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Letter to the Editor

Neuronal intranuclear inclusion disease can mimic MELAS

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With interest we read the article by Kutsuna *et al*. about 63 years-old male who was diagnosed with neuronal intra-nuclear inclusion disease (NIID) based upon the clinical presentation (urinary retention, gait disturbance), magnetic resonance imaging (MRI) findings, and a skin biopsy showing typical eosinophilic, hyaline, intra-nuclear inclusions [1]. The patient was admitted for a stroke-like episode (SLE) manifesting as acute-onset speech disturbance and dysgraphia during two hours that had already disappeared on arrival to the hospital [1]. Multimodal MRI revealed a diffusion-weighted imaging (DWI) hyperintense subcortical lesion bilateral that was isointense on apparent diffusion coefficient (ADC) maps [1]. Perfusion-weighted imaging (PWI) revealed hypoperfusion of the left temporo-parietal area that normalised after six days [1]. The study is appealing, but raises concerns that require further discussion.

We disagree with the notion that the patient suffered a stroke-like episode (SLE) [1]. The term SLE is a pathognomonic feature of mitochondrial encephalopathy, lactic acidosis, and stroke-like episode (MELAS) syndrome but more rarely occurs in other mitochondrial disorders (MIDs) as well [2]. The morphological equivalent of a SLE on imaging is the stroke-like lesion (SLL) [3]. On MRI SLLs are characterised by hyperintensity on T2/FLAIR, DWI, and PWI [3]. SLLs are hypointense on oxygen-extraction fraction OEF-MRI, the area of a SLL shows hypometabolism on fluoro-deoxy glucose (FDG)-positron emission tomography (PET). ADC maps can be highly variable, being hyperintense, hypointense, or isointense [4]. If the authors actually mean transitory ischemic attack (TIA), the term SLE should be replace accordingly.

Because the patient had a history of arterial hypertension, we should be informed if blood pressure values were within normal limits during hospitalisation or elevated. Because the patient also had diabetes, it is crucial to know the fasting blood glucose and the HbA1c values. Particularly, hyper- or hypoglycemia could explain could explain the DWI hyperintensities and also the PWI hypointensity. There is also a need to provide creatine-kinase kinase values. If elevated, this could suggest unwitnessed seizure that could also explain the MRI lesions. A normal electroencephalography (EEG) does not rule out previous seizure activity.

We disagree with the notion in the discussion that acute manifestations of NIID could be attributed to decreased cerebral perfusion in the left temporo-parietal region [1]. Hypoperfusion in the index patient in the left temporo-occipital distribution does not concur with a vascular territory. This is why a mitochondrial disorder needs to be ruled out by mtDNA sequencing. Absence of lactic acidosis is no argument against a mitochondrial disorder.

Overall, the study carries obvious limitations that require re-evaluation and discussion. Clarifying these shortcomings would strengthen the conclusions and could improve the study. The cause of the cerebral lesions on imaging remains unexplained and may not necessarily be attributable to NIID.

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