

Three cases of PSP-PNFA, a rare type of progressive supranuclear palsy

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Abstract

We report on three patients who noticed a tendency to fall while walking. occurred by falling down at a walk. Cognitive functional disorder, lalopathy came to be outstanding with the course. In addition to parkinsonism, word remembrance difficulty, a decrease in own utterances, and prosody disorders were found. Cognitive testing showed a remarkable reduction in the frontal lobe function in all the patients. Atrophy of the mesencephalic tegmentum and atrophy of the frontal lobe were found in the head MRI. Based upon the foregoing, these cases were regarded as PSP-PNFA by clinical courses and results of examination.

Keywords: PSP, stammering symptom, freezing of gait, MRI, SPECT, PSP-PAGF

Introduction

Progressive supranuclear palsy (PSP) is a neurodegenerative disease characterized by fortune-telling fall characteristics in addition to parkinsonism, dementia. It is known that a variety of vocal disorders such as explosive speech or scanning speech developed in PSP. The symptoms of the early period of onset of the PSP patients are varied, and there is a lot of cases to made differential diagnosis from other degenerative diseases.

We encountered three patients who made non-fluency-related utterances in addition to exhibiting parkinsonism. We conducted an examination of the language symptoms..

Case report

Patient 1

67 years old, woman. The tendency to fall

when walking developed at the age of 62. Parkinson's disease was diagnosed, and treatment was started. After the age of 64 old, word remembrance was difficult. MMSE was 28 points, FAB was 12 points, and the word remembrance was five words in one minute. The lalopathy included a fall in the utterance speed, decrease of the own utterance, agrammatism, distortion of articulation, inconsistent articulation errors, and prosody disorders. Speaking was easy in the unconsciousness state.

Patient 2

74 years old, woman. Freezing of gait occurred at the age of 57. PD was diagnosed by a local doctor, and she was treated. Frequent falls, forgetfulness, and lalopathy became common from about 70 years old. MMSE was 21 points, FAB was nine points, and word remembrance was four words in one minute. The lalopathy included a fall of

the utterance speed, a decrease of the own utterance, onset of gibberish aphasia, agrammatism, distortion of articulation, articulation errors, and prosody disorders. Speaking was easy in the unconsciousness state.

Patient 3

67 years old, woman. Freezing of gait occurred at the age of 57. She had a diagnosis of PD. The gait disturbance gradually progressed from 62 years old. Fortunate fall characteristics, hypersalivation, speaking in a low voice, and decrease of the utterance became common. MMSE was 26 points, FAB was ten points, and the word remembrance was five words in one minute. The lalopathy included decrease of the own utterance, and prosody disorders. Speaking was easy in the unconsciousness state.

Discussion

It is known that PSP has five types including Richardson's syndrome, PSP-P, PSP-PAGF (pure akinesia with gait freezing), PSP-CBS (corticobasal syndrome), and PSP-PNFA (progressive non-fluent aphasia) [1]. It was thought that four cases with the stammering symptom belonged to PSP-PNFA or PSP-PAGF. Furthermore, it was necessary to consider the apraxia of speech (AOS). At first we tried to think about the differences between PSP-PNFA and AOS. In contrast, it may be said that AOS shows some of the symptoms of PNFA. On the other hand, only partial symptoms of AOS were present in our four cases. Based upon the foregoing, it was thought that we were accompanied with a language symptom in PSP-PAGF including prosody disorders such as stammering, rhythm disorder and lack of variety. We examine an involved site and the bloodstream fall part according to the type. The SPECT analysis showed there was a significant bloodstream fall in the front part of the cingulate gyrus and the frontal lobe in patients with the stammering symptom. However, a slight bloodstream fall was seen in the same part in patients who did not have

the stammering. The pattern of four bloodstream falls that showed a stammering symptom was similar to the pattern of the involved site of PSP-PNFA and AOS [2-3].

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