Alexia without agraphia associated with right occipital lesion

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SUMMARY A 78 year old, right handed man developed the syndrome of alexia without agraphia due to a right occipital thrombotic stroke. The cerebral dominance test strongly suggests that his right hemisphere is dominant. This is believed to be the first case of alexia without agraphia secondary to a right occipital lesion in a right handed person.

Alexia, the selective impairment in the ability to read words, or even individual letters, has been repeatedly described in association with left dominant hemisphere lesions. A variety of reading disability in association with right non-dominant hemisphere lesions has also been reported in the literature (Paterson and Zangwill, 1944; Kinsbourne and Warrington, 1962). According to the disconnection theory which has been applied to explain the syndrome of alexia without agraphia (Dejerine, 1892; Geschwind, 1965), the left dominant visual cortex is destroyed with involvement of the splenium of the corpus callosum in a right handed person. This paper describes a right handed patient with probable thrombotic right occipital lesion who showed the syndrome of alexia without agraphia.

Case report

A 78 year old, right handed man, a retired principal, with a past history of mild hypertension and exogenous obesity, found himself to have paraesthesia in his left limbs after a two hours nap. He also felt weak in the left leg, but was able to use his legs and arms freely. Shortly after this he found himself unable to read a newspaper so he was admitted to Kanazawa Medical University Hospital for investigation.

The family history revealed that his mother, his two children, and two grandsons were left handed. On examination the vital signs were normal except for mild hypertension (150/90 mmHg). Neurological examination showed a right handed man with normal

speech, repetition, and understanding. He was alert, and oriented in three spheres. His fundi showed normal optic discs with arteriolar narrowing. He had a left homonymous hemianopia by formal visual field testing. Extraocular movements were full without any fixation problems. His opticokinetic response was slightly diminished when he watched a test flag drawn toward his right. He showed very mild left hemiparesis with arm drift.

Sensory examinations were quite normal for pain, temperature, vibration, and position sense. Higher cortical sensation, such as two points discrimination and stereognosis, was intact. He did not show any neglect of double simultaneous stimuli. His deep tendon reflexes were rather hypoactive but the left plantar response was equivocal. No cerebellar incoordination was noted.

Clinical tests of higher cortical functions are described below. He was alert, and fully oriented. His intelligence was normal for his age. His conversation was fluent, and his language comprehension was acceptable for his educational level. He could repeat spoken language very well. On the other hand, he was unable to read at all including Kanji (ideogram), Kana-Hiragana (phonogram), alphabet, and numbers. He was able to write spontaneously and to take dictation with poor penmanship but he could not later read what he had written nor identify it as his own writing. He had difficulty in copying words and made many mistakes.

His remote memory was good but recent memory retention was somewhat poor for three digits backwards and forwards. Colour naming was disturbed with 30–50% accuracy. He could draw a straight line between two dots. Object naming on confrontation was also poor but he was able to name objects easily

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by touching them. Verbal calculation was quite normal. He did not have finger agnosia or right-left disorientation. Spatial disorientation and constructional apraxia were not seen.

Laboratory examinations showed normal complete blood counts and urinalysis except for a trace of sugar in the urine. Liver function tests were normal as were serum electrolytes. A glucose tolerance test showed a typical diabetic pattern. Lumbar puncture revealed no cells with elevated protein (1.0 g/l) in the CSF which was under normal pressure. An EEG showed some theta slowing over the right parieto-occipital region, and the photic driving was quite asymmetrical, being seen only in the left parieto-occipital leads (Fig.). A technetium brain scan showed an equivocal uptake over the right parieto-occipital area. The cerebral dominance test by Tsunoda's tapping method was strongly suggestive of right hemisphere dominance.

Discussion

This 78 year old, right handed man showed clinical evidence of typical alexia without agraphia, secondary to a right occipital thrombotic stroke.

Benson and Geschwind (1969) reviewed the 17 reported cases of alexia without agraphia with autopsy findings. In all of them, with only one exception (Gloning *et al.*, 1955), the left occipital lobe was involved. In the exceptional case, Gloning *et al.* (1955) strongly suggest that the reading difficulty was produced by visual-spatial distortion, and was not

the same as that seen in pure word blindness produced by left occipital pathology.

