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Case Report

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# The First Documented Case of Marchiafava-Bignami Disease with Concurrent Bilateral Deep Vein Thrombosis

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**Abstract**

Marchiafava-Bignami Disease (MBD) presents as a rare neurological disease characterized by demyelination and necrosis in the corpus callosum. Most MBD cases are secondary to severe alcohol abuse and hypovitaminosis. MRI and other forms of neuroradiological imaging is necessary to confirm the diagnosis. We herein present an atypical case of a MBD and history of alcohol abuse who developed deep vein thrombosis in his bilateral lower extremities. This novel case represents the first documented report of MBD with concurrent DVT. Furthermore, we hypothesize that DVT may be a long-term complication associated with MBD.

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## Introduction

Marchiafava–Bignami Disease (MBD) was first reported in 1903 in Italy by Ettore Marchiafava and Amico Bignami [1]. MBD is a chronic disease that is distinguished by demyelination and necrosis in the corpus callosum. It is common in patients who abuse alcohol and is thought to stem from a vitamin B1 deficiency [2]. The mean age of onset is 45 years. MBD has a higher reported incidence in males possibly due to higher alcohol consumption compared to females [3]. MBD prevalence is low, with only about 250 cases reported between 1903 and 2001 [4].

The pathogenesis of MBD is related to ethanol and vitamin B1 deficiency. Ethanol causes hypovitaminosis (mainly vitamin B1) and generation of free radicals in the central nervous system that directly damage the corpus callosum with consequent demyelination and necrosis. Vitamin B1 is a critical cofactor for multiple enzymes involved in carbohydrates metabolism. Deficiency of vitamin B1 compromises carbohydrates metabolism in the brain leading to neurological impairment [3]. Symptoms of MBD include seizures, neurocognitive deficits, and in the most severe cases, a comatose state or death. (Figure 1) While MBD patients typically have a poor prognosis, some recover when provided with a vitamin B1 supplement [2]. (Figure 2)

Literature regarding MBD is very limited. To date, MBD with concomitant deep vein thrombosis (DVT) has never been documented. We herein present the first documented case of a 52-year-old-male with diagnosis of MBD and concurrent bilateral DVT.

## Case Report

RR is a wheelchair-bound 52-year-old male with a current medical history of alcohol abuse, tobacco abuse, and failure to thrive. He was diagnosed with Marchiafava-Bignami Disease five years ago. At the time of diagnosis, he was in a comatose state for ten days and received a percutaneous endoscopic gastrostomy (PEG) tube and tracheostomy tube. The tubes were removed after the patient regained consciousness. He was brought to the emergency room (ER) with a complaint of bilateral leg swelling and pain for three days. No reported fever, headaches, loss of vision, chest pain, shortness of breath, abdominal pain, change in bowel movement, focal neurological weakness, or sensory loss. In the ER, a doppler venous ultrasound revealed occlusive thrombus limited to the common femoral vein on the right lower extremity. Contrastingly, the thrombus affected the entire femoral-popliteal venous system on the left lower extremity. Per admission, the temperature was 98.2 F, pulse 79 beats/min, respiratory rate 18 breaths/min, blood pressure 122/82, oxygen saturation 100% on room air.

On physical examination, the patient appeared chronically ill-appearing, frail, malnourished and looked older than the stated

age. Bilateral lower extremities exhibited swelling, warmth, and decreased range of motion. Neurological examination displayed no motor or sensory deficits. Pupils were equal, round, and reactive to light bilaterally. Complete blood count and comprehensive metabolic panel were significant for macrocytic anemia, elevated aspartate transaminase (AST), and alkaline phosphatase with normal alanine transaminase (ALT). COVID-19 rapid test was negative.

The patient started on an intravenous therapeutic dose of anticoagulant which was later switched to oral anticoagulant per hematology recommendation. The patient was hemodynamically stable and was discharged home three days after admission with a plan to follow up with a hematologist. MBD has not been resolved and associated symptomatology continues to persist in the patient.

