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Implications of Gerstmann's syndrome

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Four behavioural deficits—finger agnosia, right-left disorientation, dysgraphia, and dyscalculia—have come to be known as Gerstmann's syndrome. Generally, clinicians consider that the concurrence of these deficits implies a lesion in the angular gyrus of the language-dominant hemisphere. Gerstmann (1957) has reiterated his earlier contention that the syndrome is an entity with specific localizing significance but Benton (1961) has reported that the components of the syndrome have no stronger associative bonds than a variety of concurrent intellectual deficits. He concludes that the syndrome is an artifact of observer bias and unsystematic, incomplete examination for associated deficits.

In this study, we review some of the neurological deficits in patients with Gerstmann's syndrome and relate these deficits to the location and type of brain lesion.

METHODS

PATIENT POPULATION At Indiana University Medical Centre a large percentage of neurological and neurosurgical patients suspected of having brain damage is given an extensive battery of neuropsychological tests. Since these tests require active cooperation by the patient, severely psychotic, stuporous, or recalcitrant patients are not tested. Otherwise, the patients were unselected except that they were clinically suspected of having brain damage of some type. From 456 consecutively tested patients, 111 had one or more components of Gerstmann's syndrome.

METHOD FOR ELICITING COMPONENTS OF GERSTMANN'S SYNDROME As an integral part of the neuropsychological test battery, the Halstead-Wepman aphasia screening test (1949), as modified by Reitan, was given to each patient. The test contains 27 auditory and visual stimuli, presented in identical order and with identical instructions to each patient.

Examination for finger agnosia First a system for designating the fingers was agreed upon by the patient and the examiner. Usually the patients reported by number but sometimes other designations were used. After the patient's vision was obstructed by a blindfold or shield, his outstretched fingers were touched lightly by the examiner. The patient was instructed to identify the finger as soon as it was touched. Occasionally, a patient needed practice with his eyes open to develop sufficient skill to report reliably during the test. This precaution eliminated errors due to misunderstanding or lack of alertness. No patient was included whose tactile sensitivity was sufficiently impaired to preclude perception of the tactile stimulus. All mistakes of identification were recorded for each patient. The patient was considered to have finger agnosia if he made incorrect responses in a minimum of 20% of the trials.

Examination for right-left disorientation (a) The patient was shown a card with the printed instruction, 'Place left hand to right ear'. He was requested to read the command aloud. If his reading was incorrect, the examiner communicated the instructions to the patient verbally. Then the patient was requested to execute the instruction. (b) The patient was then requested to place his left hand on his left elbow. If he became confused in executing part (a) or failed to recognize the impossibility of part (b), further commands of a similar nature were given to verify the presence of right-left disorientation. The results of previous testing of the patient's ability to comprehend visual and auditory instructions provided a basis for distinguishing specific errors of right-left disorientation from global receptive dysphasia.

Examination for dysgraphia (a) The patient was shown a drawing of a clock and requested to write the name without first saying it. (b) He was shown the printed letters, SQUARE, and asked to transform the stimulus material to script. (c) After having read the word, SEVEN, and repeated it following oral presentation by the examiner, he was asked to write the word. (d) The patient was instructed to repeat the sentence, 'He shouted the warning', explain its meaning, and write it. In additional instances the patient was asked to write the names of common geometric figures after having copied their shapes. Provided that the patient's previous educational achievement indicated that he should be able to perform correctly, failure to effect proper formation of successive letters was considered to indicate dysgraphia. Simple spelling errors were excluded. The Figure shows examples of dysgraphia. It is apparent that these patients had their principal difficulty with the sentence, 'He shouted the warning' but the second instance in the Figure illustrates the occasional occurrence of extreme difficulty with individual words.

Examination for dyscalculia (a) The patient was required to copy '85 − 27 = ' from a card and compute it.

