Vanishing Lung Syndrome, or Idiopathic Giant Bullous Emphysema, with Pneumothorax, and Subcutaneous Emphysema in a 58-Year-Old Female Smoker with Chronic Obstructive Pulmonary Disease

ABEFG 1
Haris Sohail

ABEFG 1
Yassine Kilani

BEFG 2
Julieta Osella

BEG 3
Ashna Syeda Fatima Kamal

BEFG 1
Barkha Kumari

EFG 1
Daniel Emnet Keftassa

EG 1
Mubarak H. Yusuf

EG 1
Mohammad Aldiabat

EG 1
Ali Horoub

EG 1
Shekar Murthy

Corresponding Author: Haris Sohail, e-mail: sohailh@nychhc.org

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Objective: Unusual clinical course

Background: Vanishing lung syndrome (VLS), also known as idiopathic giant bullous emphysema, is a rare manifestation of chronic obstructive pulmonary disease (COPD) and usually occurs in middle-aged smokers. This report presents a 58-year-old female smoker with COPD and VLS who presented with spontaneous pneumothorax. The pneumothorax was managed with a chest tube and was later complicated by massive subcutaneous emphysema.

Case Report: A 58-year-old woman with a past medical history of long-term smoking and COPD presented with worsening shortness of breath. Upon initial evaluation, she had tachypnea and hypoxia (SpO₂ 93%). Chest radiography revealed a new right-sided pneumothorax on top of extensive bullous disease, which the patient already had. The drainage of the pneumothorax was successful with a pigtail catheter. However, during the following night, after insertion of the pigtail catheter, the patient developed massive subcutaneous emphysema, which was confirmed with imaging. The patient remained hemodynamically stable, and diffuse subcutaneous crepitus was present on examination. The pigtail catheter was repositioned, resulting in complete resolution of the subcutaneous emphysema in the following 2 weeks.

Conclusions: This case highlights the importance of a timely diagnosis and management of the possible presentations and complications of VLS. Complications such as pneumothorax are life-threatening and require urgent management, taking precedence over the curative treatment for VLS, surgical bullectomy.

Keywords: Emphysema • Pulmonary Bullae Causing Pneumothorax • Subcutaneous Emphysema

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1 Department of Internal Medicine, Lincoln Medical Center, Bronx, NY, USA
2 Department of Pulmonary and Critical Care, Spectrum Health – Michigan State University, Grand Rapids, MI, USA
3 Department of Internal Medicine, Springfield Memorial Hospital, Springfield, IL, USA
**Background**

Vanishing lung syndrome (VLS), also known as idiopathic giant bullous emphysema (GBE), is a rare form of irreversible damage to the pulmonary parenchyma resulting in giant bullae. Patients with this condition have a long history of cigarette smoking and/or marijuana use and chronic obstructive pulmonary disease (COPD) [1]. Other risk factors include alpha-1 antitrypsin deficiency, HIV infection, Ehlers-Danlos syndrome, and sarcoidosis. Presenting features include progressive dyspnea, declining exercise tolerance, hypoxia, and decreased breath sounds. Complications include pneumothorax, subcutaneous emphysema, and respiratory failure [2-4]. High-resolution computed tomography scan (HRCT) is the criterion standard for the diagnosis of VLS and its complications. VLS, or GBE, is defined by the presence of giant bullae, involving one or both upper lobes of the lungs, occupying at least one-third of the hemithorax, and compressing the surrounding parenchyma, with mediastinal contralateral displacement [3,4].

We present a rare case of VLS in a 58-year-old woman with a history of smoking and COPD who presented with worsening shortness of breath due to a new spontaneous pneumothorax. It is challenging to differentiate between pneumothorax and bullae in patients with VLS, as described by Yousaf et al [4], because one can mimic the other. Our case describes the importance of timely diagnosis and management of complications that can arise in patients with VLS.

**Case Report**

**Chief Concerns**

A 58-year-old woman with a past medical history of long-term smoking (60 pack years), COPD (on 2 L home oxygen), bullous emphysema, obstructive sleep apnea, type 2 diabetes, and hypertension presented with a 1-day history of worsening shortness of breath, which was not relieved by her oral prednisone and as-needed inhaled albuterol. She denied any chest pain.

**Physical Examination**

On initial examination, her pulse rate was 96 beats per min, respiratory rate was 22 breaths per min, pulse oximetry was 93% with supplemental oxygen via nasal cannula at 2 L per min, and temperature 36.9°C. Her body mass index was 56.93 kg/m². The patient had anxiety and tachypnea, with prolonged expiration. Without accessory muscle use, the trachea was midline, with decreased breath sounds in all lung zones, which was more prominent on the right side. There was no cyanosis or peripheral edema. The rest of the examination was unremarkable.

**Laboratory Workup**

Her arterial blood gas showed acute on chronic hypoxic and hypercapnic respiratory acidosis, with an arterial pH of 7.37 (range: 7.35-7.45), PCO₂ of 54.5 mmHg (range: 33-46 mmHg), oxygen saturation of 98% (range: 95-98%) on 2 L, and bicarbonate level of 31.7 mmol/L (range: 22-29 mmol/L). Pro-BNP and inflammatory markers (procalcitonin) were in the normal range.

