MARCHIAFAVA-BIGNAMI DISEASE: A CASE WITH COMPLETE AND SUSTAINED RECOVERY

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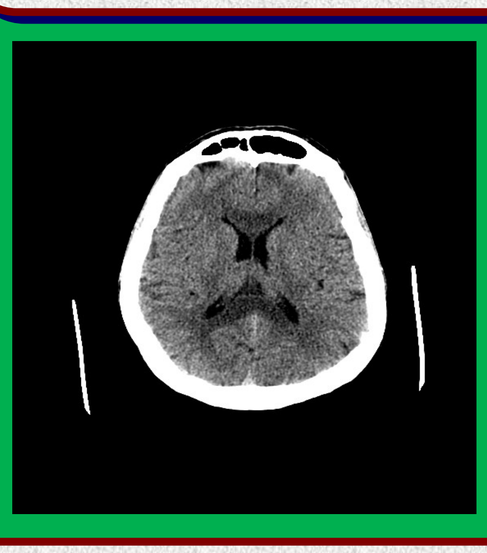
Introduction

Marchiafava-Bignami Disease (MBD) , originally described in Italy in 1903, is a neurological disorder characterized by demyelination and necrosis of corpus callosum, usually associated with chronic alcohol abuse and/or chronic malnourishment. MBD is associated with high mortality and severe neurological impairment in the survivors. An effective therapy has not been established yet for this condition. We described the case of a 46-year-old male patient with complete and persistent recovery.

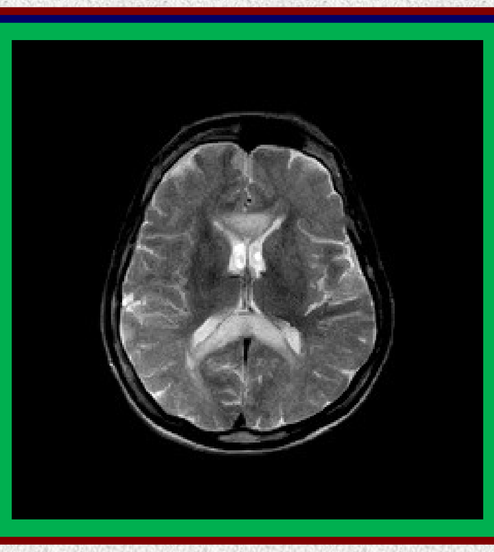
Case Report

This patient had a history of 30-year-long alcohol abuse and came to our observation for progressive confusional state, disturbances of deambulation and loss of weight . His general examination showed severe malnourishment (BMI 16). He presented with confusion , loss of memory, poor orientation in time and space, ataxic gait, hyperreflexia without Babinski sign, impaired vibration and position tests in the lower limbs , intact ocular movements. Blood tests revealed macrocytic anemia , hypoalbuminemia and high transaminase level. T2- weighted MR images , performed at admission, detected high signal intensity of splenium and genu of corpus callosum. EEG showed marked slowing of cerebral electric activity.

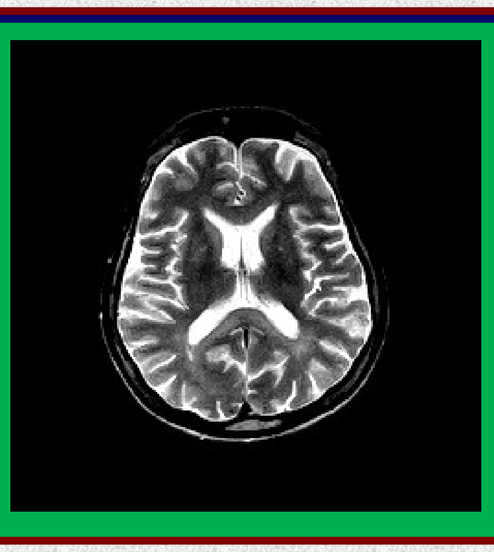
High i.v. doses of vitamin B complex were started immediately after admission and continued for 21 days. Adequate hydration and nutrition supplements were also administered. A few days after admission, the clinical conditions of the patient progressively and gradually improved. MRI performed at day 14 revealed nearly complete resolution of imaging abnormalities. At day 22, the patient was discharged from the hospital. He was alert and oriented, mentation was slightly slowed and minimal short- term memory deficit was present . The deambulation was normal. At 6-month follow-up, the patient continued to abstain completely from drinking alcoholic beverages. Under nutritionist advice he had reached a normal weight and had normal MRI and clinical examination.



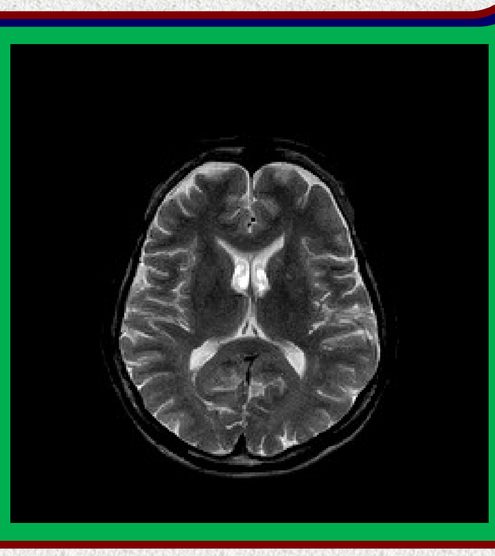
CT at onset



MRI at onset



MRI after 14 days



MRI after 1 year

Discussion and Conclusions

MBD is a rare condition with poor prognosis, since approximately only 8% of patients showed a positive outcome. A recent classification of MBD described a subtype characterized by severe neurological impairment and poor prognosis with rarely reversible MRI lesions not confined to the splenium, and a subtype with less severe neurological signs and good outcome associated with partial and reversible involvement of corpus callosum, possibly caused by underlying oedema rather than demyelination. The prompt and sustained recovery in the present case favors the latter subtype. The distinction, based on clinical and neurological features, of the two subtype seems of paramount importance in defining the diagnosis and prognosis of patients.