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Case report: Williams-Campbell syndrome

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Summary

Background:	Williams-Campbell syndrome is a rare type of bronchiectasis that is due to deficiency or absence of cartilage in the fourth- to sixth-order bronchi.
Case Report:	The paper presents the case of a patient with large, bilateral bronchiectasis caused by defect of cartilage in the fourth- to sixth-order bronchi referred to as Williams-Campbell syndrome.
Conclusions:	Williams-Campbell syndrome should be taken into consideration in differential diagnosis of bronchiectasis. Both inspiratory and expiratory high-resolution computed tomography should be performed to establish the diagnosis.
Key words:	bronchiectasis • Williams-Campbell syndrome
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Background

Williams-Campbell syndrome was first described in 1960. It is a rare type of bronchiectasis that is due to deficiency or absence of cartilage in the fourth- to sixth-order bronchi [1,2]. In vast majority of cases it is a congenital disease manifested in early childhood by productive cough, dyspnea and recurrent lower respiratory tract infections.

Fewer than 100 cases of the disease, including three cases of familial occurrence of the syndrome, have been described to date [3]. Single reports concerning acquired form of the disease due to adenovirus infection have been described [4]. Subclinical forms, sometimes diagnosed as late as in adulthood, are encountered as well [2].

The paper presents a case of Williams-Campbell syndrome in a 27-year-old female with recurrent pneumonias and bronchiectasis.

Case Report

A 27-year-old female was admitted to the Department of Thoracic Surgery at the National Tuberculosis and Lung Diseases Research Institute with a suspicion of Hodgkin lymphoma based on computed tomography performed outside the Institute, which revealed a tumor-like structure arising from the mediastinum. Parasternal mediastinotomy

based on the scan was performed. Histopathological assessment of the collected material did not reveal the presence of tumor cells, but only the signs of „mucoid impaction”, i.e. inflammatory changes in the bronchi filled with a large amount of mucoid discharge.

The patient's history was analyzed thoroughly and the diagnostics was extended.

The onset of symptoms had been observed at 9 years of age, with productive cough, dyspnea and frequent lower respiratory tract infections. The patient had been hospitalized many times because of recurrent pneumonias, diagnosed with bronchial asthma, treated with glyocorticoids and periodically with antibiotics.

Radiograms performed during numerous hospitalizations revealed bilateral parabronchial lesions and inflammatory opacities. Bilateral bronchiectasis was confirmed by bronchography. In bronchoscopy, abundant discharge, sometimes very thick, causing bronchial obstruction was observed.

Chest CT was repeated in the Tuberculosis Institute. The tumor-like mass adjacent to the mediastinum was assessed as atelectasis of the left upper lobe due to bronchial impaction. On the background of atelectatic changes – present in both lungs, oval hypodense areas with various attenuation coefficients, which might correspond to severe bronchiecta-

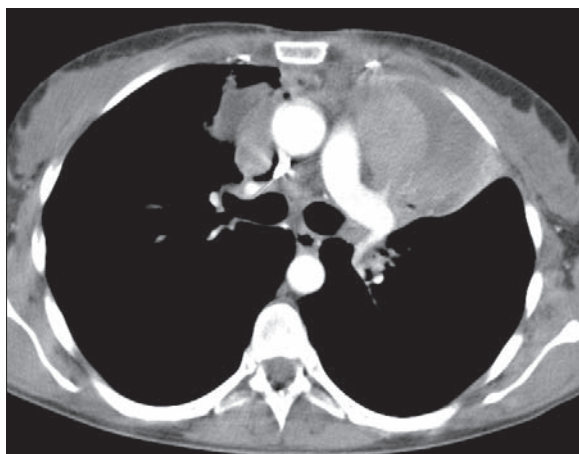


Figure 1. Contrast-enhanced CT, transverse section. Atelectasis of the left superior pulmonary lobe with oval areas of inhomogeneous density visible on that background. Smaller changes of similar character on the right side.

