Case Report

Ischemic and Hemorrhagic Strokes due to Eosinophilic Granulomatosis with Polyangiitis

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Background: We report a case of ischemic and hemorrhagic strokes occurring almost simultaneously in a patient diagnosed with eosinophilic granulomatosis with polyangiitis (EGPA) previously known as Churg–Strauss vasculitis. To our knowledge, this is the first known case. *Methods:* A 59-year-old man presented with a 2-month history of bilateral leg weakness, difficulty ambulating, and dyesthesias. While in the hospital, he developed acute right hand weakness, and magnetic resonance imaging of the brain revealed multiple, bilateral ischemic infarcts. After a few days, he acutely became unresponsive and was found to have a large left frontal hematoma and underwent emergent hematoma evacuation. His weakness was unexplained by the infarcts based on location and so a peripheral process was suspected. *Results:* Nerve conduction studies showed severe axonal sensorimotor neuropathy. A sural nerve biopsy showed necrotizing vasculitis, consistent with EGPA. *Conclusions:* EGPA is the rarest of the antineutrophilic cytoplasmic antibody vasculitides. The peripheral nervous system is frequently involved, but the central nervous system can also be affected. The vasculitis damages the vessel walls, which leads either to stenosis or to dilatation, resulting in ischemic or bleeding consequences which can occur simultaneously, such as in this case. Caution should be exercised when prescribing antiplatelet therapy to such patients. **Key Words:** Stroke—vasculitis—hemorrhagic—ischemic.

Eosinophilic granulomatosis with polyangiitis (EGPA), formerly known as Churg–Strauss syndrome, is a rare systemic disease characterized by necrotizing eosinophilic vasculitis of medium- to small-sized blood vessels and extravascular eosinophilic granulomas. EGPA is the most infrequent of the antineutrophilic cytoplasmic antibody (ANCA) vasculitides. The peripheral nervous system is frequently involved, but the central nervous system is rarely affected in EGPA.

A 59-year-old man presented with a 2-month history of bilateral leg weakness, difficulty ambulating, and dyesthesias. His medical history was significant for asthma, hypertension, and diabetes. While in the hospital, he developed acute right hand weakness, and magnetic resonance imaging of the brain revealed multiple, bilateral ischemic infarcts in the right lateral thalamus, left corona

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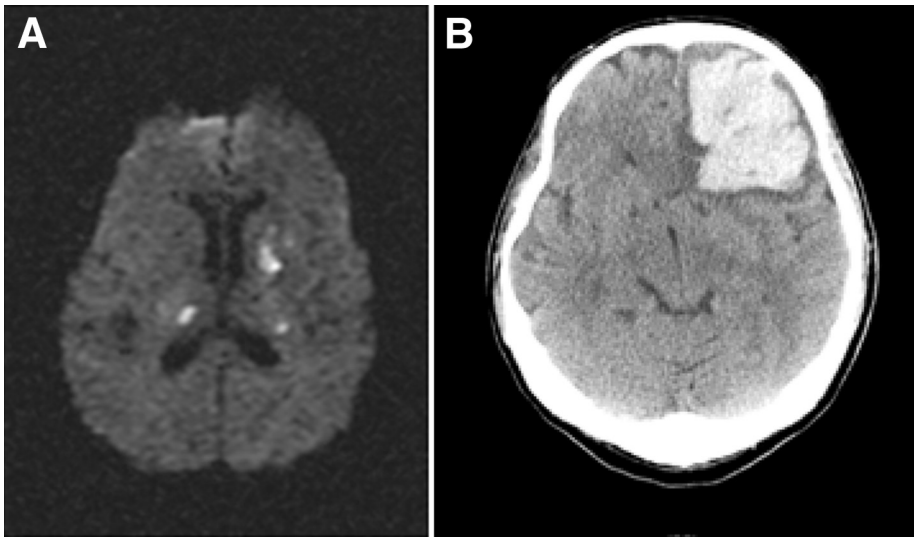

