ORIGINAL SCIENTIFIC ARTICLES

Sight Lost, Insight Kept: Cortical Blindness Without Visual Anosognosia after Bilateral Occipital Infarcts: a Case Report

John Lorenze C. Datinguinoo, MD[a] , Vicente G. Rosales, Jr., MD, FPNA, FPPA[a] and Johnny K. Lokin, MD, FPNA[a, b]

ABSTRACT

Cortical blindness, a consequence of bilateral occipital lobe lesions, typically manifests with partial or complete visual loss. It is often associated with the intriguing phenomenon of visual anosognosia, wherein patients paradoxically deny their profound visual loss, which would often lead to confabulation. This constellation of clinical findings points to Anton's Syndrome. Bilateral occipital infarcts are the most common cause of cortical blindness, and the co-occurrence of visual anosognosia is frequently reported in these cases. We present a unique case of a 65-year-old right- handed Filipino male who experienced sudden, acute cortical blindness resulting from simultaneous bilateral occipital infarcts of likely cardioembolic origin secondary to atrial fibrillation. Despite the cortical blindness, the patient explicitly acknowledged his blindness and was not demonstrating signs of denial or confabulation. The patient's neurological examination was otherwise notable only for the visual impairment, without other focal deficits. This clinical presentation stands in contrast to the typical features of Anton's syndrome. Cranial magnetic resonance imaging revealed acute infarcts in both occipital lobes and the right pons. This unusual presentation underscores the heterogeneity in the clinical expression of posterior circulation strokes. This case contributes valuable insights into the complex neural pathways involved between visual information processing, its awareness, and speech production following bilateral occipital infarction. This is the Philippines' unprecedented case of bilateral visual loss after simultaneous acute bilateral occipital infarcts occurring without accompanying visual anosognosia.

Keywords: *Bilateral Visual Loss, Bilateral Cortical Blindness, Bilateral Occipital Stroke, Cardioembolic Stroke, Stroke, Anton's Syndrome, Case Report*

INTRODUCTION

Neurological visual impairment refers to visual disturbance as a result of injury to the brain itself rather than eye abnormalities. This encompasses a broad spectrum of conditions, including cerebral visual impairment, visual neglect, visual agnosia, lack of facial recognition, and cortical blindness.[1] Cortical blindness refers to the partial or total loss of vision in the presence of normal pupillary light reflexes, and in the absence of extraocular movement abnormality and funduscopic pathology, secondary to bilateral lesions in the occipital lobes.[2] The pupils remain reactive to light because all elements of the pupillary light reflex remain intact. Bilateral posterior cerebral artery occlusions or trauma to the bilateral occipital lobes may produce cortical blindness. Other causes include severe hypoxic-ischemic brain injury, hypertensive encephalopathy with

preeclampsia, trauma of the occipital region, migraine, occipital seizures, and multiple sclerosis.[3,4] Nevertheless, the most common cause of cortical blindness is bilateral occipital lobe infarctions in the vascular territory of the posterior cerebral arteries.

Stroke can be classified either as ischemic or hemorrhagic. Ischemic strokes are usually secondary to atherothrombotic or cardioembolic causes. The most common source of infarction of the posterior circulation is cardiac embolism.[6] Anatomically, the occipital cortex has a dual blood supply from both the posterior cerebral arteries and the superior tempo-occipital sylvian artery from the middle cerebral arteries. Moreover, the temporo-occipital lobe white matter connections are supplied by the posterior cerebral arteries. However, the most common cause of cortical blindness is caused by the occlusion of one or both of the posterior cerebral arteries. According to the study of Ziaul et al. (2024), only 5-10% of strokes are represented by a posterior cerebral artery infarction.[7]

Anton's syndrome refers to visual anosognosia or the denial of loss of vision associated with confabulation in the setting of obvious visual loss and cortical blindness. Confabulation is defined as the emergence of memories of events and experiences that never took place. Due to the phenomenon of confabulation, patients would usually describe their surroundings and would deny that they could not see. Anton's syndrome is usually caused by infarction of the bilateral occipital lobes.Kondziella,& Frahm-Falkenberg (2011) described possible mechanisms leading to visual anosognosia, which they pertain to as disconnection syndrome.[8] Visual anosognosia is the condition where the patient would have cortical blindness, but is fully unaware or would even deny their blindness. They postulated that visual anosognosia is more often caused by parietal white-matter injury. As a result, some areas of the parietal cortex that integrate visual sensory information are separatedfrom inter- and intrahemispheric association pathways. Maddula et al. also supported this and further stated that the visual anosognosia could be secondary to damage to the visual association cortex and speech-language areas.[1]

