

Bilateral Cortical Blindness – Anton Syndrome: Case Report

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ABSTRACT

The aim of this work is the specify of rare neurological disorder, bilateral cortical blindness and Anton syndrome. It is about loss of vision in the presence of intact anterior pathways and a form of visual anosognosia, with resulting patient denial of blindness. This is a case of 72-year old man with history of diabetes and hypertension. Diagnosis is based on the exclusion of disease in the anterior visual tract by history and complete neuro-ophthalmological and radiological evaluation.

Keywords: Anton syndrome, bilateral, cortical blindness, case report

Introduction

Cortical blindness, a rare neurological disorder is characterized by loss of vision caused by unilateral or bilateral lesions and presence of intact anterior visual pathways. The lesions are most often binocular with preserved papillary light reflexes. They are results of an insult in the occipital lobe cortex.

Anton syndrome, a form of anosognosia, is a rare complication of cortical blindness, resulting from insult to the visual association cortex. It result with denial of blindness by patient who is clinically unable to see. They may offer excuses for their symptoms (»there is not enough light to see«), or they may endanger themselves »proving« that their vision is intact. Their affect is often described as inappropriate and they may confabulate during visual examinations, although complete indifference to the blindness. Without functioning visual association centers, these patients become detached from the concept of sight and are unable to acknowledge their loss¹.

Case

A 72-year-old man was admitted to our emergency department. He was accompanied by relatives because of »strange and confused behavior«. The patient was disorientated in the space with weakness of the left arm and leg. In his medical history there were diabetes and hypertension, but did not reveal any visual problem

prior to the accident. His initial physical examination was unremarkable. In his neurological examination we found lower position of the left mouth angle, and left arm and leg slowly fell to the base, with left weakened plantar response. There were not inequalities in myostatic reflexes and muscular tonus. The patient was mildly psychomotorically disturbed, conscious, alert and oriented to persons, place and date but exhibited no distress regarding his loss of vision.

Ophthalmologic examination proved blindness, but the patient denied his deficit. Eye movements in all directions were normal, with normal pupil light reflex. Fundoscopy and intraocular pressure were normal. Visual response to threat and optokinetic nystagmus could not be elicited.

A cranial CT scan demonstrated normal anterior visual pathways but reveled bilateral occipital infarcts (old) with small hemorrhagic conversions (new) (Figure 1).

Discussion

In etiology studies of cortical blindness case reports have included description of associations with cerebral venous thrombosis, pulmonary embolus, in pregnancy (preeclampsia and eclampsia), and cerebral vascular accidents²⁻⁴. The common pathologic component is ischemia of the occipital cortex, either as a result of local event (hemorrhage and embolism), or more commonly

