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Acute hemorrhagic leukoencephalitis: A Fatal Adult Case Highlighting the Diagnostic Value of MRI

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ABSTRACT

Acute hemorrhagic leukoencephalitis (AHLE) is a rare, fulminant hemorrhagic demyelinating disorder regarded as the most severe form of acute disseminated encephalomyelitis. A previously healthy 22-year-old woman presented with 2 days of fever, headache, vomiting, and meningeal symptoms, followed by drowsiness and signs of raised intracranial pressure. Emergency computed tomography showed diffuse sulcal effacement and a left parieto-occipital white-matter hypodensity containing punctate hyperdense foci. Magnetic resonance imaging demonstrated predominantly unilateral T2/FLAIR hyperintense lesions involving the left parieto-occipital white matter, middle cerebellar peduncle, cerebellar hemisphere, and vermis. The lesions were associated with marked vasogenic edema, susceptibility foci consistent with hemorrhage, mild parenchymal enhancement, and patchy leptomeningeal enhancement, without restricted diffusion. The clinicoradiological pattern supported a diagnosis of probable AHLE. According to the treating neurologist, immunotherapy was initiated; however, the specific agents, dosage, timing, and duration could not be retrieved from the available medical records. The patient's neurological condition continued to deteriorate, and she died during hospitalization. The immediate cause of death could not be established.

This case emphasizes that AHLE may show a predominantly unilateral supra- and infratentorial distribution and that susceptibility-weighted imaging is essential for identifying its hemorrhagic component.

KEYWORDS

acute hemorrhagic leukoencephalitis; Hurst disease; demyelination; susceptibility-weighted imaging; intracranial hypertension

MAIN ARTICLE

INTRODUCTION:

Acute hemorrhagic leukoencephalitis (AHLE), or Hurst disease, is an exceptionally rare inflammatory demyelinating disorder characterized by rapidly progressive encephalopathy, necrotizing small-vessel injury, and hemorrhage within white-matter lesions. It is generally considered the most fulminant form of acute disseminated encephalomyelitis (ADEM) [1,2]. A 2025 review of 152 published cases found a mortality rate of 58%, illustrating the severity of the disease [3]. Diagnosis is challenging because the initial presentation may resemble infectious meningoencephalitis, cerebral venous thrombosis, vasculitis, posterior reversible encephalopathy syndrome, or a tumefactive lesion. We report a fatal case in a young adult with predominantly unilateral supratentorial and infratentorial involvement, highlighting the contribution of multimodal MRI, particularly susceptibility-weighted imaging (SWI).

CASE REPORT:

A 22-year-old woman with no relevant medical history was admitted after two days of fever, headache, vomiting, and meningeal symptoms. On admission, her temperature was 41°C, blood pressure 100/70 mmHg, and Glasgow Coma Scale score 12/15. She was conscious but mildly drowsy. Neck stiffness was marked, with positive Kernig and Brudzinski signs. Fundoscopy demonstrated grade III papilledema. No focal motor, sensory, or cranial nerve deficit was identified.

Available initial laboratory investigations, including complete blood count, C-reactive protein, renal function, liver function, and coagulation tests showed no significant abnormalities. Lumbar puncture was not performed because of raised intra cranial pressure. Emergency brain CT performed before and after contrast administration showed diffuse supratentorial and infratentorial sulcal effacement. A left parieto-occipital white-matter hypodensity with vasogenic edema contained scattered punctate hyperdense foci, raising concern for a hemorrhagic inflammatory or infectious process (Fig. 1 A,B).

Brain MRI obtained the day after demonstrated poorly margined T2/FLAIR hyperintense lesions involving the left parieto-occipital white matter, left middle cerebellar peduncle, left cerebellar hemisphere, and vermis. SWI showed multiple intralesional susceptibility foci consistent with hemorrhage. Diffusion-weighted imaging showed no convincing restriction. After gadolinium, the lesions enhanced mildly and heterogeneously,

with patchy leptomeningeal enhancement along the left occipital sulci and cerebellar folia. There was extensive surrounding edema and local mass effect (Figs. 1 and 2).

