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## A case of inferior frontal gyrus infarction manifesting Gerstmann syndrome

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### ABSTRACT

A 48-year-old female suffered from cerebral infarction involving the left inferior frontal gyrus. This was due to ischemic complications of endovascular treatment for subarachnoid hemorrhage. She exhibited severe acalculia, agraphia, finger agnosia, and right-left disorientation (the four features of Gerstmann syndrome), but aphasia was scarcely noticeable. Single-photon emission tomography revealed hypoperfusion in the left inferior frontal area and also in the left parietal area. It is possible that Gerstmann syndrome was caused in the present case by disruption of the association fiber connecting the inferior frontal area with the inferior parietal area.

### ARTICLE HISTORY

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### KEYWORDS

Cerebral infarction; inferior frontal gyrus; Gerstmann syndrome; SPECT; disconnection syndrome

### Introduction

The combination of finger agnosia, right/left disorientation, acalculia, and agraphia is known as Gerstmann syndrome (Gerstmann, 1940). This is well known but remains controversial in clinical neurology because the four symptoms are not always present together and because of the issue of localization of the lesion.

There is controversy over whether Gerstmann syndrome is caused by a focal cerebral lesion or by disruption of the cortical network. Concerning the focality of the syndrome, the left angular gyrus, included in the inferior parietal lobule or its adjacent parietal subcortical white matter (Long et al., 2019; Mayer et al., 1999; Morris et al., 1984; Roeltgen et al., 1983) is believed to be the region responsible. It has nevertheless been asserted that the Gerstmann tetrad is unlikely to arise from a single lesion, but rather from a disturbed cortical network involving subcortical parietal white matter (Rusconi et al., 2010), where the association fibers join different cortical and subcortical regions.

To our knowledge, cerebral infarction of the left inferior frontal gyrus with Gerstmann syndrome has scarcely been reported in the English-language literature (João et al., 2017; E. Lee et al., 2016). We report here a patient with infarction in the left inferior frontal lobe (so-called Broca's area) manifesting the four features defining Gerstmann syndrome, and we discuss its pathophysiological mechanism. This report supports the view that Gerstmann syndrome is a disconnection syndrome.

### Case presentation

A 48-year-old and corrected right-handed female (Edinburgh handedness inventory score +68.4, indicating right-handedness in this test) (Oldfield, 1971) was transferred to

our emergency room with a complaint of severe headache and blurred vision lasting for 5 days. Consciousness and orientation were not disturbed, and no focal neurological deficits were present. CT of the brain disclosed a diffuse subarachnoid hemorrhage. An emergent cerebral angiogram was conducted, but failed to reveal the source of the bleeding. Ventriculomegaly then developed and ventriculostomy was performed via the right frontal horn of the lateral ventricle. Repeated angiogram showed a gradually increasing aneurysmal bulge on the medial wall of the left supraclinoid internal carotid artery. Stent-assisted coil embolization was performed (B. H. Lee et al., 2009) 12 days after admission. Rebleeding from the aneurysm was prevented, but ischemic complications following the procedure resulted in the infarction in the left frontal lobe.

Initially, after the intervention, the patient displayed disturbance of consciousness and severe hemiparesis on the right side. Those symptoms diminished gradually, and on the postoperative day (POD) 3 she became independent in her mobility and oral intake. She was fully awake and oriented. No motor weakness was observed. She initially showed mild expressive aphasia but could obey verbal commands correctly. Her speech capability improved further on POD6.

On POD11, neuropsychological assessment revealed very mild aphasia. She could name common objects from pictures shown to her with an accuracy of 80%. Her word fluency was slightly impaired (categorical fluency test: eight words per minute). Repetition was not disturbed. She could obey verbal commands and read simple words accurately. But she was unable to read even a simple sentence. Agraphia was severe and was characterized by difficulty writing letters and words spontaneously, dictated, or copied. Ideomotor apraxia for transitive and intransitive gestures was not observed. Acalculia for low complexity addition and subtraction sums was also obvious and severe. Interestingly, she could read

aloud the numerical characters. Finger agnosia (difficulty recognizing the fingers of one's own hand and the examiner's hand, inability to follow commands to move or imitate finger movements or gestures), and right-left disorientation, (e.g., when requested to point to her left ear with her right hand) were also seen.

