

THE GERSTMANN SYNDROME - A REVIEW OF LITERATURE

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REZUMAT

Neurologul Gerstmann a conferit statusul de sindrom unei tetrade de simptome constând în acalculie, agrafie, agnozie digitală și confuzie stânga-dreapta. Scopul acestei lucrări este de a reconstitui o jumătate de secol de controverse privind aceste simptome pentru o mai bună înțelegere a implicațiilor lor.

Cuvinte cheie: sindrom Gerstmann, acalculie, agrafie, agnozie digitală, confuzie stânga-dreapta

ABSTRACT

The neurologist Gerstmann conferred the status of syndrome to a tetrad of symptoms consisting in acalculia, agraphia, finger agnosia and left-right confusion. The aim of this paper is to retrace the half century of controversies around these deficits for a better understanding of their implications.

Key Words: Gerstmann syndrome, acalculia, agraphia, finger agnosia, left-right confusion

HISTORY OF LESIONS AND SYMPTOMS

In 1924, J. Gerstmann described the case of a 52 year-old women complaining of memory and writing difficulties. Neurological examination showed a right hemianopia, calculation impairment, writing disability, right-left disorientation and loss of ability to recognize, identify, differentiate, name, select and orient the individual fingers of her hands. The latter deficit was called finger agnosia.¹ Gerstmann interpreted finger agnosia as the selective impairment of the body image “as though the optic-tactile-kinesthetic image pertaining to the fingers were split off from the total body image”.¹

In 1927, he presented another two cases with

acalculia, agraphia, left-right confusion and finger agnosia. These patients had no hemianopia, but presented additionally constructional apraxia and impaired colour perception.² Gerstmann called the agraphia and finger agnosia “Kardinalsymptome” (cardinal symptoms) and the other deficits “Randsymptome” (accompanying symptoms). In 1930, he described acalculia, agraphia, left-right confusion and finger agnosia as a new syndrome, called since then the Gerstmann’s syndrome.³

In 1957, Gerstmann writes that the left-right confusion occurs “with special reference to the hands and fingers”, the differentiation of fingers is necessary for writing and fingers have an important role in the first arithmetical calculations of children and in counting in primitive populations, concluding that the four symptoms have a common psychoneurological basis. He attributed the left-right confusion to an impairment of body image, even though errors were also present at another person’s body, arguing that for understanding another body, it was necessary to understand one’s own body. Gerstmann also concludes that the syndrome has a high localizing value for the transition between left angular gyrus to the second occipital convolution.⁴

In 1926, Hermann and Potzl stated that “finger apraxia” was responsible for Gerstmann’s syndrome.⁵

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Schilder, in 1931, doubting the unity of finger agnosia, distinguished five types of disturbances: finger agnosia (disturbance of the finger schema), finger aphasia (impairment in naming the fingers and in indicating the fingers named by examiner), visual finger agnosia (non-differentiation on one's own or another person's fingers presented visually), apraxia of finger choice (impairment in imitating finger movements), and constructive finger apraxia (disability to imitate finger positions given by the examiner with preserved movements to verbal instructions). He hypothesized a different cerebral localization for each of these symptoms.⁶

Other authors added to the Gerstmann syndrome one or more symptoms. Stengel in 1944, Hecaen and Ajuriaguerra in 1952 considered constructional apraxia as an integral part of Gerstmann syndrome.^{7,8} Stengel stated that "constructional apraxia and Gerstmann syndrome, when occurring in isolation, are incomplete or abortive appearances of the syndrome".⁷

Benton challenged the existence of Gerstmann syndrome in a study of a large sample of brain-damaged patients showing that acalculia, agraphia, left-right confusion and finger agnosia correlated as closely with each other as they correlated with other deficits, as constructional apraxia or reading impairments.⁹

