Marchiafava-Bignami Syndrome: Rare Case Report and Literature Review

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Abstract
Marchiafava-Bignami disease is a rare neurological condition seen most commonly in middle-aged, chronic alcoholic males. It is characterized by demyelination of the Corpus Callosum, which is visible on CT scan and MRI. Diagnosis is usually made with a combination of radiographic and clinical findings. Symptoms of Marchiafava-Bignami disease are similar to other neurologic diseases related to chronic alcoholism, and include ataxia, confusion, memory loss, and slurred speech. This case illustrates that, while classically seen in males, Marchiafava-Bignami disease can be seen in females, and its clinical presentation can be variable. The patient is a young, middle-aged female who presented to the Emergency Department with fever, slurred speech, tachycardia, and chills. She has a past history of substance abuse, including alcohol. T2 weighted MRI demonstrated areas of hyperintensity in the posterior corpus callosum classically seen in Marchiafava-Bignami disease. Together with the clinical history, a diagnosis of Marchiafava-Bignami encephalopathy was made. The Emergency Room physician was made aware of the diagnosis, and the patient was counseled appropriately.

Introduction
Marchiafava-Bignami disease is a rare neurological condition in which segments of the Pons-Cerebellar junction, as well as the Corpus Callosum, begin to demyelinate. It is an extremely rare neurological finding seen on MRI. There have been at least 250 cases reported in the literature since it was originally described in 1903. Most cases are seen in chronic alcoholics, though there have been a few non-alcoholics noted. Most cases are seen in men.

While theories exist as to the exact etiology of the condition, all sources agree that MRI findings are as a result of demyelination. It is associated with other neurological conditions tied to chronic alcohol use, including Wernicke’s encephalopathy. Treatment for this condition is similar to treatment seen for chronic alcoholics, with improvement seen with administration of thiamine and alcohol cessation. Some sources also report improvement with corticosteroids.

There is no “classical” physical examination findings associated with Marchiafava-Bignami other than the radiologic findings. Symptoms vary, and range from confusion and altered mental status in an alcoholic, to memory-loss and loss of muscle strength. Most cases reported have been under 60 years old. There is a general consensus in the literature that if the patient does not cease consuming alcohol, the condition is almost uniformly fatal in less than 5 years.

It has been suggested by some sources that the incidence of this disease is much higher than the literature leads one to believe, but the condition is underdiagnosed because it is a more rare neurological condition.

Thus, physicians must maintain an awareness of this disease in patients presenting with altered mental status with a history of chronic alcohol use.

Clinical Presentation
Our patient presented to the ED via ambulance with fever, chills, slurred speech, and tachycardia. The patient also presented with a history of chills all over their body for the last day. The patient was clearly inebriated, with alcohol on her breath, and was confused. She had an altered mental status, and an unsteady gait.

The patient had a history of asthma, and admitted to previous substance abuse, including a long history of alcohol abuse. Due to her altered mental status, obtaining a more adequate history was not feasible. Due to the neurologic complaints, the attending physician decided to do a CT scan and MRI of the brain. T2 weighted MRI showed hyperdense lesions in the posterior portion of the corpus callosum, as well as 2 hyperdense lesions in the pons-cerebellar junction. Together with the clinical history, a diagnosis of Marchiafava-Bignami encephalopathy was made. The Emergency Room physician was made aware of the diagnosis, and the patient was counseled appropriately.

Discussion
Treatment for Marchiafava-Bignami starts with alcohol cessation. It is generally recommended that patients be hospitalized until they are sober and can be properly counseled on their diagnosis and prognosis. While thiamine supplementation and steroid use have been shown to be beneficial in reversing the demyelination of the corpus callosum, unless the patient ceases alcohol consumption, their neurological condition will gradually deteriorate until it results in permanent impairment or death.

There are no reported classical osteopathic findings associated with Marchiafava-Bignami. Osteopathic findings in patients with this condition may relate more to the symptoms of the underlying condition, and may include Chapman’s reflexes in the 5th and 6th intercostal spaces due to nausea from alcohol abuse. Though it has never been reported, patients could also theoretically present with CRI dysfunction secondary to the underlying neurological condition.

References

Figures 1 and 2: T2 weighted MRI shows areas of hyperintensity in the corpus callosum indicating focal demyelination

Figure 3: T2 weighted MRI showing demyelination at the cerebellar-pontine junction