BRONCHIECTASIS DISEASE: ETIOLOGY, PATHOGENESIS, MODERN DIAGNOSIS AND TREATMENT

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Annotation: *According to various authors, the prevalence of bronchiectasis has significant variability - from 1.2 to 30 per 1000 population. Bronchiectasis occurs in 0.5-1.5% of the population, developing mainly in childhood and young age (from 5 to 25 years). Common causes of bronchiectasis are cystic fibrosis, immune disorders, and recurrent infections, although some cases are idiopathic. The course of bronchiectasis alternates with periods of exacerbation and remission. Diagnosis is based on history and physical examination; confirmed by HRCT. Conservative treatment includes airway hygiene, sanitation of the bronchial tree, antimicrobial therapy, specific treatment for individual causes of the development of the disease, correction of hypoxia, etc. Surgical treatment is in most cases a radical method of treating bronchiectasis. The operation is indicated for bronchiectasis with frequent exacerbations, and with its complications (hemoptysis, frequent pneumonia), when it is not possible to achieve stable remission by conservative methods.*

Key words: *bronchiectasis, bronchiectasis, prevalence, clinical manifestations, diagnosis, treatment*

БРОНХОЭКТАЗЫ: ЭТИОЛОГИЯ, ПАТОГЕНЕЗ, СОВРЕМЕННАЯ ДИАГНОСТИКА И ЛЕЧЕНИЕ

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Аннотация: *По данным различных авторов распространенность бронхоэктазов имеет значительную вариабельность - от 1,2 до 30 на 1000 населения. Бронхоэктатическая болезнь встречается у 0,5-1,5 % населения, развиваясь преимущественно в детском и молодом возрасте (от 5 до 25 лет). Частые причины бронхоэктаза – муковисцидоз, иммунные нарушения и рецидивирующие инфекции, хотя некоторые случаи являются идиопатическими. Течение бронхоэктатической болезни чередуется периодами обострения и ремиссии. Диагноз ставится на основании анамнеза и объективного обследования; подтверждается КТВР. Консервативное лечение предусматривает гигиену дыхательных путей, санацию бронхиального дерева, антимикробную терапию, специфическое лечение при отдельных причинах развития болезни, коррекцию гипоксии и др. Хирургическое лечение является в большинстве случаев радикальным методом лечения бронхоэктазов. Операция показана при*

бронхоэктатической болезни с частыми обострениями, и при ее осложнениях (кровохарканье, частые пневмонии), когда не удается консервативными методами добиться стойкой ремиссии.

Ключевые слова: *бронхоэктазы, бронхоэктатическая болезнь, распространенность, клинические проявления, диагностика, лечения*

BRONXOEKTAZ KASALLIGI: ETIOLOGIYASI, PATOGENEZI, ZAMONAVIY DIAGNOSTIKASI VA DAVOLASH

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Annotatsiya: *Turli mualliflarning fikriga ko'ra, bronxoektazning tarqalishi sezilarli o'zgaruvchanlikka ega bo'lib har 1000 nafar aholiga 1,2 dan 30 gacha boradi. Bronxoektaz kasalligi aholining 0,5-1,5 foizida uchraydi, asosan bolalar va yoshlarda (5 yoshdan 25 yoshgacha) rivojlanadi. Bronxoektazning keng tarqalgan sabablari mukovistsidoz, immunitetning buzilishi va takroriy infektsiyalardir, ba'zi holatlarda esa idiopatik. Bronxoektaz kasalligining kechishi qo'zish va remissiya davrlari bilan almashadi. Tashxis anamnez yig'ish va fizik tekshiruvga asoslanadi; yuqori aniqlikdagi kompyuter tomografiyasi tomonidan tasdiqlanadi. Konservativ davolash nafas yo'llarining gigiyenasi, bronxial daraxt sanatsiyasi, antibakterial terapiya, kasallikning rivojlanishining individual sabablarini o'ziga xos davolash, gipoksiyani korreksiyalash va boshqalarni o'z ichiga oladi. Xirurgik davolash ko'p hollarda bronxoektazlarni davolashning radikal usuli hisoblanadi. Tez-tez qo'zib turadigan bronxoektazlar va uning asoratlari (qon tupurish, tez-tez pnevmoniya), konservativ usulda davolash bilan barqaror remissiyaga erisha olinmagan hollar operatsiyaga ko'rsatma bo'ladi.*

Kalit so'zlar: *bronxoektaz, bronxoektaz kasalligi, tarqalishi, klinik ko'rinishi, diagnostika, davolash*

Bronchiectasis - irreversible expansion of the branches of the bronchial tree, due to destruction (decomposition) bronchial walls and / or breach the wall muscle layer due to bronchial tone inflamed - Nia, sclerosis, dystrophy, hypoplasia.