Heimburger and colleagues, studying a large cohort of brain-damaged patients, found that the symptoms of Gerstmann syndrome had some localizing value related to the left side of the lesion and to the posterior parasyllian area, but the lesions were spreaded "widely over the parietal, temporal and occipital lobes". Also, the syndrome was accompanied by "numerous other neurological deficits, notably dysphasia" and all patients were aphasic.¹⁰ Moreover, Poeck and Orgass observed that the four cardinal symptoms are more frequent in aphasic patients than in non-aphasic ones. They argued that these symptoms are rather consequences of aphasia, due to their direct relationship with language.¹¹

The existence of Gerstmann's syndrome without language disturbances was documented in other studies. Kinsbourne and Warrington observed that the majority of their patients with Gerstmann syndrome were not aphasic and concluded that the association of the four symptoms is more than coincidental.¹²

Other authors described cases of Gerstmann syndrome without aphasia, but with other associated impairments such as: constructional apraxia, limb apraxia, visual or sensory right-sided deficits, intellectual deterioration.¹²⁻¹⁹

The first case of pure Gerstmann syndrome,

resulting from a well-described lesion of superior angular gyrus that extended into the supramarginal gyrus and minimally into the superior parietal lobule, documented by a CT scan was described by Roeltgen and colleagues.²⁰ After that, several cases of pure Gerstmann syndrome due to a lesion of angular gyrus were described.^{18,19,21}

Gerstmann's syndrome (associated or not with other symptoms) was described in many brain lesions: left parieto-temporo-occipital, left frontal posterior, left thalamic, right parietal in left handed patients, subcortical region underlying the left angular gyrus, diffuse subcortical HIV-1 induced encephalopathy, diffuse cerebral atrophy following systemic lupus erithematosus.^{10,16,17,22-28}

Stimulation studies in epileptic patients undergoing surgery showed that the Gerstmann syndrome can be induced by electrical stimulation of the transition zone between the angular gyrus and supramarginal gyrus, and the finger agnosia and acalculia sites were found in the supramarginal gyrus or close to the intraparietal sulcus.

THE FOUR SYMPTOMS

1. Acalculia

Acalculia is a heterogeneous deficit of Gerstmann's syndrome. Only a few cases have been fully investigated, but these studies showed that there are frequent syntactic difficulties in the comprehension and the production of Arabic digits.^{12,26} The impaired syntax is also illustrated by weaker performance when reading three-digit numbers compared to one and two digit numbers.¹⁸ The reading of number words is generally preserved, but the comprehension of the syntactic relation between number words is impaired.²⁶ The misalignment of digits in a number causes frequent errors in written calculation and written calculations are highly impaired especially when it requires carrying and borrowing strategies.^{12,13,19} The complex mental calculations are impaired, whether they are presented orally or visually.^{13,18,31}

The status of arithmetical facts is still unclear. Dehaene and Cohen predict that in Gerstmann's syndrome should be affected the processing of number magnitude and the complex operations (ie. subtraction) but the arithmetical fact retrieval (ie. small addition and multiplication) should be intact because the latter relies on language circuits.^{17,32} There are studies in agreement with these predictions, some patients with Gerstmann syndrome showing a relative preservation of small addition and multiplications.^{12,31,33} But there

are also cases of Gerstmann syndrome with loss of arithmetical facts.^{13,18,20,34}

Some of the patients were unable to compare numbers and to position them correctly on a straight line, but several others performed well the number comparison and the number positioning.^{18,26,31,33,35-38}

The existence of arithmetical difficulties despite preserved ability to recognize the arithmetical signs was described in some cases.^{21,38}

The enumeration of dots can be impaired, occasionally erroneous or slowed down.^{18,26,38} An inability to recite number series was described in one case whose numerical abilities were limited to range 1 - 4.³⁵ Incorrect recitation was reported with the alphabet, the days of the week and the months.^{26,35}

2. Agraphia

Interestingly, in Gerstmann syndrome, a peripheral dysgraphia is often present, but central agraphia (with phonological or lexical disturbance) is never observed.²⁵ There are two types of peripheral deficits that can be found together or separately.

