ORIGINAL RESEARCH PAPER

Volume-7 | Issue-2 | February-2018 | PRINT ISSN No 2277 - 8179

INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

"A RARE CASE OF ANTON'S SYNDROME"

General Medicine

Dr. Vasista Palnati Post Graduate, Sri Ramachandra University, Chennai.

Dr. Mohini Singh* Associate Professor, Sri Ramachandra University, Chennai. *Corresponding Author

Dr. S. R. Ramakrishnan Professor, Sri Ramachandra University, Chennai.

ABSTRACT

Introduction: Anton's Babinski syndrome is an extension of cortical blindness, a rare neurological condition in which the patients are unaware and deny their objective evidence of visual loss. On the contrary, in cortical blindness, patient is aware of his blindness and does not deny it. Bilateral occipital lobe brain damage results in blindness and patients starts to confabulate to fill in the missing sensory input.

Case Presentation: In this case report we present a case an Indian male, with cortical blindness and Anton's syndrome. Despite his obvious blindness, which is evident from his history of walking into walls and hitting into objects, patient denies visual loss and demonstrated confabulation in his accounts of surroundings, consistent with the diagnosis of Anton's syndrome.

Conclusion: A suspicion of cortical blindness and Anton's syndrome should be considered in patients with atypical visual loss and with evidence of occipital lobe injury. Here, we describe the causes and major clinical manifestation of this syndrome and review the current medical literature. Any condition that may result in bilateral occipital lobe damage can lead to Anton's syndrome. Recovery of visual function will depend on the etiology. Management of this condition should accordingly focus on secondary prevention and rehabilitation.

KEYWORDS

Anton's syndrome, Cortical blindness, Confabulation, Occipital lobes.

INTRODUCTION:

Visual anosognosia, or denial of loss of vision, associated with confabulation with presence of obvious visual loss and cortical blindness is known as Anton's syndrome. Originally, the syndrome is named by Gabriel Anton, who described patients with objective blindness and deafness but showing lack of self-perception of their deficit. Later Joseph Babinski used the term anosognosia to describe this phenomenon. Bilateral occipital brain damage results in cortical blindness. But the patients start to confabulate to fill in the missing sensory input. The reason for their denial is not known, but many theories has been proposed. Although visual anosognosia is frequently believed to represent cortical phenomenon, it is probably more often caused by parietal white matter injury leading to a disconnection syndrome. In this paper, we present a case of patient with Anton's syndrome due to bilateral occipital ischemic lesions due to cerebrovascular accident.

CASE PRESENTATION:

A 56-year-old Indian male, presented to SRMC OPD, with history of decreased vision for 2 days as noticed by his wife. He was a known hypertensive, diabetic, old CVA with residual right hemiparesis and takes regular medications for the same. Patient's wife gives history that the patient had visual defects in the form of walking into walls and hitting into objects. The patient was not aware of the visual loss. In particular, the visual loss was observed for the first time as the patient stumbled when he was asked to sit in a chair. On asking to reach for a glass of water, he was searching for the glass in the wrong direction. There is no history or complaints of headache, vomiting, blurring of vision, trauma and seizures.

On examination patient was conscious, coherent, oriented, moderately built and nourished. Patient was not maintaining eye contact with the examiner. On examination of the central nervous system, Glasgow coma scale was 15/15 and higher mental functions were intact. Visual acuity was 3/6 bilaterally. Pupils were bilaterally equal and reacting to light. Extra Occular Movements were intact and there was no ptosis or nystagmus. Blink reflex was absent bilaterally. Corneal reflex was intact. Residual right hemiparesis was present. All routine laboratory investigations were normal. MRI brain, which was done to rule out any acute cerebrovascular event, showed acute non hemorrhagic infarct in bilateral occipital region, chronic infarct with gliotic changes involving left temporal and parieto occipital region and chronic lacunar infarcts in pons and right corona radiata region. MR Angiogram showed severe occlusion of left middle cerebral artery. Carotid-vertebral artery Doppler, done to rule out stenosis of the arteries, was normal. Visual Evoked Potential test showed bilateral anterior visual pathway defect due to bilateral occipital region infarct. Fundoscopy was unremarkable. Patient was treated with clopidogrel, antihypertensive, antidiabetic and statin drugs. Physiotherapy was given for residual hemiparesis. The patient was regularly followed up on an outpatient basis. The patient's neurological symptoms were static, but the visual impairment partially improved, as evident from a visual acuty of 4/6 when examined, during the course of follow up.