In the absence of inputs from these areas, dysfunctional speech areas would result in confabulation. Genetics may also play a role as a predisposing factor in the development of visual anosognosia after bilateral occipital infarct. In the study of sen Tanrikulu et al. (2016), MTHFR gene mutation was associated with the risk for the development of visual anosognosia.[4] In the study of Atallah et al. (2024), 90.3% of patients with Anton syndrome would present with visual anosognosia.[8] Furthermore, they stated that since 1965, only 28 cases of Anton Syndrome have been formally reported. To date, there has not been a published journal article in the Philippines. This is the first reported case with bilateral cortical blindness due to simultaneous bilateral occipital infarcts but without visual anosognosia in the Philippines.

CASE PRESENTATION

A right-handed 65-year old male arrived at the emergency room of our institution with a chief complaint of sudden bilateral vision loss 5 hours before the consultation. The patient was driving alone from home towards a restaurant to buy food when he noticed a sudden blurring of vision in both eyes. No noted focal neurologic deficit of weakness, numbness, slurring of speech, nor facial drooping. The patient also did not report headache, nausea, or vomiting. No difficulty breathing, chest pain, nor chest heaviness were noted. He stopped the car and immediately called his son to pick him up. When the son arrived and saw him, he did not observe any slurring of speech, facial asymmetry, nor focal weakness. They decided to bring the patient to the emergency room immediately. No behavioral or cognitive changes were noted. No visual anosognosia was observed.

Past Medical History

The patient had an episode of transient ischemic attack in 2017, which presented as sudden right-sided weakness. He noted that he had difficulty in ambulation, with dragging of his right leg. No slurring of speech was noted, nor was there any facial asymmetry. There was a resolution of the symptoms even before he was brought to the hospital. He was then admitted for only three days. He also had a prior history of myocardial infarctions in 2019 and 2022, whereby both presented with gradually worsening chest heaviness, with associated easy fatigability and orthopnea. He underwent cardiac angiography with angioplasty and stent placement for both myocardial infarctions. He was also hypertensive and diabetic. He claimed to be compliant with his maintenance medications, including his antihypertensives, anticoagulant, and other cardiovascular medications.

Family History, and Personal and Social History

There is a family history of hypertension and diabetes on both maternal and paternal sides. His mother also had a stroke. He was a previous cigarette smoker with 22 pack years. He claimed to be a non-alcoholic beverage drinker and denied illicit drug use.

Physical Examination and Neurologic Examination upon Admission

He was referred to the neurology service. Upon referral, his vital signs showed blood pressure of 120/90 mmHg, heart rate of 69 bpm with irregular rate, temperature of 36.6 °C, respiratory rate of 15 cpm, and spO2 of 95% at room air. Neurologic examination showed that he was oriented to three spheres and able to follow commands. His pupils were isocoric and both equally responsive to light. There was no noted deficit in all cardinal gazes. Pursuit eye movements could not be evoked, and reflexes to visual threats and optokinetic nystagmus were absent. No facial asymmetry, impairment in gross hearing, or dysarthria were noted. The examination of the visual acuity revealed that he could only do counting fingers bilaterally. He had normal manual muscle testing and denied any sensory deficits on light touch, pain, vibration, and position sense. Deep tendon reflexes were normal. No pathologic reflexes were present.

Course in the ward

12L-ECG changes on admission showed atrial fibrillation with rapid ventricular response. No noted derangement on his metabolic work-ups. A Plain Cranial CT Scan was done, which showed hypodensities in bilateral occipital areas. A plain cranial MRI with MRA (Figure 1) was requested 24 hours after the initial Cranial CT scan, which showed areas and scattered foci of restricted diffusion seen as DWI hyperintensity and corresponding signal drop on the ADC map, are demonstrated in both occipital lobes and right side of the pons.

