SHORT REPORT

Pure apraxic agraphia with abnormal writing stroke sequences: report of a Japanese patient with a left superior parietal haemorrhage

Mika Otsuki, Yoshiaki Soma, Toshiko Arai, Atsuko Otsuka, Shoji Tsuji

Abstract
A 67 year old Japanese male patient had pure agraphia after a haemorrhage in the left superior parietal lobule. He developed difficulty in letter formation but showed no linguistic errors, consistent with the criteria of apraxic agraphia. He manifested a selective disorder of sequencing writing strokes, although he was able to orally state the correct sequences. The patient’s complete recovery after 1 month, without new learning, showed that he had manifested a selective disorder of writing stroke sequences. These findings indicate that the final stage of the execution of writing according to acquired sequential memory shown as a stroke sequence can be selectively disturbed, and should be considered to be distinct from the ability of character imagery and the knowledge of the writing stroke sequence itself. This case also indicates that the left superior parietal lobule plays an important part in the execution of writing.

Keywords: pure apraxic agraphia; left superior parietal lobule; writing stroke sequences

Pure agraphia is an impairment in writing unaccompanied by any other relevant language disorder, which occurs in association with lesions in the frontal lobes, superior parietal lobule, temporal lobes, left caudate nucleus and internal capsule, and left thalamus. Regarding the type of pure agraphia and the lesions, the parietal lobe is considered a candidate site of the causative lesion for disorders of spatial and kinesthetic movements of writing; however, the relation between the type of agraphia and other lesions remains uncertain. We herein report a case of pure agraphia after a haemorrhage in the left superior parietal lobule in a Japanese patient, who showed a characteristic impairment of writing, and we discuss the symptoms and the mechanism of pure agraphia after a left parietal lesion.

Case description
A 67 year old, right handed, university educated Japanese male bank clerk was admitted to Takeda General Hospital on 12 October 1993. He had been in good health without any relevant history of disease, but was subsequently found to have hypertension. On the evening of 12 October he noted sudden weakness in his right arm while he was at his office. His colleagues noted at that time that he answered their questions irrelevantly and they brought him to the hospital. A neurological examination showed him to be alert, but disoriented as to time and place. The cranial nerves were all intact, and he showed no paresis in the limbs, and no pathological reflexes or sensory deficits. By the next day he had become well oriented, but he became aware of an impairment in his writing. Brain MRI performed 22 days after onset (fig 1) disclosed a circumscribed haemorrhage in the left superior parietal lobule. Seventeen days after onset, N-isopropyl-(123I)-p-iodoamphetamine single photon emission computed tomography (SPECT) disclosed decreased blood flow corresponding to the region showing a haemorrhage, as disclosed by MRI.

Neuropsychological assessment
From the onset, the patient’s speech was fluent and well articulated, and he showed no orofacial apraxia. The western aphasia battery,21 Japanese version22 was administered within 1 week after onset, and disclosed no aphasic findings or reading disability except for a writing impairment. On the revised Wechsler adult intelligence scale (WAIS-R), administered 13 days after onset, the patient showed a verbal IQ of 109, performance IQ of 100, and full scale IQ of 105. Regarding the praxis examined during the first week after onset, he was able to make meaningful gestures such as beckoning, waving goodbye, tooth brushing, pretended tool use, and pantomime on verbal command and on imitation; he was also able to imitate meaningless gestures made by the examiners, and he could use tools accurately, which indicated that he had no impairment in praxis such as ideomotor apraxia or ideational apraxia. He was able to imitate simple finger patterns using either hand, and he showed no clumsiness. He was able to copy cubic and Rey-Osterreith’s designs without any problem, with the normal strategy employed to copy, and
superior parietal lobule is seen. Scattered white matter lesions indicate the ischaemia within the white matter after hypertension.

Figure 1 T2 weighted brain MR axial images (TR=3000, TE=90) of the patient obtained 22 days after onset. A circumscribed haemorrhage in the left superior parietal lobule is seen. Scattered white matter lesions indicate the ischaemia within the white matter after hypertension.

We administered writing tests 18 days after onset: the dictation and transcription of educational Kanji (morphograms\textsuperscript{12}), and the same words written in Kana (syllabograms\textsuperscript{12}). We used the easiest Kanji, selected at random from among educational samples for pupils at elementary school level.

