Correspondence Arrhythmias in MELAS syndrome

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Letter to the Editor

With interest we read the article by Thomas et al. about a 44 year old female with MELAS syndrome due to the m.3243A>G mutation, which manifested cardiologically as hypertrophic cardiomyopathy and episodes of supraventricular tachycardia requiring atenolol [1]. We have the following comments and concerns.

MELAS may be associated with sudden cardiac death [2]. Did the authors consider implantation of a loop-recoder to monitor if there were also ventricular arrhythmias requiring implantation of an ICD?

The authors describe hypertrophic cardiomyopathy as progressive [1]. What was the systolic function and wall thickness at the last follow-up? Figure 1B suggests that there is noncompaction of the left ventricular myocardium [1]. Did the authors look for this unclassified cardiomyopathy, which may occur in addition to hypertrophic cardiomyopathy, goes frequently along with late gadolinium enhancement [3], and is most frequently associated with mitochondrial disorders? [4]

A heteroplasmy rate of 25% of the m.3243A>G mutation is low. In which tissue was it determined? Blood, muscle, saliva, or urine? Were other tissues investigated for the heteroplasmy rate as well? Was a muscle biopsy carried out?

Recurrent stroke-like episodes (SLEs) are reported during the 11 years since diagnosis [1], which are a clinical manifestation of cerebral stroke-like lesions [5]. How many SLEs did the patient experience, in which cerebral region, and why could the remnants not be seen on cerebral MRI, which only showed atrophy and leucencephalopathy?

Concerning the initial fall, did the patient loose consciousness, did she experience a tongue bite, or did she experience a secessus urinae/alvi? What were the results of the EEG? Did the patient always experience seizures during SLEs? Were antiepileptic drugs administered? Were other causes of the fall excluded?

Overall, this interesting case should be supplemented by results about the heteroplasmy in more than a single tissue, long-term ECG results, and the medication she was taking at last follow-up.

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