

Effects of short-term intensive rehabilitation admission in a PSP-PGF patient with severe gait freezing

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Abstract

Among various types of progressive supranuclear palsy (PSP), pure akinesia with gait freezing (PAGF) (PSP-PGF, based on MDS-Clinical Diagnostic Criteria for PSP, 2016) causes freezing of gait (FOG) early after onset, markedly reducing activities of daily living (ADL) and the quality of life (QOL). We report a PSP-PAGF patient with severe FOG in whom short-term intensive rehabilitation admission reduced FOG.

[Case] The patient was a 67-year-old male with PSP-PAGF. In 2015, stiffness of the right hand and gait disturbance appeared. In 2016, levodopa-/carbidopa-containing tablets and a Neupro patchTM (rotigotine) were prescribed, but there was no efficacy. Although caregiving was not required in daily living, FOG was marked, making moving difficult. FOG appeared at the start of gait, on changing the direction, in a narrow place, immediately before an object, and even on going straight ahead. Furthermore, FOG exacerbated in the presence of cognitive load or mental stress. The new freezing of gait questionnaire (New FOG-Q) score was 26 points. As a rehabilitation approach, external stimulation (vision/hearing/somatic sensation) was introduced (cue exercise). Learning of external cue methods reduced FOG, decreasing the New FOG-Q score to 22 points. In particular, FOG at the start of gait was relieved. However, FOG appeared in a narrow place or in the presence of mental stress, and it was impossible to adopt external cue in some cases.

[Conclusion] Learning of external cue was effective for FOG in a patient with PSP-PAGF. However, it was impossible to adopt external cue under some circumstances; therefore, comprehensive approaches, such as environmental adjustment and mental support, may be necessary. Tottori J. Clin. Res. 9(1), 38-46, 2017

Key words: progressive supranuclear palsy (PSP), pure akinesia with gait freezing (PAGF) (PSP-PGF), rehabilitation, cue exercise

Introduction

PSP is a neurodegenerative disorder characterized by supranuclear vertical gaze palsy, dysphagia, speech disturbance, impaired postural reflexes, nuchal dystonia, falling-prone features, and frontal cognitive dysfunction. In 1964, it was first reported by Steele, Richardson, and Olszewski¹. In 2005, Williams et al.^{2,3} reported that pathologically diagnosed PSP could be clinicopathologically classified into two subtypes: typical PSP (PSP-Richardson's syndrome: PSP-R), which was described by Steele et al.¹), and PSP-parkinsonism (PSP-P),

which is difficult to differentiate from Parkinson's disease (PD). In 2007, Williams et al.⁴ reported the third subtype of PSP: PSP-pure akinesia with gait freezing (PAGF). In this subtype, pure akinesia, for which L-dopa is ineffective, FOG, or festinant speech appears prior to other neurological signs, persisting for a long period. Furthermore, the morbidity rate is very low. Although pure akinesia had first been reported by Imai and Narabayashi⁵ as a new syndrome "dopa-unresponsive pure akinesia or freezing", which might have been a new disease entity and been discussed as a subtype or an initi-