

Large Acute Cerebral Hemorrhage Presenting With Pure Alexia

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Alexia (the acquired inability to read) is an uncommon presenting complaint in the emergency department (ED). It is usually associated with a lesion in the brain located within the dominant hemisphere near the parieto-occipital junction, with some involvement of the splenium of the corpus callosum. Our review of the literature revealed multiple distinct causes for the finding of alexia, and a majority of the cases uncovered also involved agraphia (the acquired inability to write) and frequently visual field defects. We present the case of an otherwise healthy 35-year-old white man who arrived at our ED with the chief complaint of having difficulty reading. He was, however, able to write, and he exhibited no defects in his visual fields on gross testing in our ED. The patient was found to have a large, acute, intraparenchymal hemorrhage in the right posterior/inferior parietal cortex, very near the occipital lobe. We present this case, followed by a brief discussion, to heighten awareness of the complaint of alexia with or without agraphia as a possible presenting symptom of intracranial hemorrhage, or ischemic cerebrovascular accident. [Ann Emerg Med. 2007;49:428-430.]

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INTRODUCTION

Alexia was first discussed in the medical literature in a case report by the French neurologist Joseph Jules Dejerine in 1891.^{1,2} The finding of alexia is typically associated with a lesion in the brain, located within the dominant hemisphere (the left hemisphere in the majority of patients) near the parieto-occipital junction, selectively involving the splenium of the corpus callosum.^{1,3} Dejerine theorized that a disconnection between the primary visual cortex and the angular gyrus is responsible for the syndrome. The angular gyrus is the portion of the brain that provides integration of visual and language information to allow reading, so any lesion here may result in alexia.^{1,4} Later researchers have suggested that a disruption of inter- and intrahemispheric pathways is responsible for the development of this symptom.⁴⁻⁶ At any rate, the brain fibers connecting the areas of the brain that receive visual input to the areas of the brain allowing language comprehension are somehow disrupted. Pure alexia patients have normal language skills and are able to write but are unable to recognize their own written words.^{1,2} Most reported cases of alexia have also involved agraphia or associated visual field defects.

The most common cause of isolated alexia is an infarct in the dominant occipital cortex. Alexia is a rare complaint in association with a thrombotic or hemorrhagic infarct and usually involves a lesion in the posterior cerebral artery distribution.⁷ Only about 5% to 10% of all ischemic infarcts involve the posterior cerebral artery distribution.⁷ Furthermore,

only a small percentage of these will produce isolated alexia without other findings suggestive of cerebrovascular accident.

CASE REPORT

A 35-year-old white man presented by private automobile to our emergency department (ED) with the chief complaint of having difficulty reading. On arrival, he denied any visual changes or blurred vision. The patient reported that the previous evening he experienced a right frontal headache that he believed to be consistent with previous headaches associated with his seasonal allergy symptoms. After taking an over-the-counter allergy medication, the headache resolved. Later that evening, the patient began reviewing some work in his home office and thought he could see clearly but could not interpret the subject he was viewing and even stated that he could not “make sense” of the ruler he used daily on his job. He attributed this difficulty to being “tired and stressed,” so he retired to bed for the evening. On awakening on the day of presentation to the ED, the patient felt well, completed his morning routine, and drove to work without difficulty. After arriving at work, the patient discovered that he was unable to read his e-mail or computer screen and immediately decided to seek medical care. He was left-hand dominant, with a medical history significant only for intermittent seasonal allergies, treated with over-the-counter medications. He had no known drug allergies, he worked as an architect, and a complete review of his systems yielded no physical abnormalities, except as stated above.



Figure. Noncontrast computed tomography of the brain revealing an intraparenchymal hemorrhage in the right posterior inferior parietal lobe, measuring 3.8 cm by 1.8 cm at its largest dimension, with surrounding edema.

