



Inadvertent Discovery of Gerstmann's Syndrome During a Skull MRI and Its Anatomical Basis: A Case Report

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Abstract: A rare neurological disorder in which an individual is unable to perform tasks related to language, spatial recognition, mathematics, and cognition is known as Gerstmann's syndrome(GS). Gerstmann hypothesised that the four symptoms that makeup Gerstmann's syndrome are caused by a shared cognitive problem (Grundstörung). Gerstmann asserted that it is a body schema disorder that only affects the hand and fingers. Since then, the existence of a Grundstörung has been disputed. The study proposed a shared psychoneurological element, but it should be connected to changes in mental pictures rather than the bodily schema. The significant facts appear to be the common simultaneous impairment of writing, computation, and bodily awareness after left parietal lesion, beyond the diagnostic label of "pure" and "non-pure" Gerstmann's syndrome instances. Some functional domains can be linked to patients' symptoms despite being heterogeneous. A 65-year-old male was brought to the hospital with a history of injury to the head and complaints of confusion, speech difficulty, inability to perform simple calculations, severe headache, and drowsiness. The patient was sent to the radiology department of Sree Balaji Medical College and Hospital, Chennai, for a Magnetic Resonance Imaging of Skull (MRI). The MRI study revealed chronic ischemic changes in periventricular and subcortical white matter in the parietal lobes. Certain ischemic changes were also observed in the frontal lobe, suggesting age-related changes and Gerstmann's syndrome. By cutting off various portions of the inferior parietal cortex, a lesion affecting subcortical and subangular bundles of fibers affects many circuits with distinct roles. Therefore, the deficits seen in the patient are best explained by this anatomical explanation.

Keywords: Neurological disorder, Cognition, Gerstmann's syndrome, Magnetic Resonance Imaging, Grundstörung.

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I. INTRODUCTION

An uncommon neurological condition called Gerstmann's syndrome impairs a person's capacity for carrying out specific cognitive activities connected to language, math, and spatial perception. It bears Gerstmann's Syndrome after the Austrian neurologist Joseph Gerstmann discovered it in 1924. Gerstmann's biographical summary is given to place the historical significance of his scientific contribution. Joseph Gerstmann earned his medical degree in Vienna in 1912, where he also underwent his training. In 1930, he was appointed chief of the Maria-Theresien-Schlossel Neurological Institute. He immigrated to the United States in 1938 to avoid Nazi persecution following the Anschluss of Austria. He worked as a neurologist until his death, first in Springfield, Ohio, then in Washington, D.C. The case of a 52-year-old woman who was admitted to the Vienna Psychiatric Department due to the abrupt onset of schizophrenia was described by Gerstmann (1924; see Rusconi & Cubelli, 2019 for the English translation of the original case report and critical analysis). Right hemianopia, calculation deficits (inability to handle numbers, comprehend them, or calculate)¹, writing deficits (jumble of letters and unreadable scrawls), right-left orientation/discrimination disorders of her own body, low confidence in distinguishing right from left in the outside environment, and impaired ability to recognize, identify, differentiate, and name fingers (a deficit that was later called finger agnosia) are all symptoms she had.^{2,3} There were no indications of apraxia or aphasia identified. According to Gerstmann, this anomaly was caused by a selective body schema impairment that only affected the representation of hands and fingers.^{4,6} Agraphia (the inability to write), acalculia (the incapacity to execute mathematics), finger agnosia (the incapacity to differentiate fingers), and left-right confusion are the key symptoms of Gerstmann's syndrome.⁷ These signs are thought to result from damage to the brain's parietal lobe, which organizes movement and integrates sensory data.⁸ Agraphia, or the inability to write,

