

Right Parietal Stroke With Gerstmann's Syndrome

Appearance on Computed Tomography, Magnetic Resonance Imaging, and Single-Photon Emission Computed Tomography

Matthew R. Moore, MD; Jeffrey L. Saver, MD; Keith A. Johnson, MD; Jorge A. Romero, MD

• We examined a patient who exhibited Gerstmann's syndrome (left-right disorientation, finger agnosia, dyscalculia, and dysgraphia) in association with a perioperative stroke in the right parietal lobe. This is the first description of the Gerstmann tetrad occurring in the setting of discrete right hemisphere pathologic findings. A well-localized vascular lesion was demonstrated by computed tomography, magnetic resonance imaging, and single-photon emission computed tomographic studies. The patient had clinical evidence of reversed functional cerebral dominance and radiologic evidence of reversed anatomic cerebral asymmetries.

(*Arch Neurol.* 1991;48:432-435)

The localization of focal cerebral lesions causing the tetrad of symptoms—finger agnosia, left-right disorientation, agraphia, and acalculia—has been debated since Gerstmann's name was first attached to the syndrome.^{1,2} In almost all cases with focal pathologic appearance, the lesion has been localized to the left parietal cortex (angular and second occipital gyri,^{1,3} supramarginal gyrus, and superior parietal lobule⁴; superior angular, supramarginal, and inferior parietal gyri⁵; and posterior perisylvian region⁶). Strub and Gesch-

wind,⁷ reviewing all reported cases in the world literature through 1983, found only four instances in which discrete right hemisphere lesions were suspected. All of these cases, however, were reported prior to the computed tomographic (CT) era, and precise neuroanatomic localization based on their reported clinical features is difficult.

We present anatomic and metabolic studies demonstrating a right hemisphere lesion in a sinistral woman who exhibited Gerstmann's syndrome in association with a well-localized vascular lesion confirmed by CT, magnetic resonance imaging (MRI), and single-photon emission computed tomography (SPECT), indicating regional cerebral perfusion and metabolism. This case is unique to the literature in its extensive imaging of an underlying right parietal stroke.

REPORT OF A CASE

A 57-year-old left-handed white woman was seen 2 days after the operative placement of a single coronary artery bypass graft and primary closure of an atrial septal defect. During the first day postoperatively, her recovery seemed uneventful, although she did not speak very much. On the second postoperative morning, the staff noted that she seemed to be having some difficulty expressing herself, particularly with word finding, and a neurological consultation was requested.

Her medical history was notable for an inferior wall myocardial infarction 1 year prior to admission and mild hypertension. She had had no history of cerebrovascular accidents, transient ischemic attacks, or seizures and was neurologically intact on admission. The patient had a college education. The family history was notable for left-handedness in the patient's daughter.

The general medical examination was notable for a midline sternotomy incision. No carotid bruits or cardiac murmurs were observed.

On neurologic examination, the patient was alert, oriented, and cooperative. Attention was relatively intact (digit span, six forward and four backward, and preserved ability to name months backward). Language testing revealed occasional hesitations in spontaneous speech, with mild word-finding difficulties and occasional literal and verbal paraphasic error. Prosody was preserved.

Repetition was also mildly impaired, with some hesitation and occasional paraphasic substitutions. Confrontation naming of objects in the room and elements of the examiner's clothing and body parts was performed with better than 90% accuracy. Comprehension was unimpaired, as demonstrated by appropriate verbal responses to questions and correct execution of simple and multiple-step directives.

