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Case Report

Marchiafava–Bignami disease in chronic alcoholic patient

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ABSTRACT

Marchiafava–Bignami disease is a rare toxic encephalopathy seen mostly in chronic alcoholics due to progressive demyelination and necrosis of the corpus callosum. It may involve adjacent white matter and subcortical regions. We present here the magnetic resonance imaging findings of Machiafava–Bignami disease in a chronic alcoholic patient. In 1903, Italian pathologists Marchiafava and Bignami described 3 alcoholic men who died after having seizures and coma. All 3 patients were chronic alcoholics and had consumed considerable amounts of red wine.

History

A 45-year-old chronic alcoholic patient who was taking alcohol for 20 years and presented with altered sensorium and seizures.

Patient's family reported that he had consumed a daily average of 250 mL of liquor for 2 weeks. The patient was in poor physical condition and seemed malnourished. The patient is diagnosed case of liver cirrhosis since 5 years. On examination, the patient was confused. Although there was no weakness in his extremities, he demonstrated lack of motor coordination. Electroencephalogram showed diffuse slow waves of 5-8 Hz without epileptiform discharge.

Laboratory test results revealed hyperosmosis of 370 mmol/L (normal, 290 mmol/L), high blood creatinine (2.1 mg/dL; normal, 0.5-1.2 mg/dL), and high blood urea nitrogen (40.0 mg/dL; normal, 8-23 mg/dL). Sodium and potassium levels were, respectively, 146 mmol/L and 4.3 mmol/L. Cerebrospinal fluid studies revealed no abnormalities. There are low serum levels of vitamin B12, folic acid, and albumin seen.

Imaging findings

Cranial computed tomography showed diffuse corpus callosal and bilateral periventricular hypodensity.

Protocol

Magnetic resonance imaging (MRI) was performed on a 1.5 T magnet (GE Signa, 1.5 T, General Electric). Fast spin-echo T1-weighted (repetition time [TR]: 650 ms, echo time [TE]: 14 ms), T2-weighted (dual-echo TR, 2014 ms; TE, 30 and 100 ms), and fluid attenuation and inversion recovery (FLAIR; Turbo Spin Echo; turbo factor 11; TR, 5496 ms; TE, 100 ms; inversion time, 2000 ms) images in axial and sagittal planes were acquired. Diffusion-weighted imaging (DWI) scans were acquired with diffusion gradients along each of the 3 principal axes with 3 different values (0, 187, and 757 s/mm^2). Postcontrast T1-weighted (TR, 650 ms; TE, 14 ms) images were acquired after intravenous administration of 0.2 mL/kg body weight of

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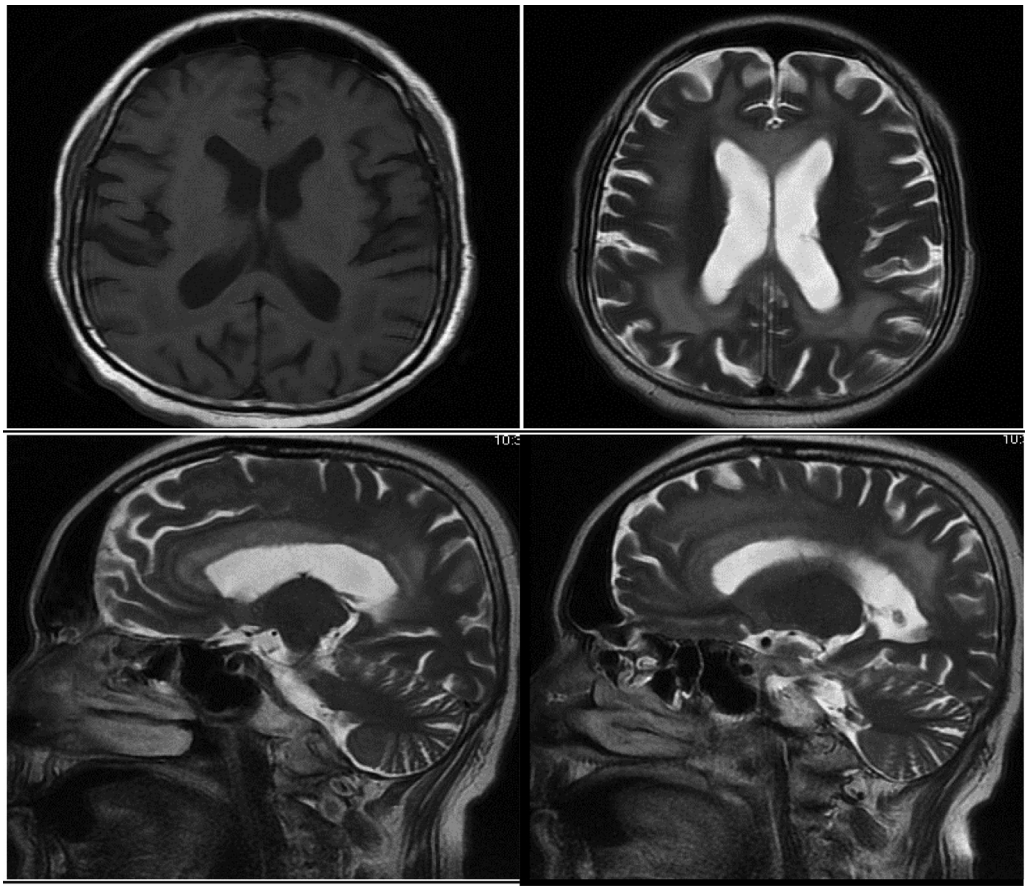

