

## VANISHING LUNG SYNDROME: A RARE ENTITY

**Khanduri S, Bhagat S, Shokeen P, Usmani T**

Department of Radiodiagnostics, Era's Lucknow Medical College, Uttar Pradesh, India

### **Abstract**

Idiopathic giant bullous emphysema or vanishing lung syndrome is a very rare condition associated with severe forms of emphysema in which the lungs appear to be disappearing on radiograph. It is a progressive condition which usually affects young male smokers and characterised by the presence of giant emphysematous bullae in lungs asymmetrically. The authors hereby describe a rare case of an 18 year old female non smoker, presented with a history of chest pain of two weeks duration. Her chest radiograph showed large bullae in both lungs, more on the left. HRCT thorax confirmed giant bullae in both upper, middle, lingula and apical segments of lower lobes compressing the surrounding lung parenchyma. Diagnosis of idiopathic giant bullous emphysema or vanishing lung syndrome was made based on imaging, for which the patient underwent surgical resection.

**Keywords;** Idiopathic, Giant Bullae, Emphysema

**Corresponding Author:** Bhagat S

**Copyright:** Bhagat S ([dr.sauravbhagat@gmail.com](mailto:dr.sauravbhagat@gmail.com))

## Introduction

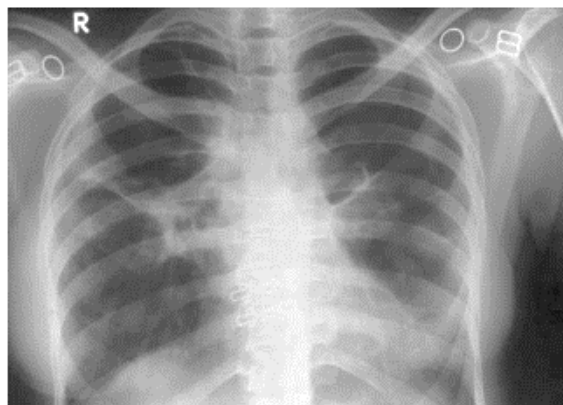
Emphysematous disease is one of the commonest respiratory diseases encountered in the hospital setting. Idiopathic giant bullous emphysema or vanishing lung syndrome (VLS)) is characterised by giant emphysematous bullae, which occupy at least one-third of the hemithorax in an asymmetric manner and is mostly seen in the upper lobes. It is a progressive condition that is also associated with several forms of emphysema. 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<sup>1</sup>. 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 radiograph<sup>2</sup>.

Most of the patients of VLS are young males<sup>3</sup>. The risk factors are smoking, typically marijuana abuse, alpha-1-antitrypsin deficiency, systemic diseases like Marfan’s syndrome or Ehlers- Danlos syndrome<sup>4,5</sup>. Extensive paraseptal emphysema coalesce to form giant bullae which compress the normal lung parenchyma often displacing it centrally. The bullae vary in size from a few centimetres to giant bullae which nearly fill the hemithorax. Computed tomography (CT) is an important investigation for the diagnosis of this bullous disease. Surgical resection of giant bullae is the treatment of choice<sup>5,6</sup>.

This is a rare case report of vanishing lung syndrome in an eighteen yearold female without known risk factors as described previously.

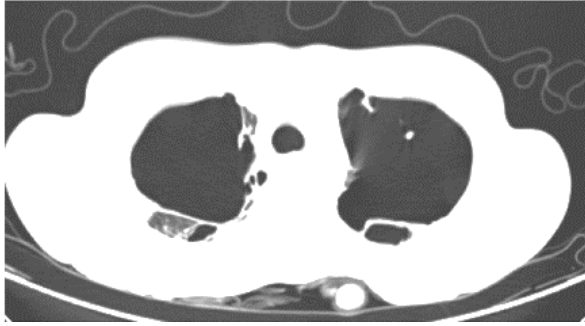
## Case Report

An 18 year old female non smoker presented with a history of chest pain of two weeks duration and this was her first presentation to a hospital. On examination she appeared to be of average built. She was afebrile, pulse rate was 90 per minute, respiratory rate was 20 per minute and blood pressure was normal. On respiratory tract examination, the chest appeared bulging. Percussion note was hyper resonant and the breath sounds were diminished on both sides. Rest of the systemic examination was normal. Laboratory tests including full blood count, complete metabolic profile, cardiac enzymes, Alpha-1 antitrypsin , and electrocardiogram were within normal limits.

