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Apply the Hirano or Japanese Criteria to Diagnose MELAS

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LETTER TO THE EDITOR

We read with interest the article by Wong, *et al*., about a 46-year-old male with hypertrophic cardiomyopathy as a feature of late-onset mitochondrial encephalopathy, lactic acidosis, and stroke-like episode (MELAS) syndrome due to the m.3243A>G variant in *MT-TL1* with a heteroplasmy rate of 14% in blood [Wang, A. *et al*., 2023]. The patient benefited from empagliflozin and dapagliflozin for diabetes and from coenzyme Q10, vitamin B1, vitamin B2, vitamin B6, niacin, and calcium pantothenate for the mitochondrial disorder (MID) [Wang, A. *et al*., 2023]. The work is compelling, but some points should be discussed.

We disagree with the diagnosis MELAS [Wang, A. *et al*., 2023]. MELAS is diagnosed according to the Hirano criteria or the Japanese criteria. According to the Hirano criteria, MELAS is diagnosed when stroke-like episodes (SLEs) occur before the age of 40 years, and the presence of seizures or dementia, lactic acidosis or ragged-red fibers, normal early development, recurrent headache, or recurrent vomiting [Hirano, M. *et al*., 1992]. According to the Japanese criteria, MELAS is diagnosed when at least two of the criteria A (headache with vomiting, seizures, hemiplegia, cortical blindness, acute focal lesion on imaging) and at least 2 of the laboratory criteria B (elevated serum or cerebrospinal fluid (CSF) lactate, mitochondrial abnormalities on muscle biopsy, MELAS-related gene mutations) are fulfilled [Yatsuga, S. *et al*., 2012]. The patient suffered from a causative MELAS variant, diabetes, hypoacusis, lactic acidosis, ischemic stroke, hypertrophic cardiomyopathy, and ragged-red fibers on muscle biopsy [Wang, A. *et al*., 2023]. He never had a SLE [Wang, A. *et al*., 2023]. The focal lesion on imaging did not correspond to a stroke-like lesion (SLL), the morphological equivalent of a SLE, but was rather ischemic in nature [Wang, A. *et al*., 2023]. Therefore, he did not meet the Hirano nor the Japanese criteria for MELAS. The strongest argument against MELAS is that the patient has never experienced SLE, which is pathognomonic for MELAS.

There is a discrepancy between the statement that the patient had no history of arterial hypertension and the current medication, which included metoprolol. Did he take the beta-blocker for arrhythmias, heart failure, or another indication?

The patient had experienced chest tightness for two years prior to presentation [Wang, A. *et al*., 2023]. What was the reason why he did not undergo a cardiology examination sooner?

Since the history was only positive for diabetes and chest tightness, we should know why the patient was still taking aspirin, clopidogrel, atorvastatin, and metoprolol.

Cardiac MRI revealed an abnormal signal in the anterior wall [Wang, A. *et al*., 2023]. Did it correspond to dyskinesia, hypokinesia, or akinesia on echocardiography? What was the nature of this lesion? Was it fibrosis, a previous myocardial infarction, or an artifact?

The patient underwent a lactate stress test, which was found to be abnormal [Wang, A. *et al*., 2023]. What protocol was used to perform the lactate stress test, specifically did he exercise at a fixed load or at a fraction of his maximum possible load? We should also know whether the lactate stress test used has been validated. What was the sensitivity and specificity of the test?

Overall, the study has limitations which challenge the results' interpretation. Addressing these limitations could further strengthen and reinforce the statement of the study. Specified criteria should be met for the diagnosis MELAS,

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guidelines. This article is based on previously conducted studies and does not contain any new studies with human participants or animals performed by any of the authors.

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