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Case Report

Vanishing Lung Syndrome – What Happens When you Stick a Drain into a Giant Bulla?

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Abstract

Vanishing lung syndrome, also known as Giant Bullous Emphysema, primarily affects young male smokers and is often misdiagnosed as pneumothorax. Detailed radiological workup aids in accurate diagnosis and appropriate management.

Case Report: We present the case of a 25-year-old female who experienced three episodes of hemoptysis and was initially misdiagnosed with right-sided pneumothorax based on clinical and chest radiographic findings. Subsequently, an intercostal drain was inserted on the right side at an outside hospital. Upon persistent right pneumothorax, the patient was referred to our hospital, where further imaging studies revealed giant bullae occupying the right hemithorax. Following comprehensive investigations, the patient underwent Video-Assisted Thoracoscopic Surgery (VATS) and bullectomy.

Conclusion: This case report underscores the significance of clinical and radiological assessment in achieving a timely diagnosis and appropriate management of vanishing lung syndrome, thereby minimizing potential complications.

Keywords: Giant Bullae; Pneumothorax; Video-Assisted Thoracic Surgery; Pulmonary Disease

Abbreviations

VATS: Video-Assisted Thoracoscopic Surgery; VLS: Vanishing Lung Syndrome; HRCT: High Resolution Computed Tomography; USG: Ultrasonography; DLCO: Diffusing Capacity of the Lungs for Carbon Monoxide; ICD: Intercostal Chest Drain; CXR: Chest X Ray; CT: Computed Tomography

Introduction

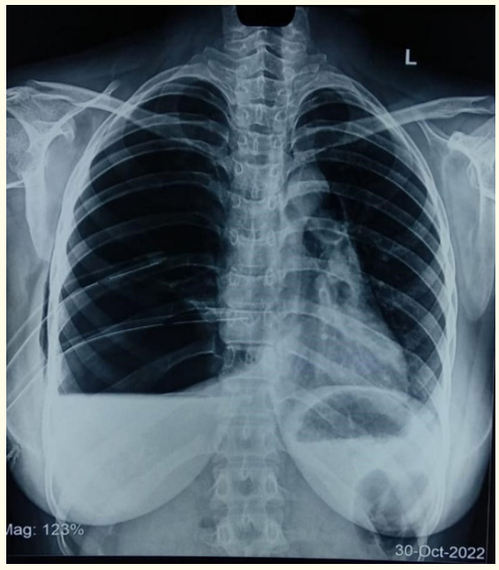
"Vanishing Lungs" has been described as early as 1937 in a 35-year-old man by Burke [1], who had respiratory failure, radiographic and pathologic findings consistent with giant bullae occupying two-thirds of bilateral hemithoraces. Robert., *et al*. [2] described the radiological criteria for Vanishing Lung Syndrome (VLS) in 1987 as the presence of giant bullous emphysema mainly involving either unilateral or bilateral upper lung lobes and occupying about one-third of the hemithorax or hemithoraces. These bullae compress the surrounding parenchyma of the remaining lung lobes and/or mediastinum. Usually, patients are young males with risk factors of smoking, alpha-1-antitrypsin deficiency, and marijuana abuse [3]. Pathogenesis of the disease involves the destruction of the alveolar walls forming subpleural blebs and into giant bullae [4]. VLS is usually misinterpreted as pneumothorax. Below we present a case of a young girl who presented with Vanishing Lung Syndrome on the right side. She was misdiagnosed as a case of massive pneumothorax and had 2 chest drains inserted at a treatment center. We describe our management of this patient.

Case Report

A 25-year-old female presented to the hospital with 3 episodes of hemoptysis. She underwent a chest roentgenogram and the treating doctors diagnosed her as having a case of massive right pneumothorax. She underwent right chest drain insertion. Followed by which there was persistent air leak with subsequent chest x-rays showing persisting pneumothorax. Hence, another chest drain was inserted into the right pleural space (Figure 1a). The patient subsequently was referred to us. On examination of the drain, both chest drains had a profuse air leak. HRCT chest revealed giant bullae with most of the right lung being collapsed and destroyed with hydro-pneumothorax on the right side and mediastinal shift to the left and a few small cystic spaces in the left lung parenchyma. The first chest drain was inside the giant bullae (Figure 1b). The second chest drain was within the pleural space. Bronchoscopy showed

