

Gerstmann's syndrome in a patient with left thalamic hemorrhage

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Abstract

A 78-year-old right-handed man presented with mild right hemiplegia and impaired deep sensation on his right side. He had no aphasia, but showed Gerstmann's syndrome which is characterized by four primary symptoms: finger agnosia, right-left disorientation, agraphia, and acalculia. A cranial CT revealed hemorrhagic lesions in the left thalamus, and single photon emission CT showed decreased regional cerebral blood flow in the superior portion of the left temporoparietal lobe, especially angular gyrus area. It is inferred that in this case, corticofugal fibers from the thalamus to the parietal lobe had been damaged by hemorrhage, secondarily causing decreased function of the cerebral cortex.

INTRODUCTION

Gerstmann's syndrome is characterized by four symptoms - agraphia, acalculia, finger agnosia, and right-left disorientation.¹ This syndrome has been well known as a focal symptom of the angular gyrus region in the dominant hemisphere. It is reported that lesions in the thalamus also cause cerebral cortical symptoms, but most of them are unilateral spatial neglect or aphasia. We present a case of Gerstmann's syndrome caused by hemorrhage in the left thalamus.

CASE REPORT

K.T. is a 78 year-old right-handed man with 12 years of school education. He had a history of hypertension for 5 years, but no prior history of neurological problems. He experienced a sudden onset of weakness of his right arm and leg on September 20, 2005, and was admitted to our hospital on the following day. On admission he was alert, but disoriented as to time and place, and showed impaired attention. He also presented with mild right hemiplegia and impaired thermal nociception, and impaired deep sensation on his right side. His visual acuity and visual field were normal, and there were no other abnormalities in the central nervous system.

A cranial CT performed on the day of onset showed hyperdense areas expanding from inside to outside in the posterior left thalamus and this was diagnosed as hemorrhage in the left thalamus (Figure 1). A magnetic resonance imaging (MRI)

performed 10 days after onset also showed lesions at the same site, and no other clear lesions were observed. Single photon emission CT (SPECT) showed decreased regional cerebral blood flow (rCBF) in part of the left temporal lobe and in the parietal lobe, especially in the left angular gyrus area (Figure 2).

His spontaneous speech was fluent without paraphasia. His performance in the naming of objects, repetition of words, and auditory language comprehension was good. However, he had difficulty in writing letters (Figure 3), characterized by paraphasia with misspellings and syntax errors, and was slow in reading, although eventually he could read the text correctly. He was confused in recognizing his right and left and his fingers, and his ability to select the same side and the same finger as that of the examiner was impaired, as well as making mistakes in distinguishing right from left or naming the fingers, and in following directions. In Benton's Right-Left Orientation Test and Finger Localisation Test, he scored 10/20 and 34/60, respectively. He managed to perform four single-digit, arithmetic operations if he took his time, but he could not do more than 2-digit arithmetic operations. No other apraxia or agnosia was observed. He needed supervision and directions to lead a daily life.

He scored 18/30 in the Mini-Mental State Examination (MMSE), with declining performance in orientation, attention and calculation, and reproduction. The WAIS-R scores were VIQ=85,