Striking right-left confusion was noted. Any task that required right-left discrimination led to hesitation, frustration, and repeated errors in execution. She was unable to identify digits on either hand; she could not reliably name the digits and made frequent errors in trying to follow commands instructing her to identify or utilize a specific digit on her own hand or on the hand of the examiner. She was aware of her difficulty and was frustrated by it. This finger-naming difficulty was in sharp contrast with her only mildly impaired ability to name objects and colors on confrontation, including low-frequency items. She was unable to perform any calculations, exclaiming "I can't visualize the math." Even very simple calculations, such as adding 12 + 13 or determining the number of nickels in 75 cents, could not be carried out. She appeared to understand the task and began calculating mentally; she then became confused, lost track of her work, and needed to start over. Her performance did not improve with the use of pencil and paper.

Reading was significantly impaired. The patient read sentences out loud slowly, ef-

Accepted for publication October 3, 1990.

From the Departments of Neurosurgery (Dr Moore) and Neurology (Drs Saver and Romero), Brigham and Women's Hospital, and the Department of Neurology (Dr Johnson), Massachusetts General Hospital and Harvard Medical School, Boston, Mass.

Reprint requests to Department of Neurosurgery, Children's Hospital Medical Center, 300 Longwood Ave, Boston, MA 02115 (Dr Moore).