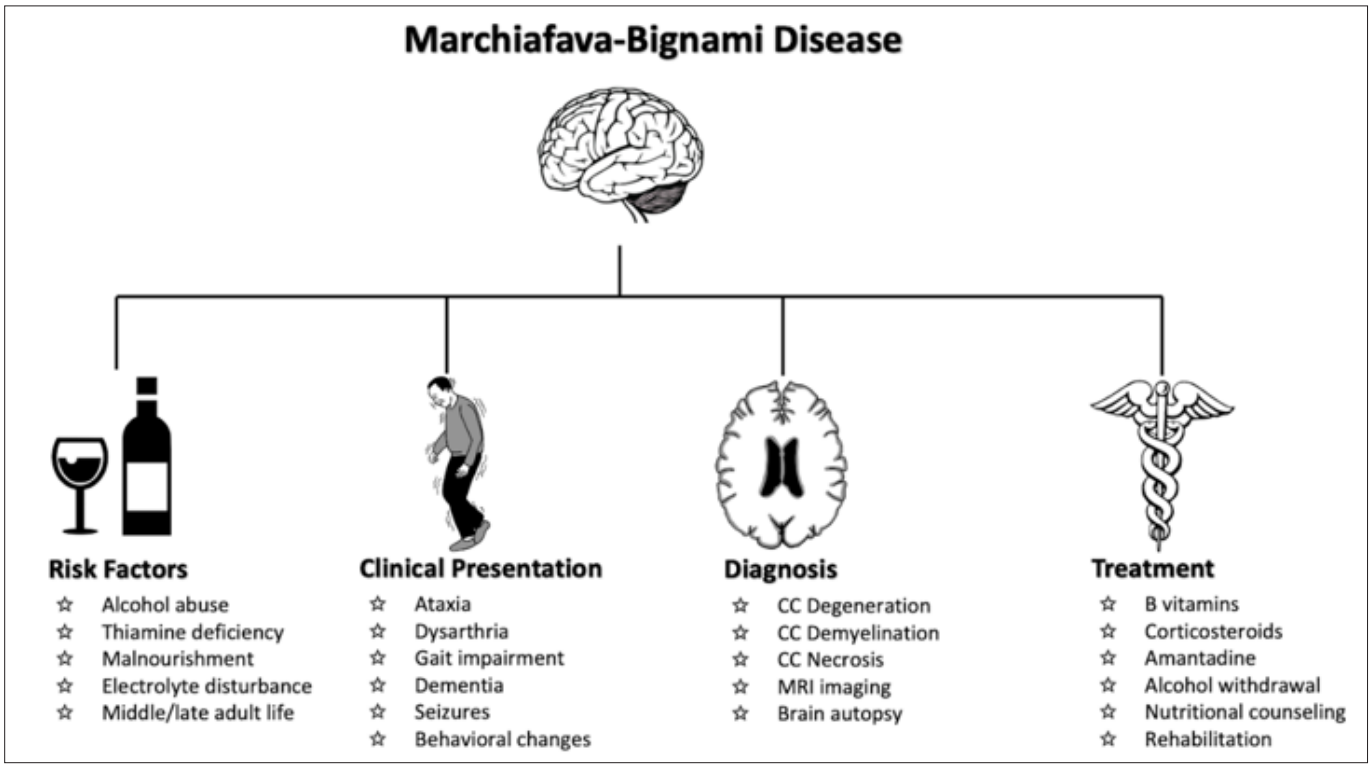


**Figure 1:** Overview of Marchiafava-Bignami Disease
CC = Corpus callosum

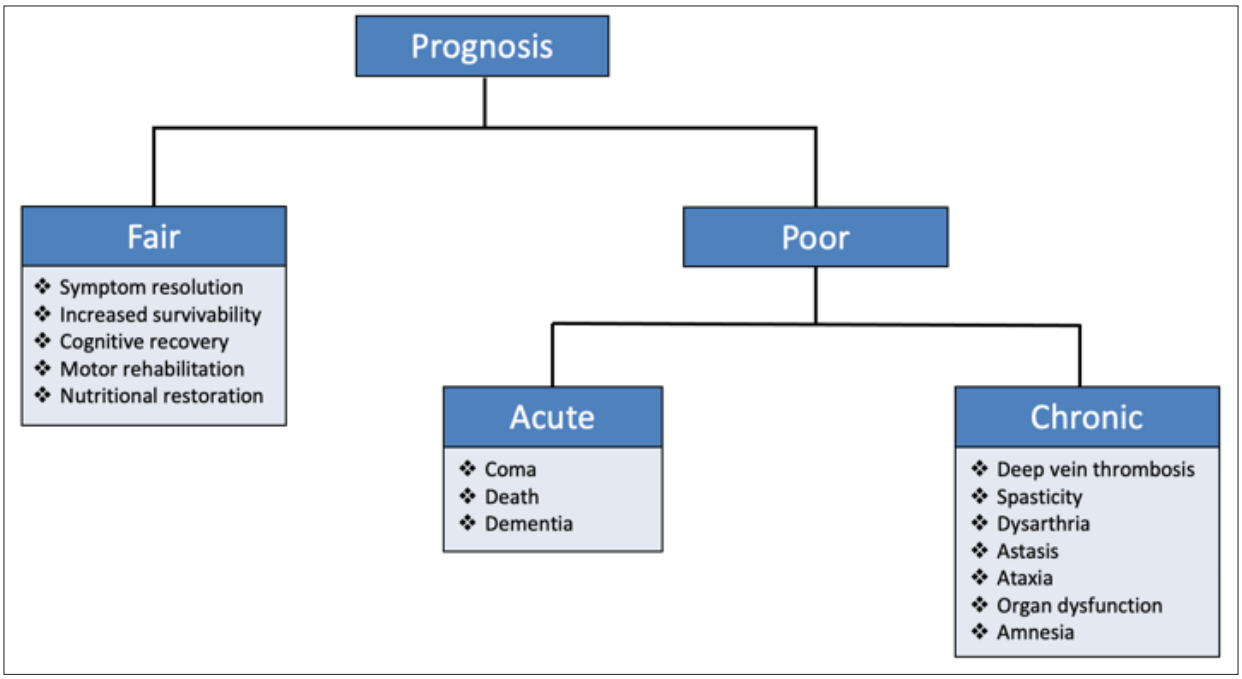


**Figure 2:** Prognosis of Marchiafava-Bignami Disease

The prognosis of MBD is variable. Some patients may be symptomatic and survive for years, recover fully, or die from end-organ damage.

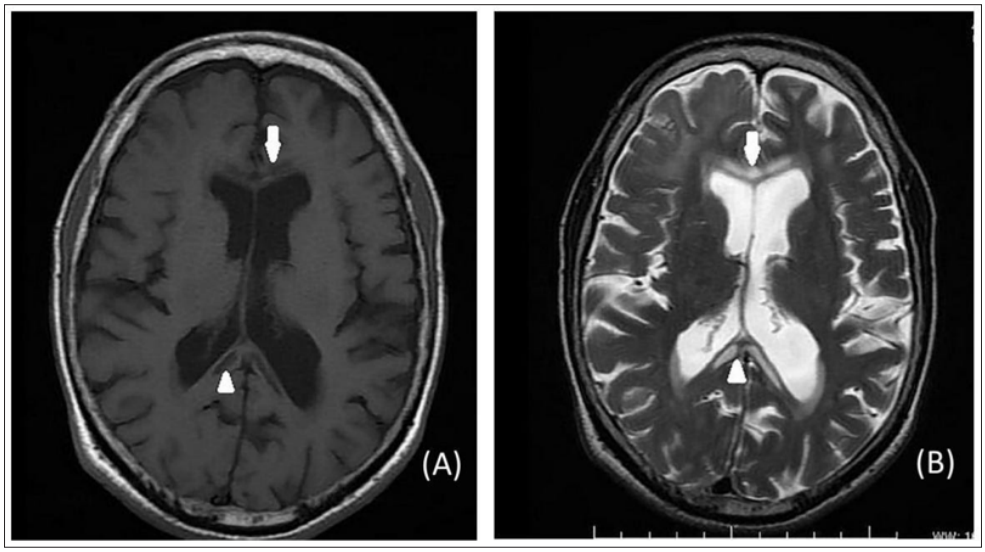


**Figure 3:** Axial images of brain MRI
**A**. T1-weighted imaging demonstrating hypointensity of the genu (white arrow) and the splenium (white arrowhead) of corpus callosum.
**B**. T2-weighted imaging showing hyperintensity of the genu (white arrow) and the splenium (white arrowhead) of corpus callosum.

The following original report is credited [5]:

**Discussion**

Marchiafava-Bignami Disease is a rare neurological complication of alcoholism characterized by targeted demyelination of the corpus callosum [6]. However, several cases of MBD have been reported in patients with poorly controlled diabetes mellitus without a history of alcohol abuse [7-9]. The precise pathomechanism of MBD remains unknown. It is postulated that the toxic effects of alcohol and vitamin B deficiency are major contributors to the necrosis and demyelination of the corpus callosum [3]. Our patient's long history of alcohol abuse and secondary nutritional deficiencies likely contributed to the pathogenesis of the disease.

Marchiafava-Bignami Disease is classified into two subtypes, according to clinical status and neuroradiological findings: [3]

1. Type A presents with acute or subacute clinical findings which may include altered level of consciousness, cognitive deterioration, dysarthria, aphasia, hypertonia, seizures, hemiparesis, hemihypoesthesia. Neuroradiologically, Type A typically is characterized by extracallosal lesions with swelling of the entire corpus callosum. Type A is typically associated with a poor prognosis. The patient's history and presentation fit the diagnostic criteria of Type A MBD.

2. Type B is a milder form than Type A. Type B manifests as a progressive and insidious onset of MBD. Patients commonly present with normal or slightly impaired levels of consciousness, gait abnormalities, and dysarthria. Compared to Type A, MRI findings in Type B reveal partial involvement of the corpus callosum, sparing the extracallosal regions in most cases. Likewise, Type B has a favorable prognosis.

