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CASE REPORT

A Tetrad Cognitive Presentation of Gerstmann Syndrome: A Rare Manifestation of Stroke

SITI NUR IZNI SMA, KEVIN WCS, AHMAD KHALDUN I

Department of Emergency Medicine, Faculty of Medicine, Universiti Kebangsaan Malaysia Medical Centre, Jalan Yaacob Latif, Bandar Tun Razak, 56000 Cheras, Kuala Lumpur, Malaysia

ABSTRAK

Sindrom Gerstmann merupakan gejala neurologi yang jarang berlaku akibat kecederaan di bahagian otak kiri yang dominan dan tanda-tanda gejala ini adalah agrafia, akalkulia, jari agnosia serta disorientasi kiri-kanan. Kami ingin melaporkan kes melibatkan pesakit berumur 52 tahun lelaki, dengan dominan tangan kanan yang datang ke Jabatan Kecemasan dengan keadaan kekeliruan selama sehari. Beliau tidak mengenali masa, tempat dan orang. Hasil pemeriksaan tertumpu kepada sistem neurologi mendapati beliau mempunyai tanda strok atipikal iaitu tanda-tanda Sindrom Gerstmann. Oleh yang demikian, protokol strok telah diaktifkan dan pesakit telah menjalani ujian imbasan otak. Keputusan imbasan otak menunjukkan kawasan penumbra yang besar di bahagian 'left middle cerebral artery' teritori. Manakala, ujian susulan 'computed tomography' angiogram mendapati terdapatnya thrombus (darah beku) yang panjang di saluran 'left internal carotid artery'. Prosedur trombektomi tidak dapat diteruskan kerana terdapatnya trombus yang keras. Seterusnya beliau dirawat mengunakan rawatan anti-platlet. Beliau telah dibenarkan keluar dari hospital setelah tiga hari dan diberikan rawatan susulan sebagai pesakiy luar. Jadi, kami ingin menekankan betapa pentingnya mengenalpasti gejala Sindrom Gerstman sebagai salah satu tanda strok akut yang tidak tipikal.

Kata kunci: kecemasan, sindrom Gerstmann, strok

ABSTRACT

Gerstmann Syndrome is a rare neurological presentation due to a lesion in the left dominant parietal lobe, manifesting as a tetrad of agraphia, acalculia, finger agnosia and right-left disorientation. We present a case of a 52-year-old man, right hand

Address for correspondence and reprint requests: Ahmad Khaldun bin Ismail. Department of Emergency Medicine, Faculty of Medicine, Universiti Kebangsaan Malaysia Medical Centre, Jalan Yaacob Latif, Bandar Tun Razak, 56000 Cheras, Kuala Lumpur, Malaysia. Tel: +6013-3276273 Email: khaldun_ismail@yahoo.com

dominant, who presented to the Emergency Department with acute confusion for one day. He was disorientated to time, place and person. Focused neurological examination revealed atypical signs of acute stroke, comprising agraphia, acalculia, finger agnosia and right-left disorientation. Stroke protocol was activated and he underwent computed tomography brain perfusion, which confirmed a large penumbra area at the left middle cerebral artery territory. Computed tomography angiogram of the carotid arteries showed a long segment thrombosis in the left internal carotid artery. Emergency thrombectomy was abandoned due to impenetrable hard thrombus. He was treated with dual antiplatelet therapy and discharged after three days with partial improvement in terms of following commands, simple calculations and reduced finger agnosia. He was scheduled for outpatient follow up. This case highlights Gerstmann syndrome as an atypical manifestation of acute stroke.

Keyword: emergency, Gerstmann syndrome, stroke

INTRODUCTION

Stroke is characterised by neurological deficit of more than 24 hours as a result of acute focal injury to the central nervous system by vascular causes, including infarct, intracerebral hemorrhage and subarachnoid hemorrhage. Stroke has become a major cause of morbidity and mortality worldwide (Sacco et al. 2013). Notable risk factors for stroke include hypertension, diabetes, dyslipidemia, metabolic syndrome, smoking, alcohol, atrial fibrillation, large artery artherosclerosis and hypercoagulability. The clinical manifestation of stroke which results from cerebral artery occlusion depends on the affected brain territory of the supplied artery. A focused history and neurological examination may localise the site of the brain lesion (Ghiasian et al. 2019).