Upon physical examination, the patient's vital signs were normal. He was a well-appearing man, well groomed, and in no acute distress. His pupils were equal, round, and reactive to light. Extraocular movements were intact, and his cranial nerve examination result was unremarkable. The patient's speech was articulate without slurring, his gait was steady and normal, he displayed full strength in all extremities (proximal and distal), and his cerebellar examination result was unremarkable (heel/shin, finger/nose/finger). His visual fields were fully intact, and he was able to write, identify letters, and follow multistep commands without difficulty. However, his examination revealed great difficulty in reading typed or written words; he pronounced every word slowly and phonetically. Unfortunately, no formal visual acuity test was documented, nor was the absence or presence of optokinetic nystagmus.

A computed tomography scan was ordered and revealed a right posterior inferior parietal lobe intraparenchymal hemorrhage, very near the occipital lobe, described by the radiologist as a "large hyperdense lesion with surrounding edema" (Figure).

The patient was admitted to the Stroke Service for observation and further evaluation of the source of intraparenchymal bleeding. He was eventually found to have a right occipital arteriovenous malformation of less than 1 cm in the distribution of the right posterior cerebral artery. He was treated with a γ -knife procedure, underwent successful rehabilitation, and was without sequelae at 6 months of follow-up.

DISCUSSION

Our literature search, performed via PubMed, located several reported cases of alexia, both with and without agraphia, as a result of either thrombotic or hemorrhagic lesions.^{4,8-13} In most of these case reports, the subject with alexia had associated visual field deficits, in light of the syndrome's involvement of the visual cortex. These defects involved the contralateral visual world to the area of the lesion. In that the majority of lesions associated with reported cases of alexia have occurred in the left hemisphere, the right visual world is affected in the majority of patients. The documented deficits were homonymous hemianopsia (most common), as well as superior or inferior quadrantanopsia.¹⁴ Our patient did not experience associated visual field deficits, and we believe this to be the first case report of pure alexia without agraphia or visual field defects as a presenting complaint for intracranial hemorrhage in the emergency medicine literature.

Although this is an uncommon chief complaint in the ED setting, our review of the literature reveals multiple distinct causes for the finding of alexia. As stated above, the most common cause seems to be an ischemic cerebrovascular accident in the posterior cerebral artery distribution, followed by an intracranial hemorrhage in this region as a result of a vascular abnormality, similar to our patient.^{4,8-13} Other causes documented in the literature include internal carotid artery dissection that extended to the posterior cerebral artery, preeclampsia with multiple punctuate hemorrhages, multiple sclerosis, frontal lobe hemorrhage with mass effect that caused compression of the posterior cerebral artery, cerebral venous thrombosis, toxoplasmosis, and one case of new-onset alexia after the biopsy of an intracranial mass.^{5,13,15-19} These lesions appear to affect the periventricular white matter of the dominant occipital lobe and the splenium of the corpus callosum and interfere with both inter- and intrahemispheric visual pathways.⁶ This area, including the fusiform gyrus of the dominant occipital lobe, is believed to be the area of the brain where word recognition occurs. Clearly, the language comprehension areas of our patient's brain were affected because he could interpret letters but not words. The majority of these cases are documented in the neurology and neuroradiology literature, and the majority of the lesions tend to occur in the left hemisphere of the brain. However, rare cases of right hemispheric lesions have been reported in left-hand-dominant patients (such as our patient) and have been attributed to right hemispheric dominance for language.⁹

Without a heightened awareness of the severe pathology that can be associated with the complaint of isolated alexia, cases such as this would undoubtedly be misdiagnosed at their acute presentation. The complaint of this non-ill-appearing young adult man without a significant medical history or risk factors could have been minimized because of the complete lack of verifiable findings on physical examination. Our patient admitted to an increased feeling of stress at work and home during the previous months, and psychiatric diagnoses were on

our differential diagnosis list, to be considered if our search for other pathologic conditions proved to be unrevealing. Our patient's lack of medical history, lack of previous visits to our ED, and lack of psychiatric history increased our concern and lowered our threshold for a more detailed medical evaluation. This evaluation led to the surprising discovery of a large intracranial hemorrhage. We have presented this case to heighten awareness that this serious condition can be associated with the rather unusual chief complaint of pure alexia. We recommend that the diagnosis of intracranial hemorrhage and cerebrovascular accident be considered in all patients who present to the ED with this complaint and that the appropriate diagnostic steps subsequently be taken.

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