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 \textit{MTTL1} gene, diagnostic of mitochondrial myopathy, encephalopathy, lactic acidosis, and stroke-like episodes (MELAS).\textsuperscript{1} LETM can be seen extremely rarely in MELAS.\textsuperscript{2}
Teaching NeuroImages: Longitudinally extensive transverse myelitis in MELAS
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