Gerstmann syndrome (GS) is a neurological disorder characterised by a tetrad of acalculia, agraphia, finger agnosia and right-left disorientation. This syndrome was named after Joseph Gerstmann, an Austrian-American Neurologist in 1924, who first recognised this four complex of neurophysiological deficit among his patients and hypothesised that the cause was due to focal ischemic lesion over the subcortical region, particularly the inferior part of left angular gyrus (Gerstmann 1940). Here we present a case of acute atypical stroke, which manifested as Gerstmann syndrome.

CASE REPORT

A 52-year-old right-hand dominant man was brought alone by taxi to the Emergency Department (ED) complaining of acute confusion for a duration of one day. He was unable to recall recent events and the reason for the hospital visit. Further history obtained from his sibling who arrived

two hours later confirmed the patient's abnormal behavior of forgetfulness and repetitive questioning. The sibling noted he was speaking appropriately the day before. There was no history of trauma, fever, facial asymmetry, visual impairment, slurred speech, weakness or numbness of limbs, imbalance or seizures. He had underlying hypertension and diabetes mellitus for the past few years and was taking antihypertensive and diabetic medication regularly from a general practitioner. He was also an active smoker and drank alcohol occasionally, but denied substance abuse.

Upon presentation, his vitals included a blood pressure of 210/120 mmHg, heart rate of 100 beats/minute, respiratory rate of 16 breaths/minute, afebrile and oxygen saturations in room air was at 99%. His Glasgow Coma Scale (GCS) was 14 (E4V4M6). He was disorientated to time, place and person. Upon neurological examination, his visual acuity and visual field were not impaired. Motor and sensory examination of the upper and lower limbs were normal. Cerebellar signs were negative and all cranial nerves were intact. Pronator drift was negative and his gait was grossly normal. However, he had a receptive aphasia with impaired attention and repetitive questioning. Systemic examination was unremarkable. He was hyperglycemic with a random capillary blood sugar of 18.0 mmol/L; however, there was no ketosis. Electrocardiogram (ECG) showed sinus rhythm with a left ventricular strain pattern.

Differential diagnosis included acute stroke, hypertensive encephalopathy and electrolyte imbalance. The stroke team graded a score of 5 on the National Institute of Health Stroke Scale (NIHSS) score and requested an urgent computed tomography (CT) scan of the brain with perfusion.

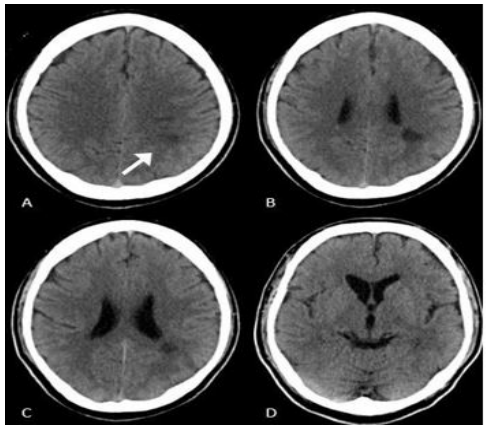

Figure 1: Non-contrast CT brain showing a hypodensity on the A:left mid-parietal region (white arrow), B & C: hypodensity of the left angular gyrus, D: loss of sulci and gyri indicates cerebral edema, which is consistent with left middle cerebral artery territory infarct.

A CT brain scan (Figure 1) revealed hypodensities at the left parietal lobe with a large penumbra area at the left middle cerebral artery territory. There was no intra-cerebral bleeding. Subsequent CT angiogram of carotid arteries (Figure 2) showed long segment thrombosis and stenosis of the left internal carotid artery (ICA) extending from the carotid bulb superiorly until the upper cervical segment.

