Striatal Encephalitis: Potential Inflammatory Vasculopathy in Systemic Lupus Erythematosus

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We report two patients who developed striatal (basal ganglia) encephalitis as a rare neuropsychiatric manifestation of systemic lupus erythematosus (SLE), both of whom had neuroimaging findings indicating an inflammatory vasculopathy as the underlying etiology.

A 42-year-old female (Patient A) presented with altered mental status. Past medical history was significant for SLE meeting 2019 European League Against Rheumatism (EULAR)/American College of Rheumatology (ACR) classification criteria[1] diagnosed 20 years prior. At presentation, she was taking prednisone 5 mg PO OD. Bloodwork demonstrated elevated erythrocyte sedimentation rate (ESR), elevated double-stranded DNA, and low C3. Anti-phospholipid antibody testing was negative. Cerebrospinal fluid (CSF) evaluation demonstrated zero white blood cells (WBCs)/μl, elevated protein of 156 mg/dL (reference range: 15–35 mg/dL), normal glucose, negative oligoclonal bands, and negative testing for N-methyl-D-aspartate receptor antibodies. Brain magnetic resonance imaging (MRI) (Figure 1) revealed striatal T2-hyperintensity, as well as multifocal punctate hyperintensities on diffusion-weighted imaging (DWI). Corresponding apparent diffusion coefficient (ADC) map hypointensities confirmed true diffusion restriction, compatible with infarctions. Brain magnetic resonance angiography (MRA) was normal. The patient was started on prednisone 60 mg PO OD and cyclosporine 100 mg PO BID. She had clinico-radiographic improvement documented 4 months later.

A 25-year-old female (Patient B) presented with fever and altered mental status. Past medical history was significant for SLE meeting 2019 EULAR/ACR classification criteria[1] diagnosed 2 years prior, juvenile rheumatoid arthritis, and anti-phospholipid syndrome. At presentation, she was taking prednisone 60 mg PO OD, hydroxychloroquine 400 mg PO OD, and mycophenolate mofetil 1000 mg PO BID. Bloodwork demonstrated elevated ESR, elevated double-stranded DNA, low C3, and low C4. CSF evaluation demonstrated two WBCs/μl, elevated protein of 154 mg/dL (reference range: 15–35 mg/dL), and normal glucose. Brain MRI (Figure 1) revealed striatal T2-hyperintensity, as well as multifocal hyperintensities on DWI. Corresponding ADC map hypointensities confirmed true diffusion restriction, compatible with infarctions. Gradient echo (GRE) revealed superimposed hypointensities, compatible with petechial hemorrhages. Brain MRA (Figure 2) demonstrated multifocal arterial stenosis. She was treated with dexamethasone 200 mg IV OD for 3 days and cyclophosphamide 500 mg IV, followed by continuation of prednisone 60 mg PO OD. She had clinico-radiographic improvement documented 1 month later.

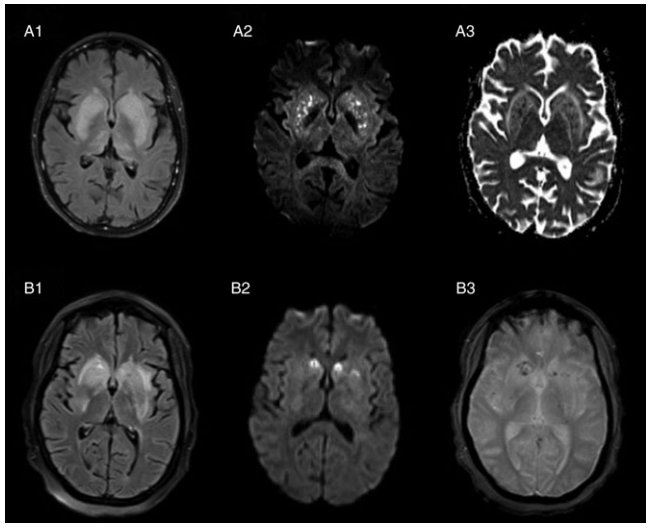

Figure 1: *Brain MRI of striatal encephalitis in systemic lupus erythematosus. Both patients had bilateral striatal T2-hyperintensity (A1 and B1) and DWI hyperintensities (A2 and B2) with corresponding ADC hypointensities (shown for Patient A, A3), compatible with infarctions. In Patient B, GRE revealed superimposed hypointensities, compatible with petechial hemorrhages (B3).*

Previous reports of striatal encephalitis in SLE without restricted diffusion have proposed an antibody-mediated pathophysiology.[2] The imaging findings of multifocal restricted

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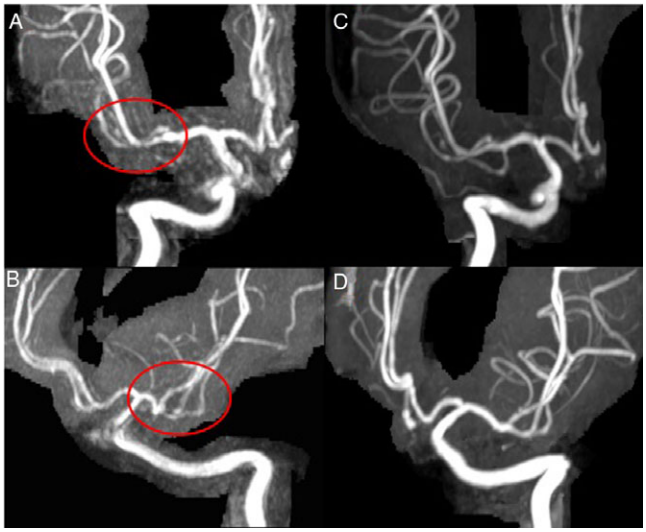

Figure 2: *Brain MRA of striatal encephalitis in systemic lupus erythematosus. Patient B had multifocal middle cerebral artery stenosis bilaterally (A, B, circles) that resolved 10 months later (C and D), compatible with an inflammatory vasculopathy.*

diffusion compatible with infarctions, superimposed petechial hemorrhages and cerebral arterial stenosis we report herein, however, are among the strongest evidence to date indicating an inflammatory vasculopathy in some cases.[3] Recognition of this unique clinico-radiographic syndrome is important to facilitate further study into disease mechanisms, as well as to ensure appropriate diagnosis and management.

Disclosures

The authors have no conflicts of interest to declare.

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