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**Clinical Case**

# Cortical Blindness Revealed by Stenosis of Basilar Trunk

## *Cécité corticale révélée par une sténose du tronc basilaire*

Kassoula Batomaguela Nonon-Saa[1, 3], Dadjo Amouzou[1,2*], Nidain Maneh[1,2], Koffi Didier Ayena[1,3], Lama Agoda-Koussema[1]

**Affiliations**
[1] Faculty of Medicine and Pharmaceutical Sciences, [1] Department of Ophthalmology, Faculty of Health Sciences, University of Lomé, Togo.
[2] Department of Ophthalmology, CHU Campus, Lomé, Togo.
[3] Department of Ophthalmology, CHU-Sylvanus Olympio, Lomé, Togo.

***Corresponding author**: Dadjo Amouzou
Phone: +22891989587. Email: dadjooson@gmail.com
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**ABSTRACT**

We report the case of a well-controlled 75-year-old hypertensive patient, brought for emergency care by his son for non-specific visual disorders. Clinical assessment evoked cortical blindness. A cerebral computed tomography (CT) and an angio-scanner of the supra-aortic vessels were requested revealing ischemic lesions on the territories of the posterior cerebral arteries related to basilar trunk stenosis. The peculiarity of this case is the occurrence of cortical blindness despite the good control of high blood pressure, which appears to be the only risk factor, and the absence of vision recovery after 13 months of follow-up, which is exceptional according to the literature.

**RÉSUMÉ**

Nous rapportons le cas d'un patient de 75 ans hypertendu bien contrôlé amené en urgence par son fils pour des troubles visuels non spécifiques. L'évaluation ophtalmologique a permis d'évoquer une cécité corticale. Une Tomodensitométrie cérébral et une angio-scanner des vaisseaux supra-aortiques ont été demandés révélant des lésions ischémiques sur les territoires des artères cérébrales postérieures en rapport avec une sténose du tronc basilaire. La particularité de ce cas est la survenue de cécité corticale malgré la bonne maîtrise de l'hypertension artérielle, qui semble être le seul facteur de risque, et l'absence totale de récupération fonctionnelle après 13 mois de suivi, ce qui est exceptionnel selon la littérature.

## INTRODUCTION

Cortical blindness (CB) is defined as loss of visual sensation in the entire visual field, without any ophthalmological causes and with normal pupillary light reflexes due to bilateral lesions of the striate cortex in the occipital lobes [1]. CB is a part of cerebral blindness, defined as loss of vision secondary to damage to the visual pathways posterior to the lateral geniculate nuclei. It is a rare pathology that is defined primarily by its specificity compared to other ophthalmological, neurological or psychiatric disorders [2]. In contrast to peripheral blindness, it does not affect the eyeball, the pupillary light reflexes, the retina or the optic nerve. The blink reflex is absent, in contrast to psychogenic blindness including malingering [1]. It is noted that cerebral blindness has replaced the term "cortical blindness", which was too restrictive because damages may be cortical or subcortical [3, 4]. We report the case of a well-controlled 75-year-old hypertensive patient with a CB. This is a special case in which there was no medical history or any risk factor other than hypertension, which was otherwise well controlled. Furthermore, patient showed no visual improvement after thirteen months of treatment and follow-up. This case is exceptional according to the literature [5].

## CASE PRESENTATION

A 75-year-old male patient was received in emergency for sudden bilateral visual impairment. According to the history, his son noticed that his father has haggard gaze, so he immediately took him to an ophthalmologist, while himself did not complain. Further clinical evaluation did not reveal any other ophthalmological or neurological history, but revealed a high blood pressure that was present since fifteen years and it was well controlled on treatment with a good compliance. There was no other risk factor such as diabetes or alcoholism. The patient has never been hospitalized.
The ophthalmological assessment revealed loss of visual perception and patient behaves like a blind (he cannot

move alone without help or a guide) with unquantifiable visual acuity. There was no blinking reflex and there was evidence of the abolition of the optokinetic-nystagmus reflex. Slip lamp examination noted a normal pupillary light reflex in both eyes, with no relative afferent pupil defect (RAPD). There was no lens opacity and the remaining anterior segment was normal in both eye.The fundus examination (FO) after full dilation revealed clear vitreous, normal optic disc with regular margin and non pathological cup. Macula and macular region were healthy. There were some copper wiring of the narrowed arterioles, and an arterio-venous crossing signs in both eyes. There was no bleeding and no exudate. Ocular motility and the corneal sensation were also normal. In addition, the patient had a good general condition and an undisturbed state of consciousness. Controlled blood pressure at the same day, has noted in both arms: Systolic Pressure of 145 mm Hg and Diastolic pressure of 90 mm Hg at rest.