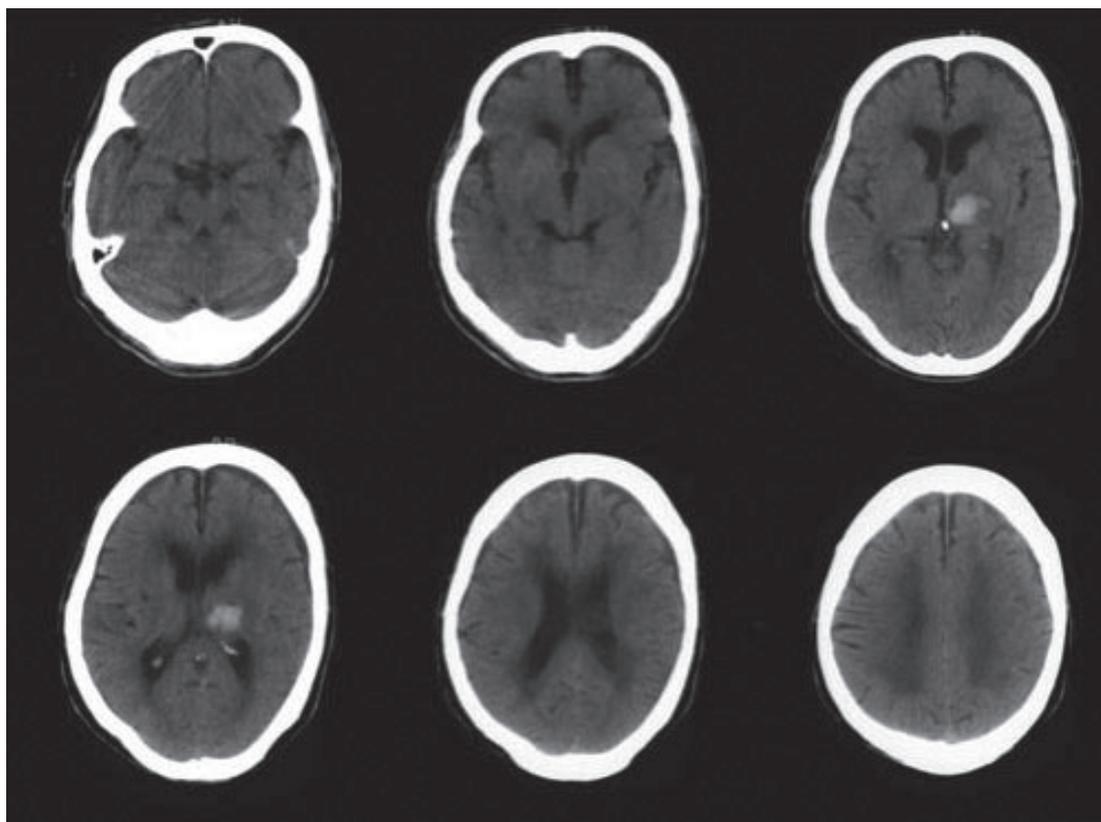


Figure 1: A cranial CT performed on the day of onset showed high density areas expanding from inside to outside in the posterior left thalamus and this was diagnosed as hemorrhage in the left thalamus.

PIQ=94, and Full scale IQ=89. On the auditory verbal learning test, the immediate recall (2, 3, 4, 1, 1/15) and delayed recall (0/15) were decreased. The score on the Raven's Colored Progressive Matrices (RCPM) was 19/36, on Frontal Assessment Battery (FAB) 8/18, and the word fluency test where the patient was required to produce exemplars from given categories resulted in scores of 2, 0, and 0 for the categories "animal," "fruit," and "vehicle," respectively, and where he was required to produce words that begin with given letters resulted in scores of 0, 0, and 0 for the letters "shi," "i," and "re," respectively. The level of executive functioning was classified as "impaired" on the basis of the scores of Behavioral Assessment of the Dysexecutive Function (BADS), i.e., overall profile 5, standardized 27, and age-corrected score 44.

DISCUSSION

In our patient, finger agnosia and right-left disorientation were prominent symptoms. His agraphia and acalculia also was severe.

These features commonly are associated with Gerstmann's syndrome.¹ There is a possibility that the four symptoms of Gerstmann's syndrome are caused secondarily by the disturbance of consciousness, disturbance of attention, aphasia, and so forth. In our case, however, the patient had good movement when transporting and transferring, and had high communication ability, good enough to undergo numerous neuropsychological tests. Therefore, as Alexander *et al.*² pointed out, it seems unreasonable to elucidate cerebral cortical symptoms such as Gerstmann's syndrome only with respect to arousal level decreases due to the lesions in the thalamus, or disturbance of attention. Although our patient did show mild word amnesia and impairment of understanding at the early stage of onset, he had abnormal results for right-left orientation and finger naming in nonlinguistic tests as well. It seems that this syndrome cannot be explained by linguistic factors alone, but factors such as disturbance of spatial perception and body image, and mental rotation may be involved.

