Rapid emergence of temporal and pulvinar lesions in MELAS mimicking Creutzfeldt-Jakob disease

Figure MRI and spectroscopy

(A) The diffusion-weighted image displays bitemporal neocortical hyperintense signals. (B) The fluid-attenuated inversion recovery image 2 days after the initial MRI scan reveals newly emerging symmetric lesions in the pulvinar thalami. (C) Magnetic resonance spectroscopy displays a strong lactate signal.

Hyperintense areas in the pulvinar in fluid-attenuated inversion recovery images and greater hyperintensity in cortical areas in DWI are typical of sporadic Creutzfeldt-Jacob disease (sCJD).¹ A 59-year-old woman developed confusion, progressive aphasia, mutism, and fluctuations of vigilance within 2 weeks. MRI revealed abnormalities consistent with sCJD (figure). CSF showed normal cell counts, negative PCR for HSV, elevated lactate (4.6 mmol/L), and increased levels of 14-3-3 and tau protein (1,300 pg/L). There were no periodic sharp-wave complexes on EEG recordings. Magnetic resonance spectroscopy revealed a lactate signal indicative of mitochondriopathy² and genetic analysis confirmed the MELAS A3243G mutation. Mitochondriopathies should be considered if otherwise typical features of sCJD rapidly emerge.

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