Research Article

Fulminant Acute Hemorrhagic Leukoencephalitis (Hurst Disease) Mimicking Hemorrhagic Septic Emboli

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Abstract

Acute hemorrhagic leukoencephalitis (AHLE), or Hurst disease, is a rare fulminant inflammatory demyelinating disorder characterized by hemorrhagic white-matter injury, marked cerebral edema, and rapid neurological deterioration. A previously healthy 27-year-old man presented with altered mental status after a 7-day febrile illness and rapidly developed stroke-like deficits, quadriplegia, coma, and hypoxemia. Laboratory evaluation showed marked peripheral leukocytosis, elevated D-dimer, prolonged activated partial thromboplastin time, and inflammatory cerebrospinal fluid abnormalities with pleocytosis, mildly elevated protein, and oligoclonal-band positivity. Blood cultures were sterile and sputum examination for acid-fast bacilli was negative. Non-contrast CT demonstrated numerous bilateral asymmetric hemorrhagic parenchymal lesions involving the supratentorial and infratentorial compartments, with extensive surrounding hypoattenuating edema and diffuse cerebral swelling; the basal ganglia were relatively spared. Septic embolic disease with hemorrhagic transformation was initially considered. Brain biopsy demonstrated vasculocentric hemorrhagic inflammatory injury with perivascular hemorrhage, fibrinoid vascular necrosis, neutrophil-predominant inflammation, and acute perivenular demyelination, establishing AHLE. Despite intensive supportive care, the patient died on the third hospital day. AHLE should be considered in rapidly progressive febrile encephalopathy when multifocal non-territorial hemorrhagic lesions are accompanied by disproportionate edema.

Keywords: Acute Hemorrhagic Leukoencephalitis; Hurst Disease; Hemorrhagic Demyelination; Septic Emboli; Cerebral Edema; Computed Tomography.

Introduction

Acute hemorrhagic leukoencephalitis is an uncommon, frequently fatal inflammatory demyelinating disorder generally regarded as the hyperacute hemorrhagic end of the acute disseminated encephalomyelitis spectrum. It often follows an infectious prodrome and may progress from encephalopathy to focal neurological deficits, coma, and death within days. Imaging typically shows multifocal white-matter abnormalities with edema and variable hemorrhage, but the appearance may overlap with infectious encephalitis, septic emboli, vasculitis, venous infarction, and coagulopathy. We report a rapidly fatal case in a young adult whose striking CT appearance initially suggested hemorrhagic septic embolic disease.

Case Presentation

A 27-year-old man with no significant previous medical history presented with altered mental status following 7 days of fever. His blood pressure at presentation was 150/80 mmHg. Neurological status deteriorated rapidly with development of stroke-like focal deficits, quadriplegia, and subsequent loss of consciousness. Oxygen saturation later declined, requiring intensive-care admission and cardiopulmonary support.

Laboratory studies showed severe leukocytosis, markedly elevated D-dimer, and prolonged activated partial thromboplastin time. Cerebrospinal fluid analysis demonstrated marked pleocytosis, mildly increased protein, and low-level oligoclonal-band positivity. Blood cultures remained sterile, and sputum examination for acid-fast bacilli was negative.

Non-contrast CT of the brain showed numerous bilateral, variable-sized hyperattenuating intraparenchymal hemorrhagic foci distributed asymmetrically throughout the supratentorial cerebral parenchyma, with additional hemorrhagic lesions in the infratentorial compartment. The lesions were surrounded by prominent

hypoattenuating edema. Diffuse cerebral edema with generalized sulcal effacement was also present, while the basal ganglia were relatively spared. In the clinical setting of fever and marked leukocytosis, septic embolic disease with hemorrhagic transformation was the leading initial differential diagnosis; hemorrhagic encephalitis, central nervous system vasculitis, and coagulopathy-related hemorrhage were also considered.

Because of rapid deterioration and persistent diagnostic uncertainty, brain biopsy was performed. Histopathology demonstrated a severe vasculocentric hemorrhagic inflammatory process with characteristic perivascular ring-and-ball hemorrhages, fibrin exudation and fibrinoid necrosis of small vessels, neutrophil-predominant perivascular inflammation, acute perivenular demyelination, and parenchymal necrosis, consistent with AHLE. Despite intensive supportive treatment, the patient continued to deteriorate and died on the third hospital day.

