Agraphia in Mobile Text Messages in a Case of Amyotrophic Lateral Sclerosis with Frontotemporal Dementia

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Abstract

We herein describe the case of a woman with amyotrophic lateral sclerosis (ALS) showing errors in her choice of Japanese kana characters in her mobile text messages and agraphia of the kana in her handwriting in spite of the absence of weakness, ataxia, or apraxia of her hands. Magnetic resonance imaging showed the atrophy of the frontal lobes. Single-photon emission computed tomography revealed hypoperfusion of the frontal lobes including Exner’s area. Although patients with bulbar-onset ALS have been reported to show agraphia of handwriting, in this case the basis of her agraphia might have been the disturbance of the pathway converting phones to graphemes in series, by which errors of spelling or writing would appear in any modality of output.

Key words: amyotrophic lateral sclerosis, agraphia, frontotemporal dementia

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Introduction

Patients with amyotrophic lateral sclerosis with frontotemporal dementia (ALS-FTD) sometimes show language problems, including agraphia in their handwriting (1) and apraxia of in their speech (2). The agraphia seen in these patients is considered to result from the involvement of the frontotemporal language-related areas of the brain, especially Exner’s area. We herein describe the case of a patient showing agraphia both in handwriting and in mobile text messages.

Case Report

A right-handed 68-year-old woman presented with speech disturbance and dysphagia, which had been deteriorating for three months. Magnetic resonance imaging performed at another hospital detected atrophy of the frontal lobes (Fig. 1). The patient was alert, but her speech showed dysarthria. Her auditory comprehension and reading of written Japanese sentences containing both kanji (ideograms) and kana (syllabograms) were good. Agraphia was found in her handwriting. On neurological examination, there was no evidence of facial palsy or disturbance of ocular movements. Atrophy and fasciculation was observed in her tongue. None of the muscles of her limbs were atrophic, and their strength was normal. No involuntary movement was observed. There was no limb or truncal ataxia. The patient’s superficial and deep sensations were normal, as were her tendon reflexes, including the pathological reflexes. The results for complete blood cell counts, liver and renal function tests, and levels of serum glucose, lipids, thyroid hormones, and creatine kinase were all normal. The patient was negative for anti-acetylcholine receptor antibody. The cell count and the levels of glucose and protein in her cerebrospinal fluid were 5/3 mm³, 67 mg/dL, and 29 mg/dL, respectively. A nerve conduction study revealed normal findings. An electromyography revealed decreased interference in the tongue. Single photon emission computed tomography (SPECT) using ⁹⁹ᵐTc-ethyl cysteinate dimer revealed hypoperfusion at the frontal lobes and the right parietal lobe (Fig. 1).

The standard language tests for aphasia revealed decreased scores for “Writing kana words” (2/5), “Narrative writing” (3), “Dictating kana words” (2/5), and “Dictating sentences” (2/5). Interestingly, the score for “Writing single

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Figure 1. Magnetic resonance images of the patient’s brain (upper panel) and a three-dimensional stereotactic surface projection of single photon emission computed tomography (lower panel). Frontal lobes were atrophic on T1- and T2-weighted images. The areas of hypoperfusion (Z-score > 2) are colored in the lower panel. Exner’s area (the caudal area of the left middle frontal gyrus) was involved in the area of hypoperfusion.

The patient was diagnosed with ALS (progressive bulbar palsy) with FTD. As was noted previously, her agraphia of kana was complicated. Kana errors were found in her mobile text messages as well as her handwriting. When making sentences in mobile text messages, people have only to select kana letters on the display and to press the corresponding buttons as opposed to handwriting where people must remember the configurations of the letters. The kana errors observed in her mobile text messages were not due to weakness, ataxia, or apraxia, because our patient did not show any of these dysfunctions. The errors in her mobile text messages differed from dystypia, which is a typing error that may be made by patients without aphasia, apraxia, agraphia, or visuospatial impairment (3). Since she understood how to use the message functions on her phone, her errors were not due to dementia. Strictly speaking, the errors in the patient’s mobile text communications cannot be described as agraphia, but common mechanisms could have been behind the errors in her handwriting and mobile mail text messages.

In handwriting, there are three successive stages of con-
distinction. The first stage is the selection of the allograph, the appropriate font and case of the letter to be written (allographic system). The second stage is the selection of the direction, relative size, position, and order of strokes. The third stage is the execution and control of neuromuscular commands. Disturbance of spelling and writing is divided into central processes (‘sublexical’, or ‘phonological’ and ‘lexical’ routes) which appear in any modality of output (handwriting, oral spelling, typing, etc.) and peripheral processes which are specific to one particular output modality (4). In this patient, the “phone to grapheme” pathway could have been normal, since the score of “Write single kana letters” was perfect. However, there may have been disturbance in how the patient was treating kana letters in series. This discrepancy between kana dictation and kana word writing has been noticed in ALS-FTD patients (5). There is a feature of agraphia in Japanese ALS patients. Although kanji agraphia has been reported, kana agraphia is predominant in ALS-FTD patients. The most frequent writing error was the omission of kana letters whereby she produced a partial response. The second most frequent error was a phonological error that involved the substitution of kana letters and the usage of incorrect syntax (1, 5). Our patient’s kana agraphia might have resulted from converting disorder from a disorder in her ability to convert from phones to graphemes brought about by an inability to treat words as single phones, possibly due to a dysfunction of the central processes. Hypoperfusion of the left frontal lobe, which includes Exner’s area, might have been responsible for her errors in handwriting and mobile text messages (6, 7).

In the advanced stage, ALS patients cannot avoid using communication tools. In patients with agraphia, it would be difficult to use such tools because of difficulties in the selection of letters.

The authors state that they have no Conflict of Interest (COI).
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