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# BEFORE ATTRIBUTING MALNUTRITION IN MELAS TO SUPERIOR MESENTERICA ARTERY SYNDROME, ALL DIFFERENTIALS MUST BE EXCLUDED

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## KEYWORDS

MELAS, stroke-like episode, malnutrition, gastroparesis, superior mesenterica artery syndrome

## LETTER TO THE EDITOR

We read with interest Horna et al.'s article about a 22-year-old male with mitochondrial encephalopathy, lactic acidosis and stroke-like episode (MELAS) syndrome, which was suspected to have manifested with superior mesenteric artery syndrome (SMAS) leading to severe malnutrition[1]. Home parenteral nutrition increased the reduced body mass index to 17 after 12 months[1]. It was concluded that SMAS is a rare manifestation of MELAS, and that adequate energy supply is the mainstay of treatment[1]. The study is impressive, but several points require discussion.

The first point is that the diagnostic criteria by which MELAS was diagnosed were not stated. Was MELAS diagnosed according to the Hirano or Japanese criteria? MELAS in the index patient began in childhood and therefore relatively early[1]; MELAS usually starts in the second decade of life. What was the initial clinical presentation of MELAS?

The second point is that the genetic cause of MELAS has not been reported[1]. Was MELAS due to an mtDNA or nDNA mutation? If it was due to an mtDNA variant, what were the heteroplasmy rates in the affected tissues?

The third point is that no family history was provided[1]. Did MELAS occur sporadically, or was it inherited? When due to an mtDNA variant, it can be inherited in up to 75% of cases[2]. Was the family history positive for mitochondrial disorders? In particular, was the index patient's mother clinically affected or not? Have any of the first-degree relatives undergone genetic testing?

A fourth point is that perimyocarditis is an unusual phenotypic feature of MELAS[2]. We should know whether myocarditis was diagnosed by cardiac MRI or by endomyocardial biopsy, and whether it was classified as immune myocarditis or infectious myocarditis. Myocarditis has rarely been reported in association with MELAS.

The fifth point is that alternative causes of hyponatraemia have not been sufficiently ruled out. In addition to medications, hyponatraemia in MELAS can also be due to pituitary dysfunction, suprarenal dysfunction, renal insufficiency or be nutritionally related. Have pituitary adenoma, hypocorticism, adrenal adenoma and renal involvement been appropriately excluded? Was there decreased salt absorption due to the gastrointestinal involvement? Which anti-seizure drug (ASD) was said to have caused hyponatraemia, valproic acid, eslicarbazepine, lamotrigine, lacosamide or carbamazepine?

The sixth point is that vomiting in the presence of

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gastroparesis is incomprehensible. Vomiting requires contraction of the stomach. Was the gastroparesis temporary or incomplete? Is it conceivable that vomiting was due to a central nervous system cause? Did the patient have a stroke-like episode (SLE) while vomiting? SLEs can manifest with vomiting[3]. Did the patient experience ketoacidosis or lactic acidosis?

A seventh point is that the rationale for treating the patient with L-arginine is not supported by evidence. There are no adequate studies demonstrating that L-arginine is effective in preventing progression of MELAS, preventing SLEs or reducing the disease severity. Data on L-arginine in MELAS are based on case reports, case series, uncontrolled studies or meta-analyses[4,5].

A final point is that no long-term follow-up was provided. Did the patient lose weight again after stopping home parenteral nutrition?

In summary, the excellent study has limitations, which complicate the interpretation of the results. Addressing these limitations could strengthen and reinforce the statement of the study. Before malnutrition in MELAS can be attributed to SMAS, alternative causes such as depression, psychosis, SLEs, medications, malabsorption, poor nutrition, pancreatitis, hepatopathy or diarrhoea must be thoroughly ruled out.

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