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Case Report

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Bilateral cortical blindness due to bilateral occipital infarcts without anosognosia

Abstract

Bilateral cortical blindness refers to the total loss of vision in the presence of normal pupillary reflexes and in the absence of Ophthalmological disease resulting from bilateral lesions of the striate cortex in the occipital lobes. In most cases, these patients deny their blindness and their behavior is as if they have an intact vision. We report the case of an 84-year-old man with bilateral cortical blindness resulting from bilateral occipital lobe infarcts. The patient presented this infrequent clinical condition after acute bilateral infarction of the occipital lobes possibly due to cardiac embolism resulting from atrial fibrillation of unknown duration. Subcutaneous administration of low molecular weight heparin in therapeutic doses resulted in neurological improvement in the first four days. Interestingly, the visual symptoms were complicated neither by anosognosia nor by memory impairment. Cortical blindness and Anton's syndrome should be considered in patients with atypical visual loss and evidence of occipital lobe injury. Cerebrovascular disease could be the background in a bilateral cortical blindness, as in our patient. To our knowledge, this is the first reported case with bilateral cortical blindness due to simultaneous bilateral occipital infarcts but without anosognosia.

Keywords: Anosognosia, Bilateral cortical blindness, Bilateral occipital infarcts

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Introduction

Cerebral blindness is defined as bilateral loss of vision secondary to disruption in the visual pathways posterior to the lateral geniculate nuclei.[1] Cortical blindness, a particular type of cerebral blindness, is defined as the total loss of vision in the presence of normal pupillary reflexes and in the absence of ophthalmological disease,[2] resulting from bilateral lesions of the striate cortex in the occipital lobes.[1]

The most common cause of cortical blindness is bilateral occipital lobe infarctions in the vascular territory of the posterior cerebral arteries (PCA). PCA infarction is most often secondary to emboli from the heart or vertebrobasilar circulation, is less frequently due to prolonged hypotension or hypoxia and leads to homonymous hemianopia, often without other neurologic deficits. Simultaneous bilateral infarctions can be seen in a variety of clinical settings, such as hypertensive crisis, cerebral hypoperfusion and cardiac embolism to the basilar artery or transtentorial herniation. The overall prognosis of bilateral occipital lobe infarcts is poor.[1]

Gabriel Anton, in 1899, described patients with bilateral occipital lobe lesions who were completely blind and did not report any disturbance in their vision, leading sometimes to confabulation. The latter phenomenon has been referred to as Anton's syndrome, which is the most striking form of anosognosia and it has subsequently been reported by numerous investigators.[3]

We report the case of an 84-year-old man with bilateral cortical blindness but without anosognosia.

Case presentation

An 84-year-old right-handed man was transferred by his relatives to the emergency department of our hospital because of sudden bilateral loss of vision. The patient reported that 2 hours ago (at 14:00 in the afternoon), when he was getting up from his bed, he had an acute episode of vision and hearing loss with simultaneous instability and dysarthria, all of which resolved completely after a few minutes, except for vision loss. Past medical history included hypertension under medical treatment and surgery for gastric ulcer 40 years ago.

On admission, the patient's Glasgow Coma Scale score was 15 out of 15. Pupillary reflexes were bilaterally intact and fundoscopy yielded unremarkable findings except cataract in both eyes. The most striking clinical feature on examination was the complete loss of vision; as stated by the patient "In the morning I could see, and now I cannot see anything". Pursuit eye movements could not be evoked and reflexes to visual threats and optokinetic nystagmus were absent. He did not move his eyes to explore the room visually. His neurological examination was unremarkable. Blood pressure was 120/80mmHg and electrocardiogram showed atrial fibrillation of unknown duration with a ventricular response of 70bpm.

