Case Report

Clinical Case of Tracheobronchopathia Osteochondroplastica

Natalia V. Chaplynska, Mykola M. Bagrii, Victoria T. Rudnyk, Mariana Y. Koniuska, Svitlana L. Filipova, Liubomyra O. Yacenta

Abstract
Tracheobronchopathia osteochondroplastica (TO) is a rare chronic disease of the airways characterized by the growth of cartilage and/or bone tissue in the submucosal layer of the trachea and large bronchi with varying degrees of their lumen narrowing. In a significant percentage of cases diagnosis is made only posthumously due to the late onset of clinical manifestations and the considerable complexity of life-time diagnostics.

The objective of the research was to conduct the clinical observation of OT.

Materials and methods. A patient Sh., 49 years of age, was admitted to the department of differential diagnostics of regional clinical pulmonology center to determine the cause of prolonged cough and bronchial obstruction syndrome. Only fiber-optic bronchoscopy (FBS) was the only informative method of examination. On the mucous membrane of the trachea and right main bronchus the dense white protuberance was found. The biopsy material was taken. Histological examination revealed areas of calcified cartilage located in subepithelial layer.

Results. Taking into account the reassessment data, the TO was suspected. The disease has a benign course for many years and is asymptomatic at initial stages. However, deformation and narrowing of the lumen of the trachea and the large bronchi with epithelial atrophy causes a disturbance of phlegm discharge. This promotes the development of infectious and inflammatory processes in the lung tissue. Patients complain of cough, hemoptysis, shortness of breath, chest pain. X-ray examination of the lungs may detect the infiltrative changes. The indicators of lung function are normal or correspond to the type of obstructive disorders. CT radiograph and virtual bronchoscopy determine the deformation and narrowing of the trachea and the large bronchi due to the elements of cartilage and bone tissue. Definitive diagnosis is possible only according to the results of FBS and histological examination.

Conclusions. Awareness on such rare pathology as TO is essential, especially in patients with chronic cough. In case of absence of specific clinical and radiographic changes, the computed tomography of the chest and FBS with histological examination sampling should be performed to confirm the diagnosis.

Keywords
chronic cough; tracheobronchopathia osteochondroplastica

Problem statement and analysis of the recent research

Tracheobronchopathia osteochondroplastica (TO) is a rare chronic disease of the airways characterized by the growth of cartilage and/or bone tissue in the submucosal layer of the trachea large bronchi with varying degrees of their lumen narrowing. It was first described in detail in case of autopsy in 1855 by Carl von Rokitansky, a professor of pathological anatomy at the University of Vienna. The first description of in vivo established changes (during the laryngoscopy) was made by HS Muskelton in 1909. Only 90 observations of TO were described in the world medical literature until 1947, and 340 descriptions of this form of disease were made until 1993. Disease occurs at the age of 23-81 years, but more often in people over 60 years of age, with roughly equal frequency in men and women [2, 3, 8]. Isolated cases of the disease in children were described. In a significant percentage of cases diagnosis is made only posthumously due to the late onset of clinical manifestations and the considerable complexity of life-time diagnostics. Accidentally TO may be diagnosed during the tracheal intubation, computed tomography (CT) of the chest and fiber-optic bronchoscopy (FBS) performed according to other indications [5].

TO etiology remains unknown. The main hypotheses of trachea and bronchi elastic tissue metaplasia in bone and cartilage are such as congenital origin of these changes, violation of the acid-base balance and subsequent inflammation of the trachea and bronchial tree leading to the transformation of histiocytes into osteocytes. They may occur due to chronic bronchitis or specific inflammation (tuberculosis, syphilis).
The tumor growth as multiple osteoma type is not excluded. TO may also be the final stage of primary respiratory amyloidosis [1, 4, 7].

