RESEARCH PAPER | Medical Science | Volume : 4 | Issue : 1 | Jan 2014 | ISSN - 2249-555X

Atypical Contrast MRI Findings in Marchiafava-Bignami Disease

KEYWORDS: Marchiafava Bignami disease, corpus callosum demyelination, CT, MRI

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ABSTRACT *Marchiafava Bignami disease or syndrome is a rare neurological condition associated with chronic alcoholism with demyelination and necrosis primarily affecting the corpus callosum. This case shows us the atypical contrast MRI findings of Marchiafava bignami disease.*

INTRODUCTION

Marchiafava bignami disease is a very rare demyelinating condition affecting the corpus callosum. It is seen mostly in chronic alcoholics which results in progressive demyelination and necrosis of the corpus callosum .We present the CT and MR imaging findings in a patient who presented acutely and review the features of the disease.

CASE REPORT

A 58 year old man who was a known alcoholic and smoker since 40 years presented with sudden onset of slurred speech, giddiness, and tremors of the hand. He used to consume country liquor around 150-200 ml daily. On examination he had slurred speech, dysarthria and dysdiadochokinesia. Romberg's test and finger nose test were positive. His blood glucose was normal. He was subjected to plain and contrast CT scan which revealed nonenhancing hypodense corpus callosum (Fig 1). MRI of the brain demonstrated hypointense areas in the corpus callosum on T1 weighted image (Fig. 3A). The same areas appeared hyperintense on T2 weighted & FLAIR images (Fig 2A& B) with central enhancement on post contrast scan (Fig. 3B). The findings of MRI were consistent with degenerative necrosis of the corpus callosum except for the pattern of contrast enhancement which was central and not peripheral. On the basis of clinical history, physical examination and imaging findings, the diagnosis of Marchiafava bignami disease was made.

DISCUSSION

Marchiafava Bignami disease is a fatal disorder consisting of demyelination of corpus callosum which occurs due to chronic alcohol consumption.[1] It was first described by two Italian pathologists Marchiafava and Bignami .Initially it was thought to occur in individuals who lived in Italy and consumed cheap "Chianti" red wine. However it is now known worldwide that it occurs in malnourished alcoholics.[2]

It is generally accepted that the disease is mainly due to a deficiency in the vitamin B

Complex, and although many patients improve following administration of these compounds, others do not, and some die from the disease. Most patients are male, between 40 and 60 years of age, and have a history of chronic alcoholism and malnutrition.[3]

The body of the corpus callosum, genu, and finally the splenium are affected in descending order.[4]The entire corpus callosum may be also be affected. Other white matter tracts such as the anterior and posterior commisures and the cortico-spinal

Tracts may be involved. Lesions may be also found in the hemispheric white matter and in the middle cerebellar peduncles. The subcortical U-fibers tend to be spared. The corpus callosum degenerates and splits into three layers also known as layered necrosis or 'sandwich sign'.[1]

Diagnosis is made on the basis of clinical history, physical examination and imaging findings.[5] There are three clinical forms of the disease; acute, sub acute and chronic forms. Acute Marchiafava bignami disease presents with mental confusion, disorientation, neurocognitive deficits, and seizures. Muscle rigidity and facial trismus may be severe. Most patients presenting with the acute type of disease will go into coma and eventually die. It may be difficult to clinically distinguish from Wernicke encephalopathy and may occur together with it. Patients with Wernicke encephalopathy have ataxia, ophthalmoplegia, nystagmus, and confusion. However in wernickes encephalopathy, MR imaging shows abnormal signal intensity and contrast enhancement in the mamillary bodies, periaqueductal region, and the walls of the third ventricle. Sub acute type of disease is characterized by dementia, dysarthria and muscle hypertonia. They may survive for years. Chronic type of the disease is characterized by a chronic dementia. In this latter form, the differential diagnosis includes Alzheimer disease, multi-infarct dementia, and Pick disease.

