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 demyelinative process, small vessel destruction, microhemorrhage and perivascular fibrin deposition.[1,5-7]
- MRI expedites diagnosis and improves outcomes. Susceptibility-weighted imaging (SWI) is a useful adjunct sequence as it best illustrates associated iron deposition and microhemorrhage.[7,8]

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 upgoing plantar reflexes bilaterally.

He had a peripheral leukocytosis of 13.16 k/uL with neutrophilic predominance. CSF: pleocytosis with white blood cell count of 41/uL (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 extensive, 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) showed scattered foci of susceptibility artifact**. (Figure 1).**

The patient was diagnosed as having AHLE based on clinical presentation and SWI findings. He received five doses of Methylprednisolone 1000mg IV daily followed by a prolonged prednisone taper, and five concomitant plasma exchange treatments. The patient began opening his eyes spontaneously on hospital day 5 after two plasma exchanges. On day 7, he opened his eyes to command and tracked the examiner. The following day, he made meaningful eye contact and was extubated successfully. He was hypophonic, cognitively impaired, and hemiparetic for several days thereafter. Intravenous immune globulin (IVIg) was started on hospital day 11 (0.5g/kg daily X 3 days). The patient discharged to rehab on hospital day 16 and made an exceptional recovery.

Repeat imaging on hospital day 20 revealed reductions of gadolinium enhancement, areas of confluent FLAIR hyperintensity, and amount of SWI artifact with complete resolution of MRI findings on 3 month follow up **(Figure 1)**. At 7 months, the patient was without remaining neurological residuum and was able to return to work.

Figure 1

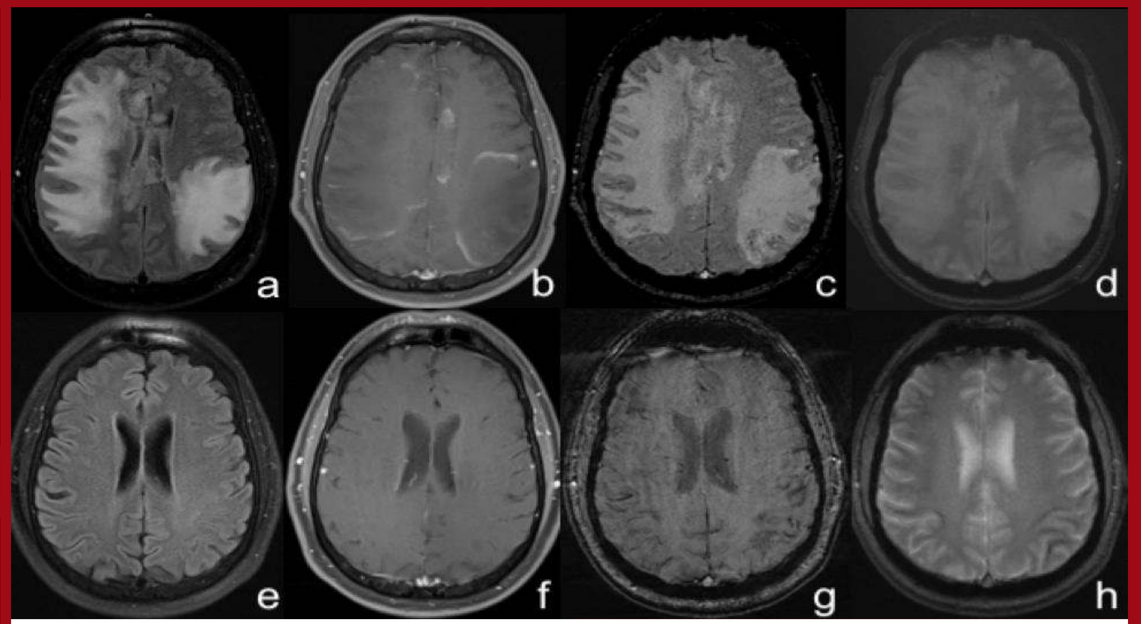

Figure 1: MR Imaging of acute hemorrhagic leukoencephalitis and resolution in a 44-year-old man.
(a) FLAIR hyperintensities on admission, (b) Post-contrast sequence on admission showing rim enhancement, (c) susceptibility-weighted imaging (SWI) on admission showing hypointensities consistent with AHLE, (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.

Influenza A Virus Ab IgG	5.88 IV	1.11 IV or greater: Positive
Influenza A Virus Ab IgM	0.97 IV	0.90 - 1.10 IV: Equivocal
Influenza B Virus Ab IgG	3.40 IV	1.11 IV or greater: Positive
Influenza B Virus Ab IgM	0.95 IV	0.90 – 1.10 IV: Equivocal
Coxsackie B Virus Type 2	1:80 (7/2/18), 1:160 (8/29/18)	
Coxsackie B Virus Type 3	1:40 (7/2/18), 1:80 (8/29/18)	
Coxsackie B Virus Type 4	1:320 (7/2/18), 1:640 (8/29/18)	

Table 1: Serologies and their interpretive data.

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[7,8]
- SWI helps differentiate AHLE from other demyelinating disorders including less severe forms of ADEM.[9]
- 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

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