Bronchiectasis (BE) - the disease is characterized by irreversible changes in the bronchi (expansion, deformation), which are accompanied by functional inferiority and the development of a chronic purulent-inflammatory process in the bronchial tree.

Suppurative inflammation of the bronchi dramatically enhanced deformed with infiltrative and sclerotic changes in okolobronhialnom space, leading to a further optionally - invertible extension with infringement of bronchial drainage function - tion, developing atelectasis, emphysema, cirrhosis parenchyma (tissue) Lung.

For the first time he formulated and defined bronchiectasis by Rene Laennek. In 1948, the Russian professor A.Ya. Tsygelnik published a unique monograph entitled «Bronchiectasis», in which the author summarized the clinical experience of bronchiectasis during wartime and in the pre-antibacterial era. In 1948, the Russian professor A.Ya. Tsygelnik published a unique monograph entitled «Bronchiectasis», in which the author summarized the clinical experience of bronchiectasis in wartime and in the pre-antibacterial era [1].

EPIDIMIOLOGY: According to various authors, the prevalence of bronchiectasis has significant variability - from 1.2 to 30 per 1000 population. Bronchiectasis occurs in 0.5-1.5% of the population, developing mainly in childhood and young age (from 5 to 25 years). The greatest prevalence of bronchiectasis was found in ecologically unfavorable regions of residence (regions of the Far North, Primorye), as well as in people with bad habits (tobacco smoking).

The epidemiology of BE is not well understood; in different regions of the world, it has a different degree of prevalence. Countries with a high prevalence of TB patients have a higher prevalence of BE. CT plays an important role in the detection of BE. In hospitals where this method of image-diagnostics is widely implemented, BEs are relatively common. In the United States, the National Institutes of Health conducted a study on the epidemiology of BE and found that more than 110,000 people in the country suffer from BE. With age, the number of people with BE increases to 272 per 100,000 population, while among the young population, BE does not exceed 4 patients per 100,000; women get sick more often than men [2].

Common causes are cystic fibrosis, immune disorders, and recurrent infections, although some cases are idiopathic.

Bronchiectasis is a polyetiological (several causes) process. The main reason for the development of bronchiectasis in childhood and adolescence is productive inflammation of the airways. Destructive (destructive) changes in muscle and elastic layers are accompanied by bronchial wall dilatation (extension) and the function - tional work bronchial disorders. Stagnation of mucus causes a hacking cough with races - stringing wall, obstruction of terminal bronchioles (the final section «bronchial tree»). In parallel with ventilation disorders, the perfusion (filling with blood) of the lung tissue decreases. The formation of bronchiectasis is often combined with chronic bronchitis and emphysema. Among patients with COPD (chronic obstructive pulmonary disease) - detected in 18-27% of cases [3].

Damage to the bronchi occurs in individuals with genetic defects or a predisposition to pathology. The primary include bronchiectasis in congenital anomalies and genetic diseases of the lungs. The secondary may be due to any reasons, defiant - E lung injury. Divided by the clinical course (Remis - Sia or acute phase) of prevalence (lung injury segments share the whole lung, reversible process), the shape - cylindrical, spindle, saccular, cystiform mixed. Among other lung diseases, bronchiectasis accounts for 10 to

30% [4]. With fluorography of the chest organs, they are detected in 1-2 out of 1000 people, and when examined using bronchography - in 0.5% of the population . According to the data of pathological studies, BE is diagnosed in 2 - 4% of the adult population. Bronchiectasias in 2/3 of patients are detected before the age of 20 years. At the same time, at the age of up to 10 years, BE is equally often observed in boys and girls, and among adults, men get sick 1.5 - 3 times more often than women. In recent years, the number of patients with BE in a specialized clinic, despite the increase in diagnostic capabilities, has decreased. This is probably due to an increase in the effectiveness of treatment of respiratory diseases in childhood, including pathologies that contribute to the development of BE [5, 31].

Secondary bronchiectasis, developing against the background of other bronchopulmonary diseases (pulmonary tuberculosis, lung abscess, chronic bronchitis, pneumofibrosis), are more common. They get sick mainly in childhood and young age, more often men [6].