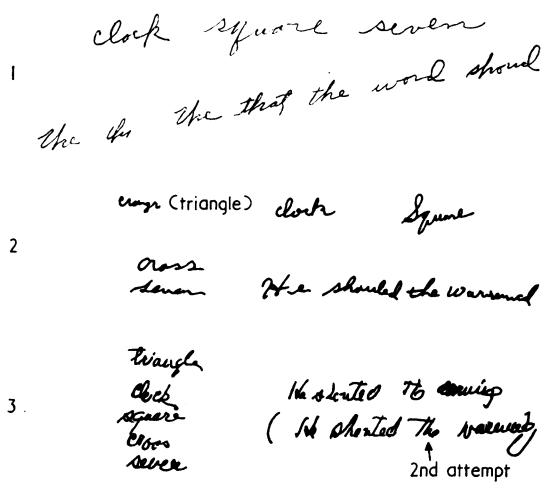

FIG. 1. *1 Example by 45-year-old white male with 10 years' education; 2 of 42-year-old white male with seven years' education; 3 of 71-year-old white female with 14 years' education.*

(b) He was required to multiply 17 by 3 in his head without seeing the figures written down. If these problems were not correctly solved, similar problems were presented for verification of dyscalculia.

METHOD FOR LOCALIZING BRAIN LESIONS Each of the patients with one or more components of Gerstmann's syndrome was evaluated by review of the medical history, physical and neurological examination, and, when available, skull radiographs, neuroradiological contrast studies, electroencephalograms, surgical notes, and necropsy findings. From the neurological information, a drawing was made on brain diagrams of the presumed site and extent of brain damage, and the type of lesion or diagnosis was recorded. The patients were then classified according to aetiology, lateralization of lesion, and intrahemispheric location of the lesion. In preparing the brain lesion charts, the results of the neuropsychological test battery were ignored, but all other clinical and anatomical information was utilized. At the time of the original case review, the referee did not know the Gerstmann classification of the patient as derived from the neuropsychological battery.

Five years after the initial study was completed, the records of the 111 patients with one or more Gerstmann components were again reviewed to check the course of patients whose diagnosis or localization of lesion had been obscure initially. This precaution minimized errors in diagnosis or localization of the lesion.

RESULTS

Of 456 consecutive neuropsychologically tested patients, 111 had one or more components of Gerstmann's syndrome (Table I). Among the 111 patients, 33 had only one of the components of the syndrome, 32 had two components, 23 had three components, and 23 had all four components of the syndrome. We will refer to these groups as group I, II, III, or IV, depending on the number of Gerstmann components.

The one common characteristic of the 111 patients is that all had organic (non-emotionally determined) neurological disorders. The evidence for the organic neurological diagnosis in each patient was derived from the neurological work-up, independently of the number of Gerstmann components. According to the type of lesion or diagnosis, the patients were classified into five categories as follows (Table I): 1 Cerebrovascular disease, including cerebral infarction, intracerebral haematomas, and generalized arteriosclerotic brain disease; 2 brain tumours, including gliomas, meningiomas, metastatic neoplasms, and abscesses; 3 trauma, including open and closed head injuries; 4 diffuse or symmetrical multifocal cerebral disease, including encephalitis, multiple sclerosis, pre-senile degenerative diseases, carbon monoxide or drug intoxication, and syphilis; 5 convulsions, including only patients considered to have idiopathic epilepsy. (Only three patients were diagnosed as having idiopathic epilepsy, and they exhibited only one or two components of Gerstmann's syndrome.)

TABLE I

NUMBER, AGE, AND TYPES OF LESION FOR PATIENTS WITH ONE THROUGH FOUR COMPONENTS OF GERSTMANN'S SYNDROME

Group	*Number of Patients in Group*	*Mean Age (yr.)*	*Age Range (yr.)*	*% of Group over 40 Years Old*	*Lesion Type (% of Group)*				
					Vascular	*Tumour*	*Trauma*	*Multifocal or Diffuse*	*Idiopathic Epilepsy*
I	33	42	21-68	55	21	18	33	21	6
II	32	40	11-71	41	25	22	22	28	3
III	23	45	17-68	61	30	35	13	22	0
IV	23	50	16-77	74	48	43	4	4	0
P< I vs IV	—	—	—	—	0·10	0·10	0·025	0·20	0·70

