Apraxic Agraphia With Neglect-induced Paragraphia

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 Agraphia may result from the loss or unavailability of the memory of movements necessary to form written letters. For this mechanism to be invoked, it must first be demonstrated that there is no language deficit and that there is no disconnection between language and motor areas. A left-handed patient demonstrated bilateral agraphia and apraxia, but not aphasia, following a right parietal infarction. Preservation of the ability to type with the left hand demonstrated that the right hemisphere was not disconnected from language input. In addition, paragraphic errors while typing were shown to be secondary to left hemispatial nealect.

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 $H^{\rm eilman}_{\rm handed\ patient\ in\ whom\ agraph-}$ ia, but not aphasia, developed following a large lesion in the middle cerebral artery territory of the right hemisphere. The patient had a left hemiplegia but, in addition, he had severe apraxia of his right hand and he could not write with his right hand, even though he had always done so prior to his stroke. Heilman et al¹ proposed that in this patient, the left hemisphere was dominant for language but, as the patient was left handed, the right hemisphere was dominant for fine motor control. For the patient to write, he had to transfer information from the left (language) hemisphere to the right (motor) hemisphere. To write with the right hand,

the right hemisphere had to direct the left motor cortex to make the appropriate movements. Despite the coexistence of apraxia and agraphia, the authors' were reluctant to attribute the patient's agraphia to apraxia since it was possible that the defect in writing resulted from the unavailability of linguistic input to the right hemisphere rather than from the unavailability of the engrams for forming the letters. Since the patient had a left hemiplegia, the right hemisphere could not be tested to see if it had access to linguistic input. Heilman et al² subsequently reported an analogous case in which the same problem in interpretation arose. In this case, a large lesion in the left hemisphere of a right-handed patient produced a dense right hemiplegia, but no aphasia. Apraxia and agraphia of the patient's left hand developed. Again, the right hemiplegia prevented testing the possibility that the left hemisphere was cut off from linguistic input from the language-dominant right hemisphere.

We now describe a patient in whom agraphia and apraxia, but not aphasia, were produced from a lesion restricted to the right parietal lobe. Since the patient was not hemiparetic, the right hemisphere could be tested for its access to linguistic information.

REPORT OF A CASE

The patient was a 74-year-old retired executive vice-president with a tenthgrade education. He was left handed: writing, throwing a ball, using scissors, combing his hair, and using a knife (among other activities) were all performed with his left hand. There was no family history of left-handedness. He had adult-onset diabetes, which was treated with small doses of insulin, and he had suffered three myocardial infarctions during the previous 20 years.

He had no symptoms of neurological dysfunction until May 23, 1977, when he awakened with a headache. His wife noted slurred speech and "confusion" and brought him to a local hospital where he was thought to be hypoglycemic. His serum glucose level, however, was 300 mg/dL. His speech and "confusion" improved and he was discharged after two days. He remained unable to dress himself and unable to write, and he was therefore referred to the Shands Teaching Hospital, Gainesville, Fla, for further evaluation.

When the patient was examined on June 8, 1977, his blood pressure was 140/55 mm Hg, his pulse rate was 50 beats per minute and regular, and he was afebrile. The general physical examination was without abnormalities except for a soft systolic murmur heard only over the aortic area. His heart was not enlarged, his chest was clear, and there was no peripheral edema.

Neurological examination demonstrated extinction of bilateral simultaneous visual, auditory, and tactile stimuli on the left. Cranial nerve examination showed full visual fields and normal fundi. There was minimal left facial weakness, but no other cranial nerve deficit. There was no weakness of his extremities. There was upward ("parietal") drift of the extended left upper extremity with the eyes closed. Sensory examination showed normal vibration and proprioception in both hands. The muscle stretch reflexes were more brisk on the left than the right. There were bilateral Babinski responses. There were no grasp. root, or suck reflexes.

He was alert, attentive, and fully oriented. His digit span was seven numbers forward. He recalled three of three items after five minutes. Remote memory was normal. Topographical localization was done well.

He was mildly dysarthric, but there was no other disturbance of spoken language; language comprehension, repetition, and object naming were all absolutely normal. He could read aloud and for comprehen-

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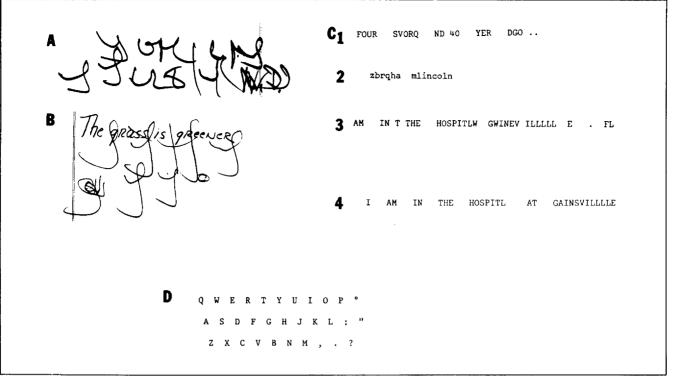


Fig 1.—A, Spontaneous writing: patient attempting to write, "You are a doctor." B, Copying. C, Typewriting. C, 1 to 3, Typewriter directly in front of patient. C, 4, Typewriter moved to patient's right side. D, The typewriter keyboard. Note that most of letters missed (A, S, and E) are on left side of keyboard.