In recent years, the role of non-tuberculous mycobacteriosis has been discussed, especially in patients with BE localized in the middle lobe or in the lingular segment of the left lung. In modern clinical guidelines, attention is drawn to the need to resort to molecular genetic research methods to confirm the mycobacterial nature of BE [7].

PATHOGENESIS. Pathogenesis The pathogenesis of BE is based on several mechanisms that ultimately lead to the development of traction, dilated BE. Another mechanism for the development of BE is a weakening of the elastic traction of the bronchial wall. The lungs are in a straightened state due to negative intrapleural pressure and the presence of cartilaginous rings of the trachea, large and medium bronchi. Fibrous changes in the lung tissue, which can be observed in diseases such as sarcoidosis, interstitial pneumonia, etc., lead to a fixed traction of the bronchi. Pulsating bronchial dilatation is more associated with infectious recurrent respiratory processes. A typical example is the development of this type of BE in aspergillosis. The inhalation route of penetration of aspergillus leads to the formation of a viscous secretion in the lumen of the respiratory tract, which has a compressive effect on the lumen of the bronchus. An important role, of course, is assigned to the inflammatory process of the bronchial wall. The third pathogenetic mechanism is associated with a weakening of the elastic traction of the airway lumen. Most often, this type of BE occurs with a previous infectious disease of the bronchopulmonary system. A decrease in the elastic traction of the airways is also noted in some hereditary forms of diseases, for which one of the characteristic signs is the development of BE. Thus, BE can be observed in the syndromes of Marfan, Williams – Campbell, Munier Kuhn. In BE during bronchoscopy, collapse of the bronchi can be observed, which reflects a violation of the elastic traction of the bronchial wall. Below we consider the so-called vicious circle [8] of the inflammatory process, which is given a central role in the pathogenesis of BE. The airway is in direct contact with the environment; the system of protection of the respiratory tract from the penetration of

viruses, bacteria, fungi and their associations into the human body has evolved. Defense mechanisms include macrophages, DCs, neutrophils, whose function is to maintain the immune defenses of the lungs. Other defense mechanisms are the cough reflex, the mucociliary escalator mechanism, and antimicrobial peptides such as lysozyme, defensins, leukocyte proteinase inhibitors, secretory IgA, and T-effector lymphocytes. A certain pathogenetic role in the development of BE is played by the formation of a viscous secretion in the lumen of the airways and a violation of the mechanisms of its clearance. This mechanism may play a central role in the colonization of pathogens such as Aspergillus and Pseudomonas aeruginosa on the surface of the bronchial mucosa. In the lumen of the bronchus, inflammatory cells are deposited: neutrophils, lymphocytes, mononuclear cells. Microbial colonization correlates with biological markers such as IL-8, -1β, tumor necrosis factor; the systemic nature of the inflammatory reaction is indicated by an increase in the secretion of E - selectin, adhesive molecules-1. Ultimately, the transmural inflammatory process leads to a violation of the anatomical architecture of the bronchial wall. In cystic fibrosis (CF) and primary ciliary dyskinesis (PCD), the main pathogenetic mechanisms of BE development are the accumulation of viscous inflammatory secretion in the airway lumen and a violation of the mechanisms of its escalation. The destruction of the bronchial wall occurs under the influence of the high proteolytic activity of neutrophil elastase and metalloproteinases. Micro-abscesses can form in the wall of the bronchus, in which pathogenic microorganisms colonize. In inflammatory foci, favorable conditions are created for the further growth of microorganisms, since their opsonization and clearance of phagocytosis products from the inflammatory areas of the bronchial wall are impaired. Necrotic neutrophils release a large amount of DNA, which affects the viscosity of bronchial secretions. This mechanism plays a key role in the development of BE in CF. Efferocytosis is a process of phagocytosis by neutrophils of apoptotic cells. It is realized through cell phosphatidylserine in the cycle of apoptosis and phosphatidylserine receptors on the surface of macrophages. However, in patients with BE, these mechanisms are inhibited by neutrophil elastase, which destroys phosphatidylserine. Thus, a deeper process of inflammatory damage to the bronchial wall is carried out [9].

CLASSIFICATION . In 1950, Eleanor Marie Reid classified BE in nature into cylindrical, cystic (saccular), and fusiform (varicose). According to her description, cylindrical BEs include diffuse edema of the bronchial mucosa, which are dilated and have straight, regular contours and end straight and abruptly. Although such characteristics are not always clinically demonstrable, morphological forms provide insight into the pathophysiology and the necessary descriptive terminology. Research has also shown that cystic morphology is most likely associated with Pseudomonas colonization and higher sputum production than other forms of BE.

