IMAGEM EM NEUROLOGIA/IMAGE IN NEUROLOGY

A Look into Familial Hemiplegic Migraine

A Imagem de uma Enxaqueca Hemiplégica Familiar

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We present an 8-year-old girl, with family history of familial hemiplegic migraine (FHM) type 2, with confirmed *ATP1A2* mutation in the mother. She had a previous episode of headache with associated motor deficit, but the reminder of her prior medical history was unremarkable.

She presented to the emergency department with a history of fever and left unilateral headache, accompanied by right-sided weakness and speech disturbances with progressive worsening for two days.

At admission, neurological examination revealed mild hemiparesis and moderate motor and sensory aphasia, as well as an attention deficit.

Analytical study and lumbar puncture were normal. Electroencephalography revealed slow activity in the left hemisphere.

Magnetic resonance imaging (MRI) showed diffuse cortical swelling and T2/FLAIR hyperintensity in the left hemisphere. Areas of restricted diffusion were identified in the left postcentral cortex and in the subcortical white matter of the left temporal lobe. Contrast enhanced imaging, angiographic or perfusion studies were not performed.

A presumptive diagnosis of a severe FHM attack was established and treatment was initiated with 100 mg of methylprednisolone for five days. The symptoms progressively resolved within a week after the start of treatment, but the aphasia had not resolved completely at the time of discharge. Migraine prophylaxis with 25 mg of lamotrigine daily was initiated.

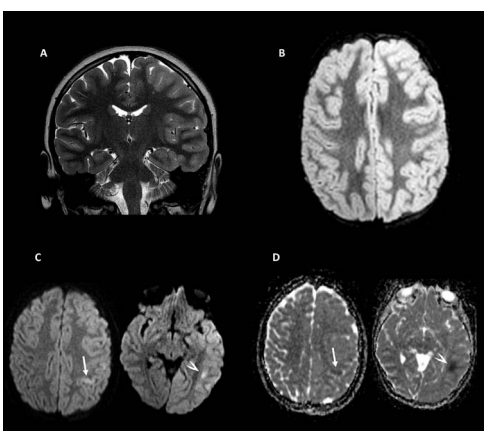

Figure 1. Cerebral MRI during migraine attack: (A) coronal T2-weighted and (B) axial T2 FLAIR images showing diffuse cortical swelling and hyperintensity in the whole left hemisphere; (C) Diffusion weighted imaging with (D) ADC map confirming restricted diffusion in the postcentral cortex (arrows) and subcortical white matter of the temporal lobe (arrowheads).

A follow-up MRI nine months later revealed a normal exam, with complete resolution of all the previous findings. By this time her neurological exam was also normal, with complete resolution of all clinical findings.

Genetic testing confirmed mutation in *ATP1A2* gene in our patient.

Due to its rarity, imaging findings in FHM are not well established. Imaging studies between attacks are typically normal. Cortical swelling and T2/FLAIR hyperintensity are the most often described findings during attacks and are normally reversible.[1]

Although ours and other published cases suggest that reversible restricted diffusion on MRI might occasionally occur in hemiplegic migraine attacks, the presence of restricted diffu-

sion as a definite characteristic of FHM is still at issue: reversible restricted diffusion has been reported, including in cases of children with pathogenic ATP1A2 variants,[2-4] but cases with normal diffusivity or even increased diffusivity in the ADC maps are also described.[5] In the absence of a permanent lesion in the follow-up exam, we disfavor the hypothesis of the restricted diffusion in our patient being due to infarction. Diffusivity anomalies in FHM may be related to perfusion changes during attacks,[6] and vasospasm may also play a role here,[5] but further investigation is needed. ■

Contributorship Statement / Declaração de Contribuição

JSS: Conception and writing of the manuscript and preparation of the images.

VSA: Acquisition and interpretation of data for the work.

DC: Acquisition and interpretation of data for the work

JPF: Critical review with intellectual contribution.

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