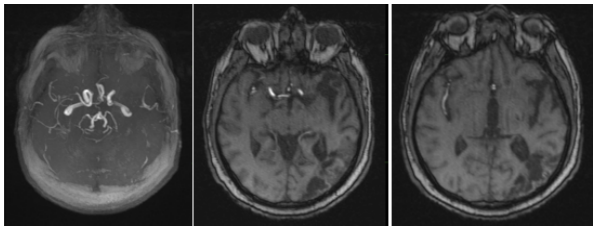

Figure 1a, 1b & 1c: MR Angiogram images revealing occluded Left Middle Cerebral Artery.

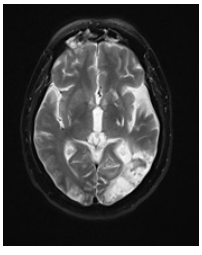

Figure 2: Selective MRI axial image of the brain showing acute non-hemorrhagic infarct in the bilateral occipital lobes (right > left) and left parieto-occipital region.

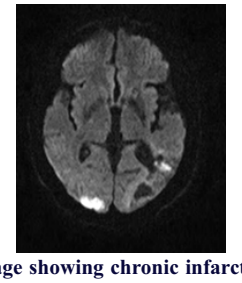

Figure 3: MRI image showing chronic infarct with gliosis in the parieto-occipital region and the left occipital lobe.

DISCUSSION:

The French renaissance writer Montaigne (1533-1592), described in his second book of *Les Essais*, a case of a nobleman, who did not believe he was blind, despite his blindness.(1) This was the first ever description of not perceiving one's own blindness in the absence of psychiatric illness or underlying cognitive impairment.

A few hundred years later, an Austrian neuropsychiatrist Gabriel Anton (1858-1933) described patients with objective blindness and deafness, with a lack of perception of their impairment.(2) Joseph François Babinski (1857-1932) later used the term anosognosia to describe this phenomenon.(3)

Confabulation is viewed as a memory disturbance characterized by verbal statements or actions that do not accurately reflect the facts and evidence.(4) Some have described it as "honest lying" because the person usually has no intention of deceiving the people they are talking to and are usually unaware that the things they are saying are false. Whereas, anosognosia is unawareness of severe bodily impairment or disability.(5)

Cortical blindness refers to loss of vision caused by bilateral occipital lobe infarcts (supplied by the posterior cerebral arteries), with presence of intact anterior visual pathway. It matches the following clinical criteria: loss of all visual sensations, including the perception of light; loss of menace reflex; preservation of light and accommodation pupillary reflexes; a normal fundoscopic examination and preservation of ocular movements.(6)

Anton syndrome (visual anosognosia) is a rare complication of cortical blindness, usually involving both the primary visual cortex (bilateral occipital lobes) and visual association area (areas of temporal & parietal lobes). The ability to recognize visually presented objects and words depends on the integrity not only of the visual pathways and primary visual area of the cerebral cortex (area 17 of Brodmann) but also of those cortical areas that lie just anterior to area 17 that is areas 18 and 19 of the occipital lobe and area 39 - the angular gyrus of the dominant hemisphere (visual association areas).

Anton's syndrome is the denial of loss of vision (visual anosognosia), associated with confabulation, in the presence of obvious vision loss and cortical blindness. Usually patients with lesion in the occipital lobes bilaterally, also have damage to their visual association cortex, which may account for their lack of awareness. (7) Also, damaged visual areas are effectively disconnected from functioning areas, such as speech and language areas. In the absence of input, functioning speech areas often confabulate a response.

Theories explaining Anton Babinski Syndrome: 1) Disconnection phenomenon: Conscious Awareness System (CAS) which is located on the parietal lobes, monitors all the information received from the senses. This system connects with the frontal lobes, which integrates the information, in order to perform complex cognitive tasks.(8) In Anton-Babinski syndrome, damage of association pathways between visual cortex and CAS would be responsible for the lack of awareness of the visual deficit. Additionally, as suggested by Anton, damaged visual areas are effectively disconnected from functioning areas, such as speech-language areas. In the absence of input, functioning speech areas often confabulate a response.