A first emergency computed tomography scan of the brain without administration of contrast agent (Figure 1A), obtained on admission, showed periventricular leukoencephalopathy, evidence of ischemic lesions in progress in the left occipital lobe and evidence of diffuse cerebral atrophy. Treatment with low molecular weight heparin (enoxaparin) and rosuvastatin was initiated. Four days later, the patient said that his vision had improved. Visual field analysis was not conclusive due to the patient's inability to cooperate effectively with the analyzer, but by a neurological and clinical evaluation, patient had tubular vision, as he could not see peripherally and he had to turn his head all the time to focalize on the subject that the doctor asked him to describe. He could recognize colors, shapes and faces, but he might have sequence agnosia, as although he could recognize parts of an image, he had difficulty describing the whole picture when he was asked to. A follow up brain CT with administration of contrast

agent (Figure 1B), was performed the fifth day of his admission, showed evidence of ischemic lesions in progress in the left occipital lobe, although these findings were not conclusive due to the presence of multiple small ischemic lesions, atrophy and periventricular leukoencephalopathy. The patient underwent magnetic resonance imaging of the brain twelve days after the episode, which showed diffuse cerebral atrophy, periventricular leukoencephalopathy and areas of increased signal intensity in both occipital regions, suggestive of ischemia (Figure 1C & 1D).

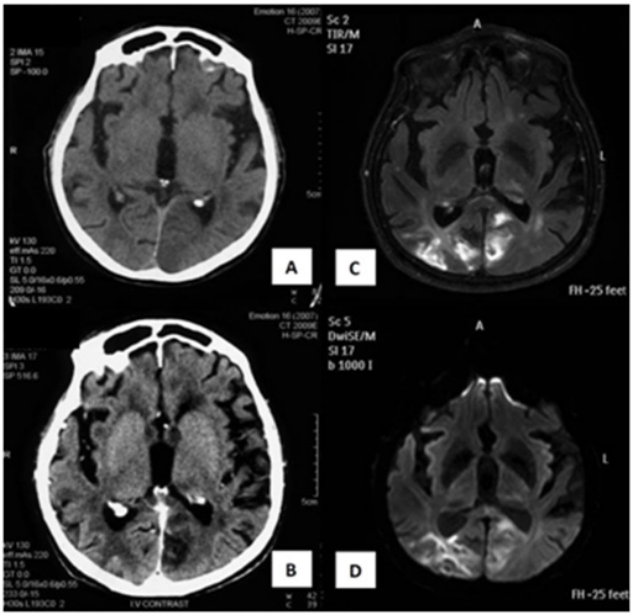

Figure 1 A-D. Emergency CT without contrast agent (A) and follow up CT with contrast agent (B) showed evidence of ischemic lesions in progress in the left occipital lobe. FLAIR MRI (C) and DWI MRI (D) showed areas of increased signal intensity in both occipital regions, suggestive of ischemia.

Discussion

Cortical blindness has been associated with bilateral lesions of the primary visual cortex of the occipital lobes secondary to hypoxia, cardiac arrest, vasospasm, cardiac embolism,[3] head trauma, intracranial hemorrhage, occipital lobe epilepsy,[4] hyponatremia, severe hypoglycemia,[2] treatment with antiepileptic agents, CT contrast media, Creutzfeldt-Jacobs disease, rapid onset of dementia, infection, congenital abnormalities of the occipital lobe,[5] eclampsia and rarely, pre-eclampsia. Sometimes, cerebral blindness can be transient when due to infective endocarditis,[6] epileptic seizures, trauma, toxins or hypertensive encephalopathy.[1]

Occipital cortex receives its blood supply from both middle and posterior cerebral arteries. This dual blood supply protects this area from ischemic damage in case of occlusion of one of these vessels.[2] However, the most common cause of cortical blindness is ischemia of the occipital lobe caused by occlusion of one or both of the posterior cerebral arteries. Complete cortical blindness is much less common than incomplete blindness.[7]

Bilateral occipital lobe infractions in the vascular territory of the posterior cerebral arteries are mostly secondary to emboli from the heart or the vertebrobasilar circulation. Additionally, prolonged hypotension or hypoxia can lead to watershed infarcts at the parieto-occipital junction between the middle and posterior cerebral arterial territories. Bilateral infarctions can also be seen in a variety of other clinical settings, including hypertensive crisis, cerebral hypoperfusion, basilar artery embolism or trans-tentorial herniation. The overall prognosis of bilateral occipital lobe infarcts is poor.