can show itself as problems with spelling or letter formation. Agraphia patients may struggle to write their names, copy text, or express themselves in writing.⁹ This can be a very irritating symptom for those affected, especially those who rely on writing communication in their professional or personal lives. The inability to do simple mathematical operations like addition, subtraction, multiplication, and division is known as acalculia. This may impair one's capacity for handling money, balancing checks, or figuring out tips in a restaurant. Acalculia can also make it difficult for a person to understand algebraic or calculus principles.¹⁰ When a person cannot discriminate between different fingers, they may find it difficult to recognize their or other people's fingers. The execution of daily actions like buttoning a garment, tying shoelaces, or using utensils may be impacted by this. Using one's fingers to point at things or describe particular digits to other people can also be challenging. The propensity to confuse left and right is known as left-right confusion, and it can be challenging for someone to navigate their environment, follow instructions, or carry out duties that call on spatial orientation. This can be particularly difficult for people who need to process and react to spatial information fast, like drivers, pilots, or sports. A stroke, an accident, or a disease that damages the brain's parietal lobe frequently results in Gerstmann's syndrome, a relatively uncommon ailment. A neurological examination, imaging scans, and a battery of neuropsychological tests¹¹ are frequently used to make a diagnosis. Occupational therapy, speech therapy, and physical therapy are frequently used as treatments for Gerstmann's syndrome to assist patients in developing their cognitive and motor abilities. To address the underlying causes of the syndrome, it may occasionally be advised to provide drugs or undergo surgical procedures. Gerstmann's syndrome, a rare neurological condition, can adversely impact the capacity to execute daily tasks involving language, algebra, and spatial recognition.¹² Although there is no cure for the syndrome, several measures can help those with it enhance their quality of life and control their symptoms.



Injury to Parietal Lobe,
resulting in Gerstmann's
syndrome

Fig 1: The image represents the cerebral cortex with the highlighted parietal lobe, the main area affected by Gerstmann's syndrome. (Representation created using Dall-E -Open AI software)

2. METHODS

2.1. Case Report



Fig 2: The image from the MRI scan with an area marked in red representing the affected parietal lobe.

A 65-year-old male patient with a history of injury to the head and complaints of confusion, speech difficulty, inability to perform simple calculations, poor hand grip, severe headache, and drowsiness for the past 3 days was referred to

the radiology department of Sree Balaji Medical College and Hospital, Chennai. The patient had a history of hypertension and was a known case of diabetes mellitus. The patient exhibited certain neuropsychiatric manifestations.

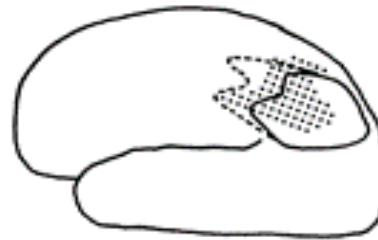


Fig 3: Schematic representation of Gerstmann's syndrome where the continuous line marks the lesion area and the dashed line the oedema area

2.2 Neuroradiological Study¹³

The patient was advised to have an MRI brain, and during the scan, the patient was unable to understand the instructions, the speech wasn't clear, and the patient exhibited confusion and lack of coordination. The patient was conscious; however, he was disoriented by the place and time. The limb muscles of the patient exhibited weakness, and the patient's grip on the upper limb was poor. The patient exhibited the classical tetrad symptoms, i.e., agraphia (inability to write), acalculia (inability to perform arithmetic), finger agnosia (inability to distinguish fingers), and left-right confusion on examination. However, there were no tremors, rigidity, or abnormal movements. The other systemic examinations were normal. The MRI study revealed chronic ischemic changes in periventricular and subcortical white matter in the parietal lobes. Certain ischemic changes were also observed in the frontal lobe, suggesting age-related changes. MRI scans and the patient's other examinations suggest Gerstmann's syndrome.

2.3 Anatomical Basis of Gerstmann's syndrome¹⁴

The parietal lobe, situated in the cerebral cortex's upper back region, is harmed in cases of Gerstmann's syndrome.

The parietal lobe combines sensory data from diverse body areas and translates it into understandable perceptions and actions. The superior and inferior parietal lobules, which comprise the two primary parts of the parietal lobe, are each further split into various zones with specific functions. The four main signs of Gerstmann's syndrome, Agraphia, Acalculia, Finger Agnosia, and Left and Right Confusion, are believed to result from harm to particular parietal lobe areas.

