

# Sign language aphasia due to left occipital lesion in a deaf signer

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## ABSTRACT

Localization of sign language production and comprehension in deaf people has been described as similar to that of spoken language aphasia. However, sign language employs a visuospatial modality through visual information. We present the first report of a deaf signer who showed substantial sign language aphasia with severe impairment in word production due to a left occipital lesion. This case may indicate the possibility of other localizations of plasticity.

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## GLOSSARY

ASL = American Sign Language; JSL = Japanese Sign Language; WAB = Western Aphasia Battery.

Sign language is a fundamental means of communication for deaf people, in the same manner as spoken language for hearing people. Recent studies have revealed that localization of sign language production and comprehension is similar to that of spoken language.<sup>1-3</sup> However, sign language has its own linguistic structure involving the use of hand signals and motions, and employs a visuospatial modality. This may indicate that visual-related areas are involved in sign language comprehension and production, in the same manner as the auditory-related cortex is involved in speech comprehension and production. We present the first report of a deaf signer who showed substantial sign language aphasia with severe impairment in word production due to a left occipital lesion.

**CASE PRESENTATION** A 72-year-old right-handed Japanese man experienced cardiac embolization. He presented with right homonymous hemianopia with otherwise intact motor and sensory systems. Brain MRI showed infarction of the left occipital lobe including the hippocampus (and surrounding area), part of the medial temporal lobe, and the left corpus callosum (figure 1).

**DEVELOPMENTAL HISTORY** The patient became deaf at 18 months of age before developing speech. His parents were hearing. He learned Japanese Sign Language (JSL) in a community of deaf people in his school days, in addition to his formal education of lip reading and Japanese phonograms (kana) and morphograms (kanji). He could communicate fluently with hearing people using reading and writing.

**NEUROPSYCHOLOGICAL ASSESSMENT** The patient was well oriented and cooperative. He could perform all his daily routines as before onset. This implied that no severe impairment of general intelligence or memory was present. The patient showed no clumsiness, no abnormalities of praxis, and no visual agnosia. He could imitate finger patterns such as the peace sign, but displayed misconnection of lines in copying a cube, indicating probable mild constructional disturbance. He could sometimes produce conversational sign language comprising a few sign words. Figure 2 shows details of JSL.

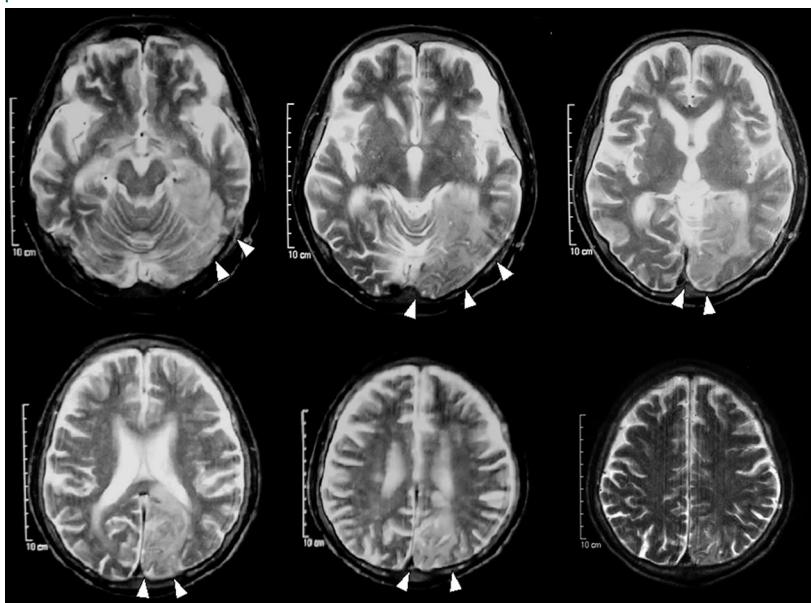
**Sign language comprehension.** The patient could sometimes follow simple instructions involving a few sign words, but never followed those involving finger-spelled kana. He could point to objects among six visually presented items after receiving indications in sign language (12 of 60 [20%] for sign words, and 4 of 60 [7%] for finger-spelled kana words).

**Sign language naming.** The naming task in the Western Aphasia Battery (WAB) was performed by naming 20 objects (60 points; 16 of 60 points [26%] for sign words, and 0 of 60 points [0%] for finger-spelled kana). Errors of sign words and finger-spelled kana included two types. First was the substitution of different-meaning words or letters similar to the originals. These substitutions seem equivalent to verbal paraphasia and phonologic paraphasia of oral language, individually. The

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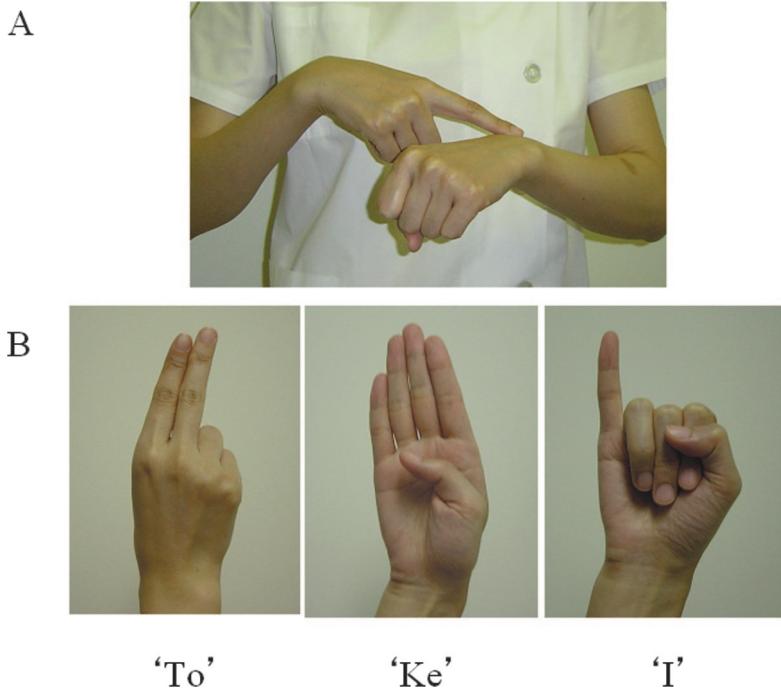
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**Figure 1** Axial T2-weighted imaging



Axial T2-weighted imaging showing infarcts of the left occipital lobe including hippocampus (and surrounding area), part of the medial temporal lobe, and corpus callosum in the territory of the left posterior artery (arrowhead).

