

Pure agraphia of kanji due to thrombosis of the Labb  vein

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Abstract

A case is described of a 56 year old Japanese male with pure agraphia of kanji (the Japanese morphograms) due to haemorrhagic infarction of the left temporal lobe caused by the rare condition of cortical vein thrombosis of Labb . Writing kanji was severely impaired without disturbed consciousness, aphasia or apraxia. On the other hand, writing kana (the Japanese syllabograms), and reading kanji and kana were almost intact. This suggests that the process of writing kanji involves a different pathway from that of reading kanji in the left temporal lobe. Pure agraphia of kanji is considered to be similar to lexical agraphia in Indo-European languages, in that the writing system with a poor or irregular phoneme-grapheme transformation is impaired by the left temporal lesion. This case indicates the necessity for considering thrombosis of the Labb  vein when a subcortical haematoma is detected in a temporal lobe on computed tomography of the brain.

The Japanese language has two different writing systems, kana and kanji, and Japanese sentences usually consist of combinations of both. Generally, kanji are used for writing most nouns and roots of verbs, adverbs and adjectives, while kana are mainly used for inflectional endings, conjunctions and postpositions. Kana are the simple syllabograms with unique phonetic readings. Kanji are the structurally complex morphograms introduced from China, which have several phonetic readings. All kanji can be represented by kana. Japanese can be written exclusively with 104 kana, which are learnt in the first year of primary school. On the other hand, the 2000 commonly used kanji should be learnt by the end of the ninth grade.

It has been suggested that the processing of kanji and kana involves a different intrahemispheric mechanism, as judged mainly through studies on patients with alexia and agraphia.¹⁻⁴ We report a patient with thrombosis of the left Labb  vein who developed pure agraphia of kanji and compare it with agraphia in Indo-European languages.

Case report

A 56 year old right handed man, educated through high school, was admitted to another hospital for convulsion and disturbed consciousness following dysarthria and slight right