Imaging appearance of Marchiafava Bignami disease on CT is quite subtle. The lesions appear as hypodense areas which are difficult to visualize.[6] The differential diagnosis for hypodense lesions includes infarct, demyelinating disorder and shear injury. On MR, patients show areas of low T1 signal intensity and high T2 and FLAIR signal intensity in the body of the corpus callosum at times extending into the genu and the adjacent white matter. These lesions do not have mass effect and may show peripheral contrast enhancement during the acute phase. Eventually, the lesions cavitate and become well marginated.[7]MR spectroscopy has been used to evaluate patients with Marchiafava Bignami disease. The initial MR spectra shows a mildly increased choline level, low *N*-acetyl aspartate and the presence of lactate.[8]

Thus the diagnosis of Marchiafava Bignami disease can be made accurately in light of correct clinical setting and imag-

Volume : 4 | Issue : 1 | Jan 2014 | ISSN - 2249-555X

ing appearance which can help in the survival of the patients.

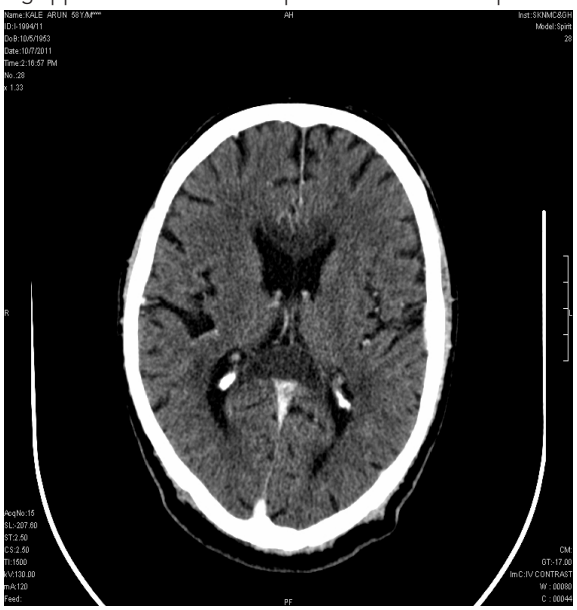

Fig 1: Contrast Enhanced CT scan shows nonenhancing hypodensity involving the genu and splenium of corpus callosum.

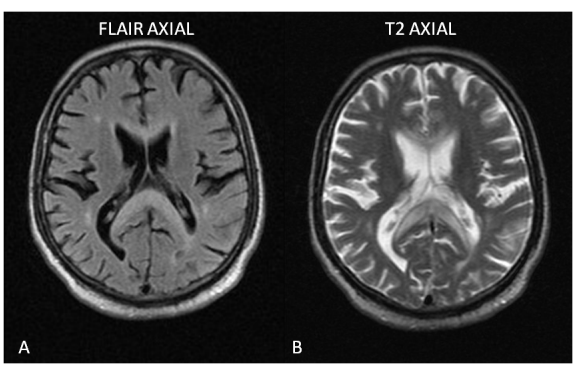

Fig 2 A&B: Axial FLAIR (A) & Axial T2 images showing hyperintese signals in the genu and splenium of corpus callosum.

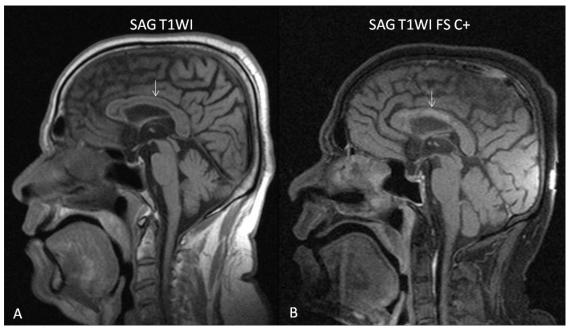

Fig 3 A&B: Plain Sagittal T1 weighted image (A) showing hypointese signals in corpus callosum involving rostrum, genu, body and splenium..Post contrast sagittal T1WI Fat Saturated image showing enhancement especially in the genu and body of corpus callosum.

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