Rivaroxaban and his maintenance medications were continued. He was discharged as a case of Acute Infarcts, Bilateral Occipital, Right Pons, Left Cerebellar Hemisphere, with Left Occipital hemorrhagic conversion, Basilar artery in vascular territory, cardioembolic in origin secondary to Atrial Fibrillation in AVR, CHADSVASc7, HASBLED 3, NIHSS 2. His visual acuity upon discharge was 20/400 OS and 20/200 OD. Perimetry testing showed right homonymous superior quadrantanopia (Figure 2).

On follow-up after 1 month, the patient still has the visual acuity at the time of discharge. He was maintained on anticoagulant and was scheduled to follow up every 3 months.

DISCUSSION

This 65-year-old Filipino male patient presented with cortical blindness due to an acute bilateral occipital infarct of

Figure 1. Plain Cranial MRI with MRA showing the sequences with areas and scattered foci of restricted diffusion seen as DWI hyperintensity and corresponding signal drop on the ADC map are demonstrated in both occipital lobes

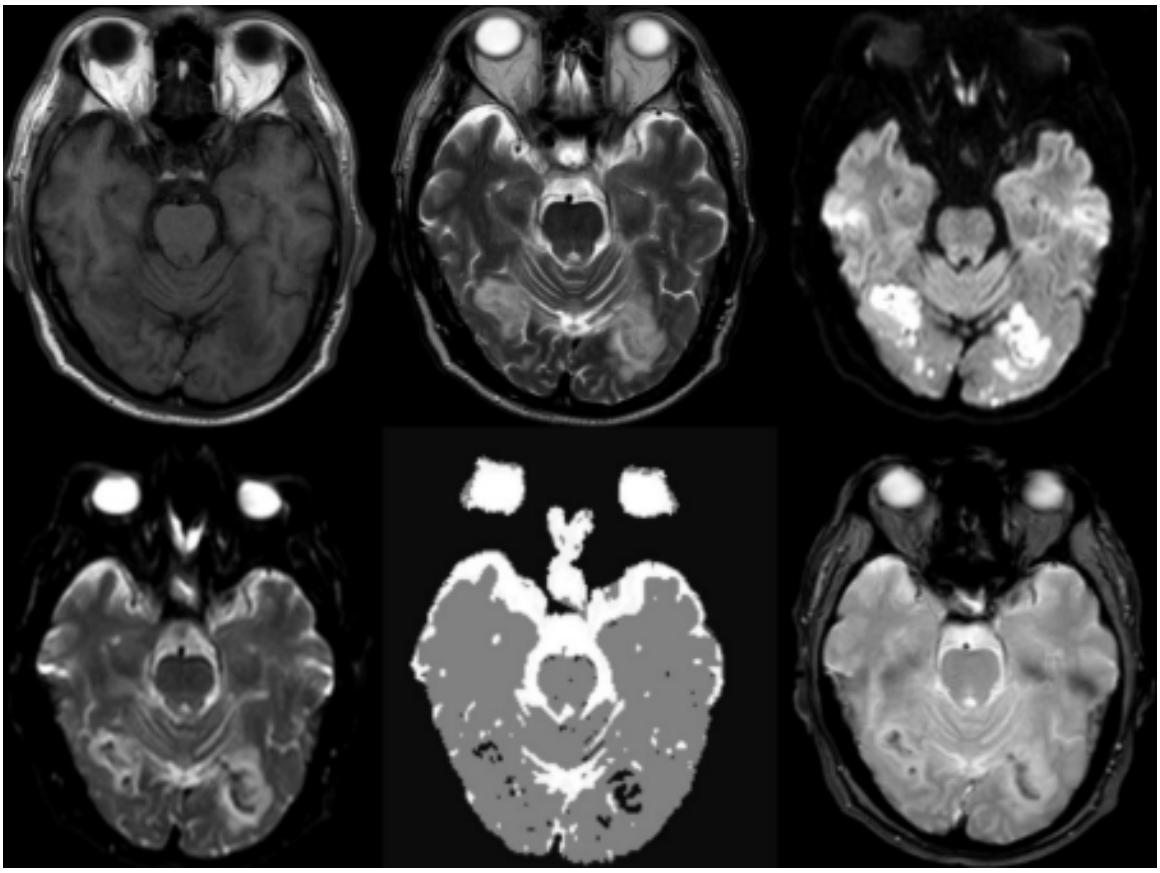
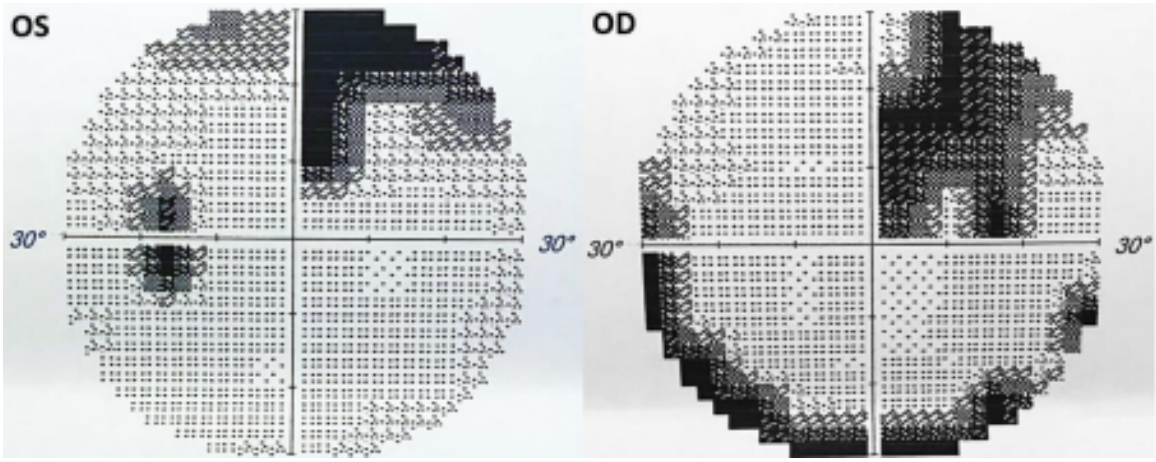