A fluid-attenuated inversion recovery image (FLAIR) MRI of the brain (Figures 1, 2) showed the ischemic lesion mainly to be in the inferior frontal gyrus, including the insular cortex, but not in the parietal area on the left side. The hyperintense lesion in the right frontal lobe shows the pattern of ventricular drainage.  $^{99m}\text{Tc}$ -single photon emission computed tomography (SPECT) disclosed reduced cerebral blood flow (CBF) in the left inferior frontal area and also in the left inferior parietal area (Figure 3).

Intensive rehabilitation was provided in a specialized ward. On POD 60, finger agnosia and right-left disorientation improved, but acalculia and agraphia persisted. She was able to resume an independent pattern of daily life.

## Discussion

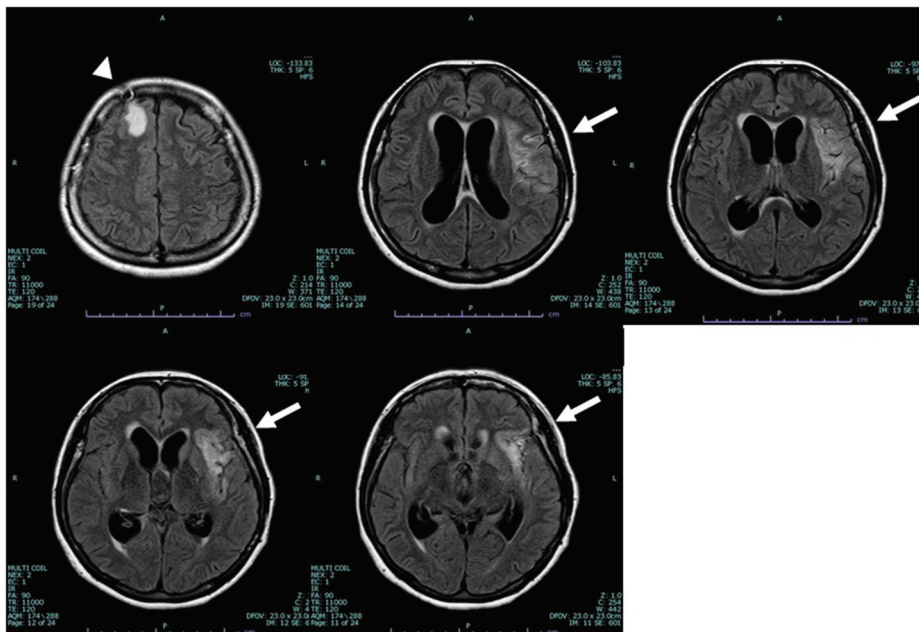
This case showed the definite cerebral infarction involving the left inferior frontal gyrus, so-called Broca's area, and exhibited the complete form of Gerstmann syndrome, which is usually a manifestation of dominant inferior parietal lobe lesion. Our CBF study demonstrated hypoperfusion not only in the left inferior frontal area but also in the left parietal area.

Gerstmann syndrome involves four symptoms: finger agnosia, right/left disorientation, acalculia, and agraphia. A focal lesion in the dominant angular gyrus or its adjacent subcortical white matter is generally accepted as the cause of this syndrome (Gerstmann, 1940; Long et al., 2019; Mayer et al., 1999). Some researchers doubt that focality produces all four