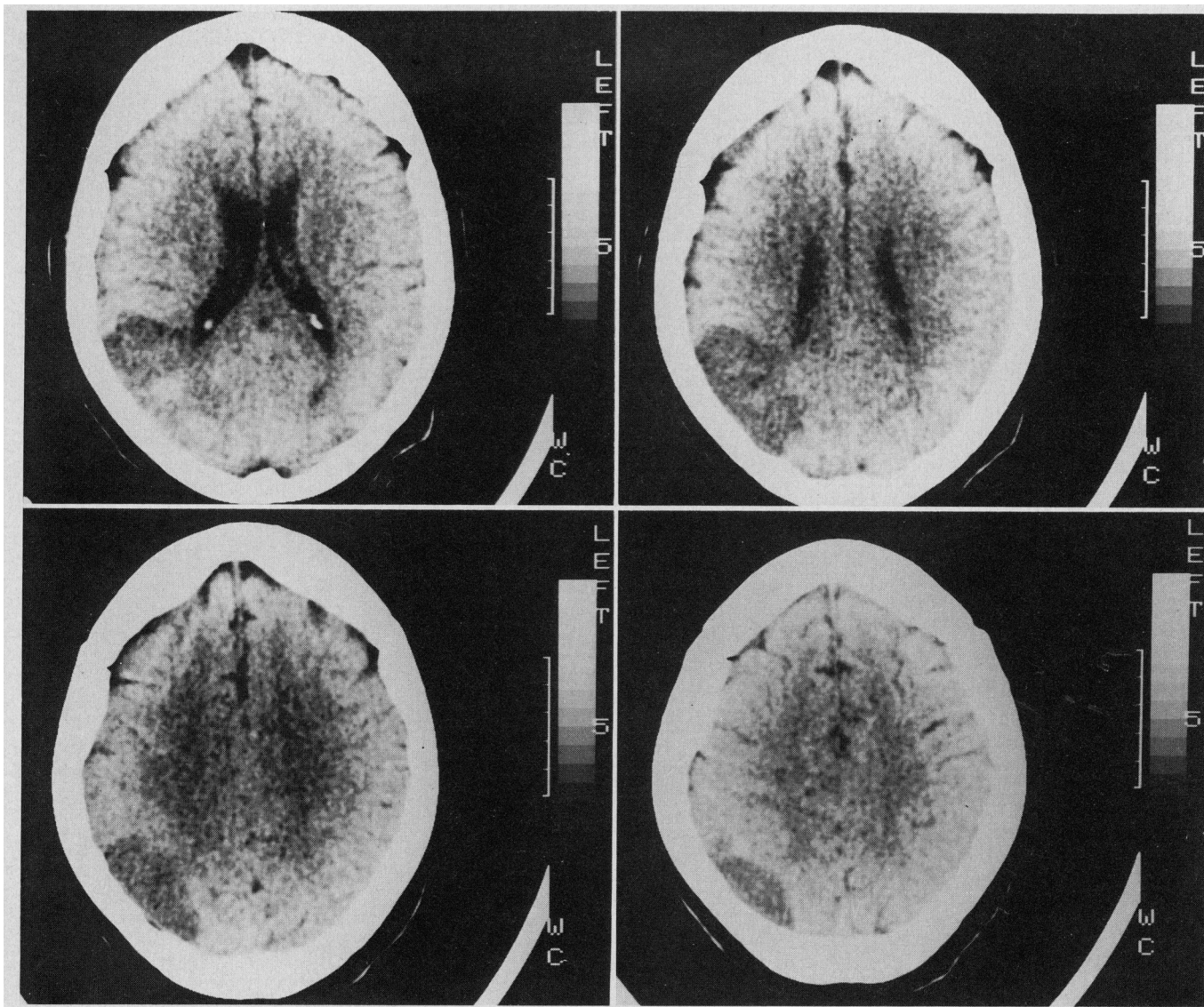


Fig 1.—Computed tomographic scans without intravenous contrast (axial, four adjacent 8-mm spaced images) taken on postoperative day 3 showing the right parieto-occipital low density in the angular gyrus (top left) extending superiorly just into inferior parietal gyrus and medially to the optic radiations.

fortfully, and monotonously, as if reading an unrelated list of words. Her comprehension of written passages was negligible. She was able to write only slowly and effortfully, with very poor penmanship, spelling errors, and omissions with drifting of the written line down and to the right on the page. When writing, she held her left hand in a “noninverted” posture. She evidenced no difficulty with constructions, drawing a clockface, and copying a cube with internal details accurately. Memory was unimpaired. She visually explored both sides of space, and there was no extinction to tactile simultaneous stimulation.

Results from the cranial nerve examination were notable for a left homonymous hemianopic visual field defect, denser in the inferior quadrant, and mild weakness of the lower face on the left side.

Examination of the extremities showed mild pronator drift of the left upper extrem-

ity but good strength on individual muscle-group testing and normal tone. Fine-motor control was unimpaired. Sensory testing to pin, touch, vibration, position, graphesthesia, and stereognosis was normal. Finger-to-nose, heel-to-shin testing was normal. Gait was normal. Reflexes were 1+ and symmetrical. Plantar responses were flexor.

The patient scored -1.0 on the modified Oldfield handedness battery.

A perioperative cerebrovascular accident in the right middle cerebral distribution was suspected. A CT scan without contrast (Fig 1) on postoperative day 3 revealed a discrete right parieto-occipital wedge-shaped hypodensity (5 × 2.5 cm) in the angular gyrus. The area of lucency began at the level of the ventricular atrium and extended superiorly through three adjacent 8-mm axial CT images to the inferior parietal lobule and medially to the optic radiations.

On postoperative day 4, the patient's

speech was fluent without paraphasic errors, and she had only minimal difficulty in repeating long grammatically complex sentences. Her facial asymmetry had resolved, and the field defect had improved to a moderate left inferior quadrant anopsia.

The patient underwent SPECT neuroimaging studies on postoperative day 7 with hexamethyl propylenamine oxime (technetium Tc 99m-HMPAO). The isotope uptake at 20 minutes after the injection shows a defect on axial view (Fig 2) of equivalent size to the hypodensity on CT scan. The three-dimensional surface reconstruction (Fig 3) places the hypoperfused region in the superior right parieto-occipital cortex (posterior angular gyrus) and underlying white matter.

