Bronchial asthma with systemic sclerosis – a case study

Współistnienie astmy oskrzelowej z twardziną układową – opis przypadku

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Słowa kluczowe: astma oskrzelowa, twardzina układowa, układ immunologiczny.

Abstract

Asthma affects from 1% to 18% of people depending on the country, and systemic sclerosis affects 0.025%. Asthma is a chronic inflammatory disease of the respiratory tract. Inflammation is induced by a plurality of cells and their release of mediators. Systemic sclerosis is a connective tissue disease belonging to the so-called major systemic diseases characterised by vascular damage, immune disorders, and excessive accumulation of extracellular matrix components in various internal organs and skin. As the disease progresses, they lead to multi-organ failure, and eventually death of the patient. It was formerly thought that there was no connection between diseases within the circle of allergies and autoimmunology. Currently, it is assumed that the cause of both disease groups is a disorder of the immune system. An example of this type of relationship is described in our clinical case where in one patient bronchial asthma coexists with systemic sclerosis.

Streszczenie

Na astmę oskrzelową choruje od 1% do 18% ludności w zależności od kraju, natomiast na twardzinę układową ok. 0,025%. Astma jest przewlekłą chorobą zapalną dróg oddechowych. Zapalenie to jest wywołane przez liczne komórki i uwalniane przez nie mediatory. Twardzina układowa to choroba tkanki łącznej należąca do tzw. dużych chorób układowych, która charakteryzuje się uszkodzeniem naczyń, zaburzeniami immunologicznymi i nadmiernym gromadzeniem się składników macierzy pozakomórkowej w różnych narządach wewnętrznych oraz w skórze. W miarę postępu choroby prowadzą one do niewydolności wielonarządowej, a w końcu do śmierci chorego. Dawniej uważano, że pomiędzy schorzeniami z kręgu alergii i autoimmunologii nie ma powiązań. Obecnie przyjmuje się, że przyczyną obu grup schorzeń jest zaburzenie funkcjonowania układu immunologicznego. Przykładem tego typu powiązań jest opisany w niniejszej pracy przypadek kliniczny, gdzie u jednego chorego współistnieje astma oskrzelowa z twardziną układową.

Introduction

Asthma is a global problem. Estimates indicate that it affects approx. 300 million people. Asthma affects from 1% to 18% of the population depending on the country [1]. In Poland, from 5% to 10% of the population are affected. Asthma is a chronic inflammatory disease of the respiratory tract. Inflammation is induced by a plurality of cells and their release of mediators. The most important cells are ongoing inflammation eosinophils, mast cells, and Th2 lymphocytes. However, in severe asthma an important role is played by neutrophils [1]. Clinical signs of the disease can be controlled with appropriate treatment. Correctly applied (depending on severity) early treatment reduces the likelihood of irreversible changes. We know that in most patients with asthma, rhinitis coexists. Inflammation of the lower airways is accompanied by inflammation of the upper respiratory tract. This con-

firms the concept of a single disease, which is, among other things, related to the similarity of rhinitis and bronchitis [2–4]. This indicates the involvement of the whole of the mucosal disease process, and thereby a part of the natural immune mechanism.

Systemic sclerosis is a disease of the connective tissue belonging to the so-called major systemic diseases characterised by vascular damage, immune disorders, and excessive accumulation of extracellular matrix components in various internal organs and skin. As the disease progresses, they lead to multiple organ failure, and ultimately death of the patient [5]. The course of scleroderma may also lead to changes in the respiratory tract – usually to interstitial pulmonary fibrosis [6]. Nonetheless, large-scale clinical trials have shown that causal treatment of systemic sclerosis does not exist, and disease-modifying therapy is still ineffective [5]. This is also due to the very diverse and virtually impossible to predict clinical course. In addition, these difficulties are due to the fact that systemic sclerosis is a relatively rare disease, affecting only 0.025% of the general human population [5]. According to estimates, in Poland scleroderma has been diagnosed in about 10,000 people [5]. It was previously thought that the circle between diseases and allergy autoimmunology has no ties. Currently, it is assumed that the cause of both groups of diseases is of the immune system disorder [7–9]. For example, one of the key cells involved in the development of allergic reactions – a mast – is also involved in scleroderma, autoimmune, and Th17 cells, and the interleukin family of IL-17 is involved in both types of lesions [7]. In contrast, non-atopic asthma has also been found to involve autoantibodies located in the respiratory tract [7, 9].

