

Neuropathological Findings in Essential Tremor

S. CHATZIKONSTANTINO¹, D. KAZIS¹, F. PETRIDIS¹,
I. MAVROUDIS^{2, 3, *}

¹ Third Department of Neurology, Aristotle University of Thessaloniki, Thessaloniki, Greece

² Department of Neurology, Leeds Teaching Hospitals, Leeds, UK

³ University of Cyprus, Medical School

* Corresponding author e-mail: ioannis.mavroudis@gmail.com

Abstract.

In this paper we are mainly describing some recent and relevant neuropathological findings in essential tremor, considering its pathophysiological importance, by mainly referring to Purkinje cells' pathology and also on the Lewy bodies in the brainstem.

Keywords: Essential Tremor, Purkinje cells' pathology, Lewy bodies in the brainstem.

Introduction

Essential tremor (ET) is a chronic progressive, neurological syndrome of heterogeneous clinical phenotypes, clinically characterized by involuntary tremor on hands or arms and progressively on head, jaw, and voice (1-4), while some patients manifest more extensive and complex deficits (5-10).

The pathophysiology of essential tremor is not clearly defined. Many studies on familial cases of essential tremor have proposed a genetic predisposition, provoked by certain DNA mutations (11-14). Postmortem neuropathology studies have revealed certain alterations at the histological and cellular level, in brain tissues of affected patients. Various neuropathological findings have been reported in ET. It is unclear which findings are well established, since not all of them reflect large series of patients. However, according to recent meta-analyses of the neuropathological evidence on ET, these could be categorized under two main groups; the first regarding Purkinje cell changes and the second the presence of Lewy bodies in the brainstem. Further sub-categorization is possible, in these main groups (15).

Apart from these main hypotheses, some studies have investigated the role of the dentate nucleus in the pathogenesis of ET. Severe neuronal loss and atrophy, microglial clusters, and reduction in the number of efferent fibers, along with reduced GABAA and GABAB receptors, have been reported.