Teaching NeuroImages: Transient cytotoxic edema in a child with a novel ATP1A2 mutation

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Figure 1 MRI of the brain obtained within the first week of symptoms

(A) Axial diffusion-weighted imaging demonstrates right hemispheric patchy restricted diffusion that crosses vascular territories and associated cerebral edema. (B) Magnetic resonance angiography shows prominent right leptomeningeal vessels suggesting hemispheric hyperperfusion.

A 7-year-old boy with prior episodes of hemiplegia and family history of hemiplegic migraine presented with fevers, waxing and waning encephalopathy (lethargic and poorly interactive), and left-sided weakness persistent throughout his 5-week hospitalization.

Figure 2 Repeat neuroimaging 1 week after presentation

(A) Axial fluid-attenuated inversion recovery demonstrates persistent diffuse right hemispheric cerebral edema with (B) corresponding areas of hyperperfusion on arterial spin labeling perfusion imaging. There is interval resolution of restricted diffusion, suggestive of transient cytotoxic edema rather than true infarct.
Diagnostic testing revealed a suspected pathogenic mutation in ATP1A2 (c.2285G>C; p.Gly762Ala), a gene associated with a broad phenotypic spectrum encompassing familial hemiplegic migraine type 2 (FHM2) and alternating hemiplegia of childhood. Previous case series illustrate that FHM2 attacks can be prolonged, debilitating, and associated with impaired consciousness and fever. Cortical edema has been described, but transient diffusion restriction can be another radiologic feature (figures 1 and 2).

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**Disclosure**
The authors report no disclosures relevant to the manuscript. Go to Neurology.org/N for full disclosures.

**References**

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