The aim of the study was to show the links between diseases from allergic and autoimmune diseases on the basis of the coexistence in one patient of bronchial asthma with systemic sclerosis.

Case report

We analysed a case of 54-year-old man who, 11 years ago, volunteered to a pulmonary clinic with shortness of breath, coughing, tightness in the chest, and "wheezing". On the basis of the following studies: history, physical examination, spirometry, X-rays – chest X-ray, and high-resolution computed tomography (HRCT) he was diagnosed with asthma, allergic rhinitis; we also found nodular lesions in the lungs. For this reason, the patient was hospitalised on several occasions in the pulmonology unit.

The HRCT studies performed during subsequent stays in the ward showed no signs of progression of nodular lesions. Four years ago, the patient went through right lung inflammation of the pleura pleural empyema. At that time he was also hospitalised for thoracic surgery in order to perform a thoracotomy and decortication. During this period, the patient noticed progressive hardening of the skin of the face, arms and legs, occurring at home. For this reason, he was sent for rheumatological consultation. In 2013 the department rheumatology, based on the results of a physical examination (hardening and thickening of the skin: forehead, cheeks, nose and chest, forearms, thighs and drumsticks), laboratory tests (rheumatoid factor levels, elevated muscle enzymes), imaging (in HRCT fibrotic lesions and areas with an opaque glass), and capillaroscopy (changes for the active form of scleroderma) was diagnosed in a patient-generalised form of systemic sclerosis. Currently the patient is under multidisciplinary care: pulmonary, allergy, rheumatology, and cardiology. He is located in a stable state without progression of any diagnosed disease. Treatment of the patient is, however, multidrug. This includes not only inhaled corticosteroids (ciclesonide used in large doses), characteristic of bronchial asthma and nasal used in rhinitis, but also a system (encorton) implemented due to systemic disease of the connective tissue, and immunosuppressive therapy (mycophenolate mofetil). A previous attempt at immunomodulatory therapy with azathioprine was stopped due to deep pancytopaenia. The recently completed study of respiratory function showed no significant deviations – without obstruction, only features a mild restrictions. In the study, blood gas analysis showed no signs of respiratory failure.

Discussion

We found no reports of coexistence in one patient of bronchial asthma and systemic sclerosis in the literature as well as the world. There is very little literature on this topic. This is probably related to the rare occurrence of systemic sclerosis (0.025% of the population) and even rarer interaction of these two units together. However, it draws attention to the fact that the immune system is involved in both of these diseases [1, 4, 5]. Common diseases in both groups impairment of their immune system indicates a relationship between the diseases from allergic and autoimmune diseases. This confirms the participation of the two groups of ailments of the same cells: mast cells, lymphocytes, and the presence of Th17, especially asthma endogenous autoantibodies located in the airways [7–12]. Recent observations suggest that autoimmune diseases and allergies are more connected with each other than believed so far. Both groups of diseases characterised by disorders of immune tolerance. Furthermore, in patients suffering from chronic atopic diseases the presence of autoantibodies IgE has been demonstrated [13, 14]. Several studies indicate a close relationship between polymorphisms of HLA genes (histocompatibility antigens), and the development of allergies and autoimmune diseases. Some of the polymorphisms of the gene may act as risk factors other as protective

agents [15]. Currently, it is assumed that the cause of both diseases groups is a disorder of the immune system, while the above mechanisms indicate the possibility of mutual relations between these disorders [5, 16, 17]. There are also publications emphasising that the two groups of diseases are not mutually exclusive but can coexist [18]. Also, research in animals show us the possibility of their mutual influence on each other [19]. An example of such a link is described by our clinical case where in one patient bronchial asthma coexisted with systemic sclerosis. These correlations still require further research. There are also publications emphasising the protective role against the development of asthma and autoimmune diseases [20].

Conclusions

On the basis of this case, it seems to be that since the cause of both groups of diseases includes immune disorders they can coexist with each other, but even the group of allergic diseases may predispose to the development of the disease from the autoimmune group (and vice versa). It can be assumed that these cases are more frequent but are often not fully recognised, and the mutual influence on other illnesses of these two groups requires further study.

Conflict of interest

The authors declare no conflict of interest.

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