A Proposed Reinterpretation of Gerstmann’s Syndrome

Alfredo Ardila*

Department of Communication Sciences and Disorders, Florida International University, Miami, FL, USA

*Corresponding author at: Department of Communication Sciences and Disorders, Florida International University, 11200 SW 8th Street, AHC3-431B, Miami, FL 33199, USA. Tel.: +1-305-348-2750; Fax: +1-305-348-2710.

E-mail address: ardilaa@fiu.edu (A. Ardila).

Accepted 9 October 2014

Abstract

Gerstmann’s syndrome includes the clinical tetrad of finger agnosia, agraphia, acalculia, and right–left confusion. Some disagreement remains with regard to the exact localization of the syndrome, but most probable it involves the left angular gyrus with a subcortical extension. Several authors have suggested that a defect in mental spatial rotations could simultaneously account for acalculia, right–left disorientation, and finger agnosia. It has been also suggested that semantic aphasia is always associated with acalculia; as a matter of fact, left angular gyrus has a significant involvement in semantic processing. In this paper, it is proposed that Gerstmann’s syndrome should include: acalculia, finger agnosia, right–left disorientation, and semantic aphasia, but not agraphia. When the pathology extends toward the superior parietal gyrus, agraphia can be found. A fundamental defect (i.e., an impairment in verbally mediated spatial operations) could explain these apparently unrelated clinical signs.

Keywords: Gerstmann’s syndrome; Angular gyrus syndrome; Semantic aphasia; Acalculia; Finger agnosia

Introduction

Gerstmann (1940) described a clinical syndrome associated to lesions in the angular gyrus of the left hemisphere which included four different signs: finger agnosia, right–left disorientation, agraphia, and acalculia. Agnosia is understood as an inability to recognize and understand external information in the absence of decreased sensory acuity; agraphia is an impairment in the ability to write; an acalculia is usually defined an impairment in numerical abilities as a result of brain pathology (Matsumoto, 2009). The existence of Gerstmann’s syndrome (also referred as “angular gyrus syndrome”), however, has been controversial (Benton, 1977, 1992; Botez, 1985; Poeck & Orgass, 1966; Strub & Geschwind, 1983). The disagreement is partially due to the fact that this syndrome usually unfolds as either an “incomplete” tetrad or it is associated to other cognitive deficits, including aphasia, alexia, and some perceptual disorders (Frederiks, 1985) (for a comprehensive review of its history, see Rusconi, Pinel, Dehaene, & Kleinschmidt, 2010).

Some authors have attempted to estimate the frequency of Gerstmann’s syndrome; for instance, in a recent study Zukic, Mrkonjic, Sinanovic, Vidovic, and Kojic (2012) found in a sample of 194 acute stroke patients that 59 had alexia, agraphia and acalculia, or different combinations of these disorders. Only two patients had agraphia and acalculia associated with other part of tetrad of Gerstmann’s syndrome: finger agnosia and left–right disorientation. They concluded that Gerstmann’s syndrome is a quite unusual clinical entity.

Localization

Some disagreement remains with regard to the exact localization of the syndrome. The appearance of a Gerstmann’s syndrome with electrical stimulation of the cerebral cortex in the posterior parietal area would support its angular localization (Morris, Luders, Lesser, Dinner, & Hahn, 1984). Other reports throughout the history have supported the left angular gyrus localization;
for instance, Mazzoni, Pardossi, Cantini, Giornetti, and Arena (1990) described a case of a ‘pure’ Gerstmann’s syndrome associated with an angular gyrus traumatic damage. More recently, new cases of Gerstmann’s syndrome resulting from left angular gyrus pathology, without other associated deficits, have been reported (e.g., Carota, Di Pietro, Ptak, Poglia, & Schneider, 2004). Some other authors, however, have found pure Gerstmann’s syndrome associated with subangular focal subcortical white matter (e.g., Martory et al., 2003; Mayer et al., 1999).

