Neurol Sci (2014) 35:1143–1145
DOI 10.1007/s10072-014-1705-9

LETTER TO THE EDITOR

Corticosteroid treatment in a patient with Marchiafava–Bignami disease

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Received: 25 September 2013 / Accepted: 25 February 2014 / Published online: 9 March 2014

Dear Editor,

We report a case of acute Marchiafava–Bignami disease (MBD) with a favourable outcome after corticosteroid treatment. MBD is a rare disorder characterized by acute demyelination and necrosis of the central part of the corpus callosum (CC), mostly associated with chronic alcoholism [1]. Pathological features include layered necrosis, degeneration and cystic cavitations [2]. MRI is helpful to diagnose MBD and its use allowed identifying patients in an early stage of disease. In the last decade, MBD can be effectively treated with steroids as suggested, although no therapeutic regimen has been established yet [3–5].

A 55-year-old man, with a history of severe daily alcohol consumption in the last 25 years (about 500 ml/day), treated with sodium oxybate, presented a sudden onset of disturbed consciousness and myoclonic jerking of upper limbs, with a slight predominance on his right arm. On admission to our ward, a neurological examination revealed a severe stupor state without any focal signs. Routine blood tests showed elevated liver transaminases, just above the upper limit of normal, and a slightly reduced proteins level. Blood glucose level, electrolytes, hepatitis B and C, VZV, HSV, HIV, and thyroid function were within normal range or negative. Cerebrospinal fluid examination was normal.

MRI, performed on the second day of admission, revealed hyperintense changes in the CC on T2WI and FLAIR (Fig. 1a), without contrast enhancement. These changes were hyperintense on DWI, indicating restricted diffusion in the same areas (Fig. 1b). The CC showed slight swelling. A diagnosis of MBD was made on the basis of MRI results, and the patient's clinical course, along with a history of chronic alcohol abuse as well. A treatment with dexamethasone (8 mg/day), vitamins B1, and B12 was immediately started. Patient's state improved remarkably by the day after. His consciousness changed from a stupor state to somnolence, with episodes of psychomotor agitation. At day 15 he showed a better face-to-face interaction, started moving all four limbs, but he was not able to start his activity daily living. Findings on MRI, 12 days after symptoms onset, corresponded to this clinical improvement, showing on T2WI and DWI a reduction of hyperintense signal in the entire CC, with cystic degeneration areas (Fig. 2a, b). MR spectroscopy displayed significantly lower *N*-acetyl aspartate concentration with an increased choline level in the CC.

The patient was discharged after 3 weeks and sent to a rehabilitation facility. After 2 months, neurological and neuropsychological evaluations were both normal, and steroid administration had gradually decreased and eventually discontinued.

MBD is a rare toxic disorder that can have an acute form with severe consciousness impairment, which often leads to death; a chronic form, with interhemispheric disconnection which may last several years, and an intermediate form with an acute onset followed by a chronic stage. Although most cases of MBD are associated with chronic

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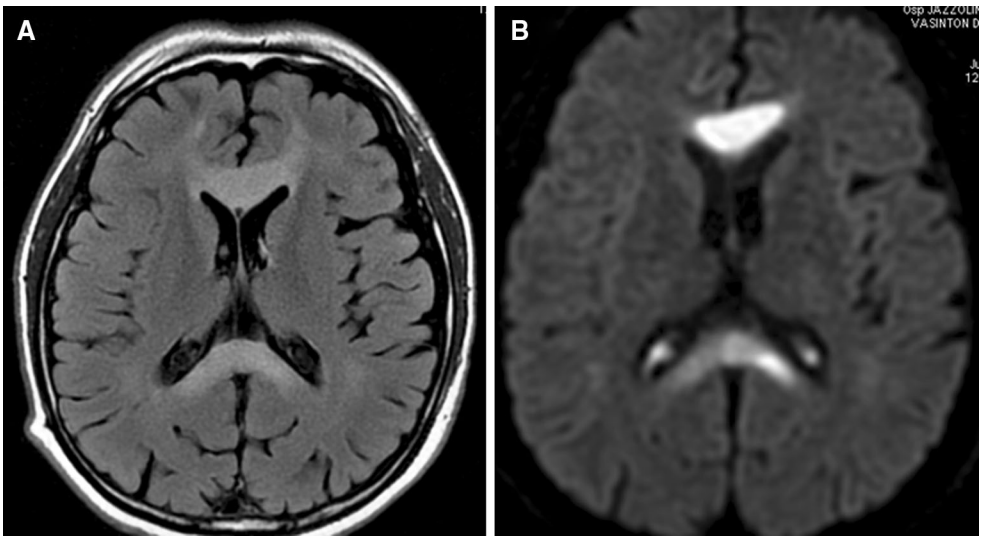

