**CARE Checklist (2013) of information to include when writing a case report **

**Title 1** Acute Marchiafava-Bignami Disease manifested as entire corpus callosum lesions: a case report Page 1

**Topic Item Checklist item description Reported on Line**

**Key Words 2** Acute Marchiafava-Bignami Disease, magnetic resonance imaging, entire callosal lesions, prognosis Page 3

**Abstract 3a** Introduction: Acute Marchiafava-Bignami Disease (MBD) is a rare alcohol-related disorder Abstract, paragraph 1

 **3b** The main symptoms : progressively developed bilateral lower limb weakness and slurred speech.

 Main clinical findings : a 40-year history of chronic and heavy alcohol abuse and typical MRI findings. Abstract, paragraph 2

**3c** The main diagnoses: Marchiafava-Bignami Disease

Therapeutics interventions: vitamin B complex (vitamin B1, B6, B9, and B12)(vitamin B1 was administered through the stomach tube at a dose of 60mg/d, the others were administered intravenously). The eperisone, citicoline and donepezil was also administered to improve the muscle tone and cognitive function.

 Outcomes: the patient's symptoms did not relieve significantly Abstract, paragraph 2

 **3d** Conclusion: MRI findings are critical for the early diagnosis of MBD in the acute stage, which prevent disease progression from irreversible callosal lesion Abstract, paragraph 3

 **Introduction 4** MBD is a very rare and typically fatal disease associated with chronic, heavy alcohol consumption and malnutrition, and is characterized by callosal lesions. Acute lesions were frequently found in the body of the corpus callosum, the genu , and the splenium. However, the entire corpus callosum is rarely involved. Many experts have concluded that complete corpus callosum lesions are one of the poor prognostic indicators. MRI findings, especially DWI, are critical for the diagnosis of acute MBD. Page 3, 4, 5

**Patient Information 5a** Zheng Chunlin , Han nationality, borns on July 29, 1961, he has retired.

**5b** Progressively developed bilateral lower limb weakness and slurred speech for three days. Page 5, 6

**5c** Medical history: The patient had a history of haematemesis and was diagnosed with erosive gastritis two years ago without any treatment. He had no history of poison contact, drug abuse , head trauma, recent fever, or infection. Page 5, 6

Family history: There was no similar family. Page 5, 6

Psychosocial history : There was no psychosocial history Page 5, 6

**5d** Relevant past interventions and their outcomes N/A

 **Clinical Findings 6** Neurological examination revealed abnormal mental status and cognitive deficits. No obvious  abnormalities were found in 12 intracranial nerves. The muscle power of the bilateral upper and lower limbs was only grade 4+ with lead-pipe rigidity of muscle. Deep tendon reflexes in the lower limbs were increased, and Babinski reflexes were positive bilaterally. Hoffmann reflexes and the forced grasp reflex were negative. He displayed no abnormalities in his Case presentation, autonomic system except sphincter dysfunction. . paragraph 1

 **Timeline 7** He had 40-year history of heavy alcohol abuse. He had a history of haematemesis and was diagnosed with erosive gastritis without any treatment in 2016. On December 14, 2018, he began to have a slurred speech when he woke up (he had drinked 250g 6 hours ago), with bilateral lower limbs weakness. Simultaneously, he presented symptoms of dysarthria, nausea, and incontinence. The symptoms were persistent without remission. Subsequently, these symptoms and signs progressed to altered mental status with confusion, bradyphrenia, as well as deterioration in standing capacity within the next two days. On December 17, 2018, he was admitted to hospital Case presentation, for treatment. paragraph 1

**Diagnostic Assessment**

**Therapeutic Intervention**

**Follow-up and**

**Outcomes**

**8a** Diagnostic methods: abnormal mental status and cognitive deficits; positive pyramidal tract symptoms; blood coagulation tests; serum level of folic acid, vitamin B12; CT; MRI. Page 6, 7

**8b** Diagnostic challenges : The serum level of vitamin B1 could not be detected due to the .

restriction of condition Page 7

**8c** Diagnostic reasoning including a differential diagnosis:

 The immediate brain computed tomography(CT) showed strongly hypodensity in the genu and splenium of the corpus callosum ,which suggested an acute cerebrovascular disease.

