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Respiratory Medicine

VANISHING LUNG SYNDROME RESEMBLING PNEUMOTHORAX - A CASE REPORT

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ABSTRACT Vanishing lung syndrome (VLS) or idiopathic giant emphysematous bullae is a rare condition characterized by giant emphysematous bullae located primarily in the upper lobes of the lungs. It is often misdiagnosed as pneumothorax. Here is a case of a 33-year-old male who presented to the hospital with shortness of breath for 2- month duration reduced effort tolerance, and pleuritic left-sided chest pain for one-month duration. He was initially diagnosed with left-sided pneumothorax based on clinical examination and chest radiograph findings. Further imaging with a high-resolution computed tomography (HRCT) of the thorax confirmed left giant emphysematous bulla. In this report, the clinical presentations, radiological features, management of Vanishing lung syndrome and also the differentiating features of VLS from a pneumothorax will be discussed.

KEYWORDS :

BACKGROUND

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 affects young males who are smokers. The pathogenesis of the disease is due to the destruction of the alveolar walls which results in the formation of subpleural blebs that coalesce to form a giant bulla [1, 3]. VLS is frequently misdiagnosed as pneumothorax [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].

Case Report

A 33-year-old manual labourer presented to the emergency department with shortness of breath ,reduced effort tolerance, and pleuritic chest pain for one month duration. There were no other associated symptoms. The patient's history was positive for tobacco abuse, with one pack of beedi per day over the past 15 years. There was a history of exposure to pulmonary tuberculosis and completed six months of treatment for the same, three years back.

On examination, his pulse rate was 88 bpm, respiratory rate was 20 breaths per minute, and pulse oximetry was 94% on room air. He was normotensive and afebrile. There was no peripheral or central cyanosis and no finger clubbing. On respiratory examination, the trachea was shifted towards the right, and there was a hyper-resonant note on percussion of left lung fields. There were reduced breath sounds and vocal resonance over the upper zone and lower zones left side. The rest of the physical examinations were normal.

His blood investigations revealed normal white cell count, C-reactive protein, and erythrocyte sedimentation rate. Investigations for hepatitis B, hepatitis C and human immunodeficiency virus (HIV)were negative. Serologic testing for connective tissue disease, including the anti-nuclear antibody (ANA) and extractable nuclear antibodies (ENA), were also negative.

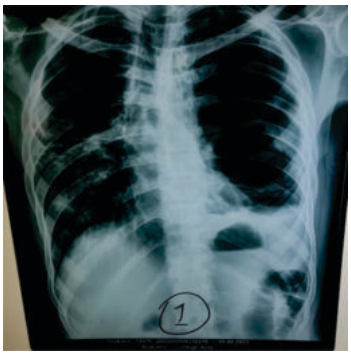

Figure 1: Chest radiograph (PA view) showing bullous changes involving bilateral upper lobe with severe bullous emphysematous changes particularly in the left hemithorax

On the initial chest radiograph, there was an absence of lung markings of the left upper and lower zone (Fig. 1) based on which, a diagnosis of left-sided pneumothorax was made. An urgent HRCT thorax revealed the left giant bulla of the upper and lower lobes, compressing the lung parenchyma inferiorly. The patient was managed conservatively and discharged three days later.

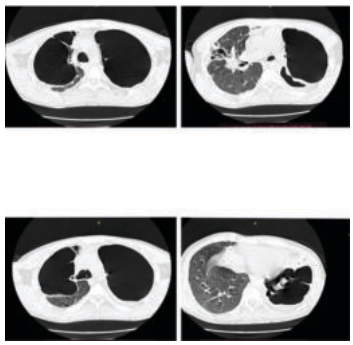

Figure 2: Axial section of CT showing severe bilateral emphysematous changes with a giant bulla involving left upper lobe, compressing left lower lobe parenchyma.

DISCUSSION

Differentiating giant emphysematous bullae from pneumothorax poses a challenge to less experienced clinicians owing to its rarity and the similarities of findings on physical examination and chest radiographs. VLS commonly presents insidiously where as pneumothorax, 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. Patients are usually young males with a history of cigarette smoking, inhaled drug use such as marijuana, and intravenous drug abuse. Certain conditions such as alpha-1antitrypsin deficiency, Marfan and Ehlers-Danlos Syndrome, and HIV infection have also been associated with VLS [3,4].

On chest radiography, the shape of the lung parenchyma associated with a giant bulla is concave in contrast to pneumothorax where it is convex in shape. [7]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]. The absence of both “comet-tail” and “lung sliding” signs on ultrasound are suggestive of pneumothorax

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 which allows re-expansion of the remaining lungs.

CONCLUSION

As Respiratory physicians whenever a patient presents with hyperlucent Chest X-Ray we should suspect possibility of VLS apart from pneumothorax.History taking, physical examination, and thorough chest radiograph evaluation is essential in the diagnosis of vanishing lung syndrome.In vitally stable patients, delaying invasive interventions such as chest drain insertion till CT thorax will improve the quality of management because VLS can be managed conservatively. In symptomatic cases, bullectomy is the mainstay of treatment.

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