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Vanishing Lung Syndrome: A Rare Entity

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Abstract
Vanishing lung syndrome is characterised by giant bullae that compress the underlying lung. Differentiating between bullous emphysema and pneumothorax and choosing the right treatment modality can be challenging. The present report describes a case of a giant pulmonary bulla in a young male patient. A 20-year-old male was admitted with complaints of shortness of breath, chest pain, sweating, and tachycardia. Chest X-ray and chest CT images revealed the presence of giant bullae occupying the left hemithorax and pneumothorax. A chest tube was inserted percutaneously under local anesthesia into the pleural cavity and then we performed bullectomy using thoracoscopic surgery. Residual lung re-expansion yielded good postoperative results without complications. It can be very difficult to distinguish a pneumothorax from a giant bulla and preoperative assessment of the extent and distribution of the bullae is vital for the following procedures.
Keyword: Giant Bulla; Pulmonary Emphysema; Pneumothorax.

Vanishing Lung Sendrom: Nadir Bir Antite

Özet
Vanishing lung sendrom nadir görülen ve dev bül veya büllerin akciğerin büyük oranda kollapsına neden olduğu bir antitedir. Bu çalışmada solunum sıkıntısı, terleme, çarpıntı şikayeti ile acil servise başvuran 20 yaşında, radyografik olarak sağ hemitoraksın tümünü kaplayan dev bül ve çevresinde pnömotoraks saptanan bir erkek hasta sunuldu. Hastaya tüp torakostomi uygulandı ve daha sonra torakoskopik cerrahi ile bül eksizyonu yapıldı. Postoperatif dönemde komplikasyon gelişmedi. Akciğer ekspanse olan olgu postop 6. günü şifa ile externe edildi. Vanishing lung sendromunda en önemli sıkıntı bül, pnömotoraks ayrımının yapılmasında ve altta yatan akciğer parankiminin ne derece sağlıklı olduğunun değerlendirilmesinde yaşanılan zorluktur. Uygun vakalarda bül eksizyonu ile kollabe akciğerin ekspanse olur ve tam bir klinik düzelme sağlanır.
Anahtar Kelimeler: Dev Bül; Pulmoner Amfizem; Pnömotoraks.

INTRODUCTION

"Vanishing lung syndrome" (VLS) is characterised by one or more giant bullae filling almost all regions of a hemithorax while collapsing underlying lung tissues. PA defines a radiological entity in which lung tissue can hardly be seen in chest X-ray. Clinically and radiologically it is difficult to make the differential diagnosis between bullous emphysema and pneumothorax.

In this article, we present the case of a patient who was admitted with respiratory distress symptoms and was diagnosed with secondary "vanishing lung" with a giant pulmonary bulla in radiological assessment.

CASE REPORT

A 20-year-old male patient was admitted with shortness of breath that had been continuing for seven days, chest pain, sweating, and palpitation. The patient did not have any coughing and fever complaints. He had been smoking eight pack-year. Routine biochemical and hematological tests showed normal results. PA chest radiograph revealed increase in radiolucency in the right hemithorax, expansion in the intercostal space, flattening of the right hemidiaphragm and minimal mediastinal shift to the left hemithorax (Figure 1).

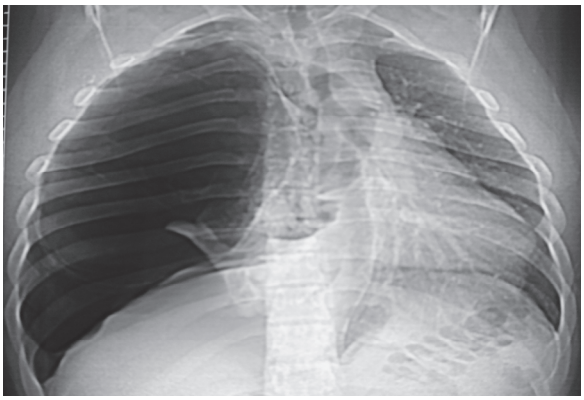

Figure 1. PA chest radiography image showing the hyperlucent view in the right hemithorax, expansion in the intercostal space, flattening of the right hemidiaphragm and minimal mediastinal shift.