Several researchers have expressed the opinion that the co-occurrence of the four clinical signs is rather coincidental; for instance, Wingard and colleagues (2002) state that the four cognitive functions impaired in Gerstmann’s syndrome do not share a common neuronal network, and their co-occurrence with dominant parietal lobe injuries may be related to the anatomical proximity of the different networks mediating these functions. Kleinschmidt and Rusconi (2011) proposed that a focal lesion in left parietal white matter provides the only tenable explanation for pure Gerstmann’s syndrome. Such a lesion would affect not only a single fiber tract but crossing different fiber tracts and hence disconnect separate cortical networks. Similarly, Rusconi and colleagues (2009) proposed that pure forms of Gerstmann’s tetrad do not arise from lesion to a shared cortical substrate but from intraparietal disconnection after damage to a focal region of subcortical white matter. Rusconi and colleagues (2010) further consider very unlikely that damage to the same population of cortical neurons should account for all the four symptoms. They propose that a pure form of Gerstmann’s syndrome might arise from disconnection, via a lesion, to separate but co-localized fiber tracts in the subcortical parietal white matter.

Despite that there is not a definitive answer, it could be tentatively concluded that the most probable localization of Gerstmann’s syndrome involves the left angular gyrus with a subcortical extension. The angular gyrus is extensively functionally and structurally connected with diverse brain areas including the ventral premotor areas, ventrolateral and ventromedial prefrontal cortex, posterior cingulate, and hippocampus (Uddin et al., 2010).

**Components of Gerstmann’s Syndrome**

It has been traditionally assumed that Gerstmann’s syndrome includes finger agnosia, right–left disorientation, agraphia, and acalculia, but not aphasia. Commonly, investigators have reported the presence of Gerstmann’s syndrome without aphasia as one of its components (Roeltgen, Sevush, & Heilman, 1983; Varney, 1984). Some other authors (e.g., Benton, 1959, 1977, 1992), however, have proposed a role for aphasia in many cases of Gerstmann’s syndrome; this idea has been supported by different clinical studies (e.g., Heimburger, Demyer, & Reitan, 1964; Poeck & Orgass, 1966).

The potential existence of semantic aphasia, nonetheless, has been rarely explored (Ardila, Lopez, & Solano, 1989). Head (1926) proposed a language impairment characterized by an inability to recognize simultaneously the elements within a sentence; Head called this disorder as “semantic aphasia.” In the following years, several authors referred to this type of aphasia (e.g., Conrad, 1932; Goldstein, 1948; Zucker, 1934). Luria (1966, 1973, 1976) restated this syndrome and analyzed it extensively. Since then, only some few references have appeared in the neuropsychology literature (Ardila et al., 1989; Benson & Ardila, 1996; Brown, 1972, Hier, Mogil, Rubin, & Komros, 1980; Kertesz, 1979). The International Neuropsychological Society Dictionary of Neuropsychology (Loring, 1999) defines semantic aphasia as an “Aphasia subtype described by Henry Head and Aleksandr Luria in which there is an impaired capacity to draw inferences beyond the literal meaning of the word” (p. 18). Luria (1966, 1973, 1976) considered that language deficiencies of semantic aphasia are evident in the following conditions: first, sentences with a complex system of successive subordinate clauses; second, reversible constructions, particularly of the spatial and temporal type; third, constructions with double negative (observed in some languages such as Russian and Spanish); fourth, comparative sentences; fifth, passive constructions; sixth, constructions which include transitive verbs; and seventh, constructions with diverse attributive relations. He also stated that the spatial disorders are not coincidental; they represent the semantic aphasia itself. Semantic aphasia represents a deficit in the perception of simultaneous structures transferred to a linguistic level. In other words, patients with semantic aphasia have difficulty understanding the meaning of words having a spatial or semi-spatial meaning, such as place adverbs and prepositions.