Fig. 1 – In a 45-year-old chronic alcoholic patient presented with seizures, altered sensorium, and bilateral lower limb paresis, T1W (upper row right) image shows hypointense corpus callosum and periventricular white matter with corresponding T2 (upper row left and middle row) and FLAIR (lower row right images shows hyperintense signal with true restricted diffusion in DWI image (lower row left). There is no postcontrast enhancement seen (last image).

gadodiamide (Omniscan) at a rate of 4 mL/s with a delay of 10 minutes in the axial and sagittal planes.

Figure 1 shows on T2-weighted and FLAIR images show diffuse hyperintensity in corpus callosum and bilateral periventricular white matter with corresponding T1-hypointense signal. On DWI images restricted diffusion seen in bilateral peritrigonal white matter and middle layer of splenium of corpus callosum with corresponding drop in signal on apparent diffusion coefficient seen. There is no evidence of increased abnormal signal in the subcortical white matter seen. There is no evidence of postcontrast enhancement.

Marchiafava–Bignami disease (MBD) was diagnosed based on the clinical and imaging features.

Discussion

MBD is a rare complication of chronic alcoholism characterized by demyelinization and necrosis of the entire length and middle layer of corpus callosum with extension into hemispheric white matter [1,2]. Occasionally, other structures of the central nervous system may be involved, such as the optic chiasm and tracts, putamen, cerebellar peduncle, and anterior commissure. Cortical gray matter and subcortical *U* fibers are involved rarely [3,4].

Most accepted etiologic factor is the in multiple vitamin B deficiency [5].

Clinical manifestations [6]:

(1) Acute state: seizures, alterations of consciousness, and death may occur.
(2) Subacute state: characterized by mental confusion, behavioral disorders, memory deficits, cerebellar signs, and interhemispheric disconnection.
(3) Chronic state: mild dementia.

Clinicoradiological subtypes of MBD [6,7]:

(1) Type A: characterized by alterations of consciousness and diffuse swelling of the entire corpus callosum on imaging.
(2) Type B: mild impairment of consciousness and small callosal lesions associated with good prognosis.

On MRI images [8], T1W shows confluent hypointense signal intensity of corpus callosum.

T2 and FLAIR shows hyperintense signal in middle layer of corpus callosum (Sandwich sign).

