

FEATURES OF THE CLINIC, DIAGNOSIS AND METHODS OF TREATMENT OF COMPLICATED BULLOUS LUNG DISEASE

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Abstract

Bullous emphysema affects more than 5% of the world's population, with almost 12% among adults over 30 years of age. It also ranks as the third leading cause of death in the US and kills over 120,000 people a year. It is also known that this disease causes spontaneous pneumothorax in 70-80% of cases. Currently, there is no consensus on the etiology, pathogenesis, clinic, diagnosis and treatment of bullous lung disease. According to the literature, surgical reduction of lung volume in the development of complications of bullous disease shows a high percentage of good immediate and long-term results by improving the physical capabilities of patients, eliminating shortness of breath, improving the quality of life and survival. Nevertheless, the search for effective minimally invasive methods of surgical treatment of patients with bullous pulmonary emphysema continues.

Introduction

Bullae (false pulmonary cysts) are pathological air cavities in the lungs (from the English blebs - "bubbles"), which can occur due to mechanical damage to the parenchyma, an infectious and inflammatory or other disease. Excess air in these sac-like cavities, changes in the structure of the pulmonary matrix, a reduction in the area of the functional areas of the respiratory organ lead to persistent respiratory failure, and the consequences of such pathological changes can be irreversible. The formation and enlargement of bullae in the lungs leads to a decrease in the gas exchange function of the lungs, and in the case of a rupture of a large bulla, it can lead to a life-threatening condition - pneumothorax.

Bullous lung disease (pulmonary emphysema) is a disease of the respiratory tract characterized by pathological expansion of the air spaces of the distal bronchioles, which is accompanied by destructive morphological changes in the alveolar walls; one of the common forms of chronic nonspecific lung diseases.

EPIDMYOLOGY. Bullous emphysema affects more than 5% of the world's population, with almost 12% among adults over 30 years of age. It is also the third leading cause of death in the United States, killing more than 120,000 people a year. It is also known that this disease causes spontaneous pneumothorax in 70-80% of cases. In foreign literature, bullous lung disease is defined by authors as "vanishing lung syndrome" (vanishing lung syndrome) and indicate a tendency towards an increase in the incidence and complicated course of this pathology, while noting the absence of a decrease in the rates of unsatisfactory treatment results due to relapse of the disease [1, 2, 3].



ETIOPATHOGENESIS and CLINICAL FEATURES. There is currently no consensus on the etiology, pathogenesis, clinical features, diagnosis, and treatment methods of bullous lung disease. The two most common causes of bullous emphysema are smoking and alpha-1- antitrypsin deficiency (A1AD or AATD), an inherited autosomal dominant genetic condition. Less common causes of emphysematous destruction of the lung parenchyma are smoking marijuana, crack cocaine, or intravenous drug use, leading to inflammatory or destructive damage to the alveoli [4, 5].

Bullous emphysema is characterized by progressive respiratory symptoms and involves persistent airspace expansion in the distal terminal bronchioles due to destruction of alveolar sacs, loss of elastic tissue, airway collapse, and impaired gas exchange [6]. The pathophysiology of bullous emphysema involves valvular broncho block, which allows air to enter but not exit the cystic space. The morphological basis of this disease is air cavities (bullae) in the lung parenchyma. In foreign literature, it is customary to distinguish between blebs - air cavities less than 1 cm in size, located in the interstitial and subpleural, and bullae - air formations more than 1 cm in diameter, which developed due to emphysematous destruction of the lung parenchyma, the walls are lined with alveolar epithelium. Thus, bullae are formed as a result of the destruction of the interalveolar walls due to chronic or, less often, acute stretching injury with increased interalveolar pressure [4, 7].

A giant emphysematous bulla is defined as an air-filled space that occupies more than one-third of the hemithorax and develops in a lung with generalized emphysema [5, 8]. Emphysema is characterized by airflow limitation associated with loss of distal lung architecture with persistent expansion of the alveolar space, also distal to the terminal bronchiole [9].

Giant bullae usually form slowly with gradual filling with air, but rapid enlargement and spontaneous deflation are also possible. The distribution of giant bullae is usually unilateral and asymmetric; however, bullous emphysema has bilateral involvement. There are no known factors that determine whether the lesion is unilateral or bilateral.

Giant bullae may be asymptomatic, with dyspnea or, rarely, hemoptysis [10]. Diagnosis is radiological, with an X-ray showing a bulla occupying more than 30% of the hemithorax. Sometimes, a chest computed tomography scan is required to differentiate giant bullae from pneumothorax.