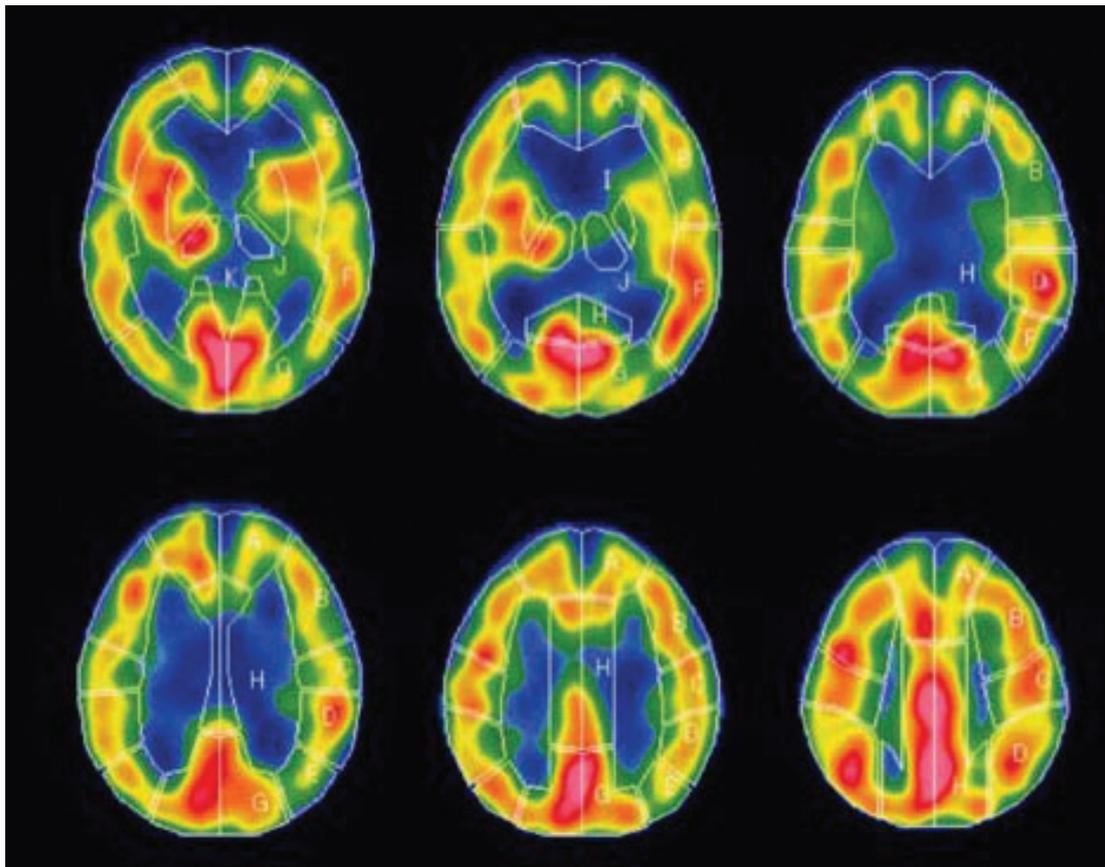


Figure 2: Single photon emission CT showed decreased regional cerebral blood flow in part of the left temporal lobe and in the parietal lobe, especially in the left angular gyrus area.

	時	新	鉛	灰	切	毛	と	し	え	は	き	け
	計	聞	筆	皿	手	糸	け	ん	ん	い	つ	い
Dictation	時 計	新 聞	鉛 筆	灰 皿	切 手	毛 糸	と けい	し ん ぶ ん	え ん び つ	は い ざ ら	き つ て	け い と
Copy	時 計	新 聞	鉛 筆	灰 皿	切 手	毛 糸	と けい	し ん ぶ ん	え ん び つ	は い ざ ら	き つ て	け い と

Figure 3: Sample of the dictation and copy in this patient showing difficulty in writing letters, although eventually the patient could copy them correctly.

Since the 1960s, there have been some reports that raised doubts as to the existence of a distinct syndrome.³ Furthermore, there have been numerous reports of Gerstmann's syndrome without language problems, most of which are accompanied by other symptoms such as constructional apraxia, impaired visual fields, impaired sensation, neglect, and mental retardation^{4,5}, and pure cases presenting all the four symptoms described by Gerstmann are rare indeed.⁶ He reported one of the few pure cases, in which culprit lesions were found in the supramarginal gyrus and the upper parietal lobe, in addition to the superior angular gyrus. Morris⁷ applied electric stimulation to the left parietal lobe cortex in patients with epilepsy, and demonstrated that the four symptoms of Gerstmann's syndrome are caused by damage to a very confined area of the angular gyrus. He also showed that the damage to a site lower than that area causes symptoms such as nominal aphasia, alexia, and constructional apraxia, in addition to the four symptoms. There is also a recent report that each of the four symptoms of Gerstmann syndrome was caused independently due to the lesions in the left lateral parietal lobe.^{8,9} From these findings, it is considered that, although it is rare for patients to present only with the four symptoms of Gerstmann's syndrome without any complications, Gerstmann's syndrome is important in locating the damaged nerve as a combination of symptoms suggestive of lesions in the left parietal lobe. The appearance of all four symptoms is not important. Benson *et al.*⁹ also pointed out that lesions in the left angular gyrus cause a series of cortical symptoms such as nominal aphasia, agraphia, and alexia as well as Gerstmann's syndrome, and that Gerstmann's syndrome reflects highly reliable information concerning the location of the damaged cortex. In our case, Gerstmann's syndrome, which is suggestive of the presence of lesions in the left parietal lobe, is caused by lesions in the left thalamus, which we consider makes this case worth reporting. The possible reason for this is as follows. The thalamus is a relay point for various projection fibers, and it connects by the fibers between posterior medial thalamus and area 39 (the so-called "angular gyrus"). In fact, SPECT of our patient showed decreased regional cerebral blood flow in the parietal lobe, which is known as classical lesions of Gerstmann syndrome. From these findings, we infer that in our case, corticofugal projection fibers from the thalamus to the parietal lobe had been damaged, and thus

secondarily caused decreased function of the cerebral cortex in area 39.

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