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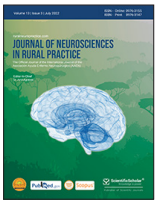

Letter to Editor

Non-cirrhotic Non-Wilsonian degeneration in antiphospholipid syndrome: Association or chance

Sunil Kumar[1], Sanyukta Hepat[1], Sourya Acharya[1], Sachin Agrawal[1], Vijay Kota[1]

[1]Department of Medicine, Jawaharlal Nehru Medical College, Datta Meghe Instititute of Medical Sciences, Wardha, Maharashtra, India.

Dear Editor

A 31-year-old non-alcoholic male patient presented with complaints of hematemesis and melena in conscious but disoriented state. On admission, his vital was stable. Abdominal examination revealed soft, non-tender spleen, and without hepatomegaly. Other system was normal. On upper gastrointestinal endoscopy, he had large esophageal varices which were treated effectively with band ligation. His magnetic resonance imaging of brain revealed bilateral symmetrical T1W1 hyperintensity in basal ganglia suggestive of Non-Wilsonian (acquired) hepatocerebral degeneration (AHD), as shown in [Figure 1]. Kayser–Fleischer rings were absent on slit-lamp examination. Cerebrospinal fluid analysis as well as serum copper, manganese, and ceruloplasmin level normal. Twenty-four-hours urinary copper level was also within the normal range. The patient had elevated serum ammonia levels (124 mg/dL). On further workup, he was found to be antiphospholipid antibody IgG positive (36 U/mL). Anticardiolipin antibody and lupus anticoagulant were negative. All routine laboratory parameter were within normal limit.

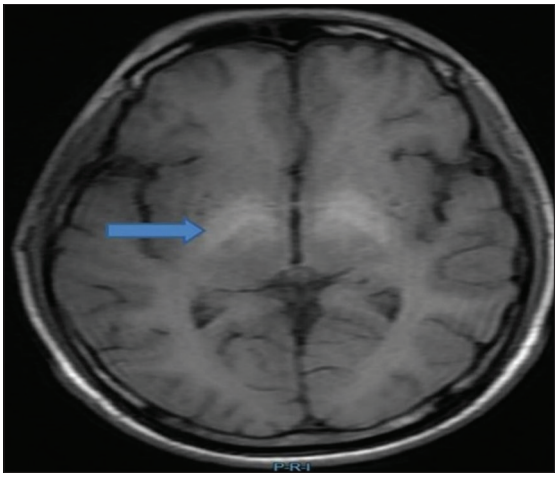

Figure 1: Magnetic resonance imaging revealing bilateral symmetrical T1W1 hyper intensitiy in basal ganglia suggestive of acquired hepatocerebral degeneration (blue arrow).

The pathophysiology of Non-Wilsonian AHD is not very well understood; however, ammonia or manganese accumulation in the basal ganglia and other parts of brain tissue has been reported.[1] The most important factor in the development of this condition is the development of porto-systemic shunting which leads to toxic substances like ammonia to enter into the systemic circulation. This crosses the blood–brain barrier and leads to the accumulation of ammonia in the brain and causing AHD.[2] Due to its effect on astrocyte function, hyperammonemia plays an important role in the development of AHD. Through glutamine synthetase, astrocytes play a homeostatic role in glutamate uptake and release. When there are higher ammonia levels in the brain, glutamine synthetase activity is inhibited leading to the accumulation of glutamine and brain edema. Higher ammonia concentration also activates nitric oxide synthase signaling leading to further nitrosative stress in the central nervous system. Increased ammonia levels may interact with pro-inflammatory cytokines and toxic metals like manganese to produce synergistic effects.[3] When the disease subsides, the neurological symptoms fade away leaving no residual biological damage. In our patient, it was antiphospholipid antibody syndrome that had led to portal thrombosis which, in turn, led to non-cirrhotic portal hypertension, leading to increased ammonia levels in systemic circulation which crossed the blood–brain barrier and caused AHD. As AHD is commonly seen associated with liver cirrhosis, in case, this is found in a patient with non-cirrhotic liver

***Corresponding author:** Sunil Kumar, Department of Medicine, Jawaharlal Nehru Medical College, Datta Meghe Instititute of Medical Sciences, Wardha, Maharashtra, India. sunilkumarmed@gmail.com

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think of other associated conditions like antiphospholipid antibody syndrome.

Declaration of patient consent

Patient's consent not required as there are no patients in this study.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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