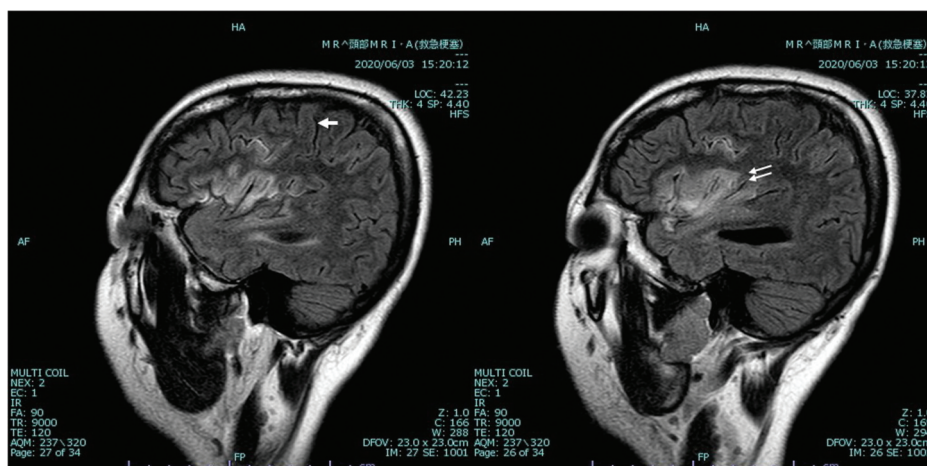
symptoms of this syndrome, however. Rusconi et al. (2009) investigated healthy subjects using functional MRI (fMRI) and the fiber tracking method, and showed that Gerstmann syndrome is due to disruption of subcortical associative fibers underlying the inferior parietal area. They did not mention the role of frontoparietal connection, even in their review article (Rusconi et al., 2010). In their article, however (Figs. 3 and 5 in Rusconi et al. (2009), the connecting fibers between the left inferior parietal lobule (including angular gyrus) and the left inferior frontal gyrus were depicted.

Regarding the involvement of frontal lobes in Gerstmann syndrome, only three articles including a total of five patients have been reported in the literature (João et al., 2017; H. Lee et al., 2013; E. Lee et al., 2016). Those cases manifesting the four symptoms of Gerstmann syndrome had lesions in the inferior and/or middle frontal gyrus with sparing lesion in the angular/supramarginal gyri. These authors speculated that this syndrome was caused by the disconnection of associative fibers connecting the frontal and parietal lobes; this hypothesis matches our view. They did not make a CBF study showing the disturbance of parietal areas, as in our case.

A reduction in the cerebral blood flow in the left parietal lobe, distant from ipsilateral inferior frontal gyrus, indicates the diaschisis mechanism (Feeney & Baron, 1986). This ipsilesional functional depression would be mediated via associative fibers connecting the distant cortical regions. In this regard, it is speculated that the long association fibers could play some role. Among them, the superior longitudinal fascicle (SLF), together with arcuate fascicle, constitutes the dorsal language stream, which controls phonological (word production) processing of speech (Friederici, 2015; Yagmurlu et al., 2016). The frontoparietal network, called the SLF, connects the posterior language areas to the precentral gyrus, and but to Broca's area



