ORIGINAL RESEARCH PAPER

General Medicine

AN IDIOPATHIC BULLOUS EMPHYSEMA MIMICKING PNEUMOTHORAX

KEY WORDS:

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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 60-year-old man with a 50 packs per year smoking history presented with worsening shortness of breath, non-productive cough, wheezing, and generalized weakness for the past few days. He was initially diagnosed with pneumothorax based on clinical examination and chest radiograph findings. However, further imaging with a high resolution computed tomography (HRCT) of the thorax confirmed giant emphysematous bullae, we referred this case to the cardiothoracic team for video-assisted thoracoscopic surgery (VATS) and bullectomy with pleurodesis. 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.

INTRODUCTION

Idiopathic giant bullous emphysema also known as Vanishing lung syndrome represents a rare form of irreversible damage to the pulmonary parenchyma often due to chronic obstructive pulmonary disease (COPD)[1]. It commonly develops in the upper lobes and occupies at least one-third of one or both hemithorax[1] .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[2]. Patients with this condition typically have a long history of smoking or COPD but may also be younger with a history of marijuana use or have alpha-1 antitrypsin deficiency[1]. VLS is frequently misdiagnosed as pneumothorax by clinicians ,Idiopathic giant bullous emphysema may mimic the presentation of pneumothorax with worsening dyspnea, hypoxia, and a chest radiograph revealing an absence of pulmonary markings[3]. However, in the case of a true pneumothorax, a white line representing visceral pleura that has separated from the chest wall is often seen.

CASE REPORT

A 60-year-old man with a 50 packs per year smoking history presented with worsening shortness of breath, non-productive cough, wheezing, and generalized weakness for the past few days.

GENERAL PHYSICAL EXAMINATION-

Patient was conscious oriented to time, place and person Blood pressure-140/80mm/hg, Pulse rate-102/min, Respiratory rate-22/min, Spo2-90% on room air, Pallor, Icterus, Clubbing, Cyanosis, Lymphadenopathy, Edema are Negative.

RESPIRATORY EXAMINATION-

INSPECTION-

1.Trachea-shift to right side
2.Shape-barrel shaped
3. Movement-decreased in left side.

PALPATION-

1.Trachea-shifted to right
2.Apex beat-cannot appreciate.
3. Respiratory movements-left sided mammary, infraclavicular movements are decreased compare to right.
4. Dimension-AP diameter-24cm,Transverse diameter-34cm.
5. Chest circumference-
Inspiration- 86cm, expiration: 83.5cm, chest expansion-2.5cm,

Right hemithorax
Inspiration- 42cm, expiration -40cm, Right hemithorax expansion-2cm,

Left hemithorax
Inspiration- 44cm, expiration 43.5cm, left hemithorax expansion-0.5cm.

PERCUSSION-
Hyper resonant in left mammary and left infrascapular area.

AUSCULTATION-
Diminished breath sound and vocal resonance in left mammary and left infrascapular area.

ECG-

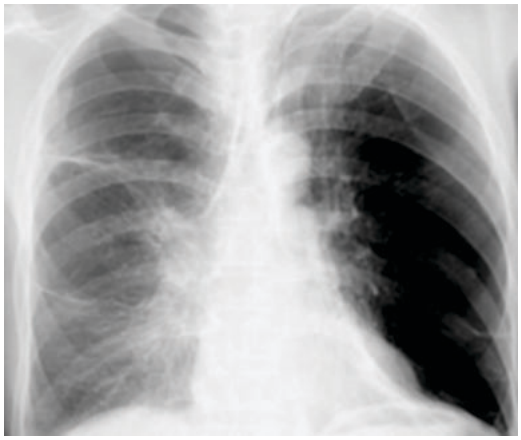
No abnormality detected.