Many classifications have been proposed that subdivide BEs by shape (cylindrical, varicose, cystic), content (dry, wet, filled), mechanisms of occurrence (obstructive,

destructive, traction, iatrogenic after radiation therapy, aggressive antibiotic therapy, etc.) [10, 11].

WM Thurbeck and MD Iseman [12, 13, 31] reflect the modern trend in the development of problems of BE classification most fully, their classification is based on etiopathogenetic principles.

Etiopathogenetic classification of BE:

- post-infectious (lower respiratory tract infections in children, abscessing pneumonia in adults, tuberculosis; adenovirus infection, whooping cough, measles; other respiratory tract infections);
- obstructive (foreign body, tumors, external compression of the airways);
- inhalation damage (inhalation of toxins, irritating gases, vapors, smoke, including thermal damage);
- aspiration (gastroesophageal reflux, aspiration pneumonia, sanitation procedures);
- genetically determined (cystic fibrosis, ciliary dyskinesia syndrome, Ewing's syndrome);
- congenital anomalies - dysplasia (agenesis, hypoplasia, sequestration, shunts, dwarfism);
- primary immune disorders (humoral defects, cellular or mixed disorders, neutrophil dysfunction);
- deficiency or abnormalities of α1-antitrypsin;
- chronic diffuse lung diseases of known or unknown etiology (idiopathic pulmonary fibrosis, collagenosis, sarcoidosis);
- idiopathic inflammatory disorders (ankylosing spondylitis, inflammatory bowel disease, recurrent polychondritis);
- other causes (allergic bronchopulmonary aspergillosis / mycosis, HIV infection / AIDS, yellow nail syndrome, radiation damage).

Questions remain about congenital and acquired causes of BE, their primary and secondary nature. Probably, bronchial damage occurs in individuals with genetic defects or a predisposition to pathology, since the vast majority of BE is formed in childhood and adolescence. The primary include BE in congenital anomalies and genetic diseases of the lungs. Secondary BE can be caused by any cause of lung damage.

The division of BE according to the clinical course (phases of remission and exacerbation), according to prevalence (lesion of segments, lobes of the lung, bilateral process) remains relevant. Expansion of endoscopic capabilities led to the subdivision of BE into central (proximal bronchi, including subsegmental) and peripheral (distal to subsegmental, from 5th to 16th gradation).

Depending on how the altered bronchi are dilated, bronchiectasis is divided into:

- cylindrical - the changed section of the bronchus looks like a cylinder;
- saccular - the wall of the bronchus is blown out in some weak point in the form of a «bag»;

• fusiform - the bronchial wall swells evenly, but more captures the bronchus along its length, which is why this area resembles a spindle;

• mixed.

Depending on the presence of atelectasis, bronchiectasis is divided into:

• atelectatic;

• without atelectasis.

Depending on the severity, there are:

• mild bronchiectasis - fully compensated;

• a pronounced form of bronchiectasis - the symptoms increase, but compensatory mechanisms partially cope with the disease;

• severe bronchiectasis - compensatory mechanisms cannot cope with the disease;

• complicated form of bronchiectasis.

According to the prevalence of bronchiectasis, there are:

• one-sided;

• bilateral.

There are two phases of the disease:

• aggravation - the height of the process;

• remission - morphological changes in the altered bronchi remain the same, but the clinical picture subsides [fourteen].

To choose the tactics for treating BE, it is advisable to single out the forms of their course:

1. Intermittent current.

2. Complicated course:

- pulmonary bleeding;

- pathology of the pleural cavity: spontaneous pneumothorax; empyema of the pleura;

- pathology of lung tissue: abscess and gangrene; cirrhosis;

- sepsis;

- extrapulmonary processes: stomach and duodenal ulcers; amyloidosis [15].

The symptoms of BE may be accompanied by a picture of the developed complications: hemoptysis and pulmonary hemorrhage (in 19.3% of patients), spontaneous pneumothorax (0.7%), abscess formation (1.8%), pleural empyema and pyopneumothorax (0.4%); in addition, sepsis, aspiration, erosive gastroduodenitis, stomach and duodenal ulcers, gastrointestinal bleeding, etc. are possible [16, 20].

