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CASE REPORT

A patient with Marchiafava–Bignami disease type A transported by ambulance with impaired consciousness and malnutrition was successfully treated after early diagnosis by MRI

Yoshitaka Ooya M.D., Ph.D.[1] | Mayumi Ishida C.P., Ph.D.[2] | Nozomu Uchida M.D.[2,3] | Ryota Sato M.S.[2,4] | Akira Yoshioka M.D., Ph.D.[5] | Izumi Sato MPH, Ph.D.[6] | Hideki Onishi M.D., Ph.D.[2]

[1]Departments of Emergency and Acute Medicine, Saitama Medical University International Medical Center, Saitama, Japan

[2]Department of Psycho-oncology, Saitama Medical University International Medical Center, Saitama, Japan

[3]Department of General Medicine, Ogano Town Central Hospital, Saitama, Japan

[4]Department of Pharmacy, Maruki Memorial Medical and Social Welfare Center, Saitama, Japan

[5]Department of Medical Oncology and Palliative Care, Mitsubishi Kyoto Hospital, Kyoto, Japan

[6]Department of Clinical Epidemiology, Graduate School of Biomedical Sciences, Nagasaki University, Nagasaki, Japan

Correspondence
Hideki Onishi, Department of Psycho-oncology, Saitama Medical University International Medical Center, 1397-1 Yamane, Hidaka City, Saitama 350-1298, Japan.
Email: honishi@saitama-med.ac.jp

Abstract

Marchiafava–Bignami disease (MBD) is a very rare disorder characterized by demyelination and necrosis of the corpus callosum.

A 53-year-old male was transported to the emergency room with impaired consciousness. On his arrival, he was quite emaciated. CT examination revealed no hemorrhagic lesions whereas MR images showed hyperintense areas throughout the corpus callosum, leading to a diagnosis of MBD. His impaired consciousness improved with treatment, including parenteral thiamine administration.

When examining patients with impaired consciousness because of malnutrition, MBD should be taken in consideration and the incorporation of head MR imaging into the examination protocol enables early diagnosis and treatment, and may improve the prognosis.

KEYWORDS
Emergency Medicine, Neurology, Marchiafava-Bignami disease, MRI

1 | INTRODUCTION

Marchiafava–Bignami disease (MBD) is a very rare disorder characterized by demyelination and necrosis of the corpus callosum.

The clinical manifestations are diverse and not disease specific. Although previously a fatal disease, advances in neuroradiology have enabled early diagnosis and reduced mortality.[1]

The disease can be categorized into two types; type A and B, which have entire and partial findings in the corpus callosum, respectively.[1]

Disease onset is reported to occur mainly in people with poor nutritional status and heavy alcohol consumption; however, alcohol is not the sole cause as some cases have occurred in nonalcoholic patients.[2,3]

As mentioned above, MBD is caused by a combination of factors; however, as it is a rare disorder of unknown pathophysiology, the addition of knowledge regarding its clinical and neuroradiological findings toward future clarification of its pathophysiology is essential.

Herein, we report a case in which a patient transported to an emergency center with impaired consciousness. He was diagnosed with MBD type A based on clinical symptoms, blood biochemistry, and neuroradiological examination, and successfully treated.

2 | CASE REPORT

A 53-year-old man was found unconsciousness at home by a visiting friend and taken to an emergency center. According to the information received from the ambulance service, he was unemployed and had no relatives. His room was littered with garbage, but there was no reason to suspect heavy drinking or heavy drug use. The patient was able to live alone until 2 days prior to hospitalization.

TABLE 1 Laboratory findings.

	Day 0	Day 7	Day 17	Reference range	Units
TP	7.4	5.7	6.6	6.6–8.1	g/dL
ALB	4.2	3.0	3.5	4.1–5.1	g/dL
CK	2758	853	80	59–248	U/L
AST	120	87	43	13–30	U/L
ALT	63	34	23	10–42	U/L
LDH	380	333	258	124–222	U/L
ALP	206	136	140	38–113	U/L
γGTP	495	249	126	13–64	U/L
ChE	110	86	133	240–486	U/L
Cr	3.20	0.71	0.88	0.65–1.07	mg/dL
eGFR	17.4	90.3	71.4		
UA	17.9	3.5	6.8	3.7–7.8	mg/dL
BUN	44.1	8.2	10.6	8–20	mg/dL
Ammonia	<10	na	na	12–66	μg/dL
Alcohol	<5	na	na	0.0–5.0	mg/dL
Blood glucose	225	na	na	73–109	mg/dL
Na	141	143	141	138–145	nmol/L
Cl	92	102	103	101–108	nmol/L
K	4.5	4.2	4.1	3.6–4.8	nmol/L
Mg	1.9	na	na	1.8–2.4	mg/dL
Ca	8.3	na	na	8.8–10.1	mg/dL
IP	6.7	na	na	2.7–4.6	mg/dL
T-Bil	0.9	0.5	0.4	0.4–1.5	mg/dL
CRP	17.49	3.12	0.32	0–0.14	mg/dL
WBC	17,560	4830	4270	3300–8600	/μL
RBC	3.57	2.72	3.08	4.35–5.55	$\times 10^6$/μL
Hb	14.7	10.7	11.5	13.7–16.8	g/dL
HCT	40	31.1	34.5	40.7–50.1	%
MCV	112	114.3	112	83.6–98.2	fL
MCH	41.2	39.3	37.3	27.5–33.2	pg
MCHC	36.8	34.4	33.3	31.7–35.3	g/dL
Plt	176	229	266	158–348	$\times 10^3$/μL
Vitamin B1	28	na	na	24–66	ng/dL
Free T3	1.1	na	na	2.3–4.0	pg/mL
Free T4	2.3	na	na	0.5–5.0	μIU/mL
TSH	0.8	na	na	0.9–1.7	ng/dL