LABORATORY WORK UP-

Complete blood count-normal
Renal function test-normal,
Liver function test -normal,
Viral serology -HIV, HBV, HCV-normal
Serum alpha 1 antitrypsin level-normal

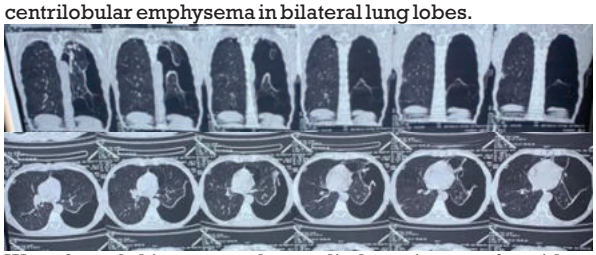
CHEST XRAY –

Shows left side absence of pulmonary marking concerning for pneumothorax.



CT CHEST-

Shows idiopathic giant bullous emphysema (vanishing lung syndrome)involving left upper and lower lung lobe associated with paraseptal emphysema, bullae and centrilobular emphysema in bilateral lung lobes.



We referred this case to the cardiothoracic team for video-assisted thoracoscopic surgery (VATS) and bullectomy with pleurodesis.

DISCUSSION

COPD is common preventable and trustable disease characterized by persistent respiratory symptoms with airflow limitation due to airway alveolar abnormalities caused by exposure to noxious particles or gases, COPD includes emphysema and chronic bronchitis, emphysema is irreversible enlargement of air spaces distal to terminal bronchiole, accompanied by destruction of their walls. Chronic bronchitis is persistent cough with sputum production for at least 3 months in at least 2 consecutive years in the absence of any other identifiable cause. COPD is 3rd leading cause of global mortality. Bullous emphysema refers to emphysematous lung with bullae, which are air-filled spaces within the parenchyma that are 1 cm or larger in diameter and consist of a thin wall of visceral pleura with remnants of alveolar and interlobular septa inside. Bullae are formed by destruction of interalveolar walls by chronic or less commonly acute stretch injury with increased interalveolar pressure. If bullae occupy >30% of a hemithorax, they are termed giant bullae. These typically form slowly by gradual filling with air, but rapid enlargement and spontaneous deflation are also possible. As giant bullae occupy spaces in the thorax, they can compress other lung parenchyma and affect gas exchange. Enzymatic destruction of the alveolar wall from elastase produced by alveolar macrophages and neutrophils plays a crucial role in the pathogenesis of bullae formation Distribution of giant bullae is usually unilateral and asymmetric; however, bullous emphysema has bilateral involvement. There are no known factors that determine whether formation is unilateral or bilateral, the major cause of giant bullae formation is cigarette smoking, but they are also associated with intravenous use of methadone, methylphenidate, or talc-containing drugs. Patients with Marfan's syndrome, Ehlers-Danlos type IV, polyangitis with granulomatosis, Sjogren's syndrome, and sarcoidosis can develop bullae but not typically giant bullae. For both symptomatic and asymptomatic patients, workup includes pulmonary function testing and a serum alpha-1-antitrypsin level. a chest radiograph revealing an absence of pulmonary markings. However, in the case of a true pneumothorax, a white line representing visceral pleura that has separated from the chest wall is often seen. 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. 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 [8]. However, ultrasound use is operator dependent and requires expert skills. Stapled bullectomy, excision, ligation, and endocavitary drainage are different operative techniques to treat bullae or giant bullae. Indications for bullectomy include severe dyspnea, pneumothorax, pain, infection, or hemoptysis. Contraindications include significant comorbid disease, pulmonary hypertension, and poorly defined bullae. Based primarily on data regarding lung volume reduction surgery, relative contraindications for bullectomy include poor forced expiratory volume in 1 second (<30%), hypercapnia, cor pulmonale, and severe hypoxemia.

CONCLUSIONS

Differentiating VLS from pneumothorax is essential to determining the best plan of care. Acute respiratory distress with hypoxemia may be the early manifestation of pneumothorax due to ruptured giant bullae, and prompt chest tube placement is critical. In VLS, lung volume reduction surgery improves patients' symptoms, pulmonary function, quality of life, and the need for re-hospitalization. In general, patients should be counseled to stop smoking tobacco and marijuana. Those who are poor candidates for surgical resection may benefit from conservative management or palliation.

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