Teaching *NeuroImages*: Mitochondrial encephalopathy, lactic acidosis, and stroke-like episodes

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Figure Brain MRI

Cortical hyperintense signal on T2-weighted imaging during the first episode at age 24 (A), fluid-attenuated inversion recovery images during the second episode at age 25 (B), and diffusion-weighted imaging sequences during the last 2 episodes at age 33 (C, D).

A 33-year-old man, with a history of seizures, hearing loss, migraine, and diabetes mellitus, presented with recurrent episodes of fever, seizures, and neurologic deficit associated with transient, cortical hyperintense MRI signal (figure). No family history of neurologic illness was reported. Mitochondrial DNA analysis revealed m.3243A>G tRNA^{Leu} (UUR) mutation causing mitochondrial encephalopathy, lactic acidosis, and stroke-like episodes (MELAS). Brain MRI in MELAS classically shows lesions, predominantly affecting posterior regions, which cross

vascular territories.¹ Stroke-like episodes are probably a result of mitochondrial dysfunction in the capillary endothelial cells.² Heteroplasmic mutation may account for the selective distribution of brain lesions in MELAS.

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