Figure 2. Perimetry of the patient showing right homonymous superior quadrantanopia (OS: Left eye; OD: Right eye)



cardioembolic origin secondary to atrial fibrillation but without visual anosognosia. This is a case of a rare presentation of Anton's syndrome wherein there is no visual anosognosia appreciated. Our patient has cardiac arrhythmia and a previous history of two episodes of myocardial infarction, predisposing him to cardioembolic stroke, which was the most common mechanism for bilateral occipital infarcts leading to cortical blindness, according to studies. Another important risk factor for the patient is also the history of a transient ischemic attack. It is well-established that a transient ischemic attack is a strong predictor of subsequent stroke.

As mentioned in the paper of Atallah et al., 90.3% of patients with bilateral occipital infarction would present with cortical blindness with associated visual anosognosia.[8] The majority of the published studies regarding Anton's syndrome are case reports and case series due to its rarity. Early published case series include the study of Satija et al. (1999), whereby all patients presented with the classical manifestation of Anton's syndrome.[10] The most recent published case report showing an atypical type of Anton's syndrome was the paper done by Kakaletsi et al. last 2016, wherein they described an 84-year-old man with cortical blindness but without visualanosognosia who had bilateral occipital infarct secondary to cardioembolic origin due to atrial fibrillation.[2] This case report of Kakaetsi et al. fits with the clinical presentation of our reported patient.[2]

Typically, patients with Anton's syndrome would deny their blindness due to the postulated disconnection phenomenon. This postulation states that visual anosognosia is due to the disconnection of the visual processing system and the centralized "conscious awareness system," which is localized in the inferior parietal lobes, which would convey inputs to the executive system found in the frontal lobe. Areas mediating speech production are also being disconnected from the damaged visual association cortex, which would explain the confabulation of these patients with bilateral occipital infarct. Maddula et al. (2009), in their paper, mentioned that patients who frequently have damage to the bilateral occipital lobes would also have concomitant damage to the visual association areas located on the occipital lobes,mwhich would account for the lack of awareness of these patients.[11] The visual association cortex plays a role in transmitting signals to the consciousness awareness system located in the bilateral parietal and frontal lobes.[12] The bilateral occipital damaged areas also become disconnected from functioning areas such as the speech-language areas. With the absence of inputs from the damaged occipital areas to the intact speech-language areas, the functioning areas would respond with confabulation. This is supported by the NOR-OCCIP study conducted by Tharaldsen et al. (2023), which concluded that higher infarct volumes led to more severe visual field defects and visual impairment.[13]

