

Alexia without Agraphia - A Case Report

Chanda¹, Prakash V. Suranagi², L. Krishnamurthy³, Deepak S.V.⁴

¹Post graduate, ²Professor and HOD, Department of Ophthalmology, SSIMS & RC, Davangere.

³Professor and HOD, Department of Neurology, SSIMS & RC, Davangere.

⁴Consultant Physician, Department of Medicine, Government Hospital, Kadur.

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Abstract :

Pure alexia, following an infarction in the distribution of the left posterior cerebral artery. is attributed to damage of the left occipital lobe and the splenium of the corpus callosum. We describe a case of pure alexia in a 64 year old woman with infarction of the left posterior cerebral artery and the splenium of the corpus callosum, a variation on this classic disconnection syndrome.

Purpose : A rare case of alexia without agraphia.

Methods : A 64 year old male non diabetic ,non hypertensive presented with acute onset of difficulty in reading.He was able to name individual letters or numbers but cannot read words or phrases.He retained the ability to speak, repeat speech, write and spell aloud. There was no history suggestive of coronary artery disease, trauma and seizure activity.The patient did not give any history of loss of consciousness, fever, headache, vomiting, dysphagia, dysarthria, gait ataxia or any weakness..

Results : General physical examination and higher mental functions were normal except for alexia. Ophthalmic evaluation revealed right hemianopia. Visual acuity,pupils,intraocular pressure and ocular fundi were normal. Patient had difficulty in reading and in naming colours but was able to match colours and shades. Blood investigations were under normal limits.

Conclusion : The classic syndrome of pure alexia without agraphia is caused by lesion in left occipital cortex and posterior sector of corpus callosum(the splenium).

Introduction : Alexia is defined as loss of reading ability in a person due to an acquired brain lesion .Pure alexia also known as “dejerine syndrome” after Joseph Jules dejerine who described it in 1892.¹ It is also called as 'pure word blindness', optic alexia. The selective impairment in the ability to read words, or even individual letters, has been repeatedly described in association with left dominant hemisphere lesions. A variety of reading disability in association with right non-dominant hemisphere lesions has also been reported.² Pure alexia almost always involves an infarct to the left posterior cerebral artery which perfuses the splenium of the corpus callosum and left visual cortex. The resulting deficits with pure alexia. This is because the left visual cortex has been damage leaving only the right visual cortex able to process visual information but it is unable to send this information to the language area (Broaca's ares ,Wernicke's area) in the left brain because of the damage to the splenium of corpus callosum.³

Case Report : A 64 year old male non diabetic ,non hypertensive presented with acute onset of difficulty in reading. He was able to name individual letters or numbers but cannot read words or phrases. He retained the ability to speak ,repeat speech, write and spell aloud. He was able to write a paragraph but not able to read it back. There was no history suggestive of coronary artery disease, trauma, blood transfusion, joint pains ,rashes, photosensitivity and seizure activity. The patient did not give any history of loss of consciousness, fever, headache, vomiting, dysphasia, dysarthria, gait ataxia or any weakness.

General physical examination and higher mental functions were normal except for alexia. Ophthalmic evaluation revealed right hemianopia. Visual acuity, including pupils, intraocular pressure and ocular fundi were normal. Patient had difficulty in reading and in naming colours but was able to match colours and shades. He was able to understand the nature and use of objects presented visually and recognise faces. Both upper and lower limb bulk, power, reflex and plantar response were normal. Sensory and cerebellar examination were also normal.

Complete blood count, blood sugar, electrolytes, coagulation profile, lipid profile were within normal

Address Correspondence to :

Dr. Chanda

Post Graduate, Department of Ophthalmology

SSIMS & RC, Davangere

Mob. : 94493 00070

E-mail : cdchandad@gmail.com



limits. Elisa for HIV was non reactive, carotid color Doppler, electrocardiogram, echocardiogram and chest x ray were normal.

MRI of brain shows acute left posterior cerebral artery territory infarction involving the occipital and postero lateral temporal lobe and left side of splenium of corpus callosum as seen in figures 1-4. Magnetic resonance angiography of circle of Willis showed no haemodynamically significant stenosis or occlusion.

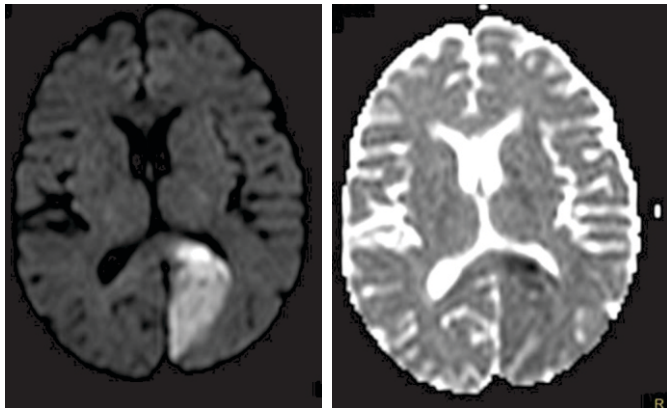


Fig 1

Fig 2

ADC axial

DWI axial

- Fig 1: Very high signal intensities in the medial aspect of left occipital lobe and left side of splenium of corpus callosum.
- Fig 2: Shows corresponding areas of low ADC value.

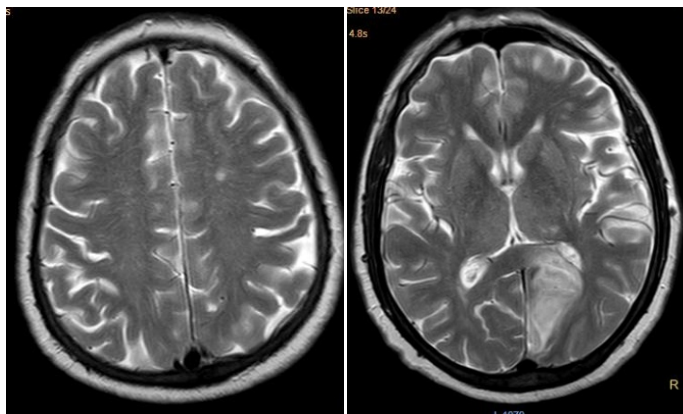


Fig 3

Fig 4

Flair axial

T2 axial

Fig 3 & Fig 4: Sharply defined inhomogeneous high signal intensified region as seen in the medial aspect of left occipital lobe and left side of splenium of corpus callosum

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Discussion :

In a classic syndrome of pure alexia without agraphia, patients can write but cannot read their own writing. Speech, auditory comprehension and repetition are normal. Naming deficits especially for colors may be present. Associated deficits include right hemianopia.

Dejerine and others concluded that loss of visual input from both occipital lobes to the dominate language area results in the inability to read without affecting the language area itself or the output from language region to the motor region similar to our study.[1] This 64 year old showed clinical evidence of typical alexia without agraphia, secondary to a left occipital infarction. Benson and Geschwind reviewed the 17 reported cases of alexia without agraphia with autopsy findings.[2,4] In all of them, with only one exception (Gloning et al., 1955), the left occipital lobe was involved as seen in our study[5]. Our patient had difficulty in identifying colours properly and this has also been well documented in cases of alexia without agraphia, a finding consistent with Genjiro's study.[6]

Conclusion : The classic syndrome of pure alexia without agraphia is caused by lesion in left occipital cortex and posterior sector of corpus callosum (the splenium) thus interrupting the flow of visual input in to the language network. There is usually a hemianopsia, but the core language network remains unaffected. Pure alexia may or maynot be associated with colour naming and visual object agnosia.

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