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Hyperlexia and ambient echolalia in a case of cerebral infarction of the left anterior cingulate cortex and corpus callosum

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We report the case of a 69-year-old woman with cerebral infarction in the left anterior cingulate cortex and corpus callosum. She showed hyperlexia, which was a distinctive reading phenomenon, as well as ambient echolalia. Clinical features also included complex disorders such as visual groping, compulsive manipulation of tools, and callosal disconnection syndrome. She read words written on the cover of a book and repeated words emanating from unrelated conversations around her or from hospital announcements. The combination of these two features due to a focal lesion has never been reported previously. The supplementary motor area may control the execution of established subroutines according to external and internal inputs. Hyperlexia as well as the compulsive manipulation of tools could be interpreted as faulty inhibition of preexisting essentially intact motor subroutines by damage to the anterior cingulate cortex reciprocally interconnected with the supplementary motor area.

Keywords: Hyperlexia; Ambient echolalia; Anterior cingulate cortex; Corpus callosum; Cerebral infarction.

INTRODUCTION

The term hyperlexia was first defined and used in the early research of Silberberg and Silberberg (1967) to describe children with highly developed word recognition skills but little or no comprehension of the words recognized. It was defined as an isolated ability in reading words within a context of severe language delays, inappropriate or absent interpersonal relations, and behavioral fragmentation (Goldberg, 1987). Case studies reported hyperlexia to be more prevalent in children with pervasive development disorders including autism

(Elliot & Needleman, 1976; Goldberg & Rothermel, 1984; Huttenlocker & Huttenlocker, 1973; Mehegan & Dreifuss, 1972). There are only a few reports about 'acquired' hyperlexia (Assal, 1985; Furumoto, Matsuda, & Yoshiyama, 1996; Hadano, Sakata, Tanaka, Hamanaka, & Toda, 1987; Suzuki et al., 2000; Vuilleumier, Staub, & Assal, 1997), some of which are related to diffuse brain dysfunction (Hadano et al., 1987; Vuilleumier et al., 1997) or bilateral brain lesion (Assal, 1985). No case of 'acquired' hyperlexia has been reported to be related to a focal unilateral region of infarction.

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Fisher (1988) introduced another phenomenon known as 'ambient echolalia', in which demented patients borrow words or phrases emanating from unrelated conversations around them or from a TV program. He explained that incoming words might automatically drive the speech apparatus in mentally deteriorated patients. Ambient echolalia has been reported only rarely (Della Sala & Spinnler, 1998; Endo et al., 2001; Fisher, 1988; Hadano et al., 1987), and this phenomenon with hyperlexia due to a focal lesion has never been reported.

We report here a patient with a cerebral infarction in the left anterior cingulate cortex and corpus callosum, who read words present in front of her and also repeated words emanating from unrelated conversations around her.

CASE REPORT

A 69-year-old right-handed Japanese woman with grade 9 education working at the front desk of a public bathhouse was admitted with numbness in her right arm and leg and some difficulty in walking. She had a past history of hypertension and diabetes. She was alert and fully oriented. She had no diminished spontaneous speech and her speech comprehension was good. The degree of right hemiparesis was slight and it improved within 2 weeks. In addition to visual groping (Seyffarth & Denny-Brown, 1948), compulsive manipulation of tools (Mori & Yamadori, 1982) was noted in her right hand. Recognizing these tendencies, she tended to restrain the movement of the right arm by holding it with the left arm. Ideomotor apraxia and agraphia were noted in her left hand which indicated callosal disconnection syndrome.

Brain magnetic resonance imaging (MRI) conducted on the day following the onset revealed the presence of an acute infarct in the left anterior cingulate cortex and the left trunk of the corpus callosum with some involvement of the subcortical superior frontal gyrus (Figure 1a-c). A technetium-99m-ethyl cysteinate dimer (99mTc-ECD) single-photon emission computed tomography (SPECT) conducted 1 month after the onset demonstrated a decrease in regional cerebral blood flow (rCBF) in the left frontal region in accordance with the MRI findings (Figure 1d-f).