Based on the fulminant clinical course and the presence of hemorrhagic white-matter lesions, a probable diagnosis of AHLE was considered. According to the treating neurologist, immunotherapy was initiated. However, the specific agent or combination of agents, dosage, timing, duration, and clinical response could not be retrieved from the available medical records. Her neurological status deteriorated to coma, and she died on the third day after admission. The immediate cause of death was not documented, and no autopsy was performed. Her neurological status nevertheless deteriorated to coma and she died 3 days after her admission. Autopsy was not performed.

DISCUSSION:

The present case illustrates the diagnostic challenge posed by AHLE, a fulminant inflammatory demyelinating disorder whose neuroimaging findings may initially mimic infectious, vascular, or neoplastic diseases. Early recognition of its characteristic MRI features is crucial because prognosis depends on prompt diagnosis and treatment. (2,4).

The most recent comprehensive PubMed-based literature review identified only 152 published cases between 1955 and 2024, more than one-third of which were reported after 2020 (13). In this cohort, the mean age was approximately 36 years, with a slight male predominance, and the overall mortality rate reached 58%. The median interval from symptom onset to treatment initiation or death was only 2 days, illustrating the hyperacute nature of the disease (13). An earlier PRISMA-based systematic review reported an overall mortality rate of 46.5% (14). Historically, mortality rates as high as 70% have been reported, with death frequently occurring within the first week after neurological symptom onset (15).

This underscores the importance of maintaining a high index of suspicion and recognizing the distinctive imaging features of AHEM, as early diagnosis remains pivotal in this potentially fatal condition.

The underlying mechanism of AHEM is thought to involve an aggressive autoimmune response triggered by a preceding infection (5,6), vaccination, or, less frequently, trauma. The pathological hallmark consists of perivenular demyelination associated with necrotizing vasculitis of small vessels, fibrinoid necrosis, and perivascular hemorrhage. This vascular

injury explains the hemorrhagic foci typically observed on susceptibility-weighted imaging (SWI) and distinguishes AHLE from classic ADEM.

Clinically, patients present with rapid neurological deterioration, fever, altered consciousness, and often intracranial hypertension (3).

Compared with ADEM, AHLE demonstrates a more aggressive course and worse prognosis (2,4).

Non-contrast CT usually demonstrates ill-defined hypodense white matter lesions associated with diffuse cerebral edema and sulcal effacement. Hemorrhagic foci may be subtle or absent initially but, when present, strongly support the diagnosis in the appropriate clinical context.

After contrast administration, CT may reveal leptomeningeal enhancement, patchy or mild parenchymal enhancement and rarely, ring enhancement (less typical than in abscesses).

MRI is the imaging modality of choice because it accurately depicts the distribution of demyelinating lesions, hemorrhagic components, and associated edema. Typical findings include large asymmetric T2/FLAIR hyperintense white matter lesions containing hemorrhagic foci on susceptibility-weighted imaging, variable gadolinium enhancement, and marked mass effect. Involvement of the cerebellum or brainstem further supports a severe form of the disease.

The presence of hemorrhagic foci within demyelinating lesions is a defining imaging characteristic. Involvement of the cerebellum, brainstem, or deep white matter may occur and is often associated with severe clinical manifestations. Compared with ADEM, lesions in AHLE tend to be larger, more edematous, and hemorrhagic, frequently producing significant mass effect.

From a neuroradiological perspective, this case illustrates how multimodal imaging can substantially narrow the differential diagnosis in patients presenting with fulminant encephalopathy. The coexistence of asymmetric white matter lesions, intralesional hemorrhage on susceptibility-weighted imaging, marked vasogenic edema, and patchy leptomeningeal enhancement strongly favors AHLE over more common infectious, vascular, or neoplastic disorders. Recognition of this characteristic imaging pattern is particularly important because early diagnosis may expedite immunomodulatory treatment in a disease associated with a high mortality rate.