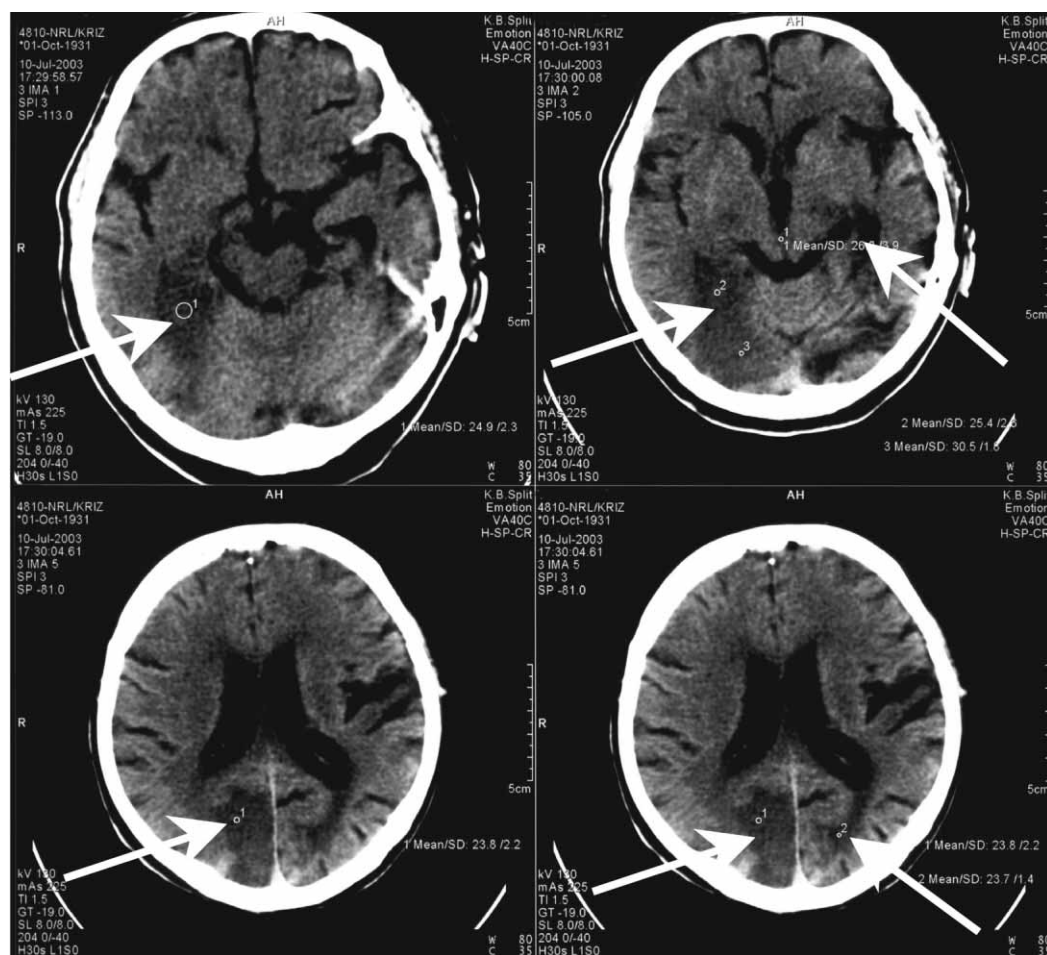


Figure 1. Cranial CT scan revealed bilateral occipital infarcts (old) with small hemorrhagic conversions (new).

as a result of global process. Stiller et al⁴ have postulated that the occipital cortex is especially sensitive to systemic hypoxia because of its relatively distal location from the central cerebral vasculature.

Alternate etiologies include migraine headache, occipital trauma (head injury with subdural or epidural hematoma^{5,6}), meningitis, carbon monoxide or other poisoning, and neoplasm⁷. Cortical blindness is sometimes difficult to differentiate from hysterical blindness since the pupils may still react to light. The diagnosis of cortical blindness is based on the exclusion of disease in the anterior visual tract by history and complete neuro-ophthalmologic evaluation. Pupillary response to light and corneal reflexes are intact because these functions are independent of cortical integrity. Functions dependent on the optic cortex, such as the blink response to threat and optokinetic nystagmus, are absent¹.

Computed tomography is useful in the exclusion of hemorrhage or neoplastic process. Although not pathognomonic, the presence of either low-attenuation areas in the occipital lobes or cerebral edema lends support to the diagnosis of cortical blindness¹. Magnetic resonance imaging is recommended by some authors^{8–10} as the di-

agnostic imaging technique of choice in these patients. Benefit of magnetic resonance includes superior detection of subtle vasogenic edema in the brain (characteristic of vasoconstriction and ischemia) as well as detailed evaluation of the venous sinuses and anterior visual tracts.

Conclusion

In this case the occipital lobes were affected bilaterally due to cerebral vascular accidents, old and new ones. A detailed history and physical, neurological and ophthalmological examination should be performed in a meticulous manner to avoid treatable intracranial pathology¹⁰. Computed tomography and magnetic resonance are useful in detailed evaluation of brain damage in cortical blindness.

The prognosis for patients with cortical blindness depends on the cause, severity, duration, speed of initial recovery, age, and medical history. In this case, expectation on poor visual prognosis considering his age, history of diabetes and hypertension, unfortunately was correct.

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BILATERALNA KORTIKALNA SLJEPOĆA – ANTON SINDROM: PRIKAZ SLUČAJA

SAŽETAK

Cilj ovoga rada je prikaz rijetkog neurološkog oboljenja, bilateralne kortikalne sljepoće i Anton sindroma. Radi se o gubitku vida s prisutnim intaktnim prednjim vidnim putevima i formi vidne anosognozije koja rezultira negiranje sljepoće od strane bolesnika. Ovo je prikaz slučaja 72-godišnjeg muškarca s dijabetesom i hipertenzijom u anamnezi. Dijagnoza se zasniva na eliminaciji oboljenja u prednjim vidnim putevima pomoću anamneze i potpune neuro-oftalmoške i radiološke obrade.