

Churg-Strauss syndrome in a case of eosinophilia

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ABSTRACT

A number of allergic, infectious and idiopathic diseases are associated with an increased number of eosinophils in blood. We report the case of a woman who was referred to our outpatient clinic due to asthma that had first developed three months previously and papular rash. Laboratory investigations revealed eosinophilia (23 %; 2,162 cells/ml).

An allergic cause was ruled out by anamnesis, skin prick-test and specific IgE determination. Another frequent cause of eosinophilia is infestation by parasitic helminths, but serologic studies and studies of parasites in feces were negative. Chest radiography and computed tomography scan revealed diffuse infiltrates. The diagnosis was confirmed by transbronchial and skin lesion biopsies. The patient was finally diagnosed with Churg-Strauss syndrome and was treated with oral corticosteroids.

A delay in the diagnosis and treatment of this syndrome increases the risk of death from the complications of vasculitis. Because of the importance of an early diagnosis in this disease, its presence should be suspected in cases of eosinophilia after ruling out more frequent causes.

Key words: Churg-Strauss syndrome. Eosinophilia. Asthma. Sinusitis. Vasculitis.

RESUMEN

Diversas enfermedades alérgicas, infecciosas e idiopáticas se asocian con un aumento del número de eosinófilos en sangre. Presentamos el caso de una mujer remitida a nuestra consulta por asma de tres meses de evolución y rash papular que en la analítica realizada presentaba eosinofilia del 23 % (2.162 eosinófilos).

La historia clínica, prick-test y determinación de IgE específica descartaron una causa alérgica. Otra causa frecuente de eosinofilia son las infecciones por parásitos helmintos pero el estudio de parásitos en heces y los estudios serológicos fueron negativos. Los estudios radiológicos y scanner de tórax reveló infiltrados difusos. El diagnóstico fue confirmado por biopsia transbronquial y de las lesiones cutáneas. Nuestro caso fue diagnosticado de síndrome de Churg-Strauss, y la paciente recibió tratamiento con corticoides orales.

Un retraso en el diagnóstico y tratamiento aumenta el riesgo de muerte por complicaciones de la vasculitis. Debido a la importancia de un diagnóstico precoz en el diagnóstico de esta enfermedad, debe sospecharse su presencia en casos de eosinofilia, tras descartar el resto de causas más habituales.

Palabras clave: Síndrome de Churg-Strauss. Eosinofilia. Asma. Sinusitis. Vasculitis.

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INTRODUCTION

Eosinophilia is a frequent cause of consultation in our speciality. A number of allergic, infectious, neoplastic and idiopathic diseases are associated to eosinophilia. We report the case of a woman who

was referred because non productive cough, dyspnea, and rash papular for several weeks. Eosinophilia had been detected in the analyses performed (23 %; 2.162 cells/ml).

The Churg-Strauss syndrome is a rare disease, occurring in an estimated two to four patients per million patient-year^{1,2}. This syndrome has been divided into 3 distinct phases, which may or may not be sequential. The prodromal phase is characterized by asthma with or without allergic rhinitis. The second phase is marked by a peripheral blood eosinophilia and eosinophilic tissue infiltration producing a picture similar to Loeffler syndrome, chronic eosinophilic pneumonia, or eosinophilic gastroenteritis. The third, vasculitic phase may involve any organ: heart, lung, central nervous system, kidney, lymph nodes, muscle, and skin. Skin involvement occurs in more than two-thirds of patients³.

CASE REPORT

A 47-year-old woman presented dyspnea and non productive cough for tree months. Her history revealed that she had perennial rhinitis, poliposis and sinusitis. Two weeks later, she developed a pruritic rash papular involving her back, chest and lower extremities. She had no history of other lung diseases, fever tobacco use, allergies or a change in her home. The patient's symptoms improved during treatment with bronchodilatadors and inhaled corticosteroids, but the dyspnea recurred when the dose was stopped after four weeks. The patient recalled that the had similar skin lesion on her legs two years earlier, but they had resolved when she was treated with prednisone.

The triad of asthma, sinus disease, and eosinophilia is suggestive of the Churg-Strauss syndrome, but other possible diagnoses include allergic bronchopulmonary aspergillosis, cancer, helminthic infection, eosinophilic pneumonia and hypersensitivity pneumonitis.