The main differential diagnoses include: (7,8)

<u>Differential diagnosis:</u>	<u>Radiological features supporting the diagnosis:</u>	<u>Radiological features arguing against the diagnosis:</u>
Classic ADEM	Bilateral, asymmetric, poorly defined white-matter lesions.	Hemorrhagic foci, necrosis, marked edema, and significant mass effect are unusual.
Infectious meningoencephalitis	Cortical or subcortical lesions, diffusion restriction, leptomeningeal enhancement, possible hemorrhage.	Predominant white-matter involvement, intralesional hemorrhagic foci, absence of a characteristic temporal or insular distribution, and lack of convincing diffusion restriction.
Herpes simplex encephalitis	Hemorrhagic lesions with diffusion restriction.	Absence of mesial temporal, insular, and orbitofrontal involvement.
PRES	Bilateral posterior white-matter edema with possible hemorrhage.	Extensive necrosis, marked mass effect, and prominent diffusion restriction are atypical.
Primary CNS vasculitis	Multifocal ischemic or hemorrhagic lesions in different vascular territories.	Absence of territorial infarcts.
Cerebral venous thrombosis	Hemorrhagic edema or venous infarction.	Absence of a venous territorial distribution.
Tumefactive demyelination	Large white-matter lesions with edema, mass effect, and incomplete ring enhancement.	Multiple hemorrhagic foci and widespread necrosis are uncommon.
High-grade glioma	Necrosis, hemorrhage, enhancement, and mass effect.	Multifocal supra- and infratentorial distribution, and absence of a dominant infiltrative tumor mass.
Toxic or metabolic leukoencephalopathy	Bilateral white-matter lesions with diffusion restriction.	Hemorrhage, enhancement, marked edema, and mass effect are unusual.

LIMITATION:

This report has several limitations. The diagnosis was established on the basis of the characteristic clinical presentation, multimodal neuroimaging findings, and exclusion of alternative diagnoses. The absence of histopathological confirmation reflects a common limitation in fulminant cases, where rapid clinical deterioration may preclude biopsy or autopsy.

CONCLUSION:

AHLE should be considered in a rapidly progressive febrile encephalopathy when MRI demonstrates edematous white-matter lesions containing hemorrhagic foci. A predominantly unilateral distribution does not exclude the diagnosis. SWI, diffusion imaging, post-contrast sequences, and vascular imaging are central to distinguishing AHLE from infectious, vascular, and neoplastic mimics. Recognition of this characteristic imaging pattern may facilitate earlier diagnosis and timely immunomodulatory therapy in this life-threatening disease.(3,8)

FIGURES :