CLINICAL CHARACTERISTICS. The course of bronchiectasis alternates with periods of exacerbation and remission. Usually exacerbations occur against the background of ARVI, in the off-season (spring, autumn). The course of bronchiectasis is considered optimal, in which exacerbations occur no more than 1 time in several years.

The most frequent and leading clinical symptom of bronchiectasis is cough with sputum. Deformation and expansion of the bronchus creates favorable conditions for

disrupting the discharge of sputum from the respiratory tract. Excessive production of sputum in the inflamed areas of the bronchus leads to stagnation. Sputum in bronchiectasis is «like in a bathroom». A change in body position (tilt, turn, transition from a horizontal position to a vertical one) leads to the movement of sputum and the occurrence of a reflex cough. Coughing up sputum can be mucous (white, transparent), mucopurulent (light yellow), purulent (yellow, light green, green). Sputum congestion and chronic inflammation can lead to bad breath, which is aggravated by coughing.

A sputum-producing cough may be absent with dry BE. In other cases, during the period of remission, cough with sputum is permanent or periodic, usually in the morning, not exceeding 50 ml of mucopurulent odorless sputum per day. During the period of exacerbation, the amount of sputum from the very beginning increases rapidly, reaching 300 - 500 ml or more.

Chest pain. A frequent symptom of bronchiectasis. The pain is provoked by coughing, chronic inflammation, deformation of the bronchial tree. Usually the pain corresponds to the side of the lung lesion by bronchiectasis. With bilateral bronchiectasis, the pain is bilateral. The intensity of pain increases during the period of exacerbation.

It is believed that the pain syndrome sometimes mimics the picture of pulmonary embolism. Pain syndrome often manifests itself in two forms. In the first, the pain is pleural in nature (a local manifestation with intensifications during a deep breath), in the second, the pain does not have a clear localization, but is felt in the form of a feeling of compression, distention, shortness of breath.

Dyspnea. In bronchiectasis, shortness of breath may be the result of the proliferation of fibrous tissue (pneumosclerosis) in the lungs and impaired gas exchange. At first, shortness of breath worries patients only during the period of exacerbation of bronchiectasis, but then it occurs constantly, increasing with physical exertion. In severe cases, there are signs of respiratory failure: cyanosis (cyanosis of the skin and mucous membranes), rapid breathing, the participation of auxiliary muscles in the act of breathing, fingers in the form of drumsticks. General weakness, fatigue, and decreased physical capabilities are noted.

Body temperature often rises. During the period of remission, an increase in temperature to a subfebrile level is episodic. During the period of exacerbation, there is a prolonged fever in combination with weakness, sweating, and sometimes chills.

Hemoptysis. This is a dangerous symptom of bronchiectasis. Hemoptysis speaks of deep damage to the bronchial wall and the involvement of bronchial arteries in the process.

The frequency of hemoptysis, according to the literature, ranges from 10-15% to 25-34% [18]. Pulmonary bleeding is noted in 10% of patients [19].

The disease is chronic in nature with periodic exacerbations of the process and the possible development of complications. In 82.6% of patients, BE is localized in one lung, including the right one in 23.7% of cases, and the left one - 58.9%. With right-sided

localization of the process, the lower lobe is most often affected (45.9%), and with left-sided localization, the lower lobe (58%) and its combination with lingular segments (26%). In bilateral BE, the lower lobes are most often affected (41.8%) [17].

DIAGNOSTICS.

Anamnesis. In the diagnosis of BE, it is important to take into account the characteristic signs, indications in the history of chronic, with exacerbations, for a long time infectious diseases of the lower respiratory tract (bronchitis, pneumonia), family history with information about risk factors. The motive for the examination may be a message about «asthma», hemoptysis.

The reason for the examination for the diagnosis of BE may be the pulmonary pattern in the lower pulmonary fields, reinforced with deformation, revealed by X-ray, reaching the peripheral parts of the lungs, the presence of atelectasis in the lower lobes, middle lobe, and reed segments.

Diagnosis of BE is aimed at identifying the disease, risk factors for the onset and development of the process, the form of the disease and its features, knowledge of which is necessary for adequate treatment.

Physical research. With a prolonged course of the disease, thickening of the terminal phalanges («drum sticks») and deformation of the nails («watch glasses») can be observed. Children may be lagging behind in mental and physical development. Deformation of the chest is possible, depending on the development of pulmonary fibrosis (decrease) or emphysema (barrel-shaped) and the prevalence of the latter (symmetric or asymmetric deformities).

