Maternally Inherited Diabetes and Deafness (MIDD) Due to the m.9155A>G Mutation

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LETTER TO THE EDITOR
In a recent article, Adema et al. [1] reported about a 28 years old white female with maternally inherited diabetes and deafness (MIDD) due to the mutation m.9155A>G in the ATP6 gene [1]. In addition to MIDD the patient manifested clinically with renal insufficiency, arterial hypertension, and hypercholesterolemia [1]. We have the following comments and concerns.

The patient is reported to have been treated with a statin over years despite the presence of muscle cramps [1]. Why was the statin not discontinued earlier than after diagnosing MIDD? It is well established that statins may induce muscular manifestations or may trigger or enhance myopathy, why it should be avoided in case of muscular manifestations, including muscle cramps.

MIDD may not only affect the pancreas and the inner ear but also other organs, such as the heart (hypertrophic cardiomyopathy, non-specific cardiomyopathy) [2], the kidneys [1,3], other endocrine organs (hypothyroidism) [4], the eyes (cataract, pigmentary retinopathy) [5], or the cerebrum (cognitive dysfunction, cerebellar ataxia and atrophy, white matter changes, basal ganglia calcification, elevated CSF lactate) [6]. Did the patient undergo echocardiography and long-term ECG to exclude or confirm cardiac involvement? Was cerebral imaging carried out to look for central nervous system involvement in MIDD? Patients with a mitochondrial disorder (MID) due to a novel mutation should be extensively investigated to learn most about the clinical manifestations and the phenotypic variability between different mutation carriers.

MIDD patients may develop lactic acidosis [7].Were serum lactate levels measured, and was serum lactate elevated? In case of normal serum lactate, a lactate stress test could be performed for diagnostic purposes [8]. Did serum lactate levels increase upon mild standardised exercise? Though most treating physicians take a nihilistic standpoint concerning treatment of MIDD, it must be stressed that various symptomatic and supportive measures may alleviate symptoms and signs of MIDD patients. Additionally, patients with a MID may profit from administration of co-factors, vitamins, or antioxidants. Which type of treatment was applied to this particular MIDD patient? Listing of the last medication with its dosages is missing or did the patient receive only the antidiabetic treatment?

Overall, this interesting case merits application of more widespread investigations to exclude or confirm multiorgan involvement, provision of the drugs the patient was taking at his last visit to see which drugs had been prescribed, and if any of these compounds had a mitochondrial-toxic effect.

REFERENCES

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