

Two Cases with Cerebral Infarction in the Left Middle Frontal Lobe Presented as Gerstmann's Syndrome

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Abstract

Gerstmann's syndrome is a neuropsychological disorder characterized by four symptoms, namely, acalculia, finger agnosia, left-right disorientation, and agraphia suggesting the presence of a lesion in the inferior parietal lobule of the dominant hemisphere, especially at the angular gyrus. Several descriptions of Gerstmann's syndrome have been reported in association with a lesion to the left frontal lobe, but none of these reports fulfilled the full tetrad of diagnostic criteria. Herein, we report two cases with all four symptoms of Gerstmann's syndrome associated with an uncommon area in the left middle frontal lobe without an angular gyrus lesion. This apparent discrepancy could be explained by disconnection of functional fiber between the frontal and parietal cortex. Moreover, left frontal cortex or the subcortical area may be an important functional area which is closely connected with the parietal lobe in Gerstmann's syndrome.

Keywords: Gerstmann's syndrome; Frontal lobe; Angular gyrus; Parietal lobe; Disconnection syndrome

an uncommon lesion in the left frontal lobe without angular gyrus lesion.

Introduction

Josef Gerstmann first described a group of symptoms, namely, acalculia, finger agnosia, left-right disorientation, and agraphia, and interpreted this association as a specific syndrome resulting from damage to the left parietal lobe, especially the angular gyrus [1]. He suggested that a common cognitive denominator, or Grundstörung, essentially underlies to these four characteristic cognitive faculties.

However, it remains debatable whether Gerstmann's syndrome can be induced by a focal lesion without other symptoms or whether the angular gyrus is the functional structure in cognition for the tetrad [2,3]. Although many reports have mentioned that Gerstmann's syndrome can present in different neurologic disorders and is associated with an extensive lesion in the left hemisphere or other locations, there are many other well-documented cases with pure Gerstmann's syndrome and reports that demonstrated transient neuropsychological abnormalities including the tetrad of Gerstmann's syndrome which was produced by electrical stimulation in the posterior-perisylvian area [4-6]. These previous papers provide us evidences that Gerstmann's syndrome does exist and has a localizing value.

Recent functional imaging and diffusion tensor tractography studies indicate that Gerstmann's syndrome develops when the white matter tract connecting various cortical and subcortical regions is disrupted at an area underlying the parietal cortex, especially the angular gyrus, in which different functional connections exhibit maximal spatial proximity [7-9]. Furthermore, several case reports on Gerstmann's syndrome caused by a lesion to the left frontal lobe support this disconnection hypothesis, but none of these cases exhibited the full tetrad of the diagnostic criteria [10-12]. In this report, we present two cases with all four symptoms of Gerstmann's syndrome each caused by

Case Report

Case 1

The patient was an 83-year-old right-handed female with six years of school education. She had a 5-year history of diabetes mellitus, but no prior history of a neurological problem. She experienced word finding difficulties of sudden onset and was admitted to our hospital. On admission she was alert and cooperative. On neurological examination, she was disoriented with respect to time and place. Her visual acuity and visual field were normal, and no other central nervous system abnormalities were detected. Motor examination of the extremities and sensory testing to pin prick, touch, vibration, and position sense were normal. Finger-to-nose and heel-to-shin testing were normal, and gait was grossly normal. Language testing revealed impairment of spontaneous speech when the patient explained her daily life or described a complex picture. Repetition was relatively intact, even long sentence repetition. The Korean version of the Boston naming test and the confrontation naming test revealed naming was also impaired. Object knowledge and word comprehension were better preserved than naming, as demonstrated for the latter by appropriate verbal responses to questions and correct execution of commands. Further neuropsychological examination showed right-left disorientation, finger agnosia, agraphia, and acalculia. She was unable to perform the command to move the same finger on the other hand that has just been touched by the examiner. Furthermore, any task requiring right-left discrimination led to hesitation, frustration, and repeated errors in execution. In addition, despite preserved number processing, she was unable to perform any calculations, even very simple calculations, such as, subtracting 7 from 100. She was unable to draw or write letters or words during spontaneous writing, dictation, or copying. Magnetic resonance imaging (MRI) performed on

admission showed diffusion restriction lesions at the subcortical area of the left middle frontal lobe in the dominant hemisphere (Figure 1a).

