



Recurrent Stroke as the Clinical Onset of POEMS Syndrome

Zigao Wang^a
Lu Xiong^b
Hengbing Zu^a

^aDepartment of Neurology,
Jinshan Hospital, Fudan University,
Shanghai, China

^bDepartment of Anesthesiology,
Tinglin Hospital, Shanghai, China

Dear Editor,

POEMS syndrome is a rare paraneoplastic syndrome caused by a plasma cell disorder that is clinically characterized by polyneuropathy, organomegaly, endocrinopathy, monoclonal plasmacell proliferative disorder, and skin changes.¹ Ischemic diseases of the coronary and lower limb arteries have often been reported in patients with POEMS syndrome.^{2,3} However, acute ischemic stroke is uncommon in POEMS syndrome. Herein we describe a unique patient in whom recurrent ischemic stroke was the presenting feature of POEMS syndrome.

A 65-year-old man presented with the sudden onset of dysarthria. His medical history was noteworthy for an ischemic stroke 1 year previously that involved the left caudate nucleus and subcortical areas (Fig. 1A and B). A physical examination revealed generalized hyperpigmentation, splenomegaly, and edema in both ankles. A neurological examination showed dysarthria and symmetrically reduced sensation in the distal limbs. The deep tendon reflexes were diminished in all limbs. The findings of laboratory tests were normal except for thrombocytosis ($452 \times 10^9/L$) and elevated vascular endothelial growth factor (VEGF, $>2,000$ ng/L). IgA lambda gammopathy was found by serum immunofixation. Brain MRI scans showed infarctions in the border zone between the left anterior cerebral artery and the middle cerebral artery (Fig. 1C and D). Brain computed tomography angiography revealed no major vessel occlusion or focal high-grade stenosis (Fig. 1E). Nerve conduction studies showed mixed demyelinating and axonal neuropathy. Examinations of the bone marrow demonstrated plasmacytosis insufficient for a diagnosis of multiple myeloma ($<5\%$ plasma cells). A diagnosis of POEMS syndrome was made, and the patient was treated with melphalan and prednisone. During a follow-up of 24 months, no ischemic stroke was observed and the neuropathy symptoms improved significantly.

The acronym POEMS captures several dominant (but not all) features of the syndrome. Patients with POEMS syndrome are at increased risk of arterial and/or venous thromboses during their course, with nearly 20% of them experiencing one of these complications.⁴ Dupont et al.⁵ found that about 10% of POEMS patients had a cerebral infarction, with a 5-year risk of cerebral infarction of 13.4%, and a median time between the onset of peripheral neuropathy symptoms and the cerebrovascular event of 23 months. It should be noted that ischemic stroke may precede the onset of peripheral neuropathy.⁶ POEMS-associated strokes tend to be end-artery border-zone infarctions, but those in the major intracranial arterial distribution have also been reported.^{5,7} The mechanisms underlying ischemic stroke in POEMS syndrome remain unclear. Thrombocytosis, polyglobulia, hyperfibrinogenemia, and high levels of circulating proinflammatory cytokines such as VEGF are believed to trigger the hypercoagulable state.^{1,5,7}

Effective strategies for the secondary prevention of stroke in POEMS syndrome remain to be established. No case of cerebral infarction after successful treatment of the underlying condition has been reported.^{5,8} Consistent with this observation, our patient had no further stroke attacks after being treated with melphalan and prednisone. We emphasize that POEMS syn-

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Correspondence

Hengbing Zu, MD, PhD
Department of Neurology,
Jinshan Hospital, Fudan University,
1508 Longhang Road,
Shanghai 201508, China
Tel +86-21-34189990
Fax +86-21-67226910
E-mail hbzyy8@sina.com

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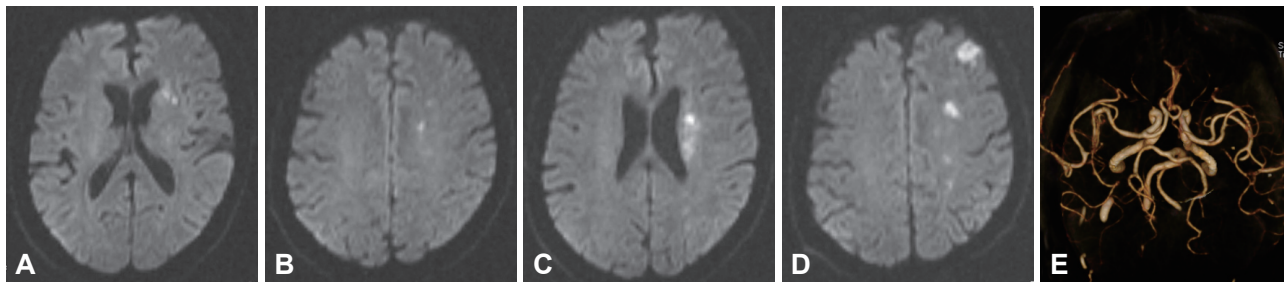


Fig. 1. Neuroimage findings of this patient. A and B: DWI MRI during the first episode showed high signal intensities in the left caudate nucleus and subcortical areas. C and D: DWI MRI during the second episode showed high signal intensities in the border zone between the left anterior cerebral artery and the middle cerebral artery. E: Brain CTA revealed no major vessel occlusion or focal high-grade stenosis. CTA: computed tomography angiography, DWI: diffusion-weighted imaging.

drome is a rare etiology of ischemic stroke, and the treatment of the underlying disease is the best approach for decreasing the risk of ischemic stroke in these patients.

Conflicts of Interest

The authors have no financial conflicts of interest.

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