In most cases, patients with bilateral occipital cortex lesions develop simultaneously acute visual impairment, cortical blindness and anosognosia. This constellation of abnormalities, termed Anton's syndrome, is a form of anosognosia, in which patients with very poor or no vision deny the deficit even when confronted with evidence of the contrary, which in some cases leads to confabulation.

There are several brain disorders in which patients appear to have no awareness of neurological and neuropsychological disturbances that are typically associated with their condition. This has been reported in patients with Huntington's disease, Parkinson's disease, Alzheimer's disease, frontotemporal dementia and postacute severe traumatic brain injury.[8]

There is incomplete understanding of why patients with Anton's syndrome deny blindness, but two main theories have been proposed, implicating a disconnection phenomenon. The first implicates a disconnection between the visual processing system and the centralized "conscious awareness system", which is localized in the inferior parietal lobes and is responsible for monitoring modality-specific information and is linked to the other independent executive system, localized in the frontal lobes, that monitors and integrates multimodal information for more complex cognitive tasks.[9,10] The second theory suggests that a damage to a modality-specific "visual monitor", possibly located in the visual association cortex, or alternatively a disruption of the pathways connecting this visual monitor with areas mediating speech production,[11,12] so that patients are unable to verbally report what they see and thus confabulate a response. In this case, the visual monitor might not be destroyed, but might instead receive false feedback from a secondary visual system that is mediated by the superior colliculus, pulvinar, and temporoparietal regions and is functional enough to allow patients with severe hemianopsia due to lesions in their geniculocalcarine system to localize targets. If this system is intact, patients may feel as if they can see even if they actually cannot.

Additionally, some patients may have an impairment in emotional reactivity or memory. Others may have an intact primary visual cortex, but damages in the visual association cortex responsible for recognition and interpretation of the visual input. A converse of Anton's syndrome, Blindsight, may occasionally occur in patients with cortical blindness, allowing them to localize targets or motion, likely secondary to intact subcortical or extrastriate visual pathways.

Our patient had this unusual presentation after acute simultaneous bilateral infarction of the occipital lobes; bilateral vision loss without anosognosia or memory abnormalities. To our knowledge, this is a rare case with bilateral cortical blindness due to simultaneous bilateral occipital infarcts without anosognosia which could be attributed to either intact "conscious awareness system" or "visual monitor" and their connecting pathways, as we described above. These bilateral infarctions were possibly due to cardiac embolism resulting from atrial fibrillation of unknown duration. Prompt administration of low molecular weight heparin resulted in symptomatic improvement within the first four days.

Two weeks after admission, the patient was started on oral anticoagulation with Acenocoumarol, aiming at an international normalized ratio between 2.0 and 3.0. In subsequent evaluations after 1 and 2 months, the patient's status was stable without any further improvement in his vision. He could carry out most of his needs inside the home, but he needed help when he had to go outside of his house.

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A follow up visual field analysis was performed, but it was again inconclusive due to the patient's inability to cooperate effectively, but clinical evaluation of patients showed that had no change in his visual ability.

In conclusion, cortical blindness and Anton's syndrome should be considered in patients with atypical visual loss and evidence of occipital lobe injury. Cerebrovascular disease is the most common cause of bilateral cortical blindness, as in the patient reported herein. However, it can be presented atypically, which could be attributed to the complex and not completely known yet, the role of the primary visual cortex in its contribution to the visual awareness process.[13]

Acknowledgments

None.

Conflicts of Interest

The authors declare that they have no conflict of interest.

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