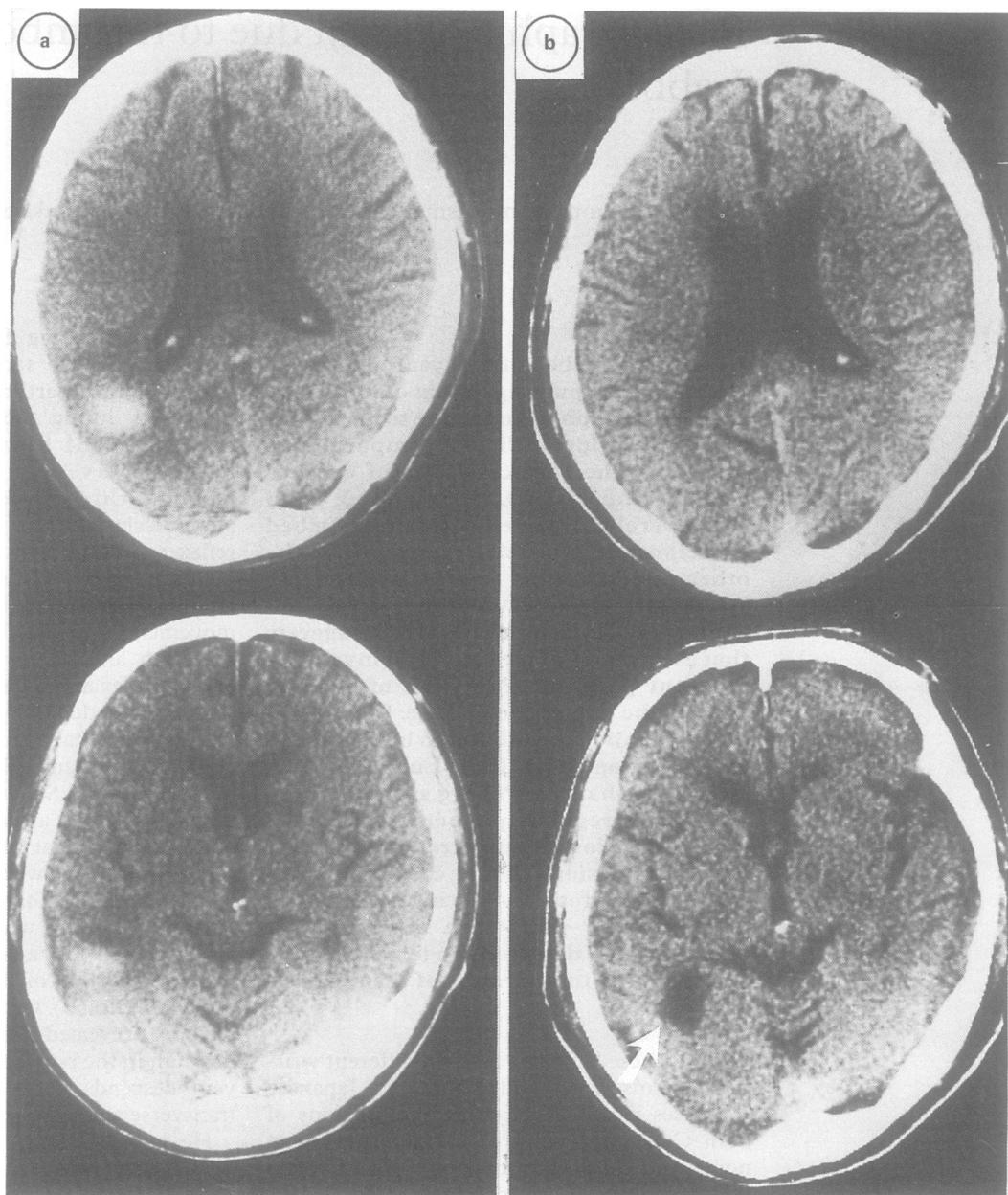
hemiparesis lasting for a few hours on 24 August 1984. He soon recovered consciousness, and his dysarthria and right hemiparesis improved during the next four days. On 7 September 1984, he was admitted to our hospital having developed agraphia of kanji for the first time. On admission, neurological examination disclosed slightly increased deep tendon reflexes on the right side with an extensor plantar reflex. Consciousness, orientation, cranial nerve functions, including ocular movements and visual fields, muscle strength, sensation and gait were normal. Neuropsychological examinations disclosed slightly disturbed immediate ability to recall, name and read kanji and kana but with severely disturbed ability to write kanji. Brain computed tomography (CT) on 7 September 1984, disclosed a high density area around the posterior portion of the left temporal lobe and angular gyrus (fig 1A). In three weeks, neuropsychological problems other than the writing of kanji improved rapidly when the haematoma was absorbed leaving a low density area in the posterior portion of the left temporal lobe on brain CT (fig 1B). Cerebral angiography on 22 October, revealed a filling defect of contrast medium in the portion in which the left Labb  vein descends between the junction of the transverse and sigmoid sinuses (fig 2).

He had a second attack of consciousness disturbance and right hemiparesis, which improved in a few days in March 1987, and he was admitted to our hospital for a second cerebral angiography in June 1987. The neurological findings were normal. The neuropsychological findings were as follows: his Wechsler Adult Intelligence Scale was average (Verbal IQ = 118, Performance IQ = 114, Full Scale = 117). Neither hemineglect (bisection of a line, Albert's line-crossing test and copying the picture of a flower) nor constructional apraxia were noted. Limb and facial praxis to command, facial recognition, left-right discrimination and finger naming were intact. His spontaneous speech was fluent and well articulated. On spontaneous writing he used more kana instead of kanji than would be expected from his intelligence level (fig 3). His speech characteristics were normal on the Western Aphasia Battery (WAB).⁵ He made no mistakes in the WAB tests of spontaneous speech, auditory comprehension, repetition and naming. In reading tasks the scores were maximum except for the following two sections: in Japanese-translated WAB, the "spelled word recognition" and "spelling" sections were changed for kanji and kana.

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Figure 1 CT scan (14 days after the first attack) (A), axial view. A high density area with a vague margin was detected around the angular gyrus (top) and left posterior temporal area (bottom). Two months after the attack (B), a low density area was recognised in the left posterior temporal area (arrow) without abnormality in the angular gyrus (top).