A neuropsychological evaluation (Table 1) revealed neither aphasia nor dementia to be present. She scored 24 points on the Japanese Mini-Mental State Examination (MMSE) (Mori,

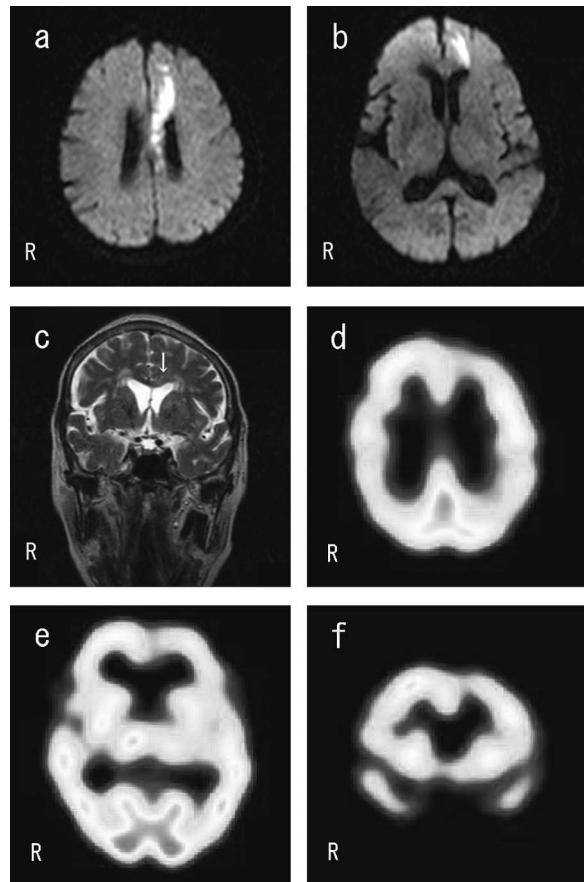


Figure 1. (a, b) Diffusion-weighted axial section magnetic resonance imaging (MRI) scan demonstrating a circumscribed lesion localized to the left anterior cingulate cortex with some involvement of the subcortical superior frontal gyrus. (c) T2-weighted coronal section MRI scan demonstrating a hyperintense lesion in the left trunk of the corpus callosum (arrow). (d, e, f) Technetium-99m-ethyl cysteinate dimer (99mTc-ECD) single-photon emission computed tomography (SPECT) showing perfusion defects in accordance with MRI findings.

Mitani, & Yamadori, 1985), which indicated no cognitive impairment. Her verbal and performance intelligence quotient (IQ) based on the Japanese Wechsler Adult Intelligence Scale-Revised (WAIS-R) (Shinagawa, Kobayashi, Fujita, & Maekawa, 1990) were in the low average range: 89 and 82, respectively. She did particularly poor regarding digit span, object assembly, and digit symbol. She performed within one standard deviation of the mean on all items excluding word fluency in the Standard Language Test for Aphasia (SLTA) (Nirayama Conference, 1975). On Japanese Wechsler Memory Scale-Revised (WMS-R) (Sugishita, 2001), she obtained scores of 93 and 62 for the verbal and visual memory, respectively,

TABLE 1
Neuropsychological profile

| <i>Neuropsychological test</i> | <i>Performance</i> | <i>Percentile/range</i> |
|--------------------------------|--------------------|------------------------------------|
| MMSE (Japanese) | 24 ^a | No cognitive impairment |
| WAIS-R (Japanese) | | |
| Information | 8 ^b | 25th percentile |
| Digit span | 6 | 9th percentile |
| Vocabulary | 8 | 25th percentile |
| Arithmetic | 8 | 25th percentile |
| Comprehension | 9 | 37th percentile |
| Similarities | 11 | 63rd percentile |
| Picture completion | 10 | 50th percentile |
| Picture arrangement | 7 | 16th percentile |
| Block design | 9 | 37th percentile |
| Object assembly | 4 | 2nd percentile |
| Digit symbol | 6 | 9th percentile |
| Verbal IQ | 89 | Low average |
| Performance IQ | 82 | Low average |
| Full-scale IQ | 85 | Low average |
| WMS-R (Japanese) | | |
| General memory | 80 ^c | 9th percentile |
| Verbal memory | 93 | >25th percentile <37th percentile |
| Visual memory | 62 | <1st percentile |
| Attention and concentration | 65 | 1st percentile |
| Delayed recall | 91 | >25th percentile <37th percentile |
| SLTA | | |
| Auditory word comprehension | 10 ^a | =Mean* |
| Sentence comprehension | 10 | >Mean |
| Follow verbal commands | 10 | >Mean |
| Kana letter discrimination | 10 | =Mean |
| Object naming | 20 | >Mean |
| Word repetition | 10 | >Mean |
| Describe behaviors | 10 | >Mean |
| Explain picture story | 6 | >Mean |
| Sentence repetition | 5 | >Mean |
| Word fluency (listing) | 7 | <Mean – 1 standard deviation |
| Read aloud kanji words | 5 | =Mean |
| Read aloud kana words | 10 | =Mean |
| Read aloud kana letters | 5 | =Mean |
| Read aloud short sentences | 5 | >Mean |
| Kanji word-picture matching | 10 | >Mean |
| Kana word-picture matching | 10 | =Mean |
| Sentence picture matching | 9 | >Mean – 1 standard deviation <Mean |
| Follow written commands | 10 | >Mean |
| Write kanji words | 5 | >Mean |
| Write kana words | 5 | >Mean |
| Narrative writing | 5 | >Mean – 1 standard deviation <Mean |
| Dictate kana letters | 10 | >Mean |
| Dictate kanji words | 5 | >Mean |
| Dictate kana words | 5 | >Mean |
| Dictate short sentences | 5 | >Mean |

