A CASE OF MARCHIAFAVA-BIGNAMI DISEASE: CLINICAL AND MRI CORRELATION.

F. De Marchi, C. Varrasi, G. Tondo, D. Mittino, R. Cantello
Department of Neurology, University of Eastern Piedmont (Novara)

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INTRODUCTION

Marchiafava-Bignami disease (MBD) is a rare, often fatal, toxic neurological disease mainly characterized by demyelination of the corpus callosum, with or without associated lesion of hemispherical white matter. Both the clinical and the imaging features can provide useful information as to evolution; ; however, the prognostic significance of these data is still debated. We present a case of MBD with a mixture of favorable and unfavorable hints that had an unexpected fatal outcome, even after the overcoming of the acute phase of the illness.

CASE REPORT

A 44 year-old Italian man with a 25-year history of alcohol abuse. In the past 2 weeks he had been increasingly showing asthenia, mental slowing and gate impairment and weight loss of 4 kg in 10 days, secondary to complete fasting. 7 days prior to admission, he stopped drinking voluntarily.

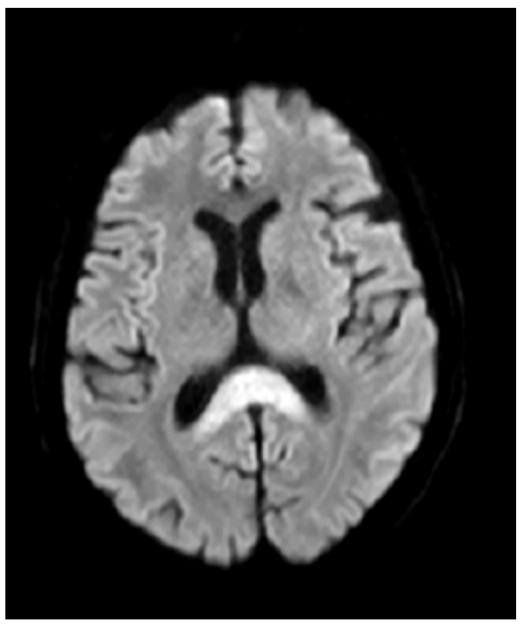
Neurological examination: cachectic and dehydrated; alert, but confused and disoriented in S/T; slow from the psychomotor point of view and unable to execute complex orders. Ataxic gait, diffuse muscle wasting and weakness. Face hypo-mimic, speech dysarthric and slow. The patient occasionally complained of visual hallucinations (animals and people in the room). There were no clear signs of hemispheric disconnection.

MRI with diffusion-weighted sequences: showed restricted water diffusion in the splenium and the body of corpus callosum (fig. 1,2). T2 and FLAIR weighted sequences confirmed hyperintense lesions (fig. 3) in the splenium and the body of corpus callosum, with relative sparing of the dorsal and ventral layers, which represented the typical “sandwich sign” (fig. 5). In FLAIR images, bilateral frontal cortical lesions were evident as hyperintense areas (fig.4).

On the basis of the above-mentioned features, a diagnosis of **Marchiafava-Bignami disease** was made.

Therapy: thiamine, hydration and nutrition.

10 days after admission, we noticed a progressive improvement of the ideomotor performance. The patient was able to walk slowly without support. He continued being disoriented in S/T. Visual hallucinations persisted. One month later, he died hit by a motor car during a sudden escape. His relatives and caring personnel reported that, in spite of the ongoing treatment, he had clearly worsened, with uncontrollable agitation and confusion.