A variety of reading disability associated with a right non-dominant hemisphere lesion is reported by Kinsbourne and Warrington (1962). Certainly optic axis distortion secondary to a non-dominant hemisphere lesion has been described in the past (McFie *et al.* 1950; Bender and Jung, 1955). But in our case, there was nothing to suggest visual fixation problems nor optic distortion. Our patient could not read letters, words, or numbers at the beginning of the illness. Within three weeks, he started to read a little by using his fingers to trace the outline of each letter in the air. This type of reading by non-visual stimulation was reported as early as 1877 by Charcot, and was strongly emphasised in the study of Goldstein and Gelb (1928). Somaesthetic reading in alexia without agraphia is well documented in the literature.

The present patient has shown no other disturbances in perceiving his environment or using language. He was unable to read at all including Kanji (ideogram), Kana-Hiragana (phonogram), alphabet, and numbers. The dissociation of Kanji and Kana processing, with much better Kanji performance, has been recognised as a characteristic of aphasia among Japanese (Asayama, 1914; Imura, 1943). Integration of visual and auditory-oral systems is necessary for decoding and encoding phonographic character, whereas ideograms are less bound to direct auditory-oral control, and can be well processed within the visual system. This explains the characteristic in Japanese aphasics.

Figure EEG showed theta slowing over the right parieto-occipital area and asymmetrical photic driving, being almost absent over the right occipital region.

The explanation for alexia without agraphia is a disconnection between those areas responsible for visual semantic processing and those responsible for motor activity. In pure word blindness, the necessary written information is not even received in the dominant hemisphere for linguistic processing. Therefore, there is usually no dissociation of Kanji and Kana-Hiragana processing in Japanese patients with alexia without agraphia.

Our patient had difficulty in identifying colours properly and this has also been well documented in cases of alexia without agraphia. From the above descriptions, our patient displayed the syndrome of alexia without agraphia in essentially pure form. He had a left homonymous field defect which was rather congruous. All the neurological examinations indicated that he had a right occipital lesion. The brain scan and EEG changes support this localisation.

To explain his alexia without agraphia in association with right occipital lobe pathology, his dominant hemisphere should be on the right side. It is of interest that there is a strong family history of left handed members, including his mother, two children, and two grandsons.

To determine the cerebral dominance, we used Tsunoda's tapping method (1971, 1975) which is based on the delayed auditory feedback tapping method devised by Chase (1959). Tapping patterns such as four continuous taps with interruption followed by two successive taps (.... .) are practised before testing. The patient is asked to tap rhythmically a key which is connected to the trigger system of a sound source. When he taps, he can hear through the earphone the interrupted sounds triggered by his tapping. Meanwhile the tapping motion is recorded on paper. While there is no delay between the key action and sound, no disturbing effect can be observed in his rhythmically tapping patterns. When the test tone is suddenly delayed as much as 200 milliseconds, he finds disturbing effects on his own key tapping.

Various sounds, such as pure tones, white noise, buzzer sounds, and human vowels were used by Tsunoda to determine the dominant cerebral hemisphere. He found that non-verbal sounds lateralised in the right hemisphere, and vowel sounds, such as 'ah', in the left hemisphere. Tsunoda and Oka (1971), tested 92 normal subjects to determine their cerebral dominance. They found that 79.3% of the subjects had left side dominance for vowel and verbal sounds, and right side dominance for the pure tone. Left side dominance for the pure tone and right side dominance for vowel and verbal sounds was found in 7.6% of subjects. These figures in normal controls are similar to the results by Wada's test and Kimura's dichotic listening method.

In the present case, this test strongly suggested that

the right hemisphere was dominant for vowel and verbal sounds. Hence it is considered that this patient's dominant hemisphere was probably the right hemisphere.

Tadanobu Tsunoda, MD, Assistant Professor, Tokyo Medico-Dental University, Medical Research Institute, kindly performed the tapping test on this patient.

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