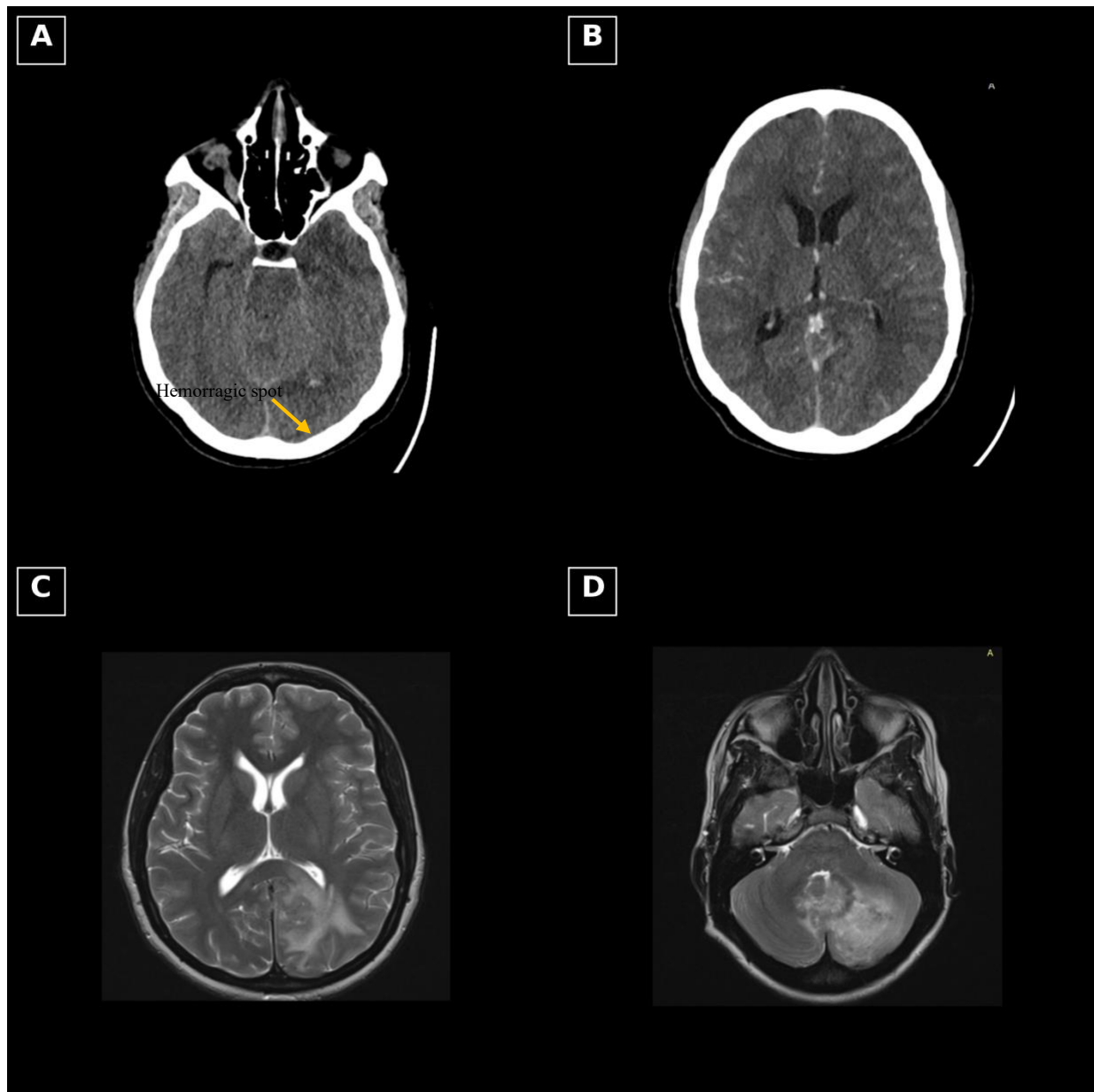


Figure 1. Initial CT and T2-weighted MRI. (A, B) Axial CT images demonstrate diffuse sulcal effacement; the complete examination showed left posterior white-matter hypoattenuation with scattered punctate hyperdense foci suspicious for hemorrhage. (C) Axial T2-weighted image shows a poorly marginated left parieto-occipital white-matter hyperintensity with marked edema. (D) Axial T2-weighted image demonstrates involvement of the left cerebellar hemisphere and middle cerebellar peduncle.