Ardila, Concha, and Rosselli (2000) reported a 58-year-old right-handed man, with an ischemic lesion to the left angular gyrus, who showed substantial difficulties in understanding logic-grammatical relationships, comparison adverbs (e.g., bigger—smaller, younger—older, etc.), place adverbs (e.g., over—below, on—beneath, etc.), and time adverbs (e.g., before—after), compatible with a semantic aphasia. Writing difficulties were minimal. In addition, he had important impairments in finger agnosia, right—left discrimination, and in understanding numbers, using numerical concepts, and performing arithmetical operations. That is, he presented kind of “incomplete” Gerstmann’s syndrome plus semantic aphasia.

Acalculia represents one of the four clinical signs included in the Gerstmann’s syndrome. The term “acalculia” was introduced to refer to the disturbances in mathematical abilities observed in cases of brain pathology (Henschen, 1925). Frequently, two types of acalculia are distinguished: primary and secondary (Berger, 1926). Secondary acalculia refers to a calculation defect resulting from an associated cognitive deficit in attention, executive functions, or language for example. Several different acalculia subtypes...
are described in contemporary literature (e.g., Ardila & Rosselli, 2002; Hittmair-Delazer, Hittmair-Delazer, Semenza, & Denes, 1994). Primary acalculia (also named as anarithmetia) is observed in cases of left angular gyrus pathology (Ardila & Rosselli, 2002; Benson & Weir, 1972; Grafman, 1988; Harvey, Goldstein, & Spiers, 1993; Hécaen, Angelergues, & Houiller, 1961; Rosselli & Ardila, 1989, 1997). Furthermore, it has been observed that the left angular gyrus mediates the retrieval of arithmetic facts during problem solving (Grabner et al., 2009, Grabner, Ansari, Koschutning, Reishofer, & Ebner, 2013). Dehaene, Molko, Cohen, and Wilson (2004) have proposed that the most crucial area for the representation of quantities is the intraparietal sulcus. Luria (1966, 1973) emphasized that semantic aphasia, that is the inability to use verbally mediated spatial and quasi-spatial concepts, is always associated with acalculia.

Calculation abilities seem to develop from counting; in child development counting begins with the sequencing of fingers (correspondence construction) (Hitch, Cundick, Haughey, Pugh, & Wright, 1987). Finger names are usually sequenced in a particular order (Ardila, 1993; Cauty, 1984; Levy-Bruhl, 1910/1947). Contemporary numerical systems are derived from finger knowledge and naming. In fact, in different contemporary languages, a 10- or 20-base system is found, simply because we have 10 different fingers. From the Latin digitus, the word ‘digit’ can mean both number and finger. Consequently, a strong relationship between numerical knowledge and finger gnosiss is evident and some commonality in brain activity and anatomy can be expected. It can be speculated that for prehistorical man finger agnosia and acalculia could have represented just the same deficit. Finger agnosia encompasses the inability to distinguish, name, or recognize the fingers not only in their own hands, but also in the examiner’s hand or in a drawing of a hand. Most evident errors are observed with the index, middle, and ring fingers. Interestingly, finger agnosia may be associated with toe agnosia as well, but the last one is rarely explored (Tuch, Steup, Christian, & Lange, 1997).

Right–left disorientation refers to the disturbances in the ability to identify right and left in one’s own body and in that of the examiner. It consequently includes not just linguistic but also spatial components. Right–left disorientation is also observed in cases of left posterior parietal damage and is included in the Gerstmann’s syndrome.

