Teaching NeuroImages: Mitochondrial encephalopathy, lactic acidosis, and stroke-like episodes

A 33-year-old man, with a history of seizures, hearing loss, migraine, and diabetes mellitus, presented with recurrent episodes of fever, seizures, and neurologic deficit associated with transient, cortical hyperintense MRI signal (figure). No family history of neurologic illness was reported. Mitochondrial DNA analysis revealed m.3243A>G tRNALeu (UUR) mutation causing mitochondrial encephalopathy, lactic acidosis, and stroke-like episodes (MELAS). Brain MRI in MELAS classically shows lesions, predominantly affecting posterior regions, which cross vascular territories.1 Stroke-like episodes are probably a result of mitochondrial dysfunction in the capillary endothelial cells.2 Heteroplasmic mutation may account for the selective distribution of brain lesions in MELAS.

REFERENCES

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