In contrast to bullae, pneumothorax is defined as the presence of air in the pleural space and is clinically classified by whether it developed spontaneously or traumatically. Additionally, spontaneous pneumothorax is classified as primary spontaneous pneumothorax if there is no known lung disease or secondary to chronic lung disease. It is called tension pneumothorax and is associated with varying degrees of hypotension, hypoxia, chest pain, and dyspnea.

The pathophysiology of bullous emphysema is closely related to chronic inflammation of the distal airspaces, leading to destruction of the alveolar walls and subsequently, persistent airspace enlargement. This may ultimately lead to decreased gas exchange and airflow limitation due to decreased elastic recoil of the chest wall. There is often an increased number of goblet cells in the airways with mucous gland hyperplasia, fibrosis, and airway collapse due to loss of attachment from alveolar destruction. This decreased gas exchange and airflow limitation ultimately leads to chronic hypoxia and hypocapnia in these patients.



CLASSIFICATION

Emphysematous lung may demonstrate a homogeneous or heterogeneous (regional) pathological focus, which may differently affect the lung parameters characteristic of bullous emphysema (e.g., dynamic lung volume) [11, 12]. Today, depending on the level of damage to the acinus, it is customary to divide bullous emphysema into:

- centrilobular emphysema - the affected areas are localized mainly in the upper parts of the lungs. It is most closely associated with smoking and is the result of the expansion and destruction of the respiratory bronchioles.
- pan lobular emphysema - found mainly in the lower lobes and is often associated with genetic (alpha1-antitrypsin) deficiency.
- Para septal emphysema - occurs on the periphery of the lobules, especially in the subpleural region.

Chronic exposure to harmful particles (cigarette smoke, air pollution) leads to oxidative stress, proteinase-antiprotease imbalance, increased apoptosis and chronic inflammation, which all lead to the gradual destruction of lung tissue [13, 14].

Progressive destruction of healthy lung tissue results in the classic physiologic characteristics of severe emphysema: lung hyperinflation, loss of elastic recoil, loss of surface area for gas exchange, and flow limitation [15]. Emphysema causes decreased elastic recoil pressures and increased lung compliance. This in turn causes static and dynamic lung hyperinflation, which limits airflow and leads to clinical outcomes of lower functional capacity, higher levels of dyspnea, and limited exercise capacity.

Bullous disease can lead to complications such as pneumothorax, infection and bleeding. The most serious and frequent complication of bullous disease is recurrent pneumothorax, the mechanism of which is due to an excessive increase in intrapulmonary pressure in the bullae (due to physical exertion, lifting weights, severe coughing, straining). At the same time, most authors do not classify spontaneous pneumothorax as an independent disease and consider it a constant companion of complications of bullous lung disease. Clinical signs of spontaneous pneumothorax are sharp pain in the chest radiating to the neck, collarbone, arm, shortness of breath, inability to take a deep breath, paroxysmal cough, forced position [16, 17, 18].

DIAGNOSIS

The objectives of examination for bullous disease are as follows:

- 1) exclusion of other diseases, primarily diffuse cystic ones, such as lymphangioleiomyomatosis, Langerhans cell pulmonary histiocytosis, Birt-Hoog - Dube syndrome [19];
- 2) establishing the possible cause of the bullae: smoking, marijuana use, HIV, connective tissue diseases;
- 3) determination of indications and contraindications for surgical treatment, exclusion of complications.

Patients undergo computed tomography (CT) of the chest organs and functional studies, including body plethysmography and a study of the diffusion capacity of the lungs. Bullous emphysema is characterized by an obstructive pattern of respiratory dysfunction, while bullous disease is more likely to be restrictive. A marked decrease in the diffusion capacity of the lungs indicates the presence of diffuse emphysema, and not just bullae. To assess the severity of respiratory failure and clarify the indications for surgery, it is necessary to study the oxygen saturation and gas



composition of arterial blood. All patients with giant bullae should have their α 1-antitrypsin levels measured [20].

Bullae are detected by CT scan of the lungs. This study allows you to measure the number of bullae, their size and shape. CT will also allow you to assess changes in lung tissue, the presence or absence of emphysema, bronchiectasis or other cavities. [26].

CT of the lungs allows to exclude cystic diffuse lung diseases. They may resemble bullous deformation.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis of bullous emphysema, which may be confused with the disease, includes asthma, which is excluded by pulmonary function tests, bronchiectasis, which may be visible on imaging, and congestive heart failure [21, 26].

