RAGE expression in motoneurons from human ALS

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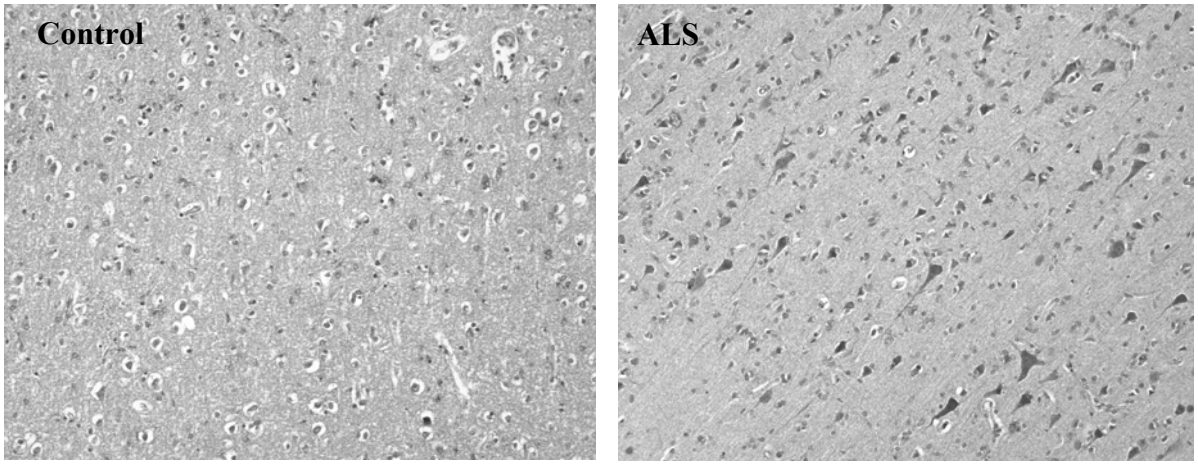
Amyotrophic lateral sclerosis (ALS) is a neurodegenerative disorder resulting in the progressive loss of motoneurons and during which neuroinflammation occurs. RAGE (Receptor for Advanced Glycation Endproducts) is a multiligand member of the immunoglobulin superfamily of cell surface molecules which binds S100A6 and S100B [1]. S100A6 and S100B differentially modulate cell survival by interacting with distinct RAGE Ig domains [2]. RAGE signalling induces, through $NF_{\kappa}B$, the production of pro-inflammatory cytokines. In this work we have looked at the RAGE expression within the human spinal cord of ALS patients. In human frontal cortex, RAGE is not or weakly expressed on normal human motoneurons but highly expressed in motoneurons from ALS patients. Considering that S100A6 and S100B are also overexpressed in the CNS from ALS patients [3], our results reinforce the concept that neuroinflammation and RAGE are playing an important role during ALS ethiopathology.



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