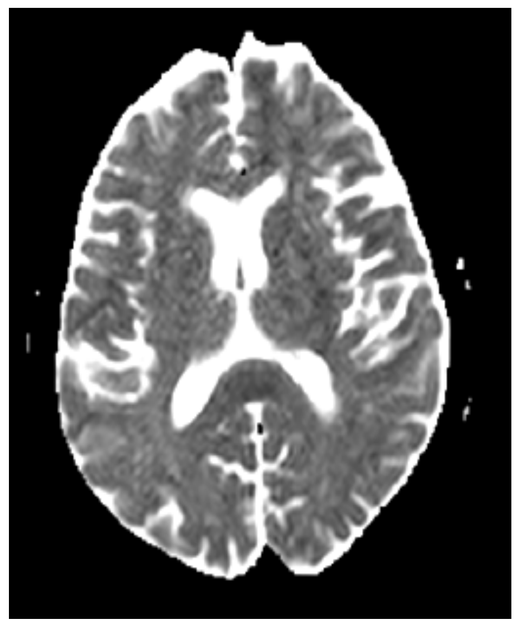
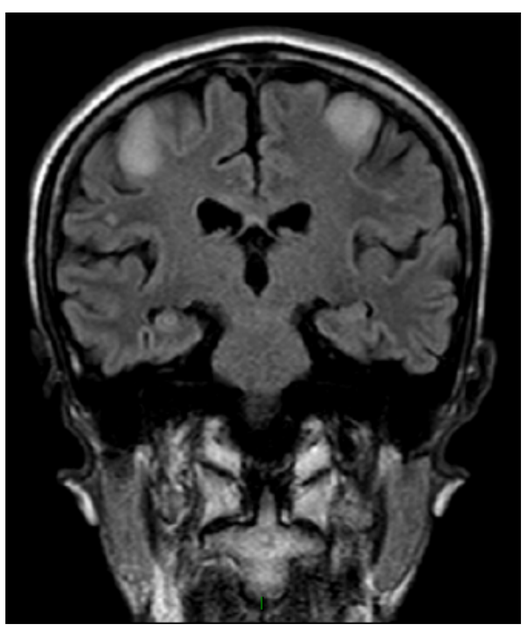


Fig. 1: DWI show hyperintensity of the corpus callosum; fig. 2: ADC images show a corresponding less prominent hypointensity in the splenium.



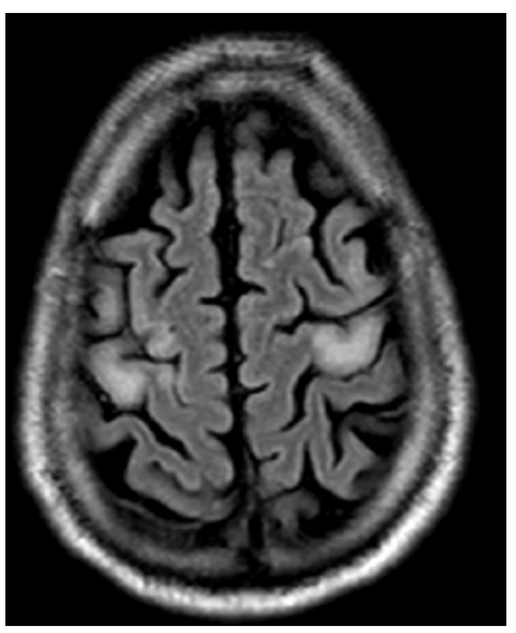


Fig. 3, 4: FLAIR coronal and axial images show frontal lesion as hyperintense areas.

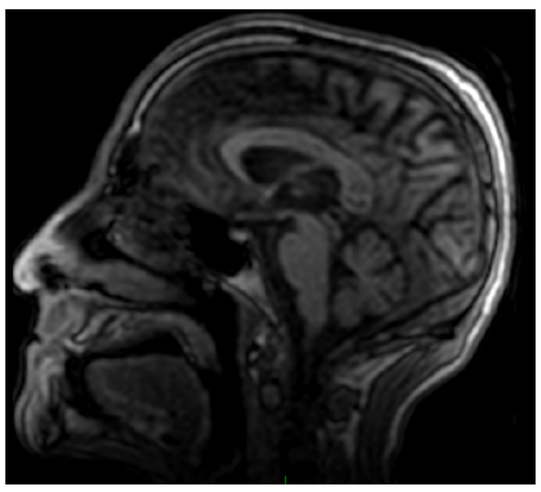


fig. 5: hyperintensity of corpus callosum with characteristic “sandwich sign”.

DISCUSSION

In MBD lesions are often located across the entire corpus callosum. Cortical lesions can be associated (frontal and temporal lobe).

A partial lesion of corpus callosum is related with a good outcome. While a complete injury to the corpus callosum and the extracallosal lesions, are related with an aggressive course and poor outcome.

In the patient of ours, MRI showed a just partial involvement of the corpus callosum (positive sign) but also the high signal intensity in T2 sequences, the diffusion restriction in DWI sequences, and the cortical lesions (negative signs). Bilateral frontal cortical lesions are somewhat uncommon in MBD, usually associated with complete involvement of corpus callosum.

CONCLUSION

Recent advances in neuroimaging favor an early MBD diagnosis, leading to proper treatment. Some MRI indexes were proposed as prognostic indicators. Partial sparing of the corpus callosum would predict a more favorable outcome. If cortical lesions coexist, they may however represent an outweighing negative finding.

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