In most patients, the writing is slow and illegible, with misaligned and scrawled letters.^{18,20,25,28} Cursive letters are more impaired than the printed ones and copying may be preserved.^{19,34} There may be difficulties in drawing geometrical shapes.³⁹ This pattern is reminiscent of apraxic agraphia, hypothesized to result from a loss or a failure to access the motor graphic patterns from memory.³⁹

Several cases present letter omissions, perseverations or substitutions, sometimes substitutions being restricted to the pairs p-b and q-d, tending to occur between similar letters.^{13,20,21,25,34} Ordering or selecting mobile letters to form a word or to type on a keyboard can be impaired.^{12,31,34} It is still uncertain if these errors are determined by the kinesthetic or by the visual factors implicated in the allographic level where letter identity is accessed for motor production.

The recognition of graphic movements can also be impaired, the patients being unable to recognize the letters presented in tactile modality (ex.: the examiner traces a letter in the patient's palm) and visual modality of movements (ex: the examiner traces a letter in the air).³⁴

Some patients failed to answer questions that require visual imagery of the letter form.^{25,31}

3. Finger agnosia

The patients with Gerstmann syndrome often fail to recognize the fingers. Some of them are unable to name or identify by the name the fingers.^{18,20,21,24,28,34}

Under visual control, some patients are capable to point or name their finger in response to verbal or tactile stimulation, but some commit more errors without visual control, are unable to raise the same finger as the examiner or to indicate the examiner's finger corresponding to their finger.^{13,19,25,31} Most evident errors are observed in the index, middle and ring fingers.^{19,21,25,40}

Some authors proposed that finger agnosia represents a mild form of autotopagnosia, but autotopagnosia and finger agnosia can appear dissociated and consequently would represent different defects.⁴⁰

In literature, cases with toe agnosia were reported, suggesting that the body schema disorder is not limited to the fingers.^{21,25}

4. Left-to-right confusion

The patients with Gerstmann syndrome make left-to-right confusions in orientation with respect to their own body, with visual control, but especially without visual control.^{12,18,19,25,34} The left-to-right confusion is generally more pronounced in orientation to the examiner's body, with a larger number of errors when the examiner faces the patient than when the patient and the examiner's body are oriented in the same direction.^{19,21,25,31,41}

The left-to-right confusion is exacerbated by two-stages commands (ex.: touch your left ear with your right hand).^{20,21,25,28} In some cases, the impairment is detectable only when the patient is asked to point to the left or right side of the examiner's body with his right or left own hand.¹³

Some patients had the ability to name or point to the right or left parts of the body intact, but their performance was always poor when mental rotation to a command was required.⁴¹

There have been reported cases with patients with impairment in deriving the relative position of objects along the horizontal axis.⁴¹ The data concerning the vertical axis (ie. up-down) are contradictory. There are evidences of cases with difficulties in recognizing the spatial relationships (ie. up-down, over-below) and cases with correct judgments on the vertical axes.^{7,21,40}

CONCLUSIONS

The pure Gerstmann syndrome is not very common. Since the first description of the syndrome, it has been suggested a common psychoneurological factor which could explain the symptomatology, but none of these hypotheses was entirely satisfactory.

