



Review Article

Vanishing lung syndrome: A Systemic Review

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ABSTRACT:

Vanishing lung syndrome is a rare condition, also known as idiopathic giant bullous emphysema. It is commonly seen in young smokers, alpha 1 antitrypsin deficiency and in persons with Marijuana abuse. It is often confused as pneumothorax and ICD being placed. It is progressive disease and signs and symptoms vary from asymptomatic presentation to exercise intolerance, worsening dyspnoea and spontaneous pneumothorax. CT scan of chest is the investigation of choice and limited bullectomy has shown better outcome.

Key Words: Vanishing lung, Pneumothorax, Bullae, Emphysema, Smoking, marijuana.

INTRODUCTION:

Vanishing lung syndrome was first described in 1937 by Burke [1] in a 35 year old male patient with history of progressive dyspnoea and giant bullae that occupied two third of both hemithorax. Vanishing lung syndrome is idiopathic giant bullous emphysema, characterized by large bulla occupying at least one third of the hemithorax [2] which on chest X-ray lung appears to be disappearing [3]. It is also known as Primary bullous disease of the lung or Type-1 bullous disease.

DISCUSSION:

Prevalence is not known but unilateral hyperlucent lung is not an uncommon radiological finding [4]. Bullae on chest X-ray defined as sharply demarcated areas of emphysema, exceeding 1cm in diameter with wall thickness of less than 1mm [5]. Giant bullae defined as one or more bullae that occupy more than one third of hemithorax [6]. The bullae in VLS are asymmetrical, sub pleural and predominantly involving upper lobes. It is also seen at bases and many patients predominantly have large para septal emphysema [7].

Radiological criteria for diagnosing VLS have been proposed by Robert's and colleagues [8]. These include giant bulla in one or both upper lobes, occupying at least one third of hemithorax and compression of surrounding normal lung parenchyma. Histopathology of bullae [9] has shown areas of emphysema interspersed with normal lung parenchyma.

Vanishing lung syndrome (VLS) commonly seen in males [10,11] and have been reported in both younger [12] and older age group [13]. The size of the bulla varies from few centimeters to giant bullae nearly filling hemithorax. Risk factors includes Smoking [12,14], Alpha 1 antitrypsin deficiency [15] Marijuana abuse [11] and connective tissue diseases such as sarcoidosis and SLE [16]. It has also been reported in non smokers [7]. In case of Marijuana abuse it is seen even in earlier age group [17,18].

Most patients are asymptomatic and usually diagnosed accidentally during routine chest X-ray for some other reason.

The bullae formation was initially thought to be due to ball-valve mechanism (air entering the lesion but could not escape) but recent evidence suggests that intra luminal pressure of bulla is negative, similar to pleural pressure. It is being ventilated during inspiration and air trapping occurs due to failure of elastic recoil.

This is a progressive condition and patients usually present with worsening dyspnoea. In case of spontaneous rupture of bulla, they present with chest tightness, shortness of breath and cold sweats. On examination these patients will have decreased breath sounds over the involved hemithorax and with hyper resonant note on percussion. Differential diagnosis includes Pneumothorax, Emphysema, Large bullae, airway obstruction and radiological artifact.

Initial investigation includes complete blood picture with differential count, CXR, ABG, SpO₂, ECG, BNP, Cardiac enzymes, Urine toxicology, PFT and HRCT chest. Definitive investigation is HRCT Chest [19,20]. Uses of HRCT chest in these

patients include, to quantify extent and distribution of the bullous disease and to look for any coexisting condition such as infected cyst, bronchiectasis, pulmonary artery enlargement and pneumothorax [19]. CT chest usually show extensive para septal emphysema coalescing into giant bullae [3,20].

Treatment is mainly surgical. Indications for surgical removal of bulla are Spontaneous pneumothorax [21], Infection of the bulla [22] and worsening dyspnoea [23]. Standard thoracotomy and video assisted thoracoscopic surgery are the two surgical options without much difference in outcome among these surgeries [24].

Limited bullectomy has shown some good outcome [25,26]. Thoracoscopic treatment has been used to reduce morbidity [27]. Bullectomy has shown only short term improvement in dyspnoea, gas exchange, pulmonary function and exercise capacity. More significant VLS has best results. These benefits have been declined after 3 years [28] with persistence of FVC (Forced vital capacity) after 10 years [29] where as decline in FEV₁ and dyspnoea grading.

COMPLICATIONS:

Include spontaneous pneumothorax [13] and secondary infection [20]. Accidental placement of ICD (Intercostal device) in case of suspected pneumothorax can cause hemothorax, hemorrhagic shock or death.

CONCLUSION:

Vanishing lung syndrome is often confused as pneumothorax since it is a rare condition and routine chest X-ray cannot differentiate between pneumothorax and giant bulla (VLS). Accidental placement of ICD can cause iatrogenic pneumothorax, hemothorax or death. Hence high suspicious of Vanishing lung syndrome along with HRCT chest can help in definitive diagnosis along with limited bullectomy and quitting of smoking can reduce morbidity and mortality.

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