vanishing lung syndrome masquerading as bilateral pneumothorax: a case report

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ABSTRACT

vanishing lung syndrome (VLS) is a rare condition characterized by giant emphysematous bullae. It is frequently misdiagnosed as pneumothorax. We describe a case of a 30-year-old male who presented with shortness of breath, reduced effort tolerance, and pleuritic chest pain for three months. He was initially diagnosed with bilateral pneumothorax based on clinical examination and chest radiograph findings. However, further imaging with a high resolution computed tomography (HRCT) of the thorax confirmed bilateral giant emphysematous bullae. Our patient subsequently underwent video-assisted thoracoscopic surgery (VATS) and bullectomy. In this report, we discuss the clinical presentations, radiological features, and the management of VLS. We also highlight the differentiating features of VLS from a pneumothorax.

1. Introduction

vanishing lung syndrome (VLS) or idiopathic giant bullous disease is a rare condition characterized by giant emphysematous bullae. It was first described by Burke et al., in 1937 [1]. It commonly develops in the upper lobes and occupies at least one-third of one or both hemithorax [2]. It mainly affects young males who are smokers. The pathogenesis of the disease is due to the destruction of the alveolar walls that results in the formation of subpleural blebs that coalesce to form a giant bulla [1, 3].

VLS is frequently misdiagnosed as pneumothorax by clinicians [4-6]. Distinguishing VLS from pneumothorax requires careful interpretation of the chest radiograph and requires confirmation with high-resolution computer tomography (HRCT) of the thorax [3]. We report a case of VLS, which was misdiagnosed as bilateral pneumothorax. In this report, we highlight the differentiating clinical and radiological features of VLS from pneumothorax. We also discuss the treatment and outcomes of this condition.

1.1. Case report

A 30-year-old farmer with 20 pack-years of smoking history presented to the emergency department with shortness of breath, reduced effort tolerance, and pleuritic chest pain for three months. There were no other associated symptoms. There was no exposure to pulmonary tuberculosis or other infections.

On examination, he was cachectic with a body mass index (BMI) of 19. His pulse rate was 88 bpm, respiratory rate was 20 breaths per minute, and pulse oxymetry was 88% under room air. He was normotensive and afebrile. There was no peripheral or central cyanosis and no finger clubbing. On respiratory examination, the trachea was central, and there were hyper-resonant notes on percussion of both lung fields. There were reduced breath sounds and vocal resonance over the middle and upper zones bilaterally. The rest of the physical examinations were unremarkable.

His blood investigations revealed normal white cell count, C-reactive protein, and erythrocyte sedimentation rate. His arterial blood gas showed type 1 respiratory failure. Investigations for secondary causes of bullous disease were negative for hepatitis B, hepatitis C and human immunodeficiency virus (HIV). Serologic testing for connective tissue...
disease, including the anti-nuclear antibody (ANA) and extractable nuclear antibodies (ENA), were also negative. The serum Alpha-1 antitrypsin level was within the normal range (1.66g/L; N: 0.9–2.0 g/L).

On the initial chest radiograph, there was an absence of lung markings of both middle and upper zones (Fig. 1). A diagnosis of bilateral pneumothorax was made, and two 17-gauge chest drains were inserted. Despite the bilateral chest drain insertion, the patient remained breathless, and there was no clinical improvement. The repeat chest radiograph did not show lung re-expansion. An urgent HRCT thorax revealed bilateral giant bullae of the upper and lower lobes, compressing the lung parenchyma inferiorly. The chest tubes were abutting the pleura’s parietal wall without puncturing the bulla (Fig. 2).

We referred this case to the cardiothoracic team, and the patient subsequently underwent bilateral video-assisted thoracoscopic surgery (VATS) and bullectomy with bilateral pleurodesis (Fig. 3). Post-operatively, the patient showed a significant clinical improvement with partial lung re-expansion. He was discharged home with bilateral portable one-way valve chest drainage (Pneumostat). He also underwent intensive outpatient chest physiotherapy. Upon clinic follow-up, the patient showed significant recovery, and the serial chest radiographs revealed marked improvement of lung expansion. The chest drain was removed after two weeks.

2. Discussion

Differentiating giant emphysematous bullae from pneumothorax poses a challenge to the less experienced clinicians owing to its rarity and the similarities of findings on physical examination and chest radiograph. VLS commonly presents insidiously in contrast to pneumothorax, which typically presents acutely [2]. Patients may present with a range of symptoms such as cough, dyspnoea, chest pain, and some may be asymptomatic [6]. VLS may be idiopathic or secondary. It has been implicated with cigarette smoking, inhaled drug use such as marijuana, and intravenous drug use. Certain conditions such as alpha-1 antitrypsin deficiency, Marfan and Ehlers-Danlos Syndrome, and HIV infection have also been associated with VLS [3,4].

![Fig. 1. Chest radiograph upon initial presentation to emergency department shows giant bullae in bilateral lungs occupying more than two third of the lungs leaving small area of normal parenchymal inferiorly.](image1)

![Fig. 2. CT Thorax in axial view shows the tip of the right chest tube (arrow) is at the anterior wall of the right giant bullae. It is located in between the parietal and visceral wall of the pleura without puncturing the bullae.](image2)

![Fig. 3. Showing both the right(above) and left bullae (below) that were removed during VATS.](image3)
with a giant bulla is concave as opposed to convex in a typical pneumothorax [7]. The inability to see the entire pleural line separated from the chest wall on chest radiography should raise a concern of an alternative diagnosis. Further imaging with HRCT of the thorax is warranted to assess the lung parenchyma. The significant feature on CT is the extensive para-septal emphysema coalescing into giant bullae. The “double-wall sign,” characterized by visualization of air on both sides of the bulla wall, is present when there is a concomitant pneumothorax [5]. Ultrasound examination may have a role to provide a quick assessment, especially in patients whose conditions are not stable enough to undergo a CT examination. The absence of both “comet-tail” and “lung sliding” signs on ultrasound are suggestive of pneumothorax [6]. However, ultrasound use is operator-dependent and requires expert skills.

A conservative approach may be appropriate in certain cases, especially in asymptomatic giant bullae or those deemed unfit for surgical interventions. However, bullectomy is the definitive treatment for symptomatic patients in order to allow re-expansion of the remaining lungs. The postoperative outcome is primarily determined by the size of the bullae and the condition of the remaining lung segments [5]. Patients without underlying diffused emphysema are expected to have a favorable long-term outcome [10]. As for patients who are not candidates for surgery, there have been recent reports on successful intervention using the bronchoscopic approach [11].

3. Conclusion

A thorough history taking, detailed physical examination, and knowledge in chest radiograph interpretation are essential in establishing the diagnosis of vanishing lung syndrome. In a clinically stable patient, it may be appropriate to delay any invasive intervention such as bilateral chest drain insertion until further assessment with a CT thorax. It is crucial to confirm the underlying pathology to avoid invasive interventions that may pose a risk to the patient. Although an asymptomatic giant bullous disease can be managed conservatively, bullectomy is the mainstay of treating symptomatic giant bullae. Advancement in the newer intervention method is still under research.

Author statement

Informed consent has been obtained from the patient for publication of the case and the related images.

Declaration of competing interest

The authors have declared that no competing interests exist.

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