TO often affects middle and lower third of the trachea and its bifurcation, main, partial and segmental bronchi. Macroscopically dense whitish nodules 1-3 mm in diameter with irregular contours are detected. They are protruding above the surface of the mucosa (“typical picture of “metal grater”), located exclusively in the cartilage of the trachea and never in its membranous part. Microscopically, insular proliferations of trabecular bone and cartilage are detected in the atrophied submucosal layer of the trachea and bronchi. Bone tissue of these islands has a mature lamellar structure and may contain elements of the marrow. Cartilage islands tissue can be fibrous, elastic or hyaloid; some parts of the newly formed cartilage are located under the epithelium [1, 2, 7].

**The objective of the research** was to conduct the clinical observation of OT.

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### 1. Materials and methods of the research

Patient Sh., 49 years of age, was admitted to the department of differential diagnostics of regional clinical pulmonology center 08/01/2016 to determine the cause of bronchial obstruction syndrome. The patient complained of paroxysmal cough with the release of a small amount of muco-purulent sputum, dyspnea of mixed nature after excessive exercise, sweating, general weakness.

The patient was ill during 6 months, when the annoying cough had appeared. He had never previously addressed for medical care. The patient suffered from cold-related diseases and had acute sinusitis in 1996. The patient did not smoke.

At the time of admission, the patient’s condition was satisfactory. Skin and visible mucous membrane were of normal color, clean. Peripheral lymphatic nodes were not enlarged. Thorax was normoesthenic. Respiratory rate (RR) = 19/min. SpO2 = 95%. Percussion lung sounds were clear, lungs borders were within normal limits. During the auscultation the breathing was harsh. During the forced expiration dry wheezing could be heard in both lungs. Heart sounds were clear and rhythmic. Heart rate constituted 86/min. BP was 120/80 mmHg. The abdomen was of normal configuration, involved in breathing. It was soft, not painful during the palpation. The liver did not protrude from the edge of the right costal arch, was not painful. Pasternatsky’s symptom was negative on both sides.

Laboratory studies were conducted. Complete blood count, urinalysis and biochemical blood assay did not detect any pathological changes. General sputum analysis provided the following results (08/02/2016): 20-30 leukocytes per HPF; 3-6 elements of bronchial epithelium per HPF; fibrin (+); atypical cells were not found. Bacterioscopic sputum analysis did not detect Koch’s bacillus.

The instrumental research was started with spirometry. The indicators of respiratory function were within normal limits. According to chest X-ray (08/01/2016) the irregular thickening of lung pattern in the lower lobes of the lungs was detected. The roots of the lungs were structural. Sinuses were free. Heart size was normal. Medical report: bronchitis.

Taking into account a history of sinusitis, the radiography of the paranasal sinuses was conducted (08/01/2016): the deformation of the nasal passages, nasal cavities, nasal septum. Deviated nasal septum was confirmed, chronic rhinitis was diagnosed during the otolaryngological examination (08/10/2016).

Computed tomography of the chest (08/9/2016) detected the irregular thickening of the walls of the trachea and right main bronchus with single calcifications (Figure 1).

Considering the fact that according to clinical and X-ray examination no changes were detected except chronic bronchitis and bronchial obstruction was present despite treatment with bronchodilators, fiber-optic bronchoscopy (FBS) was performed on 08/08/2016. Dense white protuberances 2-8 cm in diameter arranged circularly were found on the mucous membrane of the trachea. The biopsy was taken from a similar formation on the medial surface of the right main bronchus. Histological material (Figure 2) was represented by small fragments of mucosa with signs of basal epithelial hyperplasia (of reactive genesis), hemorrhage (of secondary mechanical nature) and slightly marked subepithelial lymphocytic-macrophage infiltration. Large areas of calcified cartilage were present in subepithelial layer. The above changes were a manifestation of the tracheobronchial chondro-ostopathy.

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### 2. Results of the research and their discussion

TO was suspected according to the results of examinations. The disease has a benign course for many years and is asymptomatic at initial stages, rarely causing critical airways stenosis. However, the reduction of their diameter and deformation of lumen with epithelium atrophy cause the violation of sputum allocation. This promotes the development of infectious and inflammatory processes in the lung tissue [1]. Lung cancer...
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Figure 2. Tracheobronchial chondroosteopathy. Biopsy material of the mucous membrane of the right main bronchus.