TREATMENT:

When managing patients with bullous lung disease, various conservative treatment methods are usually used, which do not lead to long-term positive results and are characterized by a large number of complications and relapses (20-50%). In this regard, when complications develop, surgical intervention is indicated, where specialists offer many procedures and in which the choice of target lung tissue remains a difficult task [4, 5, 7, 22].

The goal of surgery is to remove giant bullae, allowing the remaining lung to expand and restore respiratory function. An enlarging giant bulla may also be considered for surgical resection, even if it is asymptomatic [23].

Research has shown that removing the most damaged and no longer functioning parts of the lungs (volume reduction surgery) improves the function of the remaining lung by:

- increasing the elastic recoil pressure, thereby increasing the flow of exhaled air,
- reduction of the degree of hyperinflation, which leads to improvement of the mechanics of the diaphragm and chest wall,
- reduction of heterogeneity of regional ventilation and perfusion, which leads to improvement of alveolar gas exchange and increase in ventilation efficiency while maintaining blood gas levels [24].

Zoumot (2015) adds that surgical lung volume reduction may result in a reduction in the asynchronous movement of different compartments of the chest wall, leading to improved ventilation mechanics [25, 27].

Group authors from the Basil Hetzel Institute for Translational Health Research (Australia), van Agteren et al. (2016) presented an analysis of the results of operations (1760 patients with bullous lung disease) to reduce the volume of lungs (LVL) – lung volume reduction surgery (LVRS), which leads to an increase in the mechanical efficiency of healthy areas of the lung and, as a result, to more efficient gas exchange [17]. At the same time, several objectives were set: to study the postoperative period in terms of lung function and quality of life of patients, to determine the morbidity (relapse) and mortality rates, to study the cost-effectiveness of LVRS and to determine which surgical methods lead to better results in these patients [27]. In particular, the effect of suture line strengthening on the effectiveness of LVRS was considered, the traditional approach to LVRS was compared with a surgical approach that did not involve removal of the affected area of the lung (resection).



At the end of the follow-up period, mortality rates were lower in participants who had undergone OVL than in those who had received standard care. Participants who had poor lung function due to the distribution of diseased tissue in the lungs were at higher risk of death at three months and throughout one large study. One study identified a group of participants who responded better to OVL than other participants, making them particularly suitable for this treatment [17].

A higher percentage of good results was observed in patients with predominant upper lobe emphysema and low exercise capacity. The reduction of 13.6–14.7 units in the SGRQ respiratory questionnaire clearly exceeds the minimal clinically important difference (reduction of 4 points) for this questionnaire [27, 30]. More recent studies show that careful patient selection and performance of procedures in specialized centers lead to a significant reduction in the risk of death overtime [31, 32].

McNulty et al (2014) noted only palliative effects of OUOL [30]. This fact, among other things, has prompted the development of minimally invasive techniques that can help achieve the benefits of OUOL without the risk of death and associated costs.

The latest British Society of Thoracic Surgeons guidelines (2010) for the management of primary spontaneous pneumothorax stated that after the first recurrence, treatment should involve surgery (bullectomy-electomy followed by a pleural adhesion induction procedure). Therefore, a surgical approach is considered the best treatment to minimize the risk of recurrence in patients who have experienced primary spontaneous pneumothorax. The video-assisted thoracic surgery approach has been shown to have greater benefits in terms of pain and patient respiratory function compared with thoracotomy incisions. As an alternative to standard multiport VATS, a single-port, single-incision or uniportal approach has been developed. The uniportal technique has been shown to be safe and effective not only for lung resection and biopsy but also for lobectomy. From this point of view, evidence has shown that the minimally invasive approach should be preferred, confirming the advantages over traditional methods [29, 33, 34, 35].

According to Vanucci et al. (2019) VATS from uniportal access (Uniportal video - assisted Thoracoscopy is a feasible and safe method for performing bullectomy, with results at least comparable to other methods, resulting in symptom resolution, improved lung function and improved quality of life [8].

CONCLUSION

Chronic lung diseases, including bullous emphysema, are the third leading cause of death worldwide. Representing a heterogeneous group of pathologies, they are characterized by persistent airflow limitations, hyperinflation, decreased elasticity of the airways as a result of structural degradation and inflammation of the lung tissue, and disrupt effective gas exchange between the alveoli and blood.

According to the literature, surgical reduction of lung volume in the development of complications of bullous disease shows a high percentage of good immediate and remote results due to the improvement of physical capabilities of patients, elimination of dyspnea, improvement of quality of life and survival. Nevertheless, the search for effective minimally invasive methods of surgical treatment of patients with bullous emphysema of the lungs continues.



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