![Figure 1](image.png)  
*Figure 1. Chest radiography of (A) previous admission and (B) day 1 of the current admission. Extensive bullous disease of the right upper lobe is seen in both images (blue star), and is associated with a new-onset small right-sided pneumothorax (red arrow in image B).*
Figure 2. High resolution computer tomography (HRCT) with (A, B) axial and (C) coronal views on day 3, showing a large right-sided pneumothorax (red arrow) and a large bulla at the superior segment of the right upper lobe (blue star), with total collapse of the right middle and the right lower lobes (blue arrow). There is a right to left mediastinal shift (red star).

Imaging

The initial chest radiography (CXR) showed a new right-sided pneumothorax, with extensive bullous disease, especially in the apex (Figure 1A), similar to a previous CXR (Figure 1B). To further characterize the lesions, a chest CT scan was obtained and showed a new large right-sided pneumothorax, with collapse of the right middle and lower lobe and a large right-sided bulla in the superior segment of the lung (Figure 2).

Management and Prognosis

On day 3 of admission, following confirmation of pneumothorax on HRCT scan, a pigtail catheter was placed in the right fifth pleural space, and drainage was initiated. A repeat CXR showed complete resolution of the pneumothorax (Figure 3). However, the following night, the patient was observed to have dysphonia and difficulty in swallowing, with swelling of the face. She was hemodynamically stable, with no respiratory distress. On examination, she was found to have subcutaneous
crepitus and a positive Hamman’s sign, with an extensively swollen face (Figure 4A). A repeat CXR confirmed extensive subcutaneous emphysema (Figure 4B), which was confirmed on a repeat chest HRCT (Figure 4C, 4D). The most likely causes of the subcutaneous emphysema in our patient were spontaneous rupture of the bullae or malpositioning of the pigtail catheter. Therefore, the pigtail catheter was repositioned for management of the pneumothorax and subcutaneous emphysema. The patient stayed in the Medical Intensive Care Unit with close monitoring of respiratory status by CXR. On the following days, the subcutaneous emphysema resolved gradually, and the pigtail catheter was removed on day 14. A CXR prior to discharge showed complete resolution of the subcutaneous emphysema, with stable large blebs in the right upper segment of lung (Figure 5), consistent with her prior imaging.

**Discussion**

VLS, or GBE, is a rare form of irreversible damage to the lung parenchyma resulting in giant bullae. It poses a great challenge to clinicians to differentiate a new pneumothorax from previous giant bullae. Risk factors include a long history of smoking, marijuana use, COPD, and alpha-1 antitrypsin deficiency [1]. VLS usually affects patients over 45 years old; however, in marijuana users and patients with alpha-1 antitrypsin deficiency, it tends to have an earlier onset [1,5,6]. Patients typically present with progressive dyspnea, decreased exercise tolerance, hypoxia, and decreased breath sounds, bilaterally. Any acute deterioration in the respiratory function associated with chest pain should prompt physicians to evaluate for a pneumothorax. Other complications
include respiratory failure and subcutaneous emphysema, as seen in our patient [2-4].

VLS is defined by the presence of GBE, predominantly involving 1 or both upper lobes of the lungs, occupying at least one-third of the hemithorax, and compressing the normal surrounding parenchyma [4]. The diagnosis is confirmed with CXR with or without HRCT. CXR findings include sharply demarcated areas of hyperlucency. HRCT gives a better anatomical description and can be helpful in diagnosing bullae [7,8], as in our case. HRCT findings include multiple large bullae ranging from 1 to 20 cm [3]. HRCT is also useful to determine the etiology of VLS, assess for coexisting lung diseases (infected cysts, bronchiectasis, pneumothorax), differentiate from pneumothorax, and perform preoperative assessment [7-10]. HRCT should be done urgently in case of the clinical suspicion of a pneumothorax, especially with a negative CXR [7].

Urgent management of life-threatening complications such as pneumothorax takes precedence over curative treatment of VLS. The management of VLS is typically surgical. Bullectomy via thoracotomy or video-assisted thoracoscopic surgery is the mainstay of therapy, and nonsurgical candidates can benefit from intrabulosal adhesion pexia [11], endobronchial valve insertion on transbronchial deflation. Surgical treatment of bullous lung disease has been shown to improve dyspnea, quality of life, and pulmonary function tests (FEV1, FVC, and FEV/FVC) and to decrease the risk of a pneumothorax. However, there is no proven benefit on mortality [12]. Giant bullectomy has been shown to provide functional improvement in the early postoperative period and for up to 3 years; however, this benefit declines with time [11,13]. At long-term follow-up, the FEV1 and FVC were found to decline to near-baseline values [14].

Many authors have previously explained the challenge to differentiate between a pneumothorax and a bulla. Management of these entities is different and, a delayed diagnosis can be detrimental for the patient. Yousaf et al [4] described an interesting case in which VLS presented like a pneumothorax and, only after CT scan, was the patient correctly diagnosed with VLS and did not require any invasive intervention, which is usually needed for a pneumothorax. Similar cases have been presented by authors, in which VLS presented like a pneumothorax and patients underwent invasive interventions that did not help the patient [15,16].

Conclusions

This case highlights the importance of a timely diagnosis and management of the possible presentations of VLS. Complications such as a pneumothorax are life-threatening and require urgent management.

Declaration of Figures’ Authenticity

All figures submitted have been created by the authors who confirm that the images are original with no duplication and have not been previously published in whole or in part.

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Figure 5. Day 15, chest radiography showing complete resolution of the subcutaneous emphysema and pneumothorax, with stable large blebs in the right upper lobe of the lung (red arrow).
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