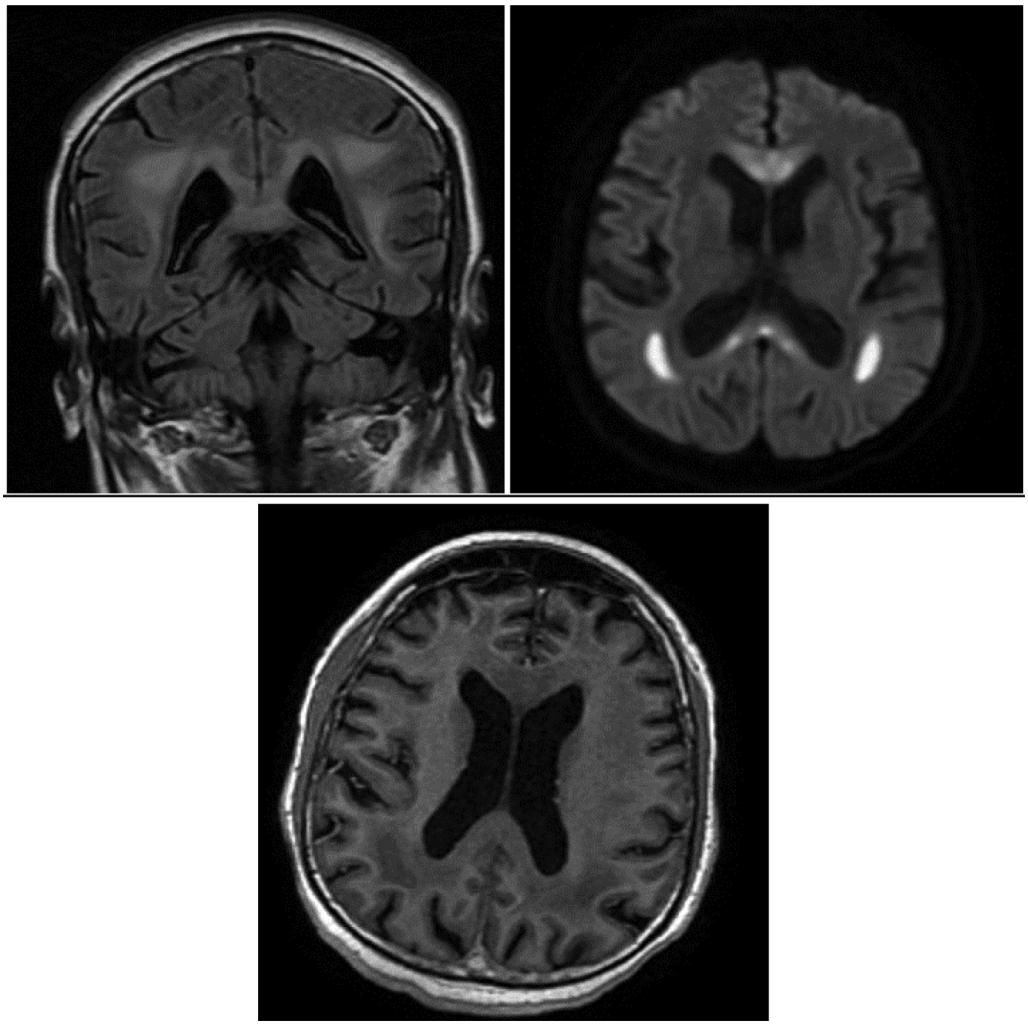

Fig. 1 – (*continued*).

T1 + C has no significant contrast enhancement.

DWI, in the acute phase, shows restricted diffusion because of cytotoxic edema caused by increase of extracellular glutamate that binds NMDA (N-methyl-D-aspartic acid) receptors inducing calcium entry and finally apoptosis without brain ischemia.

Apparent diffusion coefficient was the low apparent diffusion coefficient.

Early diagnosis and treatment can improve clinical outcome. MRI findings can help to differentiate MBD from other corpus callosal lesions and also from alcohol-related disorders.

MBD is commonly associated with other manifestations of chronic alcohol abuse such as

(1)Wernicke's encephalopathy,
(2) Central pontine myelinolysis, and
(3) Morel's laminar sclerosis.

The genu is usually the most involved structure followed by the splenium. The entire corpus callosum may also be involved. In case of chronic stage, corpus callosum degenerates and separates into 3 layers with necrotic cavities mainly in the middle layer. Cortical involvement is extremely rare and when present, it is usually localized in lateral-frontal regions. [2]

In conclusion, even if it is not possible to identify pathognomonic characteristics of MBD lesions, the clinical aspects and neuroimaging pattern may helpful for the diagnosis.

Treatment

No specific, proven treatment is available for MBD. Various treatments similar to those commonly administered for Wernicke–Korsakoff syndrome or for alcoholism in general have been given to patients with MBD.

Vitamins

The most commonly given treatments are B complex vitamins including thiamine, folate, and other B vitamins (especially vitamin B-12). Folate is commonly given with B-12.