Discussion

AHLE differs from conventional ADEM by its abrupt course, prominent hemorrhage, vascular injury, and high mortality. In a systematic review of 43 adult cases, 67% of patients were male, mean age was 38 years, mortality was 47%, CSF protein was elevated in 87%, and CSF leukocytosis occurred in 65% [1]. The clinical sequence in our patient—a febrile prodrome followed by encephalopathy, focal deficits, quadriplegia, coma, and death within three hospital days—fits the fulminant end of the reported spectrum.

MRI is the imaging modality of choice for characterizing AHLE. FLAIR sequences define the burden of inflammatory white-matter abnormality, while susceptibility-sensitive sequences are particularly useful for detecting petechial and microhemorrhagic components [2,3]. CT, however, may be the first practical examination in critically ill patients. Gibbs et al. described extensive bilateral hypoattenuation, mass effect, and microhemorrhage on early CT in biopsy-confirmed AHLE [4]. Infratentorial involvement is also well documented in severe disease [5].

The main radiological dilemma in this case was septic embolic disease. Septic emboli may produce multifocal infarcts, hemorrhagic transformation, and microabscesses in febrile patients with leukocytosis. The extensive non-territorial distribution, edema out of proportion to the hemorrhagic foci, relative deep-gray sparing, unrevealing available microbiological studies, and subsequent biopsy findings favored AHLE. Negative blood cultures alone cannot exclude septic emboli, making clinicoradiologic-pathological correlation essential. Acute necrotizing encephalopathy is another febrile encephalopathy but typically shows symmetric thalamic or other deep-gray involvement. Primary hypertensive hemorrhage was less likely given the patient's age, modest presenting blood pressure, and multifocal distribution outside the usual deep perforator territories.

The pathological substrate of AHLE explains its imaging appearance. Perivascular hemorrhage, fibrin deposition and small-vessel necrosis, neutrophil-rich inflammation, perivenular demyelination, and tissue necrosis have been repeatedly described [1,4,6]. These changes account for the coexistence of hemorrhage and extensive edema. Early recognition is important because survival has been reported with aggressive immunomodulatory treatment, although no standardized treatment regimen exists because of the rarity of the disorder [1].

Conclusion

AHLE should be included in the differential diagnosis of a young patient with a recent febrile illness, rapidly progressive encephalopathy, and widespread multifocal hemorrhagic brain lesions with marked surrounding edema. Although MRI provides superior tissue characterization, CT may supply the first diagnostic clue in an unstable patient. A non-territorial hemorrhagic pattern, disproportionate edema, rapid clinical decline, and supportive tissue findings help distinguish AHLE from hemorrhagic septic emboli and other vascular or infectious mimics.

Figure

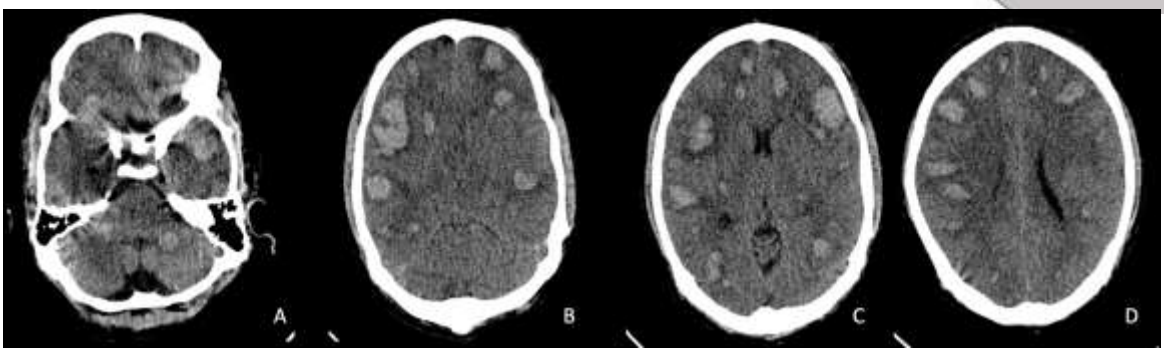

Figure 1. Non-contrast CT findings in acute hemorrhagic leukoencephalitis (Hurst disease). Axial images at sequential levels (A-D) show numerous bilateral, multifocal, variable-sized hyperattenuating hemorrhagic lesions involving the supra- and infratentorial brain parenchyma, with prominent surrounding hypoattenuating edema. Associated diffuse cerebral edema with sulcal effacement is present. The basal ganglia are relatively spared. In the setting of acute febrile encephalopathy and leukocytosis, the multifocal hemorrhagic appearance initially raised suspicion for septic embolic disease with hemorrhagic transformation.

References

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