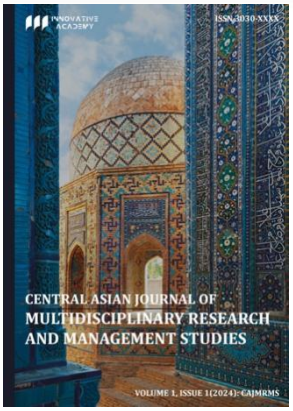

Modern technologies of treatment of bronchiectasis in children.

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ABSTRACT

bronchiectasis is characterized by chronic and abnormal dilation of the airways and is caused by impaired clearance of various microorganisms and recurrent infection. In 26–53% of cases, bronchiectasis has an unknown cause, which means that the issues of etiology and pathogenesis of bronchiectasis are very relevant for study. This review summarizes current understanding of the epidemiology, pathophysiology, clinical features and management of children with bronchiectasis. Recently, new data have emerged to improve our understanding of this heterogeneous disease, for example the role of the interaction of the immune system and microbiota in the pathogenesis of bronchiectasis.

Bronchiectasis is an acquired chronic disease of the bronchopulmonary system, characterized by a purulent-inflammatory process in dilated deformed bronchi with infiltrative and sclerotic changes in the peribranchial space [1]. It can be assumed that chronic airway inflammation is the main cause of bronchiectasis observed with recurrent pulmonary infections. But it is now believed that bronchiectasis is a multi-etiological pathology, the pathogenesis of which involves a complex interaction between the body, respiratory pathogens and environmental factors. This interaction leads to a vicious cycle of repeated infections, inflammation of the airways and tissue remodeling, contributes to impaired clearance, destruction of the structural elements of the bronchial wall, and forms dilatation and obstruction of small bronchi.

Epidemiology

The true time of development of bronchiectasis in children is difficult to establish due to the delay in diagnosis verification. There is currently no reliable information about the prevalence

of bronchiectasis. But studies indicate variation in incidence data depending on geographic regions. The incidence in children aged 0–14 years living in developed countries is considered low and ranges, for example, from 0.5 per 100,000 children in Finland to 3.7 per 100,000 children in New Zealand. However, among Aboriginal children from Central Australia, incidence rates are as high as 200 per 100,000 children [2]. In India, due to the unsatisfactory level of medical care in children under 4 years of age after pneumonia, bronchiectasis is diagnosed in 212–2646 cases per 1 million children per year [3].

Thus, bronchiectasis remains an important problem affecting socially disadvantaged populations, especially children living in developing countries where overcrowding, poor hygiene and limited access to health care occur [5].

Etiology

Bronchiectasis is a polyetiological disease associated with other disorders. According to the literature, the following causes of bronchiectasis are more common: previous pneumonia (19%), primary immunodeficiency (17%), recurrent aspiration, including foreign body inhalation (10%) and primary ciliary dyskinesia (7%). However, more than 30% of cases remain idiopathic [3, 6].

Congenital causes of bronchiectasis

Developmental defects:

• respiratory tract abnormalities;

• laryngotracheobronchomalacia;

• tracheoesophageal fistula;

• laryngeal cleft;

• congenital cartilage anomalies;

• pulmonary sequestration.

Violations of bronchopulmonary clearance:

• primary ciliary dyskinesia;

• cystic fibrosis;

• alpha-1-antitrypsin deficiency.

Primary immunodeficiency:

• hypo- and agammaglobulinemia;

• common variable immune deficiency;

• phagocyte defects: chronic granulomatous disease;

• hyperimmunoglobulinemia E syndrome;

• ataxia-telangiectasia (Louis-Bar syndrome).

Acquired causes of bronchiectasis

Post-infectious complications:

• severe pneumonia;

• measles;

• whooping cough;

• tuberculosis.

Environmental reasons:

• inhalation of toxins.

Airway obstruction:

• foreign body aspiration;

• intra- and extracompression of the bronchus by a lymph node, tumor masses, cyst, vascular ring.

Allergies and inflammation:

• allergic bronchopulmonary aspergillosis;

• rheumatic and autoimmune disorders.

In addition, A. Horani et al. established the presence of a connection between bronchiectasis and certain gene variants, as well as an increased prevalence of this disease in some ethnic groups, which indicates the influence of genetic factors.

However, at the moment there are no large studies examining the relationship between genome variations and bronchiectasis [10, 11].

A special group are patients with primary immunodeficiency. They have a significantly increased risk of developing bronchiectasis. As shown in a study [12], 37.5% of patients with bronchiectasis were diagnosed with defects in the antibody-mediated immune response.

According to R. Yazdani et al., in 26%–53% of cases, bronchiectasis has an unknown cause [12].

Identifying the etiology of bronchiectasis significantly influences patient management and prognosis of the disease. For example, administering immunoglobulin replacement therapy to patients with primary immunodeficiency may prevent the progression of irreversible lung damage.