With the development of pneumofibrosis, vocal tremor is enhanced, percussion - shortening of pulmonary sound, auscultatory - bronchophonia, and with severe emphysema, vocal tremor is weakened, percussion sound with a box shade, auscultatory - breathing is weakened. During auscultation, localization (zone of wheezing detection), process activity (number and variability of wet wheezing), volume and viscous consistency of bronchial contents (nature of dry wheezing), dynamics of the process (intensity, quantity, nature of wheezing) can be determined. With gross changes in the volume of the lung, a displacement of the heart can be physically detected.

Laboratory research. Blood tests should be aimed at determining the severity of the inflammatory process (level of leukocytosis, C-reactive protein, platelets, ESR), fungal lung disease (presence of eosinophilia), chronic infection (anemia).

Sputum examination provides for the determination of the type of pathogen (culture), nonspecific inflammatory process, mycobacteria, the presence of fungi, as well as the sensitivity of microflora to antibiotics. Bacteriological examination of sputum: to assess the sensitivity of bacteria to drugs; periodically (1-2 times a year) to exclude infection with mycobacteria and, if necessary, Aspergillus fumigatus.

Instrumental research.

Imaging methods: RG of the chest - the picture may correspond to the age norm in uncomplicated cases; in severe cases, an increase in the pulmonary pattern is determined due to the bronchial component: expansion of the lumen of the bronchi at various levels, thickening of the walls of the bronchi, their partial filling with sputum, with the formation of focal shadows, deformation of the pulmonary pattern, the formation of peribronchial muffs due to edema and fibrosis. In severe cases, areas of infiltration and atelectasis of the lung tissue of various lengths are visualized. High-resolution computed tomography (HRCT) clearly confirms the diagnosis (typical symptoms: expansion of the lumen of the bronchi and thickening of their walls, no decrease in the diameter of the bronchi along their length, visibility of the bronchi at a distance of <1 cm from the chest wall, sign of a ring).

Normally, the width of the bronchus at the height of a normal inhalation is quite stable and varies from 10 mm at the level of the lobar branches to tenths of a millimeter at the periphery. It is impossible to visualize unchanged bronchioles in the carapace layer of the lung even with the use of high-resolution CT. Consequently, the detection of the lumens of the bronchioles always indicates the existing pathology of the airways [21].

EBs, unless they are very large, are usually not visible on chest x-ray. In advanced cases of the disease, thickening of the walls of the airways, bronchiectasis and areas of compaction or collapse of the lungs are sometimes revealed. CT the chest organs is a much more sensitive method of examination, in which thickened dilated airways are detected.

Radiation symptoms of BE on a plain radiograph may be as follows:

1. Deformation and strengthening of the pulmonary pattern due to peribronchial fibrous and inflammatory changes; looped pulmonary pattern in the region of the basal segments of the lungs.

2. Thin-walled annular shadows (cavities), sometimes with a liquid level. This symptom most often occurs with saccular (cystic) bronchiectasis.

3. A decrease in the volume of the affected segments of the lungs, which can be manifested by a triangular shadow of post-telelectatic cirrhosis of the lower lobe that occupies the cardio-diaphragmatic sinus (the so-called basal triangles). In these cases, the mediastinal shadow is usually displaced towards the lesion with exposure of the opposite edge of the spine, the descent of the lung root, vicarious emphysema, and sometimes high standing and limited mobility of the corresponding dome of the diaphragm.

4. Atelectasized (cirrhotic) middle lobe on the lateral radiograph looks like a darkening band 2-3 cm wide, going from the root to the anterior costophrenic sinus.

5. Increased transparency (emphysema) of healthy segments of the lungs.

6. Symptom of «amputation» of the lung root [1].

High-resolution CT and CT are necessary for the diagnosis of bronchopulmonary pathology and are able to more or less distinguish some of the main causes of BE formation. In this regard, there are morphological provisions that need to be known for the diagnosis of BE:

- normally the bronchus is visualized within 1 cm from the pleural surface, which is true for the normal gradation of the bronchi, which narrow and almost adjoin the parietal pleura; the most reliable sign in the early formation of cylindrical BE is the absence of narrowing;

- an increase in bronchioloarterial relations: normally, the diameter of the bronchus is no more than 0.65–1.0 times the diameter of the adjacent branch of the pulmonary artery; an increase of 1 and 1.5 diameters is observed in healthy individuals, especially in those who live at high altitudes; an increase of 1.5 times or more indicates bronchiectasis.

