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Case Report

When Bullae Deceive: A Rare Case of Vanishing Lung Syndrome Misdiagnosed as Pneumothorax

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Abstract

Background: Vanishing lung syndrome (VLS), also referred to as giant bullous emphysema, is an uncommon form of lung disease in which massive bullae replace a substantial portion (typically more than one third) of the hemithorax, leading to compression of surrounding healthy lung tissue. It typically occurs in young male smokers but may also arise in non-smokers with biomass exposure, making diagnosis and management particularly challenging.

Case Presentation: We describe a case involving a 51-year-old non-smoking woman with long-term exposure to biomass fuel, who presented with dry cough, chest discomfort, and gradually worsening shortness of breath. An initial chest X-ray was interpreted as a left-sided pneumothorax, for which an intercostal chest drain was placed; however, clinical and radiological improvement was not achieved. Subsequent high-resolution computed tomography revealed a large emphysematous bulla occupying the left lower lobe and causing a contralateral mediastinal shift, findings consistent with vanishing lung syndrome. The patient underwent successful surgical management via left posterolateral thoracotomy and bullectomy, resulting in marked clinical improvement.

Conclusion: This case highlights the critical need to distinguish vanishing lung syndrome from pneumothorax, particularly in individuals with non-traditional risk factors like prolonged biomass smoke exposure. Early recognition and surgical intervention can lead to substantial symptomatic and functional recovery.

Keywords: Vanishing lung syndrome, Pneumothorax, Mimic, Giant bullous emphysema, Biomass exposure, Bullectomy, Hyperlucent lung.

Introduction

Vanishing lung syndrome (VLS), also referred to as giant bullous emphysema, is an uncommon form of lung disease in which massive bullae replace a substantial portion i.e. typically more than one-third of the hemithorax, leading to compression of surrounding healthy lung tissue. The condition was initially identified and characterised by Roberts and colleagues in 1987,[1] VLS typically presents in young male smokers but can occasionally affect non-smokers, particularly those exposed to biomass fuels or other environmental pollutants.[2] Clinically, it may mimic spontaneous pneumothorax or other causes of hyperlucent hemithorax, often leading to misdiagnosis and inappropriate initial management.[3]

High-resolution computed tomography (HRCT) is the gold standard for diagnosing and differentiating between true pneumothorax and large, non-functioning bullae.[4] Management strategies depend on symptom severity and the extent of lung parenchymal compression. Surgical resection through bullectomy is the best treatment in symptomatic patients having preserved lung function as well and has been shown to offer significant symptomatic and functional improvement.[5,6]

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We report an uncommon presentation of vanishing lung syndrome in a non-smoking woman with chronic exposure to biomass smoke, who was initially misdiagnosed with pneumothorax and ultimately treated successfully through surgical bullectomy.

Case Presentation

A 51-year-old non-smoking female, a homemaker and chronic biomass fuel user, presented to the outpatient department with complaints of dry cough and left-sided chest pain for one month, and progressive dyspnea over 15 days, now MMRC Grade 2. There was no history of fever, hemoptysis, tuberculosis, substance use, or significant weight loss.

On examination, the patient appeared poorly nourished (BMI: 19.1 kg/m²) with pallor. Respiratory evaluation revealed tracheal shift to the right, asymmetrical chest expansion, hyperresonance, and reduced breath sounds on the left side of the chest. Chest skiagram (PA view) showed a large hyperlucent area on the left lung field with ipsilateral mediastinal shift, suggestive of a pneumothorax (Figure 1).

An intercostal chest drain (ICD) was inserted on the left side. Although initial partial re-expansion was noted (Figure 2A), the left lung collapsed again with the subsequent development of subcutaneous emphysema. Conservative measures, including oxygen supplementation and ICD manipulation, failed to resolve the issue (Figure 2B)

Blood investigations showed mildly reduced haemoglobin levels. Infective workup, including sputum AFB and CBNAAT, was negative. HIV and HBsAg tests were non-reactive. 2D-ECHO showed preserved ejection fraction (55–60%).

A high-resolution CT (HRCT) chest revealed a giant bulla (14.6 × 10 × 17.5 cm) in the left lower lobe, causing contralateral mediastinal shift and compression of adjacent lung parenchyma, consistent with VLS (Figure 3).

The patient was referred to the cardiothoracic surgery department. Owing to the inability to perform pre-operative spirometry and DLco, surgical risk was clinically assessed. The patient was taken up for surgery under general anaesthesia. A left posterolateral thoracotomy was performed through the fifth intercostal space. Intraoperatively, a large, thin-walled, airless bulla was noted occupying the majority of the left hemithorax and compressing the underlying lung. Adhesionolysis was performed meticulously to free the bulla from the pleural and mediastinal structures. The bulla was resected using an 80 mm linear stapler (GIA), along with a margin of compressed adjacent lung tissue. Hemostasis was ensured, and an intercostal drain was placed before layered thoracic closure. The patient was extubated postoperatively and transferred to the ward in stable condition (Figure 4).

