Journal of Medical Biomedical And Applied Sciences
Doi : 10.15520/jmbas.v6i10.158
Received : 1 October 2018
Accepted : 4 October 2018

Do Focal Seizures Trigger Bilateral Stroke-Like Episodes Which Manifest As Transient Cortical Deafness?

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Keywords
mitochondrial DNA, epilepsy, seizures, electroencephalogram, MELAS, antiepileptic drugs, stroke-like episode, antioxidants

Letter to the Editor

In a recent article, Pittet et al. reported about an 11 years old male with mitochondrial encephalopathy, lactic acidosis, and stroke-like episodes (MELAS) syndrome due to the tRNA(Leu) mutation m.3243A>G in whom a stroke-like episode (SLE) with stroke-like lesions (SLLs) in the temporal lobes bilaterally manifested with bilateral, transient cortical deafness[1]. We have the following comments and concerns.

Since seizures are generally regarded as a trigger of SLEs we should be informed about the phenomenology of the nocturnal focal seizures prior to the onset of deafness. Was the phenomenology of nocturnal seizures the same as that of the focal seizures occurring before the current event? Did the patient experience SLEs already before and how did they manifest clinically? Which types of antiepileptic drugs (AEDs) did the patient receive for seizure control? Valproic acid, carbamazepine, phenytoin and phenobarbital are regarded as mitochondrion-toxic[2]. Did he receive any of these AEDs and was the dosage changed shortly before the current event? Were less mitochondrion-toxic AEDs, such as ethosuximide, oxcarbazepine, topiramate, felbamate, zonisamide, lamotrigine, levetirazetam, or gabapentine ever tried and in which dosage? Did the individual history reveal another trigger for the nocturnal seizures, such as fever, infection, sleepiness, or flickering light? Did focal seizures recur during the SLE and did the patient receive an AED treatment in addition to L-arginine and citrulline and the AEDs he was regularly taking? Which was the dosage of citrulline? Since epilepsy was classified as refractory we should be informed if a ketogenic diet was ever tried, if implantation of a vagus stimulator was considered, and if epilepsy surgery was planned? Which AEDs have been tried for how long so far and why was epilepsy classified as refractory? Was the family history positive for epilepsy, was the individual history positive for birth trauma, meningitis, traumatic brain injury, or childhood epilepsy? For how long was L-arginine administered?

In the acute stage, SLEs usually manifest on MRI as vasogenic edema (hyperintensity on DWI and hyperintensity on ADC) but occasionally also with cytotoxic portions (hyperintensity on DWI and hypointensity on ADC)[3]. Which were the findings on the ADC maps and which were the MRI findings at the follow-up MRIs?

Since MELAS follows a maternal trait of inheritance in two thirds of the cases[4], it would be interesting to know if the patient had inherited the mutation from his mother and if any other family members were affected by MELAS? Was the mother or other relatives tested for the m.3243A>G mutation?

Revision of the cerebral MRI suggests that the subcortical insular region was also affected by the SLE[1]. Did the patient experience any arrhythmias during the SLE? Since MELAS may also manifest as dilative or hypertrophic cardiomyopathy, Wolff-Parkinson-White syndrome, AV-conduction defects, or intra-ventricular conduction defects[5], the authors should present the results of the cardiologic investigation during the SLE and prior to the SLE.

Overall, this interesting case merits a comprehensive description of epilepsy management, a more profound family screening for MELAS and epilepsy, and alternative treatment of refractory seizures. The patient might also profit from administration of a cocktail of vitamins, cofactors, and antioxidants.

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