We presented a patient with an unusual presentation of cortical blindness after acute simultaneous bilateral occipital infarction, whereby no visual anosognosia was evident. This could be attributed to an intact visual association cortex and its connecting pathways. The exact location of the lesion and the extent of the infarcted area in the visual pathway could be important factors whether there would be accompanying visual anosognosia in a bilateral occipital infarct. For our patient, there is an intact visual association cortex and its intricate neural pathways involving the inferior parietal lobule, specifically the angular gyrus, and the white matter tracts such as the temporoparietal segment of the arcuate fasciculus and other multimodal association pathways. This would explain the atypical presentation of Anton's syndrome of our patient, wherein there is intact awareness of visual impairment and lack of confabulation despite the extent of infarcted occipital areas, which would make this case a

rare event. The affected and intact pathways involved in the typical and atypical presentation of Anton's Syndrome are presented in Figures 3 and 4.

In terms of neurorehabilitation, there is a higher degree of improvement of the visual defects in patients with larger occipital infarcts. Reported cases of bilateral occipital infarcts are mostly secondary to

Figure 3. Disconnection Phenomena showing the involved pathways in typical Anton's Syndrome

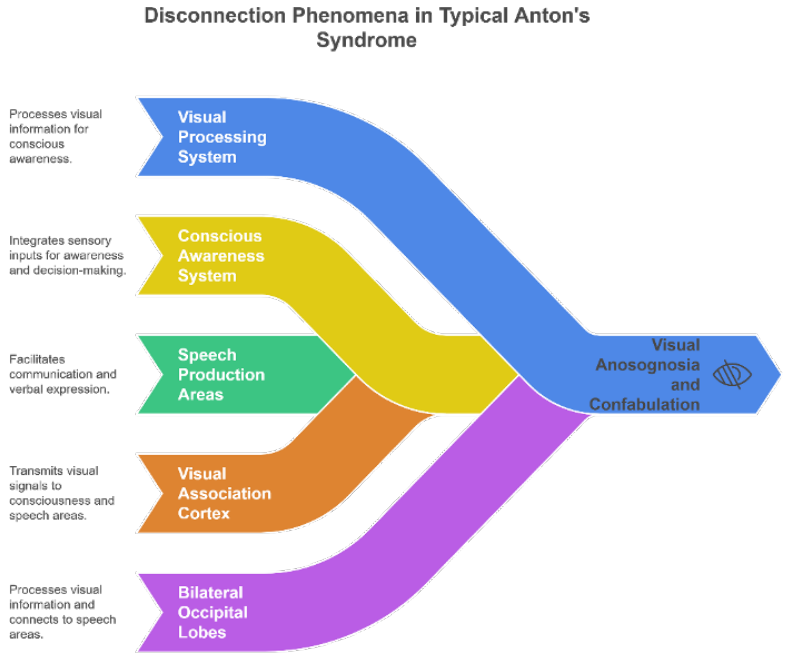
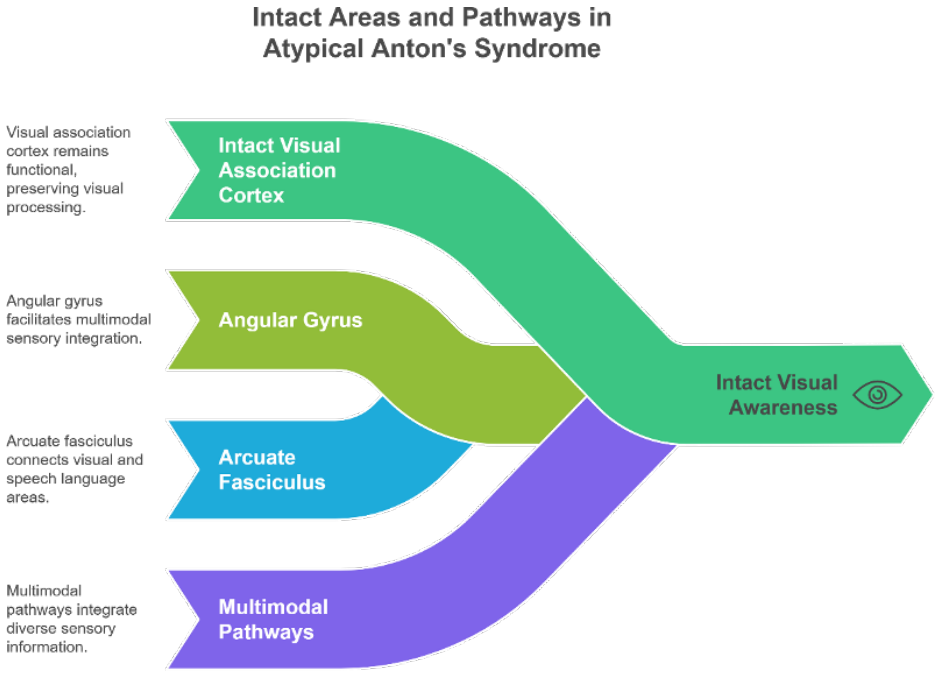


