EDORIUM Journals

Int J Case Rep Images 2022;13(2):91–95.
www.ijcasereportsandimages.com

CASE REPORT PEER REVIEWED | OPEN ACCESS

Alexia without agraphia in a young adult with ischemic stroke: A case report

Fasihah Irfani Fitri, Heru Pranata, Iskandar Nasution

ABSTRACT

Introduction: Ischemic stroke in young adults is less common than in older adults, but its occurrence warrants further investigation to determine the cause and appropriate treatment to improve outcome. One of the stroke manifestations is a disconnection syndrome, such as alexia without agraphia, a condition in which a patient cannot read but the ability to write remain relatively intact. Pure alexia is associated with the lesion in the medial occipitotemporal lobe in the dominant hemisphere, also known as the visual word form area (VWFA).

Case Report: Here we report a case of a 30-year-old male, who presented with an acute complaint of inability to read without any difficulty in writing, accompanied by right homonymous hemianopia which occurred two days before admission. Initial brain scan was normal. However, the repeated brain scan two weeks after admission showed infarction in the left occipitotemporal lobe. He had no known previous vascular risk factors other than obesity. The evaluation during admission showed high blood pressure, dyslipidemia, and suggestive of autoimmune disease, all of which might contribute to the ischemic stroke occurrence. During follow-up he was referred for the neuropsychological assessment and rehabilitation as well as to the outpatient clinic for use of antithrombotic as secondary prevention. He showed gradual improvement in his symptoms after sixth months follow-up.

Conclusion: Stroke in young adults is an increasing problem nowadays due to its rising incidence and more diverse pathogenetic mechanisms and related high morbidity. One of the neurological deficits found in stroke is a disconnection syndrome such as alexia without agraphia in which there is an acquired inability to read with relatively preserved writing ability. Pure alexia found in this case was due to an infarction of the occipitotemporal lobe that disconnects primary language areas from incoming visual information.

Keywords: Alexia, Ischemic, Pure, Stroke, Young adult

How to cite this article

Fitri FI, Pranata H, Nasution I. Alexia without agraphia in a young adult with ischemic stroke: A case report. Int J Case Rep Images 2022;13(2):91–95.

Article ID: 101333Z01FF2022

****

doi: 10.5348/101333Z01FF2022CR

Fasihah Irfani Fitri[1], Heru Pranata[2], Iskandar Nasution[1]

Affiliations: [1]Staff of Department of Neurology, Faculty of Medicine, Universitas Sumatera Utara, Medan, Indonesia; [2]Resident of Department of Neurology, Faculty of Medicine, Universitas Sumatera Utara, Medan, Indonesia.

Corresponding Author: Fasihah Irfani Fitri, Staff of Department of Neurology, Faculty of Medicine, Universitas Sumatera Utara, Jalan Dr. Mansyur No. 5, Medan 20155, North Sumatra, Indonesia; Email: fasihah.irfani@usu.ac.id

Received: 31 May 2022
Accepted: 25 July 2022
Published: 02 September 2022

INTRODUCTION

Stroke is defined as an acute neurological dysfunction presumed to be caused by ischemia or hemorrhage, persisting 24 hours or until death and ischemic stroke is an episode of neurological dysfunction caused by focal cerebral, spinal, or retinal infarction [1]. Stroke has been thought as the disease affecting older adults, since its incidence increases with age. However, demographic and technological transitions have resulted in changes in stroke incidence, where advances in technology can now cause a lack of physical activity in most people which can trigger the occurrence of one of the risk factors for vascular disorders such as obesity [2]. Nowadays, it is reported that the number of stroke patients at a younger age is increasing, which is statistically significant in various studies [3]. It is reported that stroke in children

and young people accounted for at least 15% of the total number of ischemic stroke cases [4]. This matter is now raising many questions regarding the age shift in the incidence of stroke, which initially affects elderly but now can also affect younger age which involves several risk factors that are related to genetic, congenital, metabolic, infectious, and autoimmune [5].