Pathogenesis

At the moment, our knowledge about the mechanisms responsible for initiating the process leading to bronchiectasis is still limited. It is known that infection causes infiltration of the mucosa by neutrophils and T-lymphocytes, which in turn leads to an increase in the concentration of inflammatory elements such as neutrophil elastase, interleukin-8 and tumor necrosis factor-α. The complex interaction between these mediators leads to constant dilatation and thickening of the walls of the airways, i.e. to bronchiectasis [3]. Narrowing of the bronchial lumen promotes increased bacterial colonization and infection, creating a “vicious circle” of infection and dysregulation of respiratory tract inflammation. This leads to progressive destruction of the bronchial wall as a result of pulmonary dilatation and obstruction [2, 13]

Moreover, a high bacterial load in the airways causes a decrease in the concentration of secretory leukoprotease inhibitor, which can cause uncontrolled elastase-dependent airway damage [14]. An increase in elastase concentration leads to hypersecretion of mucus, weakening of ciliary function, destruction of immunoglobulins, decreased phagocytic activity,

and also contributes to epithelial damage. It is important to note that, according to the literature, a decrease in mucociliary clearance occurs as a result of local damage, characteristic only of the affected area and not associated with initially insufficient protection of the respiratory tract [2]. At the same time, studies of bronchial secretions in children and adults with bronchiectasis revealed the presence of pathogens of a viral and bacterial nature and an increase in the level of inflammatory mediators in the lower respiratory tract, which indicates the ineffectiveness of clearance mechanisms in the pathogenesis of bronchiectasis [15–19].

A study of biopsy specimens of the bronchial mucosa showed infiltration of neutrophils and mononuclear cells with increased expression of IL-8, hypertrophy of the mucous glands and increased contractility of smooth muscle muscles [20].

Although the historical model of neutrophilic airway inflammation has been established, the role of eosinophils in the pathogenesis of bronchiectasis has also been described. V. Goyal et al found an increase in the number of activated eosinophils in bronchial biopsies and sputum of adult patients with bronchiectasis [2]. Similarly, elevated levels of eosinophils were reported in bronchoalveolar lavage fluid in 34% of children diagnosed with bronchiectasis [21]. Since eosinophils promote the recruitment of neutrophils, goblet cell hyperplasia and damage to epithelial cells, an increase in their number in the airways of bronchiectasis may contribute to a more severe course of the disease [22, 23].

Mannose binding lectin (MBL) and fecolin are complement-activating proteins that may also play a pathogenetic role in the development of bronchiectasis. They are chemotaxis factors, attracting neutrophils to the site of infection, promoting the release of potentially tissue-destroying azurophilic granules of neutrophils - myeloperoxidase, defensins, neutrophil elastase, proteinase-3 and cathepsin G [24].

It is noteworthy that the regulation of the immune response can lead to the formation of bronchiectasis. Proteolysis of mediators, pro-inflammatory molecules and increased secretion of mucus damage the structures of the lung tissue and contribute to a delay in the elimination of the pathogen [25].

Clinical manifestations and diagnosis

The clinical manifestations of bronchiectasis are diverse and directly depend on the extent of damage to the lung tissue, the prevalence and severity of inflammation in unaffected areas of the organ. In children, the course of the disease has special clinical symptoms, which are determined by the form of bronchiectasis. Common manifestations: cough (95%), shortness of breath (82%), expectoration (66%), fever (62%), wheezing (53%), repeated pneumonia (46%), hemoptysis (15%) and failure to thrive (70%) [3].

Bronchiectasis should be excluded in children with any of the following clinical symptoms [14]:

- persistent wet cough that lasts more than 8 weeks and persists between cold episodes;
- asthma not responding to appropriate treatment;
- persistent course or recurrence of symptoms of pneumonia;
- whooping cough that does not resolve even after 6 months;
- constant crepitus in the chest for no apparent reason;

• repeated breathing disorders associated with respiratory diseases of the upper respiratory tract;

• hemoptysis for no apparent reason.

If bronchiectasis is suspected, radiation methods remain the main diagnostic tool. Chest X-ray is not specific. X-ray changes include the known bronchovascular markers: dilated bronchus, loss of lung volume, and peribronchial thickening. In most cases, plain radiography only allows one to suspect bronchiectasis [3].

Currently, the diagnosis is confirmed using high-resolution CT [8]. Findings on high-resolution CT in bronchiectasis are dilation of the airway lumen greater than the diameter of the adjacent vessel; lack of narrowing at the periphery; thickening of the bronchial wall; cylindrical, varicose and/or saccular changes with local or diffuse lung damage.

It is known that patients with primary immunodeficiency (namely with defects in DNA repair) and some patients with common variable immune deficiency have an increased sensitivity to radiation. A radiation-free alternative to CT or chest radiography may be magnetic resonance imaging [9].

In addition to detecting bronchiectasis, it is advisable to determine the etiological factor of the disease.

Bronchoscopy reveals that in 3–14% of cases the main cause of bronchiectasis is malformations of the respiratory tract. Fiberoptic bronchoscopy helps identify airway abnormalities and obtain samples of the bronchial wall [4].

Treatment

The main goals of therapy for bronchiectasis are to improve the patient's condition and prevent/slow the progression of the disease. There is currently no consensus on the management of patients with bronchiectasis. Based on clinical recommendations, it is advisable to carry out treatment according to the established etiology of the disease. Conservative treatment consists of antibacterial and mucolytic therapy to improve sputum discharge. It is also recommended to consider the issue of prescribing inhaled bronchospasmolytic drugs (β2-agonists) in case of a positive test during a pulmonary function test and in case of clinical effectiveness [4].

When the possibilities of conservative therapy have been exhausted, the method of choice is surgical removal of the pathological area of the lung. Indications for surgical intervention are localized, uncomplicated, cylindrical, saccular and mixed bronchiectasis, which is a source of frequent exacerbations of lower respiratory tract infections and significantly worsens the patient's quality of life. In case of dangerous bleeding (more than 200 ml/day) or hemoptysis (uncontrolled by conservative therapy) from the local affected area, surgical intervention is also performed. An alternative to resection in the latter case is bronchial artery embolization [4, 7].

Conclusion

It is important to note that significant gaps remain in our understanding of the underlying disease mechanisms that initiate and maintain the pathophysiological changes leading to bronchiectasis. The role of immune system factors and the influence of microbiota on it has not been fully studied.

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