

SHORT REPORT

Toe agnosia in Gerstmann syndrome

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Abstract

The following case report presents a patient exhibiting Gerstmann syndrome accompanied by toe agnosia. A 72 year old right handed woman had a focal lesion in the angular gyrus of the left hemisphere which was caused by a glioblastoma multiforme. The first symptom she had complained of was severe headache. Standardised neuropsychological tests of intelligence, memory, attention, fluency, apraxia, and language functions as well as tests for the assessment of agraphia, acalculia, right-left disorientation, and digit agnosia were performed. The patient displayed all four symptoms of the Gerstmann syndrome—namely, agraphia, acalculia, right-left disorientation, and finger agnosia. The patient did not display aphasia, constructional apraxia, or any other neuropsychological impairment. In addition to the four symptoms of the Gerstmann syndrome an agnosia of the toes was found. Further studies should determine whether finger agnosia in Gerstmann syndrome is usually accompanied by toe agnosia. Finger agnosia in the context of this syndrome may be better named digit agnosia.

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In the 1930s Josef Gerstmann described a syndrome consisting of four symptoms: finger agnosia, right-left disorientation, agraphia and acalculia, which is attributed to dominant hemisphere lesions affecting the angular gyrus.^{1–3} Subsequently numerous case reports based upon this description were published.^{2–9} The facts that not all observed cases with the Gerstmann syndrome displayed all four components of the quadrima, that the patients showed other clinical manifestations—for example, constructive apraxia or aphasic disturbances—that the number of symptoms seemed to depend on the size of the lesion and that some patients with lesions in the left posterior parasyllian area were free of any of the Gerstmann symptoms raised doubts as to the existence of a distinct syndrome.^{5 9–13} The

debate concerning the Gerstmann syndrome has been described elsewhere.^{10 14–16} We describe a patient who had a focal lesion in the angular gyrus of the left hemisphere which was caused by a glioblastoma multiforme. The patient exhibited Gerstmann syndrome accompanied by toe agnosia.

Case report

A 72 year old right handed woman who had had no history of neurological or psychiatric diseases was admitted to the Department of Neurosurgery for treatment of a glioblastoma multiforme in the left parietal lobe. The first symptom she had complained of was severe headache. The patient had worked as an accountant until the age of 64.

A cranial CT scan disclosed a tumour situated in the areas P01, P02, and P05 according to the classification of Damasio and Damasio and which included the angular gyrus, supramarginal gyrus and extended to the inferior parietal lobule of the left hemisphere (figure).¹⁷

On neurological examination no deficits were found. On neuropsychological examination, the patient was alert, cooperative and well oriented. Her intellectual functions were average (four subtests of the Wechsler adult intelligence scale¹⁸). She showed no deficits in biographical memory, everyday memory, short, medium and long-term memory or in working memory for verbal material (subtests of the Wechsler memory scale¹⁹). Her performance in a recognition task (recurring figures²⁰) as well as her short term memory and working memory for non-verbal material were also normal (subtests of the Wechsler memory scale¹⁹). She had no difficulty in copying the complex figure of Rey^{21 22} or in drawing a cube and clock faces. Her motor speed and performance in a complex conceptual tracking task were normal (trail making test²³), and she showed no buccofacial apraxia or ideomotor apraxia of the limbs and body.²⁴ In a detailed examination of language functions (Aachen aphasia test²⁵), including spontaneous speech, token test, repetition of syllables, words and sentences, reading and writing words, spelling words and sentences with cards on which letters and words were printed, naming objects, colours and situations as well as tests of comprehension, no deficits were found. Only her performance in writing sentences was impaired: spelling mis-

takes, especially omissions of single letters, were found. Her performance in the other writing tasks was good; her overall performance in writing was at a normal level. Her performance in a verbal (S-words) and figural fluency (5-point-test²⁶) task was normal, although she showed a raised rate of repetitions in the figural fluency task. In a computerised examination of handwriting movements which was performed with a digitising tablet (WACOM IV),^{27, 28} no impairment was revealed. Her reaction time in a computerised reaction task (TAP-alertness²⁹) was reduced. On further neuropsychological examination she displayed all four symptoms of the Gerstmann syndrome. All of these symptoms could be seen throughout the entire duration of her stay in hospital.

