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Giant Pulmonary Bullae: Diagnosis, Complications and Treatment

Charles Joseph Haddad, Judella Haddad-Lacle

Department of Community Health and Family Medicine, University of Florida, Jacksonville, USA

Email address

Charles.haddad@jax.ufl.edu (C. J. Haddad)

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Abstract

This case report outlines a 46-year-old African American patient who presented with cough and decreased exercise tolerance. Chest X-ray revealed a giant pulmonary bullae. Giant Pulmonary Bullae typically occurs in young thin males and is seen most frequently in smokers. Diagnosis is usually confirmed with chest radiograph or chest CT scan. Giant Bullae can be seen in patients with other underlying conditions and it may have a genetic predisposition. Complications include pulmonary hemorrhage, infection, pneumothorax and lung cancer developing in the wall of the giant bullae. Treatment is dependent on the severity of the symptoms with the goals to decrease dyspnea and shortness of breath, improve lung function and improve overall quality of life for the patient. Treatment may include observation, bronchodilators, bullectomy, pleurodesis and one-way valve placement.

Keywords

Giant Pulmonary Bullae, Vanishing Lung Syndrome, Bullous Emphysema, Bullectomy

1. Introduction

Giant Pulmonary Bullae or vanishing lung syndrome typically occurs in young thin males and found most frequently in smokers. Giant bullae take up greater than one third of one or more lobes of one lung. [1]. On chest radiograph the giant bullae appear as a unilateral hyperlucency and may mimic a pneumothorax. [2]

2. Case Presentation

A 46-year-old thin African American male presented to our Family Practice office with a chief complaint of two to three month history of nonproductive cough associated with mild shortness of breath with exertion. The patient denied fever or chills or any travel outside of the United States. The patient had decreased exercise tolerance with inability to maintain his usual cardiovascular fitness program that consisted of jogging 2-3 miles 3-4 times per week, and found that after about a mile, he would have to slow down from his usual pace and have to walk because of fatigue and shortness of breath. The patient admitted to a smoking history of six-pack years, but had quit smoking 3 years before his symptoms began.

His physical exam, including his pulmonary exam was completely unremarkable, and his pulse oximetry was 95% at rest on room air. Laboratory studies including a complete blood count, comprehensive metabolic panel, and thyroid studies were normal.

A chest X-Ray was performed and reported giant pulmonary bullae measuring 13 centimeters. Conservative treatment was started with bronchodilators and inhaled corticosteroids and avoidance of tobacco products. The patient noted considerable decrease in cough and improvement in his exercise tolerance. Upon reviewing more aggressive treatment options, the patient was satisfied with his level of function and declined any treatment other than periodic monitoring of the bullae.

Over the ensuing 2 years, radiographs were performed every 6 months to monitor any potential change in size of the bullae, but no significant change occurred. The patient continues to do well since the initial diagnosis and has continued conservative measures.

3. Figure

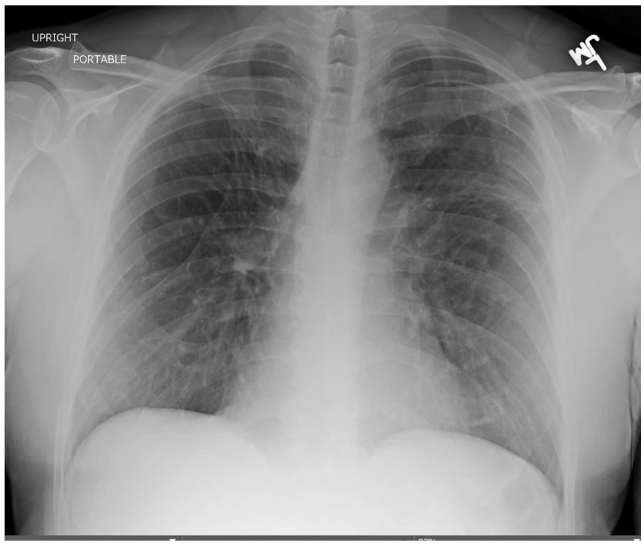

Figure 1. *Giant Pulmonary Bullae in right lobe.*

4. Discussion

Giant pulmonary bullae can be found in patients with emphysema, chronic bronchitis, pneumoconiosis, acquired cysts of the lungs, and granulomatous disease such as tuberculosis. Some studies have shown that there may be a genetic predisposition to develop the giant bullae, which appears to be autosomal dominant and autosomal recessive inheritance. [3]