Focused neurological examination revealed neurophysiological abnormality. He had difficulty in recognising numbers and an inability to do any simple mathematic calculations including addition and subtractions (acalculia). He was unable to write

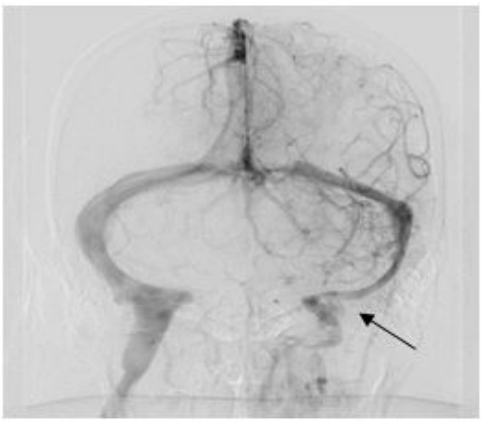

Figure 2: CT angiogram of neck and cranium shows non-opacification of the left internal carotid artery from the carotid bulb until the upper cervical segment (black arrow). All of the segments of left middle cerebral artery (MCA) are smaller in caliber due to stenosis and many collateral vessels are seen in the area of the brain supplied by the left MCA.

letters and draw during spontaneous writing or copying (agraphia). He had concomitant impairment in discriminating fingers (finger agnosia). He was also confused in distinguishing his right or left limbs and body parts, which led to hesitancy and frustration for the patient, due to right and left disorientation. Based on these neurological findings, the diagnosis corresponds with GS.

Meanwhile, his blood investigations, which included full blood count, serum electrolyte, renal function test, liver profile and coagulation test, were normal. The decision for an urgent thrombectomy was made by the neurologist, however, the procedure was not successful as there was difficulty in advancing the guidewire through the filling defect due to the presence of hard thrombus within the left ICA. He was admitted to the stroke ward and started on oral cardiprin 100 mg, clopidogrel 75 mg once daily, oral atorvastatin 40 mg on night, oral metformin 1 gm twice daily, oral gliclazide 60 mg once daily and omeprazole 20 mg once daily. On day two of admission, neurological examination demonstrated partial improvement of cognitive function. He was able to obey simple commands, perform simple calculations and able to recognise fingers. However, the other two features of this syndrome, agraphia and right-left disorientation persisted. He was discharged on day three of admission with neurology outpatient clinic follow-up.

DISCUSSION

This patient had an acute occlusion of the left internal carotid artery resulting in an acute infarct in the territory supplied by the left middle cerebral artery (due to artery-to-artery thromboembolism), specifically at the dominant parietal lobe which manifested as GS. This syndrome may manifest as an atypical presentation of acute stroke.

Gerstmann syndrome is a rare neurological disorder characterised by four cognitive symptoms which include agraphia (loss of ability to express thoughts in writing), acalculia (loss of ability to perform simple arithmetic problems), finger agnosia (loss of ability to recognise or indicate one's own or another's fingers) and right-left disorientation (loss of ability to distinguish between right and left side of one's body) (Gerstmann

1940; Tucha et al. 1997; Mayer et al. 1999; Osawa & Maeshima 2009). This set of clinical signs was first described by Joseph Gerstmann, an Austrian-American neurologist in 1924 when his patient presented with difficulty writing, performing simple calculations and difficulty moving fingers when examiner requested and right-left disorientation. Two similar cases were also reported by him three years later with additional symptoms of constructional apraxia and anomia. His findings were published in 1940 and the syndrome became globally recognised. Following the advancement of imaging technology, this clinical finding of cerebrovascular syndrome with a dominant left hemisphere involvement was attributed to specific lesions located at the angular gyrus of the left parietal lobe (Mayer et al. 1999; Long et al. 2019).