Tested kanji letters consisted of two elements, which had their own meanings and phonetic readings. "Spelled word recognition" section was changed to the recognition of kanji by hearing the structure of them, for example, the names of the elements and their positional

relation between the elements. The "spelling" section was changed to the test of explaining the structure of kanji. In these two sections the scores were low (50%, each). In writing tasks the scores for writing kanji was low (50–62.5%) except for copying kanji. On the other hand, the scores for writing kana and numbers were maximum. We performed special examinations for reading and writing kanji and kana. He was asked to read and write on dictation 218 kanji and 104 kana, which he had learnt in the first two years of primary school. He could not write 82 kanji out of 218 trials (37.6%). He tended to fail kanji characters which were more complex and had been learnt later. There was neither partial omissions, simplification nor confusion with other characters. On the other hand, he made no mistakes in reading kanji or in reading and writing kana letters. In contrast to the prompt responses in writing kana, and reading kana and kanji, he often took some time to write kanji, even when he eventually proved able to write them. When he could not write some kanji, he reported that he knew but could not recall them. In fact, he was able to write

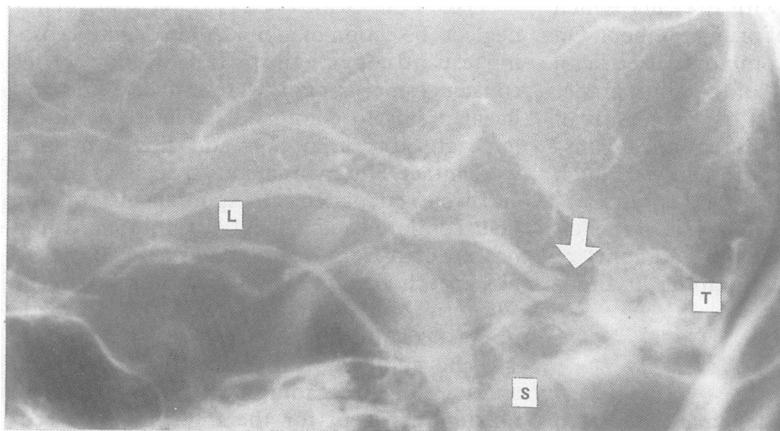


Figure 2 Angiography of the left carotid artery, venous phase, lateral view. A filling defect of contrast medium (arrow) was recognised in the Labbe vein (L) falling into the junction between transverse (T) and sigmoid (S) sinuses.

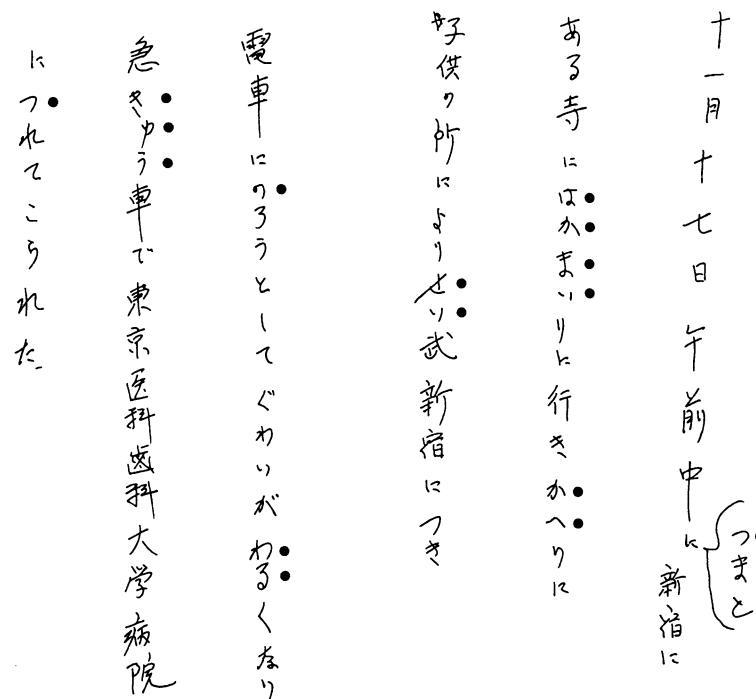


Figure 3 Spontaneous writing. He was requested to use kanji as much as possible. He wrote the corresponding kana (circle) instead of kanji, which were learnt in primary school. On the other hand, kana were written correctly. There were no paragraph errors. The meanings of the sentences are; "On 17 November I went to Shinjuku and visited the grave in a temple with my wife, and on the way home I dropped into my child's house, and I arrived at Seibu-Shinjuku station, and I fell ill when I got on the train, and I was brought to Tokyo Medical and Dental University Hospital by ambulance."