Results

The results are summarised in the table and writing samples by the patient are shown in figure 2. Before any further description, it is necessary to explain the Japanese writing system. There are two systems of writing in Japanese; Kanji (morphograms\textsuperscript{12}) and Kana (syllabograms\textsuperscript{12}). Kanji often have very complicated shapes, achieved with as many as 10–20 strokes, and many Kanji thus require a particular writing stroke sequence. Figure 2, A, B, and C are examples of Kanji. Thus it is a fundamental educational subject for Japanese pupils to learn the stroke sequences of Kanji. They learn them by practicing the sequences repeatedly, not only through knowledge but also using sequential movement memory. Kana are much simpler than most Kanji; Kana are generally two to four stroke symbols of mora (in the Japanese language, the ultimate minimum unit of a sound corresponds to a mora, which in turn correspond to one Kana letter) such as “ro” or “bo” (fig 2, D shows two Kana). Examples of both the correct sequences of strokes for Kanji or Kana, which were shown by the patient after his recovery, and the patient’s writing with unusual stroke sequences during his morbid period are shown in fig 2.

In addition to the ill shaped letter formation, the patient characteristically showed an unusual sequence of strokes for forming Kanji or Kana, although he was able to eventually arrive at the correct formation. The patient complained of his impairment of writing as follows: “My hand slips, although I know how to write.” This symptom was observed in the writing of both Kanji and Kana, equally on spontaneous writing, dictation, and transcription. He himself noticed that his stroke sequences were strange, and he was able to express orally without any difficulty how the sequence of strokes should proceed and how he used to perform the sequence before the haemorrhage. Therefore, his knowledge of the sequence of writing strokes seemed to be preserved. He sometimes succeeded in correcting his mistakes, but not always. He showed no spelling errors in his writing, and he was perfectly able to select correct Kanji or Kana from among the samples and make words and sentences.

At the re-examination 1 month after onset, in the same writing tests administered earlier, the patient was able to write correctly all of the Kanji and Kana tested with the correct sequence of writing strokes, without new learning, and he has never shown an unusual stroke sequence in writing since then. In a

<table>
<thead>
<tr>
<th>Kanji</th>
<th>Kana*</th>
</tr>
</thead>
<tbody>
<tr>
<td>50 (100%)</td>
<td>50 (100%)</td>
</tr>
</tbody>
</table>

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Results of reading and writing examinations

<table>
<thead>
<tr>
<th>Reading (administered 17 days after onset):</th>
<th>Kanji</th>
<th>Kana*</th>
</tr>
</thead>
<tbody>
<tr>
<td>Administered numbers</td>
<td>50</td>
<td>50</td>
</tr>
<tr>
<td>Correct answers</td>
<td>50 (100%)</td>
<td>50 (100%)</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Writing (administered 18 days after onset):</th>
<th>Kanji</th>
<th>Kana*</th>
</tr>
</thead>
<tbody>
<tr>
<td>Administered numbers</td>
<td>50</td>
<td>50</td>
</tr>
<tr>
<td>Correct answers</td>
<td>38 (76%)</td>
<td>38 (76%)</td>
</tr>
<tr>
<td>A</td>
<td>10 (20%)</td>
<td>9 (18%)</td>
</tr>
<tr>
<td>B</td>
<td>1 (2%)</td>
<td>3 (6%)</td>
</tr>
<tr>
<td>C</td>
<td>1 (2%)</td>
<td>0 (0%)</td>
</tr>
<tr>
<td>D</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

(A) Correct shapes with a correct sequence of strokes; (B) correct shapes with an unusual sequence of strokes; (C) bad formation; (D) no recall

* Regarding the examinations of Kana, the administered numbers are not the numbers of Kana letters but are Kana words, which consist of several Kana letters. We assessed the patient’s performance according to the existence of mistakes, which means that even if the patient showed an incorrect response in one Kana letter, we assessed the entire word as an incorrect response, and the patient was assessed as giving a normal answer only when he was able to complete whole words with completely correct letter formation. We used Kana for the word examinations because the number of Kana letters is limited, only 46 in all, compared with the standard elementary school vocabulary level of 1000 Kanji. We also hoped to examine the patient’s ability to write sequential movements which demand a certain flow, so it seemed preferable to adopt words to each letter. Thus, the patient’s scores on the Kanji and Kana examinations results cannot be directly compared for the degree of the writing disorder.
re-examination of the WAIS-R, the patient showed a verbal IQ of 92, a performance IQ of 105, and a full IQ of 98.