Stroke is the most common cause of acquired alexia, difficulty in reading. Alexia and agraphia are forms of brain disorders in which the patient experiences problems in reading and writing, respectively. These disorders are acquired as a result of brain damage, not attributed to blindness, paralysis, or other neurological deficits [6]. Pure alexia is associated with the lesion in the medial occipitotemporal lobe in the dominant hemisphere, also known as the visual word form area (VWFA), that is mainly involved in rapid word recognition and fluent reading. Alexia without agraphia is a disconnection syndrome which was first reported by Dejerine in 1892 [7]. It is commonly caused by an infarction of the medial aspect of the left occipital lobe, often with involvement of the splenium of the corpus callosum, following the occlusion of the left posterior cerebral artery (PCA) [8, 9]. We report a case of alexia without agraphia in a young adult with ischemic stroke.

CASE REPORT

A 30-year-old male right-handed patient presented at the emergency room with headache and a sudden difficulty in reading for two days before admission. He also complained of losing vision in the right half of visual field of both eyes and experienced difficulty in communication because of difficulty in finding the right words. There was no history of slurred speech, no difficulty in swallowing, and no history of limb weakness. He had no other known previous vascular risk factors other than obesity. Family history revealed stroke in his father and brother.

He was alert and oriented on the day of admission, had high blood pressure of 140/90 mmHg. His other general physical examinations were relatively normal without any remarkable findings. Both pupils were normal and reactive to light. Both optical discs were normal. He had right-sided homonymous hemianopia in both eyes. He did not have weakness of the limbs. The rest of his neurological examinations including sensory, coordination/cerebellar function, and reflexes were normal.

Cranial computed tomography (CT) scan examination done in the emergency room was reported to be normal (Figure 1). He was admitted into neurology ward for treatment with standard stroke protocol and to find out the cause of the stroke, considering his relatively young age. During his admission, several additional examinations other than the basic stroke assessments—such as chest X-ray, electrocardiography, and basic laboratory including complete blood test, lipid profile, blood glucose, renal and liver function tests—were done including assessment of autoimmune disease and several coagulation factors. There was an increase in the level of antinuclear antibody (ANA) test as much as 4 times from the normal value of 20 IU/mL, where the result was 82.9 IU/mL. There were also increase in both fasting and post-prandial blood glucose levels, which were 174 mg/dL (normal range is 70–105 mg/dL) and 200 mg/dL (normal range of 76–140 mg/dL), respectively. He had also dyslipidemia with increase in total cholesterol which was 284 mg/dL (normal value is <240 mg/dL) and in low density lipoprotein (LDL) cholesterol which was 240 mg/dL (normal value is <160 mg/dL). The basic cardiac assessment and the chest X-ray were normal. Neuropsychological examination revealed alexia, and impairments in working and episodic memory. He could read the actual letters but he had difficulty putting them together as words and he also had difficulty in understanding what he was reading. The writing ability remained intact, both for spontaneous writing and in response to dictation. He had mild slowing of speech with anomia but no apraxia of speech nor agrammatism were found. He showed difficulty in naming colors but was able to name and understand the use of objects presented visually, including faces. At this point the finding suggested lesion affecting the left occipital lobe within the left PCA territory so another cranial CT scan was requested. Brain magnetic resonance imaging (MRI) cannot be performed due to technical difficulty (he could not fit into the MRI machine). He was also referred for digital subtraction angiography (DSA) examination. The repeated CT scan two weeks after admission showed infarction in the left occipitotemporal lobe (Figure 2) and DSA examination showed normal cerebrovascular circulation. He was discharged home with antihypertensive, antilipidemic, and antiplatelet medications for secondary stroke prevention and was also prescribed cyclosporine as an immunosuppressant for the possible autoimmune disease. He was referred to a speech therapy rehabilitation program. On follow-up six months later his symptoms had improved greatly. He still had very mild difficulty in finding words during conversation, but the reading ability had significantly improved. He was able to read words and comprehend sentences.