Case 2

A 66-year-old right-handed female with nine years of school education presented with a sudden language disturbance, which had started four days ago. She was treated for hypertension for one year. On neurologic examination, she was alert and well oriented, but showed slightly attention impairment. Her visual acuity and visual field were normal, and there were no other central nervous system abnormality. Motor and sensory examinations of the extremities, finger-to-nose and heel-to-shin testing, and gait were normal. Her spontaneous speech was impaired by agrammatism and apraxia of speech. Syllable strings and sentence repetition were also impaired.

Confrontation naming test exhibited naming difficulty (accuracy 60%), but the commands and syntax comprehension test revealed comprehension was relatively preserved. This patient displayed all four symptoms of Gerstmann's syndrome (right-left disorientation, finger agnosia, agraphia, and acalculia). Her writing showed frequent omissions and substitutions, resulting in unintelligible sentences. When a command 'touch your left ear with your right hand' was given, she made errors and had difficulty distinguishing left from right on the examiner. She had severe anarithmia for both written and oral calculations, and was unable to identify digits on either hand and could not performed the task when we asked her to move the same finger on contralateral hand touched by the examiner. Acute infarction at the left middle frontal lobe was visualized by brain MRI (Figure 1b).

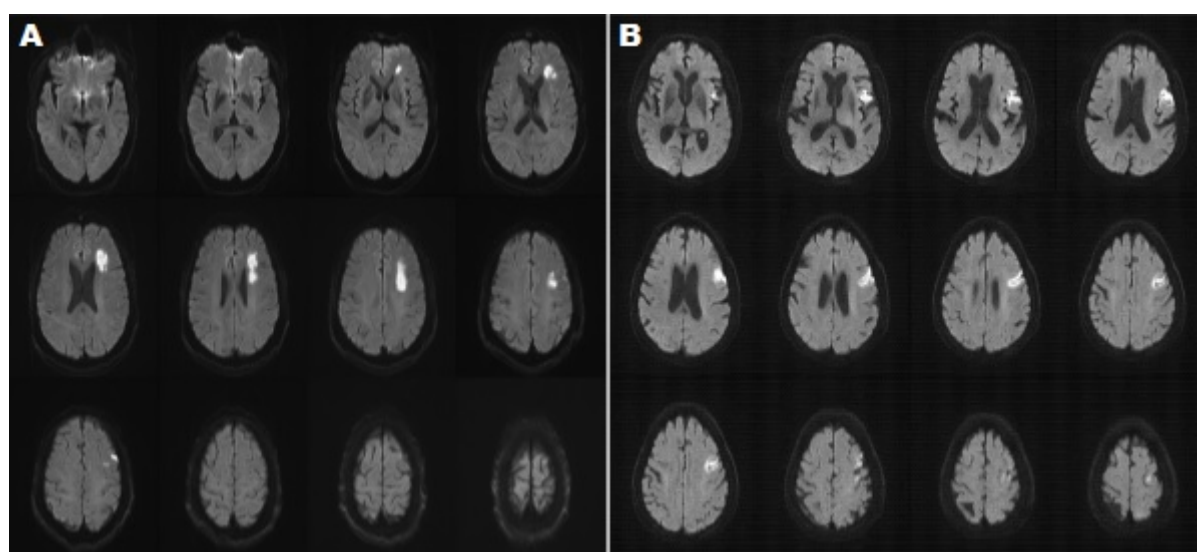


Figure 1: Brain MR images obtained at admission showing diffusion restriction lesions at the subcortical area of the left middle frontal lobe in Case 1 (Figure 1a) and at the cortex of the left middle frontal lobe in Case 2 (Figure 1b).

Discussion

In both described cases, patients showed right-left disorientation, finger agnosia, agraphia, and acalculia with transcortical motor or motor aphasia by left middle frontal lobe infarction. The acute infarction lesions were confirmed by MRI to be localized in the cortex or subcortex of left middle frontal lobe.

In both cases, patients initially showed mild disorientation or slightly decreased concentration. The lesions observed affected Brodmann area 46 and underlying white matter, which are associated with attention and working memory, meaning that a lesion in this region could disturb general cognitive function and cause secondary cognitive symptoms by disrupted attention and information-processing capacity. However, as Alexander et al. [13] pointed out that it does not seem proper to explain specific cortical symptoms like those associated with Gerstmann's syndrome based only on the disturbance of attention or arousal.