Diagnosing patients with MBD based on clinical presentation may be challenging for providers. Clinical symptoms associated with Type A and Type B MBD are non-specific and may manifest in other alcohol-related conditions such as alcohol withdrawal syndrome, Wernicke's encephalopathy, and delirium. Furthermore, dementia, epilepsy, encephalitis, and vascular disturbances may also present with symptoms seen in Type A and Type B MBD [10-12]. Therefore, radio neurological imaging must be implemented to confirm MBD diagnosis. Characteristic MRI findings of the brain in patients with MBD consist of T1 hypointensity and T2 hyperintensity involving the central portion of the corpus callosum with possible extension to the genu and splenium. (Figure 3)

Currently, no management guidelines exist for the treatment of MBD. However, several case reports have shown clinical improvement with high-dose thiamine and vitamin B complex supplementation [12, 13]. Additionally, high-dose corticosteroids and amantadine have demonstrated favorable outcomes [14]. We recommend that patients with a severe history of alcohol abuse consider enrollment in alcohol rehabilitation programs and ensure optimal nutrition. Periodic blood level testing of thiamine and other B vitamins may help identify individuals at highest risk for the development of MBD. Anticoagulation therapy may be indicated as a prophylactic measure for immobile patients.

**Conclusion**

In conclusion, we present a novel case of MBD with concomitant DVT. Such a case has yet to be reported in the scientific literature. This case report underscores the clinical significance of DVT as a potential long-term complication of MBD. Additional research is needed to establish an association between MBD and DVT development. Furthermore, management guidelines for patients with MBD are recommended to improve mortality rate, prognosis, and quality of care.

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**References**

1. Marchiafava A, Bignami A (1903) Sopra un'alterazione del corpo calloso osservata in soggetti alcoolisti" Rivista di Patologia Nervosa e Mentale 8: 544-549.
2. Dong X, Bai C, Nao J (2018) Clinical and radiological features of Marchiafava-Bignami disease. Medicine (Baltimore) 97: e9626.
3. Tian TY, Pescador Ruschel MA, Park S, Liang JW (2021) Marchiafava Bignami Disease. In: StatPearls. Treasure Island (FL): StatPearls Publishing.
4. Namekawa M, Nakamura Y, Nakano I (2013) Cortical involvement in Marchiafava-Bignami disease can be a predictor of a poor prognosis: a case report and review of the literature. Intern Med 52: 811-813.
5. Paidipati Gopalkishna Murthy K (2014) Magnetic resonance imaging in marchiafava-bignami syndrome: a cornerstone in diagnosis and prognosis. Case Rep Radiol 2014: 609708.
6. Bachar M, Fatakhov E, Banerjee C, Todnem N (2018) Rapidly Resolving Nonalcoholic Marchiafava-Bignami Disease in the Setting of Malnourishment After Gastric Bypass Surgery. J Investig Med High Impact Case Rep 6: 2324709618784318.
7. Suzuki Y, Oishi M, Ogawa K, Kamei S (2012) A patient with Marchiafava-Bignami disease as a complication of diabetes mellitus treated effectively with corticosteroid. J Clin Neurosci 19: 761-762.
8. Boutboul D, Lidove O, Aguilar C, Klein I, Papo T (2010) Marchiafava-Bignami disease complicating SC hemoglobin disease and Plasmodium falciparum infection. Presse Med 39: 990-993.
9. Pérez Álvarez AI, Ramón Carbajo C, Morís de la Tassa G, Pascual Gómez J (2016) Marchiafava-Bignami disease triggered by poorly controlled diabetes mellitus. Diabetes mellitus mal controlada como desencadenante de un caso de enfermedad de Marchiafava-Bignami. Neurologia 31: 498-500.
10. Zuccoli G, Pipitone N (2009) Neuroimaging findings in acute Wernicke's encephalopathy: review of the literature. AJR Am J Roentgenol 192: 501-508.
11. Sullivan EV, Marsh L, Mathalon DH, Lim KO, Pfefferbaum A (1996) Relationship between alcohol withdrawal seizures and temporal lobe white matter volume deficits. Alcohol Clin Exp Res 20: 348-354.
12. Helenius J, Tatlisumak T, Soinne L, Valanne L, Kaste M (2001) Marchiafava-Bignami disease: two cases with favourable outcome. Eur J Neurol 8: 269-272.
13. Nemlekar SS, Mehta RY, Dave KR, Shah ND (2016) Marchiafava: Bignami Disease Treated with Parenteral Thiamine. Indian J Psychol Med 38: 147-149.
14. Staszewski J, Macek K, Stepień A (2006) Odwracalna demielinizacja ciała modzelowatego w przebiegu choroby Marchiafavy-Bignamiego [Reversible demyelinisation of corpus callosum in the course of Marchiafava-Bignami disease]. Neurol Neurochir Pol 40: 156-161.