On the basis of the medical history, long history of drinking, findings on physical examination and Case presentation, laboratory tests, imaging features, the suspicious diagnosis was MBD. Meanwhile, MBD was paragraph 1, 2, 3, 4; differentiate from infarction of the corpus callosum, paraneoplastic syndrome (PNS) , multiple Discussion and sclerosis (MS) ,Wernicke's encephalopathy (WE). Based on the evidence mentioned above, Conclusion, the final diagnosis of acute MBD was reasonable. paragraph 8

**8d** Prognostic characteristics:

 The patient in this case study can be classified as type A which is characterized by alterations Discussion and of consciousness and diffuse swelling of the entire corpus callosum on imaging. Type A has a Conclusion, worse prognosis. Paragraph 4, 5

**9a**  Types of intervention: agathuban, atorvastatin, edaravone. vitamin B complex (vitamin B1, B6, B9, and B12) and vitamin C. The eperisone, citicoline and donepezil was also administered to improve the muscle tone and cognitive function. Abstract, paragraph 2

**9b** Administration of intervention: Before the results of the various tests, agathuban, atorvastatin, edaravone were administered to treat acute cerebrovascular disease for 2 days . Vitamin B1(20mg, three times a day) was administered through the stomach tube for 8 days. Vitamin B9 (20mg, three times a day) was administered through the stomach tube. Vitamin B6 (300mg a day), B12 (5mg, three times a day) and vitamin C (1g a day) were administered intravenously. Baclofen(the initial dose was 5mg, then increased by 5mg every three days until 15mg) was administered through the stomach tube for 9 days. Citicoline(0.2g, three times a day) and donepezil(10mg, one time a day) was also administered through the stomach tube to improve Case presentation, cognitive function. paragraph 6

**9c** Changes in intervention: Based on the results of medical history, long history of drinking, findings on physical examination and laboratory tests,as well as imaging features, the initial diagnosis of “acute cerebrovascular disease” was excluded. So we stopped giving patients agartreban, Case presentation, atorvastatin and edaravone. paragraph 5,6

**10a** Clinician and patient-assessed outcomes: Upon discharge, his mental status improved slightly , but other symptoms remained constant.. A phone follow-up ,four months later, revealed little improvement in his Case presentation, symptoms, except for a decrease in muscle tone. paragraph 6

**10b** Important follow-up diagnostic and other test results N/A

**10c** Intervention adherence and tolerability (How was this assessed?) N/A

**10d** Adverse and unanticipated events N/A

 **Discussion 11a** Discussion of the strengths and limitations : The strength is that this is a very rare and typical case involving the entire corpus callosum. There are a few limitations: 1. The serum level of vitamin B1 could not be detected due to the restriction of condition. 2. The mode of administration (stomach tube) and dosage (60mg/d) of vitamin B1 (personal opinion). 3. Upon discharge, further cranial MRI and related laboratory examinations cannot be scheduled due to the patient's physical condition.

 **11b** Discussion of the relevant medical literature: The corpus callosum is vulnerable to a variety of external and internal risk factors, like alcoholic toxicity and thiamine deficiency. Acute lesions were frequently found in the body of the corpus callosum, the genu , and the splenium. However, the entire corpus callosum is rarely involved. Other sites which have been described include the periventricular white matter, the basal ganglia, the internal capsules, subcortical areas, the cerebral cortex, optic tracts, as well as cerebral and cerebellar peduncles. Among these, complete corpus callosum lesions , which can be classified as Type A, are one of the poor prognostic indicators. Hyperintense signal lesions on T2 weighted imaging (T2WI) , fluid attenuated inversion recovery (FLAIR) and DWI within the corpus callosum are critical for the diagnosis Discussion and of MBD. DWI ,which displays a limited extent of diffusion ,is sensitive to cytotoxic edema. During the acute Conclusion, phase, the involved regions become oedematous. paragraph 2,4,6

 **11c** The rationale for conclusions: The patient in this case study can be classified as type A from a clinicoradiologic point of view (acute onset with suspected expressive aphasia, alterations of mental status, pyramidal tract syndromes and the strong affection of the complete corpus callosum). Considering his short medical history, clinical manifestations, and hyperintense signal lesions on DWI, the patient was likely in the acute stage. Poor prognosis may be related to the brain areas involved in Discussion and the MBD. When the entire corpus callosum is involved, the function of the corpus callosum cannot be Conclusion, compensated(personal opinion), which may explain the poor prognosis of our patient. paragraph 5, 9

**11d** The primary “take-away” lessons of this case report : From this case, I understand the pathogenesis and clinical manifestations of MBD, as well as the diagnostic methods and classification. The last but not the least, I learned about the multiple reasons that affect the prognosis of MBD. Prevention is the key to disease. Later treatment can only partially relieve the patient's symptoms . Therefore, we should Discussion and pay attention to health education. At the same time, doctors should make a diagnosis as soon as possible Conclusion, and take drugs early to prevent the disease from worsening. paragraph 10

**Patient Perspective 12** When appropriate the patient should share their perspective on the treatments they received **Yes**

**Informed Consent 13** Did the patient give informed consent? Please provide if requested . . . . . . . . . . . . . . . . . . . . . . . . . . . . . . . . . . . . . **Yes**√. **No**