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Prikaz slučaja/
Case Report

DIFFERENTIAL DIAGNOSIS BETWEEN VANISHING LUNG SYNDROME AND PNEUMOTHORAX – DOUBLE WALL SIGN - *Case report*

DIFERENCIJALNA DJAGNOZA IZMEĐU „NESTAJUĆIH PLUĆA" I PNEUMOTORAKSA - ZNAK DUPLE LINIJE - *Prikaz slučaja*

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Ključne reči
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Abstract
Giant bullous emphysema, or vanishing lung syndrome, typically occurs in young, thin male smokers with large bullae in one or more upper lobes occupying at least one-third of the hemithorax. Giant bullae often mimic pneumothorax on radiographic appearance.
We present here a rare case of giant bullous emphysema in a mid-age smoking male who was treated as exacerbation of chronic obstructive pulmonary disease and has live for four years on oxygen at home. In this case we highlight the radiology sign which could help the distinction between vanishing lung tumor and pneumothorax, etiology and presentation, since the fact that treatment of these disease is very different.

INTRODUCTION

Giant bullous emphysema (GBE) also referred as vanishing lung syndrome (VLS) are entity with large bullae that encompass more than one-third of the lung volume. They are uncommon entity but when present can lead to compression of adjacent normal lung tissue. The presence of emphysema associated with large bullae is referred to as bullous emphysema. It is either congenital without general lung disease or a complication of chronic obstructive lung disease with generalized lung disease[1,2]. Clinical presentations of GBE usually include dyspnea, hemoptysis, chest pain, or spontaneous pneumothorax. GBE needs to be differentiated from pneumothorax [2, 3]. Giant bullae can easily be misdiagnosed as a pneumothorax, but the management of the two conditions is vastly different. A major complication of VLS is pneumothorax, which classically involves a history of acute deterioration in respiratory function associated with chest pain [2,3]. Distinguishing between the two may require CT scan [1,3].

Case report: A 46-years-old man presented to the Emergency Department with complaints of shortness of breath, cough, and increased sputum production for 2 days without fever or chills. His past medical history was significant for coronary artery disease and chronic obstructive pulmonary disease (COPD). He was not on home oxygen. He had smoked for many years and currently smoked two packs per day.His past medical history was significant for coronary artery disease and chronic obstructive pulmonary disease (COPD) without follow-up by a specialist. He was initially hypoxic and required 2 liters of oxygen. The patient had no occupational exposures or family history of lung disease. Physical examination revealed cachexia, with decreased lung breath sounds and increase in resonance to percussion in left lung. Initially laboratory tests including complete blood count with differential, complete metabolic profile, cardiac enzymes, brain natriuretic peptide, and electrocardiogram were within normal limits. His heart rate was 110 beats/minute and his blood pressure was 110/80mm Hg, and he required 2 L/min oxygen via nasal cannula to achieve a peripheral capillary oxygen saturation of 91%.The initial arterial blood gas analysis on 2litres oxygen showed Ph 7.47, PO2 130, and PCO2 37mmHg. Chest radiography showed large bullous lesion occupying approximately 95%

of left lung, compressing surrounding parenchyma, mildly displacing his upper mediastinum to the right,which was suggestive of vanishing lung syndrome [Figure 1A]. Lung CT scan was performed with addition volumetric rendering technique and revealed the diagnosis of giant bullous emphysema (GBE) [Figure 1B]. Additionally, during hospitalization, he checked his alpha-1-antitrypsin level which was 181 mg/dL (normal range, 90–200 mg/dL).Thoracic surgery was consulted and recommended against surgical management, as the patient had minimal residual lung and would be at high risk for developing a bronchopulmonary fistula. He was treated for a COPD exacerbation with systemic steroids, antibiotics, and nebulized bronchodilators with symptomatic improvement and was discharged home with oxygen. He was scheduled for pulmonary clinic follow-up at 1 week, when he felt well. He is being regularly followed up for four years and he has no symptoms using oxygen at home. Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Discussion: Bullous emphysema refers to emphysematous lung with bullae, which are air-filled spaces within the parenchyma that are 1 cm or larger in diameter and consist of a thin wall of visceral pleura with remnants of alveolar and interlobular septa inside [4]. The natural history of bullous lung disease is usually progressive enlargement as the bullae fill with air and loss of lung function because the irregular, fibrous membranes result in poor gas exchange. If bullae occupy >30% of a hemithorax, they are termed giant bullae [5,6].

Giant bullous emphysema has also been called vanishing lung syndrome or idiopathic giant bullous emphysema (6). It typically occurs in young, middle-aged thin male smokers, who often present clinically with acute shortness of breath. In this setting, difficulties arise in distinguishing pneumothorax from progression of the underlying bullous emphysema [6, 7].

The clinical signs of pneumothorax in patients with giant bullous emphysema are unreliable [7, 8]. A complex and distorted radiographic appearance of the lungs in these patients could bring difficulties to detect of, and may even falsely suggest, pneumothorax [7]. The distinction between the pleural line of a pneumothorax and the bulla wall can be difficult. The advantage of CT over conventional radiography in aiding diagnosis and treatment of pneumothorax has been described, but air in the pleural space may still be a challenge to diagnose with certainty in these patients[6, 7].

