Case 16106

Less commonly reported but fairly pathognomonic radiological appearance of acute severe hepatic encephalopathy on MRI brain.

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Section: Neuroradiology
Area of Interest: Neuroradiology brain
Procedure: Imaging sequences
Procedure: Diagnostic procedure
Imaging Technique: MR-Diffusion/Perfusion
Imaging Technique: MR
Imaging Technique: CT
Special Focus: Pathology Case Type: Clinical Cases
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Patient: 34 years, male

Clinical History:

34 years old male with history of liver cirrhosis admitted for pretransplant workup. Initial workup shows negative hepatitis B/C serology, negative autoimmune and Wilson’s marker and it was labelled as cryptogenic liver cirrhosis. During the course of admission patient suddenly developed generalized tonic clonic seizures.

Imaging Findings:

Figures 1-3 Axial FLAIR and Figures 4-6 Axial DWI. Diffuse cortical hyperintensities involving bilateral cerebral hemispheres on FLAIR with characteristic sparing of the bilateral perirolandic and occipital cortex. DWI shows diffusion restrictions in the involved cortical regions. These imaging findings are typical of hepatic encephalopathy.

Discussion:

Hepatic encephalopathy (HE) or portosystemic encephalopathy is a spectrum of neuropsychiatric abnormalities often associated with hepatic failure. Primarily this condition is related to hepatic cirrhosis and portal-systemic shunts or portal hypertension, however, it could be observed in fulminant hepatic failure and, infrequently, with portal-systemic shunt without intrinsic hepatic dysfunction. While portosystemic encephalopathy is a clinical diagnosis, various radiologic modalities, such as Magnetic Resonance imaging (MRI), can aid in the diagnosis. [1] MRI can often detect the parenchymal injury at both early and late stages. In the initial stage, imaging identifies specific regions accountable for the acute onset of clinical dysfunction, however in the late phase, when the neurologic changes have become permanent; neuroimaging permits recognition of neurologic sequelae. However, the degree and amount of the neurologic abnormalities may not essentially correspond the clinical status in either occasion. Understanding of the radiological changes of several toxic brain insults may facilitate in narrowing the diagnosis in
acute encephalopathy cases. [2]
Typical MRI findings are hyperintensities in the globus pallidus on T1-weighted images, most likely related to high tissue concentrations of manganese. [3] Arnold et al [4] mentioned in a case report that diffuse cortical lesions without involvement of the occipital and perirolandic cortices, with primary involvement of the cingulate gyrus and the insular cortex in both adults and pediatric patients is highly specific MRI finding in hepatic encephalopathy. Therefore, the cingulate gyrus and insular cortex appear to be specifically susceptible to hyperammonemichyperglutaminergic encephalopathy, while the perirolandic and occipital cortices sound comparatively resistant. However, the pathophysiologic mechanism of specific regional vulnerability in hyperammonemic encephalopathy is not evidently proven. [5]
It is critical to diagnose hepatic encephalopathy as early as possible and combination of clinical signs and symptoms with neuroimaging features can be extremely beneficial in order to make prompt diagnosis and treatment planning avoiding permanent dysfunctions. [6]

"Written informed patient consent for publication has been obtained"

**Differential Diagnosis List:** Acute severe hepatic encephalopathy, Status epilepticus, Hypoxic-ischemic encephalopathy, Hypoglycemic encephalopathy, Creutzfeldt–Jakob disease, Uremic encephalopathy

**Final Diagnosis:** Acute severe hepatic encephalopathy

**References:**

**Figure 1**

*Description:* Diffusion Weighted sequence of the brain shows typical cortical diffusion restriction sparing periorolandic cortex bilaterally. *Origin:* Department of Neuroradiology, Prince Sultan Military Medical City, Riyadh, KSA.
**Description:** Diffusion Weighted sequence of the brain shows typical cortical diffusion restriction in cerebral hemispheres with sparing of cerebellar hemispheres. **Origin:** Department of Neuroradiology, Prince Sultan Military Medical City, Riyadh, KSA.
Description: Diffusion Weighted sequence of the brain shows bilateral thalamic diffusion restriction in addition to cortical involvement. Origin: Department of Neuroradiology, Prince Sultan Military Medical City, Riyadh, KSA.
Figure 2

**Description:** Diffuse cortical swelling with hyperintensity sparing periorolandic cortex on both sides

**Origin:** Department of Neuroradiology, Prince Sultan Military Medical City, Riyadh, KSA
Description: Diffuse cortical swelling and hyperintensity. Origin: Department of Neuroradiology, Prince Sultan Military Medical City, Riyadh, KSA
Description: Pronounced hyperintensity in both thalami in addition to slight signal abnormality in both basal ganglia

Origin: Department of Neuroradiology, Prince Sultan Military Medical City, Riyadh, KSA
Figure 3

Description: Follow up CT brain of the same patient 11 days after the MRI shows severe brain edema with loss of gray-white distinction. 

Origin: Department of Neuroradiology, Prince Sultan Military Medical City, Riyadh, KSA