normal bronchial tree development, but a segmental bronchus to the right middle lobe appeared missing. A lung perfusion scan showed almost no perfusion in the right lung, with normal perfusion in the left lung (Figure 2). Her alpha 1 antitrypsin levels were 192 mg/dl. Following a multidisciplinary team meeting, the patient was planned for surgical intervention, because of a persistent air leak. Subsequently patient underwent Right Uniportal VATS. Intraoperatively most of the right lung was destroyed, with relatively small right upper lobe and lower lobe remnant lung tissue. A large bulla arising from the right upper lobe, adhered to the chest wall with a chest drain inside was identified. Multiple smaller bullae were found adjacent to the large bulla. The remainder of the lung collapsed and had a thin cortex restricting it. Part of the diseased right upper lobe bearing the bullae as mentioned above including the large bulla has been excised with EndoGIA staplers (Figure 3). The thin cortex over the remaining lung tissue was removed, and the remaining lung tissue, although reduced in size, expanded after decortication. No air leak was noted. Post surgery, the patient made a successful recovery and was discharged home on the fourth post-operative day. Subsequently biopsy results indicated chronic inflammation with a giant cell reaction in the pleura. Lung specimens had sections revealed dilated alveolar spaces with focal architectural distortion due to underlying and peripheral fibrosis extensive hemorrhage and areas of fibrinoid necrosis with thick-walled proliferating blood vessels. A moderate chronic mononuclear inflammatory infiltrate composed of lymphocytes, plasma cells, histiocytes, and hemosiderin-laden macrophages deposition was noted. Additionally, bronchioles lined by ciliated columnar cells were observed as were a few cystic spaces lined by flat cuboidal cells. Focal bronchial cartilaginous and osteoid tissue was seen. No granuloma, atypia, or malignancy was seen. The patient continues in follow-up and is doing well, integrated back into daily life.

1a



1b

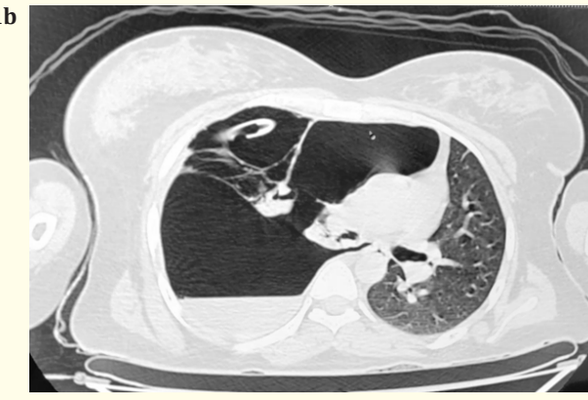


Figure 1a: Chest X ray PA view showing right sided hydropneumothorax with two chest drains insitu with mediastinal shift to opposite side.

Figure 1b: HRCT chest axial cut at the level of T4 showing right sided hydro- pneumothorax with mediastinal shift to the left with chest drain within the bullae, with underlying lung collapse. Left lung parenchyma appears normal.

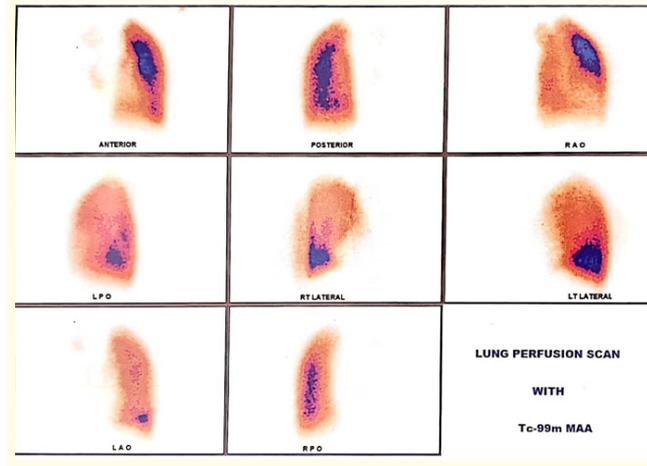


Figure 2: Lung Perfusion scan showing almost no uptake in the right lung with normal uptake in the left lung.

