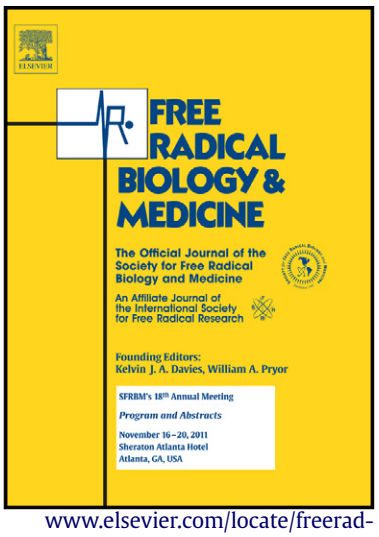

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Endoplasmic Reticulum Stress and Proteasomal System in Amyotrophic Lateral Sclerosis

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Abstract

Protein processing including folding, unfolding and degradation is involved in the mechanisms of many diseases. Unfolded protein response and/or endoplasmic reticulum stress areaccepted to be the first steps which should be completed via protein degradation. In this direction, proteasomal system and autophagy play important role as the degradation pathways and controlled via complex mechanisms. Amyotrophic lateral sclerosis is a multifactorial neurodegenerative disease which is also known as the most catastrophic one. Mutation of many different genes are involved in the pathogenesissuch as superoxide dismutase 1, chromosome 9 open reading frame 72 and ubiquilin 2. These genes are mainly related to the antioxidant defense systems, endoplasmic reticulum stress related proteins and also protein aggregation, degradation pathways and therefore mutation of these genes cause related disorders.This review focused on the role of protein processing via endoplasmic reticulum and proteasomal system in amyotrophic lateral sclerosis which are the main players in the pathology. In this direction, dysfunction of endoplasmic reticulum associated degradation and related cell death mechanisms that are autophagy/apoptosis have been detailed.

Keywords

ER stress, unfolded protein response, proteasome, amyotrophic lateral sclerosis