Discussion
We have described a patient who manifested an impairment in only writing after a circumscribed haemorrhage in the left superior parietal lobule, without any other symptoms such as aphasia, alexia, agnosia, limb clumsiness, ideomotor apraxia, ideational apraxia, or constructional disturbance. He showed no other abnormal findings which could possibly cause a writing disturbance, such as dementia or general memory disturbance. His writing impairment persisted for a month and cannot be simply ascribed to an acute confusional state and cannot be interpreted as transient and negligible, because he was able to perform other examinations without any abnormality during the same period and there is no confusional state known which shows such a selective disorder persisting for 1 month. In addition, the patient’s WAIS-R re-examination showed almost the same IQ score, indicating that the patient’s writing impairment cannot be ascribed to general mental deterioration during the morbid period. Therefore, the patient’s symptom can be interpreted as a focal sign after the onset of haemorrhage, and it was consistent with the criteria for pure agraphia.1

Previous neuropsychological assessments of pure agraphia have disclosed that it can be classed into two types: in “linguistic” agraphia, the patients manifest spelling errors, and in “apraxic” agraphia, they manifest abnormal letter formation. Although apraxic agraphia is termed “apraxic,” it is not accompanied by

Figure 2  Top: writing samples of the patient show a poorly formed Kana word. He had tried to rewrite the same word, which consists of three Kana letters, “A-i-zu” (which is the name of the city in which the patient lives) repeatedly from the top to the bottom as the arrow indicates, but he was not able to complete it (11 days after onset). Bottom: the patient’s unusual order of writing strokes at dictation (18 days after onset). The right side of each column shows the correct sequence used to make the complete character, shown at the bottom of each column. The patient executed the writing in the order indicated by the arrow in the left hand columns. (A) This character (Kanji), for aviation, was not completed correctly. (B) This character (Kanji), for fly, was completed correctly, but with an unusual order of writing strokes. (C) This character (Kanji), for part of the patient’s name, was also made with an unusual sequence. (D) These two letters (Kana), for “ro-bo”, part of the word “robot”, were eventually completed correctly, but with an unusual sequence order.
limb apraxia or a constructional disturbance. Baxter and Warrington\textsuperscript{13} used the term “pure” apraxic agraphia to identify the symptom of writing disturbance appearing in the absence of a disturbance of spelling, reading, or other general language problems, and importantly, occurring in the absence of major praxic or visual-constructional difficulties. Although the early reports of patients with apraxic agraphia disclosed the fact that writing apraxia and limb apraxia could be distinct,\textsuperscript{13,14} these patients had atypical cerebral dominance. For this reason the authors considered that the dissociation of writing apraxia and limb apraxia could be due to the atypical cerebral dominance. Subsequently Baxter and Warrington\textsuperscript{15} issued the first report describing a patient with typical cerebral dominance who seemed to have apraxic agraphia, and they postulated that their patient’s inability arose at the level which specifies the motor sequences or “graphic motor pattern,” and termed it “ideational agraphia.” Their report indicated that selective writing apraxia can also occur in a patient with typical cerebral dominance.

Our patient showed pure agraphia with a preserved ability for spelling, and he was able to select letters or characters correctly, all of which indicated that he had no linguistic impairment. He was able to make excellent copies of cubic and Rey-Osterreith’s designs and had a normal score on block design tests, results which show that he had no visual constructional impairment. He showed ill shaped letter formation without any other inability, and these findings disclosed his symptom pattern to be similar to those of the patients described as having apraxic agraphia in the previous reports.\textsuperscript{1,13,15,16}

The most characteristic of our patient’s impairment was the conspicuous, unusual, and incorrect writing sequence strokes. As shown in fig 2, Japanese Kanji have complicated stroke sequences and, thus, if people who are not familiar with Kanji, try to transcribe such a character, they would not know which line to write first and which should be second, etc. Our patient wrote Kanji and Kana with unusual stroke orders, although he was able to write the complete Kanji or Kana. He wrote each Kanji or Kana as if he were transcribing an unfamiliar foreign shape, both at dictation and transcription. Naturally, the degree of achievement in mastering the correct writing stroke order depends on the individual person, and some Japanese people fail to master them and habitually use some idiosyncratic stroke sequences. However, we cannot ascribe our patient’s unusual stroke orders to low achievement or to personal habit for two reasons; one is because he could orally state how the correct stroke sequences for each letter or character in a month.