Transcortical motor aphasia or motor aphasia in our patients could be partially responsible for the agraphia which is a part of Gerstmann's syndrome. However the other symptoms, that is finger agnosia, acalculia, and right-left disorientation could not be explained by

transcortical motor or motor aphasia because we approached the examination cautiously to avoid the influence of language impairment. The causative role of aphasia in Gerstmann's syndrome has been controversial. Heimburger et al. and Poeck and Orgass reported that in almost all of their patients with Gerstmann's syndrome had aphasia concomitantly and symptoms of Gerstmann's syndrome were directly or indirectly related to language [3,14]. In contrast, Kinsbourne and Warrington, who described five patients with complete Gerstmann's syndrome and constructional apraxia without aphasia, concluded that no causal relationship existed between aphasia and Gerstmann's syndrome [15,16]. In addition, Strub and Geschwind, who described a patient with full Gerstmann's syndrome without aphasia, concluded that aphasia was not a necessary underlying factor but rather a possibly associated factor in Gerstmann's syndrome [17]. Therefore, we believe that transcortical motor or motor aphasia in our patients does not adequately explain the symptoms of Gerstmann's syndrome.

Interestingly, both of our patients showed completely impaired calculation skills but preserved number processing. Dyscalculia has been analyzed using two distinct systems, that is, one for number processing and the other for calculation [18]. Although the majority of previously reported cases of isolated acalculia had a parieto-occipital

lesion [19], Lucchelli and Renzi described a case with a left frontal lobe lesion that showed a deficit strictly confined to calculation skill with preservation of number processing [11,12]. A recent study using functional MRI demonstrated that left inferior prefrontal cortex and bilateral angular regions are associated with exact calculation and dorsolateral and superior prefrontal regions involves in approximation [20]. Accordingly, dyscalculia which is caused by left frontal lobe infarct in our cases strongly concurs with previous reports and the recent functional MRI study.

Location of anatomical lesions associated with agraphia has been variously reported. Several authors have reported an association between agraphia and the parietal lobe and suggested the angular gyrus acts as a graphic center. However, although agraphia associated with a left frontal lesion is rare, several case reports have described a frontal lesion presenting with agraphia and explained as the interruption of intrahemispheric association fibers between the parietal and frontal cortices [10,14,21,22]. Agraphia in our cases could be explained by a left frontal cortex or subcortical lesion disrupting the functional connecting fiber from the graphic center located in the parietal lobe to the frontal cortex.

Furthermore, both of our patients showed typical right-left disorientation and finger agnosia, which are disorders of the body scheme and are typically associated with a left parietal lesion, especially the angular gyrus. However, reports on right-left disorientation and finger agnosia caused by a left frontal lobe lesion are rare. Body perception can be disrupted by a deficit in neural substrate or by deficit of the link between embodiment and multisensory processing for body parts in the frontoparietal cortex [23]. The posterior parietal and premotor cortices are involved in the coding of embodiment, and thus, lesions in these areas can lead to embodiment disturbances. Therefore, we hypothesize that a lesion of the frontal lobe may cause a defect of embodiment presenting as finger agnosia and right-left disorientation as symptoms of a disturbed body scheme.

Although several cases of a frontal lobe lesion showing incomplete Gerstmann's syndrome have been reported, our cases are rare because left middle frontal lobe lesion was associated with all components of Gerstmann's syndrome, especially right-left disorientation and finger agnosia. Because, in our cases, the same Gerstmann's syndrome was exhibited regardless of cortical and subcortical lesion in frontal lobe, moreover, which is uncommon location for Gerstmann's syndrome, this finding demonstrates that functional cortical cognitive denominator such as angular gyrus is not critically necessary for development of Gerstmann's syndrome. Therefore, we believe that the disconnection hypothesis is rather responsible for development of Gerstmann's syndrome as described in recent functional neuroimaging studies [9]. This finding also suggests that the frontal lobe may be a functionally important area connected with the parietal lobe in Gerstmann's syndrome, and patients with a frontal lobe lesion require close observation.

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