On comparing groups I-IV, we found a shift in the character of the lesions (Table I). In group I, the lesions tended to be diffuse, relatively small, and static. In group IV, the lesions tended to be large, highly destructive of tissue, and progressive or recurrent. The combined percentage of patients with either a brain tumour or cerebrovascular disease in the groups illustrates this feature clearly. In group I, 39% of the patients had either a brain tumour or cerebrovascular disease, while in group IV the combined incidence of tumour or vascular disease was 91%. Concomittantly, the combined percentage of diffuse or symmetrical multifocal disease or trauma decreased from 54% in group I to 8% in group IV. While analyses of the individual columns did not show significant differences in groups I and IV except for the trauma category, a χ^2 analysis of all types of lesion considered simultaneously indicated that the distribution of entries differed significantly from chance ($P<0.05$).

The greater severity of the lesions in group IV and the greater severity of the associated neurological deficits is reflected in numerous trends, as shown in Table II. The trends are consistent and mutually confirmatory. The P values given below each column are based upon χ^2 comparisons of groups I and IV, computed with Yates' correction for continuity. The significance of differential frequencies in each column was also evaluated with the four groups considered simultaneously and with groups I and II versus groups III and IV. The probability values for these latter comparisons are not reported because they were generally similar to those obtained in the comparisons of groups I and IV.

Some of the more striking neurological abnormalities in comparing groups I and IV are as follows (Table II): The incidence of hemiparesis (combined right or left) judged to be the result of lesions above the mid-brain is 48% in group I and 92% in group IV; right or left-sided delta focus in the E.E.G., 16% versus 60%; E.E.G. focalization in the posterior part of the left cerebral hemisphere, 20% versus 73%; known dead at five years, 21% versus 48%; and the incidence of dysphasia other than Gerstmann components, 39% versus 100%.

Every one of the 111 patients had evidence of organic neurological disease in addition to the Gerstmann components. If the patient had three or four Gerstmann components, the additional deficits tended to be grave and disabling. In accordance with Benton's observation, our results indicate that the syndrome is only one of a number of concurrent deficits rather than an autonomous entity. In fact, review of the cases previously reported in the literature, including the patients originally studied by Gerstmann himself (1924, 1927), invariably discloses evidence of considerable neurological impairment, such as difficulty with recent memory, emotional lability, and dysphasia, whenever the patients have been thoroughly studied. Thus the syndrome does not appear as an isolated cluster of deficits against an otherwise normal neurological status. The associated deficits are an integral part of the total picture of neurological disability.

Does the concurrence of the Gerstmann components have localizing significance? The question of the site of the lesion can be considered in two stages, the first of which is hemispheric lateralization, and the second of which is intrahemispheric location. The most objective direct evidence for localization of brain lesions is necropsy examination or, in lieu of necropsy, surgical operation and neuroradiological contrast procedures. Since all of these procedures were performed significantly more often in groups III and IV than in I and II, localizing conclusions based on these procedures carry a bias toward greater

TABLE II

FREQUENCY OF ASSOCIATED NEUROLOGICAL DEFICITS IN PATIENTS WITH ONE THROUGH FOUR COMPONENTS OF GERSTMANN'S SYNDROME

Group	*No. of Patients in Group*	*Hemiparesis (%)*		*Increased Intracranial Pressure (%)*	*Dysphasia (%)*	*% Known Dead in Five Years*	*% Necropsied*	*E.E.G.*				
								No.	*% Normal*	*Delta or Dysrhythmia on Left (%)*	*Predominant Delta Activity (%)*	
		R	*L*								*Right*	*Left*
I	33	18	30	21	39	21	12	25	24	20	12	4
II	32	31	19	28	75	16	6	24	13	42	13	33
III	23	52	0	26	96	35	22	17	12	59	0	47
IV	23	83	9	43	100	48	35	15	7	73	0	60
P> I vs IV		0·001	0·20	0·20	0·001	0·10	0·10		0·50	0·005	0·50	0·001