In the thorax CT, we detected a giant bulla with accompanying minimal pneumothorax which filled the right hemithorax causing a nearly total collapse of normal lung tissues and mediastinal shift (Figure 2).

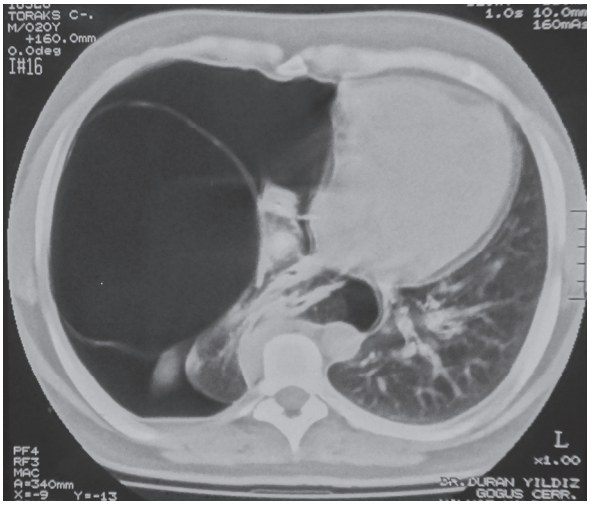

Figure 2. The thorax CT image showing a giant bulla and minimal pneumothorax covering almost the entire normal lung tissue in the right hemithorax and causing mediastinal shift.

We performed tube thoracostomy connected to closed drainage system under local anaesthesia. Although we achieved reduction in terms of respiratory distress and improvement in the mediastinal shift observed in the chest X-rays, the bullae maintained its integrity and the lung did not expand (Figure 3).

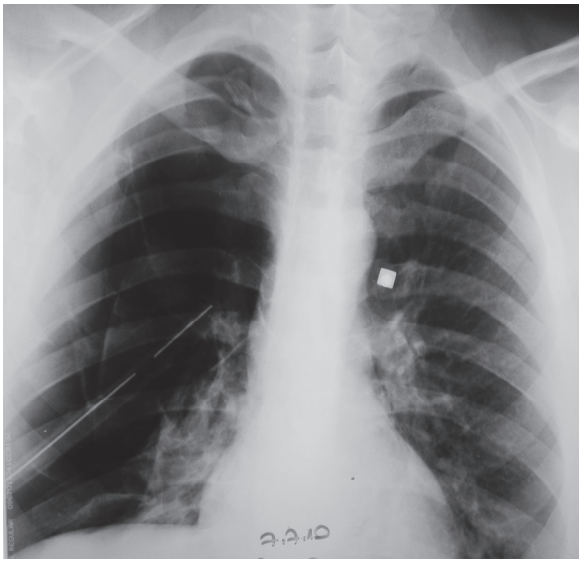

Figure 3. Following the tube thoracostomy, the chest X-ray shows that the bullae maintained its integrity and the lung did not expanse despite the improvement in the mediastinal shift.

Thereupon, we decided to perform bullectomy with video-assisted thoracoscopic surgery (VATS). All the symptoms improved in the postoperative period and the lung was fully expanded, radiologically (Figure 4).The patient was discharged on postoperative sixth day without further complications.

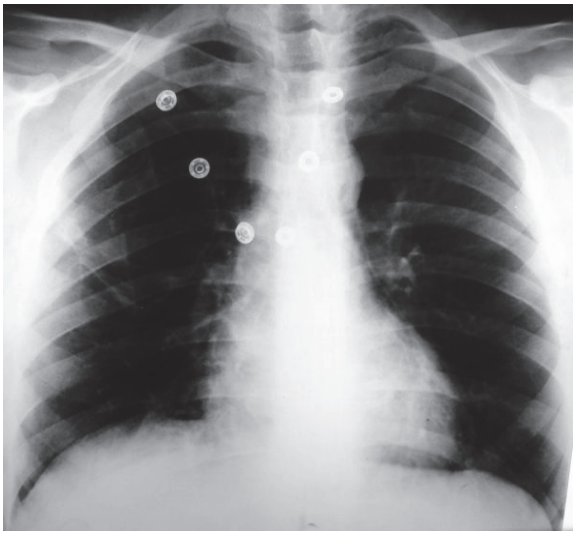

Figure 4. Fully expansed view of the lung in the postoperative period.