Post-operative recovery was uneventful. Lung expansion was achieved completely (Figure 5A), and the ICD was subsequently removed. The patient was discharged with symptomatic medications and advised to follow up. At

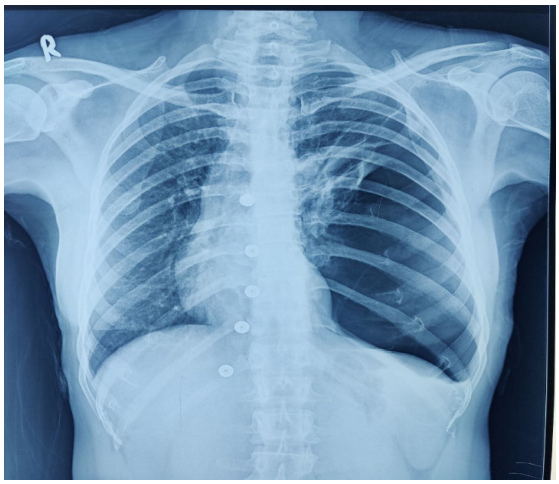

Figure 1: Chest X-ray showing a large hyperlucent area in the left hemithorax with absence of vascular markings and significant contralateral mediastinal shift. However, the lung margin is not clearly visualised

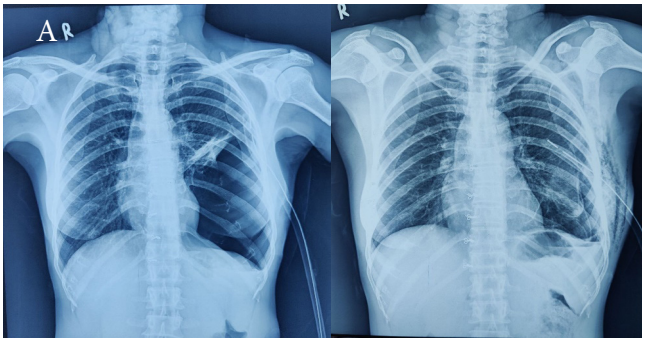

Figure 2: Serial chest radiographs after intercostal chest drain (ICD) insertion. (A) Chest X-ray (PA view) immediately following ICD insertion in the left fifth intercostal space, mid-axillary line, shows partial re-expansion of the left lung with reduction in the hyperlucent area and mediastinal shift. Surgical emphysema is also evident at lateral chest wall (B) Follow-up chest X-ray reveals re-collapse of the left lung with recurrence of hyperlucency in the left hemithorax

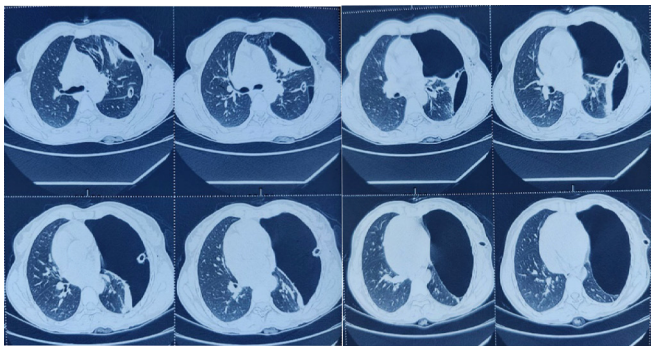

Figure 3: HRCT chest Axial HRCT image reveals a giant, thin-walled emphysematous bulla occupying the majority of the left hemithorax with significant compression of adjacent lung parenchyma, contralateral mediastinal shift, and displacement of the oblique fissure. (B) Follow-up chest X-ray reveals re-collapse of the left lung with recurrence of hyperlucency in the left hemithorax

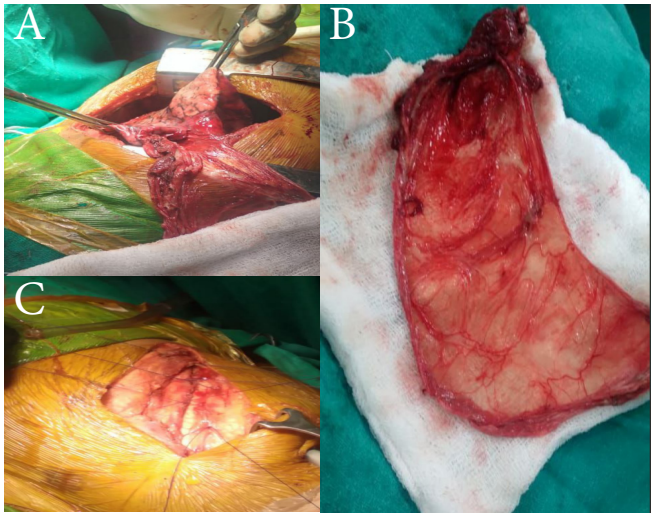

Figure 4: Intraoperative surgical findings and steps during bullectomy (A) Intraoperative image showing left posterolateral thoracotomy with identification and isolation of the giant emphysematous bulla occupying the majority of the left hemithorax. (B) Gross specimen of the excised giant bulla measuring approximately 17.5 cm in maximum dimension, thin-walled and airless, consistent with radiological findings. C) Wound closure following stapled bullectomy and intercostal drainage placement, demonstrating complete homeostasis and layered thoracic closure