There are limited information regarding the incidence of GS among stroke patients because it is seldom reported. A study of 194 acute stroke patients showed 59 (30.4%) had alexia, agraphia and acalculia or different combinations of these. Meanwhile, only two patients (3.4%) had agraphia and acalculia associated with other signs of the GS tetrad (finger agnosia and right-left disorientation). Both of the patient's cranial CTs showed ischaemic lesions in the left parietal and left temporoparietal lobe, or the angular gyrus of the dominant hemisphere (Zukic et al. 2012). However, lesions to the dominant parietal lobe which manifested as GS are not only due to cerebrovascular infarction. Various insults to the brain such as brain tumours (Tucha et al. 1997), intracerebral hemorrhage (Osawa & Maeshima 2009), sickle cell disease (Buonanno et al. 2016) and anabolic steroid usage (Long et al. 2019) may present as GS. A recent article in 2019 reported that there are many conditions related to GS etiology includes aneurysms, carotid artery dissection or stenosis, chronic subdural hemorrhage, multiple sclerosis, cortical atrophy, alcohol, anaphylactic shock, carbon monoxide poisoning and lead intoxication. Meanwhile, developmental GS may occur among paediatric age group due to brain insult and transient GS manifested in patients with parietal lobe epilepsy (Altabakhi & Liang 2019).

The main function of the parietal lobe is perception of sensation and interpretation of sensory input. Lesions specifically located in the angular gyrus manifest as agraphia and acalculia. Meanwhile, lesions in the fronto-parietal cortex are postulated to cause finger agnosia and right-left disorientation, as a result of impaired perception of body orientation. (Gerstmann 1940; Rusconi et al. 2009; Lee et al. 2016; Joao et al. 2017; Patil & Kulkarni 2019).

In a published report of two cases of GS, the ischemic lesions were identified in the left medial frontal lobe sparing the angular gyrus (Lee et al. 2016). This is an uncommon pathological location for GS and is attributed to the disconnection hypothesis. It is postulated that this discrepancy is due to the disconnection of functional fibers between the co-localised tract

of the subcortical frontal and parietal cortex secondary to ischemic injury (Lee et al. 2016; Dimitrov et al. 2018; Patil & Kurkarni 2019).

A similar case of GS reported in Malaysia occurred in a 42-years-old lady with GS and diagnosed with high grade Glioblastoma Multiforme. She underwent surgical debulking of tumor, intraoperative radiation therapy, in combination with whole brain irradiation therapy and chemotherapy. As a result, she had a favourable outcome in 2 years and functionally independent after intensive therapy (Zamzuri et al. 2012).

A tetrad signs of GS may be reversible and can be treated based on etiology that includes treatment of stroke or hemorrhages, tumor surgery, removal of epileptic foci lesion and reversal of toxicity such as carbon monoxide poisoning. However, as the component of GS increase, it indicates the brain area affected were larger and likely result in neurological impairment. Following intensive rehabilitation and treatment, patients with GS may recover (Altabakhi & Liang 2019).

Therefore, the clinical findings of GS should be evaluated via neuroimaging and abnormalities may be seen at the dominant angular gyrus of the parietal lobe. Meanwhile treatment of GS is based on its individual etiology. In this patients, part of GS has been resolved after medical therapy most likely due to reperfusion to the penumbra area supplied by the left MCA.

CONCLUSION

GS is a rare clinical entity with a tetrad of cognitive symptoms due to damage to the dominant parietal lobe. Our case highlights the importance of recognising this syndrome in suspected cases with atypical presentation of acute stroke. Hence, it is vital to have knowledge on the diverse presentation of acute stroke in order to institute early treatment.

REFERENCES

Altabakhi, I.W., Liang, J.W. 2019. Gerstmann Syndrome. *StatPearls [Internet]*. https://www.ncbi.nlm.nih.gov/books/NBK519528/ [28 June 2022]

Buonanno, F.S., Schmahmann, J.D., Romero, J. M., Makar, R.S. 2016. CASE RECORDS of the MASSACHUSETTS GENERAL HOSPITAL. Case 10-2016: A 22 year old man with Sickle Cell Disease, Headache, and Difficulty speaking. *N Engl J Med* **374**(13): 1265-75.

