Speech Disorders in Progressive Supranuclear Palsy

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Abstract: Stuttering has been defined as a disruption of the fluency of verbal expression characterized by the involuntary repetition or prolongation in the utterance of sounds and syllables. Acquired stuttering in the adult is rare and is usually associated with trauma or vascular disease. However, some patients with extrapyramidal diseases, especially with progressive supranuclear palsy (PSP) have been reported to develop stuttering in their disease course. We evaluated 22 PSP patients whether they developed stuttering or not. We observed 9 patients showed acquired stuttering and discussed pathognomonic mechanism of their stuttering.

Keywords: Progressive supranuclear palsy, stuttering, mini mental scale examination, frontal assessment battery, Montreal cognitive assessment.

INTRODUCTION

Speech disorders, especially stuttering, is a main causative factor for communicative impairment in wide variety of neurological disease and reduced their quality of life. Nontheless, for such importance, stuttering in neurological disorders has rarely been discussed.

Stuttering has been defined as a disruption of the fluency of verbal expression characterized by the involuntary repetition or prolongation in the utterance of sounds and syllables [1, 2]. Acquired stuttering in the adult is rare and is usually associated with trauma or vascular disease [3]. However, some patients with extrapyramidal diseases, especially with progressive supranuclear palsy (PSP) have been reported to develop stuttering in their disease course [4-6].

OBJECTIVE AND METHOD

According to diagnostic criteria of the movement disorder society for PSP [7], we recruited outpatiently 22 PSP patients (75.7±5.8 years old) and were given informed consent for recruitment in this study.

After recoding conversation of patients into Voice Memo (Apple Inc), we analyzed them by its software. Utterance continuance, oral diadochokinesis and reading aloud of sentences were evaluated. We also assessed the cognitive function using the mini mental scale examination (MMSE) [8], the frontal assessment battery (FAB) [9], and the Montreal cognitive assessment (MOCA) [10]. We evaluated severity of PSP by clinical rating scale for PSP [11].

For neuroimaging analyses, we conducted MRI, single photon emission computed tomography (SPECT), and dopamine transporter (DAT) image. MRI features of PSP have known as the hummingbird sign or the penguin sign characterized by a flattening or concave outline to the superior aspect of the midbrain which should be upwardly convex. In addition, PSP patients showed period- or oval-shaped uptakes were seen within the caudate nucleus head without tracer uptake in the putamen on DAT images, and some patients showed decreased uptakes in the anterior part of frontal and temporal lobes on SPECT [12-14].

RESULTS

Recruited 22 PSP patients showed characterized MR images (Figure **1a**). Nine out of 22 PSP patients showed stuttering and all of them had no history of stuttering before they manifested PSP and all of those patients showed decreased uptakes in the anterior part of frontal and temporal lobes on SPECT (Figure **1b**). Dopamine transporter (DAT) images showed decrease reduced isotope uptake in the basal ganglia (Figure **1c**). Specific binding rations in the right and left basal ganglia were 0.41 and 0.31 (normal ratio is above 4.50).

We divided patients into two groups as follows patients with (consistent with 9, mean age±SD: 75.6±8.1 years old) and without stuttering (consistent with 13, mean age±SD: 75.8±3.6 years old). Patient with stuttering showed significantly low total sores in FAB (Table **1**). It is recognised that due to small numbers the power of this study is low, but it is based on the maximum available data. We analysed using the Mann-Whitney U test (MMSE, FAB, and MOCA). Tests were considered significant at the 5% level.