For example, in 1944, Stangel included the Gerstmann syndrome into a larger syndrome including impairment of spatial orientation and constructional apraxia.⁷ This hypothesis cannot be sustained because several cases of pure Gerstmann syndrome were described in literature.^{18,20,21,25,26,31}

Gerstmann in 1957 stated that the common psychoneurological factor for the syndrome was a selective disorder of the body schema in one area of the body, the hand.⁴ The toe agnosia described in other cases with the four symptoms of Gerstmann syndrome excludes his hypothesis.^{21,25} It also excludes the assertion put forward by Critchley who suggested that the localized autotopagnosia could be due to the unique role of the hands in human activities.^{25,42}

Poock and Orgass claimed that each symptom of Gerstmann syndrome results from language impairment.¹¹ This idea can be rejected, in literature being described patients who performed correctly the language tests, with the exception of writing production.^{12,13,20,25}

The presence of a general intellectual deficit overwhelming the four symptoms, claimed by Heimburger et al can be excluded due to the existence of cases that scored well within the norms in intellectual assessment measured by Progressive Matrices and Wechsler Adult Intelligence Scale.^{10,25}

Levine and colleagues hypothesized that Gerstmann's syndrome results from a disconnection between verbal and visuo-spatial functions.³⁴ According to his theory, all tasks that require a spatial analysis in response to a linguistic stimulus should fail, but the analysis of other cases from literature does not confirm their assumptions. For example, some patients performed well in tasks where, on verbal stimulation they had to point or name their finger under visual control.^{13,19,25,31} Also, number production can be erroneous in both verbal and digital modalities and the fact that the performance in finger localization tests is worsened in the absence of visual control is not explained by this hypothesis.²⁵ Regarding agraphia, the substitutions between certain letters (eg. p-b, q-d) observed in some cases are not sufficiently explained by a defective integration between spatial and verbal skills.^{25,31} Consequently, functional verbo-visual disconnection cannot be implicated in these performances.²⁵

Gold et al suggested that the defective horizontal mapping could account for the symptoms of Gerstmann syndrome.⁴¹ The substitutions between certain letters (p-b, q-d) concerning exclusively a top-bottom letter reversal and not left-right shift do not

sustain this hypothesis.^{25,31}

Mayer and colleagues, in 1999 proposed that the four symptoms of Gerstmann syndrome are due to the impairment in the manipulation of mental images which require spatial visual processing in the form of translation, rotation or other transformations of visual mental images.²⁵ Mental rotation was tested explicitly and the results confirmed that it is impaired in Gerstmann syndrome.^{25,31} The fact that the finger agnosia was more pronounced when visual support was suppressed can be explained by the impairment in visual image transformation. Also, according to this hypothesis, the left-to-right discrimination was more impaired when the patients had to make a mental rotation.^{25,41} The substitutions limited to the letters p-b and q-d can be interpreted as incapacity to rotate the vertical stroke around the circle to form this letter.²⁵

Mayer and colleagues stated that since the numbers are learned in a fix order, they must be represented in a spatial configuration. Accordingly, an impairment of visual imagery could interfere with the spatial processing of numbers in semantic tasks. The preserved language abilities and impaired counting suggest that the latter is not entirely an automatic language sequence and it may be associated with a spatial representation as a numerical line.²⁵ The impaired counting has also been related with the impairment in spatial processing of the rank information.

Due to the positional syntax of the Arabic system, a visual imagery deficit could affect the comprehension, production of multidigit numbers and oral and written calculation. Also, a visual imagery deficit could explain the fact that perceptual estimation of quantities (which require correct inferences from visual imagery) was more impaired than the cognitive estimation of quantities.²⁵

At this time, future research is necessary to identify more precisely the cognitive deficit underlying each of the four symptoms of Gerstmann syndrome. The question if there is a common psychoneurological factor is complicated by the fact that each symptom refers to various difficulties. The hypothesis that the common denominator is the impaired visuospatial image transformation remains plausible until new cases of pure Gerstmann syndrome will be investigated for this disorder.

The four symptoms of Gerstmann syndrome can be found in absence of other impairments. Several case studies demonstrated that the presence of pure Gerstmann syndrome is highly predictive for a damage or isolation of the left angular gyrus, as Gerstmann suggested in 1957. It still remains in question if the left

angular gyrus is a part of a common neural network for calculation, writing, finger discrimination and left-to-right orientation or it corresponds to the overlapping region of distinct neural networks for each of these functions.

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