2.3.1 Agraphia¹⁵

Damage to the angular gyrus, a region at the intersection of the parietal, temporal, and occipital lobes, is linked to agraphia. Reading, writing, and other language-related skills need the integration of visual, aural, and tactile information from many brain sections, largely accomplished by the angular gyrus.

2.3.2 Acalculia¹⁶

It is linked to injury to the intraparietal sulcus, a region along the lateral surface of the parietal lobe. Processing numerical data and carrying out mathematical computations occur in the intraparietal sulcus.

2.3.3 Finger agnosia¹⁷

Finger agnosia is due to injury to the inferior parietal lobule's supramarginal gyrus. The main function of the supramarginal gyrus is to distinguish individual fingers and carry out fine motor tasks. The supramarginal gyrus also processes tactile and proprioceptive information from the hands and fingers.

2.3.4 Damage to the right parietal lobe¹⁸

Which controls spatial orientation and the processing of visual-spatial information, is linked to left-right disorientation. The ability to discern between left and right can be compromised by damage to the right parietal lobe, impairing one's ability to move around in space and carry out tasks that call on spatial orientation.

3. RESULTS

3.1 Clinical Evaluation

3.1.1 Finger Agnosia

The patient had trouble naming and recognizing the digits on the hands or when he was shown pictures of his hands. When asked to point to specific fingers, he showed hesitation and struggled to identify them correctly.

3.1.2 Left-Right Confusion

The patient frequently had trouble telling the difference between left and right, making it difficult to follow instructions or perform tasks requiring spatial orientation.

3.1.3 Agraphia

The patient had difficulty writing and had weak handwriting abilities. He needed help correctly forming letters and spelling words.

3.1.4 Acalculia

A condition in which a patient has trouble doing simple addition, subtraction, multiplication, and division operations. He needed help understanding arithmetic ideas and sequencing of numbers.

3.2 Intellectual Functioning:

3.2.1 Attention and focus

During the assessment, the patient showed intact attention and focus. He could pay attention and concentrate on the subject for the necessary time.

3.2.2 Memory

The patient showed healthy short- and long-term memory, as seen by the ability to recall knowledge from the past and retain it during the evaluation.

3.2.3 Linguistic abilities

The patient's linguistic abilities seemed to be untouched by anything. He could communicate by fluently understanding and using the right language and vocabulary.

3.2.4 Solving problems

The patient's problem-solving skills showed some mild limitations. He needed help with activities requiring logical deduction and quantitative reasoning. Visualization or mental manipulation of items in space was particularly challenging for the patient when doing visuospatial activities.

3.3 Scholastic Functioning

3.3.1 Reading

The patient showed largely intact reading abilities, but there were significant comprehension and word recognition issues, particularly when reading unfamiliar or challenging literature.

3.3.2 Writing

The patient needed help organizing his thoughts clearly and displayed bad handwriting and spelling mistakes. The patient had major problems with mathematical problems. He needed help with simple mathematical procedures and understanding and using mathematical ideas.

3.3.3 Overall Academic Performance

The symptoms of Gerstmann's illness considerably hindered the patient's academic performance. They would profit from interventions and specialized educational support that catered to their particular needs.

4. DISCUSSION

There are incomplete variants of the Gerstmann syndrome that only have two or three of the four cardinal symptoms; therefore, it is not always present. According to Heimburger et al. (1964)¹⁹, a precise anatomical correlation is impossible because as the number of symptoms rises, so does the size of the lesion. Memory problems, anomia, alexia, constructional apraxia, and other parietal injury symptoms frequently manifest. Benton (1961) showed how frequently they were connected to the cardinal symptoms. Gerstmann syndrome was described as "an artefact of defective and biased observation" since the association of the four cardinal symptoms is the same as that of the condition's cardinal symptoms. Austrian neurologist Joseph Gerstmann described the case of a 53-year-old patient who presented with agraphia, acalculia, right-left disorientation, and difficulty moving specific fingers in 1924. These clinical findings were indicative of cerebrovascular syndrome with left hemisphere involvement²⁰. Three years later, the same author described two identical cases in that they both displayed constructional apraxia, anomia for colours, and trouble reading numbers but not words²¹. However, the syndrome didn't become well-known in the international academic community until 1940, when Gerstmann's study was published in English. In the same year, the GS tetrad agraphia, acalculia, finger agnosia, and right-left disorientation²². After several years, it was noted that the angular gyrus of the parietal lobe is mainly associated with the symptoms of agraphia and acalculia²³. Poeck and Orgass (1966)²⁴ concluded that a lesion in the left hemisphere can only be predicted if at least three of the four symptoms are present. Still, they added that this has little diagnostic value because aphasia also occurs in right, left, or bilateral brain-injured patients. On the other hand, Strub and