Because of the sudden visual impairment and patient history, a vascular event was suspected. The brain computed tomographic scan allowed us to demonstrate ischemic lesions on the two posterior cerebral arteries areas **(Figure 1).**

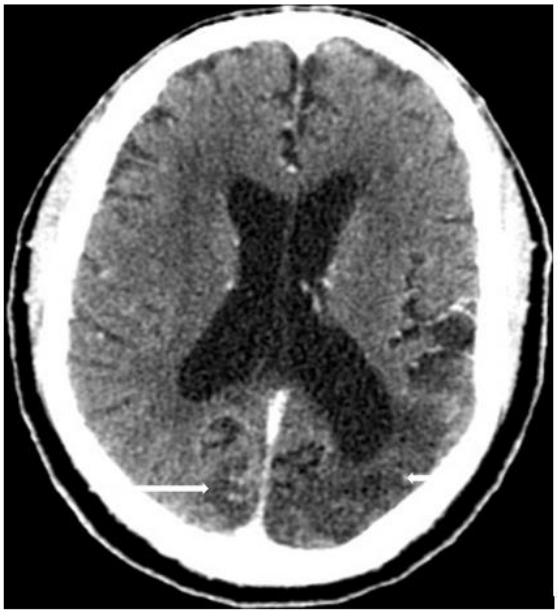

**Figure 1:** Non-contrast Axial CT scan in parenchymal window showing: bilateral occipital and subcortical hypodensity, left-most dominant with triventricular dilation **(white arrow)**

Computed tomographic angiography scans of supra-aortic trunk revealed endoluminal lacuna confirming the stenosis of the basilar trunk **(Figure 2).** The patient was transferred to the neurology department for management.


**Figure 2:** CT angiography of supra-aortic trunks in sagittal reconstruction: endoluminal lacuna in the union of vertebral arteries **(white arrow)**

## DISCUSSION

### Neurological picture

In CB, it is not a massive decline in visual acuity, but a loss of conscious visual sensation throughout the visual field. Even the rudimentary discrimination of light and darkness, movement or immobility are absent. It is still a blindness, since legally, blindness is any person whose best corrected visual acuity is lower than 3/60 and whose field of view is lower than 10° around the fixation point according to the World Health Organization (WHO) [6]. This semiological pattern was present in our patient, which made our diagnostic procedure easy.

CB can be sudden or progressive, the loss of the vision occurring in the latter case in an individual that already had a lateral homonymous hemianopsia; that is, an impairment of the contralateral visual field; to the unilateral occipital lesion [1]. Our patient is clearly in the latter situation, as the results of the brain Computed Tomographic (CT) scan revealed ischemic lesions on the posterior cerebral arteries zones at the chronic stage on the left occipital lobe and at the sub-acute stage on the right occipital lobe.

### Associated signs

Surprisingly, as it may seem, the patient with CB does not spontaneously complain of visual loss, vigorously denies his discomfort when he is defeated, and sometimes attributes to external causes his incapacity to perform certain tasks involving vision; that is anosognosia [1]. Anosognosia results in delayed diagnosis. Actually in our case, the patient never complained about visual difficulties, he was brought by his son to emergency who

had discovered his inability to see well around himself. This pattern known as visual anosognosia is feature of Anton's syndrome in cortical blindness where a person cannot see but always denies the blindness. These individuals often try to walk through the closed door or wall, and in the process of denial, they take the help of confabulation. It occurs due to lesions in the V1 (the primary visual cortex) [3, 7].

The CB can be associated with more or less severe variety of neuro-psychological disorders: memory disorders with temporo-spacial disorientation, confusions, visual hallucinations, neurological signs in keeping with the cortical attack (hemiplegia, sensory disorders, aphasic disorders) [7,8]. Our patient presented personality disorders like that, this would explain the lack of complaint during visual disturbances caused by the left occipital lesion which was the first to settle as brain CT scan suggests.