Agraphia is usually the absent clinical sign in cases of “incomplete” Gerstmann’s syndrome. It is assumed that agraphia in Gerstmann’s syndrome should not be associated with alexia (alexia with agraphia syndrome), and consequently, it corresponds to an apraxic or pure agraphia. It has been reported that apraxic agraphia is usually found in cases of left parietal pathology (Crary & Heilman, 1988; Valenstein & Heilman, 1979), probably left superior parietal damage (Otsuki, Soma, Arai, Otsuka, & Tsuji, 1999), and hence is not expect to be necessarily associated with acalculia and semantic aphasia. Using functional magnetic resonance imaging (fMRI), it has been observed that during writing to dictation significant clusters of activation are observed in left superior parietal lobe (SPL) and the dorsal aspects of the inferior parietal cortex (IPC) bordering the SPL. Localized clusters of activation are also observed in the left premotor cortex, sensorimotor cortex, and supplementary motor area (Menon & Desmond, 2001) confirming that left superior parietal area is directly involved in writing.

How to Understand Gerstmann’s Syndrome?

Gerstmann (1940) suggested that this syndrome represents a disorder of the body schema restricted to the hand and fingers. Hence, it is due to kind of disturbance in the body scheme. Some authors have considered that Gerstmann’s syndrome is just a fiction and it is simply an artifact due to incorrect observations (e.g., Benton & Meyers, 1956). Rusconi and colleagues (2010) have suggested that Gerstmann’s syndrome have been so enigmatic and problematic, because its signs lack a functional logic that would withstand more rigorous examination. They argue that Gerstmann’s syndrome does not (or if so only indirectly) reflect the functional architecture of cognition but rather the functional architecture of the brain and that it results from disconnection. According to another proposal, defective mental manipulation or transformation of images would be the common denominator; indeed, this interpretation has been supported by different authors since several decades ago (e.g., Ardila & Rosselli, 2002; Ardila et al., 2000; Carota et al., 2004; Mayer et al., 1999; Martory et al., 2003: see also Gold, Adair, Jacobs, & Heilman, 1995).

Gold et al. (1995) studied a patient with a Gerstmann’s syndrome following a focal infarct of the left angular gyrus; the patient’s right–left confusion could not be accounted for by either an aphasia or a degraded body schema; however, performance was always poor when mental rotation to a command was required. The authors suggested that a defect in horizontal translation, i.e., mental rotation, accounted for the right–left disorientation in their patient. Moreover, that acalculia and other signs associated with angular gyrus syndrome could also be due to a deficit in the performance of these mental rotations. By the same token, this deficit in mental rotations could potentially be reflected in the impaired understanding of comparisons, for example, time and place adverbs, observed in semantic aphasia. It could be conjectured that a single underlying deficit—defective mental rotations—could account for right–left disorientation, finger agnosia, acalculia, and semantic aphasia, and that their simultaneous appearance in a single clinical syndrome is not coincidental.

Notwithstanding, agraphia would still remain unexplained by this unifying underlying mechanism. It is interesting but not surprising that it is precisely agraphia that usually is missing in the “incomplete” Gerstmann’s syndrome. When agraphia is found, it corresponds to an apraxic agraphia, which correlates not exactly to angular gyrus, but to superior parietal area damage (Benson &
Cummings, 1985; Otsuki et al., 1999). Hence, agraphia will be observed only in cases of extension of the pathology to the superior parietal area.

Right–left discrimination and finger gnosis are strongly correlated; they could be interpreted as components of the autotopagnosia syndrome. Calculation ability might be regarded as a type of cognition involving in their origins at least: some type of body knowledge (autotopognosis), spatial concepts, and language (Ardila & Rosselli, 2002). Different authors have emphasized the significant association between spatial knowledge mediated through language and calculation abilities (e.g., Ardila et al., 1989; Luria 1966, 1976). When calculation defects in patients with Broca’s and Wernicke’s aphasia were studied using factor analysis, Dahmen, Hartje, Bussing, and Sturm (1982) were able to identify two different factors: numeric–symbolic and visual–spatial. Milder calculation defects found in Broca’s aphasia patients were derived from linguistic alterations, while visual–spatial processing defects significantly contribute to the calculation difficulties in Wernicke’s aphasia. Luria (1966, 1973) also emphasized the underlying spatial conceptualization deficits in the acalculia and the strong association between acalculia and so-called semantic aphasia.

