Teaching NeurolImages: 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.

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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.

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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 FMH2 attacks can be prolonged, debilitating, and associated with impaired consciousness and fever.¹ Cortical edema has been described, but transient diffusion restriction can be another radiologic feature (figures 1 and 2).²

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Disclosure

The authors report no disclosures relevant to the manuscript. Go to [Neurology.org/N](https://www.neurology.org/N) for full disclosures.

Appendix Authors

Name	Location	Contribution
Alexandra B. Kornbluh, MD	Nationwide Children's Hospital, Columbus, OH	Review of case history and EMR documentation, primary research into the literature, choosing appropriate images for figures, primary manuscript author
Melissa G. Chung, MD	Nationwide Children's Hospital, Columbus, OH	Critical revision of the manuscript for intellectual content, manuscript supervision

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