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# Fatal course of Churg-Strauss disease

## Abstract

Churg-Strauss syndrome (CSS) is an allergic vasculitis, which is commonly characterized by asthma and allergic rhinitis, eosinophila and eosinophillic infiltration of organs. Approximately 60–90% of patients with CSS are long-term survivors. We presented the case of fatal case of the disease complicated by pulmonary embolism, arrhythmia, and eventually — cerebral bleeding.

**Key words:** Churg-Strauss syndrome, allergic vasculitis, asthma, pulmonary embolism

## Introduction

Asthma is a chronic inflammatory disease [1], which in the majority of patients may be sufficiently controlled. However, in some cases, in spite of proper treatment the disease progresses and the full control of symptoms is difficult to achieve. Several factors have been identified as triggers of the course of asthma, including aspirin intolerance, cigarette smoking, GERD, infections caused by *Chlamydia pneumoniae*. In some cases an uncontrolled asthma may be associated with disorders of different than asthma origin, like Churg-Strauss syndrome (CSS), which is an allergic vasculitis.

In this paper the case of fatal course of CSS, which presented mainly as asthma, is described.

## Case report

Male patient (W.D). 63-years-old, was admitted at the Allergology Department on 11.02.2005 due to hemoptysis in the course of asthma, which was diagnosed a few years before. His asthma was poorly controlled and it exacerbated frequently with the need for oral steroids. In the early nineties he experienced pulmonary insufficiency and required respiratory management after surgical treatment of abdominal hernia. In 2005 he had intracerebral haematoma. He had a history of lung tuberculosis treated with tuberculostatics and complicated by empyema.

At the time of admission patients was mildly short of breath, with mild hemoptysis; auscultation revealed sporadic wheezing.

Spirometry showed moderate bronchial obstruction with good reversibility ($\sigma$FEV1 over 12%; 206 ml). Chest X-ray was normal, whereas ground glass opacifications were present in high resolution computed tomography. Blood eosinophils were markedly increased (1500/mm), blood cell sedimentation was 35mm/h and CRP — 33 mg/l.

Thus the preliminary diagnosis of CSS was proposed. There were no signs or symptoms of other organs involvement — blood markers of kidney function and urine analysis were normal, ECG and cardiac enzymes (CK-MD, troponin 1) were normal, otolaryngological and neurological assessment were normal. D-dimmer was beneath 500 mg/L. Some laboratory abnormalities, however, were identified

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— LDH was increased — 365 U/L, APTT — 36 sec, fibrinogen — 4.92 g/l, protein electrophoresis showed slightly increased alfa-1 protein. ANA test was moderately increased (80 U/L), ANCA were absent. The final diagnosis of CSS disease was made and patient received oral prednisone (total daily dose — 1mg/kg). On 19.02.2005 sudden severe dyspnea and critical hypotension occurred. Patient was intubated and administered to Cardiology Department, where pulmonary embolism was diagnosed and he received fibrinolytic treatment, dopamine and dobutamine, oral prednisone was changed to intravenous solu-medrol and cyclophosphamide. During following three days recurrent supraventricular extrasystolia, and eventually atrial fibrillation were observed. At that time serum level of troponine 1 was 0.66, CK-MB — 5.8, CRP — 183, D-dimmer — 785. Intravenous amiodarone and cardioversy failed to control heart rhythm. Increasing respiratory and cardiac failure required invasive ventilation and heart stimulation and patient was referred to the Intensive Care Unit (24.02.2005). During the following days his status has been improving, his cardiovascular and respiratory system become efficient two days later and he was extubated on 01.03.2005. Patient continued anticoagulative treatment,and solumedrol.

On 03.03.2005 patient deteriorated rapidly, he required re-intubation, head CT scans showed massive hematoma (9 × 6 cm) in the right hemisphere, with blood present in the brain chambers' system, left lateral herniation under the falx and massive brain oedema. Consultant of neurology affirmed the "death of the brain stem". Patient died on 06.03.2005 with signs and symptoms of DIC and multi-organ insufficiency.

## Discussion

Churg-Strauss syndrome is a multi-organ disorder. Characteristic triad of symptoms includes asthma, eosinophilia, and organs involvement due to eosinophils infiltrations of vessels.

Usually small and medium-sized arteries are involved. Paranasal sinusitis, pulmonary and renal infiltrates, mononeuritis multiplex or polyneuropathy, allergic rhinitis, gastroenteritis and skin lesions are common presentations of the disease.