A number of less reliable ray signs are also taken into account:

- thickening of the bronchial wall, which should normally be less than half the width of the associated branch of the pulmonary artery;

- accumulated mucus, air traps and mosaic perfusion (noticeable difference in lung tissue density on high-resolution CT).

So, the symptoms described on chest CT include:

1) a symptom of «tram rails, rails» - paired strips of thickened walls of the bronchi;

2) a symptom of an annular shadow;

3) the symptom of a «string of pearls»;

4) the symptom of «bunch of grapes» [1].

Bronchoscopy: in case of unilateral bronchiectasis, short duration of symptoms or hemoptysis.

Spirometry: in all cases recommended annually or more frequently; as a rule, obstructive ventilation disorders are found, the severity of which correlates with the severity of the disease; 1/3–2/3 of patients have bronchial hyperreactivity.

Other tests to find the cause: including cystic fibrosis, immune disorders, abnormalities in mucociliary clearance (eg saccharin test), allergic bronchopulmonary aspergillosis, CT scan of the paranasal sinuses.

Studies in hospitalized patients with exacerbations: microbiological examination of sputum (preferably taken before starting antibiotic treatment), chest x-ray (RG), pulse oximetry (or gasometry if indicated), blood culture (if fever is present), monitoring the amount of sputum secreted in during the day.

DIFFERENTIAL DIAGNOSTICS. Diagnosis of BE is not difficult, but sometimes it is not possible to distinguish between air cysts [29] and cystic BE. In addition, there is often a combination of these conditions in pulmonary dysplasia [22]. Filled BEs (retention cysts) can mimic tumor pathology; in the young - hamartoma, adenoma, carcinoid, in the elderly - lung cancer. The deposition of calcium salts on the walls of retention cysts raises suspicions of tuberculoma [23]. In these cases, intravenous administration of contrast agents helps to establish the nature of changes in the degree of accumulation in education or its absence. In avascular formations - retention and parasitic cysts, mature tuberculomas, lung infarctions - even a slight decrease in density is recorded after the administration of a contrast agent.

Errors in the diagnosis of broncho-bronchioectasia can be explained by artifacts from the movement of the heart, manifested by doubling of the contours of the bronchi and blood vessels, which simulates cylindrical BEs. False bronchiectasis can be caused by the summation of bronchial lumens at the bifurcation levels of tomography [30]. A picture similar to BE is observed in Langerhans cell histiocytosis, destructive inflammation (tuberculosis, septic-metastatic pneumonia), some pseudocavitary formations - Pneumocystis pneumonia, bronchioloalveolar cancer.

Hemoptysis is a common complication of BE. There is no connection between the number, size of BE and hemoptysis; often single, small BEs are a source of threatening bleeding. In such cases, CT is used in conjunction with endoscopy to localize the source of bleeding. On the side of the lesion, accumulations of blood in the bronchi and subpleural foci of aspiration pneumonia are visible.

Filled BEs (retention cysts) in patients with atopic asthma suggest possible allergic bronchopulmonary aspergillosis (ABPA). Due to a prolonged allergic reaction antigen - antibody on the mucous membrane of the bronchi, granulomatous inflammation occurs with obliteration of the lumens and subsequent stretching of mucus. Subsegmental bronchial involvement is typical.

Retention cysts are combined with impaired ventilation, regional emphysema, BE. Histologically, in ABPA, various stages of chronic nonspecific inflammation, spreading beyond the bronchi into the pulmonary parenchyma, are determined; eosinophils and mycelium of the fungus are found in the bronchial secretion. Diagnosis of the disease is based on laboratory data (eosinophilia, increased IgE levels, determination of fungal mycelium in sputum, antibodies to aspergillus in blood serum, positive skin tests), symptoms (dyspnea, asthmatic component) and is supplemented by a CT scan [24, 25]. In addition to severe asthma, ABPA often complicates cystic fibrosis.

Cystic fibrosis is a systemic disease of exocrine glandular tissue, accompanied by sinusitis, nasal polyps, bronchiectasis. This inherited disorder is transmitted in an autosomal recessive manner. The genetic defect disrupts the transport of electrolytes in the glandular epithelium [26]. In particular, the cells of the bronchial mucosa become impermeable to chlorine ions, which increases the viscosity of the secretion and complicates the work of the mucociliary apparatus. The associated infection destroys the structure of the lower respiratory tract and leads to irreversible bronchiectasis. Cystic fibrosis is observed exclusively in childhood and young age and is characterized by widespread central cylindrical bronchiectasis and multiple mucous blockages [27].

