

Vanishing Lung Syndrome: A Brief Report

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Case Report

A 50 year old smoker male, presented with complaints of insidious and progressive shortness of breath for three years and cough with expectoration for one year. Patient gave history of smoking 20 bidis/day for the last 35 years. On physical exam, blood pressure was 130/86, heart rate-98/min, respiratory rate-30/min, and O₂ Sat. of 90% on room air. Examination of the chest revealed hyper resonance to percussion in both lungs along with ronchi and decreased apical breath sounds. Examination of all other systems was unrevealing. Chest radiography showed large bullae in the upper zone of right lung and lower zone of left lung respectively (Figure 1). Spirometry revealed an obstructive pattern and a decreased DLco. Computed tomography of the chest showed the enormity of the bullous disease in both hemithoraces (Figures 2-4). Patient's alpha-1 antitrypsin levels were normal. USG of the abdomen revealed no abnormality. On the basis of his clinical and radiological findings he was diagnosed as a case of "vanishing lung syndrome".



Figure 1: X-ray chest PA view showing emphysematous chest and bullae in both Hemithoraces.

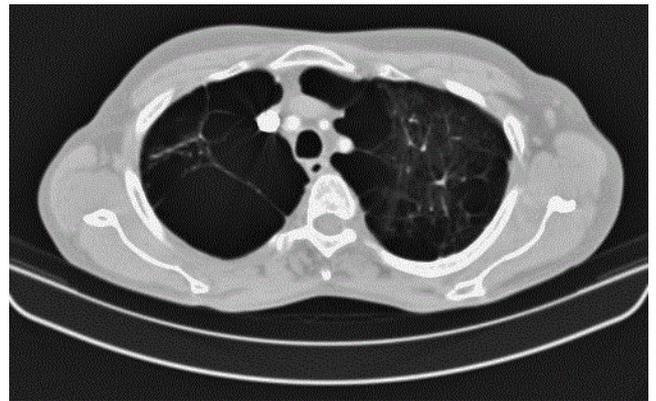


Figure 2: Cect chest showing large bullae in the right upper lobe.

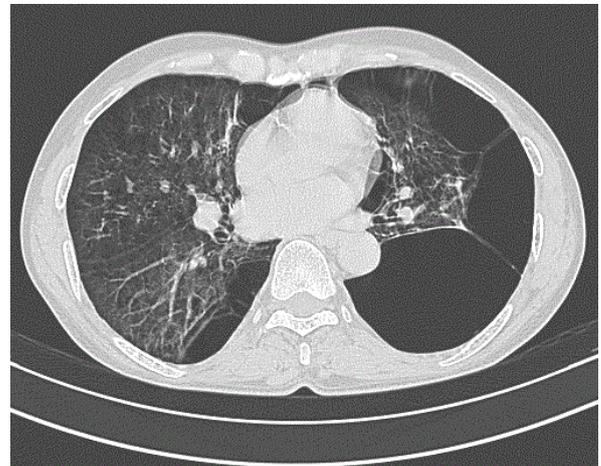


Figure 3: Cect chest showing large bullae in the left lower lobe.

"Vanishing lung syndrome" (VLS), also termed as primary bullous disease of the lung, giant bullous emphysema (GBE), or Type I bullous disease, was first described by burke in 1937 [1]. It usually occurs in association with chronic obstructive pulmonary disease and is characterized by the presence of large progressive bullae that occupy a significant volume of one or both hemithoraces and are usually asymmetrical [2]. According to the radiographic criteria for vanishing lung syndrome proposed in 1987, it is defined as 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 [2-4]. Surgery in GBE is a valid treatment option for a specific group of

patients with emphysematous bullae who present symptoms like chest pain or spontaneous pneumothorax.

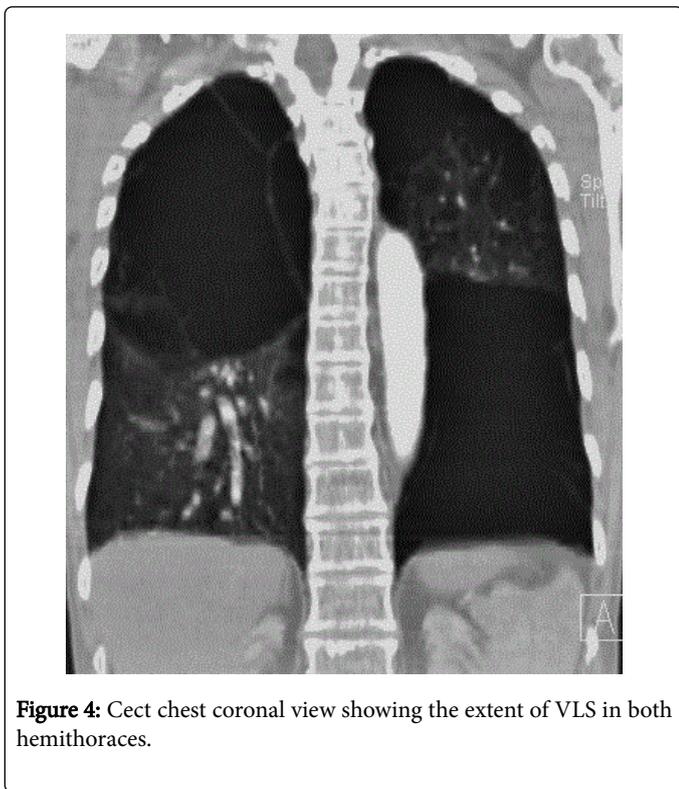


Figure 4: Cect chest coronal view showing the extent of VLS in both hemithoraces.

Different thoracoscopic techniques which were used in the past, such as thoracoscopic bullous end loop ligation, intra-cavitary bullous drainage, laser bullae ablation and bullous fibrin glue treatment, however all with variable success [5-8]. A non-excisional treatment option is the Brompton technique, first described by Monaldi. In this technique the bulla is drained percutaneously and a mushroom Foley catheter is left behind after septectomy and talcage and is put under suction for a few days after which the bronchocutaneous fistula closes spontaneously after removal [9].

In the last decade the application of VATS bullectomy with endoscopic staple resection is considered as a suitable and safe treatment of choice [10].

Operative mortality rates after bullectomy for localised bullous emphysema in patients with preserved lung function are low (from 0-2.5%). Morbidity is primarily related to prolonged air leak (53%), atrial fibrillation (12%), postoperative mechanical ventilation (9%), pneumonia (5%) and postoperative incisional pain [11].

Take home message

- Vanishing lung syndrome (VLS) is a progressive bullous disease characterised by large bullae that involve at least one-third of one or both hemithoraces.
- Pneumothorax is a potential complication in patients with VLS and an important disorder with which it can be mistaken for and wrongly treated.
- X-ray of the chest may not reveal the exact extent of the disease as in this case.

4 CECT of the chest is the investigation of choice to

- Confirm the diagnosis,
- Assess severity of the disease,
- Differentiate between pneumothorax and a large bulla,
- To plan treatment.

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