Teaching NeuroImage: Partially Reversible Widespread Leukoencephalopathy Associated With Atypical Hemolytic Uremic Syndrome

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Figure 1 Partially Reversible Widespread Leukoencephalopathy

(A) Admission brain MRI: axial T2 fluid-attenuated inversion recovery (FLAIR) sequences showing extensive hyperintensities of bilateral subcortical areas ("band-like"), basal ganglia, thalami, brainstem, and cerebellar hemispheres (arrows). (B) 1-month and (C) 3-month MRI showing partial resolution of T2 FLAIR hyperintensities and unrevealing disseminated subcortical ischemic lesions (arrows).

A 43-year-old woman presented with altered mental status and hypertension. She had a 3-day history of oliguria. The blood test detected microangiopathic hemolytic anemia, thrombocytopenia, and severe kidney injury. The absence of shiga-like toxin, ADAMTS13 autoantibodies, and normal ADAMTS13 activity were consistent with a diagnosis of atypical hemolytic uremic syndrome (aHUS) and then confirmed by a renal biopsy. Genetic tests (CFH, CFHR1-5, MCP/CD46, CFI,...

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C3, CFB, THBD, and DGKE) were unremarkable. Nevertheless, a history of anemia and kidney failure in her younger brother suggested a genetic etiology.

Brain MRI revealed extensive T2 FLAIR hyperintensities. Treatment with eculizumab and twice-weekly hemodialysis resulted in prompt mental recovery and improvement of MRI abnormalities (Figure 1).

aHUS is an ultra-rare complement-mediated kidney disease occasionally associated with neurologic involvement. Less extensive T2 FLAIR abnormalities involving every CNS structure have also been reported in diarrhea-associated HUS. Here, we presented a case of aHUS-related widespread leukoencephalopathy partially reverting on treatment with eculizumab and hemodialysis.

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References

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