Patients with lung masses may develop air-trapping leading to a one-way valve causing these giant bullae to develop.

These giant bullae take up space in the lungs and do not take part in air exchange. They compress the existing, previously functional, lung parenchyma and can cause decreased air exchange. [2, 4]

Symptoms of giant pulmonary bullae vary widely with some patients being completely asymptomatic and the bullae being found incidentally on chest radiograph or other imaging studies. Other patients may present with cough, pleuritic type chest pain, shortness of breath, or hemoptysis. Rarely patients may develop acute dyspnea, chest pain and become unstable with acute hypoxia and a picture resembling a tension pneumothorax.

Differential diagnosis of giant pulmonary bullae includes emphysematous bullae, pneumothorax, and lung cancer.

Complications that can occur either because of, or in addition to the giant pulmonary bullae include pulmonary hemorrhage, infection, pneumothorax, ipsilateral pulmonary edema, and decreased lung function. [5]

Pulmonary function tests reveal a decreased forced expiratory volume at one second (FEV1), and increased functional residual capacity (FRC).

A rare complication that can occur is an air embolism causing a major ischemic stroke due to spontaneous rupture of a giant bullae that occurred shortly after takeoff on an airline flight. [6]

There are additional reports of lung cancer developing in the wall of the giant bullae that was complicated with a pulmonary hemorrhage. [7]

Treatment of giant pulmonary bullae is partially dependent on the symptoms and varies widely depending on the severity of the symptoms. Treatment goals include decreasing dyspnea, improving lung function, decreasing exacerbations of shortness of breath, decreasing complications and improving overall quality of life. All patients with this disorder should be encouraged to discontinue smoking by providing counseling, medications such as nicotine replacement patches or gum and non-nicotine products such as Varenicline, Bupropion and other medication. In addition patients may be referred to smoking cessation classes.

For asymptomatic or mildly symptomatic patients, observation or inhaled short acting bronchodilators such as albuterol or long acting bronchodilators such as Salmeterol may be used. Adding an inhaled anti-inflammatory corticosteroid such as fluticasone may provide improvement in symptoms. [8]

For patients that are more symptomatic or in those where the bullae is enlarging, more aggressive treatments may be required. Surgical treatments for this disorder include bullae resection, pleurodesis, lung resection, unilateral endobronchial valve placement, resection lung stapling, and lung volume reduction surgery or lung transplant. There is also limited information regarding the efficacy of using autologous blood, or antibiotics being instilled into the giant bullae as a treatment method. This process is thought to cause scarring and inflammation of the bullae causing reduction in size and decreased volume of the bullae. [9]

Using video-assisted thorascopic surgery (VATS) has been widely used to diagnose and treat intrathoracic disease including giant pulmonary bullae. [10]

This technique can be used for bullectomy, pleurodesis, and one-way valve placement. VATS may also decrease the operative time and improve recovery time because of smaller incision and less peripheral damage.

One way endobronchial valve placement allows air to be expelled from the giant bullae with gradual shrinkage over time. There are some reports of reinflation of the remaining lung after bullectomy, with improved lung function over time. [11]

Some giant bullae may resolve spontaneously, and therefore in asymptomatic, or minimally symptomatic patients, watchful waiting may be an option.

Careful selection of patients is required to make a clinical decision as to what treatment is optimal. This is based on the patient's symptoms, underlying lung function, size of the giant bullae, and the patient's comorbidities.

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