AGRAPHIA

The patient was asked to write down dictated letters and words, to name words which were spelt by the examiner either orally or with lettered cards, to spell and read words, to write a short report of her relatives' last visit, to recite the alphabet and to search for spelling mistakes in a short story.

While the patient displayed no deficits in reading or spelling, in correcting spelling mistakes, in reciting the alphabet, in writing dictated letters, or in naming words which were spelt by the examiner, frequent omissions, additions and substitutions could be seen when she was asked to write down dictated words, to copy words and to write a report. These findings were supported by the results of the examination of language functions.

ACALCULIA

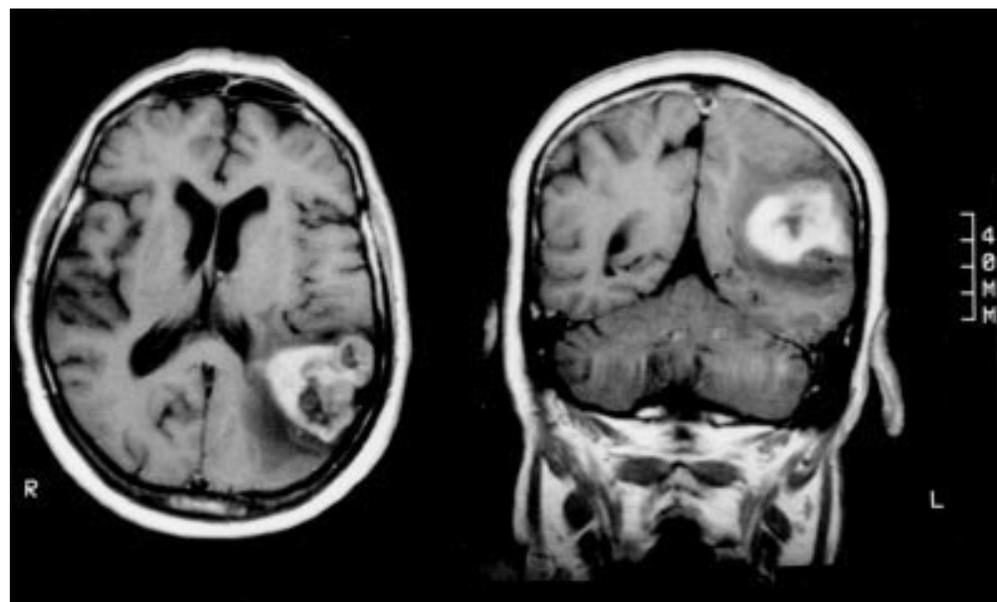
The patient was asked to count to 25, to point to numbers printed on cards (the number being determined by the examiner), to copy and read numbers, to repeat numbers which were called by the examiner, to discriminate and grade numbers, to write down dictated

numbers, to insert arithmetic operators (for example, +, −, ×, ÷) into simple calculations and to do simple mental arithmetic as well as written calculations. The patient was requested to add, subtract, multiply, and divide.

The patient had no difficulty in counting, in pointing to numbers called by the examiner or in discriminating, grading, and copying numbers. However, deficits in writing dictated numbers, in reading, and in repeating numbers were found. Furthermore, she displayed severe impairments in calculating. In writing dictated numbers and in repeating numbers she had difficulty with numbers which contained a zero and which had more than three digits—for example, when the examiner said 3011 she repeated 3111. In reading numbers she made some mistakes concerning the first digit of the numbers—for example, she said 148 instead of 248. In reading as well as in repeating numbers she made the mistake several times of splitting the numbers into thousands, hundreds, and digits, eg. she said 9000-0-9 instead of 9009. In arithmetic she made mistakes in more than 50% of the problems, even in simple calculations. The deficits affected adding, subtracting, multiplying and dividing equally. In inserting arithmetic operators into calculations her performance was much better than in the other arithmetical tasks.