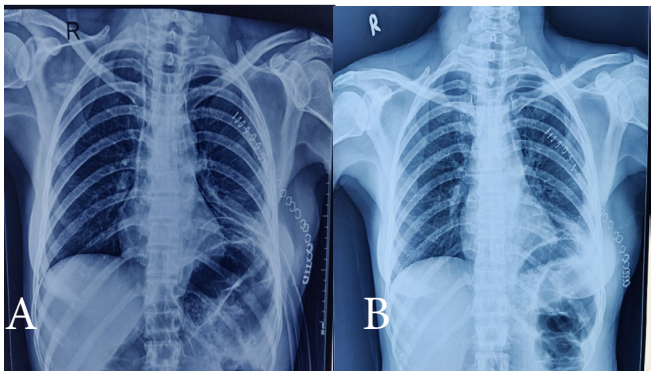

Figure 5: Post-operative and follow-up chest radiographs. (A) Immediate post-operative chest X-ray (PA view) showing expansion of the left lung with an intercostal drainage tube in situ. The previously visible giant bulla has resolved, and mediastinal structures have returned to midline. (B) Follow-up chest X-ray (PA view) after intercostal drain removal demonstrates sustained lung expansion on the left side, with no evidence of residual bullae, pneumothorax, or mediastinal shift

follow-up, the patient remained clinically stable with no recurrence of dyspnea, chest pain, or radiographic evidence of bullae or pneumothorax. Chest X-ray showed well-expanded lung fields and maintained mediastinal alignment (Figure 5B). She continued on inhaled bronchodilators and was advised to have routine follow-up.

Discussion

Vanishing lung syndrome, also known as giant bullous emphysema, is a rare pulmonary disorder defined by the formation of one or more oversized bullae that involve at least one-third of a hemithorax, resulting in compression of the adjacent functional lung tissue.[1] This condition predominantly affects male smokers between the ages of 20 and 40 years and is strongly associated with chronic tobacco use, α1-antitrypsin deficiency, and, to a lesser extent, environmental exposures such as biomass fuel combustion.[2]

Our case is noteworthy due to the occurrence of VLS in a middle-aged, non-smoking female with prolonged exposure to biomass fuel fumes which is a recognised, though underreported, risk factor for chronic pulmonary injury in developing countries.[3] The patient was initially misdiagnosed with spontaneous pneumothorax based on chest radiograph findings and underwent intercostal drainage, which failed to provide symptomatic or radiological improvement. This highlights a critical diagnostic challenge also described in literature, where bullous emphysema was mistaken for pneumothorax, leading to unnecessary invasive management with intercostal chest tube.[4,5]

The distinction between pneumothorax and a giant bulla is essential and often requires high-resolution CT imaging for confirmation.[1] CT typically demonstrates a thin-walled air-filled cavity with clear evidence of compressed adjacent lung and mediastinal shift, which distinguishes it from a true pleural air collection.

The functional impairment in VLS is variable and depends on the extent of underlying emphysema and parenchymal compression. Patients may show near-normal pulmonary function if the bullae are non-functional spaces and the underlying lung is relatively preserved.[2] However, in symptomatic patients, bullectomy offers significant clinical benefit. A prospective study demonstrated marked improvements in dyspnea scores, lung volumes, and exercise tolerance after bullectomy in appropriately selected patients.[6]

Surgical resection remains the mainstay of treatment in patients with symptomatic giant bulla with recurrent infections or suspicion of malignancy. In our case, the patient underwent successful bullectomy via posterolateral thoracotomy with stapler resection of the bulla and had significant post-operative improvement. Literature supports this approach, with VATS or open thoracotomy both yielding good outcomes depending on bulla size, adhesions, and surgeon expertise.[7,8]

Comparatively, similar cases have been reported in both smokers and non-smokers. For example, Palla *et al.*[9] described a case series having 35 patients with giant bullae, where 20% were non-smokers and biomass exposure was identified as a possible factor. This supports the growing recognition of alternative risk factors in the pathogenesis of bullous lung disease.

Importantly, spirometry and DLCO are recommended for pre-operative evaluation to assess surgical risk. In our patient, these tests could not be performed due to poor effort tolerance, yet clinical judgment allowed surgical intervention, with favourable results.

Conclusion

This case highlights the importance of differentiating VLS from pneumothorax in patients with large hyperlucent lung fields on imaging. Accurate diagnosis using HRCT and timely surgical intervention can prevent unnecessary procedures and significantly improve patients' outcomes.

Patient Consent

Informed written consent was obtained from the patient for the publication of this case report, along with any related clinical images.

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