A case of progressive supranuclear palsy with cortical myoclonus

Kazuyuki Kawamura, M.D., Yoshiharu Arii, M.D., Toshio Inui, M.D., Takao Mitsui, M.D.

Department of neurology, Tokushima National Hospital, National Hospital Organization, 1354 Shikiji, Kamojima, Yoshinogawa, Tokushima 776-8585 Japan

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Introduction

Progressive supranuclear palsy (PSP), a neurodegenerative disease with the parkinsonism, is characterized by supranuclear eye movement disorders and static reflex disorder appearing in the neck / trunk from early period of muscle rigidity and illness. Myoclonus is the lightning involuntary muscle shrinkage resulting from excessive neurologic excitement. This disease develops with various conditions of a patient including side effects of drugs, nervous diseases and systemic metabolic derangements. In the neurodegenerative disease group with parkinsonism, myoclonus develops with 15-50% frequency for Diffuse Lewy body disease, multiple system atrophy, and corticobasal degeneration; however, it is very rare in PSP [1]. We encountered one PSP patient with cortical myoclonus.

Case report

The patient was a 59-year-old Japanese man. Fortunetelling fall characteristics and sleep disturbance developed at the age of 56, in October, 2010. He came to fall down frequently, and everyday life became difficult subsequently. He was admitted to the psychiatry hospital in August, 2011 (57 years old). After hospitalization, he developed downward gaze palsy, dysarthria, slow movement, muscle rigidity of the trunk and four extremities. He was received at our hospital in February, 2012. There was no-one with similar symptoms in his family and no consanguineous marriage of parents. There were no abnormalities found in a general physical examination. His thoughts were lucid. A mini-mental state examination was 19/30 and frontal assessment battery (FAB) was 9/20. There was no aphasia / senseless deed / agnosia. He could stand up with assistance. The cranial nervous system was disturbed by dysarthria, dysphagia and external ophthalmoplegia in all directional restrictions. There was no muscle atrophy or muscle weakness of the four extremities, but there was rigidity of the trunk- dominant muscle. The left leg showed a mild tremor. His movements were slow. The sensory system was normal. The tendon reflex of the four extremities was aggravated slightly in the left upper extremity. The Myerson sign and palmomental reflex was positive. The Babinski sign was positive in the left leg. A finger-nose test and a knee-heel test showed mild decomposition of movement and dysmetria. Head MR imaging showed atrophy of the mesencephalic tegmentum, tegmentum of the pons and the frontal lobe, and expansion of the third ventricle. He was hospitalized in our hospital in April, 2012. Jerky tremors of the four extremities intermittently developed in May, 2012. Giant SEP was found in somatosensory evoked potential (SEP) of the median nerve stimulation (Figure 1). After giving clonazepam 1mg/d, the onset frequency of tremor decreased.

Correspondence to: Kazuyuki Kawamura , M.D., Tokushima National Hospital, National Hospital Organization, 1354 Shikiji, Kamojima, Yoshinogawa, Tokushima 776-8585 Japan Phone: +81-883-24-2161 Fax : +81-883-24-8661

Discussion

Myoclonus is very rare in PSP [1]. A case that developed palatal myoclonus with false hypertrophy of the inferior olivary nucleus has been reported. There has been a case in which myoclonus appeared during amantadine use [2,3]. The results of SEP indicated that tremor was cortical myoclonus. Certical myoclonus reflects cerebrocortical hyperirritability. In most cases of PSP which do not present cerebral cortex symptoms, there is an amplitude increase of median nerve stimulation SEP [4,5]. This suggests that cerebrocortical hyperexcitability lurks in PSP. Rarely, myoclonus may be induced during treatment of PSP and Parkinson's disease by amantadine and levodopa [3,6,7]. In these cases, the medicine seems to influence dopamine, serotonin, or glutaminic acid-based conduction of the nerve. As a result, cerebrocortical hyperexcitability is enhanced, and cortical myoclonus is induced. In the present case, there is a possibility that amantadine or levodopa induced myoclonus.

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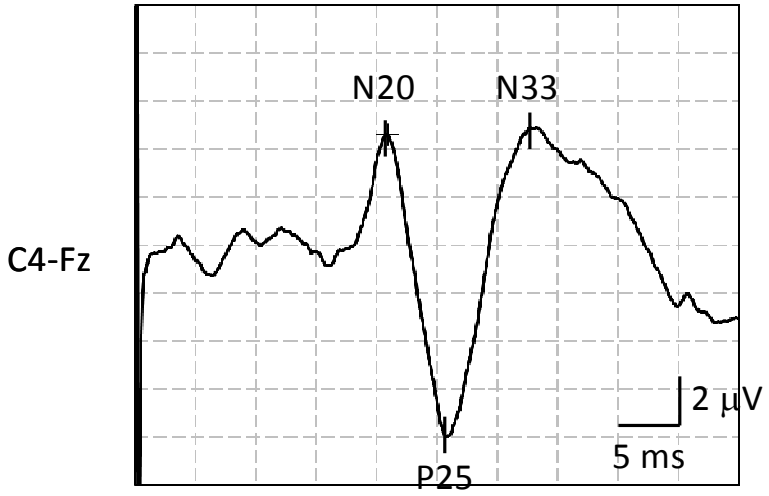

Figure 1. Somatosensory evoked potential of median nerve showing giant potential.