RIGHT-LEFT DISORIENTATION

In order to investigate a possible right-left disorientation, tests were chosen considering the reflections of Benton and Sivan.³⁰ The patient was asked to name the parts of her body which were touched by the examiner, to state which side of her body was being touched, and to point to parts of her body as well as their side (for example, left leg). These requests were based on one-stage commands. Another set of two-stage commands was also used consisting of crossed (for example, "touch your right ear with your left hand") and uncrossed com-



MRI T1W in transverse and coronal view showing a multigyrated contrast enhancing tumour mass with peritumoral oedema. Slight compression of ipsilateral lateral ventricle.

mands (for example, “touch your left knee with your left hand”) which were given first by the examiner and subsequently by the patient herself. All of these tasks were performed with eyes open and closed. Furthermore, she was asked to perform most of these tasks on a model who sat opposite or next to her.

On one-stage commands the patient displayed no difficulty in distinguishing left from right referring to her own body. The same requests referring to a model’s body caused problems regardless of the model’s spatial relationship to the patient. These difficulties were not therefore related to problems with mental spatial rotation. In all tasks comprised of two-stage commands the patient demonstrated a right-left disorientation. She was, however, able to manage tasks that were based on commands with similar complexity without including right and left. No impairment of up-down orientation (for example, orientation corresponding to the vertical body axis) was revealed.

FINGER AGNOSIA

The patient was asked to grade strips of paper of differing size, to name parts of her own and of the examiner’s body, to name or touch fingers of her own, of the examiner and of a cardboard model as indicated by the examiner, to move her own fingers on request by the examiner, to touch the same fingers of the examiner’s hand that had just been touched on the model’s hand, to touch, move, or name fingers of the hand that had just been touched by the examiner without the aid of vision, and with closed eyes to touch or move the fingers of one hand which corresponded to the fingers of the other hand which had just been touched by the examiner. Furthermore, the finger strip test, the two-point finger test, the in-between test, and the matchbox test as described by Kinsbourne and Warrington were employed.¹³ In the finger strip test the patient is requested to arrange strips of paper on which the names of the fingers are printed in the correct sequence. In the two-point finger test the fingers of the patient are touched in two places. The patient judges whether he is being touched on one or on two fingers. In the in-between test two of the patient’s fingers are touched simultaneously by the examiner. The patient is asked to state the numbers of fingers between the ones touched (possible answers: 0, 1, 2 or 3). In the matchbox test the patient’s hand is touched by the examiner in two places, either by one matchbox which is slipped between two fingers, or by two matchboxes each touching only one finger. The patient is asked to judge whether he is being touched by one or two matchboxes. The last three tests were performed with the patient’s eyes closed. All tests were performed on both the left and the right hand.

In addition, the naming and touching of toes as instructed by the examiner was examined. The toes between the big toe and the little toe were named the second, third and fourth toe. In the examination only the big toe, the third toe and the little toe were touched. The inves-

tigation comprised touching and naming toes on request by the examiner with and without sight and touching with closed eyes the toe of one foot which corresponded to the toe of the other foot that the examiner had just touched. It was not possible to perform the tests described by Kinsbourne and Warrington on the toes.¹³

The patient had no problems in naming parts of the body and in grading strips of papers which differed in size or which were labelled with the names of the fingers. However, her performance in naming and touching her own fingers, the fingers of the examiner or the model as well as her performance in the two-point finger test, in the in-between test and in the matchbox test was impaired. While she made only few mistakes in tasks in which the requested fingers were shown on a model or named by the examiner she displayed severe impairments in the tests without sight, especially when asked to touch, move, or name the fingers of one hand which corresponded to those of the other hand that had just been touched by the examiner. When the finger had been touched on the same hand, less severe impairments could be observed. This result was due to a strategy of compensation in which the patient held with her thumb the finger which had been touched by the examiner until she was asked to touch, move or name this finger. To avoid this possibility of compensation the patient was asked to shake hands with a second examiner after the examiner had touched her finger. She then worked out a similar strategy: Instead of shaking hands she quickly grabbed the finger of the examiner which corresponded to the finger of her own hand that had just been touched and made a gesture like shaking hands.

The deficits affected each finger but more often the index finger, the middle finger and the ring finger. Most frequently she mistook the ring finger for the index finger and vice versa. When she was touched three or more times in sequence on the same finger she always gave different responses. While she made only few mistakes in the matchbox test and in the two-point test, every second trial of the in-between test was wrong. The wrong responses never exceeded one digit—for example, when one finger was between the touched fingers she answered either “no finger”, “one finger” or “two fingers”.