On postoperative day 9, the patient underwent a cranial MRI scan in multiple planes for further evaluation of the vascular lesion. Figure 4 shows the coronal and axial images. The cerebral gyri are identified and show an

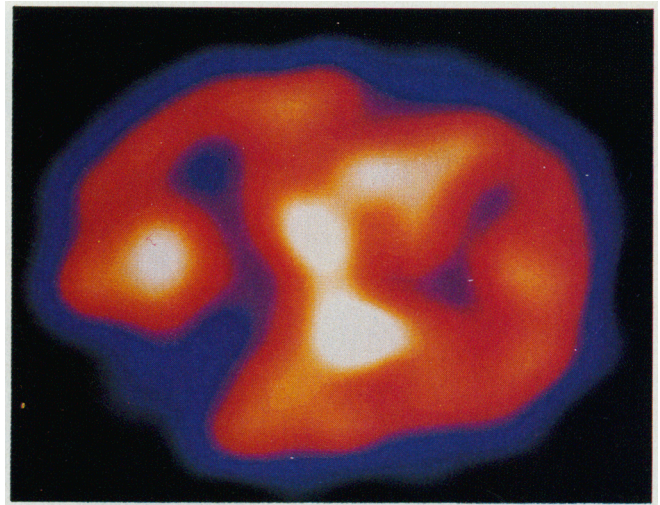
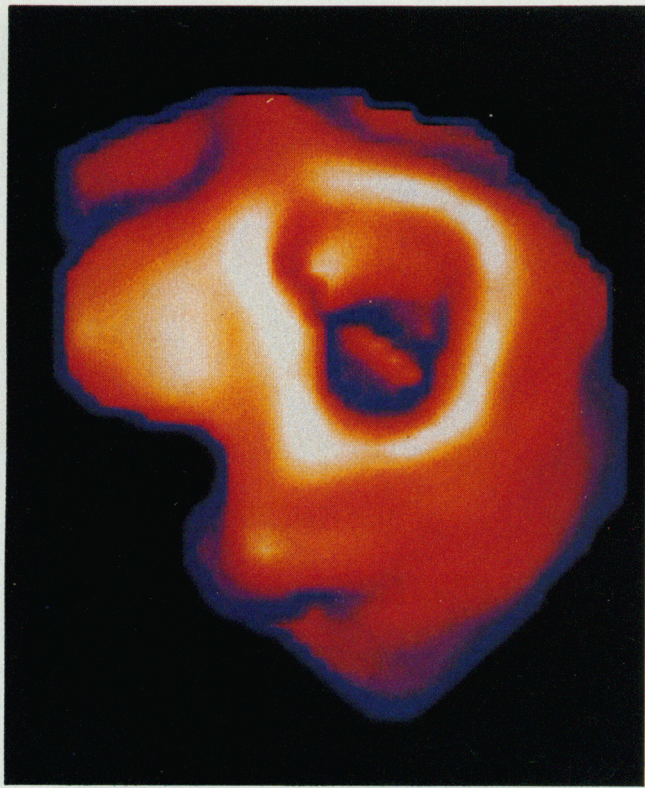


Fig 2.—Single-photon emission computed tomographic/hexamethylpropyleneamine (HM-PAO) axial image 20 minutes after injection on postoperative day 7. Note the similar perfusion defect in the right parieto-occipital region (patient's right on left of image). Color scale is such that yellow indicates regions with greatest HM-PAO concentration and dark blue indicates lowest HM-PAO activity.

Fig 3.—Three-dimensional surface reconstruction of the single-photon emission computed tomographic/hexamethylpropyleneamine images shown in Fig 2, viewed from the right posterior oblique angle. Note the shape and extensive defect inferior and posterior to the angular gyrus.