MMSE, Mini-Mental State Examination; WAIS-R, Wechsler Adult Intelligence Scale-Revised; IQ, Intelligence quotient; WMS-R, Wechsler Memory Scale-Revised; SLTA, Standard Language Test for Aphasia.

^aRaw score. ^bScaled score. ^cIndex score. *Mean of 150 non-aphasics.

thus indicating dissociation between verbal and visual memory.

Her most outstanding behavioral trait was hyperlexia. She read words written on the cover of

a book or parts of information written on bulletin boards placed in front of her during interviews and test preparations. For example, when the examiner tried to conduct a test and showed her an evaluation

manual, she suddenly whispered, 'Wais Wais'. This is because she had responded to the words 'WAIS-R Intelligence Test for Adults, Japanese version' written on the title page of the evaluation manual. She was aware of her hyperlexia, saying, 'I try to avoid looking at anything with words because I read them aloud when I see them'.

Ambient echolalia, i.e., a tendency to repeat words emanating from unrelated conversations was also present. This phenomenon occurred when she heard conversations between other patients and therapists during therapy and parts of hospital announcements heard from loudspeakers, but she did not repeat any words of conversations or questions directed toward her. She did not express anguish while repeating any words, and she did not simply repeat words she had heard one after another. For instance, when she was asked, 'How long have you been hospitalized?', she answered, 'Since September 27', and added 'ICU' after a pause. This is because she had repeated a part of the hospital announcements heard through the speaker ('Family of Mr. or Ms. XX, please come to the ICU room').

DISCUSSION

Our patient presented with a peculiar type of hyperlexia. She had a tendency to read words present in front of her and repeated words emanating from unrelated conversations around her. She was aware of the hyperlexia and tried to prevent it by avoiding looking at anything with words written on it. The callosal disconnection syndrome in this case was very similar to a case with apraxia (which may be caused by a mesial frontal lobe injury) and agraphia of the left arm described by Liepmann and Maas (1907) in which the callosal lesion would disconnect language from the right hemisphere while also disconnecting the left hemisphere motor engrams from the right hemisphere motor areas.

The compulsive manipulation of tools could be a mesial frontal form of alien hand sign, which was interpreted as faulty initiation, execution, and inhibition of preexisting essentially intact motor subroutines (Goldberg, Mayer, & Toglia, 1981). This form of alien hand sign was postulated to be dissociation between will and action (Archibald, Mateer, & Kerns, 2001). The alien hand sign was often contrasted with utilization behavior (Lhermitte, 1983), which results from an imbalance

between proposed medial and lateral motor systems. In both conditions, patients perform actions elicited by objects in the environment even though these actions are inappropriate in the wider context. However, patients with alien hand sign report the actions as unintended and complain that the movement of their hand is out of their control, whereas patients with utilization behavior make no such complaint (Pacherie, 2007).

Her hyperlexia as well as the behavior of the right hand is an alien hand sign feature. These were unintended actions, and she complained that her speech was out of her control. Yamadori (2000) explained that the compulsive manipulation of tools and hyperlexia were both interpreted in terms of the rivalry between the top-down process of inhibition and bottom-up process of stimulus processing, resulting in the predominance of the latter. The process of hyperlexia in our case may therefore be identical to that of the right hand behavior.

Before the present report, acquired hyperlexia had only been previously described in a few cases in the literature (Assal, 1985; Furumoto et al., 1996; Hadano et al., 1987; Suzuki et al., 2000; Vuilleumier et al., 1997). The clinical features of six cases with acquired hyperlexia are summarized in Table 2. This phenomenon was caused by various diseases, however, the medial frontal lobe involvement including the anterior cingulate cortex seemed to be a common finding (4/6 cases). Aphasia occurred in 4/6 cases, all with impaired intelligence. Ambient echolalia was present in the case of dementia, as well as in our case. Behavioral disorders in either the manipulation or utilization of objects was frequent (4/6 cases).

