

Pure alexia caused by separate lesions of the splenium and optic radiation

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Abstract Pure alexia is severe difficulty in reading and understanding written language but with normal oral language and writing abilities. We report a patient with pure alexia caused by two different infarct lesions in the left lateral thalamus and the left splenium of the corpus callosum. A 56-year-old right-handed man was admitted to hospital with right homonymous hemianopia associated with pure alexia. He could write kana characters but not kanji. His cranial magnetic resonance imaging revealed two different infarct lesions in the left optic radiation and the left splenium of the corpus callosum. Magnetic resonance angiography showed mild stenosis at the origin of the right vertebral artery and stenosis of the left distal posterior cerebral artery. The mechanism of developing pure alexia can be simply explained by disconnection. We assumed that agraphia of kanji was caused by the effect of ischemia and edema following transient obstruction in branches from the distal posterior cerebral artery.

Keywords Alexia · Stroke · Agraphia · Cerebral infarction

Introduction

Pure alexia is severe difficulty in reading and understanding written language but with normal oral language and writing abilities. Since the first report by Dejerine [2], many cases have been described. Most cases had involvements in the medial surface of the occipital lobe and the corpus callosum. Greenblatt [4] reported a patient with pure alexia who had a subcortical lesion in the left angular gyrus without occipital impairment, and another who had a lesion inferolateral to the posterior horn of the lateral ventricle. Both reports described pure alexia caused by a single lesion, and those associated with multiple involvements are rare [13]. We report a patient with pure alexia caused by two different infarct lesions in the left optic radiation and the left splenium of the corpus callosum.

Case report

The patient was a 56-year-old right-handed man who was a manager of his own woodworking shop with 12 years of school education. He had history of diabetes and hyperlipidemia and had been treated with oral medication.

On July 15, 2009, sudden difficulty in seeing developed in the right eye, but it disappeared in 1 h. The patient did not visit hospital. He noticed abnormal vision in the afternoon of the following day because everything looked dim even when looking at bright light. In the evening of the same day, he became aware of an abnormal sensation on the right side of the body. The next day, he visited a local practitioner with continued symptoms. Medical treatment was initiated with a diagnosis of cerebral infarction. He was referred to our hospital for further investigation on July 27.

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Neurologically, he was alert on admission. There was no abnormality in cranial nerves, including eye movement, except for right homonymous hemianopsia. Slight motor paralysis was present in the right upper and lower limbs, and mild sensory disturbance in the right upper limb.

Neuropsychological findings

His oral language was normal. He had fluent spontaneous speech without paraphasia, and could correctly name objects and repeat spoken words. His auditory comprehension was also normal. However, he had difficulty in reading aloud and understanding both written kanji and kana, even single characters. He could read aloud and understand letters or words only by tracing them with his fingers (kinesthetic facilitatory effect). Paraphilia was observed in writing kanji, but transcription ability was intact. His letters looked better and smoother when he wrote with his eyes closed compared with when writing with his eyes open. The performance in the writing task of the Standard Language Test of Aphasia was perfect, but he could only write up to 60% of kanji words (Fig. 1). His oral reading score was poor in both kana and kanji. There are 102 kana and 240 kanji characters that are required learning in the first and second grade at elementary school in Japan. The rate of correct answers in oral reading was 42.2% (43/102) for kana and 37.1% (89/240) for kanji. In contrast, the rate of correct writing was clearly different at 97.1% (99/102) for kana and 87.9% (211/240) for kanji. Color anomia was observed despite good performance in color recognition and color matching. Although Mini Mental State Examination (MMSE) decreased to 17/30, Raven's Colored Progressive Matrices score and Frontal Assessment Battery score were good at 32/36 and 16/18, respectively. The Wechsler Adult Intelligence Scale third edition (WAIS-III) score was 95 for verbal IQ (VIQ), 68 for performance IQ (PIQ), 81 for full IQ (FIQ), 88 for verbal comprehension (VC), 75 for perceptual organization (PO), 85 for working memory (WM), and 52 for processing speed (PS). His visual field defect and sensory disturbance were affected by his low PIQ score. The Wechsler Memory Scale Revised (WMS-R) score was 54 for verbal memory, 72 for visual memory, 54 for general memory, 92 for attention and concentration, and 54 for delayed recall. Additionally, he found it difficult to remember, because he could not conjure up images of letters.

