

Teaching NeuroImages: Longitudinally extensive transverse myelitis in MELAS

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A 16-year-old girl presented with acute-onset flaccid quadriparesis with urinary incontinence. Medical history unveiled generalized epilepsy since age 7 years and status epilepticus 3 years before. Multisystem compromise was absent. Family history was unremarkable. Two days after admission, generalized refractory status epilepticus occurred. Neuroimaging studies disclosed laminar cortical necrosis and longitudinally extensive transverse myelitis (LETM) in cervical and thoracic spinal cord levels (figure). Muscle biopsy disclosed subsarcolemmal mitochondrial proliferation and ragged-red fibers (figure). Genetic testing confirmed m.A3243G mutation in the *MTTL1* gene, diagnostic of mitochondrial myopathy, encephalopathy, lactic acidosis, and stroke-like episodes (MELAS).¹ LETM can be seen extremely rarely in MELAS.²

AUTHOR CONTRIBUTIONS

Dr. Souza: conception of case report project, organization of case report project, execution of case report project, writing of the first draft. Dr. Pinto: conception of case report project, organization of case report project, execution of case report project, writing of the first draft. Dr. Oliveira: execution of case report project, writing of the first draft, manuscript review and critique.

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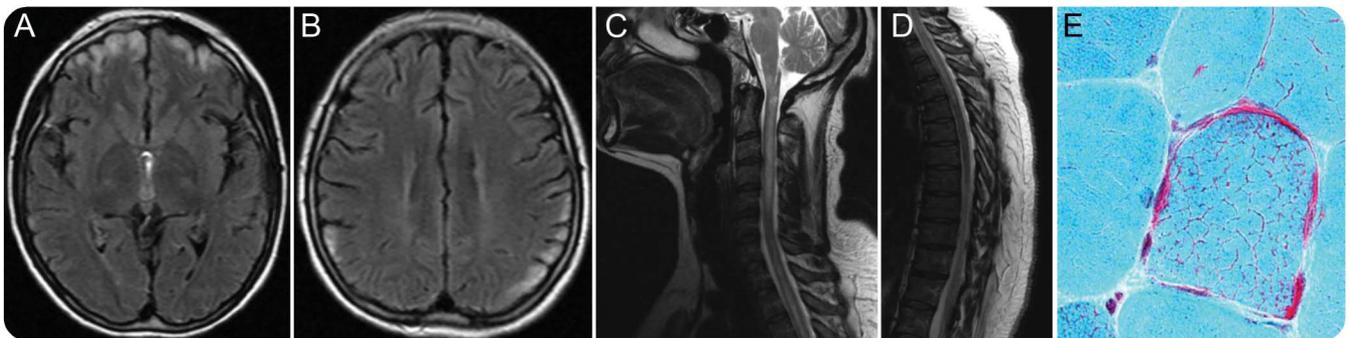
DISCLOSURE

The authors report no disclosures relevant to the manuscript. Go to Neurology.org for full disclosures.

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Figure Longitudinally extensive transverse myelitis in mitochondrial myopathy, encephalopathy, lactic acidosis, and stroke-like episodes (MELAS)



(A, B) Axial brain MRI section shows laminar cortical necrosis in frontal, temporal, parietal, and occipital lobes in fluid-attenuated inversion recovery sequences. (C, D) T2-weighted sagittal spine MRI section shows longitudinally extensive transverse myelitis in the cervical and thoracic spinal cord levels. (E) Muscle biopsy discloses ragged-red fibers in Gomori trichrome stain.

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