Scholars Journal of Medical Case Reports **ISSN 2347-6559 (Online)**
Sch J Med Case Rep 2017; 5(10):639-641 **ISSN 2347-9507 (Print)**

(An International Publisher for Academic and Scientific Resources)

Marchiafava-Bignami disease induced by chronic alcohol intake

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Article History
Received: 09.10.2017
Accepted: 15.10.2017
Published:30.10.2017

DOI:
10.36347/sjmcr.2017.v05i10.011

Abstract: A 42-year-old male admitted to a local psychiatric facility fell unconscious, and his oxygen saturation decreased to 40% on the first hospital day. The medical staff could not respond to this situation and called an ambulance. A doctor car was dispatched and docked with the ambulance. As the patient remained in a deep coma state, he underwent tracheal intubation at the rendezvous point. He had a history of drinking 360 g of ethanol per day. On arrival, he received infusion of vitamin B1. On a physical examination, both eye balls were in the abductor position. A urinary qualitative test for drugs was negative. The level of vitamin B1 was 24.0 (normal range: 21.3-81.9) ng/ml. Whole-body computed tomography (CT) revealed low density at the splenial area, left dorsal consolidation at the left lung and fatty liver changes. Magnetic resonance imaging immediately after CT revealed an isolated splenial lesion. He received a diagnosis of Marchiafava-Bignami disease (MBD), alcoholic hepatitis, alcoholic rhabdomyolysis and aspiration pneumonia. After admission to the intensive-care unit, he underwent infusion of thiamine and antibiotics. On the third hospital day, his respiratory function and consciousness recovered, an extubation was performed. At that time, he had disorientation and dysarthria. After regaining his ability to feed himself, he was transferred to the previous psychiatric facility. He ingested vitamin B1 until transportation. We herein report a rare case of MBD diagnosed based on MRI findings and the patient's history of alcohol consumption. Immediate and continuous infusion of thiamine is important for MBD even the level of vitamin B1 is the normal range.
Keywords: Marchiafava-Bignami disease; thiamine; splenium

INTRODUCTION

Marchiafava-Bignami disease (MBD) is a rare disease characterized by the demyelination and necrosis of the corpus callosum. It is typically associated with chronic alcohol intake and clinically demonstrates various symptoms and signs [1, 2]. Several cases with malnutrition or prolonged vomiting without alcohol association have also been reported [1, 2]. MBD was initially described in 1903 by the Italian pathologists Marchiafava and Bignami [3]. They observed three alcoholic men who had died after having seizures and falling into a coma. All three patients were chronic alcoholics, had consumed considerable amounts of red wine, and were characterized by demyelination and necrosis of the corpus callosum. The actual mechanisms leading to callosal damage in MBD and the clinical conditions that mimic it are still unclear [2]. We herein report a case of MBD induced by chronic alcohol intake.

CASE REPORT

A 42-year-old male admitted to a local psychiatric facility fell unconscious, and his oxygen saturation decreased to 40% on the first hospital day. The medical staff could not respond to this situation and called an ambulance. Since the physician-staffed helicopter could not make the flight, a doctor car was dispatched and docked with the ambulance. As the patient remained in a deep coma state, he underwent tracheal intubation at the rendezvous point.

He had a history of drinking 360 g of ethanol per day. On arrival, his Glasgow Coma Scale score was E1VTM1. He had a blood pressure of 104/80 mmHg, a heart rate of 104 beats per minute (BPM), an SpO_2 of 99% under 100% oxygen and a body temperature of 36.5 °C. Immediately after the blood examination, he received infusion of vitamin B1. On a physical examination, both eye balls were in the abductor position. An arterial gas analysis under 100% oxygen revealed a pH of 7.26, PCO_2 of 54.2 mmHg, PO_2 of 209 mmHg, HCO_3^- of 23.7 mmol/l and a base excess level of -3.2 mmol/l. Electrocardiogram revealed sinus tachycardia, and the heart wall motion was normal according to an ultrasound study. A urinary qualitative test for drugs was negative. The main results of a biochemical analysis of the blood were as follows: white blood cell count of 13,700/μl, hemoglobin level of 10.5 g/dl, platelet count of 23.2×10^4/μl, total protein level of 5.7 g/dl, glucose level of 215 mg/dl, aspartate