Whereas her performance in naming touched toes and touching toes which had been named by the examiner was flawless with the aid of vision, she was correct in only 30% of trials with closed eyes. In the task in which she was touched on one toe and was asked to touch the same toe of the other foot she erred in every second trial. This was also the case when responses regarding the big and the little toe were recorded. These deficits affected the left and right feet equally. They could not have been due to loss of or altered sensation since sensation of the feet had been normal on neurological examination. Sense of touch, sense of pain, position sense and perception of passive and active movement in the toes were

undisturbed. Vibration sense was slightly reduced at the toes in an age-associated fashion.

To exclude age-related impairment, six female subjects aged 67 to 75 years without neurological or psychiatric diseases were assessed on the tests relevant to the diagnosis of Gerstmann syndrome. These subjects were not impaired in any of the tasks employed; in arithmetic they made a few mistakes.

Comment

The patient described above displayed all four symptoms of the Gerstmann syndrome—namely, agraphia, acalculia, right-left disorientation and finger agnosia. The nature of the disturbances corresponds with the findings reported by other authors.^{2,5} The patient displayed neither aphasia nor constructional apraxia. Our findings therefore confirm the conclusions of a series of authors who suggested that no causal relation existed between aphasia and the Gerstmann syndrome.^{2,13,30,31} This relation was assumed to be the necessary underlying factor in the Gerstmann syndrome by Heimburger and colleagues as well as Poeck and Orgass.^{11,12} Our results also contradict the assumption made by Benson and Geschwind, Sobota and Kinsbourne, and Warrington that constructional apraxia is an additional component of the Gerstmann syndrome.^{5,9,13} We agree with Roeltgen *et al* who stated that Gerstmann's tetrad may exist without any other neurological or neuropsychological symptoms.²

In addition to the four symptoms of the Gerstmann syndrome we found an agnosia of the toes related to finger agnosia. The examination of the toes of patients with Gerstmann syndrome has until now seldom been considered. Only Benson and Denckla mentioned briefly an examination of the toes in which they found no toe agnosia.⁴ Although Critchley remarked that some neurologists include the toes in their examination he gave no further information about the authors or their findings.¹⁰ Kinsbourne and Warrington stated in their study of finger agnosia that their patients displayed no evidence of autotopagnosia and that they had no problems in naming parts of the body other than the fingers.¹³ They did, however, emphasise that the toes were not examined. The neglect of the testing of toes in the cortical stimulation study of Morris *et al* is unfortunate.⁷ The authors were able to elicit in a 17 year old epileptic patient the four symptoms of the Gerstmann syndrome on stimulation of certain loci in the perisylvian area. Furthermore, they could transiently produce alexia, anomia, constructional apraxia, conversation difficulties, and spelling errors in addition to the four symptoms of the Gerstmann syndrome.

As we found in our patient a larger error in the identification of both toes and fingers when opposite sides of the body were involved, the apparent digit agnosia might at least partly be influenced by right-left disorientation. Gerstmann placed the lesion underlying finger agnosia at the parieto-occipital junction

around the angular gyrus of the left hemisphere.³² More recent case reports support this view. Roeltgen *et al* and Varney *et al* have described non-aphasic patients with focal lesions in the posterior parietal region of the left hemisphere who exhibited bilateral finger agnosia and the other elements of the Gerstmann tetrad.^{2,3} The study by Morris *et al* showed that electrical stimulation of one locus in the transition zone between the angular and supramarginal gyri produced finger agnosia, agraphia, and acalculia.⁷ Benton, taking account of the fact that Morris *et al* could elicit distinctive combinations of deficits by stimulation of very small areas of the posterior parasyllvian cortex, postulated that the Gerstmann syndrome is only one possible syndrome of focal posterior parietal disease.^{7,14} Whether the four symptoms of the Gerstmann syndrome in combination with toe agnosia form a new syndrome as proposed by Benton or whether finger agnosia is always accompanied by toe agnosia is not clear.¹⁴ The related character of finger agnosia and toe agnosia in our patient points to the latter proposition. If future studies demonstrate that toe agnosia is a constant symptom of the Gerstmann syndrome, finger agnosia in the context of this syndrome would be better named digit agnosia. Toe agnosia should otherwise be regarded as a symptom which, in combination with the Gerstmann's tetrad, constitutes a new syndrome.

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