A Rare Case of Vanishing Lung Syndrome

Amarjit Singh Vij*, Robert James**, Akashdeep Singh***, Amrinder Singh Dhaliwal****, Ajay Chhabra†, Kamaljeet Kaur Vij‡

Abstract
Vanishing lung syndrome (VLS) is a rare radiological syndrome in which the lungs appear to be disappearing on X-ray. It is a chronic, progressive condition usually affecting young male smokers and is characterised by giant emphysematous bullae, which commonly develop in the upper lobes. We describe here a rare case of 60-year-old male patient who had a history of chronic smoking for 30 years. He had been admitted in the hospital multiple times due to spontaneous pneumothorax, type 2 respiratory failure and infective exacerbations. He was earlier diagnosed having chronic obstructive pulmonary disease (COPD) with predominant emphysema on the basis of his history and chest X-ray findings. Eventually, his CT chest revealed the diagnosis of giant bullous disease/vanishing lung syndrome. He had been surviving with his little lung tissue for about 10 years. No such case has been reported in the literature so far. He was attended last on 12th October, 2009 in medical outdoor of Christian Medical College and Hospital, Ludhiana by the first three authors. Thereafter, the patient was not traceable.

Introduction
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 hemithoraces.¹ The radiographic criteria for this syndrome defined by Roberts et al² 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. Vanishing lung syndrome (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.³

Most patients are young male,⁴⁵ the risk factors are smoking, alpha-1-antitrypsin deficiency, and marijuana abuse.⁶⁷ Extensive paraseptal emphysema coalesce to form giant bullae, compressing the normal lung parenchyma and often displacing it centrally. The bullae range in size from a few centimetres to giant bullae nearly filling hemithorax, mimicking a pneumothorax. The giant bullae may remain asymptomatic for a long time, their progression may cause worsening dyspnoea. A major complication of VLS is pneumothorax. Infection of the bulla is also common. Computed tomography (CT) is an important tool for the diagnosis of this bullous disease.⁹¹⁰

Surgical resection of giant bullae is the treatment of choice, the indications include (1) spontaneous pneumothorax,¹¹ (2) infection,¹² or (3) dyspnoea,¹³ the best results are seen following limited bullectomy.¹⁴¹⁵ Thoracoscopic treatment of giant bullae is an effective alternative to conventional thoracotomy with minimal morbidity.¹⁶
Case Report

Mr. X, 50 years old, chronic smoker for 30 years and known case of chronic obstructive pulmonary disease was admitted with right sided pneumothorax and type 2 respiratory failure, first time in March, 2000 in CMCH, Ludhiana. He was put on ventilator and thoracotomy done. He recovered after one week. He was discharged on inhaled glucocorticoid and bronchodilator combination and empirical anti-tubercular treatment for one year. He was counselled to stop smoking. In December, 2001, he developed right sided lower lobe pneumonia which resolved with intravenous (I/V) antibiotics. In January, 2002, he developed diabetes mellitus type-2. In July, 2002, he had left sided pneumothorax and recovered with conservative management. He did well for about three years except occasional episodes of mild to moderate airflow obstruction. In December, 2005, he developed systemic hypertension. He developed left sided pneumothorax and required thoracotomy again in May, 2007. In November, 2007, he came with type II respiratory failure and put on ventilator but unfortunately, he developed left sided pneumothorax. He recovered with thoracotomy. Then, he had acute infective exacerbation in December, 2007 and managed with I/V antibiotics. Contrast enhanced CT chest was done which revealed findings of vanishing lung syndrome (Figures 1, 2, 3, 4, 5, 6). Echocardiography revealed moderate tricuspid regurgitation (TR) with right ventricular systolic pressure (RVSP) of 45 mmHg. He was discharged after recovery. Then, he got admitted in March, 2009 with left sided pneumothorax and recovered with thoracotomy. He developed infective exacerbation again in October, 2009 and got treated. He was doing well when last seen in medical outpatient department on 12th October, 2009.

Discussion

This patient got admitted first time with spontaneous pneumothorax which is a common complication of COPD. The chest X-ray showed right sided pneumothorax and emphysematous lung on left side,
VLS was not suspected that time. In his subsequent admissions also, the diagnosis of VLS was not made till his CT chest done in December, 2007 made it clear. CT chest revealed multiple emphysematous bullae bilaterally ranging in size from 1 cm to as large as 14 cm. This is in close agreement with the findings in a study on series of patients with VLS done by Stern et al.3 Making the diagnosis of VLS in this case with CT chest highlights the importance of computed tomography in diagnosing this entity as reported earlier.9,10

In contrast to younger age of the patients of VLS reported in the literature,4,5 this patient was 57 year old at the time of diagnosis of VLS by CT chest and 50 year old when he had pneumothorax first time. He might be having asymptomatic emphysematous bullae at his younger age. He had history of smoking as risk factor reported but he was not tested for alpha-1-antitrypsin deficiency.

The interesting feature of this case is that he survived multiple episodes of pneumothorax and respiratory failure which is unique and no such case has been reported so far in the searched literature. He was admitted ten times in the period of about 10 years and developed pneumothorax five times, 4 times spontaneous and once iatrogenic due to mechanical ventilation. Distinction between the pneumothorax and bullae may be difficult on X-ray films, and a chest tube may be placed into the bulla by mistake.13 MacDuff A et al report that the unintentional placement of a chest tube into a giant bulla can cause iatrogenic pneumothorax, haemothorax, haemorrhagic shock or death.12 No such event occurred in our case and every time he recovered successfully after thoracotomy.

**Conclusion**

Vanishing lung syndrome is an important rare entity occurring mainly in young male who smokes, abuse marijuana or have alpha-1-antitrypsin deficiency. It can occur in non-smokers also. We should consider this entity in such patients whose chest X-ray gives the appearance of disappearing lungs or very large emphysematous bullae. Suspected patients must get CT chest which is the investigation of choice to diagnose this syndrome. With timely diagnosis of VLS, the patient can be referred to a pulmonologist/cardiothoracic surgeon for appropriate preventive/curative bullectomy which may help the patient to lead a better life with less morbidity.

**References**