Gerstmann's Syndrome

Synonyms: developmental Gerstmann's syndrome (when it occurs in children), Gerstmann tetrad

The condition should not be confused with Gerstmann-Sträussler syndrome or Gerstmann-Sträussler-Scheinker syndrome - a transmissible spongiform encephalopathy.

Description

This is a condition arising as a result of disease of the dominant parietal lobe at the angular gyrus. Possibly both superior and inferior lobes need to be affected. The specific effect of lesions of various lobes is discussed in the separate Space-occupying Lesions of the Brain article. However, in Gerstmann's syndrome in particular, the result is characterised by four components:

- Agraphia or dysgraphia
- Acalculia or dyscalculia
- Finger agnosia
- Left-right disorientation

Pure Gerstmann's syndrome is said to be without aphasia. It generally presents as either a congenital or learning disorder or as a feature of a stroke of the middle cerebral artery. Both forms are rare, especially the childhood form. It can also be a feature of neurodegenerative diseases such as Alzheimer's disease or as a result of head injury.

The childhood type may occur in those with brain damage or in isolation with otherwise good mental function. The pathophysiology in children is not understood. It does not seem to have a genetic component and is not listed in Online Mendelian Inheritance in Man (OMIM). It has been argued that developmental Gerstmann's syndrome is not a unique entity but a feature of other neurodevelopmental disorders.

In adults, the risk factors appear to be the same as those for a cerebrovascular event.

Presentation

There is loss or absence of four sensory abilities:

- Loss of the ability to express thoughts in writing (agraphia, dysgraphia).
- Inability to perform simple arithmetical calculations (acalculia).
- Inability to recognise or indicate one's own or another's fingers (finger agnosia).
- Inability to distinguish between right and left.

In addition, many adults also experience aphasia (difficulty in expression with speech, in understanding speech or in reading and writing).

The speech area is in the dominant hemisphere that is on the left in over 95% of right-handed people. It is also on the left in 75% of left-handed people; however, in the other 25% it appears to be bilateral.

Most paediatric cases are identified when children start school and they are challenged with writing and numbers. Generally, children with the disorder exhibit poor handwriting and spelling and difficulty with mathematical functions (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 have constructional apraxia, an inability to copy simple drawings. Frequently, there is also impairment in reading (dyslexia). Children with good intellectual function as well as those with brain damage may be affected.

Eliciting features of Gerstmann's syndrome

- Agraphia:
  - Illegible or very poor writing.
  - Inconsistencies in forming letters.
  - Mixture of upper- and lower-case letters or print and cursory writing.
  - Irregular letter sizes and shapes.
  - Unfinished letters.

They struggle to use writing for communication.

- Acalculia: this is tested by asking the patient to do serial subtraction of 7 from 100. This means 100, 93, 86, 79, 72, etc. It must be interpreted in the light of the educational level of the patient, including the age of a child. An easier test may be applicable, especially for children.

- Finger agnosia: finger agnosia is difficulty in distinguishing fingers on the hand. It is tested by requests like, "Touch my index finger with your index finger" and "Touch your nose with your little finger".

- Left-right disorientation: this is confusion of the right and left limbs and indicates a lesion in the dominant parietal lobe. It is tested by requests like, "Show me your left hand", "Touch your right foot" and "Touch your left ear with your right hand". A positive test is the inability to obey these commands in the presence of otherwise normal sensory and motor function.
Differential diagnosis

In adults, differential diagnosis is that of cerebrovascular events and dementia. In children, it is global brain damage and general learning disability.

Investigations

MRI scan will usually show a lesion of the angular gyrus in the left parietal lobe.

Associated diseases

As well as occurring in strokes, head injuries and developmental disorders, the syndrome has been associated with:

- Cerebral atrophy
- Alcoholism
- Carbon monoxide poisoning
- Lead poisoning
- Anaphylactic shock
- Systemic lupus erythematosus

Management

There is no cure for Gerstmann's syndrome. Treatment is supportive:

- Dysgraphia and apraxia can be helped by occupational and speech therapy. Calculators and word processors can also help school children cope with their disabilities.
- There has been a recognition of the association between finger recognition and numerical ability and there has been promising work done where improving finger gnosis through training exercises has led to improvements in the mathematical skills of young children.

Prognosis

Symptoms may lessen in adults over time. This is also thought to occur in children but it is more likely that they adapt to life with them. It has been suggested that early diagnosis and intensive treatment give a better outcome.

Further reading & references

7. Gerstmann's Syndrome; National Institute of Neurological Disorders and Stroke

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