In the proportion of patients with vasculitis antineutrophil cytoplasmic antibodies (ANCA) against myeloperoxidase (MPO) and against proteinase 3 (PR3) may be identified. The former are usually associated with CSS [2]. Goncalves et al. postulated even that antineutrophil cytoplasmic antibody (ANCA) assessment in chronic rhinosinusitis might allow for early identifying vascilitis [3]. In the presented case ANCA test was negative, however moderate increase in the level of antinuclear antibodies (ANA) was found. Although ANA test is typically positive in rheumatoid diseases rather than in CSS, some patients may have weakly increased levels of ANA and/or rheumatoid factor (RF) [4].

The American College of Rheumatology (ACR) has proposed 6 criteria for the diagnosis of CSS:

— asthma (wheezing, expiratory rhonchi);
— eosinophilia of more than 10% in peripheral blood;
— paranasal sinusitis;
— pulmonary infiltrates (may be transient);
— histological proof of vasculitis with extravascular eosinophils;
— mononeuritis multiplex or polyneuropathy.

The presence of four or more criteria yields a sensitivity of 85% and a specificity of 99.7%. In the presented case, at the time of admission to the Department of Allergology, three criteria were actually fulfilled — asthma, eosinophilia and pulmonary involvement. However, hemoptysis and CT high resolution findings indicated pulmonary angitis, the condition which required systemic treatment [5]. Hemoptysis, which occurred in patient were most probably related to pulmonary alveolar hemorrhage resulting from alveolar capillaritis.

Cardiac manifestation of the disease (28%) is not rare and symptoms are usually related to heart failure, infarction, myocarditis, pericarditis, constrictive pericarditis [6]. Thromboembolic disease is much more rare occurrence. Recently Gracia et al reported three cases of severe pulmonary embolism [7]. Interestingly in all these patients, similarly to ours, ANCA were negative. They all presented with deep vein thrombosis revealed by Doppler ultrasonography. In our patients such an assessment was not done, because of his critical condition, when massive pulmonary embolism had appeared.

Current therapeutic approach in CSS includes prednisone, usually combined with cyclophosphamide, metothrexate or azathioprine. Although with this regimen response may be frequently reached it is toxic and does not prevent relapse [8]. Long-term treatment may be the risk for thrombo-embolic disease, but it was not the case in our patient, who received steroid for a few day only before embolic incident. Vessels inflammation, particularly associated with eosinophilic infiltrates may increase the frequency of local thrombotic complications, which in turn may result in pulmonary em-

bolism. Incident of pulmonary embolism prior to steroidotherapy was previously described in 13-year-old boy with CSS [9].

Long term survival in the entire group of patients with CSS is good, approximately 60–90%.

Even patients with severe active disease may experience remission. In our case, however massive pulmonary embolism was complicated with arrhythmia, and further with severe cerebral bleeding.

## References


1. www.ginasthma.com.
2. Kallenberg CG. Pathogenesis of PR3-ANCA associated vasculitis. J Autoimmun 2008; 30: 29–36.
3. Goncalves C, Pinaffi JV, Carvallino JF et al. Antineutrophil cytoplasmic antibodies in chronic rhinosinusitis may be a marker of undisclosed vasculitis. Am J Rhinol 2007; 21: 691–694.
4. Savige J, Gillis D, Benson E et al. Inernational Consensus Staement on Testing and Reporting of Antineutrophil Cytoplasmic Antibodies (ANCA). Am J Clin Pathol 1999; 111: 507–513.
5. Alberts WM. Pulmonary manifestations of the Churg-Strauss syndrome and related idiopatic small vessel vasculitis syndromes. Curr Opin Pulm Med 2007; 13: 445––450.
6. Agard C, Rendu E, Leguern V et al. Churg-Strauss syndrome revealed by granulomatous acute pericarditis: two cases reports and a review of the literature. Semin Arthritis Rheum 2007; 36: 386–391.
7. Garcia G, Achouh L, Cobarzan D et al. Severe venous thromboembolic disease in Churg-Strauss syndrome. Allergy 2005; 60: 409–410.
8. Langford CA. Small-vessel vasculitis: therapeutic management. Curr Rheumatol Rep 2007; 9: 328–335.
9. Ikemoto Y, Kohdera U, Uraoka M et al. Pulmonary infarction and deep venous thrombosis in a 13-year-old boy with Churg-Strauss syndrome. Ped Intern 2001; 43: 441––443.