aminotransferase level of 95 IU/L, alanine aminotransferase level of 60 IU/L, gamma-glutamyltransferase level of 113 IU/L, blood urea nitrogen level of 24.7 mg/dl, creatinine level of 0.93 mg/dl, creatinine phosphokinase level of 2196 IU/l, c-reactive protein level of 9.2 mg/dl, activated partial thromboplastin time of 26.4 (27.0) seconds and international normalized ratio of prothrombin time of 0.93. The level of vitamin B1 was 24.0 (normal range: 21.3-81.9) ng/ml. Whole-body computed tomography (CT) revealed low density at the splenial area, left dorsal consolidation at the left lung and fatty liver changes (Figure 1). Magnetic resonance imaging (MRI) immediately after CT revealed an isolated splenial lesion (Figure 2). He received a diagnosis of MBD, alcoholic hepatitis, alcoholic rhabdomyolysis and aspiration pneumonia. After admission to the intensive-care unit, he underwent infusion of thiamine and antibiotics. On the third hospital day, his respiratory function and consciousness recovered, an extubation was performed. At that time, he had disorientation and dysarthria. After regaining his ability to feed himself, he was transferred to the previous psychiatric facility. He ingested vitamin B1 until transportation.

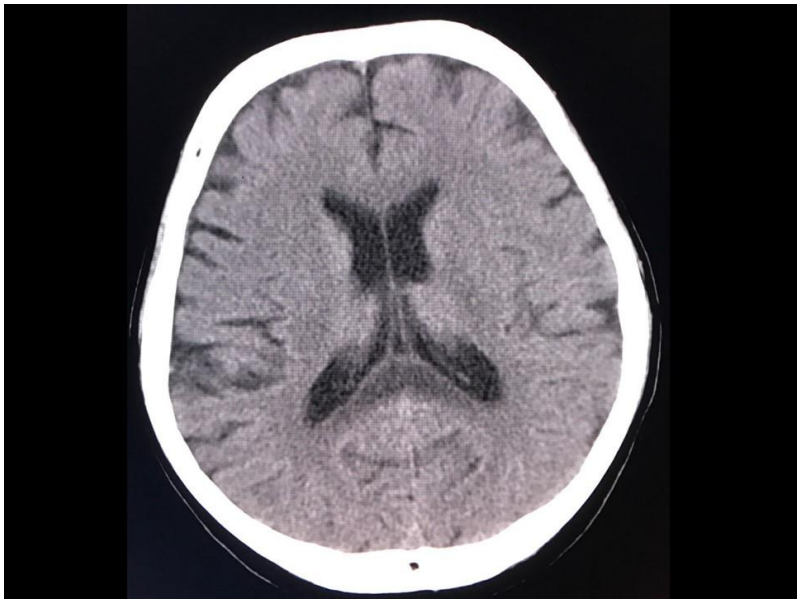

Fig-1: **Head computed tomography (CT) findings on arrival. CT reveals low density at the splenial area.**

