

Vanishing lung syndrome masquerading as pneumothorax in a smoker: Now you see me, now you do not

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ABSTRACT

A rare clinical syndrome, giant bullous emphysema, also known by the name of vanishing lung syndrome (VLS), is characterized by an X-ray of the chest showing disappearance of the lung. VLS is a chronic disease that progresses gradually and is usually seen in young male smokers, with other risk factors being alpha-1 antitrypsin deficiency and marijuana abuse. Giant emphysematous bullae are a pathognomonic presentation with a preference towards the upper lobe of the lungs. These emphysematous bullae may remain dormant for a long duration before presenting as worsening dyspnoea. Computed tomography is an essential tool to diagnose VLS. The treatment modalities include surgical and thoracoscopic resection of the bullae. We have reported a rare case of VLS in a 58-year-old chronic smoker managed conservatively.

KEY WORDS: Giant bullous emphysema, smoking, vanishing lung syndrome

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INTRODUCTION

Vanishing lung syndrome (VLS), also known as giant bullous emphysema, is a chronic disease which may be encountered in chronic smokers or those with alpha 1 antitrypsin deficiency. The underlying pathophysiological mechanism is thought to be alveolar wall destruction due to elastase. This entity can easily be misinterpreted as pneumothorax. A computed tomography of chest is a valuable tool that helps to correctly identify this entity and thereby avoid insertion of a chest tube, which can lead to development of prolonged air leaks. We hereby describe a case of a 58-year-old smoker who was referred to us with a provisional diagnosis of pneumothorax and which was ultimately diagnosed to be a case of VLS.

CASE

A 58-year-old male presented in a rural tertiary care hospital of central India with a history of breathlessness for 2 months, which was exaggerated since last 1 day. The patient was referred from primary health care centre with the provisional diagnosis of pneumothorax. The patient was a chronic smoker with a 54 packs per year smoking history. There was no history of prior hospitalization. On examination, pulse was 128 beats per minute, regular in rhythm, blood pressure was 100/70 mm of Hg, respiratory rate was 32 breaths per minute, body temperature was 37.5°C and room air saturation was 82%.

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Figure 1: Chest X-ray demonstrating bullae in bilateral upper lobes

There was reduced chest expansion bilaterally along with hyperresonant note on percussion. On auscultation, breath sounds were reduced in the bilateral inframammary region. Laboratory workup was unremarkable. Arterial blood gas (ABG) on room air showed pH 7.38, Pao₂ 54 mm Hg, and Pco₂ 36 mm Hg. Chest X-ray revealed bullous changes in bilateral upper lobes as shown in [Figure 1]. A CT scan was performed to rule out pneumothorax, which revealed giant bilateral bullae compressing rest of the lung parenchyma suggestive of VLS as shown in [Figure 2]. Alpha antitrypsin levels were within normal range.

Our patient improved with oral prednisolone and nebulization along with oxygen support and was discharged in stable condition. Spirometry at discharge revealed forced vital capacity (FVC) of 1.27 L (52% predicted), FEV₁ 0.61 L (30% of predicted) and FEV₁/FVC ratio 48.9. In view of poor respiratory reserve, the patient was decided to be managed conservatively.

DISCUSSION

Burke reported the first-ever case of VLS as a case of “vanishing lungs” in a case of a 35-year-old male with progressive breathlessness and respiratory failure. The radiologic finding showed giant bullae occupying more than two-thirds of both the hemithoraces.^[1]

Alveolar wall destruction by elastase released by macrophages in the alveoli and neutrophils are pivotal in bullae pathogenesis. By definition, these bullae are air-filled spaces having diameters more than 1 cm and up to 20 cm with an average of around 6–8 cm. Bullae are of three types: type I which occupy the upper zone, type II which occupy the middle zone and type III which occupy the lower lobe. Patients with giant bullae are divided into four groups: Group I having a single giant bulla along with normal lung parenchyma, Group II having multiple giant bullae along with normal lung, Group III having multiple bullae with underlying lung increasingly affected by emphysema and Group IV with multiple bullae with