DISCUSSION

Alexia without graphia is a classic disconnection syndrome due to lesion in the left occipital cortex and posterior aspects of the callosum body. It was first described by Dejerine in 1892 in a patient who experienced a sudden loss of the ability to read but had no other language disturbance, accompanied with a right homonymous heminaopia. Pure alexia is associated with the lesion in the medial occipitotemporal lobe in the dominant hemisphere, also known as the visual word

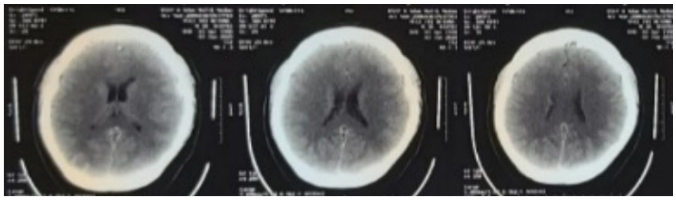

Figure 1: Cranial CT scan (normal).

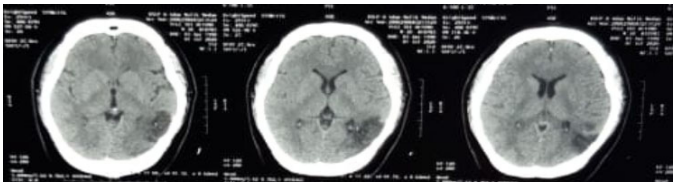

Figure 2: Repeated cranial CT scan showing left occipitotemporal infarction.

form area as the case presented here, that was associated with the occlusion if the left PCA [9, 10]. The patient had significant reading impairment while retaining the ability to write, both spontaneous writing and in response to dictation. He was able to spell letters and to comprehend spoken words. Lesions of the left occipital cortex cause right hemianopia. The spared primary language areas are disconnected from incoming visual information from the intact left visual field, due to the lesion in the splenium of corpus callosum. Meanwhile, because of the right hemianopia, words in the right visual field cannot be read. He also had color anomia as a result of disconnection between the cortical color centers from the language areas in the left hemisphere of the brain.

There are two areas of the posterior cortex that are needed for a person to be able to read fluently and automatically organize visual words. The visual word form area is located within the visual system in the fusiform gyrus in the left hemisphere. This area appears to pack the letters of a word into a visual unit. This system allows the reader to avoid scanning individual letters. A second area located close to the auditory cortex allows fluent readers to retrieve phonological word units from visual input [11, 12]. The infarction of the left occipitotemporal lobe in this case seemed to damage the VWFA, hence the patient was still able to read letter by letter but could not form the words and subsequently impaired his ability in reading and in reading comprehension.

Reading and writing are part of language modalities. The language areas of the brain are located along the sylvian fissure. Visual information is transmitted to the lateral geniculate nucleus of the thalamus to the primary visual cortex (area 17) and higher visual areas (area 18) then to the angular gyrus. The angular gyrus that is located behind Wernicke's area plays a role in processing information from the read word and then converting it into auditory forms of words in Wernicke's area. Wernicke's area is located in the posterior region of the superior temporal gyrus and is responsible for understanding auditory and visual information. Information from Wernicke's area is then projected through the arcuate fasciculus to Broca's area of the inferior frontal lobe. Broca's area processes the information that received from Wernicke's area to form detailed and coordinated vocalization patterns, then projects these patterns through the speech articulation area of the insula and then to the motor cortex to initiate the precise movements of the lips, tongue, and larynx to produce sound [6–11]. In this case the patient had no lesion in Broca, Wernicke, and arcuate fasciculus. He had only mild slowing of the speech and difficulty of finding the right words during conversation but this did not significantly impact the communication.