METHODS AND RESULTS

Skin prick-test and IgE especific with a battery of common inhalant allergens and foods were negative. Another causes of eosinophilia is an infestation by parasitic helminths, but studies of parasites in faeces and serologic test for antibodies against *Strongyloides*, *Echinococcus*, *Trichinella*, etc. were negative.

Laboratory tests showed a hemoglobin level of 14 g/dL, a hematocrit of 42 %, leukocytes of

9400/mm³, eosinophils of 2162/mm³ (23 %), platelets of 228,000/mm³, an erythrocyte sedimentation rate of 85 mm/h, and immunoglobulin E (IgE) elevated to 338 U/mL Test for antineutrophil cytoplasmic antibodies (ANCA) and antinuclear antibody were negative. Urinalysis, complement, and antinuclear antibody tests were normal or negative. Physical examination demonstrated several erythematous plaques, with ill-defined borders on the back, chest and lower extremities.

The results of pulmonary-function test were consistent with the presence of severe, partially reversible airflow obstruction. X-rays of the paranasal sinuses revealed opacification and chest roentgenogram showed a micronodular infiltrate in the right apex and inferior lobe. A thoraco CT scan revealed several small lymph nodes, and diffuse infiltrates.

The evaluation was negative for allergic bronchopulmonary aspergillosis: skin and serologic test for aspergillus-specific antibodies were negative and chest CT did not show central bronchiectasis.

A review of outpatient records revealed that the proportion of eosinophils on a complete blood count had been 10 percent four years earlier (1850/mm³) and 20 percent (2027/mm³) on a more recent count. Although eosinophilia may accompany asthma, the levels rarely exceed 800 per cubic millimeter even in severe cases. In addition, the patient's erythrocyte sedimentation rate is markedly elevated, a finding not seen in asthma.

Because the diagnosis remained uncertain, a transbronchial biopsy and biopsy of skin lesion were performed. The transbronchial biopsy revealed diffuse eosinophilic infiltrates and a biopsy of the skin demonstrated an infiltration in the dermis with a predominance of eosinophils infiltrating the vascular walls. There were no granulomas.

Treatment with 60 mg of prednisone per day was begun, and the dose was tapered over a period of several months to 5 mg per day. She showed a good response to oral corticosteroids, with total remission of symptoms and normalization of the eosinophil count, IgE levels and pulmonary-function test.

DISCUSSION

The Churg-Strauss syndrome is the major vasculitis associated with eosinophilia. In 1990, the American College of Rheumatology developed criteria for epidemiologic and therapeutic studies. These criteria are asthma, eosinophilia greater than 10 % on a differential white blood cell count, mono- or polynuropathy, nonfixed pulmonary infiltrates, paranasal sinus abnormalities, and biopsy containing a blood ves-

sel with extravascular granulomas. The presence of 4 of the 6 criteria yielded a sensitivity of 85 % and a specificity of 99.7 %⁴.

This syndrome is distinguished from other systemic vasculitis by the presence of asthma. Involvement of the peripheral nerves, gastrointestinal tract and skin is common. Perinuclear ANCA, present in most patients, should be considered as a major diagnostic criterion but have not proven useful for follow-up purposes⁵. The etiology of Churg-Strauss syndrome remains unclear; however, the presence of asthma, eosinophilia, and increased immunoglobulin E (IgE) suggest an allergic process.

We present a 47-year-old woman, with difficult-to-control asthma, rhinosinusitis and rash papular, which were the prodromal phase of Churg-Strauss syndrome. The prominent eosinophilic infiltrates of the skin and lung confirmed the presence of a systemic eosinophilic syndrome.

A rapid improvement of clinical manifestations, analytical parameters, and radiological images coincided with the administration of systemic corticosteroids. In most patients with this syndrome, the eosinophilia and symptoms respond quickly to corticosteroid therapy, but residual asthma may require continuation of low-dose prednisone, and some may benefit from interferon alpha^{6,7}. This syndrome is diagnosed in some patients who are receiving leukotriene antagonist but whether this drug causes the syndrome remains uncertain^{8,9}.

Because a delay in the diagnosis and treatment of the syndrome increases the risk of death from vasculitis complications involving the heart (cardiac involvement is the most common cause of death due to pericarditis or ischemia in the vasculitic phase),

gastrointestinal tract, or other organs¹⁰, their presence must be considered when a patient's symptoms and signs are atypical of asthma alone. Therefore, we think that the allergologist should be aware of Churg-Strauss syndrome as a cause of eosinophilia.

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