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Electrophysiological study in patients with Spinocerebellar Ataxia Type 3 (SCA3)

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Currently there is no published article on the neuro-electrophysiological findings of patients with SCA3 in China, the published articles abroad just focused on the central part of the proprioceptive system of somatosensory evoked potential (SSEP) and neglected the peripheral part. Hence, we sought to study the electrophysiological features of patients with SCA3 among the Han Chinese and explore its underlying pathogenesis. We performed clinical and electrophysiological examinations in patients with clinically diagnosed and genetically confirmed SCA3, the electrophysiological examinations comprise the electromyography (EMG), nerve conduction velocity (NCV), motor evoked potential (MEP), somatosensory evoked otential (SSEP) etc. For a few patients, the SSEP examinations consist of both the N20 and N9 in the upper limbs, both the P40 and N8 in the lower limbs. A total of 23 patients with SCA3 (12 males, 11 females, mean age 39.43 ± 8.72 years, mean disease duration 5.04 ± 3.50 years) underwent the clinical and electrophysiological examination. 30% of the SCA3 patients have EMG and NCV abnormalities suggestive of peripheral neuropathy; 80% of the SCA3 patients have SSEP abnormality, more severe in the lower limbs than in the upper limbs, suggesting the proprioceptive sensory system lesion; 50% of the SCA3 patients have MEP abnormality, also more severe in the lower limb than in the upper limb, suggesting the pyramidal tract lesion. Our studies suggest that there are lesions in the peripheral part of the somatosensory conducting system, which may serve as a therapeutic target.

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