DISCUSSION

In addition to the categorisation about being single or multiple, bullous emphysema has also been classified according to whether it accompanies diffused emphysema. According to this classification group I is characterised by a single bullae on the normal lung tissue while group II is characterised by multiple bullae on the normal lung tissue. Groups III and IV define whether bullous lesions accompany diffused emphysema and obstructive airway diseases, respectively (1).

VLS was first defined as a distinct clinical entity by Burke in 1937 in a young patient with progressive dyspnea along side with bullous emphysema in the upper lobe (2). In subsequent publications, this phenomenon has mostly been described in young and middle-aged men as a single giant bulla or multiple bullae related pathology developing on the adjacent normal lung tissues under pressure (3-9). Some authors have used the term "vanishing lung" to refer to the radiographic appearance in which lung tissue is completely destructed due to progression of emphysema (1, 2, 10, 11). Stern has stated that this syndrome may occur along with various forms of emphysema (2).

VLS has been identified both in young smokers and non-smokers alike. Our 20-year-old patient also had a heavy smoking history of four years. In the majority of cases, there are no symptoms until the bulla causes compressive atelectasis and dyspnea by suppressing the normal parenchyma. The over-extension of the bullous lesion may bring about a clinical and radiographic picture that may be confused with tension pneumothorax in some cases (12, 13). In our case, the minimal pneumothorax along with accompanying compression of the giant bulla has resulted in tension pneumothorax.

As opposed to more uniformly diffused emphysema cases, PA chest radiographs refer to large peripheral bullae. The presence of a giant bulla occupying at least 1/3 of the hemithorax in one or both of the upper zones and lung tissue compression are regarded as the most important criteria in the diagnosis (2). Complex view in radiographic images and altered parenchymal limits can prevent the detection of an existing pneumothorax. Waitches et al. have stated that "double-wall sign" is an important indicator in identifying pneumothorax in these cases (11). On the other hand, complex radiological view may indeed lead to a false diagnosis of pneumothorax (3, 4). For this reason, many authors recommend that, before they plan chest tube thoracostomy with a diagnosis of pneumothorax based on conventional radiographic findings, one should assess thorax CT results in patients with detected or suspected large bullous lesions and acute respiratory distress (4, 5, 11). In our case, we did detect the presence of "double wall sign" in the chest CT scan and reassured the pneumothorax diagnosis before the tube thoracostomy intervention.

Treatment of VLS includes conservative and surgical approaches. Localisation and distribution of bullous emphysema determine surgical methods and surgical success. It has been reported to be difficult to evaluate underlying suppressed lung tissues in bullous emphysema; however, high-resolution chest CT (HRCT) plays an important role in revealing the degree and distribution of emphysema (10). The treatment method in Group I and Group II bullous emphysema is bullectomy. Early respiratory improvement can be achieved by eliminating non-functioning dead space created by bullous lesions and by the re-expansion of adjacent lung tissues with compression atelectasis. In our case, all symptoms improved dramatically in the early postoperative period. Group III and IV cases, however, require volume reduction surgery. In these groups, selection of patients is extremely important and one should not consider surgical intervention in these cases except for those with localised emphysema (1).

As a result, the most crucial issue in VLS is to have a healthy differential diagnosis of pneumothorax and bullae and to evaluate the level of lung tissues affected. Expansion of the collapsed lung and,clinical recovery can be fully achieved via resection of the bullae, in selected cases.

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