Pathogenetic similarity due to impaired airway clearance is observed in other congenital dysfunctions of the mucociliary apparatus, for example, in Kartagener's syndrome. It was found that the mirror position of the organs «disorients» the work of the ciliated epithelium, leads to stagnation, accumulation of mucus and the addition of a secondary infection. Unlike cystic fibrosis, the middle and lower lobes are primarily affected. Recurrent inflammations cause left-sided mid-lobe syndrome with disseminated

foci and branched structures in the lower parts of the lungs, which reflects the picture of widespread infectious bronchiolitis [28].

TREATMENT: The methods of treatment remain relevant: conservative and surgical. Attempts to give one of them a dominant character did not come true. Each of them pursues specific goals at different stages of the development of the disease. Conservative treatment can be carried out with an independent task of clinical recovery and stabilization of the process, ensuring the quality of life, preventing the exacerbation of the process, its progression. On the other hand, conservative treatment can be considered as preoperative preparation, postoperative rehabilitation and prevention of disease progression and recurrence. Conservative treatment includes airway hygiene, sanitation of the bronchial tree, antimicrobial therapy, specific treatment for individual causes of the development of the disease, correction of hypoxia, etc.

1. Pulmonary rehabilitation: procedures to facilitate the release of stagnant sputum from the bronchi - postural drainage in combination with vibration, shaking and tapping of the chest, exercises for controlled breathing (including breathing through pursed lips), etc. up

2. Antibiotic therapy: in the treatment of acute infections, initially empirically (antibiotics active against H. influenzae and S. aureus - amoxicillin + clavulanate (for example, 625 mg 3 × daily), in patients with hypersensitivity to penicillins - macrolide (clarithromycin 500 mg 2 × daily, or azithromycin 500 mg 1 × daily) Large bronchiectasis and chronic colonization of H. influenzae → high dose antibiotics (eg, amoxicillin 1 g 3 × daily) In patients with P. aeruginosa colonization → ciprofloxacin After receiving antibiotic culture results → targeted antibiotic therapy, usually for 2–3 weeks.

3. Other drugs: intermittent mucolytics (only in patients with cystic fibrosis constantly dornase α), bronchodilators (β 2 -agonists, anticholinergic drugs) in case of bronchial hyperreactivity.

4. Surgical treatment: is in most cases a radical method of treating bronchiectasis. The operation is indicated in the case of the location of bronchiectasis in a limited space, with bronchiectasis with frequent exacerbations, and with its complications (hemoptysis, frequent pneumonia), when conservative methods cannot achieve stable remission. The main options for surgical treatment of bronchiectasis are lobectomy (removal of a lobe of the lung) or segmentectomy (removal of a segment of the lung). The most effective operation is for unilateral localization of bronchiectasis. An alternative method for bleeding may be bronchial artery embolization.

Indications for surgical treatment: pulmonary bleeding; pneumothorax not controlled by drainage; frequent exacerbations of the process that are not amenable to conservative treatment; inability to achieve stable remission within 2 - 3 years; cirrhosis of the lung zone with a purulent process in the sharply dilated bronchi.

Conditions for carrying out operations: stopping the process in the area of the alleged intersection of the bronchi; bronchoscopy with obturation of the regional bronchus with

significant purulent discharge; bronchoscopy at the end of the operation; limited areas of the lung with bronchiectasis; maximum removal of lung segments with bronchiectasis and preservation of intact lung tissue.

Contraindications to surgical treatment: bilateral widespread process; severe concomitant diseases; Pulmonary heart.

Conclusion. BEs are quite common in modern clinical practice, which was significantly facilitated by the introduction of image diagnostics into the modern diagnostic algorithm of respiratory diseases. The clinician is always faced with intense diagnostic and therapeutic work when making a diagnosis of BE. Progress has been achieved not only in the field of image diagnostics, but also in the field of genotyping of certain forms of BE, immunology and microbiology. However, with significant progress in understanding the nature of BE development, idiopathic BEs persist. This diagnosis is the exclusion of other very diverse and often rare forms of pulmonary tissue pathology that may manifest as the development of BE. The treatment algorithm assumes a certain sequence of both medication and non-medication methods of treatment. In a number of patients with BE, there is a need for urgent measures, including embolization of the bronchial arteries or emergency surgical treatment to remove part of the lung tissue that is the source of bleeding.

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