1 - respiratory epithelium with signs of basal cell hyperplasia;
2 - subepithelial hemorrhage;
3 - calcified cartilage in the submucosal layer.

Staining: hematoxylin and eosin.
Magnification: x200.

can occur on this background [7]. As in our case, according to the literature, TO clinical manifestation are cough with a little amount of sputum, hemoptysis (60%, may be due to ulcer or friction of tubercules), dyspnea (53%), chest pain [6]. Sometimes there is hoarseness (when the larynx is injured). Some patients have recurrent pneumonia [5].

During the X-rays examination pathological changes are usually not defined for many years. In some cases, secondary inflammatory changes in the lungs can be identified. CT scan and virtual bronchoscopy detect the discoid particles of cartilage and bone tissue. They protrude into the lumen of trachea and bronchi, deform and narrow their lumen [2].

At an early stage of the disease during the fiber-optic bronchoscopy rare whitish protuberances are visualized. They are located on the front and side walls of the trachea and the large bronchi (along the cartilage rings) and are of rocky density during the instrumental palpation. These changes give the walls of the trachea and major bronchi the view of “streaks of hardened wax”, “block stone”, “rock garden”. With further development of the disease trachea and main bronchi look stiff, rigid tubes, close to the density of the bone tissue. The mouth of the main bronchus, the mobility of which is sharply reduced, is visualized as a narrow ring with a whitish scar edges. Mucous or muco-purulent secret is in a significant amount. The mucous membrane is tightly fused to the underlying tissues; its contact bleeding is defined [2, 3, 7].

Main indicators of lung function in patients with TO may be normal as in our patient or respond to obstructive variant of curve “flow-volume”. During the test with bronchodilatators is performed functional indices are increased by the amount of not more than 15% of the initial values. It shows the prevalence of organic (narrowing of the airways by protrudences) rather than functional (bronchospasm) changes in these patients [4, 6].

TO must be differentiated from tumors of various origins, tracheobronchial papillomatosis, endobronchial sarcoidosis, tracheobronchial amyloidosis, calcified endobronchial tuberculosis, fungal lung disease, Wegener’s granulomatosis, relapsing polychondritis [1, 8]. Definitive diagnosis is possible only based on the results of histological studies [1, 7]. In order to clarify the diagnosis the patient was directed to the National Institute of Phthisiology and Pulmonology. The pathology was confirmed.

TO specific treatment does not exist [1]. Symptomatic therapy is aimed at the elimination of concomitant inflammatory process in the bronchi (ultrasonic alkaline inhalations, sanation bronchoscopy, mucolytic and antibacterial drugs) [4]. The positive effect in some cases is obtained from steroids and desensitizing agents [2]. Such surgical methods as cryotherapy, laser therapy, bouginage, removal of nodules with kelectome during FBS, application of endotracheal or endobronchial stent can be used in case of significant stenosis of the lumen of the trachea and bronchi [5, 6].

In most cases the prognosis of the disease is favorable. The disease progression may be slow. The spontaneous termination of new bone appearance and tracheobronchial calcareous formations may occur. TO significant progression is observed in approximately 17% of cases. Stenosis of the respiratory tract requiring surgical intervention is rare [2, 8].

3. Conclusions

Knowledge of such rare pathology as TO is essential, especially in patients with chronic cough. In case of specific clinical and radiographic changes absence, chest computed tomography and FBS with sampling for histological examination should be performed to confirm the diagnosis.

4. Prospects for further research

Every doctor in his medical practice sometimes encounters with patients with rare diseases, with which he has not met yet and sometimes not even heard or read. Although, according to I.P. Pavlov, “clinical casuistry forever remains a rich source of new unexpected facts” [3]. The above mentioned clinical observation is a proof of such cases and confirms the difficulties of the diagnostic process in routine clinical symptoms. All possible methods should be used in difficult diagnostic cases. Morphological verification of pathology is always the most accurate, but requires careful evaluation of clinical status of patients and the risks associated with its implementation.
References


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