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Glucocorticoids in MELAS

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In a recent article Choi et al. described a 22-year-old male with MELAS syndrome due to the mutation m.3243A>G, who experienced recurrent stroke-like episodes (SLEs) for which he received glucocorticosteroids (steroids) [1]. The authors pretend that steroids caused severe side effects (osteonecrosis) in this patient [1]. We have the following comments and concerns.

We do not agree with the diagnosis of avascular necrosis (AVN) [1]. The patient had diabetes and arterial hypertension, at least two risk factors for atherosclerosis and thus macro- or microangiopathy. Osteonecrosis of the femoral head is a well-established complication of diabetes. How do the authors exclude that the hip complication was rather due to diabetes than a side effect of steroids? Did the patient have micro- or macro-angiopathy in other vascular territories?

It is unusual to prescribe steroids in a patient with diabetes. Which was the reason why the patient nonetheless received steroids? Evidence that steroids are beneficial in SLEs is poor and it is well-established that steroids in mitochondrial disorders (MIDs), particularly Kearns-Sayre syndrome (KSS) can be detrimental [2]. Did the patient alternatively receive L-arginine for his SLEs?

The sentences "During the last 8 months, he had three consecutive SLEs" and "One month ago, the patient was admitted for eighth SLE" [1], are contradictory. What is really meant? How many SLEs were truly registered in the last three months prior to admission?

Another cause of osteonecrosis, as described in the presented patient, could be osteoporosis. Since MIDs are frequently associated with endocrine abnormalities, including hypo- or hyperparathyroidism, it would be interesting to know if calcium phosphate, parathormone, or the calcitonin serum levels were normal. Was ever osteoporosis diagnosed in the described patient?

Steroids themselves induce myopathy. Did the patient also manifest in the skeletal muscle? Did he undergo needle EMG or muscle biopsy? Did pre-existing myopathy deteriorate upon administration of steroids?

According to the case description, SLEs were diagnosed only clinically and not by magnetic resonance imaging (MRI) [1]. How can the authors be sure that SLEs were truly SLEs and that they correlated with imaging findings? Were EEGs recorded during the SLEs?

SLEs may be triggered by seizures and require treatment with antiepileptic drugs (AEDs). Did the patient receive AEDs during the many SLEs reported? Was he on a permanent AED treatment? Which AEDs were applied? Were mitochondrion-toxic AEDs avoided? Patients with epilepsy have an increased risk of bone fracture, either due to the seizures, or due to the AEDs of which some increase the risk of osteoporosis. Which AEDs were applied and since when did the patient take these AEDs?

Nothing is reported about the family history of the index case. In the majority of the cases, however, MELAS is maternally inherited. Were any of his first-degree relatives clinically affected?

Overall, this interesting case requires more widespread considerations and investigations concerning the cause of his hip fractures. Seizures, endocrine involvement, and side effects of AEDs need to be excluded as alternative causes of hip fractures before making corticosteroids responsible for the problem. The family history needs to be outlined.

Key Words: Mitochondrial, mtDNA, phenotype, genotype, lymphocytes, MELAS, lactic acidosis,

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