MARCHIAFAVA-BIGNAMI DISEASE: HAVE YOU EVER HEARD OF THIS?

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INTRODUCTION

Marchiafava–Bignami disease (MBD) is a rare disorder characterized by primary demyelination and necrosis of the corpus callosum. This disorder is mainly encountered, although not exclusively, in nutritionally depleted chronic alcoholics, resulting in usually fatal neurologic disease.

CASE REPORT

A 55- year-old male with a past medical history significant for chronic alcoholism and who had consumed about 800 to 1000 grams of liquor for the past 15 years was admitted initially for evaluation of recurrent falls and new-onset abdominal pain over the last few weeks.

Further workup revealed leukocytosis, macrocytic anemia with normal vitamin B12 and folate levels, thrombocytopenia, transaminitis, hypoalbuminemia, severe hyponatremia, and mild hyperammonemia, and normal coagulation studies. CT head without contrast was normal on admission.

He was initially diagnosed and managed for acute alcoholic hepatitis. The hospital course was complicated with altered mental status, which progressed to a coma in two weeks. He was treated for hepatic encephalopathy, correction of hyponatremia, alcohol withdrawal, injection of thiamine, and empiric coverage with antibiotics for possible spontaneous bacterial peritonitis (SBP). SBP workup was normal.

MRI of the brain without contrast was performed as the patient did not improve on earlier management. MRI revealed an increased FLAIR signal in the inferior corpus callosum and mild restricted diffusion in the corpus callosum's genu and splenium, consistent with Marchiafava-Bignami disease (Figure 1).

The patient's clinical condition deteriorated, and he subsequently passed away after the family proceeded to honor his wishes for hospice care.

IMAGING

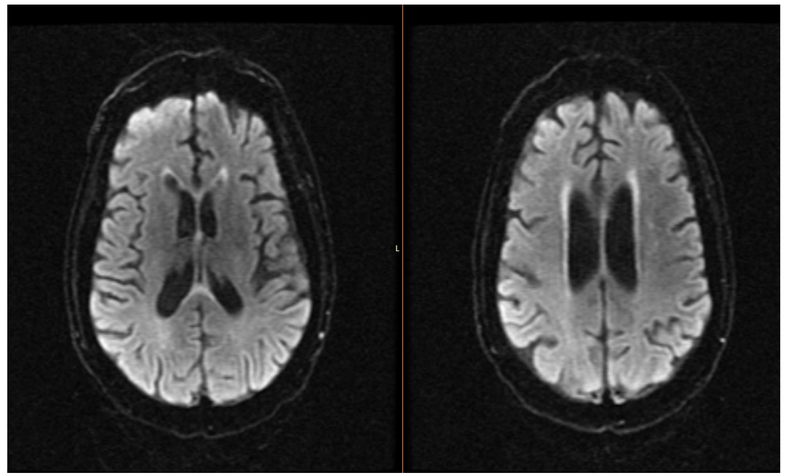

Figure 1. MRI revealed an increased FLAIR signal in the inferior corpus callosum and mild restricted diffusion in the corpus callosum's genu and splenium

REFERENCES

1. Hillbom M, Saloheimo P, Fujioka S, Wszolek ZK, Juvela S, Leone MA. Diagnosis and management of Marchiafava-Bignami disease: a review of CT/MRI confirmed cases. J Neurol Neurosurg Psychiatry. 2014 Feb;85(2):168-73.

2.]Wenz H, Eisele P, Artemis D, Förster A, Brockmann MA. Acute Marchiafava-Bignami disease with extensive diffusion restriction and early recovery: case report and review of the literature. J Neuroimaging. 2014 Jul-Aug;24(4):421-4.

3. .Matsuura H, Shindo K. Marchiafava-Bignami disease. QJM. 2018 Oct 01;111(10):755.

4. Hoshino Y, Ueno Y, Shimura H, Miyamoto N, Watanabe M, Hattori N, Urabe T. Marchiafava-Bignami disease mimics motor neuron disease: case report. BMC Neurol. 2013 Dec 21;13:208.

5. Hampel H, Teipel SJ, Alexander GE, Horwitz B, Teichberg D, Schapiro MB, Rapoport SI. Corpus callosum atrophy is a possible indicator of region- and cell type-specific neuronal degeneration in Alzheimer disease: a magnetic resonance imaging analysis. Arch Neurol. 1998 Feb;55(2):193-8.

DISCUSSION

Marchiafava–Bignami disease (MBD) or syndrome is a rare progressive neurological disease characterized by selective demyelination of the corpus callosum and cortical laminar necrosis involving the frontal or temporal lobes, subsequently resulting in atrophy.

The most important factor underlying its appearance is chronic and severe nutritional depletion.

MRI typically shows hyperintense lesions on T2-phase and FLAIR images, indicating edema and demyelination, with hypointense lesions on T1WI of the corpus callosum and sometimes even the genu and the splenium, signifying bleeding and hemosiderin deposits.

The presentations of MBD are nonspecific and include dementia, altered mental status, spasticity, dysarthria, ataxia, gait abnormalities, seizures, and coma.

Patients who develop coma carry the most significant risk of severe disability or death, whereas those with minor impairment in consciousness may recover with only mild disability.

This is a rare disorder that is difficult to diagnose as there are other possible etiologies; it is, therefore, imperative that gastroenterologists and hepatologists are familiar with the diagnostic and therapeutic approaches to this disorder and involvement of the multispecialty team early on.