Dimitrov, I., Atanasova, S., Kaprelyan, A., Ivanov, B., Nestorova, V., Drenska, K, Chuperkova, Z., Aleksandrov, I. 2018. Gerstmann's syndrome in a young man: A case report. *Trakia J Sci* **16**(3): 239-41.

Gerstmann, J. 1940. Syndrome of finger agnosia, disorientation for right and left, agraphia and acalculia: local diagnosic value. *Arch NeuroPsych* **44**(2): 398-408

Ghiasian, M., Moradi, A., Daneshyar, S. 2019. Gerstmann's syndrome in non-dominant hemisphere: a case report. *The Journal of Urmia University of Medical Science* **29**(10): 716-8.

Joao, R.B., Filgueiras, R.M., Mussi, M.L., de Barros, J. 2017. Transient Gertsmann's Syndrome as a manifestation of stroke: Case report and brief literature review. *Dement Neuropsychol* **11**(2): 202-5.

Lee, E.J., Shin, H.Y., Noh, Y., Park, K.H., Park, H. M., Lee, Y.B., Shin, D.J., Sung, Y.H., Shin D.H. 2016. Two cases with cerebral infarction in the left middle frontal lobe presented as Gerstmann's Syndrome. *J Neurol Disord* **4**(7): 1-3.

Long, N., Bassi, S., Pepito, D., Akhondi, H. 2019. Gerstmann Syndrome complicating polycythemia secondary to anabolic steroid use. *BMJ Case Report CP* **12**(6): e229004

Mayer, E., Martory, M.D., Pegna, A.J., Landis, T.,

Delavelle, J., Annoni, J.M. 1999. A pure case of Gerstmann syndrome with a subangular lesion. *Brain* **122**(6): 1107-20.

Osawa, A., Maeshima, S. 2009. Gerstmann's syndrome in a patient with left thalamic hemorrhage. *Neurol Asia* **14**(2): 161-4.

Patil. V.C., Kulkarni, A.R, 2019. Gerstmann's syndrome: A rare clinical condition with a tetrad of symptoms. *International Journal of Contemporary Medicine Surgery and Radiology* **4**(2): B173-5.

Rusconi, E., Pinel, P., Dehaene, S., Kleinschmidt, A., 2009. The enigma of Gerstmann's syndrome revisited: a telling tale of the vicissitudes of neuropsychology. *Brain* **133**(2): 320-32

Sacco, R. L., Kasner, S.E., Broderick, J. P., Caplan, L. R., Connors, J.J., Culebras, A., Elkind, M.S., George, M.G., Hamdan, A.D., Higashida, R.T., et al.; American Heart Association Stroke Council, Council on Cardiovascular Surgery and Anesthesia; Council on Cardiovascular Radiology and Intervention; Council on Cardiovascular and Stroke Nursing; Council on Epidemiology and Prevention; Council on Peripheral Vascular Disease; Council on Nutrition, Physical Activity and Metabolism. 2013. An updated definition of stroke for the 21st century: a statement for healthcare professionals from the American Heart Association/American Stroke Association. *Stroke* **44**(7): 2064-89.

Tucha, O., Steup, A., Smely. C., Lange, K.W. 1997. Toe agnosia in Gerstmann's syndrome. *J Neurol Neurosur Psychiatry* **63**(3): 399-403.

Zamzuri, I., Neuro, M.S., Rahman, G.I., Muzaimi, M., Jafri, A.M. and Ruzman, N.N. 2012. Polymodal therapy for high grade gliomas: a case report of favourable outcomes following intraoperative radiation therapy. *Med J Malaysia* **67**(1): 121-2.

Zukic. S., Mrkonjic, Z., Sinanovic, O., Vidovic, M., Kojic. B. 2012. Gerstmann's syndrome in acute stroke patients. *Acta Inform* **20**(4): 242-3.

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