IMAGEM EM NEUROLOGIA/IMAGE IN NEUROLOGY

Atypical Cerebellar Involvement in MOG Antibody-Associated Disease (MOGAD) in Early Childhood

Envolvimento Cerebeloso Atípico na Doença Associada ao Anticorpo Anti-MOG na Infância

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Acute disseminated encephalomyelitis (ADEM) is the syndrome most commonly associated to positive antibodies against myelin oligodendrocyte glycoprotein (MOG) and has an estimated incidence of 0.2-0.4 cases per 100 000 children, in the paediatric population.[1]

We present a case of a 3-year-old boy who was brought to the Emergency Department for the first time due to difficulty walking and excessive drowsiness, as reported by his parents. The child was also experiencing language disorders. The neurological examination revealed an asymmetric tetraparesis with a left-side predominance, generalized myotatic hyperreflexia and ataxic gait. This episode was preceded by a fever of unknown origin that lasted for 7 days. Brain magnetic resonance imaging (MRI) showed diffuse lesions in the white matter, with bilateral involvement of the thalamus and the brain peduncles. Spinal MRI showed lesions that extended from the medullary transition to the dorsal region (D10), with even expansive areas at the cervical level (**Fig. 1**). Electroencephalogram showed diffuse slowing and cerebrospinal fluid (CSF) examination was normal. Anti-aquaporin 4 (AQP4) antibodies were negative, but the anti-MOG were positive (titer 1:320). He was treated with intravenous pulses of methylprednisolone (30 mg/kg/day) for 5 days and fully recovered.

Three months later, the patient was brought back to our hospital with symptoms of gait imbalance, frequent falls, apathy and altered speech (according to his parents he could no longer make complete sentences). Upon examination,

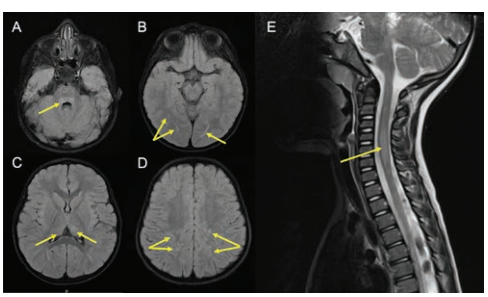

Figure 1. MRI of the brain (A-D, axial FLAIR) and spinal cord (E, sagittal T2), revealing several lesions, which allowed the diagnosis of ADEM to be established: lesions adjacent to the fourth ventricle (A, arrow), diffuse lesions of the occipital region, bilaterally (B, arrows), bilateral thalamic lesions (C, arrows) and lesions scattered throughout the supratentorial white matter (D, arrows); in E, a longitudinally extensive lesion is observed, with expansion of the cervical cord (arrow).

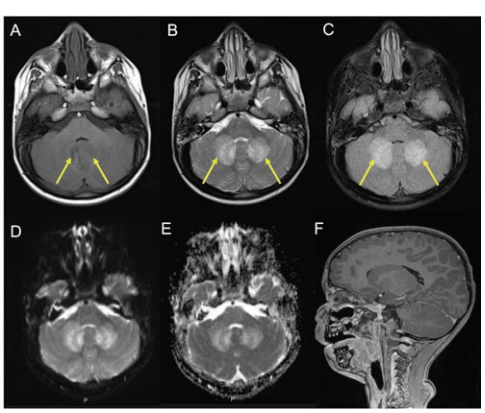

Figure 2. Cerebral MRI revealing the presence of hypointense lesions on axial T1-weighted images (A, arrows) and hyperintense on axial T2-weighted (B, arrows) and FLAIR (C, arrows), centered on the dentate nuclei of the cerebellum and involving the adjacent white matter. The lesions are relatively symmetrical and do not exhibit diffusion restriction (D, E) nor do they enhance after gadolinium administration on T1-weighted imaging (sagittal plane, F).

the patient exhibited signs of dysarthria, a slight resting tremor, a hesitant gait and a positive Romberg sign. He had been admitted to another hospital a week prior with bacterial pneumonia and received intravenous antibiotic therapy with good clinical evolution. A new brain MRI showed marked hypersignal areas on T2-weighted images of the cerebellar dentate nuclei and surrounding white matter (**Fig. 2**). The electroencephalogram was normal. Oligoclonal bands were detected in the CSF (the remaining tests were negative). AQP4-antibodies were negative and MOG-antibodies remained positive, with a titer of 1:1000. The patient was treated again with intravenous pulses of methylprednisolone (30 mg/kg/day) for 7 days with excellent clinical evolution and was discharged with oral prednisolone. A follow-up MRI one month later showed marked attenuation of the previously visualized lesions. A progressive withdrawal of oral prednisolone was performed over 8 weeks. The child remains asymptomatic and without any focal deficit.

In ADEM, typical MRI findings include bilateral diffuse lesions, mainly involving cerebral white matter and spinal cord, consistent with demyelination.[1-3] The clinical phenotype "ADEM" is increasingly being recognised as an umbrella-term and may be the translation of a very diverse set of entities. MOGAD is just one of them, with an increasing number of specificities being described, both in clinical and (mainly) in imaging terms. Even so, the lesional pattern presented by our patient is very atypical. It is true that the involvement of the cerebellar peduncles is very suggestive of MOGAD,[1,4] but the presence of relatively symmetrical lesions centered on the dentate nuclei is not common. Without prior knowledge of this child's history, other clinical entities of metabolic or genetic origin could have been suspected in the differential diagnosis. We present this case as a rare and distinctive lesion pattern in MOGAD with significant clinical and imaging significance. ■

Contributorship Statement / Declaração de Contribuição

TAL: Conception, writing and final approval.

CC, RG: Conception and final approval.

RPP: Images preparation and final approval.

FP: Conception, critical review with intellectual contribution and final approval.

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