CASE REPORT

Acute Marchiafava-Bignami disease presenting as reversible dementia in a chronic alcoholic

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SUMMARY

Marchiafava-Bignami disease (MBD) is a rare complication of chronic alcoholism. Its clinical diagnosis has considerably changed during recent times, with MRI of the brain paving way for in life diagnosis. We believe that physicians need to have a high index of suspicion, because acute onset MBD is not always fatal and complete recovery is possible, provided the diagnosis is made early and treated appropriately. We report a case of MBD who was diagnosed early in the disease course with subsequent clinical and radiological recovery on institution of appropriate treatment.

BACKGROUND

Even though considered as a rare disease, Marchiafava-Bignami disease can pose diagnostic dilemma in an appropriate scenario (such as heavy alcohol consumers/malnourished individuals) by mimicking the other common aetiologies in the context of a rapidly progressive dementia. This case was highlighted in order to generate awareness regarding this uncommon but historic complication of chronic alcoholism, which if promptly diagnosed and expeditiously treated may be reversible to a significant extent as is exemplified in our case.

CASE PRESENTATION

A 45-year-old gentleman presented with 5 days history of speech disturbances and gait abnormality. Two days prior to admission he had generalised tonic clonic seizures and altered sensorium. On examination, he was confused and disoriented without any signs of meningeal irritation. He had a mixed type of dysarthria with both scanning and spastic components. Fundus and extraocular movements were normal. There was no nystagmus and other cranial nerves were normal. Motor system examination revealed normal power in all limbs, but with spasticity and brisk reflexes with extensor plantar responses bilaterally. Gait was both spastic and ataxic. There was no evidence of disconnection syndrome. The patient had been consuming an average of 350 ml of country liquor per day for the last 20 years. The patient was worked up for acute confusional state with a background of heavy alcoholism.

INVESTIGATIONS

His baseline investigations in the form of haemogram, liver function tests, renal function tests, coagulation profile, vitamin B₁₂ levels, thyroid function tests, serum ACE level and serum ammonia were within normal limits. Anti nuclear antibody (ANA), cytoplasmic-anti neutrophil

cytoplasmic antibody (c-ANCA) and perinuclear - anti neutrophil cytoplasmic antibody (p-ANCA) were negative. Cerebrospinal fluid examination was unremarkable with no pleocytosis, normal biochemistry and sterile culture. Blood HIV and venereal disease research laboratory tests were negative. EEG showed diffuse θ range slowing but no epileptiform discharges. Gadolinium-enhanced MRI of the brain disclosed hyperintense lesions in the central portion of the corpus callosum (with sparing of upper and lower layers), subcortical white matter and putamen on T2-weighted (figure 1A,B) and fluid attenuated inversion recovery sequence (FLAIR) imaging (figure 1C,E). These lesions were hypointense on T1-weighted images (T1WI), showing restricted diffusion on diffusion weighted imaging (DWI) (figure 1D) without any contrast enhancement.

DIFFERENTIAL DIAGNOSIS

In view of the short history with background history of alcoholism, differentials were kept in line of an acute confusional state which included structural pathology (subdural haematoma/parenchymal cerebral haemorrhage/vascular event involving the posterior circulation), infective (acute meningoencephalitis), metabolic (Wernicke's encephalopathy/non-endemic pellagra) as well as non-convulsive status epilepticus.

TREATMENT

On the basis of clinical history and imaging findings, a diagnosis of Marchiafava-Bignami disease (MBD) was made and 1 ml of high-dose vitamin B complex (1000 μ m of vitamin B₁₂, 100 mg thiamine, 50 mg of vitamin B₆ and 50 mg of D-pantethol per ml) was administered daily intramuscularly for 14 days followed by once weekly regimen for 1 month.

OUTCOME AND FOLLOW-UP

With the institution of the above medications, his sensorium, speech and gait improved gradually. After 3 months, he showed complete resolution of symptoms with a normal neurological examination. Repeat Gadolinium-enhanced MRI of the brain showed significant resolution of lesions with only subtle residual T2-weighted hyper intense signal noted in the body of the corpus callosum (figure 2A,B).

