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

Author(s):
Ilaria Cani, MD1; Matteo Righini, MD2, 3; Patrizia Cenni, MD4; Matteo Foschi, MD5, 6

Corresponding Author:
Matteo Foschi, matteo.foschi@auslromagna.it

Affiliation Information for All Authors: 1. Department of Biomedical and Neuromotor Sciences (DIBINEM), University of Bologna, Bologna, Italy; 2. Nephrology and Dialysis Unit, S. Maria delle Croci Hospital, AUSL Romagna, Ravenna, Italy; 3. Nephrology, Dialysis and Transplantation Unit, IRCCS Azienda Ospedaliero-Universitaria S. Orsola-Malpighi di Bologna, Bologna, Italy; 4. Neuroradiology Unit, S. Maria delle Croci Hospital, AUSL Romagna, Ravenna, Italy; 5. Department of Neuroscience, Neurology Unit, S. Maria delle Croci Hospital, AUSL Romagna, Ravenna, Italy; 6. Department of Medical and Surgical Sciences, University of Bologna, Bologna, Italy.

Equal Author Contribution:

Contributions:
Ilaria Cani: Drafting/revision of the manuscript for content, including medical writing for content; Major role in the acquisition of data; Analysis or interpretation of data
Matteo Righini: Major role in the acquisition of data; Analysis or interpretation of data
Patrizia Cenni: Major role in the acquisition of data; Analysis or interpretation of data
Matteo Foschi: Drafting/revision of the manuscript for content, including medical writing for content; Major role in the acquisition of data; Study concept or design; Analysis or interpretation of data

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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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<thead>
<tr>
<th>Name</th>
<th>Location</th>
<th>Contribution</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ilaria Cani, MD</td>
<td>Department of Biomedical and Neuromotor Sciences, University of Bologna, Italy</td>
<td>Acquisition and analysis of neurological data, manuscript drafting</td>
</tr>
<tr>
<td>Matteo Righini, MD</td>
<td>Nephrology and Dialysis Unit, S. Maria delle Croci Hospital of Ravenna, AUSL Romagna, Italy</td>
<td>Acquisition and analysis of nephrological data</td>
</tr>
<tr>
<td>Patrizia Cenni, MD</td>
<td>Neuroradiology Unit, S. Maria delle Croci Hospital of Ravenna, AUSL Romagna, Italy</td>
<td>Acquisition and analysis of neuroradiological data</td>
</tr>
<tr>
<td>Matteo Foschi, MD</td>
<td>Neurology Unit, S. Maria della Croci Hospital of Ravenna, AUSL Romagna, Italy</td>
<td>Study design and conceptualization, analysis of neurological data, manuscript drafting and revision for intellectual content</td>
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