Fig. 1 **a** Axial FLAIR imaging shows hyperintense changes in the genu and splenium of the corpus callosum. **b** Hyperintense signal on DWI, in the same areas

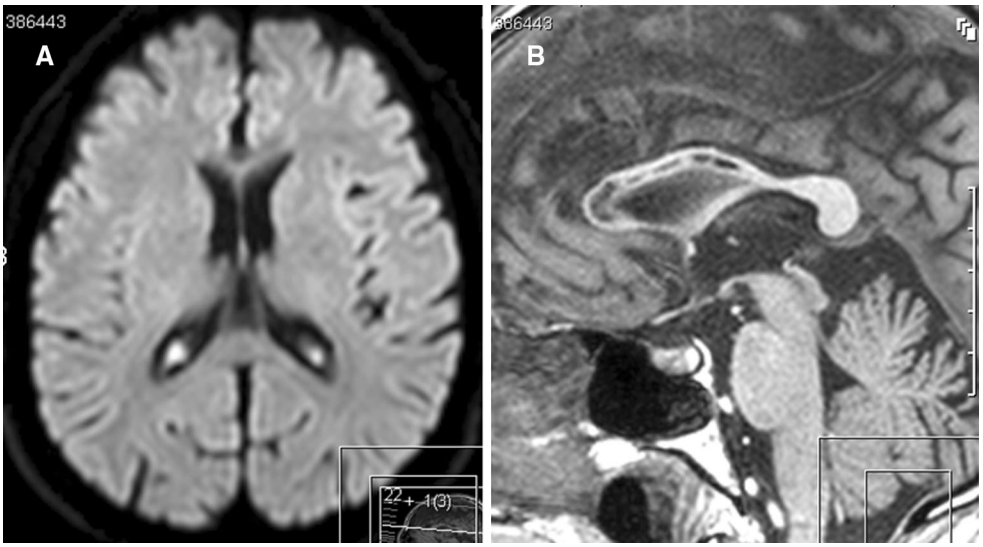

Fig. 2 **a** Twelve days after symptoms onset, DWI shows a reduction of hyperintense signal in the entire corpus callosum. **b** T1-weighted sagittal imaging shows cystic degeneration areas in the corpus callosum

alcoholism and malnutrition, its causes and exact pathophysiology are to be determined yet [6].

The main pathological feature is a degeneration of the CC, with variable degrees of damage, from demyelination to necrosis. This process may involve the entire CC and hemispheric white matter [3].

MRI demonstrates a symmetrical involvement of the CC, with hyperintense signal on T2WI and FLAIR, and hypointensity on T1WI. Early pathological changes can be seen on DWI, with high signal intensity due to reduced diffusion capacity of the surrounding tissue, caused by swollen myelin sheaths, suggesting a reversible lesion [7]. Susceptibility-weighted imaging (SWI) may show asymmetrical hypointense areas in the multiple cortico-subcortical regions, indicating the presence of cerebral microhaemorrhage [8].

A case of MBD, rapidly recovering after high-dose intravenous corticosteroids, has been reported [4]. After therapy, only mild cognitive impairment persisted. There are just a few more reports on effectiveness of steroid treatment in MBD. A patient with anorexia nervosa recovered completely after treatment with methylprednisolone [4]. A case of a malnourished woman with MBD, treated with high-dose steroid, presented a favourable

outcome in the acute stage, but worsening after 18 months, due to the development of a chronic MBD form [3]. In another case, the authors did not use high doses of steroids due to the patient's milder symptoms, but a low dose of prednisolone (30 mg/day) was equally effective [4]. We presume that oedematous change in the early stages of MBD causes impairment of the blood–brain barrier. Corticosteroids are known to stabilize blood–brain barrier by reducing vasogenic permeability, thereby decreasing inflammatory oedema. Reducing the myelin sheath swelling can let the steroids reverse MBD lesions, and determine an improvement of the patient's clinical condition [9]. Disappearance of typical MRI findings after administration of high-dose corticosteroid therapy may represent a resolution of altered BBB permeability and demyelinating changes.

We have described a case of a patient presenting a complete clinical recovery after corticosteroid therapy, and thus sustaining the beneficial effects of this treatment in MBD, strengthening the need for early recognition.

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