Confusion and personality disorders are the reasons why providers sometimes refer as first-line patients to a psychiatric center before the neurological diagnosis. It is noted that cortical blindness is sometimes diagnosed incorrectly. This misdiagnosis is even more dramatic in children, in whom cortical blindness may go undetected for decades, or even never diagnosed at all. [9, 10]. This difficulty did not arise in our case, as our patient was lucid and had been easily examined. Our patient was admitted to a neurology department and no other neurological disorders were noted. Nevertheless, we had to clinically rule out other probable diagnoses.

Peripheral blindness due to bilateral involvement of the pregeniculate visual pathways was easily eliminated by the normal pupillary function. The abolition of optokinetic nystagmus confirms the cortical site of the lesion and eliminates blindness of psychogenic origin where it is preserved. Similarly, the abolition of the blinki reflex in response to a threat or light was also used to eliminate malingering.

**Paraclinical investigations**

If the electroencephalogram (EEG) can locate a focus of suffering at the occipital level, it does not allow making the diagnosis of cortical blindness infallible [11]. Moreover, the EEG can be perfectly normal in patients with cortical blindness; Michael et al. [12] in 25 cases of CB five had normal EEG. In our case report, the EEG was normal.

Advances in neuroimaging have considerably changed the diagnosis and monitoring of CB.CT scan of the brain is the initial investigation because of its easy availability, but CT scan can miss an early stroke and small strokes. MRI (Magnetic Resonance Imaging) with gadolinium injection is superior to the CT of the brain in diagnosing stroke even at a distance from the lesion, but it is not easily available in all health care facilities[1,3].Other complementary investigations have identified risk factors to prevent further strokes.

**Etiologies and risk factors**

The risk factor associated with our case was hypertension which evolved since 15 years ago. Abalo-Lojo et al. [13] reported a 66-year-old patient with cortical blindness under background of malignant hypertension at 200/176 mm Hg. That patient urgently presented with severe frontal headaches and progressive visual loss. The lesion found on Magnetic Resonance Imaging was leukoencephalopathy in occipital cortex. The emergency treatment of hypertension had allowed recovery of vision without sequelae. Michael et al. [12], in their study, we observed that cerebrovascular diseases and cardiovascular surgeries were the most common cause found in cortical blindness. Shugang Cao et al. [14], reported a rare case of Anton's syndrome as a presentation of Trousseau syndrome involving the bilateral optic radiation. The patient had been diagnosed with gallbladder cancer 2 months previously, and he was admitted to the hospital with confusion and quadriplegia. Nicolas Burra et al. [15], in their clinical case have described a stroke occurring first to the left and creating a lateral homonymous hemianopsia , followed on the right side in the same patient, thus giving the characteristic of a bilateral CB, a case similar to ours. Kurtz et al. [16] reported nine cases of cortical blindness in whom seven were vascular and 2 had septic emboli. Michael et al. [17] found primary causes of cerebrovascular accidents in primary visual areas, and concluded that the incidence of cortical blindness would logically increase with the aging of the population. The best visual prognosis is associated with a young age for example patients less than 40 years old with no history of vascular diseases such as hypertension or diabetes mellitus [10]. This explains the poor vision observed in our advanced age patient.

**Evolution and prognosis**

Exceptionally, CB can persist completely. Most often there is a total or partial recovery [5]. This recovery can be limited to elementary visual functions (light, movement, colors). The visual field can recover with a concentric narrowing of the visual field corresponding to a macular sparing or in a fragmentary way with central or Para-central scotomas. After 13 months follow-up, no visual improvement was observed in our patient.

## CONCLUSION

The diagnosis of cortical blindness is easy in the absence of associated neuro-psychological symptoms. But the existence of these can compromise this diagnosis and its proper care.

To date there is no standardized and systematic treatment of cortical blindness even in developed countries. In our still medically limited countries, prevention, which involves early and adequate treatment of risk factors for cortical blindness, remains the most dissuasive weapon in this rare but feared pathology.

## DECLARATIONS

**Ethical statement and informed consent.** The patient has given an oral consent and written informed consent to publish the case was provided by the patient's son.

**Conflict of interest statement.** The authors declare that there is no conflict of interest**.**

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