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UNUSUAL CASE OF MARCHIAFAVA-BIGNAMI DISEASE PRESENTING AS AXIAL HYPOTONIA

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Abstract

Introduction: Marchiava-Bignami disease is a rare disorder mostly associated with chronic heavy alcohol consumption that results in progressive demyelination and necrosis of the corpus callosum.

Case report: We report a 35-year-old woman with a history of alcohol consumption and malnutrition. Neurological examination revealed axial hypotonia, dysarthric speech and lack of motor coordination. Brain MSCT imaging demonstrated hypodense lesion of the corpus callosum. On the basis of her history, clinical features and imaging studies the diagnosis of an acute form of Marchiava-Bignami disease was made. Definite diagnosis was confirmed at autopsy.
Conclusion: MBD is a medical emergency and early recognition and early aggressive treatment are critical for a good clinical outcome. To our knowledge this is the first case of MBD presented with axial hypotonia.

INTRODUCTION

Marchiava-Bignami disease (MBD) is a rare disorder mostly associated with chronic heavy alcohol consumption. MBD is characterized by primary demyelination and necrosis of the corpus callosum (1). The computerized tomography (CT) and magnetic resonance imaging (MRI) are helpful in diagnosis in the early stages of the disease (2).

CASE REPORT

We report a 35 year-old woman with a history of drinking red wine and very poor nutritional status. One month before the admission the patient suddenly developed gait ataxia and slurred speech. The patient was hospitalized because she experienced acute onset vomiting and mental confusion. Physical examination showed asthenia. Neurological examination revealed dysarthric speech, lack of motor coordination and pronounced axial hypotonia. Hypotonia was presented primarily of neck musculature with the impossibility of keeping the head and achieving a vertical position, her head kept falling off the back. Results of routine blood tests and cerebro-spinal fluid examinations were all within the normal limits. Electroencephalography (EEG) showed diffuse slow waves of 6-8 Hz without epileptiform discharge. Brain computed tomography (CT) which was performed, only in transverse plane, immediately at admission showed no significant abnormalities. Three days later, her rapidly deteriorating level of consciousness rapidly deteriorated and she became comatose (Glasgow Coma Scale–GCS was 3) with respiratory failure that required mechanical ventilation. Follow-up brain MSCT imaging performed one month after the onset of symptoms demonstrated hypodense lesion of the corpus calosum involving genu, body and splenium, in sagittal plane. (Figure 1) The diagnosis of an acute form of MBD was made. The patient was treated with high-dose of thamine vitamin, 100 mg per day, vitamin B complex. A high dose of intravenous corticosteroids was also
administered. Three days after the onset of the therapy the patient showed improvement in her consciousness and become sopor, but 11 days after the admission in hospital the patient got pneumonia and died. A clinical diagnosis was confirmed by postmortem pathologic findings. The main pathologic change was the degeneration of the corpus callosum with demyelination and fragmentation of some axons. Demyelination was accompanied by focal collections of macrophages and present proliferation astrocytes.(Figure 2) General autopsy observation included fibrinopurulent pneumonia and lung abscess on the left side.

**DISCUSSION**

Marciafava-Bignami disease (MBD) is a rare disorder mostly associated with chronic alcoholism. Although several cases of MBD have been associated with non-alcoholic patients, most instances of MBD have occurred as a result of malnutrition. Our patient had a history of malnutrition and consummation of red wine for an unknown period. It is generally accepted that the disease is due to the deficiency of the vitamin B complex, thiamin (3,4). The syndrome is in most instances seen in middle-aged to elderly men drinkers (5). Our patient was a young women with acute form of MBD.

In the acute stage, the patient often has non-specific neurologic changes such as dysarthria, seizures, confusion, coma, generalized muscular hypertonia and clinic diagnosis of MBD can be difficult (6,7). All authors discussing MBD have emphasized the difficulty of making the clinical diagnosis during life because the disease is rare and its manifestations are non-specific (8). The course of the disease in our patient was acute and presented with unusually finding such as axial hypotonia.

Magnetic resonance imaging (MRI) and computerized tomography (CT) are more useful for early diagnosis and detailed analysis of the distribution of lesions (2). Our patient presented characteristic follow-up brain MSCT imaging, performed one month after the onset of the symptoms, and then the disease was recognized.

The most important is early recognition and detection of MBD. In the era before CT scanning and MR, MBD was confirmed almost exclusively at autopsy. Our patient had an acute form of MBD and she had a rapid course resulting in death. No standardized treatment protocols have been established in MBD. The early aggressive treatment is often
associated with marked clinical improvement. Clinical improvement has been documented when using high dose corticosteroids and thiamine (9).

In summary, MBD is a medical emergency and early recognition and early aggressive treatment are critical for a good clinical outcome. To our knowledge this is the first case of MBD presented with axial hypotonia.
REFERENCE


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