Bilateral High Signal Intensity on DWI of the Precentral Cortices in Mitochondriopathy Due to *MT01* Mutation and *MT-TF* Variant

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Figure MRI Presentation of Mitochondriopathy Due to *MT01* Mutation and *MT-TF* Variant in Adults

Brain MRIs of a sister (upper line, 39 years old) and her brother (lower line, 37 years old) with *MT01* mutation and *MT-TF* variant showing cortical, bilateral, symmetric, b1000 DWI hypersignal of the precentral gyri (A, E) without ADC restriction (B, F) nor FLAIR (C, G) or T1 abnormality (D, H).

A 40-year-old woman and her 38-year-old brother developed since childhood optic neuropathy, absence epilepsy, cognitive disability, cerebellar syndrome, hypertrophic cardiomyopathy, and scoliosis, due to a mitochondriopathy associating *MT01* mutation and an *MT-TF* variant, both genes involved in mitochondrial tRNA modification.¹ Brain MRIs were normal during childhood, but cortical, symmetric hyperintensities of precentral gyri on diffusion-weighted imaging (figure) arose apart from any new symptom, different from previously reported *MT01* mitochondriopathies, only noncortical,² from status epilepticus, generally unilateral and associated with a thalamic or corpus callosum splenium hypersignal, and from Creutzfeldt-Jakob disease, usually more posterior and associated with pulvinar and basal ganglia hyperintensities.

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Appendix Authors

Name	Location	Contribution
Guillaume Dorcet	Département de Neurologie, Hôpital Pierre Paul Riquet, CHU de Toulouse, France	Case report concept, design and critical revision of content, and conduct of the case report
Vincent Fabry, MD	Département de Neurologie, Hôpital Pierre Paul Riquet, CHU de Toulouse, France	Critical revision of content

Appendix (continued)

Name	Location	Contribution
Fabrice Bonneville, MD, PhD	Service de Neuroradiologie, Hôpital Pierre Paul Riquet, CHU de Toulouse, France	Case report design, critical revision of content
Pascale Cintas, MD	Département de Neurologie, Hôpital Pierre Paul Riquet, CHU de Toulouse, France	Major role in the acquisition of data, case report concept, critical revision of the manuscript content

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