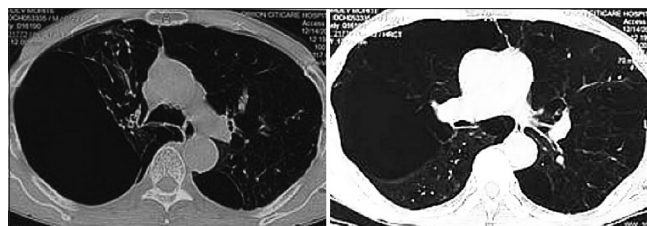


Figure 2: CT scan demonstrating giant bullae with compression of the lung parenchyma

underlying lung affected by other diseases.^[2] When these bullae are accompanied with emphysema, they are known as emphysematous bullae.^[2]

In VLS, normal lung parenchyma is rapidly replaced with bullae.^[3] They result in a poor gas exchange resulting in hypercapnia and hypoxemia.^[3]

In certain cases, these enlarging bullae may lead to compression of the surrounding lung parenchyma, with pressure effect on the mediastinum. The radiological criterion for the diagnosis of VLS is the presence of giant bullae in one or both the upper lobes of the lung, which occupy more than one-third of the hemithorax and compresses the surrounding lung tissue.^[4] Our case also showed shifting of mediastinum to left due to pressure effect of giant bulla. Investigations for a case of VLS, apart from radiological investigations, include pulmonary function test and alpha 1 antitrypsin levels along with ABG monitoring. In patients with VLS who are asymptomatic or have mild symptoms, a conservative approach is usually useful; however, in patients having severe symptoms or in the presence of pneumothorax intervention in the form of surgery, including volume reduction with video-assisted thoracoscopic surgery, bullectomy, endo-cavitary drainage, one-way endobronchial valves or lung transplant have been reported.^[5] Our patient responded well to medical management and was discharged in stable condition.

In contrast to other cases which have been reported in young individuals by Darlong *et al.*^[6] and Sood *et al.*^[7] our patient was middle aged. Our patient was denied surgical management due to poor respiratory reserve and was managed conservatively similar to another case reported by Yousaf *et al.*^[8] but in contrast to another case reported by Vij *et al.* where the patient was managed with multiple thoracotomy procedures.^[9] Hence, a case-to-case individualised approach is required for drafting a management plan in VLS.

It was important that our patient was not intubated and put on a mechanical ventilator as managing such cases without invasive ventilation is desirable. This is due to ventilator-induced pneumothorax which is a potentially lethal complication.

An important aspect of the reported case remains that the patient belonged to rural central part of India with

persistent breathlessness for 2 months for which he did not consult any health care centre till the symptoms were aggravated. This highlights the fact that there is lack of awareness in the remote areas of India as regards to when to visit a health care centre. Also, the primary health care clinicians who are often the first point of contact with these patients presenting with breathlessness should be made aware of this entity known as VLS, which can be missed easily on a chest X-ray or misinterpreted as pneumothorax leading to insertion of a chest tube and development of serious complications with a resultant increase in mortality and morbidity due to prolonged air leaks.^[10] In our case, the patient was referred with a provisional diagnosis of pneumothorax without insertion of a chest tube due to uncertainty in diagnosis, which proved to be a blessing in disguise.

Sometimes even a CT scan is not clearly able to distinguish between a giant bulla vs pneumothorax, but in our case, a straight line demarking lung parenchyma from bulla was clearly seen. However, this could be mistaken for border of collapsed lung with pneumothorax, thereby emphasizing need for precise interpretation of the CT scan of such patients. An Indian case report from a tertiary care too indicates that this diagnosis is frequently overlooked even after multiple hospital visits, hence requiring more awareness.^[9]

CONCLUSION

VLS is an uncommon diagnosis which closely mimics a pneumothorax. It should be suspected by the treating clinicians in individuals at risk such as smokers, cases with alpha-1 antitrypsin deficiency as well as marijuana users who present with progressive breathlessness. A CT scan of the chest is valuable in differentiating a bulla from a pneumothorax. It can also avoid incorrect insertion of a chest tube in such cases. An early and prompt diagnosis

can help in the successful management and prevention of morbidity and mortality.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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