

ANTON BABINSKI SYNDROME - A Rare Complication of Cortical Blindness

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ABSTRACT

In this case report, we present a rare complication of cortical blindness which is Anton Babinski syndrome. Patients with this syndrome usually disaffirm their blindness and confabulate to fill in missing visual information. The commonest etiology of cortical blindness is posterior cerebral artery infarction but in this particular report we present a case of hypertensive patient with Anton Babinski syndrome who developed bilateral cerebral hematoma confined to parieto-occiput area.

Keywords: Anton Babinski Syndrome, cortical blindness

CASE PRESENTATION

A 45 year old gentleman presented to Emergency department with complains of severe headache and blurring of vision. He was unable to walk and he was brought by his relative to ER. He could not remember what happened during the day and he was confused. No history of weakness and sensory symptoms. There was no urinary incontinence. He is known hypertensive and is on atenolol 50 mg once daily. He is not under regular follow-up with his physician. He is a barber by occupation and smokes around 3-4 cigarettes per day. No history of alcohol or drug intake.

On examination he was conscious, oriented with time, place and person but was not able to maintain eye contact. The blood pressure was 210/120 mmHg and pulse was 62/minutes and regular. He maintained that he could see our blurred faces. But on visual field examination his visual field was markedly decreased with absence of perception of torch light. Pupils were dilated and reactive to light. Fundus examination appeared normal. No sensory or motor deficit were present and both superficial and deep reflexes were intact. Other systemic examination was unremarkable.

He was given amlodipine and perindopril to lower down the blood pressure. CT brain was done which revealed bilateral occipital fresh intracerebral hematoma (Figure 1). He was initiated on mannitol and phenytoin. After neuro-surgical opinion it was decided to manage him conservatively. On day 2 his headache had reduced and vision improved slightly with perception of lights and abnormal finger counting. His blood pressure was gradually reduced to 160/80 mmHg over a period of 3 days. MRA was done which showed right parieto-occipital and small left occipital acute to sub-acute hematoma. Multiple scattered cerebral, pontine and cerebellar petechial hemorrhage was observed (figure 2,3). He was discharged after one week of hospital stay. At the time of discharge his visual acuity was 6/12 in both eyes. He wanted to go his home country for further treatment. The case study points towards diagnosis of Anton syndrome where

the patient denies blindness. In our case we did not tell his relative about his loss of vision but explained that he can see although it's blurred.



Figure 1: CT Brain showed bilateral occipital fresh intracerebral hematoma

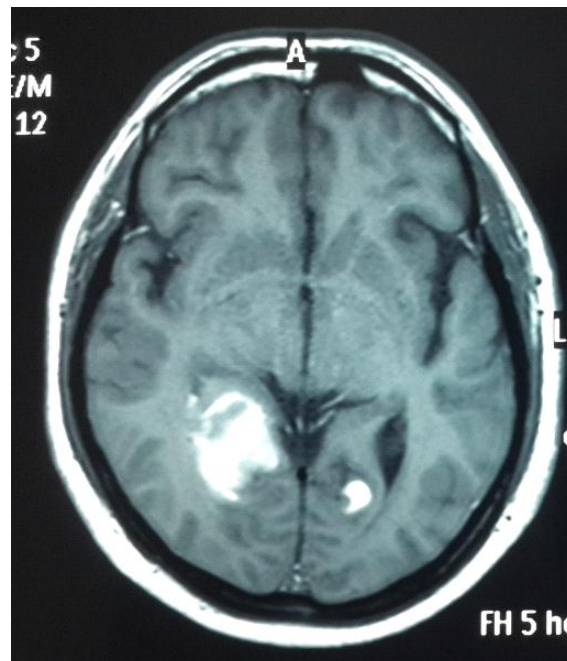


Figure 2: MRA showed right parieto-occipital and small left occipital acute to sub-acute hematoma.

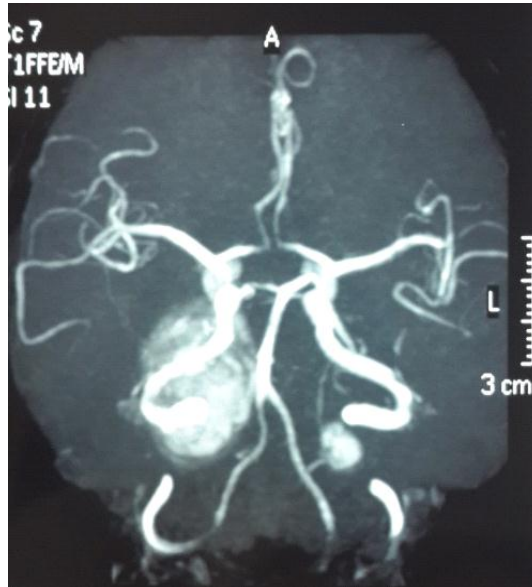


Figure 3: MRA showing no evidence of intracranial aneurysm

INTRODUCTION

Cortical blindness is one of the rare neurological condition characterized by binocular vision loss due to insult in the occipital cortex. Pupillary light reflex are preserved as anterior visual pathway is intact. One of the rare complication of cortical blindness is Anton Babinski Syndrome. This syndrome results from damage to visual association cortex (brodmann area 18, 19) along with primary visual cortex (brodmann area 17)^{1, 2}.