Figure 1. *(A) Diffusion-weighted imaging sequence showing multiple acute ischemic infarcts in the right lateral thalamus, left corona radiata, and external capsule. (B) Computed tomography scan showing left frontal hematoma found after the patient became unresponsive.*

radiata, and external capsule (Fig 1). He was transferred to our tertiary care center after he acutely became unresponsive and was found to have a large left frontal hematoma and he underwent emergent hematoma evacuation. On examination, he exhibited transcortical motor aphasia, akinesia, and severe right arm weakness distal more than proximal, left arm moderate weakness (distal more than proximal), and bilateral severe leg weakness, right more than left, with lower extremity hyporeflexia. Blood work revealed normal eosinophil count, blood urea nitrogen, and creatinine and an elevated erythrocyte sedimentation rate of 94 mm/hour. C-reactive protein was elevated at 8.6 mg/dL. His serum immunoglobulin (Ig) E levels were elevated at 4300 IU/mL (normal, 0-100 IU/mL). Clinically, we were doubtful that the scattered infarcts could account for the leg weakness, especially because his weakness was accompanied by hyporeflexia. This raised the question of a peripheral process and his nerve conduction studies showed severe axonal sensorimotor neuropathy. An enzyme-linked immunosorbent assay specific for proteinase 3 was negative but was positive against antigens for myeloperoxidase (MPO). A sural nerve biopsy showed necrotizing vasculitis with eosinophilic infiltrates, consistent with EGPA (Fig 2). He was started on prednisone 60 mg per day orally and discharged to an acute rehabilitation facility after which he was lost to follow-up.

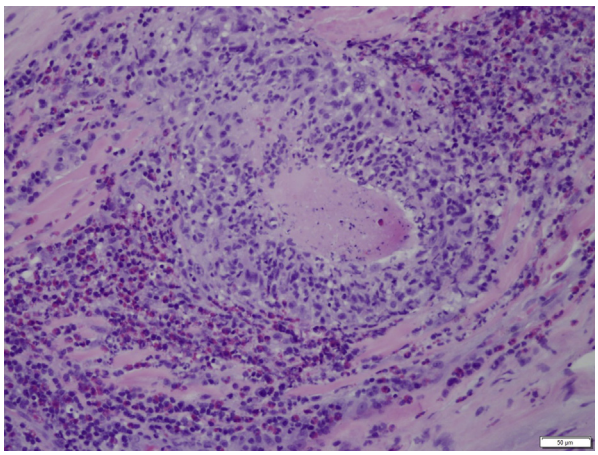

Figure 2. *Hematoxylin and eosin section of sural nerve biopsy shows vasculitis of a large arteriole with transmural acute and chronic inflammation, focal mural fibrinoid necrosis, and thrombosis. The inflammatory infiltrate at the periphery of the vessel wall and adjacent epineurial tissue contains abundant eosinophils.*

Discussion

EGPA is a multisystem disorder characterized by allergic rhinitis, asthma, and prominent eosinophilia. Its prevalence in the general population is low, reportedly between 2.4 and 6.8/1 million people, with a higher predilection in asthmatics and people between the age of 40 and 60 years.[1-3] It is one of the antineutrophilic cytoplasmic antibody (ANCA)-associated vasculitides which can affect the central nervous system in a variety of ways.[4,5] To our knowledge, this is the first report where ischemic and hemorrhagic strokes occurred almost simultaneously. The American College of Rheumatology guidelines require at least 4 of the following criteria be met for a diagnosis of EGPA: asthma, eosinophilia greater than 10% on differential white blood cell count, mononeuropathy/polyneuropathy, pulmonary infiltrates, paranasal sinus abnormality, and extravascular eosinophils on biopsy.[6] ANCA, both against proteinase 3 and MPO, can be detected in patients with EGPA and may occur variably. Approximately 50% of EGPA patients are ANCA positive with a moderate predilection for antibodies against MPO, consistent with our case.[7]

In our patient, only nerve biopsy provided definitive diagnosis. Although he had a remote history of asthma and currently experienced polyneuropathy, we could not elicit any history of eosinophilia, pulmonary infiltrates, or paranasal sinus abnormalities. A computed