A 73-Year-Old Woman With Hyperammonemic Encephalopathy Caused by Noncirrhotic Congenital Portosystemic Shunts

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The authors report no disclosures relevant to the manuscript.
A 73-year-old woman with hyperammonemic encephalopathy presented with recurrent, transient episodes of diminished memory and confusion lasted for 2-3 days. Except, ammonia levels were 208.7 μmol/L (normal range 10.71–32.13 μmol/L), the laboratory tests, including all liver tests, were normal. Neuropsychological assessment showed cognitive decline (4/30 of MMSE and 1/30 of MoCA). The EEG showed triphasic waves which disappeared along with clinical remission. MRI showed bilateral symmetrical lesions in cortex, cingulate, lentiform nucleus and insular (Figure 1). An abdominal CTA found congenital portosystemic shunts connected the superior mesenteric vein and the inferior vena cava, without liver cirrhosis signs (Figure 2).

Figure legends
Fig 1. FLAIR (A, B) showed bilateral frontal parietal cortical high signal (arrows), as well as signal abnormalities in cingulate, lentiform nucleus and insular (arrowheads). DWI (C, D) showed restricted diffusion in corresponding locations in accord with FLAIR.

Fig 2. An anomalous vessel (red arrowheads) was identified between superior mesenteric vein (white arrowheads) with inferior vena cava (yellow arrowheads). Arrows pointed pancreas.
References:


Appendix 1 Authors

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<tr>
<th>Name</th>
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