Cerebral Circulation - Cognition and Behavior 4 (2023) 100160

Contents lists available at ScienceDirect

Cerebral Circulation - Cognition and Behavior

journal homepage: www.sciencedirect.com/journal/cerebral-circulation-cognition-and-behavior

A comment on a case report about perfusion abnormality in neuronal intranuclear inclusion disease with stroke-like episode

ARTICLE INFO

Keywords
Mitochondrial disorder
NIID
Stroke-like episode
Transitory ischemic attack

Letter to the editor

We read with interest the article by Katsuna et al. about a 63 years-old male with slowly progressive neuronal intra-nuclear inclusion disease (NIID) who suddenly developed a stroke-like episode (SLE) clinically manifesting as transient speech disturbance and dysgraphia for two hours [1]. In addition to NIID, his previous history was positive for arterial hypertension and diabetes [1]. Because cerebral magnetic resonance imaging (MRI) showed hypoperfusion in the left temporo-occipital region on perfusion weighted imaging (PWI), the transient episode was interpreted as stroke mimic in connection with the NIID [1]. The study is appealing but raises concern that should be further discussed.

The main limitation of the study is that the authors mix up the terms "stroke-like episode" and "transient ischemic attack" (TIA). SLEs are the hallmark of mitochondrial encephalopathy, lactic acidosis and stroke-like episodes (MELAS) syndrome but can occur in some other mitochondrial disorders and in non-mitochondrial disorders as well. They are clinically characterized by manifestations similar to those seen in ischemic stroke but may additionally manifest with seizures, cognitive disturbances, or lactic acidosis [2]. The morphological correlate of SLEs on cerebral imaging is the so called stroke-like lesion (SLL) [3].

We disagree with the notion that the index patient had a SLE and a SLL on MRI. SLLs are characterized on multimodal cerebral MRI by hyperintensity on T2, fluid attenuated inversion recovery (FLAIR), diffusion weighted imaging (DWI), and PWI and hypointensity on oxygen-extraction fraction (OEF) MRI [3]. On fluoro-deoxy glucose positron emission tomography (FDG-PET) the area of the SLL is characterized by hypometabolism [4]. SLLs are not congruent with the territory of a cerebral artery. Additionally, SLLs are characterised by focal lactate accumulation manifesting as a lactate peak on magnetic resonance spectroscopy (MRS). Direct measurement of lactate in the cerebrospinal fluid (CSF) may show normal results. MRI images presented in Fig. 1 of the case report do not match with this definition [1].

We should know how the authors explain the perfusion abnormality in the light of the normal magnetic resonance angiography (MRA). MRA ruled out any occlusion, stenosis, dissection, aneurysm, or vasospasm. An argument against a perfusion deficit is that the patient did not develop any clinical manifestations corresponding to the area of hypoperfusion. A second argument against a perfusion deficit is that PWI hypointensity did not correlate with the hyperintensities on DWI [1]. Apparent diffusion coefficient (ADC) maps were isointense [1]. A third argument against a perfusion deficit in the left temporo-occipital area is that the DWI lesions concerned the cortico-medullary junction in both hemispheres [1]. An argument against a TIA is that there were multifocal hyperintensities on DWI. A TIA usually goes along without DWI lesions. Only a single NIID patient with a SLL has been previously reported [5].

There is no mention about the type of speech disturbance. We should know whether the patient developed aphasia or dysarthria, or both and whether the patient was right or left handed.

Further limitations are that no electroencephalography (EEG) was recorded and that no FDG-PET was carried out. To rule out a focal status epilepticus it is crucial to record an EEG. To assess if there were areas of hypo- or hyper-metabolism an FDG-PET is essential.

Overall, the interesting study has limitations that challenge the results and their interpretation. Addressing these limitations could further strengthen and reinforce the statement of the study. According to the presented data there is neither evidence for a perfusion deficit nor for a SLE. If a SLE is suspected in a patient with NIID, he should be investigated for a mitochondrial disorder.

Abbreviations: ADC, Apparent diffusion coefficient; CSF, cerebrospinal fluid; DWI, diffusion weighted imaging; EEG, electroencephalography; FDG-PET, fluoro-deoxy glucose positron emission tomography; FLAIR, fluid attenuated inversion recovery; MRA, magnetic resonance angiography; MELAS, mitochondrial encephalopathy, lactic acidosis and stroke-like episodes; MRI, magnetic resonance imaging; MRS, magnetic resonance spectroscopy; NIID, neuronal intra-nuclear inclusion disease; OEF, oxygen-extraction fraction; PWI, perfusion weighted imaging; SLE, stroke-like episode; SLL, stroke-like lesion; TIA, transient ischemic attack.

https://doi.org/10.1016/j.cccb.2023.100160

Available online 30 January 2023

Funding sources

No funding was received.

Ethics approval

Was in accordance with ethical guidelines. The study was approved by the institutional review board.

Consent to participate

Was obtained from the patient.

Consent for publication

Was obtained from the patient.

Availability of data

All data are available from the corresponding author.

Code availability

Not applicable.

CRediT authorship contribution statement

Josef Finsterer: Data curation, Supervision, Writing – original draft, Conceptualization, Writing – review & editing.

Declaration of Competing Interest

None.

Acknowledgments

None.

References

[1] F. Kutsuna, Y. Tateishi, K. Yamashita, T. Kanamoto, T. Hirayama, T. Shima, A. Nagaoka, S. Yoshimura, T. Miyazaki, J. Sone, T. Izumo, A. Tsujino, Perfusion abnormality in neuronal intranuclear inclusion disease with stroke-like episode: a case report, Cereb. Circ. Cogn. Behav. 3 (2022), 100127, https://doi.org/10.1016/j.cccb.2022.100127.

[2] J. Finsterer, Mitochondrial metabolic stroke: phenotype and genetics of stroke-like episodes, J. Neurol. Sci. 400 (2019) 135–141, https://doi.org/10.1016/j.jns.2019.03.021.

[3] J. Finsterer, R. Aliyev, Metabolic stroke or stroke-like lesion: peculiarities of a phenomenon, J. Neurol. Sci. 412 (2020), 116726, https://doi.org/10.1016/j.jns.2020.116726.

[4] J.H. Kim, M.K. Lim, T.Y. Jeon, J.H. Rha, H. Eo, S.Y. Yoo, C.H. Shu, Diffusion and perfusion characteristics of MELAS (mitochondrial myopathy, encephalopathy, lactic acidosis, and stroke-like episode) in thirteen patients, Korean J. Radiol. 12 (1) (2011) 15–24, https://doi.org/10.3348/kjr.2011.12.1.15.

[5] Q. Zhou, M. Tian, H. Yang, Y.B. Luo, Adult-onset neuronal intranuclear inclusion disease with mitochondrial encephalomyopathy, lactic acidosis, and stroke-like (MELAS-like) episode: a case report and review of literature, Brain Sci. 12 (10) (2022) 1377, https://doi.org/10.3390/brainsci12101377.

Josef Finsterer
Neurology & Neurophysiology Center, Postfach 20, Vienna 1180, Austria
E-mail address: fifigs1@yahoo.de.