

Churg–Strauss Syndrome with Concomitant Occurrence of Ischemic Stroke and Relapsing Purpura

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A 77-year-old woman suffering from chronic bronchial asthma and chronic atrial fibrillation who had had a previous ischemic stroke presented to our emergency unit with gait disturbance. She had new-onset truncal ataxia, right hemiparesis, and right sensory disturbance related to the previous stroke. Her lower legs were slightly swollen and had a reddened appearance. Her medical history included mitral valve replacement because of severe mitral valve regurgitation. Her white blood cell count was 8600/ μ L, mainly consisting of eosinophils (4480/ μ L; 52.1%). Serum nonspecific immunoglobulin E was elevated to 1600 IU/mL (normal range <170 IU/mL). She was taking warfarin for secondary stroke prevention, and on admission her prothrombin time international normalized ratio was 3.06. Diffusion-weighted magnetic resonance imaging revealed a fresh infarct in the right cerebellum. No stenosis or occlusion was shown in the cervicocephalic arteries on magnetic resonance angiography or carotid ultrasound. No emboligenic diseases, except for atrial fibrillation, were identified. On day 3, an extensive itchy, purpuric rash appeared on her lower limbs. The rash remitted and recurred spontaneously for several weeks. A skin biopsy specimen of the purpuric lesions revealed massive eosinophilic infiltration of the dermis and eosinophilic vasculitis involving small vessels. We diagnosed the patient with Churg–Strauss syndrome (CSS). Skin lesions and eosinophilia disappeared after oral corticosteroid therapy. In this case, cerebellar infarction occurred with purpuric rash despite well-controlled anticoagulation. Patients with CSS may suffer from ischemic stroke when the condition of CSS deteriorates. **Key Words:** Churg–Strauss syndrome—ischemic stroke—purpura.

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Case Report

A 77-year-old woman suffering from chronic bronchial asthma and chronic atrial fibrillation, with previous cardioembolic ischemic stroke, presented to our emergency unit with gait disturbance. She had new-onset truncal ataxia in addition to right hemiparesis and right sensory disturbance because of the previous stroke. Her lower legs were slightly swollen and had a reddened appearance. Her medical history included mitral valve replacement because of severe mitral valve regurgitation.

Her white blood cell count was 8600/ μ L, mainly consisting of eosinophils (4480/ μ L; 52.1%). Serum nonspecific immunoglobulin E was elevated to 1600 IU/mL (normal

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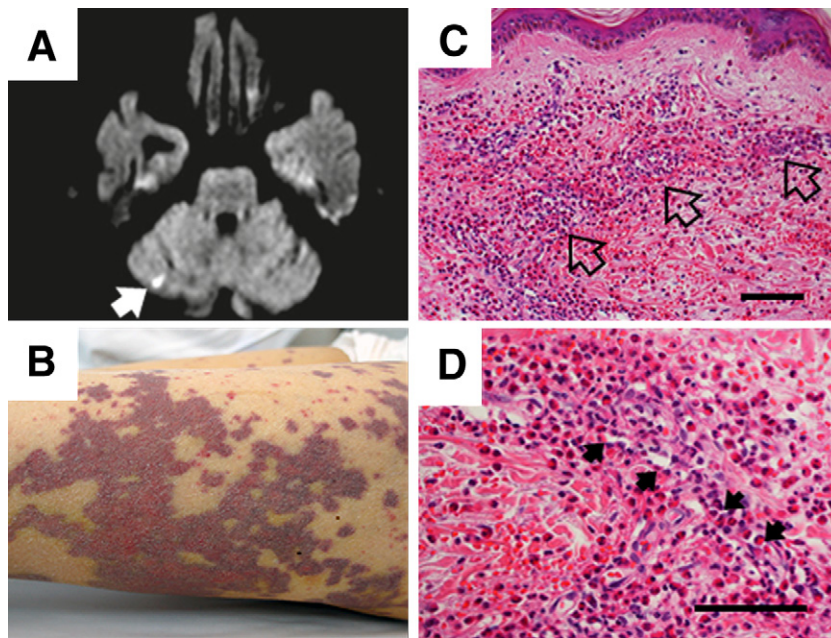


Figure 1. (A) Diffusion-weighted magnetic resonance image revealing a hyperintense region in the right hemisphere of the cerebellum (arrow). (B) Macroscopic photograph of the purpura on the left lower leg. (C) A skin biopsy specimen revealed extensive eosinophilic infiltration and hemorrhage (arrow) in the dermis. (Hematoxylin and eosin stain; scale bar = 100 μ m.) (D) Eosinophil-dominant necrotizing vasculitis (circle) of small vessels in the dermis with massive extravasation of eosinophils and red blood cells. (Hematoxylin and eosin stain; scale bar = 100 μ m.)

range <170 IU/mL). Perinuclear antineutrophil cytoplasmic antibody was negative. She was already taking warfarin for secondary stroke prevention and her prothrombin time international normalized ratio was 3.06 on admission, with a D-dimer of 1.8 μ g/mL (normal range <1.0 μ g/mL). Diffusion-weighted magnetic resonance imaging (MRI) revealed a fresh infarct in the right cerebellum (Fig 1A). Cerebral microbleeds were not detected on T₂-weighted MRI. No stenosis or occlusion was shown in the cervicocephalic arteries on magnetic resonance angiography and carotid ultrasound. No emboligenic diseases, except for atrial fibrillation, were identified.

On day 3, an extensive itchy, purpuric rash appeared on her lower limbs (Fig 1B). The rash remitted and recurred spontaneously for several weeks. Skin biopsy specimens of the purpuric lesions revealed massive eosinophilic infiltration of the dermis and eosinophilic vasculitis involving small vessels (Figs 1C and 1D). We diagnosed the patient with Churg–Strauss syndrome (CSS). Skin lesions and eosinophilia disappeared after oral corticosteroid therapy.

Discussion

CSS is a rare, small-sized vessel systemic necrotizing vasculitis that occurs in patients having asthma or an allergic history.¹ Ischemic stroke is uncommon in patients with CSS.^{2–7} Concomitant extravascular eosinophils in the dermis being compatible with CSS classification⁸ has not been pathologically proven in reported cases. In this case, cerebellar infarction occurred with purpuric rash despite well-controlled anticoagulation. Her stroke may have been caused by small-vessel arteritis of the central nervous system⁹ or hypercoagulable state because of CSS.¹⁰ Patients with CSS may suffer from ischemic stroke when the condition of CSS deteriorates. In patients with concomitant occurrence of ischemic stroke, eosinophilia,

and extensive purpura, CSS should be considered in the differential diagnosis.

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