

Vanishing Lung Syndrome

Sharath Kumar D. Shah¹, Lokesh MR², Nischal N Hegde³

¹Professor and Head, ²Assistant Professor, ³Post Graduate. Department of General Medicine
Sri Siddhartha Medical College & Research Centre, Tumakuru, Karnataka, India.

Abstract

Vanishing lung syndrome otherwise known as idiopathic giant bullous emphysema, is a rare disorder, typically occurs in young, thin male smokers. It starts with the appearance of emphysematous bullae in the lung, which progressively enlarge, causing compression of the adjacent lung. The paucity of the disorder in the literature and rarity of the condition prompts us to report this case. A 30 year old poorly built female patient was evaluated for complaints of breathlessness and non-productive cough of 1 week duration with a past history of Pulmonary Tuberculosis. Systemic examination revealed the absence of chest movements, hyperresonance note and absence of breath sounds on the left side. Chest CT showed large multiple emphysematous bullae in the left lung suggestive of vanishing lung syndrome with an emphysematous bulla in the right upper lobe. The presence of clinical and radiological features and restrictive pattern on pulmonary function testing favours the diagnosis of vanishing lung syndrome in our patient, who was not a candidate for lung volume reduction surgery. So the patient was given only supportive treatment with beta2 agonists, Glucocorticoids, Theophylline and O₂ supplementation and is under follow-up for future course and may need further evaluation.

Key Words: Bullous emphysema, emphysematous bullae, vanishing lung, idiopathic giant bullous emphysema.

Introduction

Vanishing lung syndrome (VLS) is a rare disorder otherwise known as idiopathic giant bullous emphysema, typically occurs in young, thin male smokers. In this disorder emphysematous bullae appear in the lung, alveolar walls gradually disintegrate to form large air spaces with eventual atrophy of the lung and compression of the adjacent side.^[1-3] The patients have little or no evidence of cough or respiratory infection and finally die in respiratory failure^[1]. The paucity of the disorder in the literature and rarity of the condition, prompts us to report this case. The basic pathogenesis is not exactly known, however it has been attributed to possible respiratory myositis.^[2] There is evidence of diaphragmatic weakness, and the condition is associated with systemic lupus erythematosus in one third of cases.^[2-4] In 1937, 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 hemithoracies.^[5] The radiographic criteria for this syndrome defined by Roberts et al include the presence of giant bullae in one or both upper lobes, occupying at least one third of the hemithorax and compressing surrounding normal parenchyma.^[6] VLS is also known as type I bullous disease or primary bullous disease of the lung in which the lungs appear to be disappearing on X-ray.^[7] Surgical resection of giant bullae is the treatment of choice, the indications include spontaneous pneumothorax, infection, or dyspnoea, the best results are seen following limited bullectomy.^[8-12] Thoracoscopic treatment of giant bullae is an effective alternative to conventional thoracotomy with minimal morbidity.^[13]

Address for Correspondence:

Dr. Nischal N Hegde

Sri Siddhartha Medical College & Research Centre,

Agalkote, Tumakuru, Karnataka

Email: nisheg8825@gmail.com

Case Report

A 30 year old female patient from Belagumba, Tumakuru was evaluated for complaints of breathlessness and non-productive cough of 1 week duration. There were no symptoms pertaining to any

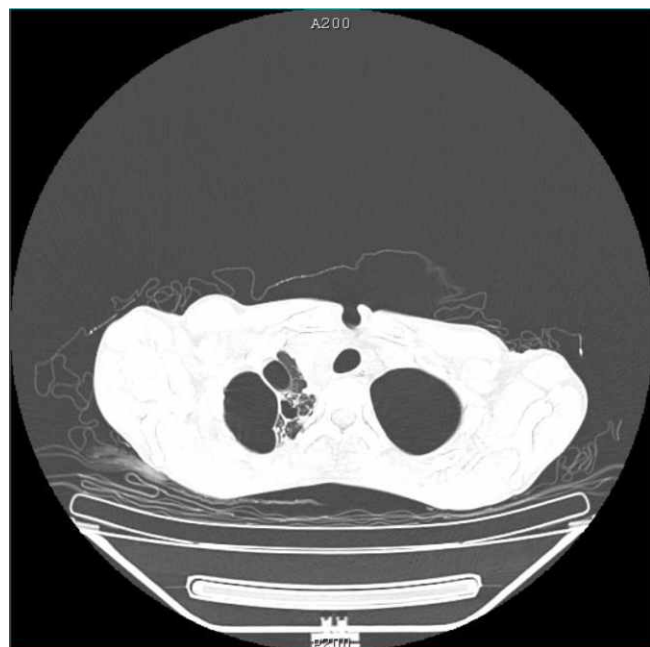


Figure 1 - Multiple emphysematous bullae seen on the right side with a large emphysematous bullae on the left side compressing the lung parenchyma.