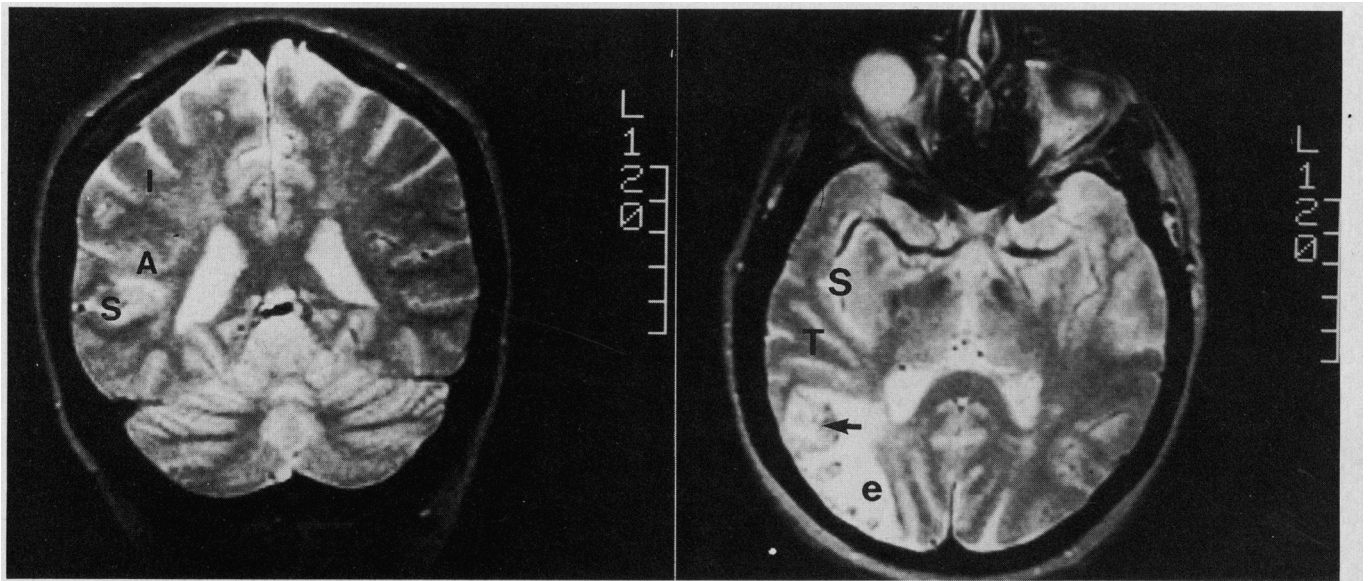


Fig 4.—Left, Magnetic resonance imaging (MRI), modified coronal plane (TR, 2000 milliseconds; TE, 35 milliseconds), anterior extent of increased signal of lesion. Inferior parietal gyrus (I), angular gyrus (A), and sylvian fissure (S) are indicated. Right, MRI, modified axial plane (TR, 2000 milliseconds; TE, 70 milliseconds), imaging through temporo-occipital lobes shows an area of heterogeneous cortical signal (arrow) in posterior inferior angular gyrus with extension of increased signal intensity (edema[e]) to adjacent occipital gyrus. The temporal operculum (T) and sylvian fissure (S) are shown.

area of heterogeneous cortical signal in the posterior inferior angular gyrus with extension of increased signal intensity (edema) to adjacent occipital gyri.

Both MRI and CT images were analyzed for anatomic asymmetries that have been proposed to correlate with functional cere-

bral asymmetry.^{9,10} Standardized methods were employed to analyze sylvian fissure course,¹⁰ fronto-occipital petalias,^{10,11} and size of planum temporale (modified).¹¹ Each of these techniques demonstrated a reversal of the most common pattern of asymmetry found in right-handed subjects: (1) the poste-

rior sylvian fissure on the right extended more horizontally and posteriorly than it did on the left (demonstrated in Fig 4, left); (2) on both MRI and CT studies, left frontal and right occipital lobes exhibited greater width and length (petalia) than did their counterparts; and (3) asymmetry of the posterior

opercular demarcation, consistent with a larger planum temporale in the right hemisphere than in the left, was noted.