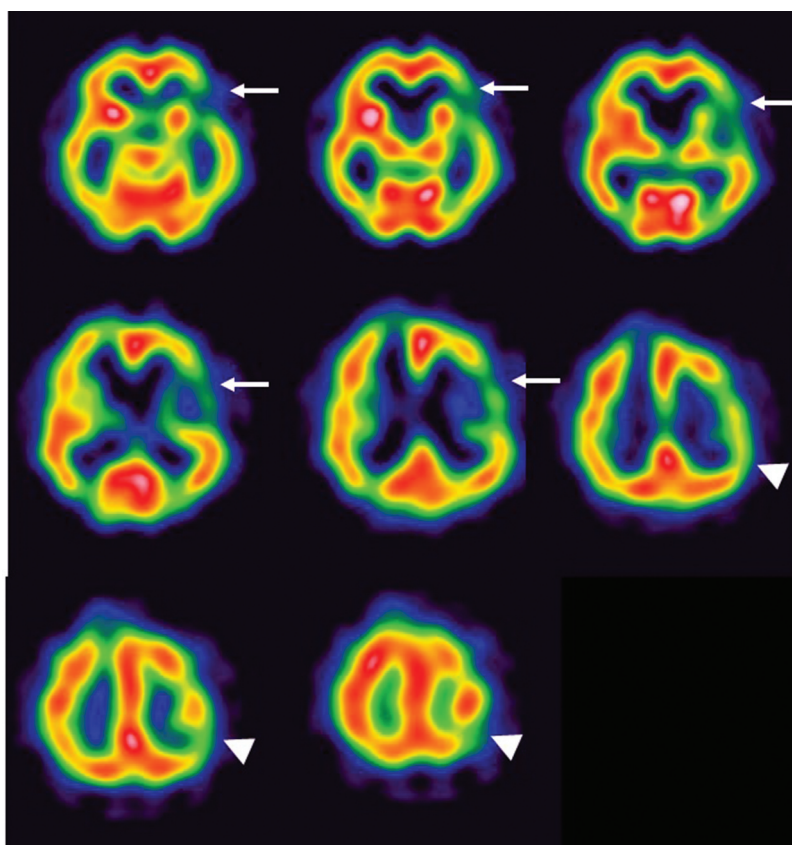
**Figure 1.** Axial view of FLAIR image of brain MRI showing cerebral infarction involving the left inferior to middle frontal gyrus, including insular cortex (white arrows) but not the parietal lobe. The high-intensity lesion in the right frontal tip area (white arrow head) indicates the pattern of external ventricular drainage at an acute stage.



**Figure 2.** Sagittal view of [Figure 1](#). These images show the cerebral infarction extending from the inferior frontal gyrus to the lower part of the precentral gyrus. White arrow: central sulcus, Double white arrow: precentral sulcus.

only marginally in about half of cases (Bernal & Altman, 2010). The SLF has three parts: SLF I (dorsal), II (middle) and III (ventral). We believe disruption of white matter fiber tracts connecting the inferior frontal lobe and the inferior parietal cortex (such as third branch of the superior longitudinal fasciculus or SLF III) caused Gerstmann syndrome in our case (Frey et al., 2008; Friederici, 2015; Yagmurlu et al., 2016).

In the acute stage, the patient showed mild aphasia. This might be consistent with the recently proposed mechanisms of Gerstmann syndrome; (1) the pathological process is located in the left parietal white matter disconnecting separate cortical networks and (2) it represents a disturbance in the ability to verbally mediate some spatial knowledge (Ardila, 2020). As regards agraphia and acalculia, the opercular part of the precentral gyrus could constitute the phonological route in writing



**Figure 3.** Cerebral blood flow study according to  $^{99m}\text{Tc}$ -single photon emission computed tomography (SPECT) shows the hypoperfusion area not only in the left inferior/middle frontal area (white arrow) but also in the ipsilateral parietal area (white arrowhead).

that conveys phonological information of syllable sequences, and its damage might cause phonological agraphia and acalculia with reduced verbal short-term memory (Sakurai et al., 2018).

## Limitations

This is a single case report, so it is not possible to generalize our interpretation. Furthermore, as mentioned in (Benton, 1961), aphasia during the acute stage would affect the symptoms. Observations of this patient nevertheless suggest that white matter fiber tracts, particularly SLF III, may be responsible for the symptomatology of Gerstmann syndrome.

We did not perform other anatomico-functional neuroimaging studies, such as fiber tracking with diffusion tensor imaging or functional MRI. The patient's handedness profile, the corrected right-handedness, might also contribute to the symptoms in this report. Further study is needed to clarify the role of this white matter tract on the mysterious combination of symptoms constituting Gerstmann syndrome.

## Conclusion

A patient with left inferior frontal lobe infarction in association with decreased CBF in the left parietal lobe exhibited Gerstmann syndrome. Disconnection of white matter fiber connecting the inferior frontal and parietal gyri on the left side may be a pathophysiological mechanism of Gerstmann syndrome.

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## Disclosure statement

No potential conflict of interest was reported by the authors.

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