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Protect the MELAS Heart not Only through Cardiac Examination, but also by Clarifying the Multisystem Component

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LETTER TO THE EDITOR

We were interested to read the article by Arriola-Montenegro, *et al*. on a 30-year-old man with mitochondrial encephalopathy, lactic acidosis and stroke-like syndrome (MELAS) due to the mtDNA variant m.3243 A>G in MT-TL1 with a heteroplasmy rate of 53% who underwent successful heart transplantation (HTX) and renal transplantation (RTX) due to dilated cardiomyopathy and heart failure (ejection fraction (EF) 5-10%) and renal failure [Arriola-Montenegro, J. *et al*., 2024]. Anaesthetic management included propofol, ketamine, midazolam, dexmedetomidine, fentanyl, and quetiapine [Arriola-Montenegro, J. *et al*., 2024]. The study is excellent, but some points should be discussed.

The first point is that we disagree with the diagnosis of apical thrombus [Arriola-Montenegro, J. *et al*., 2024]. More likely than thrombus is that the patient had left ventricular noncompaction (LVNC). LVNC is supported by the fact that it has repeatedly occurred in patients with neuromuscular disorders [Finsterer, J, 2010], including mitochondrial disorders (MIDs) [Zhu, L, 2017]. Therefore, we should know how the thrombus formation in the apex of the heart was diagnosed, whether it was due to heart failure, atrial fibrillation or other causes, and how it was treated. Has the patient received anticoagulants or heparin? Did the patient undergo cardiac magnetic resonance imaging (cirri) with contrast prior to HTX to rule out LVNC? LVNC shows up on cry as hypertrabeculation distal to the papillary muscles and often shows late gadolinium enhancement [Zhu, L. *et al*., 2023]. Was the explanted heart autopsied and was the thrombus documented on inspection or was LVNC present?

The second issue is that no cerebral imaging information was provided [Arriola-Montenegro, J. *et al*., 2024]. Since MELAS is characterized by variable central nervous system (CNS) disorders [Weiduschat, N. *et al*., 2014] and the index patient had mental retardation [Arriola-Montenegro, J. *et al*., 2024], information on the condition of the brain would have been mandatory. The most common and pathognomonic CNS abnormality in MELAS is the so-called stroke-like episode (SLE), which shows up on imaging as a stroke-like lesion (SLL) with typical features on cerebral MRI and PET [Finsterer, J, 2023]. Since most MELAS patients suffer SLLs, we should know whether the index patient also had a history of SLE or whether he suffered SLE after HTX/RTX.

The third point is that family history was not reported. Since mtDNA variants are passed on through the maternal line in up to 75% of cases, we should know whether the family history for MELAS or MID in general was positive or negative. Due to the large phenotypic heterogeneity of MELAS, it should be considered that first-degree relatives who may also be carriers of the mutation may have a completely different phenotype than the index patient.

The fourth point is that no long-term outcome has been reported [Arriola-Montenegro, J. *et al*., 2024]. Since some of the immunosuppressive drugs administered for life after HTX/NTX are mitochondrial toxic, we should know the immunosuppressive regimen, whether the patient tolerated these drugs and whether the immunosuppressive regimen had to be changed due to side effects.

In summary, this interesting review has some limitations that put the results and their interpretation into perspective. Taking these limitations into account could strengthen the conclusions and increase the validity of the review. Prior to surgery, MELAS patients need to be screened for multisystemic disease, as brain or endocrine organ involvement strongly influences the outcome of these patients.

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