Stroke is less common in young adults but carry substantial functional impairment for patients and an economic burden on society. Thus the patient's relatively young age required further assessment of risk factors, mechanisms, and causes of ischemic stroke in this patient in order to administer proper treatment and prevent another stroke in the future. The causes of ischemic stroke in young adults include but not limited to heart disease, coagulation disorders such as protein S and C deficiencies, autoimmune diseases such as antiphospholipid syndrome, systemic lupus erythematosus (SLE), malignancy, metabolic syndrome, vasculopathy and vasculitis, carotid or vertebral artery dissection, and lifestyle risk factors including tobacco use, poor diet, physical inactivity, heavy alcohol consumption, illicit drug use (methamphetamine, cocaine, and heroin) [13]. The most obvious risk factor in the patient was only obesity but then during hospitalization, he was also found out to have several vascular risk factors including high blood pressure, diabetes mellitus, and dyslipidemia. The patient's laboratory results also showed an increase in the level of the ANA test despite no previous history of autoimmune disease. He was given cyclosporine tablets 2×100 mg as an immunosuppressant due to possible autoimmune disease. The presence of vascular risk factors and the possible underlying autoimmune disease could contribute to the occurrence of ischemic stroke in this patient.

Involvement of the nervous system in autoimmune, among others, can be caused by vasculitis which is characterized by the accumulation of mononuclear cells in the perivascular, without damage to blood vessels. Small infarction may occur due to occlusion of the lumen. There are various autoantibodies which directly or indirectly affect endothelial cells, causing damage to the vessel wall [14]. The increased/positive ANA test in this patient suggested an autoimmune disease, particularly SLE. Patients with SLE are at a 2- to 3-fold increased risk of developing any type of stroke compared to the general population. Atherosclerosis is accelerated due to the increased prevalence of traditional risk factors and immune-mediated processes are associated with an increased risk of stroke in patients with SLE. Hyperlipidemia, younger age, male, smoking, hypertension, homocysteine levels, and steroid use have

been reported to be risk factors for stroke in patients with SLE [13, 14]. Most of these conditions were also found in the patient that can increase the risk of ischemic stroke.

Systemic lupus erythematosus can affect both arterial and venous vascular beds and vessels of all diameters through both multiple inflammatory and noninflammatory mechanisms. Immune complex deposition can occur on the vascular walls and subsequent action of antibodies against them can cause endothelial damage resulting in secondary vasculitis. Arterial dissection is more common and can often be the initial manifestation in SLE. Cerebral small vessel disease with lacunar infarcts, white matter hyperintensities, cerebral microbleeds, and enlarged perivascular spaces, is more common in SLE and contributes to the increased stroke burden [14].

The patient was treated with antihypertensive, antilipidemic, and antiplatelet medications for secondary stroke prevention. Initial treatment for reading disorders is based on the level of impairment, meaning whether the impairment is at the level of syllables, words, phrases, sentences, or paragraphs [15]. Exercises that can be given to patients are practicing letter pairing, pair words with pictures, match words with words based on antonym and synonym categories, match words with their definitions, complete phrases or sentences, follow written orders, answer yes or no questions to a sentence or paragraph [15]. Some patients who have difficulty reading can also practice using the patient's fingers to pretend to copy the letters on words and sentences, very accurate and reading speed increased twice after four weeks of exercise using the strategy. Exercises using a tactile-kinesthetic strategy, writing with a pen on the left stair palm of the letter to be read also increases accuracy and speed in reading [16].

Pure alexia can be treated depending on the level of the deficit, at the word level it can be done with cross-modality cueing with kinesthetic therapy by trying to write letters on the palm of the hand or using fingers to make it easier for visual letter recognition; at the sentence level, it can be done by using multiple oral re-reading and oral reading for language in aphasia. Multiple oral re-reading is done by reading aloud the text over and over again quickly and the accuracy is calculated until the target reading level is reached. It is done at least 30 minutes a day for 3–5 times a day. Patients can be assisted at first, but they can do it themselves later [15]. Oral reading for language in aphasia is done by asking the patient to repeat aloud a short sentence or paragraph, initially guided and eventually expected to be independent [16].

In this case the symptoms improved gradually and showed significant improvement after six months. Long-term follow-up is needed to find out whether there is an improvement or worsening of the patient's clinical symptoms and to find out the conditions or risk factors that cause ischemic stroke at a young age in this patient, in order to keep it under control to prevent stroke recurrence.

