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Case report of Churg-Strauss syndrome — signs and symptoms suggesting disseminated neoplasmatic disease

Abstract

Churg-Strauss syndrome (CSS) is characterized by asthma, sinusitis, hypereosinophilia and eosinophilic infiltrates, with particular pulmonary, gastrointestinal, neural and cardiovascular involvement. We presented a case of CSS complicated by weight loss, pulmonary embolism and non-traumatic rib fractures. The case confirms that the clinical course of the disease is highly heterogeneous.

Key words: Churg-Strauss syndrome, cough fracture, pulmonary embolism

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Introduction

Churg-Strauss syndrome (CSS) was described for the first time in 1951 by two pathologists, Jacob Churg and Lotte Strauss [1].

CSS is a necrotizing small vessel vasculitis characterized by the presence of asthma and sinusitis, hypereosinophilia and eosinophilic infiltrates, with particular pulmonary, gastrointestinal, neural and cardiovascular involvement [2]. Thromboembolism is considered a rare complication in the course of CSS [3].

In this paper, a case of CSS with symptoms such as weight loss, weakness, atraumatic fracture, and dorsalgia suggesting a malignant disease is described.

Case report

A female patient (RM), 53 years old, was admitted to the Department of Allergology, The Medical University of Gdansk, Poland in July 2005 because of progressive dyspnoea, chronic cough, chest pain and progressive weakness. She had been diagnosed with chronic obstructive pulmonary disease (COPD) a few years earlier. She also had a history of deep vein thrombosis in the left lower limb and hypertension. The first diagnosis was pulmonary embolism confirmed by lung scintigraphy (Figure1, 2). The diagnosis of COPD, however, was changed to asthma, since a bronchodilatation test with salbutamol was positive (Δ FEV1 over 16% predicted value, 280 ml). A peripheral blood count showed

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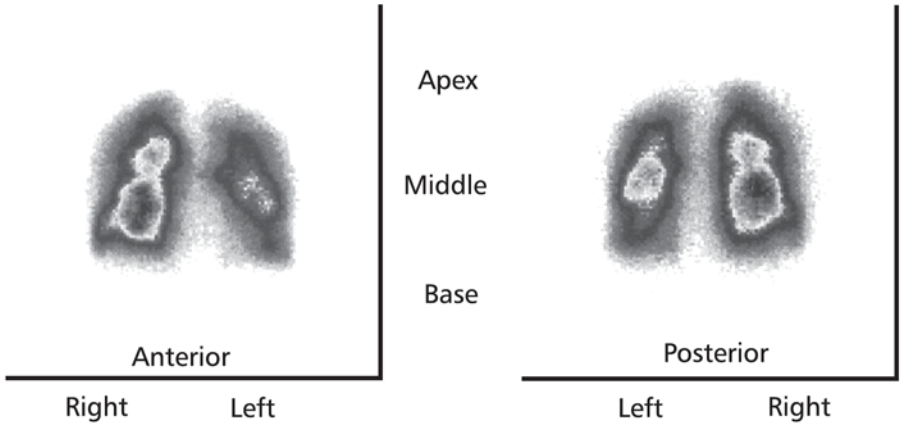

Figure 1. Lung scintigraphy (July 2005)

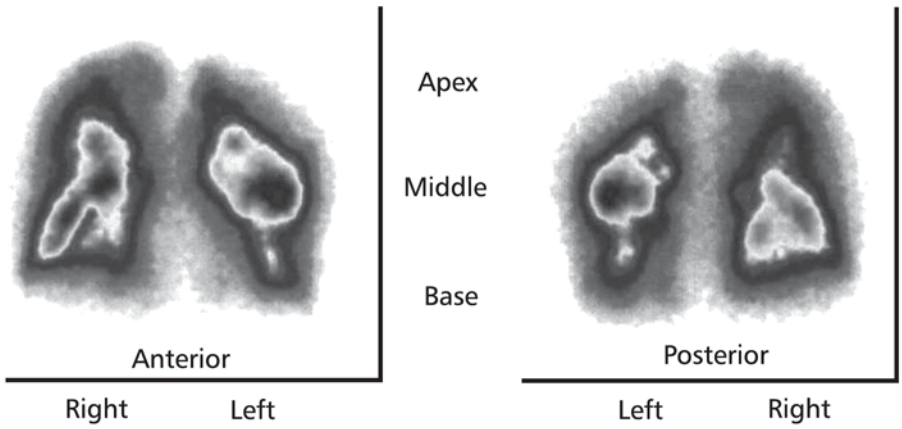

Figure 2. Lung scintigraphy (September 2005)

11% (1.3 G/l) of eosinophils. An allergic cause was ruled out by anamnesis and skin prick tests. Infestation by parasitic helminthes was ruled out by serologic studies and tests for parasites in the faeces. Eventually, the patient was successfully treated with the following: heparin, inhaled corticosteroids, long-acting β-agonists, and short-acting β-agonists.

Two months later, the patient was again referred to the clinic due to a troublesome cough, dyspnoea, recurrent weakness, fever, weight loss (10 kg), and dorsalgia. The chest X-ray revealed a sixth right rib fracture. Disseminated neoplastic disease was suspected.

Although the patient was treated with an anticoagulant the control lung scintigraphy revealed newly-developed lesions from pulmonary embolism. A peripheral blood count showed 43% (2.96 G/l) of eosinophils. High resolution chest tomography revealed ground glass opacifications (Figure 3).

Spirometry showed serious bronchial obstruction. An ENT consultation revealed nasal polyposis and paranasal sinusitis; while a neurological consultation showed mononeuritis multiplex. Despite a negative anti-neutrophil cytoplasmatic antibody (ANCA) test and lack of histological confirmation, the patient was diagnosed with CSS.

The patient received oral prednisone (a total daily dose of 1 mg/kg) and oral anticoagulation therapy was continued. A few weeks after the administration of prednisone, her clinical condition had improved markedly and the chest X-ray was normal.

Discussion

The triad of asthma, sinusitis, and eosinophilia is suggestive of CSS syndrome. Respiratory symptoms are the most commonly presented features of CSS. The site of the vasculitis process, however, is

Figure 3. High resolution computer tomography

also often outside the lungs, most commonly involving the peripheral nervous system, heart, skin, kidneys and gastrointestinal tract [4]. Thromboembolism is a rare complication of granulomatous vasculitis and of CSS in particular [5]. Thrombosis may develop at the first diagnosis and during the follow-up of CSS [3]. In the case presented, pulmonary embolism occurred at the first diagnosis of allergic vasculitis. In the pathogenesis of thromboembolism in CSS, not only does inflammation play a role, but eosinophils and eosinophil-derived proteins also significantly contribute to thrombotic activity. Thus, anticoagulants might be considered during disease exacerbation in addition to steroids [3].

In the case presented, the patient showed symptoms resembling disseminated neoplastic disease, including weight loss, weakness and non-traumatic fracture. The patient also complained of a chronic cough. The most frequent and best-documented complications of cough are rib fractures [6]. The typical locations for rib fractures are the fifth through ninth rib at the lateral part of the rib cage. These fractures are caused from opposing muscular forces in the middle of the rib at the axillary line from the serratus anterior and external oblique muscles. Other cough-induced rib fractures may be caused by a complex interplay between the inspiratory and expiratory muscles [7]. All the above-mentioned symptoms resolved after prednisone treatment, although our case confirmed that the clinical course of CSS is highly heterogeneous and may mimic other diseases.

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