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MRI で Marchiafava-Bignami 病が疑われた多発性脳梗塞の 1 剖検例

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An autopsy case of multiple cerebral infarctions under an in vivo suspicion of Marchiafava-Bignami disease based on magnetic resonance imaging

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要旨

Marchiafava-Bignami 病（MBD）はアルコール多飲者に生じ, 脳梁に限局した脱髄・壊死病変を来たす疾患である. 核磁気共鳴画像 (MRI) などの画像診断法の進歩により, 脳梁病変が容易にかつ明瞭に描出できるようになって生前診断が可能となり, MBD の臨床例が多く報告されてきている. 今回, 73 歳の大酒家男性で, MRI 画像で脳梁萎縮がみられたために MBD が示唆された例を経験した. 飲酒量の増量とともに, 意識障害はなかったが急に横になったまま動かなくなって病院を訪れた. 全般的な精神機能の低下, 右手の強制把握, 強制泣き笑い, 眼球運動障害, 運動失調, パーキンソン症状を呈し, 血圧値の変動が激しい高血圧症を有し, 身体各所の動脈硬化と高血圧性心疾患が存在した例である. MRI にて脳梁の萎縮とその近傍で側脳室前角周囲の大脳深部白質に異常信号域を認め, これは左側では一部脳梁や帯状回白質, 前頭葉皮質下白質にまで及び, その中心には嚢胞性病変が存在した. また, 両側の側脳室周囲から半卵円中心に亘る広範な異常信号域のほか, 両側の半卵円中心, 被殻, 視床, および橋に小スポット状の複数個の異常信号域を認めた. 臨床的には多発性脳梗塞と考えられ, 鑑別として MBD が挙げられた. 剖検にて多発性脳梗塞が主病変であり, 脳梁萎縮は MBD ではなく, 左脳梁近傍の前角部周囲白質を中心に存在する大小の多発性脳梗塞病変による神経線維の二次的な変性のためと考えられた. 多発性脳梗塞でも MBD が鑑別診断に挙がってくる MRI 画像を呈し得ることを示す例として重要であり, また MRI に頼り過ぎている MBD の生前診断に警鐘を鳴らす目的で報告した.

Abstract

Marchiafava-Bignami disease（MBD）is rare and has been reported to occur typically in male, middle-aged, malnourished persons, who have a long history of excessive alcoholic consumption. Necrosis and subsequent demyelination of the corpus callosum are the principal pathological features of MBD. Advances in diagnostic imaging, particularly magnetic resonance imaging (MRI) techniques have allowed for an early and precise in vivo diagnosis, and several clinical cases of MBD based on the results of MRI techniques were reported in the literature. At the present time, we saw a 71-year-old, right handed man, and considered him a case of probable MBD because of his confirmed alcoholism and marked callosal atrophy on his MRI, although he died of bronchopneumonia at the age of 73. His alcoholic consumption had gradually increased. He visited hospital, because he suddenly fell down on the floor and was unable to speak or walk without unconsciousness. On examination, he was found to exhibit generalized psychological impairments, forced grasping of his right hand, forced laughing and crying, disturbance of his ocular movement, ataxia, and parkinsonism. He had hypertension accompanied by very labile blood pressure levels, atherosclerosis throughout his body, and hypertensive heart disease. Results of an MRI showed marked atrophy of the corpus callosum, and areas of abnormal signal intensity distributed next to the corpus callosum in the deep white matter surrounding the anterior horns of the lateral ventricles. Particularly, the aforementioned areas of abnormal intensity in the left cerebrum extended up to the part of the left corpus callosum, the white matter of the cingulate gyrus, and the subcortical white matter in the left frontal lobe, as well. And cystic lesions were found in those areas of abnormal high intensity in the white matter. Small spotted areas of abnormal intensity bilaterally scattered in the centrum semiovale, the putamen, the thalamus and the pons, and diffuse areas of abnormal intensity were observed in the centrum semiovale surrounding the lateral ventricles. Based on the clinical information and findings, this patient was given a diagnosis of multiple cerebral infarctions, and the diagnosis may have involved differentiation from MBD. The autopsy revealed that multiple cerebral infarctions were the principal pathological findings in the present case. The marked callosal atrophy was due to multiple infarct lesions distributed next to the left corpus callosum in the deep white matter surrounding the anterior horn of the left lateral ventricle, but not due to MBD lesions. Those infarct lesions secondarily produced axonal degeneration, leading to the callosal atrophy. The present case strongly suggests that, even though he really has a pathological diagnosis of multiple cerebral infarctions, his MRI findings may lead us to consider MBD as an in vivo diagnosis that involves differentiation from the callosal atrophy due to multiple infarctions. At the same time, this case notifies us to be careful to depend blindly on radiological imaging techniques, including MRI, for in vivo diagnosis of MBD. *Tottori J. Clin. Res.* 1(2), 380-396, 2008

Key words: Marchiafava-Bignami 病，脳梁，アルコール中毒，多発性脳梗塞，核磁気共鳴画像; Marchiafava-Bignami disease, corpus callosum, chronic alcoholism, multiple infarction, magnetic resonance imaging

はじめに

Marchiafava-Bignami 病（MBD）はアルコール多飲者や稀に栄養障害者に生じ，脳梁に限局した脱髄性もしくは壊死性病変を来たす疾患である．1898 年，イタリアの Carducci によって最初の剖検例が記載され，その後 1903 年に同じくイタリア人の Marchiafava と Bignami によって 2 例が追加され，計 3 例の剖検例が報告されたのが最初である．従来は剖検でないと確定診断は困難であったが，MRI などの画像診断法の進歩