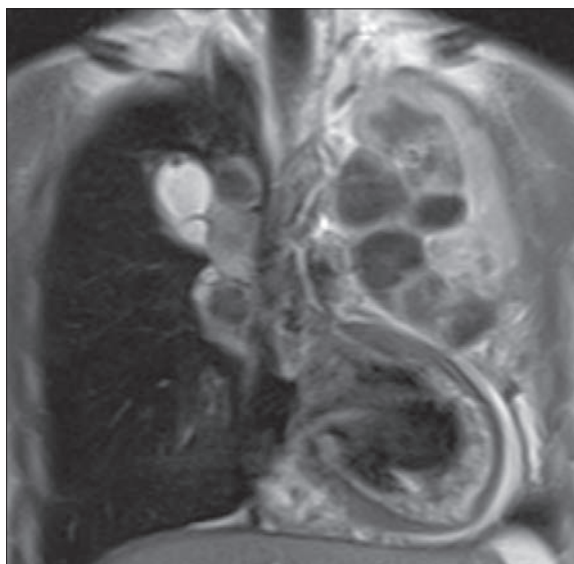


Figure 2. MRI, SS TSE (HASTE), T2-weighted image in the frontal plane. Large, cyst-like spaces corresponding to bronchiectasis, filled with discharge, visible on the background of atelectatic changes.

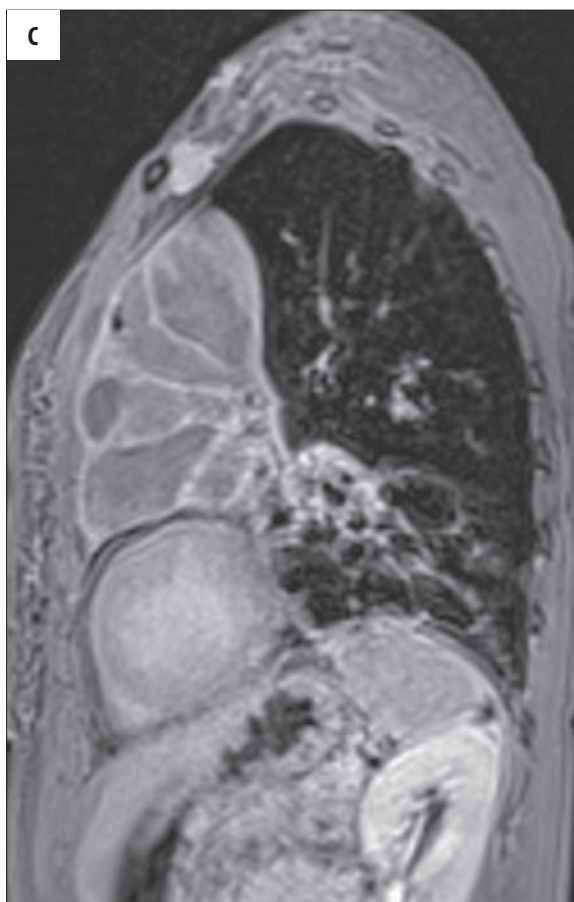
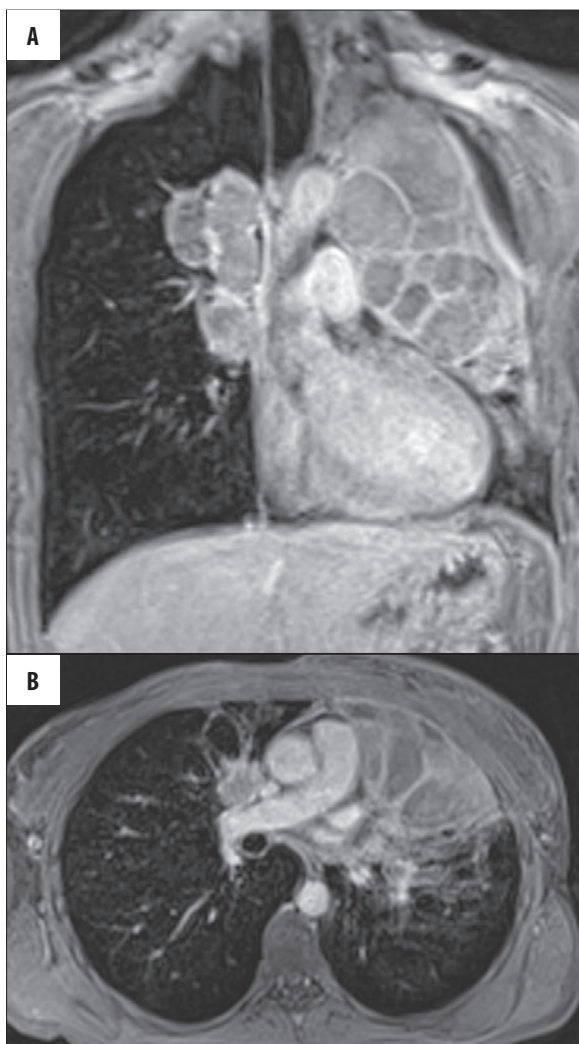


Figure 3. Contrast-enhanced MRI, GRE sequence, T1-weighted image with fat saturation (3D VIBE). (A – frontal plane; B – transverse plane; C – sagittal plane). Contrast enhancement of the atelectatic area and bronchi affected by bronchiectasis visible on its background. Bronchiectasis visible also in the remaining lung parenchyma.

sis filled with discharge, were visualized (Figure 1). Cyst-like and cylindrical bronchiectasis spaces, partially filled with mucus, were also visible in other areas of lung parenchyma.