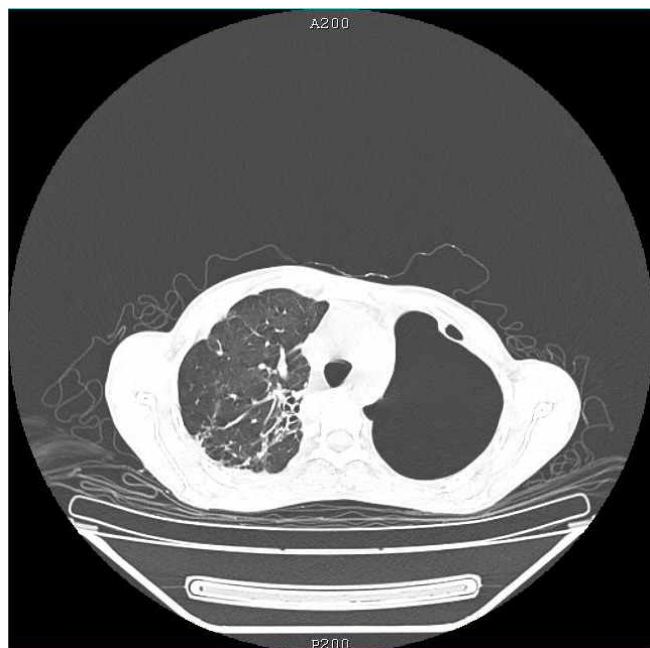


Figure 2 - Right lung showing features of Old Pulmonary Koch with bronchiectatic changes and Left lung typical of VLS.

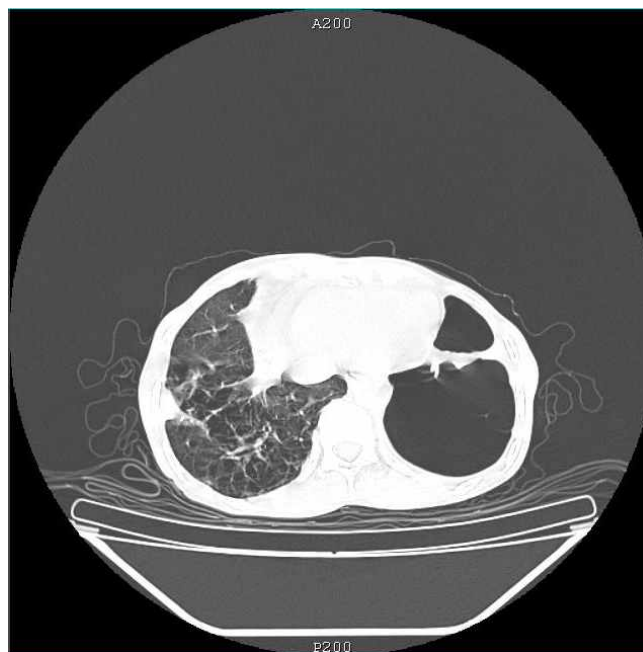


Figure 3 - Right lung showing features of Old Pulmonary Koch with fibrotic bands and left lung typical of VLS.

other body systems. She had a past history of Pulmonary TB 8 years back and was treated for 6 months for the same.

On examination she was poorly built with pulse rate of 83bpm, respiratory rate of 19 breaths/min and a normal BP of 130/90 mm Hg. The clinical examination of respiratory system revealed absence of chest movements, hyperresonance note on percussion and absence of breath sounds on the left side of the chest. Chest CT showed large multiple emphysematous bullae in left lung occupying the entire lung suggestive of vanishing lung syndrome with an emphysematous bulla in the right upper lobe with multiple patchy areas of consolidation in right lung with adjacent fibrosis suggestive of post pulmonary Koch's. (Figure 1, 2 & 3)

Ultrasonography did not show any cystic changes in the abdominal viscera and the facility for estimation of serum $\alpha 1$ antitrypsin was not available.

Discussion

Vanishing lung syndrome starts with appearance of emphysematous bullae where alveolar walls disintegrate and large air spaces are created leading to lung atrophy and progression occurs till death due to respiratory failure. The basic pathogenesis is not exactly known, however it has been attributed to possible

respiratory myositis.^[1,2] There is evidence of diaphragmatic weakness and the condition is associated with systemic lupus erythematosus in one third of cases.^[2-4] Lung function tests show a restrictive pattern. The radiographic criteria for vanishing lung syndrome were proposed in 1987 and they include giant bullae in one or both upper lobes occupying at least one third of the hemithorax and compressing surrounding parenchyma. Air–fluid levels within bullae are uncommon and raise the question of bacterial superinfection. Lung-volume–reduction surgery is considered for selected patients with vanishing lung syndrome after assessment of exercise capacity, pulmonary-function testing and smoking cessation. The disorder needs to be differentiated from few other similar conditions. Infantile lobar emphysema is an obstructive distension of one lobe in an infant, often giving rise to severe dyspnea and necessitating surgical removal. There is a strong male preponderance and about half have congenital cardiac abnormalities and left upper or middle lobe is usually involved and the disorder manifests in less than 6 weeks age. In lobar emphysema with bronchial atresia, there is complete atresia of the proximal bronchus, which may be patent peripherally. The left upper lobe is transradiant and hypoplastic.^[1] Macleod's syndrome comprises of unilateral emphysema of a lung or lobe due to localized bronchiolitis or bronchitis. The pulmonary artery on the affected side is often small and there is irregular dilatation of bronchi with failure to fill the peripheral airways. The condition often occurs six months to five years after viral bronchitis, or follows tuberculosis in the childhood.^[1] The presence of clinical and radiological features and restrictive pattern on pulmonary function testing favours the diagnosis of vanishing lung syndrome in our patient, who was not a candidate for lung volume reduction surgery and is under follow-up for future course and may need further evaluation.

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