Some authors have suggested that finger agnosia represents a mild form of autotopagnosia (e.g., Hécaen & Albert, 1978). However, dissociation between autotopagnosia and finger agnosia has been occasionally observed, and consequently they most likely should be interpreted as different disorders (De Renzi & Scotti, 1970). Finger agnosia is a relatively frequent defect, whereas autotopagnosia represents a rather unusual syndrome (Guariglia, Picardi, Puglisi Allegra, & Traballesi, 2002). The role of parietal lobe in body-knowledge and the disorders of the body scheme in cases of parietal pathology have been well established since long-time ago (e.g., Botez, 1983; Critchley, 1953). Parietal damage has been associated with asomatognosia in general, hemiasomatognosia, alloesthesia, finger agnosia, autotopagnosia, asymbolia for pain, apraxia, and the so-called Verger-Dejerine syndrome (Hécaen & Albert, 1978).

Pathogenesis of right–left disorientation is not totally clear. It is known that patients with left posterior damage present more evident difficulties than right posterior damaged patients (Ratcliff, 1979). It should be emphasized that right–left disorientation implies difficulties in the application of spatial concepts to the body’s lateral orientation. Gold et al. (1995) suggested that a defective horizontal mapping could account for right–left confusion and the other components of Gerstmann’s syndrome, because they all share a common dependency on relative spatial position.

Asymmetry in brain organization of cognition is regarded as the most outstanding characteristic of the human brain. LeDoux (1982, 1984) suggested that the primary functional distinction between human hemispheres involves the differential representation of linguistic and spatial mechanisms: while the right posterior parietal lobe is involved in spatial processing, the left is involved in linguistic processing. Spatial mechanisms are indeed represented in both the right and left parietal lobe in human ancestors including nonhuman primates, but in man language is represented in a region of the left hemisphere, the posterior parietal lobe, while spatial functions are represented in the corresponding right hemisphere (De Renzi, 1982; Lynch, 1980). In consequence, the evolution of language involved adaptations in the neural substrate of spatial behavior (LeDoux, 1984). Boles (1991) presented different tasks (recognition of words, products, locations, dichotic digits, etc.) and using a factor analysis was able to identify different lateralized parietal functions: lexical functions (e.g., word, numbers) were associated with left hemisphere, whereas spatial functions (e.g., locations of dots) were correlated with right hemisphere activity.

A Proposed Reinterpretation

It has been suggested that the angular gyrus represents a cross-modal hub where converging multisensory information is combined and integrated to comprehend and give sense to events and manipulate mental representations (Seghier, 2013). It could be proposed that Gerstmann’s syndrome (also referred as angular gyrus syndrome) is the result of a disturbance in the ability to verbally mediate some spatial knowledge (kind of verbal mediation of spatial operations), that is manifested both at the linguistic and at the numerical level. Gerstmann’s syndrome could be restated to include acalculia, finger agnosia, right–left disorientation, and semantic aphasia. Agraphia would still remain unexplained by this unifying underlying mechanism. However, when the pathology extends toward the superior parietal gyrus, agraphia can be found. As illustrated in Fig. 1, a single underlying deficit can potentially account for the simultaneous presentation of these four clinical signs.

Fig. 1. A single factor could explain the different signs of the Gerstmann’s syndrome.
This interpretation of the Gerstmann’s syndrome would make understandable the simultaneous appearance of so different clinical signs; as Rusconi et al. (2010) pointed out, Gerstmann’s syndrome have been so enigmatic and problematic because its signs lack a functional logic. The assumption that there is a single underlying common factor (“verbally mediated spatial operations”) responsible for the different clinical signs would provide this functional logic, and hence, would advance our understanding of so-called Gerstmann’s (or angular gyrus) syndrome.

References