Geschwind (1974)²⁵ stressed that the association between the syndrome and aphasia is unnecessary. It is probably due to the contiguity of the cortical area subserving each. They did this by describing a patient with the Gerstmann syndrome but without language disorders. Thus, even in situations lacking aphasia, the whole Gerstmann syndrome would continue to have high localizing lesions. In his analysis of these studies on the Gerstmann syndrome, Benton (1977)²⁶ noted that the components of the syndrome were, "in reality, collective terms for diverse types of disability of a perceptual, praxis, linguistic, and conceptual nature," and that there was no evidence that the cardinal symptoms had a distinctive neuropsychology significance. In contrast to the patients of Strub and Geschwind, our situation is not open to the same kind of criticism. The aphasia was first present on the first day and had no history or clinical signs of worsening. A CT scan revealed an incredibly localized lesion, which the trauma and the subsequent surgery had caused. According to the brain mapping, the lesion correlates to the left angular gyrus after stabilization. Following stimulation of the posterior perisylvian area, more precisely in the transition between the angular and supramarginal gyri, Morris et al. described the case of a patient with temporary symptoms consistent with GS in 1984²⁷. Using the brain mapping approach, Roux et al. in 2003 discovered problems with writing, arithmetic, and finger recognition in the left angular gyrus of 6 right-handed individuals who had undergone tumour resection²⁸. In 2014, a case of an older patient with right-left confusion, dysgraphia, and dyscalculia (without finger agnosia) was reported due to ischemia injury to the left posterior insula and temporal-parietal operculum. The patient's angular and supramarginal gyri were unaffected²⁹. According to several recent articles, the frontal lobe and GS may be related. Heymi Lee et al. published two cases in 2013 in which brain MRI revealed impairment of the middle and inferior frontal gyri, as well as the basal ganglia, as a result of an ischemic stroke with hemorrhagic transformation in the first case, and involvement of the inferior frontal gyrus, the pars opercularis, and the triangularis as a result of a glioma's compression effect in the second case. Both patients'

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neurological examinations indicated the four typical GS symptoms.²⁹ The patient reported here also exhibited ischemic changes in the frontal lobe and in the subcortical areas in the angular gyrus. The classical tetrad signs of Gerstmann's syndrome were also observed on the overall examination of the patient.

5. CONCLUSION

Gerstmann's syndrome is generally believed to result from injury to certain parietal lobe areas essential for integrating sensory data and translating it into meaningful perceptions and actions. Clinicians can more easily pinpoint the disorder's underlying causes and create focused interventions to treat its symptoms if they know the syndrome's anatomical foundation. This comprehensive assessment of a 65-year-old patient with Gerstmann's syndrome revealed deficits in various domains, including finger agnosia, left-right confusion, agraphia, and acalculia. While their intellectual functioning showed relative preservation in attention, memory, and language skills, they exhibited difficulties in problem-solving and visuospatial tasks. Scholastically, the patient struggled with reading, writing, and mathematics. It is crucial to provide appropriate educational and therapeutic interventions to support their functional abilities and enhance their quality of life.

6. AUTHOR'S CONTRIBUTION STATEMENT

Mr. Bheemesh P collected the data for this paper, while Ms. Nitya Waghray conceptualised and designed the study. Dr. Archana R evaluated the work and provided the appropriate input regarding the necessary changes. Dr. W.M.S. Johnson made significant contributions to the manuscript's design. The final draft of the work was read and approved by every author.

7. CONFLICT OF INTEREST

Conflict of interest declared none.

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