Hemicraniectomy as a life-saving measure in acute hemorrhagic leukoencephalitis, a rare variant of acute disseminated encephalomyelitis

Brian P Curry, MD
George Rymarczuk, MD
Daniel J Coughlin, MD
Patrick Cooper, MD

Walter Reed National Military Medical Center, Bethesda, MD
Disclosures

None.

The views expressed in this lecture are those of the authors and do not necessarily reflect the official policy or position of the Department of the Navy, Department of the Army, the Department of Defense, nor the U.S. Government.

We certify that all individuals who qualify as authors have been listed; each has participated in the conception and design of this work, the analysis of data, the writing of the document, and the approval of the submission of this version; that the document represents valid work; that if we used information derived from another source, we obtained all necessary approvals to use it and made appropriate acknowledgements; and that each takes public responsibility for it. Nothing in the presentation implies Federal/DOD/DON/DOA endorsement.
Introduction

- Acute hemorrhagic leukoencephalitis (AHL) is a rare, monophasic, rapidly progressive hemorrhagic demyelinating condition, a variant of acute disseminated encephalomyelitis (ADEM).

- It is frequently associated with a dismal neurological outcome, though vigilant monitoring and aggressive medical interventions, including corticosteroids, intravenous immunoglobulins, and plasmapheresis, have been associated with improved outcomes.

- Here we report a case of fulminant edema associated with AHL, for which a decompressive hemicraniectomy was performed as a life-saving measure.
Case Presentation

• A 34-year-old woman presented with rapidly progressive neurological decline. Imaging demonstrated a large, enhancing mass in the right frontal lobe.

• When laboratory and radiographic investigation failed to yield a diagnosis, she underwent stereotactic biopsy of her right frontal lobe lesion.

• Despite aggressive immunomodulatory therapy, she developed malignant edema and ultimately required a life-saving decompressive hemicraniectomy. She was ultimately discharged to acute rehabilitation.
Initial Imaging
a. H&E; b. Luxol Fast Blue; c. Neurofilament Protein; d. CD68; e. CD3; f. CD20
Hallmarks of acute hemorrhagic leukoencephalitis: a. Fibrinoid necrosis; b. Ring hemorrhage; c. Necrosis
Post-craniecytomy MRI

FLAIR | T1Pre | T1C+
Conclusions

• AHL is a rare, fulminant, rapidly progressive hemorrhagic demyelinating condition, often associated with poor outcomes.

• Medical therapies aimed at immune modulation remain the mainstay in the management of this and other variants of ADEM.

• Infrequently, emergency decompressive hemicraniectomy may be considered as a life-saving measure to treat the malignant cerebral edema associated with AHL, an intervention that has only rarely been reported in the literature in the management of this disease.