Reversible hyperintensity in middle cerebellar peduncles. An infrequent finding in Marchiafava-Bignami disease

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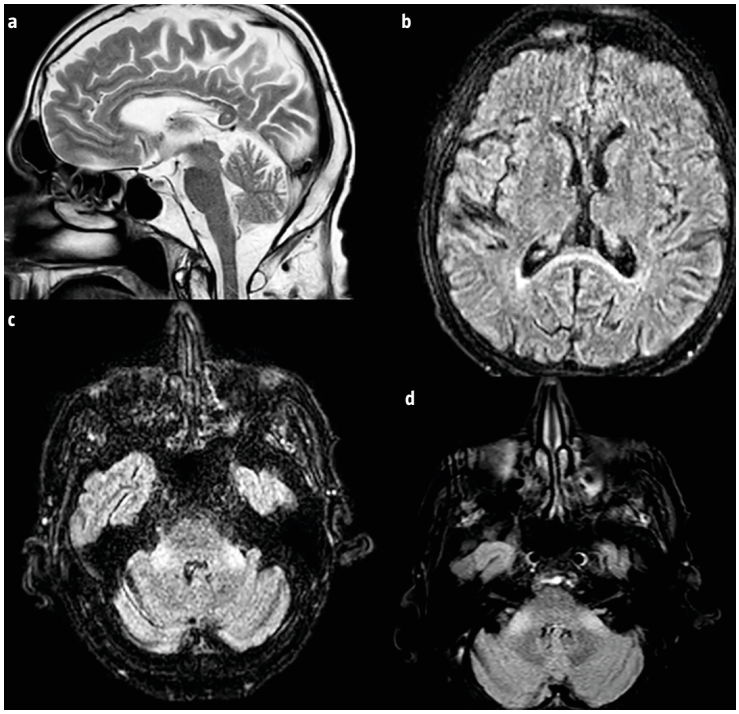

Figure 1. a) In T_2WI magnetic resonance image, we can see hyperintensity at the splenium of the corpus callosum in sagittal sequences. b) In T_2-FLAIR we also observed hyperintensity at the splenium of the *corpus callosum* in axial sequences sequences. c) In axial DWI sequences, we appreciated hyperintensities in bilateral middle cerebellar peduncles, also known as MCP-sign. d) In T_2-FLAIR we also observed the MCP-sign.

Hiperintensity in middle cerebellar peduncles in T_2 sign magnetic resonance image (MRI) (MCP-sign) is an infrequent finding, usually in the context of neurodegenerative pathologies, considered a major radiological diagnostic criteria of fragile X-associated tremor ataxia syndrome [1]. It has rarely been described in other entities like multiple system atrophy, Wilson disease, etc. We present a case of Marchiafava-Bignami disease, where reversible MCP-sign is observed.

A 59-year-old male, with 40 years history of alcoholism, presents subacute gait disturbance and cognitive deterioration, progressively worsening over 3 months, with inability to walk, requiring hospital admission. On examination, we observe somnolence, cerebellar syndrome (gait ataxia, dysarthria) and cognitive impairment (recovery-memory deficit, alteration of processing speed and executive-attentional functions). The analytical study showed deficiency of folic acid and thiamine. Brain MRI showed global thinning and T_2 hyperintensity at the splenium of the corpus callosum and the MCP-sign (Fig. 1).

The patient was diagnosed with rapidly progressive dementia and ataxia. Progressive subacute course made cerebrovascular etiology unlikely. Cerebrospinal fluid study showed only hyperproteinorrachia, with negative polymerase chain reaction, so infectious etiology is ruled out. A comprehensive analytical study ruled out immune-mediated etiology. We considered neurodegenerative etiology, multiple system atrophy is ruled out in the absence of parkinsonism or dysautonomia. Given the finding of MCP-sign, we performed a genetic study of the *FMR1* premutation, with no alterations [2].

Final diagnostic judgement was probable MBD type-B, based on clinical and radiological findings. Treatment with vitamin supplementation showed progressive clinical improvement, being able to walk with one support two months after discharge, with persistence of the cognitive symptoms. A follow-up brain MRI highlights the resolution of MCP sign (Fig. 2), but persistence of the findings in the *corpus callosum* (not shown on the image).

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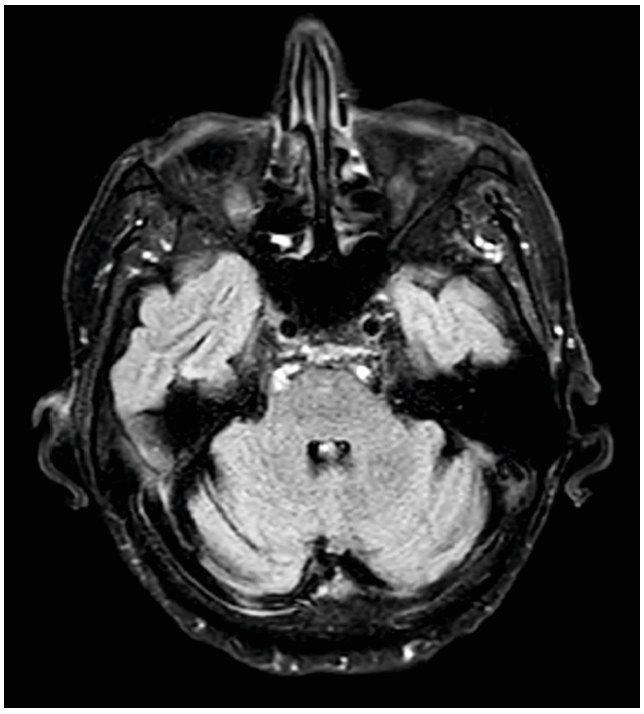

Figure 2. A follow-up brain magnetic resonance image showed the resolution of the hyperintensity in middle cerebellar peduncles (in T_2-FLAIR sequences).

Marchiafava-Bignami disease is an infrequent pathology (although probably underdiagnosed), affecting most frequently middle-aged male (40-60 years of age) [1]. Its pathophysiology is unknown, it is associated with chronic alcoholism and malnutrition (including vitamin deficiencies, especially thiamine) [2], producing demyelinating lesions involving the *corpus callosum* [1,3]. The few cases described demonstrate a variable clinical presentation [4], including ataxia, impairment of mental status, tetraparesis, etc. Given the non-specific symptomatology, the diagnosis in the past was based on *post mortem* pathological examination, and currently is based on the characteristic neuroimaging findings (symmetric lesions of the *corpus callosum*). Extracallosal lesions such as cortical/subcortical white matter lesions are well documented, but the MCP-sign is rare, with only a few cases reported [1,3,5], occasionally associating poor prognosis [1]. Treatment includes vitamin reconstitution and supportive care [4]. The prognosis is poor, with death in up to 21% of cases [2], and only 8% of cases have a favorable outcome [2,5]. It has been postulated that early diagnosis and treatment may improve the prognosis [4,5].

In our case, good clinical and radiological response stands out, with resolution of the MCP-sign after vitamin supplementation, its resolution associated with clinical improvement could indicate that MCP-sign is a marker of prognosis of the disease, although the description of more cases is necessary to draw more weighty conclusions. The presence of MCP-sign should also suggest the possibility of Marchiafava-Bignami disease, and early vitamin and nutritional assessment should be performed, being an infrequent but treatable entity, with a potentially fatal course in the absence of treatment.

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