Electrophysiological characteristics of spastic paraplegia (SP)

Affection of sensory systems, peripheral nerves and upper extremities

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Background:

Hereditary spastic paraplegia is characterised by progressive spastic gait disturbance due to degeneration of the corticospinal tract. Disease is typically restricted to the legs. Sensory affection and peripheral neuropathy occur as complicating symptoms.

Objective:

To characterize sensory and peripheral system deficits in SP and determine the frequency of upper extremity affection.

Design/Methods:

Clinical and electrophysiological analyses were performed in a cohort of 97 SP patients. Sensory deficits were stated when vibration sense, joint position sense, temperature or pinprick discrimination were impaired. Peripheral neuropathy was assumed when muscle wasting or loss of stretch reflexes were observed. Electrophysiological characterization included motor nerve conduction studies of tibial and ulnar nerves, sensory nerve conduction studies of sural and radial nerves and motor evoked potentials (MEP) using F-wave technique.

Results:

We found sensory deficits in 59% of patients and clinical signs of peripheral neuropathy in 34%. Nerve conduction studies revealed peripheral neuropathy in 38% of patients. In 21% subclinical affection of the peripheral nervous system was detected.

All patients presented with definite signs and symptoms of pyramidal tract dysfunction in the legs. However, MEPs to the abductor hallucis muscle were abnormal in only 76% of patients. Extensive prolongation of central motor conduction time (>20ms) suggesting primarily demyelinating damage was found in 48% of patients. MEPs to the arms were abnormal in 34% of patients.

Conclusions/Relevance:

Frequent subclinical affection of sensory and peripheral nervous system raises the question whether these patients should be classified as “pure” or “complicated” SP. Further genetic characterisation of our cohort will help to define genotype specific electrophysiological patterns.

Involvement of upper extremities in upper motorneuron disease often leads to classification as primary lateral sclerosis. Since one third of our patients with typical SP including 60% with familial disease had abnormal MEP to the arms, this appears an insufficient criterion.