



Visual Diagnosis

## “Black Toenail” Sign

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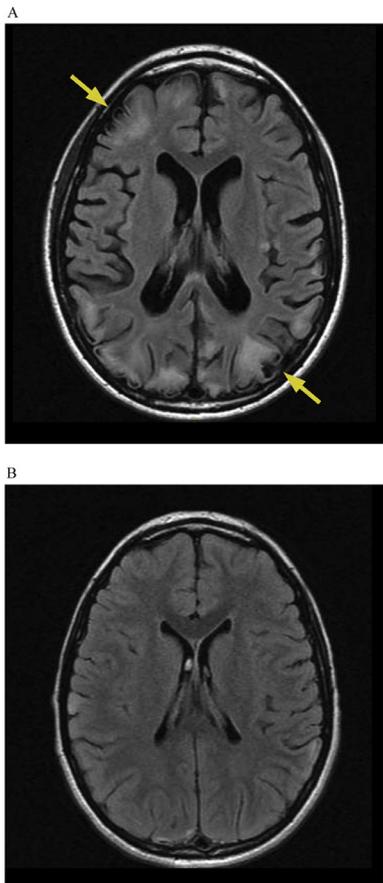
This 11-year-old boy with genetically confirmed MELAS (mitochondrial encephalopathy, lactic acidosis, and stroke-like episodes syndrome) with mutation at mt.3243A>G presented with extreme fatigue. Upon presentation, he had elevated lactic acid of 5.2 mmol/L. His brain magnetic resonance imaging showed progression of multifocal gyral necrosis and cerebral atrophy compared to the images done three years earlier (Fig). He was discharged soon after his fatigue resolved following supportive therapy.

This image finding of gyral necrosis is known as “black toenail” sign, representing the MELAS-related gyral

necrosis that is commonly seen in chronic MELAS patients.<sup>1</sup> Although the etiology of gyral necrosis has not been clearly elucidated, it is assumed that the necrosis is associated with multifactorial etiologies including hypoxic-ischemic injury, decreased substrate delivery, decreased toxin removal, and superimposed neuronal excitation leading to energy depletion.

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**FIGURE.** (A) Magnetic resonance imaging enhanced fluid-attenuated inversion recovery (FLAIR) showing “black toenail sign” (arrows) characterized with gyral necrosis and cerebral atrophy. (B) Three years prior to presentation, these abnormal findings were not present.

**Reference**

1. Whitehead MT, Wien M, Lee B, et al. Black toenail sign in MELAS syndrome. *Pediatr Neurol.* 2017;75:61–65.