullae compressing lung parenchyma. These bullae can mimic pneumothorax, and it can be difficult to make the distinction on chest radiography.

Keywords: Idiopathic giant bullous emphysema, Large bulla, Lung

How to cite this article

Farcas AM, Dangayach P, Narendra DK. A case of vanishing lung syndrome. J Case Rep Images Med 2017;3:58–60.

Article ID: 100044Z09AF2017

doi: 10.5348/Z09-2017-44-CR-16

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Acknowledgements

Jason Pelton, M.D

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Andra Malina Farcas – Conception and design, Acquisition of data, Drafting the article, Critical revision of the article, Final approval of the version to be published

Priti Dangayach – Conception and design, Acquisition of data, Critical revision of the article, Final approval of the version to be published

Dharani Kumari Narendra – Analysis and interpretation of data, Critical revision of the article, Final approval of the version to be published

Guarantor of Submission

The corresponding author is the guarantor of submission.

Source of Support

None

Conflict of Interest

Authors declare no conflict of interest.

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