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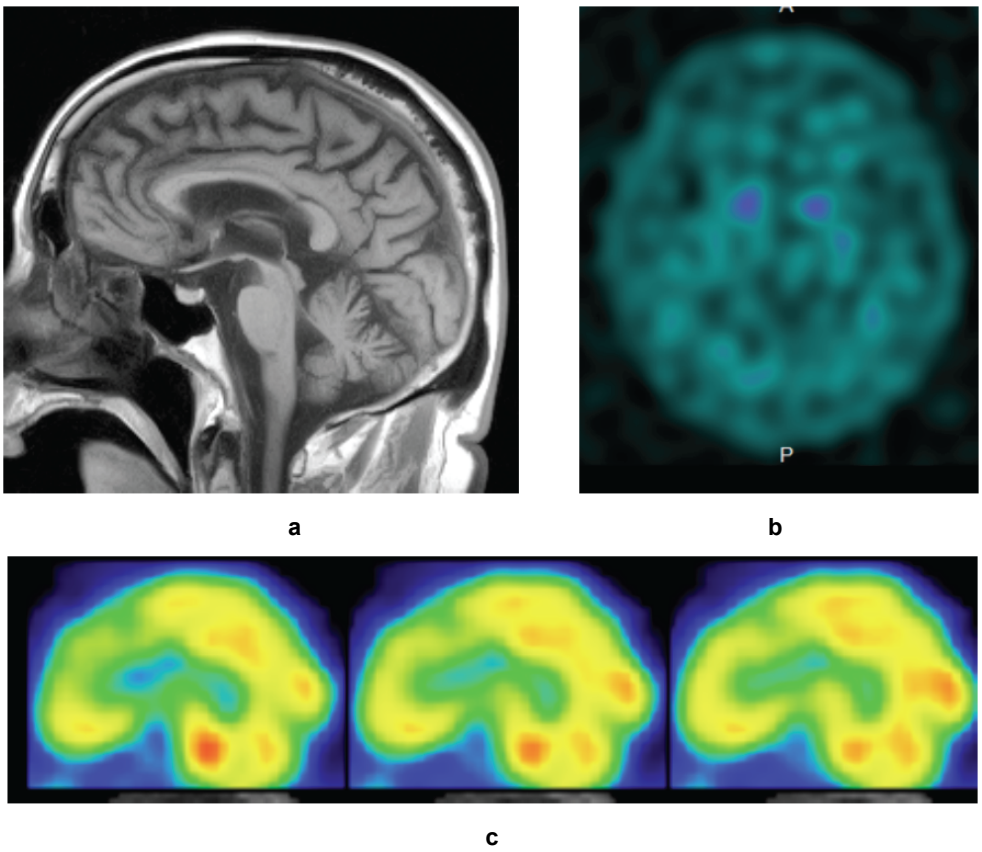

Figure 1: Representative neuroimages of a progressive supranuclear palsy (PSP) patient with acquired stuttering

a. A sagittal MRI showed the hummingbird sign or the penguin sign characterized by a flattening or concave outline to the superior aspect of the midbrain which should be upwardly convex.

b. An axial dopamine transporter (DAT) image showed decrease reduced isotope uptake in the basal ganglia.

c. Sagittal images of single photon emission computed tomography (SPECT) showed reduced isotope uptakes in the frontal lobe.

Table 1: Comparing between Patient with and Without Stuttering Concerning in Total Scores of MMSE, FAB, and MOCA

	PSP patients with stuttering	PSP patients without stuttering
MMSE (/30)	24.2±1.2	25.4±3.1
FAB (/18)	12.8±3.2*	15.9±1.7
MOCA (/30)	21.7±2.8	22.9±3.6

Patient with stuttering showed significantly low total sores in FAB.
*P<0.05.

DISCUSSION

Progressive supranuclear palsy is characterized by decreased cognition, abnormal eye movements, postural instability and falls, as well as parkinsonian features, also and speech disturbances and some PSP patients have been reported to develop stuttering [6, 7, 11].

Stuttering can be defined as a deviation of speech attracting attention of speakers or listeners because of interruption of the normal rhythm of speech by involuntary repetition, prolongation or arrest of sounds. Acquired stuttering, mainly resulting from cerebral infarcts, has been described in disorders in the corpus striatum, the dominant temporal lobe, the dominant parietal lobe, or in the dominant operculum [2, 17, 18]. Although, the pathogenesis of stuttering is considered in relations with extrapyramidal dysfunction, patients with extrapyramidal disease do not always develop stuttering. Thus, another anatomical region might be

caused stuttering in PSP patients. In our patients, patients with stuttering significantly low total scores in FAB and relatively reduced isotope uptakes in the frontal lobe comparing with patients without stuttering. Diminished initiation of conversational exchange is frequently attributable to frontal lobe dysfunction [19, 20] and is consistent with evidence of in our patients. Thus, we consider that a causative lesion for stuttering cannot be restricted to the extrapyramidal system.

There are no differences concerning pyramidal tract signs evaluated by clinical rating scale for PSP between PSP patients with and without stuttering. In addition, Brown and Marsden suggested role of extrapyramidal tract that the motor areas of the cortex (sensorimotor, lateral premotor and dorsolateral prefrontal cortices, and the supplementary motor area (SMA) and cingulate motor areas) may select and group together particular aspects of the distributed neuronal responses related to an intended movement. Thus, the basal ganglia facilitate this process, thereby focusing attention on what is relevant among the plethora of activities going on at any one time [21]. Since some patients with infarcts in SMA, where had connections with extrapyramidal systems, demonstrated uncontrollable interruption of speech or utterance, the SMA might play a significant role both in initiation and in control of spontaneous speech [22, 23]. Our patients with stuttering showed significantly reduced isotope uptake in the frontal lobe including SMA comparing with patients without stuttering that supported causative region for stuttering might include SMA.

Speech disorders including stuttering reduce communication ability of PSP patients. Since communication is indispensable for keep ability of daily livings (ADL) and quality of life (QOL), understanding pathogenesis of stuttering in PSP patients is needed to improve their ADL and QOL.

CONFLICT OF INTEREST

The author confirms that I have no conflict of interest related to the content of this manuscript.

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