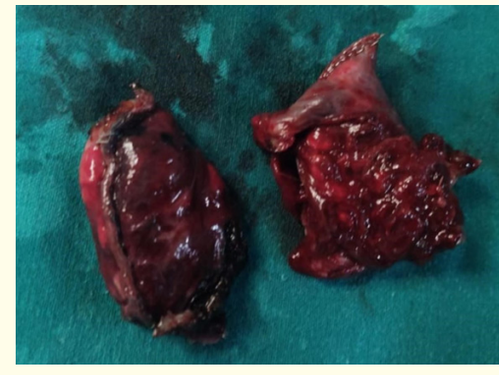


Figure 3: Wedge resection of the bullae bearing diseased right upper lobe lung tissue.

Discussion

VLS or giant bullous emphysema is a progressive bullous disease characterized by large bullae which involve up to one-third of either unilateral or bilateral hemithoraces [5]. Emphysema refers to the permanent enlargement of the airspace and destruction of alveolar walls distal to terminal bronchioles [6]. Whereas bullae are air-filled spaces within the parenchyma that are 1 cm or larger in diameter [7]. Bullae occupying more than 30% of the chest cavity are called giant bulla. Bulla originates within the emphysematous lung in 80% and 20% of normal lung tissue [8] (Table 1).

Table 1: WITZ and roeslin classification of giant bullae [8,9].

GROUP	
I	Bullae with normal underlying parenchyma, Para septal emphysema
II	Bullae with diffuse emphysema; bullae are a local exacerbation of diffuse pan acinar emphysema
III	Vanishing lung syndrome; entire lobe or lung replaced by bullae

The clinical features depend upon the size, extent, and type of bullae. As the formation of bullae is insidious patients may have no symptoms to symptoms such as cough, dyspnoea, or chest pain [9]. VLS can be due to an unknown cause or associated with conditions such as alpha- 1 antitrypsin deficiency, Ehler-Danlos Syndrome, Marfan syndrome, and risk factors such as intravenous drug usage, cigarette smoking, and marijuana use [10]**.** Diagnosis is usually radiological. Bullae appear as radiolucent thin-walled avascular areas with less than 1 mm thickness, hence may not be visible at times on chest roentgenogram which can cause misinterpretation as a pneumothorax [11]**.** On chest radiography, the shape of the lung parenchyma associated with a giant bulla is usually concave whereas convex in a pneumothorax [12]. Imaging with HRCT of the thorax shows a "double-wall sign," [13]. Whereas On USG absence of both "comet-tail" and "lung sliding" signs are suggestive of pneumothorax [14]. Giant bullae without symptoms are treated conservatively or by bronchial valves, lung volume reduction coils through bronchoscopy [11]**.** Indications for surgery in giant bullae include- 1) increasing bullae size, 2) pneumothorax, 3) infection within bullae, and 4) pulmonary insufficiency [15]. According to a study by Menconi., *et al*. VATS bullectomy with endoscopic staple resection is considered a safe treatment of choice [16]. The factors determining the postoperative period are the size of the bulla and the status of the underlying lung. The presence of almost normal DLCO in these patients indicates that the remaining lungs are not emphysematous and hence is a good prognostic marker for surgery [17].

In the present case, a young female was initially misdiagnosed elsewhere as having right pneumothorax and had chest drains placed in the right pleural space. The HRCT chest showed the presence of a giant bulla with collapsed remnant lung tissue with chest drain (ICD) within the bullae. The patient was taken up for Right Uniportal VATS surgery.

A similar case was reported by Khasawneh., *et al*. where a 44-year-elderly smoker whose chest X-ray (CXR) was diagnosed as bilateral spontaneous pneumothorax and underwent bilateral chest tube placement. Follow-up CXR showed no expansion. CT chest showed large bullae bilaterally consistent with vanishing lung syndrome (VLS). The patient then underwent a video-assisted thoracoscopy and bullectomy [5].