COMMENT

It is recognized that the four components of Gerstmann's syndrome in an individual patient may manifest to a variable degree.⁷ In our patient, finger agnosia and left-right disorientation were prominently affected. Her agraphia, characterized by paraphasia with misspellings and syntax errors and spatial dysgraphia (poor word alignment across a page), was mild. Her acalculia was also severe, and of the type, anarithmetria,^{2,7,15} that is most commonly associated with Gerstmann's syndrome. Additionally, our patient exhibited alexia and a mild posterior aphasia—deficits that have been noted to not infrequently accompany Gerstmann's tetrad (the *angular gyrus syndrome*). The major impairment in finger identification, left-right discrimination, and calculations was disproportionate to the mild deficit in oral language function. Evidence of constructional apraxia or hemispatial neglect was absent.

We believe that ours is the first patient to be described with Gerstmann's syndrome resulting from a focal right hemisphere lesion that has been fully documented by CT, MRI, and SPECT studies. Strub and Geschwind's⁷ review of all reported cases through 1983 noted only four patients suggested to have discrete right hemisphere lesions, with 68 patients with left hemisphere lesions, 20 with bilateral lesions, and four with either left hemisphere or bilateral lesions. Our review of the literature since has yielded no additional right hemisphere cases. All four right hemisphere cases preceded the advent of modern neuroimaging. The two patients described by Heimberger et al¹⁶ are listed in a table but are not clearly identified in the accompanying individual case descriptions. It appears likely that one of the cases they listed as right hemisphere was a patient described as having a large right hemisphere glioblastoma with midline shift and an electroencephalogram displaying slowing on the left side; thus, this patient almost certainly had undergone a pathologic event bilaterally. Heimberger et al¹⁶ provided no detailed description of a second patient with clinical evidence of a discrete right hemisphere lesion. In Kinsbourne and Warrington's¹⁷ case, localization was based on accompanying clinical signs and electroencephalographic findings—no confirmatory anatomic studies were provided. Hermann and Potzl's¹⁸ 1926 case had a right occipital tumor discovered at surgery, with-

out exploration or postmortem examination of the left hemisphere to exclude a bilateral process.

Gerstmann suggested that the symptom cluster of finger agnosia, right-left disorientation, agraphia, and acalculia indicated lesions in the dominant parietal lobe, situated in the great proportion of right-handers and most left-handers in the left hemisphere. We hypothesize that our patient had strongly reversed functional cerebral lateralization so that a right parietal lesion could produce the constellation of signs of Gerstmann's syndrome. Our patient was strongly left-handed (modified Oldfield handedness score, -1). A family history of left-handedness in a first-degree relative was known. (Evidence of a genetic component to determination of handedness has been reviewed by Levy.¹⁹) The patient wrote with a noninverted hand posture, which has been suggested to indicate likely contralateral hemispheric (here, right hemispheric) dominance for speech.²⁰ The patient also experienced alexia and mild posterior aphasia following her isolated right hemisphere insult, further suggesting that she had had major representation of language function in the right hemisphere.

Moreover, a number of investigators have identified anatomic asymmetries that may correlate with functional cerebral asymmetry.⁸⁻¹⁴ Our patient exhibited reversed asymmetry for several of these anatomic findings. The posterior sylvian fissure on the right extended more horizontally and posteriorly than did its counterpart, suggesting that the temporal and parietal opercula were larger on the right than on the left. The planum temporale, a major component of the temporal speech region of Wernicke, was larger in the right hemisphere than in the left. Thus, our patient exhibited both a pattern of anomalous functional motoric and linguistic dominance and a reversal of anatomic brain asymmetries that may comprise the biologic substrate of functional lateralization.

In addition to the variable deficit severity of each component in Gerstmann's syndrome,⁷ some have remarked that deficit improvement occurs at different rates longitudinally.²¹ Similarly, we noted improvement in this patient's writing ability and right-left orientation at 1 week postoperatively (postvascular event).

In this well-documented instance of a focal right hemisphere lesion producing Gerstmann's syndrome, the pathologic event was situated primarily in the posterior parietal lobe. Thus, this case provides further evidence to supplement

the findings drawn from numerous previously reported left hemisphere cases that the lesion most likely to produce Gerstmann's syndrome is placed in the dominant posterior parietal region.

