Gerstmann's syndrome

Neuropsychological deficit that is characterized by a constellation of symptoms that suggests the presence of a lesion in a particular area of the brain. (It should not be confused with Gerstmann-Sträussler-Scheinker syndrome, which is a transmissible spongiform encephalopathy.) Destruction to the inferior parietal lobule of the dominant hemisphere results in Gerstmann's syndrome.

It is named for Josef Gerstmann.

Primary symptoms

Gerstmann syndrome is characterized by four primary symptoms:

Dysgraphia/agraphia: deficiency in the ability to write

Dyscalculia/acalculia: difficulty in learning or comprehending mathematics

Finger agnosia: inability to distinguish the fingers on the hand

Left-right disorientation

Causes

This disorder is often associated with brain lesions in the dominant (usually left) hemisphere including the angular and supramarginal gyri near the temporal and parietal lobe junction. There is significant debate in the scientific literature as to whether Gerstmann Syndrome truly represents a unified, theoretically motivated syndrome. Thus its diagnostic utility has been questioned by neurologists and neuropsychologists alike. The angular gyrus is generally involved in translating visual patterns of letter and words into meaningful information, such as is done while reading. In adults

In adults, the syndrome may occur after a stroke or in association with damage to the parietal lobe.

In addition to exhibiting the above symptoms, many adults also experience aphasia, which is a difficulty in expressing oneself when speaking, in understanding speech, or in reading and writing. In children

There are few reports of the syndrome, sometimes called developmental Gerstmann syndrome, in children.

The cause is not known. Most cases are identified when children reach school age, a time when they are challenged with writing and math exercises. Generally, children with the disorder exhibit poor handwriting and spelling skills, and difficulty with math functions, including adding, subtracting, multiplying, and dividing. An inability to differentiate right from left and to discriminate among individual fingers may also be apparent. In addition to the four primary symptoms, many children also suffer from constructional apraxia, an inability to copy simple drawings. Frequently, there is also an impairment in reading. Children with a high level of intellectual functioning as well as those with brain damage may be affected with the disorder.

Treatment

There is no cure for Gerstmann syndrome. Treatment is symptomatic and supportive. Occupational and speech therapies may help diminish the dysgraphia and apraxia. In addition, calculators and word processors may help school children cope with the symptoms of the disorder.

Outcome

In adults, many of the symptoms diminish over time. Although it has been suggested that a similar diminishing of symptoms occurs in children as well, it appears more likely that most do not overcome their deficits, but instead simply learn to adjust.

Case reports

A 65-year-old right-handed man noted a sudden onset of numbness and weakness of the right hand. On the initial visit to the hospital, he showed severe acalculia, and transient agraphia (so called incomplete Gerstmann syndrome) and transcortical sensory aphasia. Brain MRI revealed a fresh infarct in the left middle frontal gyrus. The paragraphia and aphasia improved within 14 days after onset, but the acalculia persisted even at seven months after onset In an 123I-IMP SPECT study, the cerebral blood flow (CBF) was found to be decreased in the infarction lesion and its adjacent wide area, the ipsilateral angular and supramarginal gyri, and contralateral cerebellar hemisphere. Ando et al., speculate that inactivation in the infarction lesion caused the CBF decrease in the non-infarcted areas due to diaschisis. This case indicates that Gerstmann syndrome can be caused by not only dysfunction of the angular gyrus but also of the left middle frontal gyrus in the dominant hemisphere [1].

1)
Ando Y, Sawada M, Morita M, Kawamura M, Nakano I. [Incomplete Gerstmann syndrome with a cerebral infarct in the left middle frontal gyrus]. Rinsho Shinkeigaku. 2009 Sep;49(9):560-5. Japanese. PubMed PMID: 19928685.