reliability in groups III and IV than the localizing conclusions we can apply to groups I and II. Apart from the neuropsychological testing, the only other localizing procedures applied with equivalent frequency to the four groups were the neurological examination and the E.E.G. To be as objective as possible one should select from these procedures the signs which have the least dependence on the patient's cooperation and level of consciousness and would be detected and recorded with the greatest reliability by the clinician. In the neurological examination, hemiparesis is one of the most reliable lateralizing signs of hemispheric damage. In the E.E.G. perhaps the most reliable hemispheric lateralizing sign is a delta focus.

Using hemiparesis or a delta focus as lateralizing criteria, one finds that the probability of a left hemisphere lesion increases as the number of Gerstmann components increases (Table II). When group I is compared with group IV, the incidence of right hemiparesis increases from 18% to 83%, while the incidence of left hemiparesis decreases from 30% to 9%. A predominantly left-sided delta focus was found in only 4% (one of 25) of the patients in group I, while in group IV a predominantly left-sided delta focus was found in 60% (nine of 15) of the records. When one adds to the delta foci other localizing E.E.G. signs, such as focal dysrhythmia or significant amplitude asymmetries, the incidence of left-sided localization by E.E.G. is increased to 20% in group I and 73% in group IV.

When the brain lesion charts, drawn from a synthesis of all available data except the number of Gerstmann components, are examined for lateralization, the overwhelming involvement of the left hemisphere is again apparent (Table III); 78% of the patients in group IV had an area of tissue damage strongly lateralized to the left hemisphere, 9% had the area of tissue damage strongly lateralized to the right hemisphere, and 13% had destructive lesions of both hemispheres.

Although two or three Gerstmann components imply left hemispheric damage, does the concurrence of all four Gerstmann components compel the conclusion that the lesion involves the left posterior parasylvian area? Statements about the intrahemispheric location of the lesion have to be made with less certainty than about hemispheric lateralization. In the tabulation of the data for the last three columns in Table III, the left posterior parasylvian area was listed as affected only if we had strong evidence for the site of the lesion. In patients with diffuse or symmetrical multifocal disorders such as multiple sclerosis, we could not make completely reliable inferences as to the site of the lesions. All diffuse disorders and any others without several lines of evidence converging had to be classified as uncertain. When the lesion was classified as not affecting the left posterior parasylvian area, we again required strong negative evidence. If we had reservations about the left posterior parasylvian area being involved, we classified the case as uncertain. Because of the necessity for conservatism in judgment, the estimated incidence for definite involvement or definite non-involvement of the left posterior parasylvian area probably underestimates both frequencies, particularly in group I. As was the case with lateralizing the lesion, the number of Gerstmann components was not considered in assessing the intrahemispheric location.

In group I, we considered the left posterior parasylvian area definitely to be involved in 6%, not involved in 63%, and uncertain whether involved in 30%. In group IV, we considered the left posterior parasylvian area to be definitely involved in 57%, not involved in 13%, and uncertain whether involved in 30%. In the three patients of group IV known definitely not to have a left posterior parasylvian area lesion, we can base the statement on necropsy examination of the brain. One of these patients had a clivus meningioma which elevated the posterior part of the third ventricle and compressed the midbrain

TABLE III

SITE OF MAXIMAL TISSUE DESTRUCTION AND PRESENCE OR ABSENCE OF DAMAGE IN LEFT POSTERIOR PARASYLVIAN AREA IN GROUPS WITH ONE THROUGH FOUR COMPONENTS OF GERSTMANN'S SYNDROME