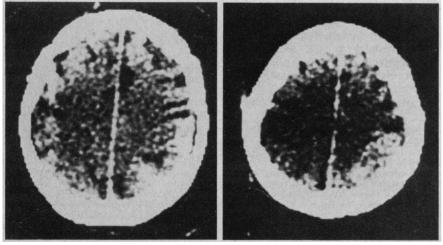


Fig 2.—Computerized tomographic scans with contrast enhancement, demonstrating moderate diffuse cortical atrophy, and a distinct lentiform area of contrast enhancement in right posterior parietal region. This area was of decreased density on noncontrast scan.

sion. He could spell accurately, and he understood words spelled to him. In contrast, he could not write even his name. Attempts at writing produced large, poorly formed letters, only a few of which were identifiable. There was no improvement when writing to dictation (Fig 1, A) or copying (Fig 1, B). He spontaneously used his left hand for writing; performance with his right hand was poorer. Typewriting with one finger of his left hand (he had never learned to touch type) was performed better than writing, but there were still many errors (Fig 1, C_{1-3}). When it was realized that nearly all of these errors were made when he tried to type letters on the left side of the keyboard, the typewriter was moved to the patient's right side. His typewriting (still with his left hand) improved sharply; in striking contrast to his writing, he was able when typing to complete whole sentences with only minor errors (Fig 1, C₄). The patient's wife reported his typewriting to be slightly worse with the right hand than with the left. The patient demonstrated remarkable apraxia, both of face and of limb movements. He understood the commands, and the limb movements produced were clumsy imitations of the correct movement. His performance did not improve with imitation. The limb apraxia was bilateral, slightly worse on the right.

Routine laboratory study results were normal, and the serum VDRL test was negative. A lumbar puncture yielded clear, colorless fluid under normal pressure. The CSF protein level was 121 mg/dL, and there were eight mononuclear cells per cubic millimeter. The CSF cultures were negative. The EEG showed generalized right hemisphere slowing in the theta range, which was most pronounced posteriorly. A computerized tomographic scan done 17 days after the onset of his illness showed a discrete area of decreased density in the right posterior parietal region, which enhanced with contrast (Fig 2). There was no mass effect. In addition, mild ventricular dilation and moderate cortical atrophy were noted. No other focal lesions were apparent.

COMMENT

In our patient, most of the common causes of agraphia could be excluded on the basis of the clinical examination. Thus, our patient was not aphasic or alexic, and he lacked the other features of the Gerstmann syndrome. There was no evidence of a mass lesion, and he was not confused.³ There was no gross visuospatial disorientation. He did have definite neglect of the left half of space. Patients with unilateral neglect may demonstrate "spatial dysgraphia": their writing does not maintain a straight line and is crowded to one side of the page. Their writing is nevertheless legible. Our patient showed only a slight tendency to write on the right side of the page; however, in contrast to patients with spatial dysgraphia, he had difficulty correctly forming even a single letter.

Agraphia may be seen in the absence of aphasia as a result of a disconnection of motor centers from linguistic control.^{5.6} The combinations of agraphic and apraxic features possible in patients with callosal lesions depend on the patterns of hemisphere dominance in the individual patients, and these have been discussed in detail by Heilman et al.¹ In the patient with left hemisphere dominance for language, a callosal lesion will produce agraphia in the left hand since the right hemisphere is deprived of linguistic input. Although, theoretically, ipsilateral pathways for crude motor activity might be available to enable the left hand to type, the patient described by Geschwind and Kaplan⁶ with a callosal disconnection could not write, type, or arrange anagram letters with his left hand although he could do all three with his right. In contrast, our patient could write with neither hand, but he could use his left hand for typewriting. The fact that he spontaneously used his

left hand for typewriting is further evidence that ipsilateral pathways from the left hemisphere were not being used (the left hemisphere should preferentially use the right hand). We conclude that his right hemisphere did have access to language and that his agraphia cannot be explained on the basis of a disconnection of language from motor areas. We propose instead that he lost the memory of the movements necessary to form written letters. His agraphia can thus be regarded as a specific type of apraxic disorder.

Apraxia is frequently seen in association with left-sided post-Rolandic lesions, including lesions in the parietal lobes.^{7.8} Recent electrophysiological work⁹ has identified single cells in areas 5 and (especially) 7 that appear to have a premotor ("command") function. Liepmann⁷ proposed that visuokinesthetic-motor engrams were encoded in these posterior regions. Handedness might reflect an asymmetric representation of these engrams. The right-hander thus has bilateral apraxia from left parietal lesions; the left-hander might be expected to become bilaterally apraxic from right parietal lesions. Our patient's lesion was well circumscribed and involved the posterior parietal cortex, which included the inferior parietal lobule and spared the postcentral gyrus and the frontal lobes entirely. It is conceivable that a lesion restricted to the frontal premotor cortex would have produced a similar disorder of praxis and writing in

this patient by interfering with the execution of these functions by the premotor and motor cortex. It is possible that this mechanism explains some of the cases of agraphia reported from frontal lesions.

It is worth noting that our patient's performance on the typewriter not only demonstrated that his right motor cortex had access to linguistic input but also brought out a spatial paragraphia that was only slightly apparent in his written productions. Neglect of the far left side of the typewriter produced errors in typewriting (neglect-induced paragraphic errors) that, if more severe, could easily have been mistaken for a linguistic deficit. The failure in the previous two patients with agraphia described by Heilman et al^{1,2} to use anagram letters and to type was thought to be on this basis. However, the defects were not studied systematically and the authors^{1.2} could not exclude the possibility that a linguistic deficit was also contributory. The simple expedient of moving the typewriter into our patient's normal field eliminated almost all of these typing errors. This maneuver was important not only for theoretical reasons; it also enabled the patient to carry on his correspondence, which he could not have done alone in any other way.

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