Neuroradiological findings

Magnetic resonance imaging (MRI) of the head revealed infarct lesions in the left optic radiation near the lateral

geniculate body, the corona radiate, and the left splenium of the corpus callosum (Fig. 2). MR angiography (MRA) of the head and neck performed on admission revealed mild stenosis at the origin of the right vertebral artery and stenosis of the left distal posterior cerebral artery. There was no stenosis or obstruction in the internal carotid artery system.

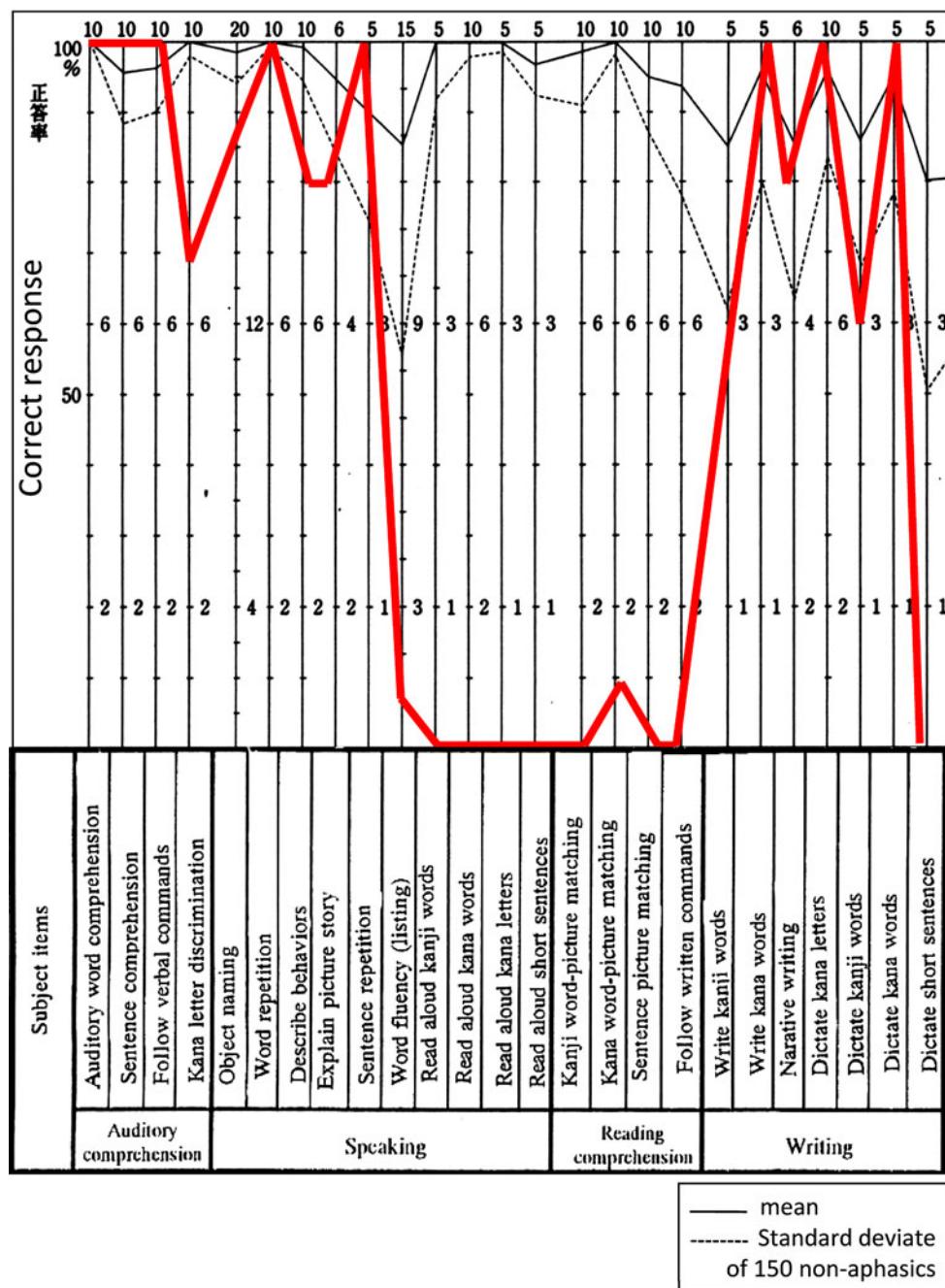
Clinical course

The patient was discharged home 2 weeks after treatment by intravenous drip infusion and oral medication. He was independent in activities of daily living and returned to work about 3 months after the onset. Difficulty in writing kanji disappeared, but the alexia symptom was still persistent. When he read the Bible in his leisure time, he could read it aloud at normal speed but could not understand the meaning. He had to stop reading it once and repeat the sentence he had just read in his mind over and over for comprehension.

Discussion

Classical pure alexia previously reported by Dejerine [2], which accompanied right homonymous hemianopsia due to damage to the inferomedial surface of the left occipital lobe, disturbed the ability to read visualized letters by disconnecting visual information from the right occipital lobe on its way to the left angular gyrus with injury in the splenium of the corpus callosum [3]. On the other hand, no cases of pure alexia reported by Greenblatt [4] accompanied right homonymous hemianopsia, and the author estimated that visual information from the left occipital lobe and that from the right occipital lobe, conveyed through the corpus callosum, were disconnected before reaching the angular gyrus. In the present case, seeing that visual information from the contralateral side was interrupted by damage to the splenium of the corpus callosum in addition to the disturbance of the optic radiation near the lateral geniculate body, the patient could be a subtype of classical pure alexia with different lesions. The two lesions were circumscribed to the subcortex, suggesting development of ischemia in the pulvinar and splenium, branches from the lateral posterior choroidal artery. Pure alexia is classified into two groups on the basis of lesion localization [10]. One of the types, called “splenium type,” involves extensive lesions including the medial occipital gyri (or optic radiation, lateral geniculate body) and splenium (or callosal radiation, forceps major). The other type, called “nonsplenium type,” involves damage to the mid-fusiform gyri, or posterior fusiform or inferior occipital gyri. According to this classification, our

Fig. 1 Standard Language Test for Aphasia (SLTA) 8 days after stroke onset. His oral language was normal, but he had difficulty in reading aloud and understanding both written kanji and kana. Performance in the writing task of the Standard Language Test of Aphasia was perfect, but he could only write up to 60% of kanji words

