

Case Report

A rare case of vanishing lung syndrome with pneumothorax: importance of computed tomography

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ABSTRACT

Vanishing lung syndrome (VLS), also known as giant bullous emphysema (GBE) is a distinct clinical syndrome usually affecting young male smokers characterised by large bullae that involve at least one-third of one or both hemithoraces. We are reporting one such case in a 40-year-old male patient, who was a chronic smoker for past 20 years. He was diagnosed having chronic obstructive pulmonary disease (COPD) on the basis of his history and chest X-ray findings 5 years back and treated symptomatically. He was never suspected of having VLS until a computed tomography of thorax done in our department revealed characteristic findings of VLS with pneumothorax and subcutaneous emphysema. Bullectomy, either via video thoracoscopy or conventional thoracotomy, is the treatment of choice. The patient is now in follow-up with pulmonary medicine department. This case exemplifies role of HRCT thorax in timely diagnosis and planning appropriate treatment of VLS.

Keywords: Vanishing lung syndrome, Bulla, Emphysema, Video-assisted thoracoscopic surgery, Computed tomography

INTRODUCTION

The first reported case of vanishing lung syndrome (VLS) goes back to 1937 when Burke described a case of “vanishing lungs” in a 35-year-old man who had progressive dyspnoea, respiratory failure, radiographic and pathologic findings of giant bullae that occupied two thirds of both hemithoraces.¹ This condition is also known as type 1 bullous disease, primary bullous disease of the lung. Fifty years later in 1987, Roberts and colleagues defined the radiographic criteria for vanishing lung syndrome as presence of giant bulla in one or both upper lobes, occupying at least one third of the hemithorax and compressing surrounding normal lung parenchyma.² The condition is associated with smokers, alpha-1 antitrypsin deficiency, and marijuana abuse.³⁻⁵ These giant bulla may mimic pneumothorax on chest radiograph in the clinical setting of acute dyspnoea which can lead to unintentional placement of a chest tube into a

giant bulla causing iatrogenic pneumothorax, hemothorax, hemorrhagic shock or death.⁶

High resolution computerized tomography (HRCT) is an important diagnostic tool to assess the extent and distribution of the bullous disease and coexisting conditions such as infected cysts, bronchiectasis, pulmonary artery enlargement, and pneumothorax.⁷ Bullectomy is the treatment of choice and interval follow up imaging is indicated because new giant bulla might develop again. Smoking is the only modifiable risk factor.

CASE REPORT

A 40-year-old male patient presented with insidious and progressive shortness of breath and cough with scanty expectoration for five years with frequent exacerbation of symptoms. He complained of acute shortness of breath

and chest pain for past 3 days. He did not have constitutional symptoms such as fever, night sweats, anorexia or weight loss. He didn't have history of significant cough, wheezing, hemoptysis, chest trauma or tuberculosis. He had no significant medical or surgical history. He was a chronic cigarette smoker for past 20 years. He had no history of drug abuse.

On examination, blood pressure was 130/90 mm Hg, heart rate-100/min, respiratory rate-30/min, and O2 saturation of 90% on room air. There was no cyanosis, clubbing or peripheral oedema. Examination of the chest revealed hyper resonance to percussion in both lungs and decreased apical breath sounds. Routine blood test, serological tests, sputum AFB was all negative. Spirometry revealed an obstructive pattern.

A chest X-ray (CXR) showed bilateral apical hyperlucencies with a paucity of vascular markings as shown in Figure 1. A HRCT of thorax showed bilateral large bullae consistent with vanishing lung syndrome as seen in Figure 2. CT scan confirmed presence of pneumothorax by showing double-wall Sign as presented in Figure 3. Bilateral subcutaneous emphysema was also evident on CT as given in Figure 4. Serum alpha-1 antitrypsin measurement could not be done because of non-availability in our institute. However we believe that chronic smoking has led to such condition.



Figure 1: Chest X-ray PA view showing bilateral upper lobe hyperlucencies with multiple bullae. It is difficult to assess extent of bullae on CXR and possibility of pneumothorax cannot be excluded.

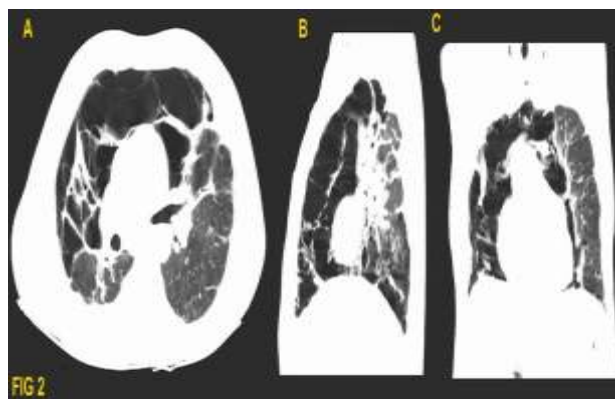


Figure 2: HRCT thorax lung window in axial (A), sagittal (B) and coronal (C) views showing the extent of bullae in both hemithoraces. Underlying lung tissue appears normal without evidence of centrilobular emphysema.



Figure 3: Double-wall sign-visualization of air external to bulla (B) i.e. air outlining both sides of the bulla. Bulla wall (arrow) is seen parallel to parietal pleura. This sign confirms presence of pneumothorax.