In another report by Ferraira., *et al*. where a 50-year-old male having a history of trauma to the chest with decreased bilateral breath sounds with hyper resonance and respiratory instability, chest drains bilaterally had been placed given suspicion of pneumothorax, but later the patient had subcutaneous emphysema with an air leak in bilateral chest tubes. CT chest revealed bilateral apical giant bullae. Negative pressure aspiration and changes in ventilator parameters failed to improve oxygenation thus right sided bullectomy was done. The air leak was resolved later VATS with right lung decortication was done as he developed empyema. But the patient succumbed due to infective complications [18].

Similarly, study reported by Sood., *et al.* [19]**,** where a 33-year-old old male presented with shortness of breath with risk factors of tobacco and marijuana abuse with CXR of right pneumothorax. Post-ICD chest X-ray remained unchanged. Upon CT chest giant bullous emphysema was present. Alpha 1 antitrypsin was negative. Thence diagnosis of vanishing lung syndrome was made. The patient was discharged with a plan of outpatient pulmonary rehabilitation with the possibility of bullectomy (Table 2).

Conclusion

"In conclusion, this case report underscores the significance of recognizing Vanishing Lung Syndrome (VLS) as a distinct clinical entity that may mimic a large pneumothorax, thus necessitating heightened awareness among treating physicians. Misdiagnosing VLS as a massive pneumothorax can inadvertently lead to complications, particularly when chest drains are inappropriately placed within the giant bullae. However, it is imperative to note that surgical interventions, notably Video-Assisted Thoracic Surgery (VATS), have demonstrated favorable outcomes in the management of VLS.

Table 2: The following table gives an insight into various case reports and the pattern of management of the patients based on the symptomatology, risk factors associated with the patient, and the presence of pneumothorax.

Case report	Gender	Age	Symptoms	Risk factors	Pneumothorax	Treatment
YU-TZU TSAO., *et al.* [20]	Male	44Y	Dyspnea, chest pain, cold sweats	Tobacco	Yes	Chest tube
KHASAWNEH., *et al.* [5]	Male	65Y	Dyspnea	Tobacco, cannabis, aids	No	Bullectomy
VAN BAEL., *et al.* [21]	Male	36Y	Dyspnea, chest pain	Tobacco	No	Bullectomy
WANG., *et al.* [22]	Female	19Y	Chest pain	Tobacco	No	Bullectomy
HUANG., *et al.* [23]	Male	59Y	Dyspnea, cough	None	No	Bullectomy
LIANG., *et al.* [24]	Male	35Y	Dyspnea, cough, chest pain	Tobacco	No	Bullectomy
CHEN., *et al.* [25]	Female	58Y	Dyspnea, cough	None	No	Bullectomy
ANILE., *et al.* [26]	Male	14Y	Dyspnea, chest pain	None	No	Bullectomy
JAISWAL., *et al.* [27]	Female	48Y	Decreased exercise tolerance	Tobacco	No	Bullectomy
SAEED., *et al.* [28]	Male	56Y	Dyspnea, chest pain	Tobacco, cannabis, aid	Yes	Heimlich valve
JUNIOR EG., *et al.* [18]	Male	50Y	Dyspnea, chest pain	Tobacco, cannabis	Yes	Chest tube, bullectomy
PRESENT CASE	Female	25Y	Hemoptysis	None	Yes	Chest tube, bullectomy

Our case report meticulously outlines the entire spectrum of this clinical scenario, starting with the misdiagnosis as a massive pneumothorax, the complications resulting from inappropriate chest drain placement, and the subsequent surgical approach that effectively addresses the compounded problem.

This situation serves as a reminder of the challenges that can arise in a physician's daily practice. We hope that this paper contributes to the collective knowledge and understanding of VLS, thereby enhancing patient care and outcomes in clinical settings".

Vanishing Lung Syndrome (VLS) is an entity that a treating physician needs to be aware of. It can often be misdiagnosed as a large pneumothorax and placing a chest drain into the giant bullae can lead to more complications. There are surgical options available for VLS, especially in the form of VATS, with good results. Our case report highlights all these starting from misdiagnosing VLS as massive pneumothorax, placing chest drains into it, and subsequent surgical solution to the compounded problem. This situation can arise in a physician's daily practice and we hope to provide a comprehensive way of managing the condition through this paper.

Conflict of Interest

No conflict of interest.

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