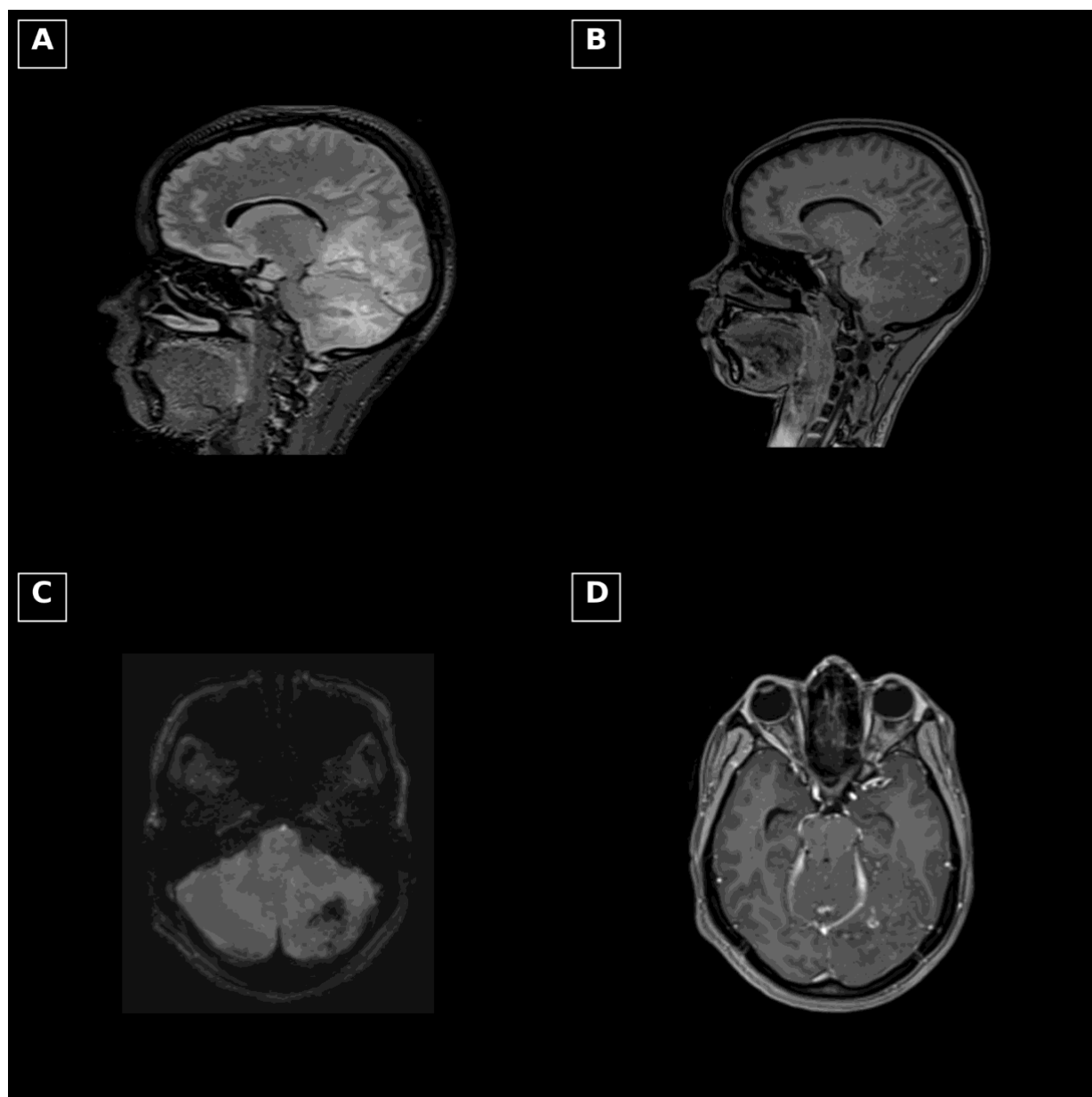


Figure 2. Multisequence MRI findings. (A) Sagittal FLAIR image demonstrates contiguous posterior supratentorial and infratentorial hyperintensity. (B) Sagittal post-gadolinium T1-weighted image shows mild focal parenchymal enhancement. (C) Axial SWI reveals intralesional susceptibility foci consistent with hemorrhage. (D) Axial post-gadolinium T1-weighted image shows patchy leptomeningeal enhancement along the left occipital sulci and cerebellar folia.

DECLARATIONS

Patient consent for publication: Written informed consent for publication of clinical information and images was obtained from the patient's legally authorized representative.

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Data availability: The data supporting this case are available from the corresponding author on reasonable request, subject to patient confidentiality.

CRedit authorship contribution statement:

Salma Tabati: Conceptualization, Investigation, Data curation, Visualization, Writing – original draft, Writing – review and editing.

Nidal El Hassani: Investigation, Data curation, Visualization, Writing – review and editing.

Fatma El Mabrouk: Investigation, Data curation, Writing – review and editing.

Kamal Hlioui: Investigation, Data curation, Writing – review and editing.

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Hamza Retal: Validation, Supervision, Writing – review and editing.

Meriem Fikri: Conceptualization, Validation, Supervision, Writing – review and editing.

Firdaous Touarsa: Conceptualization, Validation, Supervision, Project administration, Writing – review and editing.

All authors have read and approved the final version of the manuscript.

Declaration of generative AI and AI-assisted technologies in the manuscript preparation process:

During the preparation of this work, the authors used OpenAI to improve the organization, language, and readability of the manuscript. After using this tool, the authors reviewed and edited the content as needed and take full responsibility for the content of the publication.

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