Because thiamine deficiency is associated with malnutrition and prolonged vomiting in alcoholics, MBD patients with these symptoms may effective from parenteral thiamine administered within 15 days of symptom onset. [9]

Antiparkinsons drugs

With regard to more unusual treatments, a case report by Staszewski et al [10] described amantadine given together with thiamine, vitamin B-12, and folate, the patient improved.

Immunosuppressant therapy

In another case, reported by Kikkawa et al [11], administration of high-dose corticosteroids was said to precede clinical improvement. In patients who improved, the computed tomography and MRI scan findings also improved, at least somewhat.

Patients are usually admitted because they present with stupor, coma, and, frequently, seizures.

Patients who survive should receive rehabilitation and, if appropriate, alcohol and nutritional counseling.

In our patient was given high-dose vitamin B complex, injectable optineuron including 500 mg/d thiamine intravenously for 10-15 days. Patient showed gradual clinical improvement.

Differential diagnosis

To differentiate MBD lesions to other possible causes of callosal damage, we have to consider their specific localization inside the corpus callosum.

Wernicke's encephalopathy is characterized by lesions involving midline structures such as medial thalami, hypothalami, mammillary bodies, and periaqueductal gray matter. In a few cases of Wernicke's encephalopathy, cortical abnormalities are unusually restricted to the motor and premotor cortices. [12] Our patient's MR images revealed symmetrical hyperintensity in the bilateral periventricular areas. At the same time, our patient did not present ophthalmoplegia, nystagmus, or ataxia and did not show MR findings typical of Wernicke's encephalopathy, such as symmetrical hyperintense lesions surrounding the third ventricle and aqueduct.

Other differentials

Corpus callosum glioma: Produces enlargement of the corpus callosum with mass effect.

Demyelination: lesions are usually discrete and shows perpendicular arrangement called Dawson's fingers.

Vascular lesions: lesions are usually more discrete in distribution.

Final diagnosis: MBD type A in chronic phase.

Teaching point

MBD seen in chronic alcoholic patient is due to B complex deficiency On neuroimaging, it shows corpus callosal and periventricular white matter involvement.

REFERENCES

[1] Friese SA, Bitzer M, Freudenstein D, Voig K, Kuker W. Classification of acquired lesions of the corpus callosum with MRI. Neuroradiology 2000;42:795–802.

[2] Johkura K, Naito M, Naka T. Cortical involvement in Marchiafava–Bignami disease. AJNR Am J Neuroradiol 2005;26:670–3.

[3] Lechevalier B, Andersson JC, Morin P. Hemispheric disconnection syndrome with a 'crossed avoiding' reaction in a case of Marchiafava–Bignami disease. J Neurol Neurosurg Psychiatry 1977;40:483–97.

[4] Ellison D, Love S, Chimelli L, Harding BN, Lowe J, Vinters HV. A reference text of CNS pathology. 2nd ed. Philadelphia: Mosby; 2004. p. 489–90. Neuropathology.

[5] Arbelaez A, Pajon A, Castillo M. Acute Marchiafava–Bignami Disease: MR findings in two patients. AJNR Am J Neuroradiol 2003;24:1955–7.

[6] Heinrich A, Runge U, Khaw AV. Clinicoradiologic subtypes of Marchiafava–Bignami disease. J Neurol 2004;251:1050–9.

[7] Gambini A, Falini A, Moiola L, Comi G, Scotti G. Marchiafava–Bignami disease: longitudinal MR imaging and MR spectroscopy study. AJNR Am J Neuroradiol 2003;24:249–53.

[8] Gallucci M, Limbucci N, Paonessa A, Caranci F. Reversible focal splenial lesions. Neuroradiology 2007;49:541–4.

[9] 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.

[10] Staszewski J, Macek K, Stepien A. Reversible demyelinisation of corpus callosum in the course of Marchiafava–Bignami disease. Neurol Neurochir Pol 2006;40(2):156–61.

[11] Kikkawa Y, Takaya Y, Niwa N. A case of Marchiafava–Bignami disease that responded to high-dose intravenous corticosteroid administration. Rinsho Shinkeigaku 2000;40(11):1122–5.

[12] Yamashita M, Yamamoto T. Wernicke encephalopathy with symmetric pericentral involvement : MR findings. J Comput Assist Tomogr 1995;19:306–8.