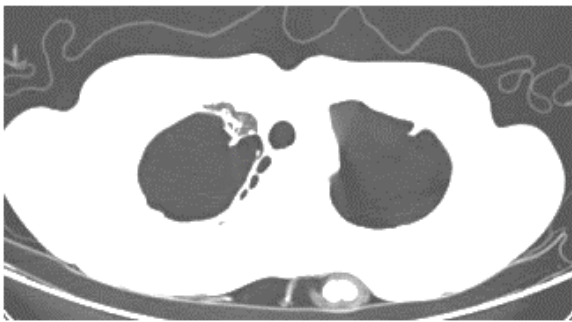


**Figure 1:** Chest X- ray (PA view ) - bullae in both upper zones and left mid zone

Her chest radiograph showed bullae in both lungs, predominantly upper zones (Figure 1). On HRCT thorax there were giant bullae in both lungs involving upper lobes occupying over 1/3 of the lung fields, middle and apical segments of both lower lobes (Figure 2). They were compressing the surrounding lung parenchyma (Figure 3). Diagnosis of idiopathic giant bullous emphysema or vanishing lung syndrome was made based on clinical features and imaging characteristics.



**Figure 2:** HRCT - multiple bullae in upper zones with intervening septae



**Figure 3:** HRCT - giant bullae in B/L upper lobes compressing the residual lung parenchyma

There was no evidence of pneumothorax. No fluid levels were seen in bullae to suggest superadded infections. Our patient underwent successful resection of bullae after careful assessment by the cardiothoracic surgeon and showed no residual symptoms on subsequent follow up.

## Discussion

Vanishing lung syndrome is also known as type I bullous disease or primary bullous disease of the lung with evidence of emphysema associated with large bullae, which commonly involves upper lobes and occupy around one-third of the hemithorax<sup>2</sup>. It is a progressive condition that is associated

with emphysematous changes, most commonly paraseptal emphysema<sup>3</sup>.

The radiographic criteria for diagnosis of vanishing lung syndrome, as defined by Roberts and colleagues, include the presence of giant bullae in one or both upper lobes, occupying at least one third of the hemithorax and compressing surrounding normal lung parenchyma. It is a chronic condition that mainly affects young male smokers<sup>3</sup>.

Most important complication of VLS is pneumothorax, in which the patient develops deterioration of respiratory function. Another common complication is infection of the bullae. It can eventually lead to respiratory failure<sup>3</sup>. HRCT is used to assess the extent of the disease and to determine suitability for lung volume reduction surgery. On HRCT, bullae are predominantly found in subpleural location, usually asymmetric with concomitant foci of paraseptal and centrilobular emphysema. Paraseptal emphysema is the predominant chest radiographic and HRCT findings which was found in our patient as well. HRCT also helps in assessing complications such as infected cysts, pneumothorax, bronchiectasis or pulmonary hypertension<sup>3</sup>. Our patient did not have any of these complications.

Surgical removal of bullae is the treatment of choice and it results in significant functional improvement<sup>8</sup>. In addition the patients are strictly advised to stop smoking. Our patient also underwent successful resection of the bullae with no residual symptoms on follow up.

This case is different from others described in the literature as it has been diagnosed in a young female without known risk factors.

## Conclusion

Vanishing lung syndrome is a rare disease which is mostly diagnosed when the patients are severely symptomatic. High resolution chest CT is the investigation of choice in these patients which should be immediately done if the patients show the features on chest radiograph. After diagnosis the patients should be referred to pulmonologist or cardiothoracic surgeon where treatment can be planned and the patients are relieved of their symptoms and the complications would be minimised.

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