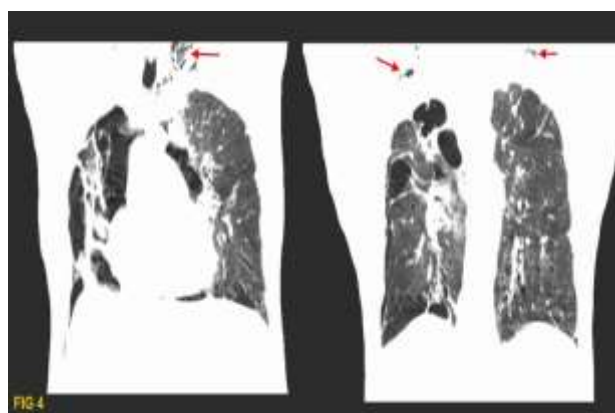


Figure 4: Note the presence of subcutaneous emphysema (arrows) which is an indirect evidence of pneumothorax due to ruptured bullae.

DISCUSSION

VLS is rare chronic, progressive condition following a downhill course, leading to respiratory failure and eventual death. It is characterised by unilateral or bilateral asymmetric upper lobe involvement with the formation of multiple bullae. It usually affects male smokers. Extensive paraseptal emphysema coalesce to form giant bullae, compressing the normal lung parenchyma.^{1,2}

Its etiology is obscure. However there is clear association with smoking, alpha-1 antitrypsin deficiency, and marijuana abuse. Cigarette smoke attracts alveolar macrophages which release chemotactic factors that provoke leukocytes to release neutrophil elastases. Neutrophil elastases causes destruction of the alveolar walls leading to emphysema. This effect is balanced by alpha-1 antitrypsin which is an antiprotease. Certain chemicals in smoking oxidize alpha-1 antitrypsin leading to unrestricted elastase activity. However VLS can also occur in non-smokers.^{3-5,8-10}

Pneumothorax is the most common complication of VLS which presents with acute dyspnoea and chest pain. Severe breathlessness can also occur due to sudden increase in size of the bulla due to air trapping. Bullae can also get infected. Giant bulla can compress on normal lung parenchyma, reduce lung compliance and increase work of breathing.^{10,11} Poor ventilation of the bullae may lead to accumulation of carcinogens in them leading to increased incidence of lung cancer. Infrequent associations with medical conditions (e.g. sarcoidosis and systemic lupus erythematosus), recreational drug abuse (e.g. intravenous methylphenidate, cocaine and marijuana) have been described.⁹

Chest radiography is the most practical method for identifying the presence of bullae and their progression. However in the clinical setting of acute dyspnoea presenting to emergency department in undiagnosed giant bullous emphysema, a chest radiograph may not be reliable enough to differentiate a pneumothorax from a giant bulla. Insertion of a chest tube into a bulla can lead to iatrogenic pneumothorax, hemothorax and death. Hence in case of equivocal clinical and conventional radiographic findings, emergency CT is advocated.^{6,9,12,13} Recognising the double wall sign on HRCT help distinguish a pneumothorax from adjacent giant bulla in the setting of giant bullous emphysema. This sign occurs when one sees air outlining both sides of the bulla wall parallel to the chest wall (the intrathoracic equivalent of the double-wall sign of pneumoperitoneum).^{12,13}

High resolution computed tomography (HRCT) plays an important role in characterizing the degree and distribution of emphysema, providing information necessary for operative planning in symptomatic individuals. On HRCT, a bulla is defined as a sharply demarcated region of emphysema 1 cm or more in diameter without discernible wall (wall thickness less

than 1 mm). HRCT has been shown to have significant correlation with pathologic grade on resected lung specimens.^{10,11} HRCT also allows assessment of coexisting conditions such as infected cysts, bronchiectasis, pulmonary artery enlargement, and pneumothorax.^{7,10,11} In addition, HRCT allows to assess extent of associated centrilobular emphysema by helping predict the behavior of the nonbullous lung following surgical resection of the bullae. Severe underlying centrilobular emphysema may preclude bullectomy. Stern et al reviewed imaging findings on chest radiography and CT of GBE in 9 patients, which include large multiple bullae ranging from 1 to 20 cm in diameter, usually between 2 to 8 cm, without a single dominant giant bulla.¹¹

Surgical resection of giant bullae is the treatment of choice (either by thoracotomy or thoracoscopy). But it is considered for selected cases only after an assessment of exercise capacity, pulmonary-function testing, and smoking cessation. The indications include (1) spontaneous pneumothorax, (2) infection, (3) dyspnoea or (4) increasing bulla size.^{9,10,14-16} Determination of the preoperative bulla volume allows the prediction of the expected increase of postoperative FEV1. There is significant improvement in all three measurements of FVC, FEV, and dyspnoea grading in the early postoperative period but all the improvements except FVC are insignificant at five to ten years. Bullectomy causes significant improvements in dyspnoea, gas exchange, pulmonary function, and exercise capacity.^{10,14} But these improvements persist for approximately 3 to 4 years and begin to decline thereafter.¹⁴⁻¹⁶ Complications of bullectomy include prolonged air leak for >7 days (53%), atrial fibrillation (12%), need for postoperative mechanical ventilatory support (9%), and pneumonia (5%). Reported early mortality rates after bullectomy are low (from 0% to 2.5%).¹⁰ Asymptomatic bullae are treated conservatively by reassurance, advice to stop smoking, avoid strenuous activities that can promote the rupture of the bullae and regular follow up. Spontaneous resolution of giant bulla have also been reported.¹⁵

CONCLUSION

VLS is a rare entity characterised by multiple giant bullae mostly affecting young smokers with a chronic progressive course. A high index of suspicion is required to diagnose VLS. Chest radiographs are not always reliable and may misdiagnose GBE as pneumothorax leading to inappropriate management. CT scans are the most accurate means of detecting emphysema, determining its type and extent and distinguishing giant bullae from pneumothorax, assessing coexisting conditions which help in deciding appropriate treatment. Surgical resection of giant bullae is the treatment of choice after careful selection of patients. Better results may be expected in patients without underlying lung disease.

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