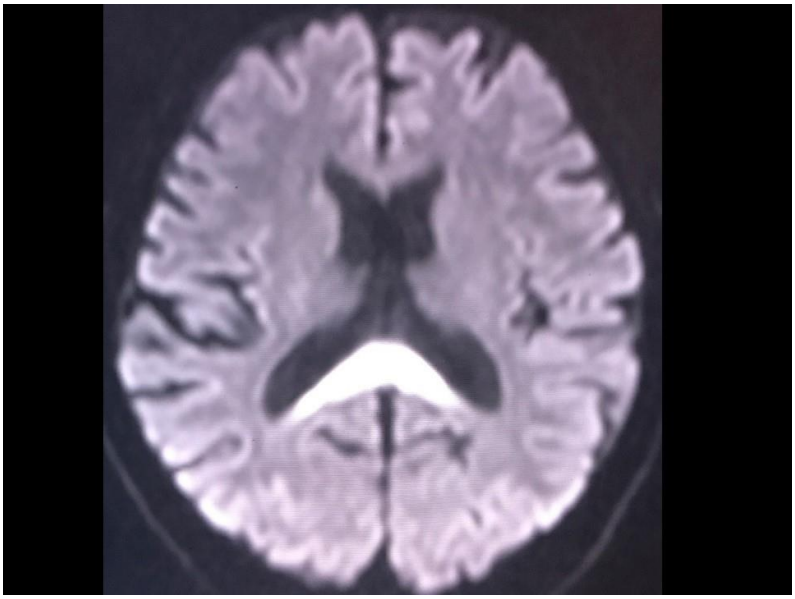

Fig-2: **Magnetic resonance imaging (MRI) findings on arrival. MRI reveals an isolated splenial lesion.**

DISCUSSION

Concerning the splenial lesions detected by MRI, Wilson et al. examined 174 cases of callosal lesions detected by diffusion-weighted MRI [5]. A total of 47% of these lesions were vascular, and 53% were nonvascular. Among the vascular cases, atypical

mechanisms of stroke (e.g. vasculitis/vasculopathy, hypercoagulable state) were most common (37%), followed by cardioembolism (28%). The vascular splenial lesions were likely due to atypical causes of stroke. The most common nonvascular etiologies were trauma (44%), tumor (22%), and demyelination (15%) with causes other than MBD. In addition, the etiology of nonvascular, non-trauma and non-tumor are mainly reported to be mild encephalitis/encephalopathy with reversible splenial lesion (MERS) and reversible splenial lesion syndrome induced by epilepsy, antiepileptic drug withdrawal, hypoglycaemia, high-altitude sickness or systemic lupus erythematosus. The present case did not have a history of trauma, convulsion, hypoglycemia or drug use. MR angiography denied the possibility of vasculopathy, and electrocardiogram did not show dysrhythmia. Accordingly, due to the alcoholism of the present case, MBD was deemed the most likely cause of his unconsciousness.

Patients with MBD may show severe signs and symptoms, including an altered mental state, impaired walking, deficient memory, dysarthria, pyramidal signs and disconnection lasting for several weeks [2]. Split brain syndrome has been reported as a characteristic feature[7], but it has lost its relevance as a diagnostic finding since callosal lesions are now detected by modern brain imaging, enabling a diagnosis in the ER setting among confused subjects, similar to the present case.

Hillbom reviewed 122 reports containing data on 153 subjects with confirmed MBD [2]. Cerebral spinal fluid (CSF) data were available for 62 MBD subjects, showed that CSF proteins and cells were increased in 5/62 (8.1%) and 0/62 MBD patients respectively. In contrast, deep white-matter lesions and cortical lesions were frequently observed in the subjects with MBD (60/121 [49.6%] and 32/104 [30.8%], respectively). The present case showed no abnormality in the CSF or isolated splenial lesion.

Patients with MBD treated with thiamine within two weeks after the onset of symptoms have been shown to have a significantly better outcome than those with delayed treatment. The dose of thiamine should be the same as recommended for Wernicke's disease [8], and the therapy should continue for as long as recovery is on-going [2]. Accordingly, the present case underwent infusion of thiamine immediately after a blood examination for the measurement of vitamin B1, and the prescription was continued until he was transported, even after the level of vitamin B1 had returned to the normal range.

CONCLUSION

We herein report a rare case of MBD diagnosed based on MRI findings and the patient's history of alcohol consumption. The diagnosis and treatment were discussed using recent reports.

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