Adult acute hemorrhagic leukoencephalitis: role of susceptibility-weighted imaging in diagnosis and importance of aggressive early immunotherapy.

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Objective

Describe a case of acute hemorrhagic leukoencephalitis, highlighting early diagnosis with susceptibility-weighted imaging and aggressive immunotherapy treatment regimen leading to substantial patient recovery.

Background

• Acute hemorrhagic leukoencephalitis (AHLE) is a rare, fulminant form of ADEM.

• Patients present days to weeks after an upper respiratory tract infection with multifocal neurologic deficits rapidly progressing to profound coma.1

• Edema with herniation is responsible for death in 68% of patients within the first week.2,4

• Pathology reveals a T cell-mediated demyelinating process, small vessel destruction, microhemorrhage and perivascular fibrin deposition.1,2,7

• MRI expedites diagnosis and improves outcomes. Susceptibility-weighted imaging (SWI) is a useful adjunct sequence as it best illustrates associated iron deposition and microhemorrhage.1,4

Case Description

44-year-old male presented with acute coma and left hemiparesis shortly following headache, fatigue and urinary incontinence. Past medical history was significant for recent bronchitis two weeks before presentation. Exam revealed stupor requiring intubation, intact brainstem reflexes, clonus in right Achilles and upgazing paretic reflexes bilaterally. He had a peripheral leukocytosis of 13.16 x10^9/L with neutrophilic predominance. CSF: pleocytosis with white blood cell count of 41 x10^6/L (63% lymphocytes). Viral studies of serum revealed elevated levels of Influenza A and B immunoglobulins as well as coxsackie B viral titers (Table 1).

MRI brain with contrast revealed widespread multifocal areas of T2/FLAIR hyperintensity within bilateral frontal, parietal, and temporal lobe white matter; bilateral cingulate gyri; and associated rim enhancement. Gradient reduced echo (GRE) was normal without significant areas of hypointensity. Susceptibility-weighted imaging (SWI) on admission showing hypointensities consistent with AHLE, (d) GRE negative on admission, (e) FLAIR at 3 months, (f) Post-contrast sequence at 3 months, (g) SWI at 3 months, (h) GRE at 3 months.

The patient was diagnosed as having AHLE based on clinical presentation and SWI findings. He received five plasma exchange treatments. The patient began opening his eyes spontaneously on hospital day 5 after two doses of Methylprednisolone 1000mg IV daily followed by a prolonged prednisone taper, and five concomitant intravenous immunoglobulins (IVIg) infusions. The patient made meaningful eye contact and was extubated successfully. He was hypophonic, cognitively impaired, and had bilateral oculomotor nerve palsy. He was discharged to rehab on hospital day 16 and made an exceptional recovery.

Conclusions

• AHLE is a rapidly progressive, highly morbid disorder without many positive outcomes reported in the literature.

• SWI was a useful tool for early diagnosis and expedited aggressive treatment. SWI is also more sensitive for hemorrhage than GRE, revealing hypointense venules with surrounding petechial hemorrhage in the areas of FLAIR hyperintensity.1,4

• SWI helps differentiate AHLE from other demyelinating disorders including less severe forms of ADEM.2,3

• Our regimen of corticosteroids with plasma exchange and IVIg was associated with an exceptional outcome.

• This case reinforces the importance of early diagnosis and prompt, aggressive immunotherapies emphasized in the existing literature.

References