Usually, the term hyperlexia is applied to children with developmental disorders, and it is characterized by superior word reading ability with deficits in comprehension and spontaneous speech. Nevertheless, there are cases of acquired hyperlexia absent from development and comprehension disorders. This indicates that the cause of 'acquired' hyperlexia is different from 'developmental' hyperlexia. A dysfunction of the frontal lobe (Furumoto et al., 1996) or the left supplementary motor area (Suzuki et al., 2000) is believed to be the cause of hyperlexia, but few reports have explained the detailed mechanism of acquired hyperlexia.

The lesion in our case was limited to the left anterior cingulate cortex and corpus callosum. The neuropsychological evaluation demonstrated the absence of dementia and aphasia. The clinical features were restricted to callosal disconnection

TABLE 2
Clinical cases of acquired hyperlexia

| Author | Age | Sex | Diagnosis | Reported sites of lesions | Intelligence | Aphasia | Ambient echolalia | Other clinical features |
|---------------------------|-----|-----|----------------------------|--|----------------------------|-------------------------------|-------------------|--|
| Present reported case | 69 | F | Cerebral infarction | L ACC L SFG L CC | WAIS-R VIQ 89 PIQ 82 | – | + | Visual groping Compulsive manipulation of tools |
| Suzuki et al. (2000) | 69 | F | Post operative brain tumor | L ACC L SMA L CC | WAIS-R PIQ 61 | Transcortical motor aphasia | – | Ideomotor apraxia |
| Vuilleumier et al. (1997) | 65 | M | Post-anoxic encephalopathy | – | Severe dementia | Anomia | – | Manual groping Compulsive sniffing Manipulation of tools Utilization of tools |
| Furumoto et al. (1996) | 66 | F | Cerebral hemorrhage | L ACC L SFG L MFG L IFG L CC | MMSE 9 | Transcortical sensory aphasia | – | Utilization behavior |
| Hadano et al. (1987) | 77 | F | Dementia | – | Dementia | Mixed transcortical aphasia | + | Constructional apraxia |
| Assal (1985) | 37 | F | Cerebral infarction | B MFL R STG R MTG | WAIS-R VIQ 85 PIQ 90 | – | – | Imitative behavior Utilization behavior |

M, male; F, female; R, right; L, left; B, bilateral; +, present; –, absent; ACC, anterior cingulate cortex; SFG, superior frontal gyrus; CC, corpus callosum; SMA, supplementary motor area; MFG, middle frontal gyrus; IFG, inferior frontal gyrus; MFL, medial frontal lobe; STG, superior temporal gyrus; MTG, middle temporal gyrus; WAIS-R, Wechsler Adult Intelligence Scale-Revised; VIQ, verbal intelligence quotient; PIQ, performance intelligence quotient; MMSE, Mini-Mental State Examination.

syndrome and compulsive disorders such as compulsive manipulation of tools. Although the anterior cingulate cortex has prominent projections to motor systems, its contributions to human behavior have been elusive because isolated cingulate lesions are rare, and spontaneous lesions of the anterior cingulate cortex, such as those observed in tumors and strokes, almost always involve adjacent areas (Devinsky, Morrell, & Vogt, 1995). The supplementary motor area may contribute to the establishment of new motor programs and control the execution of established subroutines according to external and internal inputs. This could be the area where external inputs and commands are matched with internal needs and drives to enable the formulation of a strategy of voluntary movements (Larsen, Skinhej, & Lassen, 1978). Hyperlexia as well as dissociative psychomotor disturbances, such as compulsive manipulation of tools, could be interpreted as a faulty inhibition of preexisting essentially intact motor subroutines by damage to the anterior cingulate cortex reciprocally interconnected with the supplementary motor area.

It was a remarkable and unusual finding that ambient echolalia was associated with hyperlexia in our case. A deterioration of speech (Hadano et al., 1987) or a lack of inhibition of the automatic repetition of verbal stimuli captured in the environment (Della Sala & Spinnler, 1998) was believed to cause ambient echolalia. Our patient repeated surrounding voices but did not repeat the utterances of persons with whom she was speaking. Usually, echolalia occurs in response to questions directed from a person facing the patient, who shows no interest toward auditory verbal stimuli that are completely ‘neutral’ to themselves. However, our case responded only to ‘neutral’ stimuli. The phenomenon observed in this case is believed to be an example of stimulus-bound behavior that may be associated with hyperlexia.

In conclusion, we herein described a case with cerebral infarction with a peculiar type of hyperlexia. Ambient echolalia was also present. The hyperlexia in our case could be interpreted as a faulty inhibition of preexisting essentially intact motor subroutines due to damage of the anterior

cingulate cortex, which was reciprocally interconnected with the supplementary motor area.

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