The authors thank Mathias-Victor Schromm for bibliographic assistance.

References

1. Gerstmann J. Fingeragnosie und isolierte Agraphie, ein neues Syndrom. *Z Gesamte Neurol Psychiatr.* 1927;108:381-402.
2. Gerstmann J. Syndrome of finger agnosia, disorientation for right and left, agraphia and acalculia. *Arch Neurol Psychiatry.* 1940;44:398-408.
3. Levine DN, Mani RB, Calvanio R. Pure agraphia and Gerstmann's syndrome as a visuospatial-language dissociation: an experimental case study. *Brain Lang.* 1988;35:172-196.
4. Hrebeek V. Pathophysiologic interpretation of Gerstmann's syndrome: definition of new broader syndrome of interparietal disconnection: their value for topical diagnostics of lesions in the dominant cerebral hemisphere. *Neuropsychologia.* 1977; 11:377-388.
5. Roeltgen DP, Sevush S, Heilman KM. Pure Gerstmann's syndrome from a focal lesion. *Arch Neurol.* 1983;40:46-47.
6. Morris HH, Luder H, Lesser RP, Dinner DS, Hahn J. Transient neuropsychological abnormalities (including Gerstmann's syndrome) during cortical stimulation. *Neurology.* 1984;34:877-883.
7. Strub RL, Geschwind N. Localization in Gerstmann syndrome. In: Kertesz A, ed. *Localization in Neuropsychology.* New York, NY: Academic Press Inc; 1983:295-321.
8. Geschwind N, Levitsky W. Human brain: left-right asymmetries in temporal speech region. *Science.* 1968;161:186-187.
9. Geschwind N, Galaburda AM. Cerebral lateralization. *Arch Neurol.* 1985; 42:428-459.
10. Damasio H, Damasio AR. *Lesion Analysis in Neuropsychology.* New York, NY: Oxford University Press Inc; 1989:133.
11. Pienadz JM, Naeser MA. Computed tomographic scan cerebral asymmetries and morphologic brain asymmetries. *Arch Neurol.* 1984;41: 403-409.
12. Bear D, Schiff D, Saver J, Greenberg M, Freeman R. Quantitative analysis of cerebral asymmetries: fronto-occipital correlation, sexual dimorphism, and association with handedness. *Arch Neurol.* 1986;43:598-603.
13. Kertesz A, Black SE, Polk M, Howell J. Cerebral asymmetries on magnetic resonance imaging. *Cortex.* 1986;22:117-127.
14. Stratta P, Rossi A, Gallucci M, Amicarelli I, Passariello R, Casaccia M. Hemispheric asymmetries and schizophrenia: a preliminary magnetic resonance imaging study. *Biol Psychiatry.* 1989; 25:275-284.
15. Hecaen H, Angelergues R, Houllier S. Les varietes cliniques des acalculies au cours des lesions retro-roladiques: approche statistique du probleme. *Rev Neurol.* 1961;105:85-103.
16. Heimburger RF, Demyer W, Reitan RM. Implications of Gerstmann's syndrome. *J Neurol Neurosurg Psychiatry.* 1964;27:52-57.
17. Kinsbourne M, Warrington EK. A study of finger agnosia. *Brain.* 1962;85:47-66.
18. Hermann G, Potzl O. Über die Agraphie und ihre lokaldiagnostischen Beziehungen. Berlin, Germany: S Karger AG; 1926.
19. Levy J. A review of evidence for a genetic component in the determination of handedness. *Behav Genet.* 1976;6:429-453.
20. Levy J, Reid M. Variations in writing posture and cerebral organization. *Science.* 1976; 194:337-339.
21. Varney NR. Gerstmann syndrome without aphasia: a longitudinal study. *Brain Cogn.* 1984;3:1-9.