

## LETTER TO THE EDITOR

Iran J Allergy Asthma Immunol  
June 2015; 14(3):338-340.

# Familial Churg-Strauss Syndrome in a Sister and Brother

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*Received: 1 June 2014; Accepted: 25 June 2014*

## ABSTRACT

Churg-Strauss syndrome (CSS) is a granulomatous small vessel vasculitis. It is characterized by asthma, allergic granulomatosis and vasculitis. This syndrome is rare in children. A 5 years old boy was admitted with cough, fever and dyspnea for 2 weeks. On the basis of laboratory data (peripheral eosinophilia), associated with skin biopsy, and history of CSS in his sister, this disease was eventually diagnosed. The patient had good response to corticosteroid.

In every asthmatic patient with prolonged fever, eosinophilia and multisystemic involvement, CSS should be considered.

**Keywords:** Children; Churg-Strauss syndrome; Vasculitis

There are a number of comparatively rare vasculitis disorders that affecting children like Churg-Strauss syndrome (CSS) and Kawasaki disease.<sup>1</sup>

CSS is an uncommon vasculitis in children and its etiology is unknown. CSS overlaps clinically and pathologically with several diseases. To the knowledge of authors, the occurrence of CSS in the members of the same family is rare. In this report, we describe the occurrence of CSS in sister of a boy who had this disease one year ago.<sup>2</sup>

A 5 year old boy with a history of respiratory discomfort, fever and wheeze was referred to Shiraz Imam Reza Allergy Clinic. The patient has family history, his 8 year old sister with asthma, prolonged fever and skin lesion was admitted last year

and skin biopsy revealed eosinophilic fibrinoid vasculitis, compatible with CSS.

On admission time, the patient had fever and tachypnea. In chest physical examination, intercostal muscle retraction and bilateral wheeze and ronchi were present. Submandibular lymphadenopathy 2×1.5 centimeters was palpable. Chest x-Ray showed triangular density in lower zone of right lung and thickness of right paratracheal in favor of pneumonic infiltration. A CBC showed hemoglobin 12 g/dl. White blood cell count was  $13 \times 10^9/L$  with 15% neutrophil, 51% lymphocyte and 34% eosinophils. The IgE was 644 Iu/L (normal below 88 Iu/L). ESR was 22 mm/hr. Gastric washing for acid fast bacilli stain was negative for 3 times. Stool test for ova, parasite and blood was negative. Anti neutrophil cytoplasmic antibodies (perinuclear and cytoplasmic) (p-ANCA, c-ANCA) were negative. Complement 3 and 4 were in normal range. Abdominal sonography was normal.

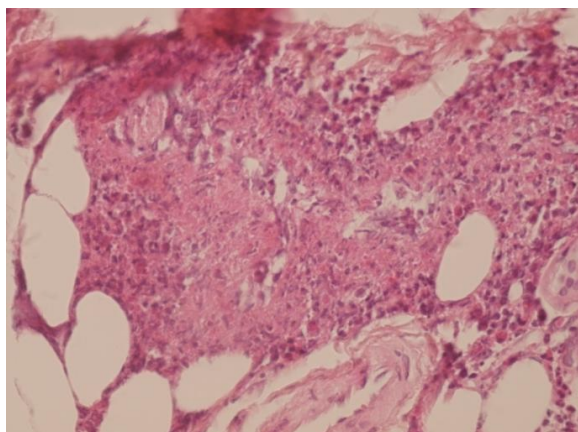
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## Familial Churg-Strauss Syndrome

Echocardiography disclosed patent foramen ovale and mild pulmonary hypertension. The patient had a few maculopapular eruptions on his trunk, so skin biopsy was done that showed heavy infiltration of eosinophils around skin vessel wall (Figure 1).

Antibiotic, prednisolon, fluticasone and ventolin were prescribed and nifedipine was added for his heart problem.

Glucocorticoid ointment was used for his skin.



**Figure 1. Heavy infiltration of eosinophils around the skin vessel wall.**

Although his condition became better, but respiratory discomfort and wheeze continued.

Prednisolon was continued for him, with relative improvement in lung problem, although wheeze persisted in follow up visits. The last CBC showed 10% eosinophilia. The patients did not continue the follow up after few months.

CSS is an uncommon disease, . CSS was described with severe asthma associated with fever, eosinophilia and vasculitis affecting various organ systems. While the clinical features of adult CSS have been well illustrated, the feature of childhood CSS are less clear.

Pulmonary disease is a central features of CSS and present with transitory patchy pulmonary infiltrates, nodular infiltrates or pleural effusion. Asthma develops in all patients and is preceded or sometimes develops concomitantly with the onset of systemic vasculitis.<sup>3</sup>

Patients with CSS usually respond well to corticosteroid therapy. Several trials including cyclophosphamide, plasma exchange and azathioprine are useful in controlling disease and relapse.

Familial clustering of systemic vasculitides has been described, usually with the simultaneous or later occurrence in the same family of a single type of

vasculitis, such as Giant Cell Arthritis.<sup>4</sup> An unusual familial clustering of 2 different systemic vasculitides, CSS and Wegner granulomatosis in a father and son has been reported too.<sup>5</sup> To our knowledge, CSS in members of the same family has not been reported, previously.

The familial association of systemic vasculitides suggests that genetic factors may confer susceptibility to these diseases, but the HLA typing studies did not give conclusive results.<sup>6</sup> Triggering factors, such as vaccination, desensitization, or exposure to leukotriene-receptor antagonists, have been suspected as contributing to the development of CSS, but its etiology has not yet been fully elucidated.<sup>7</sup>

In CSS, many patients have a history of atopy, and different putative triggering factors have been identified. Because our patients had upper and lower airway disease and both lived in a village, we can also hypothesize a role for environmental factors. The hypothesis that environmental agents may trigger the disease in genetically predisposed subjects implies variations in genes encoding for proteins crucial for the immune response regulation.<sup>8</sup>

Considering the interaction of both genetic predisposition and environmental influences, such familial clustering between siblings of the same vasculitis, seems to underline a predominant pathogenic role of the genetic factors. Further studie are needed to understand the etiology and pathogenesis of the systemic vasculitides, like CSS.

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