Case Studies

Vertebrobasilar Artery Stroke as the Heralding Sign of Systemic Lupus Erythematosus

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Cerebral ischemia because of vertebrobasilar insufficiency (VBI) rarely presents as an initial sign within the systemic lupus erythematosus (SLE) population, and there are very few case reports supporting this manifestation. This report details 3 different patients with SLE who experienced VBI as an initial manifestation. Patient 1 was a 24-year-old female who developed a bilateral pontine lesion as a consequence of basilar artery stenosis. Patient 2 was a 34-year-old male with an acute ischemic lesion on the right side of his cerebellum and pons because of significant stenosis in the distal segment of the right vertebral artery. Patient 3 was a 37-year-old female, previously diagnosed with multiple sclerosis, with multiple lesions in her cerebellum and pons bilaterally. Further investigations within this case revealed severe stenosis of the left vertebral artery. The diagnosis of SLE was based on clinical presentations such as myalgia, skin rashes, ulcers, and fatigue along with relevant laboratory findings including positive anti ds-DNA antibody and depressed levels of complement C3 and C4 proteins. In young patients with multifocal ischemic lesions or infarcts in the posterior cerebral circulation system, physicians should investigate for less common etiologies such as SLE. **Key Words:** Systemic lupus erythematosus—vertebrobasilar insufficiency—cerebrovascular events—stroke.
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Introduction

Systemic lupus erythematosus (SLE) is an autoimmune disorder characterized by microvascular inflammation within multiple organ systems secondary to the production of autoantibodies.[1] The disease primarily affects young females and is associated with vascular complications such as premature atherosclerosis and stroke.[2-4] Vascular occlusive heart diseases and associated atherogenesis are commonly reported in patients with SLE.[5,6] The relative risk for stroke has been reported about 8-10 times, and it was noted that approximately 3%-15% of these patients experience a cerebrovascular event at some point during the time course of the disease.[7] Although SLE was understood as one of the more unusual causes of vertebrobasilar insufficiency (VBI), an accurate diagnosis may be limited if stroke symptoms occur before the more common symptoms of SLE. Limited cases have been reported on this topic within published medical literature. In this report, 3 different patients with SLE with initial manifestations of VBI were presented and evaluated (Table 1).

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Table 1. *Suggestive clinical signs and symptoms and diagnostic tests for systemic lupus erythematosus among 3 patients*

Patients	Suggestive clinical signs or symptoms	Diagnostic test leading to diagnosis
Case #1	– Sudden-onset headache – Lack of risk factor for atherosclerosis – Cerebellar and other posterior neurologic signs – Severe segmental stenosis of the basilar artery	– ANA weakly positive – Positive anti ds-DNA Ab – Decreased C3 and C4 complement levels
Case #2	– Cerebellar and other posterior neurologic signs – Severe segmental stenosis of the basilar artery – ESR of 34 mm/h	– Positive anti ds-DNA Ab – Decreased C3 and C4 complement levels
Case #3	– Multiple focal ischemic lesions within the cerebellum and pons on MRI – Severe segmental stenosis of the basilar artery	– Positive anti ds-DNA Ab – Decreased C3 and C4 complement levels

Abbreviations: ANA, antinuclear antibodies; ds DNA, double stranded deoxyribonucleic acid; ESR, erythrocyte sedimentation rate; MRI, magnetic resonance imaging.

Patients

Case 1

A 24-year-old previously healthy female developed sudden onset headache, right-sided (upper or lower extremity, or both) weakness, numbness, and paresthesia along with generalized ataxia. Computerized tomography (CT) of the head showed no irregularities. Based on her clinical presentation, she was diagnosed with a vertebral artery dissection and received intravenous heparin at another hospital. Her neurologic status deteriorated while on treatment, and as a result, she was referred to our team for further neurologic management. The patient denied a history of hypertension, seizures, epilepsy, tobacco, alcohol, and recreational drug use or head trauma. The family history was unremarkable. On physical examination, blood pressure was normal. There was no evidence of skin lesions, musculoskeletal abnormalities, lymphadenopathy, splenomegaly, or hepatomegaly. On neurologic examination, cognition was normal. The patient experienced mild right-sided paresis and hypotonia along with an ataxic gait. Additionally, she required assistance with ambulation.

Complete blood count (CBC), electrolytes, thyroid and kidney function tests, erythrocyte sedimentation rate (ESR), and antiphospholipid antibodies were all normal. The serum homocysteine level was also within normal limits. Serologic tests for syphilis, human T-lymphocyte virus type I, and human immunodeficiency virus were unremarkable. Antinuclear antibodies were weakly positive. There were no renal abnormalities such as proteinuria or cellular casts in the urine analysis. Anti ds-DNA antibodies were positive, whereas C3 and C4 complement levels were decreased.

Transesophageal echocardiography results were normal. Duplex ultrasounds of superficial and deep veins in the lower limbs showed no abnormalities. A magnetic resonance imaging (MRI) of the brain showed bilateral acute ischemic lesions within the pons with left-sided prominence (Fig 1). A CT angiography showed severe basilar artery stenosis, and a digital subtraction angiography (DSA) confirmed this finding (Fig 1).

Case 2

A 34-year-old male developed occipital headaches accompanied with nausea, vomiting, vertigo, ataxia, and dysphagia, which progressed over several hours. He also developed paresthesia in his right arm and leg. His medical, social, along with family histories were unremarkable. Physical examination findings along with blood pressure readings were all within normal limits. Neurologic examination revealed right arm dysmetria, right-sided hyperreflexia, flexor plantar reflexes, and severe gait ataxia (the patient required walking assistance). Finger-to-nose and heel-to-shin performance was reduced on the right side. Cranial nerve examination revealed a reduced gag reflex. Findings on the sensory examination demonstrated right-sided hypoesthesia.

CBC, vitamin B12, immune-electrophoresis, thyroid-stimulating hormone, antinuclear antibody, electrolytes, liver enzymes, glucose, paraneoplastic panel, and anti-phospholipid antibody. Serologic testing for syphilis, human T-lymphocyte virus type I, and human immunodeficiency virus were all unrevealing. The ESR was 34 mm/h. SLE was suspected because of positive titers of antinuclear antibody in serum samples. Further analysis demonstrated positive anti ds-DNA and smooth muscle antibodies along with decreased C3 and C4 levels.

A brain MRI showed evidence of an acute ischemic lesion in the right cerebellar hemisphere along with a small lesion on the left side of the pons (Fig 2). A cerebral CT angiography and DSA showed significant and isolated stenotic lesions within the distal segment of the right