Oxidative stress is an aggravating factor in Lafora rare disease

22 May 2015

Neurodegenerative Lafora disease usually becomes apparent through seizures during adolescence and puberty and occurs as a consequence of defects in glycogen metabolism and in the cellular mechanisms that are responsible for its disposal. Researchers at the University of Valencia have led a study in which they propose that Lafora could be aggravated by oxidative stress. These ideas have been put forward in a review article recently published in the journal *Free Radical Biology and Medicine*.

Researcher Carlos Romá-Mateo, one of the lead authors of this paper, explains that oxidative stress is "an aggravating factor for the molecular processes in charge of eliminating useless substances, which are damaged in Lafora disease. This is emerging as an interesting field of study to fully understand the complex etiology of this disease."

This study is linked to a previous investigation in which these same authors proved that the levels of cellular oxidative stress were high in models of human cells and experimental animals for Lafora disease, and they proposed mitochondrial damage as a possible cause for these high levels of oxidative stress.

Romá-Mateo comments that under normal conditions, defective mitochondria are eliminated by cellular mechanisms, which are affected in Lafora disease. Consequently, as noted by this scientist of the University of Valencia, "the relationship between the defects to eliminate aberrant substances and increased oxidative stress that we observed may reflect the consequences of mitochondrial damage accumulated". This argument has been presented in the article published in the special issue of *Free Radical Biology and Medicine*, dedicated to rare diseases and oxidative stress.

More information: "Oxidative stress, a new hallmark in the pathophysiology of Lafora progressive myoclonus epilepsy." *Free Radic Biol Med*. 2015 Feb 10. pii: S0891-5849(15)00043-X. DOI: 10.1016/j.freeradbiomed.2015.01.034

"Increased oxidative stress and impaired antioxidant response in lafora disease." *Mol Neurobiol.* 2015 Jun;51(3):932-46. DOI: 10.1007/s12035-014-8747-0

Provided by Asociacion RUVID