**Figure 2** Japanese Sign Language



Japanese Sign Language (JSL) is a language system using visuospatial recognition and expression, and uses a grammatical system similar to that of American Sign Language (ASL). JSL primarily comprises sign words and finger-spelled kana (similar to the finger-spelled alphabet in ASL). In JSL, each sign word represents a word or meaning expressed by the characteristic shape of an object or motions related to an object, while each finger-spelled kana represents a single letter (kana phonogram) made using finger patterns. (A) shows the sign word for "watch" using a motion indicating the wrist, while (B) shows the same word as sequence of motions comprising three finger-spelled kana, "TO," "KE," AND "I," meaning "watch" in Japanese.

second error type involved substitutions of unknown finger patterns that did not indicate any words or letters at all. This substitution seems equivalent to distorted phonemes of oral language, such as anarthria. The impairment of sign language production did not display laterality.

**Sign language repetition.** The patient could imitate sign words and finger-spelled kana one at a time. He could perform only the first few motions, not the entire sequence.

#### Writing and reading of Japanese kana and kanji.

The patient could sometimes write short sentences comprising a few words in both kana and kanji (characters were in the correct shape), but could not read even when he had written the characters himself. He could write the names of visually presented objects (three of six items for kana, two of six items for kanji in WAB), although phonologic paraphasia was sometimes evident. He could not dictate from JSL or translate a sentence written on paper to JSL.

#### DISCUSSION

The patient showed language impairment in all modalities, including sign comprehension, production, reading, and writing, in accordance with the criteria for "sign language aphasia."<sup>4</sup>

Localization of sign language production and comprehension in deaf people has been studied using various strategies: clinical reports, and PET and functional MRI studies.<sup>1-3,5</sup> The clinical reports revealed that left hemisphere-damaged signers showed marked sign language aphasia.<sup>5</sup> Furthermore, the relationship between lesion localization and the characteristics of sign language aphasia resemble those for spoken language, such as a case with frontal and parietal lesions presenting with conduction-like sign language aphasia.<sup>6</sup> PET and functional MRI studies have also shown similar results: in production, the anterior insula, Broca area, premotor cortex, posterior parts of the superior temporal cortex and the supermarginal gyrus are activated,<sup>1</sup> while in comprehension, the lateral temporal lobe,<sup>5</sup> particularly the auditory area,<sup>2</sup> and associated areas are activated. This phenomenon is called cross-modal plasticity. Similarly, in blind people, tactile sensation in Braille reading activates the visual cortex in the occipital lobe, which is employed in visual recognition in sighted individuals.<sup>7</sup> This cross-modal plasticity shows that localizations of compensatory modalities occupy the native area of the deficient modality, such as the auditory cortex in deaf people and the visual cortex in blind people. However, no previous reports have described a left occipital lesion causing substantial changes to sign language, particularly with severe production deficits. This could not be fully explained by the localization that previous reports have showed by cross-modal plasticity. Our patient

demonstrated a different localization from previously reported cases.

Lesions in the present patient were distributed not only to the left occipital lobe, but also to part of the medial temporal lobe and corpus callosum. However, the patient showed severe impairment in word production, and this impairment could not be sufficiently explained by the medial temporal lesion, which would affect word comprehension and retrieval, but not articulation. Although the callosal lesion may have caused disconnection syndrome, aphasic symptoms in this patient could not be fully explained by such disconnection syndrome, as no laterality was shown in sign language production. Recent studies have shown that the right hemisphere may also be recruited in processing sign language, particularly when spatial properties are involved.<sup>8</sup> While left hemisphere dominance for comprehension of sign language was shown, the authors also presumed some involvement of the right hemisphere.<sup>9</sup> They inferred that lateralization depends on the degree of visuospatial information required for the task. In the present patient, the intact right hemisphere may assist in sign language comprehension, but not most of the language function.

There was a report of a patient with "sign blindness" showing severe impairment of comprehension, but trivial impairment of production following a left occipital lesion.<sup>10</sup> The authors considered the deficit of comprehension was analogous to alexia due to disconnection syndrome, although sign language was not merely an encoded reading system, but an independent linguistic system. However, they also outlined an alternative explanation in which neural reorganization occurs as a result of the visual input of linguistic information in deaf signers, and noted that more cases are needed to map out brain plasticity. Although they discussed that production deficits of mild paraphasia could be attributed to limited left thalamus involvement, the present patient did not show any thalamic lesions on MRI. We therefore suggest that this case emphasizes sign language aphasia due to a left occipital lesion.

Visual information is input to the primary visual cortex and analyzed in the associated visual cortex, which is connected to other cortical re-

gions associated with the output process. The occipital lobe is generally considered to control only input processes, not output processes of visual information. However, the present case may indicate that the occipital lobe may control not only the process of visual sense, but also input and output processes for sign language in deaf people. This case thus provides new insights into functional neural plasticity.

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