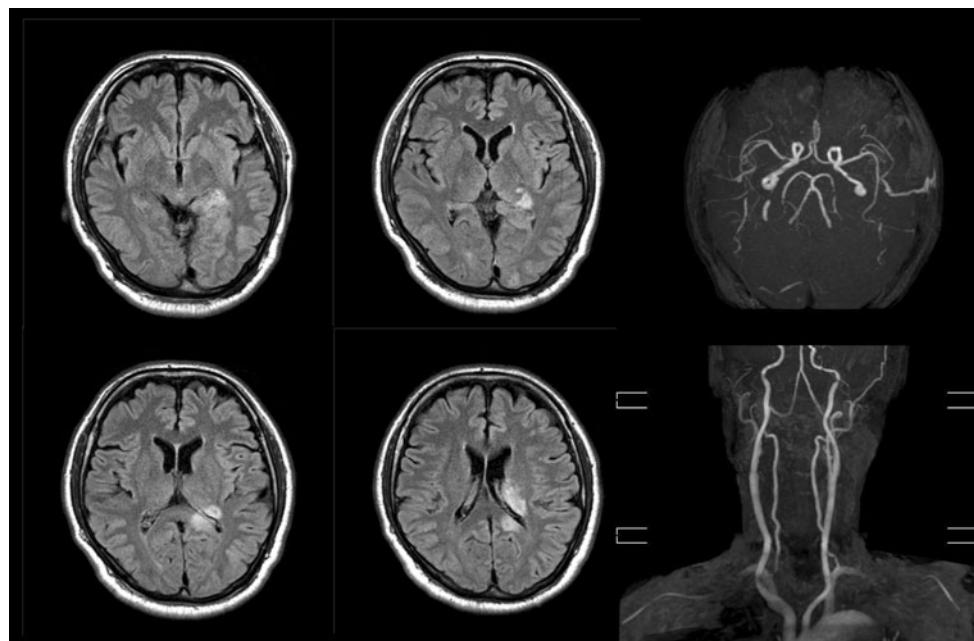


case is the “splenium type”. As described in this case, pure alexia caused by two distinct lesions disconnecting the callosal fibers and visual information to the primary visual cortex is extremely rare [5, 12].

Anatomical substrates of nonaphasic alexia and agraphia have recently been reported [10]. The difference between pure alexia and alexia with agraphia, and between pure agraphia and alexia with agraphia is a relative one; pure alexia may have some degree of agraphia, and pure agraphia may have some degree of alexia. Additionally, with

regard to Japanese patients with pure alexia, the degree of defects differs between kana and kanji. Two neural circuits run between the visual area and Wernicke’s area: one is the dorsal neural circuit passing through the angular gyrus, and the other is the ventral neural circuit passing through the posteroinferior temporal lobe. In the process of reading, the dorsal neural circuit is considered to be responsible for phonological reading and the ventral neural circuit for semantic reading [6]; in large part, the dorsal circuit enables reading of kana and the ventral circuit enables

Fig. 2 Magnetic resonance imaging (MRI) of the head revealed infarct lesions in the left optic radiation near the lateral geniculate body, the corona radiate, and the left splenium of the corpus callosum (left and center). MR angiography of the head and neck performed on admission revealed stenosis at the origin of the right vertebral artery (right lower) and stenosis of the left distal posterior cerebral artery (right upper)



reading of kanji. Sakurai et al. [12] recently described that patients with pure alexia whose lesions were confined to the fusiform gyrus showed remarkable defects in reading kanji rather than kana, while most of those with lesions confined to the posterior occipital lobe behind the fusiform gyrus showed selective reading defect for kana. The present case developed pure alexia for both kanji and kana because of damage to visual information pathways en route to the fusiform gyrus. The mechanism of developing pure alexia can be discussed not only in the context of disconnection, but also of visuoperceptual disorder [1]; nonetheless, it is interesting that this case can be simply explained by disconnection.

Our patient exhibited agraphia specific to kanji at the early stage. Agraphia selective for kanji can develop with involvement of the left posteroinferior temporal lobe [7, 8, 11], and agraphia of kanji is commonly seen in pure alexia of the Japanese language. Since the inferior temporal gyrus in the temporal lobe plays a role in form vision [9], kanji information, which is strongly related to form vision, is processed here. Iwata [6] hypothesized the process of writing Japanese letters, whereby the memory image of movement used to write the letter is evoked, corresponding to the auditory memory of the word of interest via the angular gyrus, in writing kana. For kanji writing, the visual memory image of the letter is recalled via the ventral neural circuit, and then the kanji letter is visually composed based on this visual memory image via the dorsal neural circuit. This patient might have transient obstruction in branches from the distal posterior cerebral artery. We assumed that the effect of ischemia and edema was more extensive in the acute stage.

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