Abbreviation: na, not available.

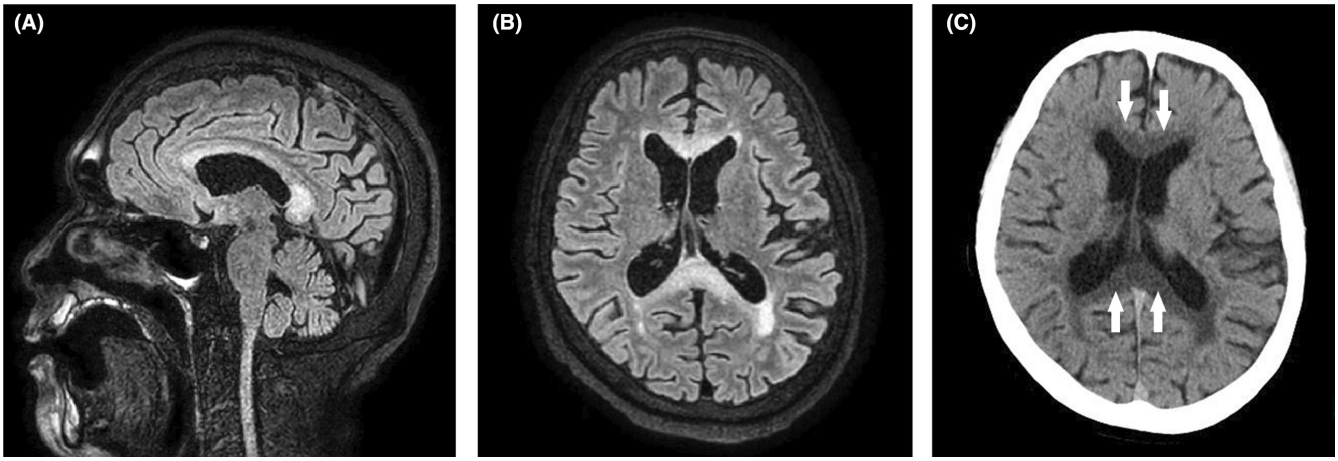

FIGURE 1 Patient MRI and CT findings. Sagittal (A) and axial (B) FLAIR images showing areas of hyperintensity throughout the corpus callosum. (C) Axial head CT images showing corpus callosum hypodensity (arrowhead)

His medical history included anxiety, insomnia, alcoholism, and pancreatitis.

At the time of admission, he was 166 cm tall, and had a BMI of 13.4, showing marked emaciation.

When he was transferred to the hospital, he was able to open his eyes voluntarily, but was indifferent to his surroundings and had a poor understanding of the situation. His level of consciousness was E4V1M6 according to the Glasgow Coma Scale (GCS), blood pressure 156/99 mmHg, pulse rate 95/min, respiratory rate 24/min, and oxygen saturation (SpO_2) 100% (room air). Neurologically, no paralysis of the limbs, muscle rigidity, nystagmus, or oculomotor disturbance were observed.

Bilateral pupils 6.0/6.0 mm with no light response. Pressure ulcers were observed on the right bulbar conjunctival hyperemia, right cheek, mandible, anterior chest, bilateral iliac regions, and both knees.

CT scans of the head were taken to identify the cause of the impaired consciousness, but no hemorrhagic lesions were found. Blood and biochemical findings included prerenal renal failure due to severe dehydration and rhabdomyolysis. Alcohol, ammonia, and thyroid function were normal (Table 1). After blood collection, an infusion containing thiamine was started.

On the same day, brain MR images taken after the CT scans demonstrated hyperintensity on fluid-attenuated inversion recovery (FLAIR) T2 sequences in throughout the corpus callosum (Figure 1A,B). A low-density area was also observed in the corpus callosum when the CT scans of the head were reviewed based on the MRI findings (Figure 1C).

