LETTER TO THE EDITOR

To The Editor

Sequential Stroke-Like Lesions in MELAS are Common and Diagnosable upon Multimodal MRI

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With interest we read the article by Chen et al. about a 53-year-old female with mitochondrial encephalopathy, lactic acidosis and stroke-like episodes (MELAS) syndrome who experienced two sequential stroke-like episodes (SLEs) within 12 months.[1] It was concluded that MELAS can mimic stroke in older patients, but is distinguished on MRI and with appropriate clinical suspicion, why unnecessary biopsy can be avoided.[1] We have the following comments and concerns.

The acronym "MELAS" does not stand for "metabolic encephalomyopathy with lactic acidosis and stroke-like episodes", as mentioned in the introduction, but for "mitochondrial encephalopathy, lactic acidosis, and stroke-like episodes".[2]

We do not agree with the wording in the title.[1] Imaging is not carried out of SLEs but of stroke-like lesions (SLLs), the morphological equivalent of an SLE on imaging.

We do not agree with the statement that MELAS is only an inherited disorder.[1] MELAS is inherited in only about 75% of the cases.[3] In the remainder, the causative mutation occurs spontaneously. Knowing the origin of the mutation is crucial for genetic counselling. We should be told about the family history, particularly if the mother or any other first-degree relative carried the causative mtDNA variant as well or if any of them manifested clinically with features of a mitochondrial disorder (MID).

We do not agree that "SLEs occur with cortical imaging abnormalities".[1] The authors obviously mean SLLs, which usually start in the cortex and usually spread to the subcortical white matter and the deep grey matter.[4] Furthermore, SLLs not only occur supratentorially but also infratentorially.[4]

The statement "SLEs do not follow single vascular territories" is contradictory to the statement that "a SLE is due to smooth muscle dysfunction within vessel walls".[1] Dysfunction of vascular smooth muscle cells (VSMCs) implies that the lesion is vascular in nature and very well follows a vascular territory.

Though it is mentioned that the causative variant m.3243 A > G occurred in a heteroplasmic distribution, the exact heteroplasmy rate was not mentioned. Knowing heteroplasmy rates in various tissues is crucial as it may determine the phenotype and outcome of an individual patient.

It is neither unique that SLLs end up as laminar cortical necrosis nor that they recur. We should be told about the outcome of the second SLL. SLLs, may not only end up as laminar cortical necrosis but also as normal brain tissue, white matter lesion, cyst, atrophy or the toenail sign.[4]

It should be added to the discussion that SLLs typically show up as T2-, DWI, PWI hyperintensity in a non-vascular distribution, which expands over time. The lesion is hypointense on OEF-MRI and is characterised by hypometabolism on FDG-PET.[4]

Missing is the trigger of the SLLs. We should know if the trigger was a seizure, physical stress, emotional stress, nutritional stress, an infection or drugs.

Overall, the interesting case report has a number of limitations, which should be addressed before drawing conclusions. Results of more MRI modalities should be presented as well as the family history, the trigger of the SLLs, the heteroplasmy rate and the phenotype and genotype of first-degree relatives. Several inconsistencies should be solved.

Conflicts of Interest

None.

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