complex kanji immediately after glancing at them, but could not recall them spontaneously. When he wrote kanji and kana with his left hand, the results did not change markedly.

Magnetic resonance imaging (MRI) revealed the lesions in the posterior part of left temporal cortex (fig 4A) and the adjacent white matter (fig 4B) with dilatation of the inferior horn of the left lateral ventricle. There were no abnormalities in the left angular gyrus. On the second cerebral angiography, the same finding, that is, the filling defect of the left Labbé vein, was seen as before.

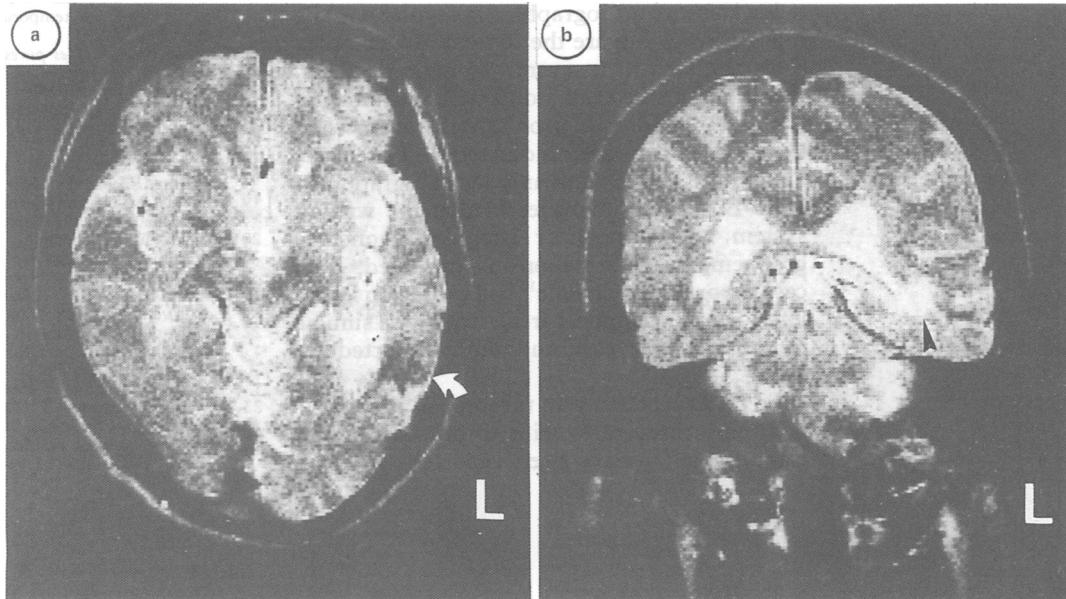
Discussion

Writing is a very complex function involving linguistic, acoustic, praxic, kinesthetic, visual and motor components. Chédru and Geschwind⁶ first indicated that writing was first disturbed in general brain dysfunction. It has been suggested that agraphia was of no clinical value in the localisation of brain damage.⁷ However, recently, much evidence has accumulated indicating that isolated writing disturbances do occur in cases of localised brain damage, which could not be explained by the disturbance of any other higher functions. Three specific areas for pure agraphia have been reported: the left posterior frontal lobe,⁸⁻⁹ the left superior parietal lobe¹⁰⁻¹² and the left temporal lobe.¹³⁻¹⁶

Our case showed no disturbance of consciousness or intelligence, or aphasic or apraxic errors in the chronic phase even after a thorough examination. The pure agraphia in our case was therefore considered to be a focal cerebral sign. The lesion was clearly demonstrated in the left temporal lobe on CT and MRI, which was probably due to the thrombosis of the Labbé vein.

