

Activity and expression of glutathione S-transferase pi in patients with amyotrophic lateral sclerosis.

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Glutathione S-transferase (GST, EC 2.5.1.18) is an enzyme responsible for inactivation of a large variety of toxic, electrophilic compounds and organic peroxides. GST activity and GST pi expression were studied in patients with amyotrophic lateral sclerosis (ALS). Studies were conducted on cerebrospinal fluid (CSF), blood serum and peripheral blood mononuclear cells (PBMC) obtained from 40 ALS patients. CSF from 30 subjects without neurological diseases and blood from 40 healthy blood donors were used as controls. GST activity assayed with 1-chloro-2,4-dinitrobenzene (substrate for transferase activity) and cumene peroxide (substrate for peroxidase activity) was significantly decreased in PBMC of ALS patients, as well as the GST pi expression on both mRNA and protein level. The mean peroxidase activity was however significantly increased in CSF and serum of ALS patients with the specificity of 80% and 73%, and the sensitivity of 78% and 85%, respectively. It can thus be concluded that the protective barrier formed by GST is originally affected in peripheral blood of ALS patients, and may increase their vulnerability to toxic effects of electrophilic compounds and organic peroxides. Studies on a larger group are needed to answer the question whether GSH-Px determination may be implicated in the diagnosis of ALS.