The radiographic criteria for vanishing lung syndrome (VLS) were proposed in 1987,by Roberts et al. [8], and they included giant bullae in one or both upper lobes occupying at least one third of the hemithorax and compressing surrounding parenchyma. The following signs have been used to detect pneumothorax in patients with giant bullous emphysema: compressed or consolidated lung, nonanatomic,hyperlucency, and immediate symptomatic relief and lung expansion at chest tube placement [8, 9]. Waitches et al. have described radiological signto distinguish pneumothorax from adjacent giant bullae: the double-wall sign [9]. This sign occurs when one sees air outlining both sides of the bulla wall parallel to the chest wall. Absence of this sign provides further evidence and increased confidence against the diagnosis of pneumothorax, which can prevent unnecessary chest tube placement. The double-wall sign may not be evident on all CT slices, particularly with compression of adjacent bullae, but careful observation of multiple images will reveal this sign when a pneumothorax is present. One potential pitfall in the appreciation of the double-wall sign of pneumothorax occurs when two large bullae are adjacent to one another. This situation can produce an apparent double-wall sign, mimicking pneumothorax. However, careful scrutiny of multiple images will show the absence of air in the pleural space and that the bulla wall is not parallel to the chest wall or parietal pleura [9, 10]. This study which performed Waitches et al. could be limited because of small sample of investigated CT scans, but very helpful in suspecting situations. Also, combing clinical presentation and suddenly onset of shortness of the breath, necessitates CT examination where presence or absence of double wall could be helpful. Our case image had not double wall sign which made easier to diagnose.

In conclusion, Vanishing Lung Syndrome is a rare condition which becomes clinically evident in a much advanced stage. Patients should be strongly counseled against any further tobacco abuse. Vanishing Lung Syndrome could easily be mixed with pneumothorax. Although not sufficiently evidence-based or controlled studies validated, there is radiological sign (double wall sign) which helps in distinguishing in these two entities.

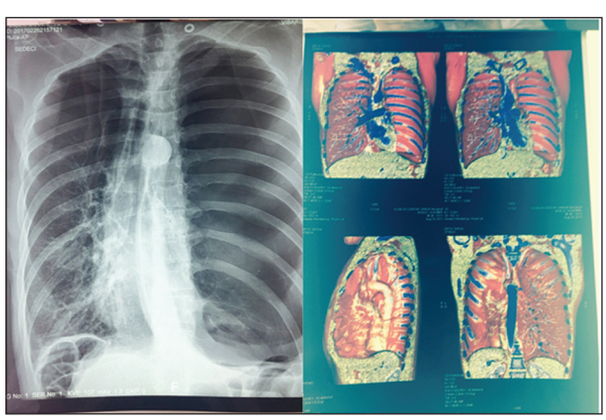

Figure 1A. *Chest X-raywith extensive bullous disease in the left lung, occupying more than 95% of the hemithorax.*
Figure 1B. *Volumetric rendering technique CT done showing a giant avascural emphysematous bullae*

Sažetak

Gigantski bulozni emfizem ili „nestajući sindrom pluća" obično se javlja kod mladih pušača, pripadika muškog roda, radiološki se prikazujući kao ogromna bula u jednom ili više lobusa, zauzimajući najmanje jednu trećinu hemitoraksa. Veliki (gigantski) bulozni emfizem se često radiografski može zameniti sa pneumotoraksom.

Naš rad predstavlja redak slučaj sredovečnog bolesnika sa ogromnom bulom koja je zauzimala ceo hemitoraks, a koji je trertiran zbog pogoršanja hronične obstruktivne bolesti pluća i poslednjih pet godina bio na dugotrajnoj oksigenoj terapiji. Ovaj prikaz je do sada prvi koji opisuje razliku između pneumotoraksa i „nestajućeg sindroma pluća" koja je često „zavarujuća", ali esencijalna, jer je lečenje iI pristup bolesniku totalno različit.

REFERENCES

1. Benito Bernáldez C, Almadana Pacheco A. Spontaneous Regression of Pulmonary Emphysematous Bulla. Arch Bronconeumol. 2017; 53(6): 346–356

2. Greenberg JA, Singhal S, Kaiser LR. Giant bullous lung disease: evaluation, selection, techniques, and outcomes. Chest SurgClin N Am 2003; 13:631–649.

3. Santini M, Fiorelli A, Vicidomini G, Di Crescenzo VG, Messina G, Laperuta P. Endobronchial treatment of giant emphysematous bullae with one-way valves: a new approach for surgically unfit patients. Eur J CardiothoracSurg 2011; 40: 1425–1431.

4. McDonough JE, Yuan R, Suzuki M, Seyednejad N,Elliott WM, Sanchez PG, et al. Small-airway obstruction and emphysema in chronic obstructive pulmonary disease. N Engl J Med 2011; 365(17):1567–1575.

5. Rennard SI. COPD: overview of definitions, epidemiology, and factors influencing its development. Chest 1998; 113(4):235S–241S

6. Lazovic B. Correlation of CRP and serum level of fibrinogen with severity of disease in chronic obstructive pulmonary disease patients. Med Arh. 2012; 66(3): 159-160

7. McDonough JE, Yuan R, Suzuki M, Seyednejad N, Elliott WM, Sanchez PG, et al. Small-airway obstruction and emphysema in chronic obstructive pulmonary disease. N Engl J Med 2011; 365(17):1567–1575.

8. Deslauriers J, Leblanc P. Management of bullous disease. Chest SurgClin North Am 1994; 4(3):539–559.

9. Roberts L, Putman CE, Chen JTT, Goodman LR, Ravin CE.Vanishing lung syndrome: upper lobe bullous pneumopathy. RadiolInteramRadiol 1987; 12:249–255-

10. Waitches GM, Stern EJ, Dubinsky TJ. Usefulness of the double-wall sign in detecting pneumothorax in patients with giant bullous emphysema.AJR Am J Roentgenol2000; 174(6):1765-8.

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