Noncirrhotic Hyperammonemic Encephalopathy Causing Bilateral Cortical Diffusion Restrictions

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Keywords

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A 32-year-old male with factor IX deficiency, untreated hepatitis C, and suspected epilepsy presented to our hospital with encephalopathy following an episode of thrashing movements like his previous episodes. He was recently started on valproic acid for suspected seizures. Clinical examination performed immediately after intubation revealed diffuse hyperreflexia, bilateral lower extremity clonus, and asterixis, in addition to the patient's comatose state. His ammonia on admission was elevated to 860 $\mu\text{mol/L}$, with normal liver function panel, coagulation panel, albumin, and valproic acid serum level. Hepatitis C viral load was 1.57 million copies/mL. Abdominal ultrasound with Doppler revealed normal liver morphology and was without abnormalities. Electroencephalogram showed diffuse suppression of activity. Brain magnetic resonance imaging (MRI) revealed symmetric areas of cortical diffusion restriction (CDR) involving frontal, temporal, and parietal lobes including cingulate gyrus and insular cortex, consistent with hyperammonemic encephalopathy (Figure 1).^{1,2} Valproic acid was held, and the patient was treated with intravenous vancomycin and an aggressive bowel regimen including lactulose with resolution of symptoms in a week. Repeat computed tomography scan of the head showed resolution of edema (Figure 2). Patients who present with acute-onset encephalopathy and symmetric CDR on MRI have a unique differential diagnosis including hypoxic-ischemic encephalopathy, seizures, hypoglycemia, and hyperammonemia.¹ Due to the similar clinical manifestations and MRI findings of the different causes of symmetric CDRs, physicians must consider rare but reversible causes such as noncirrhotic hyperammonemic encephalopathy. Accurate diagnosis is crucial as the underlying disease can be fatal if treatment is not started early.

Authors' Note

All authors have contributed substantively to the conception, design, or analysis and interpretation of the data, contributed substantively to the drafting of the manuscript or critical revision for important

intellectual content, given final approval of the version to be published, and agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

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Figure 1. Cortical diffusion restriction along the cingulate gyri (A-D), insula and parietal lobes (C and D), and temporal lobes (E and F). Minimal diffusion restriction in thalami. Mild T2 and Fluid Attenuation Inversion Recovery hypersignal intensity involving the basal ganglia which is more conspicuous than Diffusion Weighted Imaging and Apparent Diffusion Coefficient sequences. In addition, mild cortical T2 and Fluid Attenuation Inversion Recovery hypersignal intensity which correlate with diffusion restriction sparing the occipital lobes (G and H). No evidence of hemorrhage or abnormal enhancement (I and J).

Figure 2. Axial brain computed tomography (CT) show diffuse brain edema and effacement of sulci (A and C). Follow-up CT 22 days later and after treatment show resolution of diffuse cerebral edema (B and D).