Based on the above clinical and neuroradiographic findings, MBD type A was diagnosed, and fluid replacement and intravenous high-dose thiamine therapy (200 mg three times daily)[4] were immediately started from Day 0 to 3, and oral administration of 150 mg/day of vitamin B1, 150 mg/day of vitamin B6, and 1500 μg/day of vitamin B12 was started from Day 4 of hospitalization.

Tube feeding was started on Day 1 of hospitalization, and the patient's level of consciousness level fluctuated around E4V3M5 (GCS). On Day 5 of hospitalization, the patient's level of consciousness was E4V4M6 (GCS), and a certain degree of spontaneity was observed. No symptoms of carpus callosum disconnection syndromes were observed.

His thiamine level, as measured using high-performance liquid chromatography, was found at 28 ng/mL (reference range: 24–66 ng/mL).

Thereafter, the patient's level of consciousness level continued at E4V4M6 (GCS), and his general condition stabilized and remained almost constant by Day 8 of hospitalization. Dysphagia tended to improve from Day 7, and was recovered on Day 19. In terms of ADL, he was able to transfer to a wheelchair on Day 10, walk with assistance on Day 13, and walk alone on Day 20.

He was transferred on Day 28 of hospitalization. After spending about a month in a rehabilitation hospital, the patient returned home and now lives alone once more.

3 | DISCUSSION

We experienced a case in which a patient was transported to an emergency center with impaired consciousness and thereafter diagnosed with MBD type A based on living conditions at home, blood biochemical findings, and diagnostic imaging. The symptoms improved with treatment, including parenteral thiamine administration.

The initial basis for the diagnosis was the MRI findings. In the case of impaired consciousness that cannot be differentially diagnosed on the basis of CT scans, MRI may, as in this case, reveal lesions in the central nervous system. Thus, MRI is considered essential for diagnosis and establishment of treatment strategies, especially when malnutrition is present as a complication.

Wernicke encephalopathy was first suspected, and intravenous thiamine was administered immediately after blood sampling.[5] Subsequent MR images revealed MBD. Intravenous administration of thiamine within 2 weeks after the onset of symptoms is a prognostic factor for MBD.[2] Therefore, the patient could return to daily life. No symptoms of carpus callosum disconnection syndrome were observed; however, this is probably due to the effect of the early treatment.

There are certain limitations to this report.

First, no MRI findings were obtained after the treatment. It has been reported that MRI findings improve after improvements in neuropsychiatric symptoms.[6] A serial image search should have been performed to further clarify the pathology.

Second, vitamin B6, B12, and folic acid were not measured, so it was not possible to ascertain abnormalities in these vitamins. It is necessary to measure these vitamins in order to elucidate the pathology of MBD.

In conclusion, we diagnosed MBD in a patient with impaired consciousness accompanied by malnutrition. When examining patients who are malnourished and exhibit impaired consciousness, despite MBD being a rare disorder, the incorporation of head MRI to the examination protocol may allow early diagnosis and treatment, as well as improve treatment prognosis.

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CONFLICT OF INTEREST STATEMENT

The authors have stated explicitly that there is no conflict of interest in connection with this article.

CONSENT FOR PUBLICATION

Written informed consent was obtained from the patient for publication of this case report.

CLINICAL TRIAL REGISTRATION

None.

ETHICS APPROVAL STATEMENT

Our institution does not require approval of institutional ethics committee for case reports.

ORCID

Nozomu Uchida https://orcid.org/0000-0002-2357-9823
Ryota Sato https://orcid.org/0009-0001-1347-6337
Hideki Onishi https://orcid.org/0000-0003-2949-2734

REFERENCES

1. Heinrich A, Runge U, Khaw A. Clinicoradiologic subtypes of Marchiafava-Bignami disease. J Neurol. 2004;251(9):1050–9.
2. 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;85(2):168–73.
3. Bachar M, Fatakhov E, Banerjee C, Todnem N. Rapidly resolving nonalcoholic Marchiafava-Bignami disease in the setting of malnourishment after gastric bypass surgery. J Investig Med High Impact Case Rep. 2018;6:232470961878431.
4. Galvin R, Brathen G, Ivashynka A, Hillbom M, Tanasescu R, Leone MA. EFNS guidelines for diagnosis, therapy and prevention of Wernicke encephalopathy. Eur J Neurol. 2010;17(12):1408–18.
5. Sechi G, Serra A. Wernicke's encephalopathy: new clinical settings and recent advances in diagnosis and management. Lancet Neurol. 2007;6(5):442–55.
6. Hoshino Y, Ueno Y, Shimura H, Miyamoto N, Watanabe M, Hattori N, et al. Marchiafava-Bignami disease mimics motor neuron disease: case report. BMC Neurol. 2013;13(1):208.

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