DISCUSSION

Anton Babinski syndrome is a condition in which patient disaffirm their blindness (visual anosognosia) inspite of complete amaurosis of central origin (cortical blindness). In this, patient adamantly claim they are capable of seeing and/ or experiencing strange visual hallucinatory episodes, consequently resulting in confabulation¹.

This syndrome was first described by French writer Michel de Montaigne (1533-1592). In late 1890s Gabriel Anton (1858-1933), Austrian neuro-psychiatrist noted three patients, one with cortical blindness one with deafness and one with left sided hemiparesis all lacking self-perception of their deficit. Joseph Babinski (1857-1932), French neurologist in 1914 used the term 'Anosognosia' to describe hemiplegic patients with unawareness of their blindness^{1, 3}.

Patients with Anton syndrome often find excuses for their symptoms such as; there is not enough light to see or they may also put themselves in danger to prove their intact vision⁴. These patients act as though they could see, and in attempting to walk, collide with objects, even to the point of injury. They may offer excuses for the difficulties—"I lost my glasses," "The light is dim"—or may only evince indifference to loss of sight. The lesions in cases of negation of blindness extend beyond the striate cortex to involve the visual association areas⁵.

There are diverse causes of Anton syndrome, most common being bilateral occipital lobe infarction of posterior cerebral artery⁶. Other cause being obstetric complication such as pre-eclampsia and hemorrhage resulting in hypo-perfusion⁶,

Hypertensive encephalopathy⁸, MELAS (Mitochondrial myopathy, Encephalomyopathy, Lactic Acidosis and Stroke like episodes)⁹ Adrenoleukodystrophy¹⁰, cardiac surgery¹¹, head trauma¹², Angiitis (auto-immune) of central nervous system¹³. Angiographic procedures which disrupts blood brain barrier, coexisting with hypotension, embolism and vasospasm can also cause cortical blindness¹⁴. Any condition resulting in systemic hypoxia mainly affects occipital cortex because it lies distal from central cerebral vasculature¹⁵.

Why only some patients with cortical blindness, end up having Anton Babinski syndrome, is still unknown. But there are various hypotheses postulated, one stating that along with primary visual cortex (brodmann area 17), visual association cortex (brodmann area 18, 19) is also compromised. Damage of visual association area, results in disconnection with speech and language area^{16,17}. Thus, functional speech area confabulate due to missing visual input. This was explained by Dr. G. Goldenberg and colleagues in 1995. Other hypothesis states this syndrome as hallucinatory sensation.

It is diagnosed by excluding anterior visual pathway disease by proper history taking along with complete neuro- ophthalmological examination. There are five criteria for clinical diagnosis of cortical blindness, a) loss of blink reflex in response to threat b) loss of all visual impulse including light and dark c) preserved light and accommodation-pupillary reflex d) normal fundoscopic examination e) preserved extra- ocular movements [4]. Once clinically diagnosed laboratory and radiological investigation are carried out to confirm the etiology of cortical blindness.

In this patient Computed tomography of brain revealed bilateral occipital fresh intracerebral hematoma. MRA was done further, which showed right parieto-occipital and small left occipital acute to sub-acute hematoma. Multiple scattered cerebral, pontine and cerebellar petechial hemorrhage was observed.

The outcome of cortical blindness solely depend on age of patient, etiology, severity, duration, initial recovery time, medical history. This patient was managed conservatively by lowering down blood pressure. Medicines used to lower blood pressure were amlodipine, perindopril and mannitol.

CONCLUSION

Anton Babinski syndrome should always be suspected in patient with evident occipital lobe damage presenting with denial of their blindness. Emphasis should be made on secondary prevention and rehabilitation of this syndrome. Absence of focal neurological deficits combined with denial of blindness makes the diagnosis challenging like the above case report.

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A comparison of in-house Hemoglobin DNA Mutation analysis for β thalassemia by ARMS PCR with a Commercial Line Probe Assay

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ABSTRACT

This study was done with a view to find a correlation between two molecular tests for β Thalassemia, our in-house developed haemoglobin DNA mutation analysis using ARMS PCR and a Commercial Line Probe Assay. De-identified samples from known β thalassemia patients characterised by HPLC for HbA₂, Peripheral smear (Target cells) and CBC (microcytosis and erythrocytosis) were used for the study. DNA was extracted using the DTAB/CTAB method. Amplified DNA from the samples was hybridised for mutations using a line probe assay. The extracted DNA was also examined for wild type genes and mutant