CASE REPORT

A 28-year-old male was found with altered mental status and unknown mechanism of injury at the bottom of a flight of stairs. He reportedly had gone to a party the evening before, which was the last time he was seen by his wife. He regularly smoked an average of “one nickel bag” (0.5 gm/10-20 cigarettes) of marijuana daily. His Glasgow Coma Scale (GCS) on presentation was seven, and he was intubated for airway protection. A trauma workup including physical examination revealed a left scalp hematoma and some minor facial fractures. Non-contrast computed tomography (CT) imaging of the brain demonstrated a left temporal and parietal scalp hematoma but no intracranial injury. CT angiography of the neck demonstrated no vascular injury. Ethyl alcohol was undetectable in his serum; his toxicology panel revealed cannabinoids in his urine but was negative for barbiturates, benzodiazepines, and cocaine, methamphetamines, opiates, or phencyclidine. Liver enzymes were within normal limits (aspartate transaminase, AST 50; alanine transaminase, ALT 20; alkaline phosphatase 78; total bilirubin 0.9).

Over the next several days, the patient’s neurological status remained poor, with an average GCS of eight, and intermittent agitation requiring sedation. Repeat CT imaging of the brain demonstrated no new findings. His electroencephalography (EEG) was abnormal with delta slowing and left frontal spikes indicative of encephalopathy. Unenhanced MRI of the brain demonstrated restricted diffusion and hyperintense signal on fluid attenuated inversion recovery (FLAIR) imaging throughout the

INTRODUCTION

Marchiafava-Bignami disease (MBD) was first described in Italian heavy red wine drinkers in 1903. A relatively rare condition classically found in chronic alcoholics, a small number of cases of MBD has been reported in non-alcoholics. Clinical features classically include reduced consciousness or coma, neuropsychiatric manifestations, seizures, hemiparesis, dysarthria, ataxia and apraxia, and symptoms of hemispheric disconnection. It was historically confirmed by pathological diagnosis; however, magnetic resonance imaging (MRI) has become the diagnostic method of choice by easily revealing the pathognomonic features.

We describe a young non-alcoholic male who presented after being found unresponsive at the bottom of a flight of stairs, who failed to make a full neurological recovery despite having no acute traumatic intracerebral findings on brain imaging.

KEY WORDS: Marchiafava-Bignami disease, trauma, corpus callosum

CASE REPORT

Marchiafava-Bignami disease in a trauma patient

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ABSTRACT

Marchiafava-Bignami disease (MBD) is a rare pathological condition affecting the corpus callosum (CC), characterized by progressive demyelination and necrosis. While usually found in patients with chronic alcoholism, it has rarely been characterized in non-alcoholics. We describe a trauma patient with an unknown mechanism of injury, who was found to have MBD after remaining comatose for a prolonged period of time. Magnetic resonance imaging (MRI) demonstrated restricted diffusion involving the genu, body, and splenium of the CC. The patient eventually awoke but was non-communicative and uncomprehending prior to discharge to a nursing facility. We reviewed the literature and report here the first case of MBD encountered in a trauma patient. In conclusion, MBD is an extremely rare condition in non-alcoholic patients, and the use of MRI is crucial for its identification.

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Access this article online

Quick Response Code:
Website: www.onlinejets.org
DOI: 10.4103/0974-2700.150399
corpus callosum (CC) [Figure 1]. Additionally, two small areas of microhemorrhage were identified in the right hemisphere on the gradient echo sequence. The CC did not enhance on a follow-up MRI exam. MRI of the cervical spine demonstrated a small syrinx at the C6 level.

Clinical and neurological findings were consistent with MBD. High dose of thiamine was started empirically based on anecdotal support in the literature. The patient's mental status subsequently improved, with spontaneous eye opening, incoherent vocalization, and random limb movement but no ability to follow commands. Five weeks after the patient’s admission, the patient was discharged to a nursing facility.

DISCUSSION

MBD has been reported worldwide in less than 300 patients. It is a primary degenerative condition of the CC characterized by symmetrical demyelination and necrosis. While other pathologies such as ischemic stroke, diffuse axonal injury, inflammatory demyelination, brain tumors, Wernicke encephalopathy, and Wallerian degeneration can also involve the CC, they can be differentiated based on history, physical exam, and MRI. Two subtypes of MBD have been described, differentiating between either acute or chronic onset. Type A MBD is characterized by acute to subacute onset of impaired consciousness, pyramidal tract signs, limb hypertonia, seizures, hyperintense swelling on T2-weighted MR sequences of the CC, and poor prognosis. Type B MBD has a less precipitous onset and carries a more favorable prognosis. Sequential MR imaging may demonstrate reversal of lesions, suggesting underlying edema rather than demyelination. It appears that our patient’s presentation was more consistent with the acute form of MBD.

Heavy alcohol consumption has been the classic associiative factor with MBD. While the exact role of alcohol in MBD is unknown, it has been hypothesized that a toxin present in the alcohol may be responsible for demyelination in the CC. Alternatively in non-drinkers, there may be exposure from a different source. It has also been suggested that severe liver dysfunction in chronic alcoholics may lead to elevated serum ammonia levels and encephalopathy, callosal edema, and demyelination. Various vitamin deficiencies and malnutrition in general have also been linked with MBD.

Our patient was unique in that he had neither a history of alcohol abuse nor malnutrition. His family reported him to drink alcohol only occasionally. He did have a strong pattern of cannabis use, averaging up to 20 marijuana cigarettes per day. The presentation of our patient, being “found down” with a scalp hematoma, suggested traumatic brain injury. Routine urine toxicology revealed only cannabinoids but this does not exclude the possibility of other illicit drug use just before admission, not included within the screen. Further, the marijuana our patient was exposed to could have been laced with unknown potentially neurotoxic adulterants. Clinical, laboratory, and diagnostic imaging features of other substance abuse or any other cause of sudden coma were absent.

Our patient was described as being asymptomatic just hours prior to admission implying a diagnosis of type A or acute onset MBD. Additionally, it is unclear whether the patient’s improvement in neurological status two weeks into his hospital admission was due to supplemental thiamine or the passage of time and supportive medical care. Scant recovery over five weeks left the patient with severe neurologic impairment at the time of discharge.

CONCLUSION

MBD is an extremely rare neurological disorder that carries an unfavorable prognosis. To the best of our knowledge, this is the first reported case of MBD following injury in the setting of habitual marijuana abuse. Other ambiguities include the possibility of other undetected illicit drug intoxication and the possibility of an unknown injury mechanism.

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How to cite this article: Jorge JM, Gold M, Sternman D, Prabhakaran K, Yelon J. Marchiafava-Bignami disease in a trauma patient. J Emerg Trauma Shock 2015;8:52-4.
Received: 16.04.14. Accepted: 14.05.14.
Source of Support: Nil. Conflict of Interest: None declared.