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Letter to the Editor

Epilepsy in Leigh Syndrome Due To the ND3 Variant Requires Non-Mitochondrion-Toxic Anti-Seizure Drugs and the Ketogenic Diet

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In a recent article Kori *et al.*, reported about a 4 years-old female with Leigh syndrome(LS) due to the *ND3* variant m.10158T>C with a heteroplasmy rate of 82% (Kori, A. *et al.*, 2019). The study has a number of shortcomings, which require comments.

The first shortcoming is that the anti-seizure-therapy was inadequate. If only levetirazetam, carbamazepine and clobazam were given than it is not surprising that the patient deteriorated with recurrence of seizures and that cerebral lesions progressed. Seizures in patients with a mitochondrial disorder (MID) need to be aggressively treated as they may trigger stroke-like lesions (SLLs) and may become intractable (Finsterer, J., & Zarrouk, M. S. 2013). If three anti-seizure drugs (ASDs) are ineffective, they need to be replaced. When applying ASDs in MIDs only ASDs with a low mitochondrion-toxic potential should be used. ASDs with high mitochondrion-toxic potential include carbamazepine, valproic acid, phenytoin, and phenobarbital (Finsterer J. 2017). They should not be applied as first-line ASDs and only if less mitochondrion-toxic ASDs failed to be effective. If ASDs are ineffective, the ketogenic diet (KD) should be added as it may stop seizures (Paleologou, E. *et al.*, 2017). The KD attenuates neuronal injury via autophagy and other mitochondrial pathways (Wang, B. H. *et al.*, 2018).

The second shortcoming is that the nature of the cerebral lesions was not identified. Only T2- and T1-images were presented. More important would be diffusion-weighted images, apparent-diffusion-coefficient maps, perfusion-weighted images, and the oxygen-extraction fraction MRI. Additionally, results of MR-spectroscopy should be presented. More profound assessment of the MRI may unmask the described lesions as SLLs, which require appropriate treatment with nitriv-oxcide-precursors, ASDs, steroids, or the KD.

We do not agree with the notion that there was a transition from LS to MELAS. Typical MRI features of LS were still present on the last MRI and SLLs may also occur in LS.

Overall, this interesting case report would be more meaningful if the entire ASD therapy and entire results of multimodal MRI was provided.

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