Mochizuki and Ohtomo¹⁷ reported a case of infarction of the left occipital lobe and inferior temporal gyrus, who initially had the difficulty in kanji reading and writing and kana writing, but later kanji reading improved with little improvement in kanji writing. They proposed that the left inferior temporal lobe might be indispensable for writing kanji. Recently Soma *et al*¹⁵ first described "pure agraphia of kanji" in three cases with haematoma, infarction and trauma. All these cases had the lesion in the left posterior temporal area probably extending to the angular gyrus on CT. Although they had amnesic aphasia, alexia and agraphia in the acute phase, disturbances other than agraphia of kanji disappeared in a few months. The clinical course, symptoms and lesion of our case was very similar to those of Soma's cases. Our case, however, clearly demonstrated that amnesic aphasia and alexia, associated with

Figure 4 MRI (3 months after the second attack), T_2 -weighted image ($TR = 2500$ ms, $TE = 105$ ms). (A) Axial view. Low signal area in the posterior portion of left middle temporal lobe (arrow). (B) Frontal view. High signal area in the white matter of left temporal lobe (arrow head).



agraphia of kanji in the acute phase, was due to the lesion in the left angular gyrus, and that the remaining symptom of pure agraphia of kanji was caused by the left posterior temporal lesion. The haematoma, seen around the posterior temporal area and angular gyrus in the acute phase, was absorbed leaving the localised lesion in the posterior temporal area with no abnormality in the angular gyrus on CT and MRI.

In reviewing reported Japanese cases with alexia and agraphia, Iwata¹ proposed the following pathway for the writing of kanji: Wernicke's area(W)-posterior inferior temporal area(T)-occipital lobe(O)-angular gyrus(A)-anterior speech area of Broca and motor association area of hand(M). This is the pathway involved in selecting the correct kanji graphemes according to the meaning of the words, and thus recalled visual engrams of letters are sent to the angular gyrus which mediates the transformation of them into the motor engrams of letters. On the other hand, the pathway for reading of kanji was postulated to be O-T-W, for the writing of kana, W-A-M, and for reading kana, O-A-W. Our case suggested that the process of writing kanji is different from that of reading kanji or kana, or writing kana. According to Iwata's hypothesis, in our case, the writing pathway for the kanji from W to T was selectively disturbed by the lesion in the temporal white matter, sparing the reading pathway for kanji from T to W.

In Indo-European languages two different agraphias, lexical and phonological, have been discussed.¹⁶⁻²⁰ Beavois and Derouesne¹⁹ argued that writing a word can be performed either by using a direct lexical method of obtaining its spelling (the "lexical route") or by sounding-out the word into phonemes and employing phoneme-grapheme transformations. Lexical agraphia consists of a specific difficulty in writing those words which use any irregular or ambiguous phoneme-grapheme transformations, while the writing of regular words and nonsense syllables is preserved. Kana are the simple syllabograms with unique phonetic reading, and the words using kana are orthographically regular. On the other hand, kanji are the structurally complex morphograms with several phonetic readings. Kanji cannot be read aloud without knowing how they should be pronounced, even when their meanings are understood. Therefore, agraphia of kanji is similar to lexical agraphia in Indo-European languages in that the writing system with a poor or irregular phoneme-grapheme transformations is impaired. The case reported by Croisile¹⁶ had the lesion in the left posterior temporal area, which was similar to those in our case and those reported by Soma.

So far as cerebral vein thrombosis is concerned, only three cases with thrombosis of the Labbé vein²¹⁻²³ have been reported. In their

acute phase, two cases^{22,23} had sensory aphasia and the other²¹ had motor aphasia. However, there was no detailed description about agraphia in the reports. Cambria's case²¹ had carbon monoxide poisoning, and Komiyama's²² case was deficient in anti-thrombin III. In our report and that of Kawase²³ the cause of the thrombosis could not be determined. All the reported cases including ours were accompanied by lobar subcortical haemorrhages, which were located around the left posterior temporal lobe and angular gyrus. Therefore, in such situations, it is necessary to consider Labbé vein thrombosis.

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