Post-polio syndrome and amyotrophic lateral sclerosis - similarities, differences and diagnostic dilemmas.

Ewa Matyja, MD, PhD; Milena Laure-Kamionowska, MD
Department of Neuropathology, Mossakowski Medical Research Centre, Polish Academy of Sciences, Warsaw, Poland

Post-polio syndrome (PPS) and amyotrophic lateral sclerosis (ALS) are neurodegenerative disorders characterized by motor neurons (MNs) loss in the spinal cord, brain stem and motor cortex. The distinction between PPS and ALS bears important prognostic implications as ALS is a rapidly progressive lethal disease with a low survival rate, whereas PPS is regarded to be a slowly progressive syndrome. Both these motor neuron diseases (MNDs) still remain a challenge for clinical and scientific community and require better understanding of their etiology and treatment efficacy.

The patients with post-polio muscular atrophy (PPMA) develop clinical symptoms similar to the onset of ALS with weakness and muscular atrophy due to upper and/or lower MNs involvement, occasionally accompanied by respiratory or swallowing problems. Although the neuropathological features consisting of MNs loss in both PPS and ALS are similar, the histopathological details are different. Neuropathology in PPS has been reported in a few autopsy findings and revealed MNs loss of anterior horn of the spinal cord with gliosis and inflammatory B cells infiltrates, without corticospinal tracts involvement. In human sporadic and familial form of ALS a variety of neuronal inclusions are observed as Lewy-like, Bunina bodies and hyaline or ubiquitin-immunoreactive inclusions, which are not typical for PPS. The signs of inflammation in ALS cases were reported only sporadically.

The etiopathogenesis of ALS and PPS is not fully understood and an involvement of different pathological factors related with progressive MNs stress, accompanied by age dependent risk factors ought to be considered. Moreover, about 1% of patients with a history of paralytic polio have been reported to develop ALS as coincidental findings. The histopathological diagnosis in such cases is very difficult as features of PPS and/or ALS are developing on the background of primary spinal lesions of poliomyelitis. The diagnosis of PPS and ALS is based on exclusion clinical criteria since there were not specific tests and biomarkers for confirmation of diagnosis.

The basic principle of management address to both PPS and ALS patients is mostly supportive and require individually tailored training programs and lifestyle modification. However, due to the differences in the rate of natural progression of these MNDs, the management in ALS patients is more aggressive and include respiratory assistance and supplemental nutrition at early stages of the disease.

Key words: PPS and ALS; differential diagnosis.