2) Neuropsychological mechanism: the signal transmission to the visual monitor (located on the visual association cortex) comes from a secondary visual system, located on the superior colliculus, pulvinar and temporo-parietal regions. In the absence of transmission on the geniculocalcarine pathway, this secondary visual pathway would allow blind patients to confabulate i.e. in the absence of visual input, this false internal imagery may convince the monitor or speech areas to come out with a false response. Denial of blindness could be related to memory loss or confusion following the infarct.(6)

Our patient with bilateral occipital infarcts causing cortical blindness and visual anosognosia, fulfilled the description for Anton's syndrome. He strongly believed that his vision is normal despite his obvious deficit. Cause for Anton's syndrome in our patient was bilateral acute occipital lobe infarcts, with additional involvement of left temporal and parieto-occipital regions (thus leading to damage of the Visual Association Pathway). Most common cause of Anton's syndrome is ischemic cerebrovascular accident involving the occipital lobe, as in our case. The other causes leading to Anton syndrome are "MELAS [Mitochondrial encephalomyopathy, lactic acidosis, and stroke-like episodes], Preeclampsia, Obstetric hemorrhage, Trauma, Adrenloeucodistrophy, Hypertensive encephalopathy and Angiographic procedures. (9)

Correction of the causative factor may lead to resolution of symptoms. Better visual outcome was observed in (i) young patient (<40 years old), (ii) no history of hypertension and diabetes, (iii) no cognitive, language, or memory impairment, and (iv) CVA is not the causative factor & (v) short duration of symptoms. Treatment should also be aimed at secondary prevention, and to offer rehabilitation. Recovery of visual loss mainly depends on the etiology. Occipital lobe infarcts after cerebrovascular accidents are less likely to improve.

CONCLUSION:

A suspicion of cortical blindness and Anton's syndrome should be considered when the patient has denial of blindness, with atypical visual loss and with evidence of occipital lobe injury. The most common cause of Anton's syndrome is cerebrovascular accident. However, any condition that may result in bilateral occipital lobe damage can lead to Anton's syndrome. Recovery of visual function will depend on the etiology. Visual loss due to occipital lobe infarction after cerebrovascular disease is less likely to recover completely. Management of this condition should accordingly focus on secondary prevention and rehabilitation. Our case adds on to the limited references for Anton syndrome.

REFERENCES:

1. de Montaigne M. In: Book 2, Chapter 12. Langelier A, editor. 1595.
2. Anton G. Über die Selbstwahrnehmung der Herderkrankungen des Gehirns durch den Kranken bei Rindenblindheit und Rindentaubheit. Arch Psychiatrie Nervenkrankh. 1899;32:86–127. doi: 10.1007/BF02126945.
3. Babinski J. Contribution a l'étude des troubles mentaux dans l'hémiplégie organique (anosognosie) Revue Neurol. 1914;27:845–848.
4. Mechanisms of spontaneous confabulations: a strategic retrieval account. Gilboa A, Alain C, Stuss DT, Melo B, Miller S, Moscovitch M, Brain. 2006 Jun; 129(Pt 6):1399-414.
5. M. S. Aldrich, A. G. Alessi, R. W. Beck, and S. Gilman, "Cortical blindness: etiology, diagnosis, and prognosis," Annals of Neurology, vol. 21, no. 2, pp. 149–158, 1987
6. M. Maddula, S. Lutton, and B. Keegan, "Anton's syndrome due to cerebrovascular disease: a case report," Journal of Medical Case Reports, vol. 3, article 9028, 2009
7. Visual Anosognosia (Anton-Babinski Syndrome): Report of Two Cases Associated with Ischemic Cerebrovascular Disease: Juan José Romero Carvajal, Augusto Alejandro Arias Cárdenas, Germán Zamora Pazmiño, Patricio Abad Herrera.
8. Bilateral Occipital Infarction Presenting as Anton-Babinski Syndrome and Charles Bonnet Syndrome Nii Kwanchie Ankrah, Yongxing Zhou, Roger Weir and Annapurni Jayam-Trouth.
9. Cortical Blindness: Etiology, Diagnosis, and Prognosis : Michael S. Aldrich, MD," Anthony G. Alessi, MD," Roy W. Beck, MD,and Sid Gilman, MD.
10. Anton's Syndrome and Cortical Blindness: gadwalkar srIkant r, deePadv,ramam u r t h y, ravIdhar.
11. Anton's Syndrome due to Bilateral Ischemic Occipital Lobe Strokes: Sanela Zuki T, Osman Sinanovi T, Lejla Zoni T, Renata Hod Di T, Svjetlana Mujag iT, andEdina Smajlovi T.
12. Anton Babinski Syndome - A Rare Complication of Cortical Blindness: Mohammed Khalid, Mohamed Hamdy, Himanshi singh, Kiran Kumar, Altaf Basha.