Group	*No. of Patients in Group*	*Hemisphere with Maximal Tissue Destruction (%)*		*Multifocal or Diffuse (%)*	*No Independent Evidence of Structural Lesion (%)*	*Left Posterior Parasylvian Region Lesion (%)*		
		Left	*Right*			+	−	*Doubtful*
I	33	30	27	30	12	6	63	30
II	32	41	16	34	9	25	31	44
III	23	65	0	35	0	39	17	43
IV	23	78	9	13	0	57	13	30
I vs IV: P<		0·001	0·20	0·25	0·25	0·001	0·001	1·00

and aqueduct, causing moderate internal hydrocephalus. The second patient had a glioblastoma confined to the left frontal lobe. The third patient, a right-handed man, had a large glioblastoma of the right hemisphere infiltrating the posterior two-thirds. The brain was shifted from right to left, perforce indenting the medial aspect of the left hemisphere. In spite of the large tumour, the patient had been symptomatic for only one month, and he was alert and able to complete the entire battery of neuropsychological tests. Although the neoplasm was in the right hemisphere, he had a large delta focus on the left side in the E.E.G. He died postoperatively, five days after testing.

Although the left angular gyrus was affected in the majority of the 23 patients with four Gerstmann's components, the lesion was not restricted to that region in any patient. The tumours, infarcts, or haematomas were huge and greatly destructive of tissue, except in the case of the one patient previously mentioned with a clivus meningioma. Of the two patients in group IV who did not have vascular disease or a brain tumour, one had severe post-encephalitic brain damage, and the other had a bullet tract which began at the left occipital lobe and emerged from the left frontal pole, reaming out the core of the hemisphere. Because of the large size of the lesions in the patients of group IV, it seemed of interest to review the brain lesion charts with respect to involvement of structures adjacent to the angular gyrus. When this tabulation was done, the supramarginal gyrus, the posterior part of the inferior parietal lobule, the posterior part of the superior temporal gyrus, and the anterior parts of the occipital lobe all were involved with frequencies which approximate the estimated involvement of the angular gyrus.

In our original group of 456 patients, 345 had none of the components of Gerstmann's syndrome. Of the 345 patients, seven had focal destructive lesions judged to be confined to the left post-rolandic region, either in the parietal lobe or in the posterior parasylvian area. Of these seven patients, five were right handed, one was left handed, and the handedness of the remaining patient was in doubt. In three of the seven, the angular gyrus was definitely involved by the lesion. Two of the three were right handed and the third patient was the one whose handedness was in doubt. Certainly our material, together with other cases in the literature, conclusively shows that a destructive lesion of the left angular gyrus area is not a necessary condition for Gerstmann's syndrome. Because of the large size of the lesions in groups III and IV we would also suspect that a lesion completely restricted to the left angular gyrus might not be a sufficient condition to cause the syndrome.

Considerable interest has been shown in the past in the frequency of the four components of Gerstmann's syndrome. Table IV shows the frequency of the components in the groups of patients. In terms of total frequencies, finger agnosia was lowest and dyscalculia the greatest. Finger agnosia was consistently the least frequently occurring component in groups I, II, and III. Dyscalculia, however, occurred most frequently of the four components only in group I, suggesting that it tends to be dissociated most readily from the other components. When two components are present, the frequency of the six possible combinations of components is as follows: One patient had finger agnosia and right-left disorientation; two patients had finger agnosia and dyscalculia; four patients had finger agnosia and dysgraphia; seven patients had dysgraphia and right-left disorientation; eight patients had dyscalculia and right-left disorientation; and 10 patients had dyscalculia and dysgraphia. Noteworthy is the fact that the combination of finger agnosia and right-left disorientation was found in only one of the 32 patients with two components of Gerstmann's syndrome, which would not support Gerstmann's (1957) assertion that the two are 'characteristically associated'.

The four possible combinations of three components occurred as follows: One patient had finger agnosia, dyscalculia, and right-left disorientation; three patients had finger agnosia, dysgraphia, and

TABLE IV

NUMBER AND PERCENTAGE OF SUBJECTS MANIFESTING THE COMPONENTS OF GERSTMANN'S SYNDROME IN EACH GROUP

Group	*Number of Patients in Group*	*Dysgraphia* No.	*Dyscalculia* No.	*Finger Dysgnosia* No.	*Right-left Disorientation* No.
One component	33	5 (15%)	15 (45%)	4 (12%)	9 (27%)
Two components	32	21 (66%)	20 (62%)	7 (22%)	16 (50%)
Three components	23	22 (96%)	19 (83%)	8 (35%)	20 (87%)
Four components	23	23 (100%)	23 (100%)	23 (100%)	23 (100%)
Totals	111	71 (64%)	77 (69%)	42 (38%)	68 (61%)

dyscalculia; four patients had finger agnosia, dysgraphia, and right-left disorientation; and 15 patients had disgraphia, dyscalculia, and right-left disorientation.

