A 17-year-old girl presented with subacute decline in ambulation, ataxia, generalized weakness, dysphagia, and asymmetric hearing loss. MRI findings include abnormal signal in medial thalami, mesencephalon, posterior aspect of pons, and medulla oblongata (figure). Magnetic resonance spectroscopy showed elevated lactate and decrease in N-acetylaspartate (figure, F). DNA isolated from muscle biopsy showed A8344G mutation of mitochondrial DNA (tRNA [Lys] gene), associated with 80% of patients with myoclonic epilepsy with ragged-red fibers (MERRF). MERRF is a rare mitochondrial disorder with variable onset and clinical presentation. Neuroradiologic findings of MERRF are reported rarely, with brainstem and cerebellar degeneration being the main feature of MERRF.¹
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The authors report no relevant disclosures. Go to Neurology.org/N for full disclosures.

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Reference
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