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Letter to the Editor

For Patients who Die from Weston-Hurst Syndrome, An Autopsy Could be Helpful to Confirm the Diagnosis

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Letter to the Editor

We read with interest the article by Aidouni *et al.* about a 4-year-old girl who was diagnosed with Weston-Hurst syndrome (acute hemorrhagic encephalomyelitis (AHE)), who had initially been misdiagnosed with Guillain-Barré syndrome (GBS) because she first presented with ascending flaccid tetraparesis one week after a flu-like bronchopulmonary infection ^[1]. However, cerebral magnetic resonance imaging (MRI) revealed bilateral, confluent, T2- and fluid-attenuated inversion recovery (FLAIR)-hyperintense lesions in the white matter, and susceptibility-weighted imaging suggested hemorrhagic petechiae. Despite treatment with glucocorticoids and intravenous immunoglobulins, her condition continued to deteriorate, and the patient died of cerebral edema one day after admission ^[1]. The study is forward-looking, but several points warrant discussion.

The first point is that the cerebrospinal fluid (CSF) was not tested for neurotropic viruses (including severe acute respiratory syndrome (SARS)-corona virus-2 (CoV-2)), bacteria, protozoa, fungi, and parasites by means of rapid tests which provide a result within a few hours. CSF can also be analyzed using Gram staining within a few hours, allowing a bacterial infection to be either ruled out or confirmed. Even if the cell counts and protein content in the cerebrospinal fluid are normal, this does not rule out viral meningitis or encephalitis, especially at the onset of the illness ^[2]. Other differential diagnoses for AHE that were not sufficiently ruled out include acute disseminated encephalomyelitis, hemorrhagic stroke, fulminant multiple sclerosis, primary central nervous system (CNS) vasculitis, venous sinus thrombosis, and malignancies ^[3].

Finally, no reference values for blood chemistry were provided. This makes it difficult to assess what is normal and what is abnormal.

A limitation of the study is that no autopsy was performed ^[1]. Autopsy could have supported the diagnoses, could have revealed the cause of the flu-like infection that preceded the brain catastrophe, and could have revealed signs of necrotizing vasculitis, which is believed to be responsible for the typical ring-shaped hemorrhages seen in AHE ^[4].

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