

## Churg–Strauss syndrome. Report of a case.

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### Introduction

Churg–Strauss syndrome (CSS) is a form of small and medium vessel vasculitis characterized by eosinophilic infiltration of organs with necrotizing vasculitis and interstitial and perivascular granulomas. Three phases have been described in the natural history of the disease (prodromal, eosinophilic, and vasculitic phases) although they do not always occur successively. Initial records show CSS is a condition that is highly responsive to steroids. We report a CSS case that occurred in dysesthesia.

### Case report

The patient was a 65-year-old woman. On December 23, 2011, dysesthesia of the finger-tip of the right hand developed. A feeling appeared in both legs in a few minutes. On December 31, our hospital was consulted because she could not move either hand. She developed asthma when 60 years old. She was hospitalized with eosinophilic pneumonia at the age of 63. There was no abnormality neurologically in lucidity, or in the cranial nerve domain. The muscular strength decreased in the middle class in distal muscle predominance with four extremities. The right and left grip was 5.7 kg and 11.7 kg, respectively. The tendon reflex decreased at the four extremities. The sense of touch of both hands decreased. There was dysesthesia of the

tip of both legs. The urine was normal. The WBC increased to 15300 in peripheral blood. Acidophiles increased with 67.0%. The rheumatoid factor was positive. The IgE increased to 2,370 IU/ml (normal, <173). MPO-ANCA increased to 43 EU (normal, <20). The antinuclear antibody was negative. The cell count was one-third and the cerebral fluid was colorless. Protein 27.2 mg/dl (normal, 10-40), sugar 58 mg/dl (normal, 50-75). As for the examination for nerve conduction, axonopathy was found in multiple nerves.

### Discussion

Churg–Strauss syndrome was first described in 1951 by Churg and Strauss [1]. It is a rare form of systemic vasculitis (2.5 cases/100 000 adults/year) occurring exclusively in people with asthma, and is associated with blood and tissue eosinophilia. The most commonly involved organ is the lung, followed by the skin. However, CSS can affect any organ system of the body. Asthma is the main feature of CSS and precedes the vasculitic phase [2]. It presents as a chronic severe form and requires frequent or long-term courses of systemic steroids. Upper airway abnormality in the form of allergic rhinitis, recurrent sinusitis, and nasal polyposis is fairly common [3]. Involvement of the skin is a frequent feature of the vasculitic phase and presents as tender subcutaneous nodules,

palpable purpura and hemorrhagic lesions [4]. Cardiac and neurological involvement is often seen; cardiac complications in the form of infarction and arrhythmias are responsible for 50% of deaths [5]. Early diagnosis and treatment prevent organ damage and mortality. However, confirming the diagnosis is difficult as individual manifestations occur in isolation and lung parenchymal involvement is not universal. Moreover, although classified as vasculitis, ANCA positivity is seen in only 40%-60% of patients. To add to the problem there are no laboratory tests specific for CSS. The laboratory abnormalities are nonspecific and include eosinophilia, high IgE, raised acute phase reactants, and hypergammaglobulinemia. Therefore, diagnostic criteria specified by ACR are most commonly used for diagnosis. A minimum of four criteria are required for a confident diagnosis of CSS. Our patient satisfied four.

### References

1. Churg J, Strauss L. Allergic granulomatosis, allergic angiitis, and periarteritis nodosa. *Am J Pathol.* 1951;27:277–301.
2. Cottin V, Khouatra C, Dubost R, Glérant JC, Cordier JF. Persistent airflow obstruction in asthma of patients with Churg–Strauss syndrome and long-term follow-up. *Allergy.* 2009;64:589–95.
3. Bacciu A, Buzio C, Giordano D, Pasanisi E, Vincenti V, Mercante G, et al. Nasal polyposis in Churg–Strauss syndrome. *Laryngoscope.* 2008;118:325–9.
4. Kawakami T, Soma Y, Kawasaki K, Kawase A, Mizoguchi M. Initial cutaneous manifestations consistent with mononeuropathy multiplex in Churg–Strauss syndrome. *Arch Dermatol.* 2005;141:873–8.
5. Neumann T, Manger B, Schmid M, Kroegel C, Hansch A, Kaiser WA, et al. Cardiac involvement in Churg–Strauss syndrome: impact of endomyocarditis. *Medicine (Baltimore)* 2009;88:236–43.

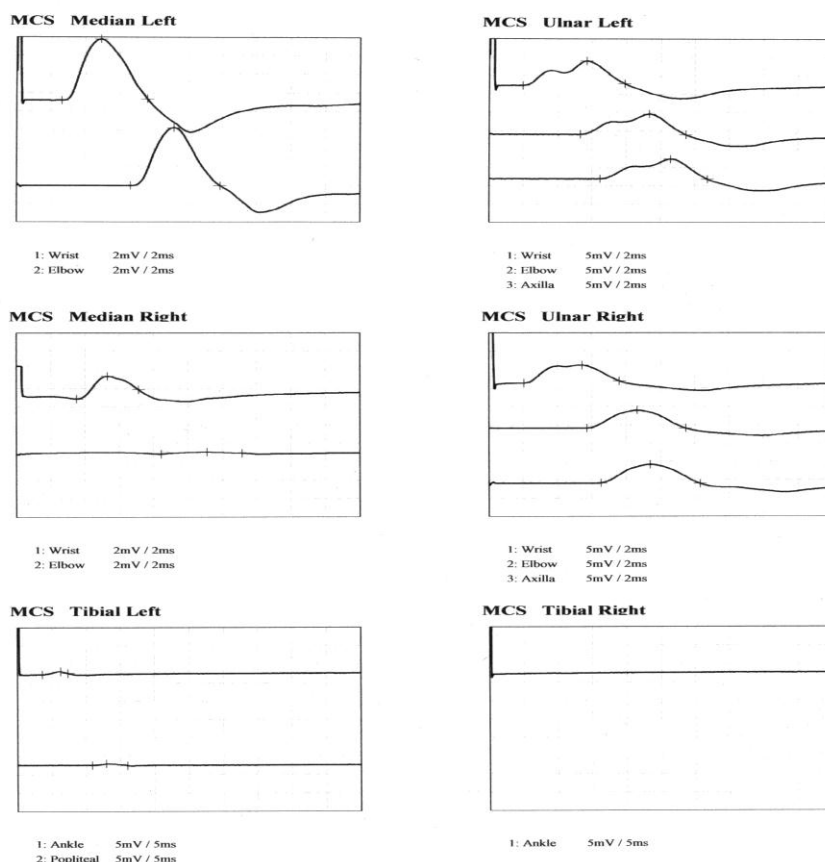


Figure 1. Nerve conduction study