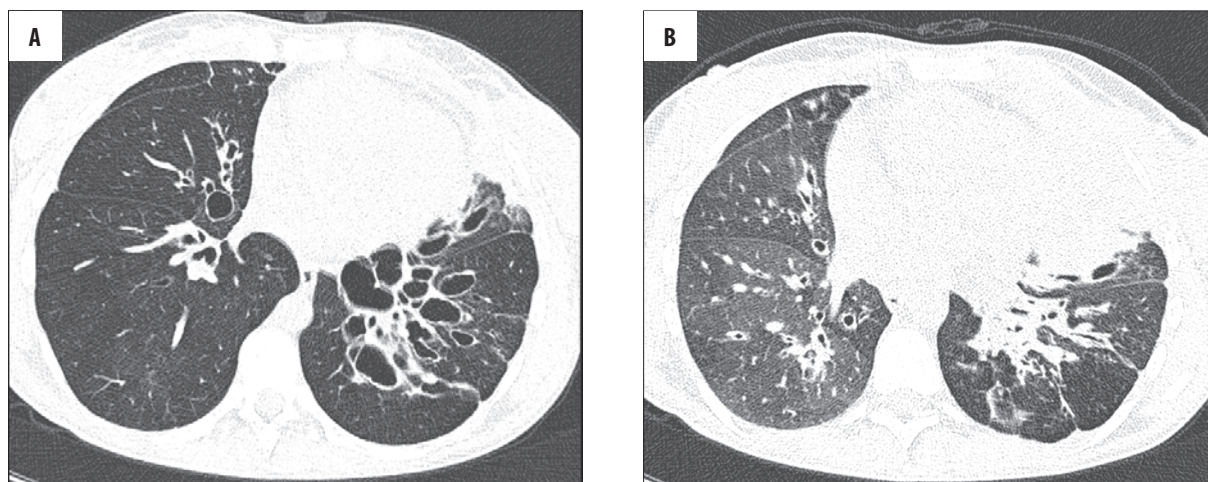


Figure 4. Chest HRCT scan (A – inspiration, B – expiration). In the inferior lobe and the lingula of the left lung, as well as in its middle lobe, cyst-like bronchiectatic areas of various sizes, distending during inspiration and collapsing during expiration, characteristic of Williams-Campbell syndrome.

On MRI, mediastinal structures were found to be normal, with better visualization of mucoid discharge filling the bronchiectasis areas than in CT (Figures 2,3A-C)

Bronchoscopy revealed marked retention of mucoid discharge in the bronchi. Mycological investigations did not reveal the presence of *Aspergillus fumigatus*. Cystic fibrosis was also excluded.

HRCT scan performed in both respiratory phases revealed significant distension during the inspiratory phase, and during the expiratory phase – characteristic signs of collapse of the airways affected by bronchiectasis, as well as signs of air trapping (Figure 4A,B).

Williams-Campbell syndrome was diagnosed in the 27-year-old patient on the basis of diagnostic examinations (especially HRCT) and anamnesis data.

Discussion

Patients with Williams-Campbell syndrome demonstrate bilateral bronchiectasis characteristically distended in the

inspiratory phase and collapsing in the expiratory phase. Thick mucoid discharge retention in the bronchi causes atelectatic changes. The diagnosis is based on bronchoscopy and chest HRCT [2,5]. Differential diagnosis includes other congenital and acquired conditions associated with bronchiectasis: sinobronchial syndrome, tracheobronchomegaly, ciliary dyskinesia, cystic fibrosis, allergic bronchopulmonary aspergillosis, immune deficiency [1,6].

The course of the disease is characterized by recurrent pneumonias and respiratory failures. No causal treatment is available, exacerbations are treated with antibiotics and glucocorticosteroids. A single attempt of lung transplantation in a patient with Williams-Campbell syndrome has been described [7].

Conclusions

Differential diagnosis of patients with bronchiectasis should take into consideration also rare Williams-Campbell syndrome. To establish the diagnosis of the syndrome, inspiratory and expiratory HRCT scan and detailed analysis of the patient's history is necessary.

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