There are few reports in the western literature of patients with pure agraphia showing an unusual stroke sequence. We suspect that the nature of the Japanese writing system disclosed the patient’s disturbance more conspicuously. In the Japanese literature, Ishiai et al\textsuperscript{17} reported pure agraphia due to a left parietal lobe infarction in a patient, who showed unusual sequences of writing strokes, which seems similar to the impairment of our patient. Our patient was able to orally state not only the correct spelling but also the correct sequences of writing strokes for all Kanji and Kana tested, which indicates that his disturbance was not one of memory of the stroke order as knowledge itself but rather a disturbance in the process of the execution of writing as sequential memory.

Rothi and Heilman\textsuperscript{18} postulated the existence of a graphemic area that is responsible for guiding motor programming in grapheme production. Roeltgen and Heilman\textsuperscript{19} described a patient who was unable to write any formed graphemes despite a preserved ability to spell aloud, whose disorder was thought to be a disconnection of output from the graphemic area. This disorder seems relevant to that of our patient. In addition to this disorder, our patient showed the unusual writing stroke sequences even when his writing did not show poor formation, and he could not improve by copying. This indicates that the disorder arose at the final stage of motor output, which involves both spontaneous writing and copying. Cray and Heilman\textsuperscript{19} presented a case of pure apraxic agraphia in which defective letter imagery was evident, and they proposed the possibility of two regulation sites in the graphemic area, one for letter imagery and the other for motor transcoding. Our patient presented evidence of a selective disorder of the second site: a disorder of motor transcription without letter imagery disorder.

Regarding the causative lesions for apraxic agraphia, Alexander et al\textsuperscript{20} reported the lesion localisation as the superior parietal lobule. Kapur and Lawton\textsuperscript{21} hypothesised that parietal agraphia is due to a memory difficulty for the motor movements associated with letters. In the parietal area, we hypothesise that a lesion restricted to the left superior parietal lobule can cause a selective impairment of the execution of writing sequence, manifested as an abnormal order of writing strokes. This would be particularly conspicuous in a Japanese patient, because Japanese characters and letters require certain complicated stroke sequences, a feature which demands much of sequential memory.

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Multiple sclerosis

The recognition of multiple sclerosis as a clinical entity different from other movement disorders or spinal paraplegias was made by Charcot in 1868. Previously multiple sclerosis had been repeatedly confused with paralysis agitans. It was Charcot who made the first definitive pathological description that distinguished the two diseases. The pathological lesions of multiple sclerosis, probably first illustrated 30 years earlier in 1838 by Robert Carswell, were those of an unnamed French patient. Carswell, a Scotsman, held the inaugural chair of pathology at London University. (Compston A. The 150th anniversary of the first depiction of the lesions of multiple sclerosis. J Neurol Neurosurg Psychiatry 1988; 51:1249-52.) Until the mid-1960s little was known about the disordered physiology produced by demyelination. In 1963 Professor Ian McDonald demonstrated that electrical signs of conduction block developed abruptly at the junction of normal and demyelinated parts of nerves. McDonald and Sears, in 1969, showed that conduction block occurred at the margin of the lesions produced in the posterior columns of cats and that portions of the nerve fibres distal to the block retained their ability to conduct impulses. One hundred and forty three years after Carswell’s illustration the ability of NMR to disclose abnormalities with multiple sclerosis on a scale not previously seen, except at necropsy, was demonstrated. The potential of NMR to assist in the diagnosis of MS was established. (Young IR et al. Lancet 1981; ii: 1063-5.)

In 1962 Monaco produced this stamp showing the sun, flowers, and a chest filled with hope (Stanley Gibbons 151, Scott 506). It was another 30 years before realistically hopeful therapies for the disorder began to emerge.

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