CONCLUSION

Stroke in young adults is an increasing problem nowadays due to its rising incidence and more diverse pathogenetic mechanisms and related high morbidity. It has a negative impact and causes a significant burden for the patient and family. One of the neurological deficits found in stroke is a disconnection syndrome such as alexia without agraphia in which there is and acquired inability to read with relatively preserved writing ability. Pure alexia found in this case was due to an infarction of the occipitotemporal lobe that disconnects primary language areas from incoming visual information. Recognition of this syndrome is important for proper treatment and rehabilitation that will lead to better outcomes.

REFERENCES

1. Sacco RL, Kasner SE, Broderick JP, et al. An updated definition of stroke for the 21st century: A statement for healthcare professionals from the American Heart Association/American Stroke Association. Stroke 2013;44(7):2064–89.
2. Griffiths D, Sturm J. Epidemiology and etiology of young stroke. Stroke Res Treat 2011;2011:209370.
3. Singhal AB, Biller J, Elkind MS, et al. Recognition and management of stroke in young adults and adolescents. Neurology 2013;81(12):1089–97.
4. deVeber G, Singhal AB, Caplan LR. Stroke in children and young adults. In: Caplan LR, editor. Caplan's Stroke: A Clinical Approach. Cambridge: Cambridge University Press; 2016. p. 511–33.
5. Alchuriyah S, Wahjuni CU. The factors that affect stroke at young age in Brawijaya Hospital Surabaya. Jurnal Berkala Epidemiologi 2016;4(1):62–73.
6. Fitri FI, Lastri DN. Gambaran sindrom afasia kronik dan perubahannya pasca terapi. The Journal of Medical School (JMS) 2019;52(3):125–32.
7. Dejerine J. Contribution l'étude anatomo-pathologique et clinique des différentes variétés de cécité verbale. Mem Soc Biol 1892;4:61–90.
8. Cohen L, Dehaene S, McCormick S, Durant S, Zanker JM. Brain mechanisms of recovery from pure alexia: A single case study with multiple longitudinal scans. Neuropsychologia 2016;91:36–49.
9. Rupareliya C, Naqvi S, Hejazi S. Alexia without agraphia: A rare entity. Cureus 2017;9(6):e1304.
10. Sharma B, Handa R, Prakash S, et al. Posterior cerebral artery stroke presenting as alexia without agraphia. Am J Emerg Med 2014;32(12):1553.e3–4.
11. Barret K, Barman SM, Boitano S, Brooks HL. Ganong's Review of Medical Physiology. 24ed. New York: McGraw Hill; 2012. p. 283–95.
12. Yeatman JD, Rauschecker AM, Wandell BA. Anatomy of the visual word form area: Adjacent cortical circuits and long-range white matter connections. Brain Lang 2013;125(2):146–55.
13. George MG. Risk factors for ischemic stroke in younger adults: A focused update. Stroke 2020;51(3):729–35.

14. Timlin H, Petri M. Transient ischemic attack and stroke in systemic lupus erythematosus. Lupus 2013;22(12):1251–8.
15. Widyastuti K, Laksmidewi AAAP, Widyadharma PE. The aspect of neurocognitive and rehabilitation on alexia without agraphia case. Asian Journal of Pharmaceutical and Clinical Research 2018;11(2):4–6.
16. Kim ES, Lemke SF. Behavioral and eye-movement outcomes in response to text-based reading treatment for acquired alexia. Neuropsychol Rehabil 2016;26(1):60–86.

****

Author Contributions

Fasihah Irfani Fitri – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Heru Pranata – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Iskandar Nasution – Acquisition of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Guarantor of Submission

The corresponding author is the guarantor of submission.

Source of Support

None.

Consent Statement

Written informed consent was obtained from the patient for publication of this article.

Conflict of Interest

Authors declare no conflict of interest.

Data Availability

All relevant data are within the paper and its Supporting Information files.