Alexia without Agraphia-report of Five Cases and Review of Literature

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Abstract
Alexia without agraphia (also called pure alexia or word blindness) was the first of the disconnection syndromes to be described. It results from the loss of visual input to the language area without involvement of the language area. The most common cause is occlusion of the left posterior cerebral artery with involvement of left occipital cortex and the splenium of corpus callosum. However, it can also be caused by any lesion affecting the splenium of corpus callosum disrupting the white matter tracts from the left visual cortex to the angular gyrus. We hereby describe five cases of alexia without agraphia, of which three are due to involvement of the left occipital cortex and splenium, and two are due to involvement of the splenium of corpus callosum alone.

Introduction
Alexia without agraphia or pure word blindness is one of the classic disconnection syndromes.¹ Patients are unable to read (even words that they have just written) but retain the ability to write. This was first described by Dejerine in 1892.²,³ This disorder results from the loss of visual input to the language area without involvement of the language area or output from the language area to the motor cortex. The most common causative lesion is a stroke in the territory of left posterior cerebral artery with infarction of the medial occipital lobe, splenium of the corpus callosum and often the medial temporal lobe.³,⁴

We hereby report 5 cases of alexia without agraphia, all resulting due to left posterior cerebral artery infarct.

Case 1
A 77 year old right handed male, with long standing history of type 2 diabetes mellitus, presented to us with complaints of acute onset of blurring of vision in both eyes, associated with headache and vomiting. On examination, he was conscious, oriented. His word output was normal, repetition and comprehension was intact, he could write normally, but could not read. He could not read the words that he had just written. He had impairment in recent memory. Examination of the cranial nerves revealed a right homonymous hemianopia. He had no motor weakness or cerebellar signs. MRI brain was done and it showed T2/FLAIR hyperintensities with diffusion restriction in the left occipital lobe and splenium of corpus callosum (Figures 1, 2), suggestive of acute infarct. Hence the diagnosis of alexia without agraphia due to a left posterior cerebral artery infarct was made. He was started on antiplatelets and statins. On follow up at 1 month, he continued to have alexia without agraphia.

Case 2
A 64 year old right handed male, with history of type 2 diabetes mellitus and ischemic heart disease developed acute onset of vertigo, vomiting and blurring of vision both eyes. On examination, he was conscious, oriented. He had impairment in short term memory. His word output, repetition and comprehension was normal. He had difficulty in naming objects, he could write but he couldn’t read even what was just written by him. He had difficulty in identifying and matching colours. He had no motor weakness.

MRI brain showed T2 and FLAIR hyperintensities in the left occipital region and splenium of corpus callosum (Figures 3, 4). On follow up his symptoms were persisting.

Fig. 1: FLAIR hyperintensities in the left occipital lobe and splenium of corpus callosum

Fig. 2: Diffusion restriction in the left occipital lobe and splenium of corpus callosum

Fig. 3: FLAIR hyperintensities in the left occipital region and splenium of corpus callosum

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Case 3

A 63 year old right handed female, hypertensive, initially visited the ophthalmology out patient department with complaints of blurring of vision of both eyes, from where he was referred to us. On examination, she was conscious, oriented, had a blood pressure of 180/110 mm of Hg. She had difficulty in naming colours. She could write normally but could not read, suggestive of alexia without agraphia. Her other language functions including word out put repetition and comprehension was normal. A CT brain revealed infarct involving the splenium of corpus callosum (Figure 5). On follow up she had persistence of alexia without agraphia.

Case 4

A 50 year old right handed male, hypertensive, diabetic, presented to us with inability to read the morning newspaper, though he had no difficulty in seeing objects. On examination, he was conscious, oriented, had a blood pressure of 140/90 mm of Hg. He could not read, though could write normally, suggestive of alexia without agraphia. His other language functions including word out put repetition and comprehension was normal. A MRI brain revealed infarct involving the the splenium of corpus callosum on the left side (Figures 6, 7, 8). On follow up he had persistence of alexia without agraphia.

Case 5

A 75 year old right handed male, smoker, with no co morbidities, presented to us with complaints of acute onset of headache and vomiting, and difficulty in seeing objects on the right side. On examination, he was conscious, oriented. His word output was normal, repetition and comprehension was intact, and he had difficulty in naming objects. He could write normally, but could not read. Examination of the cranial nerves revealed a right homonymous hemianopia. He had no motor weakness. CT brain showed a hypodensity in the left occipital lobe and splenium of corpus callosum (Figure 9) suggestive of acute infarct.
Discussion

Pure alexia without agraphia or pure word blindness is a disorder in which the patient can write but cannot read their own writing. Alexia without agraphia is a rare but “classic” disconnection syndrome. It was Dejerine, a French neurologist, who described this syndrome. He was the first to describe a patient with this syndrome, whose autopsy revealed an infarct of the left posterior cerebral artery territory involving the splenium of the corpus callosum and the medial portion of the left occipital lobe. He concluded that it is a disconnection syndrome which disrupts the visual input from the occipital lobes to the dominant angular gyrus, without involvement of the language area as such.

There are two theories in the pathogenesis of this syndrome. The dominant angular gyrus is located in the inferior parietal lobule of the cerebral hemisphere, usually the left, and is involved in phoneme processing in language comprehension and phoneme production for repetition and speech. It receives direct input from the left visual cortex. Input from the right visual cortex reaches the left angular gyrus through the splenium of the corpus callosum (Figure 10). Hence, in a lesion involving the occipital lobe and the splenium, vision (the right visual cortex) and language in the form of speech and writing (left angular gyrus and the motor cortex and its projections) are spared, but the patient is unable to read, as visual information cannot be transmitted to (has been “disconnected” from) the language area. In the reported cases of alexia without agraphia, infarction in the distribution of the left posterior cerebral artery (PCA) is the most common pathologic process. Since the visual cortex is supplied by the posterior cerebral artery, infarction of this area results in a right homonymous hemianopia, which is usually associated with this syndrome. There have been rare reports of alexia without agraphia not associated with right homonymous hemianopia. This brings in the second theory - a single lesion proximal to the language area (with sparing of the left visual cortex) affecting the fiber tracts from both visual cortices can explain these symptoms (Figure 11).

In our third and fourth case, our patients had alexia without agraphia, not accompanied by right homonymous hemianopia. MRI brain in these patients revealed an infarct adjacent to the splenium of corpus callosum, without involvement of the left occipital lobe, fitting with the second theory of this syndrome. All the other patients had infarct involving the left occipital cortex and the splenium of the corpus callosum.

References