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

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A 43-year-old woman presented with altered mental status and hypertension. She had a 3-day history of oliguria. Blood test detected microangiopathic hemolytic anemia, thrombocytopenia and severe kidney injury. Absence of shiga-like toxin, ADAMTS13-autoantibodies and normal ADAMTS13 activity were consistent with a diagnosis of atypical hemolytic uremic syndrome (aHUS), then confirmed by a renal biopsy. Genetic tests (CFH, CFHR1-5, MCP/CD46, CFI, C3, CFB, THBD, 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.¹ Less extensive T2-FLAIR abnormalities involving every CNS structure have been reported also in diarrhea-associated HUS.² Here we presented a case of aHUS-related widespread leukoencephalopathy partially reverting upon treatment with eculizumab and hemodialysis.
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-and C. 3- month MRI showing partial resolution of T2-FLAIR hyperintensities and unrevealing disseminated subcortical ischemic lesions (arrows).
References


Appendix 1. Authors

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