Statistical analysis of our data for the strength of association between the Gerstmann components, as carried out by Benton (1961), is vitiated by the small number of individuals in the groups and the fact that our data are subject only to binary classification. A further problem relates to the design of the present study, particularly insofar as one group of subjects was selected because they represented only one of the components. These subjects, then, could only diffuse the correlation between the components. Since the design of this study is not appropriate to this type of analysis and the reliability of the coefficients would be questionable, any interpretation of the resulting analysis would be of little scientific value.

Our study indicates that brain lesions can be localized about as well by ignoring Gerstmann's syndrome and using all the other data as by considering it. As a pathognomonic localizing criterion, Gerstmann's syndrome was definitely misleading as to the site of the lesion in three of 23 patients in group IV. The sum total of information available from neurological diagnostic procedures appears to provide a more confident basis for estimating sites of lesions than does the single criterion of Gerstmann's syndrome. It is, of course, no surprise that the total pattern of deficits is more reliable for localization of lesions than any fragment of information considered in isolation. The data indicate that as the number of Gerstmann components increases, the likelihood of the lesion being in the left posterior parasylvian area also increases. In this sense, Gerstmann's syndrome has about the same localizing implications as dysphasia. On the other hand at least seven of our original 456 patients had a lesion in the left posterior parasylvian area but had no components of Gerstmann's syndrome.

The high incidence of dysphasia as well as the other neurological deficits in our patients with three or four Gerstmann components supports Benton's (1961) conclusions about associated cerebral impairment; however, Kinsbourne and Warrington (1962) reported dysphasia in only three of their 12 patients with Gerstmann's syndrome. In our study and Benton's, the patients were all examined by standardized and quantified methods, whereas Kinsbourne and Warrington did not list their methods of examination or criteria for aphasia. The case protocols of the nine patients considered as not being dysphasic by Kinsbourne and Warrington record difficulty in reading, naming, and oral spelling (along with constructional dyspraxia and other deficits). The authors offer no explanation as to why they ignored these obvious language difficulties in classifying their patients as not dysphasic. The impairment of intellectual abilities other than dysphasia in our groups of patients with Gerstmann components will be dealt with in another communication.

SUMMARY

Of 465 consecutive patients subjected to a standardized battery of neuropsychological tests, 111 had one or more components of Gerstmann's syndrome. Each of these 111 patients had some evidence of organic brain dysfunction in addition to Gerstmann components. As the number of Gerstmann components increased, the responsible brain lesions tended to be larger, more highly destructive of tissue, and to cause greater neurological impairment. Every patient with four Gerstmann components had associated evidence of severe impairment of brain functions and the lesion or underlying disease was likely to compromise survival of the patient. The syndrome is not to be regarded as an autonomous entity, but merges with numerous other neurological deficits, notably dysphasia. In agreement with Benton, we find no justification for singling out the four Gerstmann components as a separate syndrome, unless one is also prepared to recognize that any other arbitrary groups of concurrent deficits are also separate syndromes.

In at least three of 23 patients with all four Gerstmann components, the angular gyrus, as shown by necropsy examination, was not involved by the lesion. However, the probability that the left hemisphere contained a lesion increased with the number of Gerstmann components, and the probability of involvement of the left posterior parasylvian area also increased with the increase in the number of Gerstmann components. With two, three, or four Gerstmann components, the lesions were never restricted to the angular gyrus but tended to spread widely over the parietal, temporal, and occipital lobes. As to localizing significance, Gerstmann's syndrome has approximately the same degree of cogency as dysphasia.

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