DISCUSSION

MBD, a rare and fatal condition associated with chronic alcoholism is characterised by demyelination and necrosis of the corpus callosum.¹ Initially described in persons who ingest red wine, it is now

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Figure 1 (A) Baseline cranial MRI mid-sagittal T2-weighted MRI depicting corpus callosum hyperintensity. (B) Axial T2 sequence showing corpus callosal and bilateral putaminal hyperintensities. (C) Coronal T2 FLAIR images showing corpus callosal hyperintensity which shows diffusion restriction on DW images (D). (E) T2 FLAIR axial images showing hyperintensities in B/L putamen and B/L temporal cortex.

known that this condition can be seen with chronic intake of other alcoholic beverages and even in malnourished non-alcoholics.² The exact pathogenesis is still unclear but it is generally attributed to deficiency in the B complex group of vitamins.³ The clinical spectrum of this disease is diverse which makes diagnosis difficult. Presentation can be acute as in our case with altered sensorium, seizures, ataxia, rigidity and dysarthria. In acute cases, prognosis is usually poor and mortality is high, even though there are case reports documenting complete recovery.⁴ Chronic presentation includes progressive dementia, behavioural abnormalities and an interhemispheric disconnection syndrome (limb apraxia, tactile agnosia, unilateral agnosia and hemialexia). An intermediate form has been described with an initial acute onset followed by regression to chronic form.^{5,6} It may be seen in combination with other manifestations of chronic alcohol abuse like Wernicke's encephalopathy, Central Pontine myelinolysis and Morel's laminar sclerosis.⁷

In the past, cases of MBD were diagnosed only at autopsy, but with the advent of CT and MRI of the brain, early and prompt diagnosis has resulted in improved survival and better prognosis. The characteristic MRI picture of acute MBD shows symmetrical lesions involving the central portion of the body of corpus callosum with sparing of dorsal and ventral layer; the 'sandwich sign'. Sometimes, lesion extends into the genu and splenium,⁸ but only rarely is the entire corpus callosum involved. The lesions are

hypointense on T1WI, hyperintense on T2WI and FLAIR showing diffusion restriction on DWI and variable reduction in apparent diffusion coefficient value.⁹ In acute MBD, T2W hyperintensity is due to both oedema and demyelination. With recovery, T2 signal intensity alteration progressively reduces over time in parallel with decreasing oedema. The corpus callosum may remain hyperintense in case of permanent myelin damage or may recover its normal signal intensity in cases of total remyelination.¹⁰ These lesions usually do not show mass effect and may show peripheral contrast enhancement. In the chronic state, the lesions may cavitate associated with atrophy of the corpus callosum.¹¹ Recent MRI studies have shown that lesions may also be found in the cerebral hemispheric white matter, cerebellar peduncles or cortical grey matter.¹² Our patient had the classical 'sandwich sign' involving the corpus callosum with additional extracallosal lesions involving subcortical white matter and putamen. Heinrich suggested the differentiation of two clinicoradiological subtypes: type A is characterised by major impairment of consciousness, T2-hyperintense swelling of the entire corpus callosum on early MRI and poor outcome. Type B shows only at the most slight impairment of consciousness, partial callosal lesions on MRI and a favourable

Figure 2 Post-treatment cranial MRI showing resolution of lesions on T2 sagittal (A) and axial (B) sequences.

Learning points

- ▶ Marchiafava-Bignami disease (MBD) should be considered as a differential for rapidly progressive cognitive decline in heavy alcoholics, even though uncommon.
- ▶ Despite being a rare and historic complication of chronic alcoholism, awareness regarding the diverse clinical presentations and neuroimaging findings are key to the diagnosis of MBD.
- ▶ The characteristic cranial MRI finding in acute MBD is the 'sandwich sign' characterised by T2 hyperintensity in the central part of body of the corpus callosum sparing the dorsal and ventral layers.
- ▶ Heinrich type B variety of MBD is associated with ultimately good outcome.

outcome.¹³ Our patient categorised into Heinrich type B in view of the clinoradiological picture and ultimate favourable outcome. The principal pathological finding in patients of MBD is degeneration of the corpus callosum that ranges from demyelination, oedema with preservation of axons to necrosis.¹⁴ There is no specific therapy for MBD. Prompt diagnosis and early initiation of treatment with thiamine, vitamin B complex and folic acid expedite clinical recovery. High-dose steroids may aid in recovery by reducing oedema.¹⁵

Competing interests None.

Patient consent Obtained.

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