Figure 4. The intact pathways in the atypical presentation of Anton's Syndrome



cardioembolic cases. Hence, secondary stroke prevention management, such as blood pressure control, anticoagulants, and control of cardiac arrhythmia and other cardiac risk factors, is recommended.

CONCLUSION

This is an unprecedented case in the Philippines of bilateral visual loss without visual anosognosia after simultaneous acute bilateral occipital infarcts. This case report shows that stroke patients may present with only acute visual changes with no other focal neurologic deficits or any subjective complaints. Patients who present with acute vision loss and who have risk factors for cerebrovascular events should be thoroughly evaluated with a detailed history and proper neurologic examination, including visual examination.

In neurology, "time is brain." Late diagnosis of stroke could lead to increased morbidity and mortality. Therefore, in patients who are at high risk of developing stroke, even if the only sign is acute loss of vision, besides ocular pathologies, it is still prudent to consider occipital infarction.

RECOMMENDATION

Future studies may consider doing scoping reviews or systematic reviews discussing the possible risk factors or predictive variables for a patient to develop or not develop visual anosognosia after bilateral occipital infarct such as location of the infarct, volume of infarct, role of genetics, demographics of the population, posterior cerebrovascular anatomy, and medical comorbidities. Moreover, utilizing functional neuroimaging could potentially reveal the neural networks involved in the development of visual anosognosia.

ETHICAL CONSIDERATIONS

This study complies with the ethical principles set out in relevant guidelines as specified in the Certificate of Agreement and Compliance in this research, as well as the National Ethical Guidelines for Research Involving Human Participants (NEGRIHP) 2022 Edition. Printed documents (i.e., consent form, printouts, case tracking sheets containing identifying information) are in a locked file cabinet when not in use. These documents are handled only by the primary investigator and corresponding co-authors. Electronic confidential data stored on transportable media such as flash drives, optical discs, and portable external drives is stored securely in the said locked file cabinet as well. Documents are also password-protected and handled only by the authorized investigators and co-authors. Data will be stored for five years, after which the hard copies will be shredded and thrown away, and the electronic files will be deleted. There are no direct or indirect benefits to the patient from the presentation of this case. However, knowledge from this case may give important insight for future studies and cases. The case was reviewed and approved by the Research Ethics Committee of the University of Santo Tomas Hospital.

INFORMED CONSENT PROCESS

A written informed consent form was obtained, understood, and signed by the patient, witnessed by the patient's son. Securing consent for the presentation and publication of the case, including other diagnostic results and images, was likewise done. The primary investigator, not the patient's primary physician, obtained the consent. They were knowledgeable and comfortable with the English language, and a written informed consent form in English was used to obtain consent. The family was also informed that the case would be published in scientific journals and may be presented at both local and international conferences to broaden knowledge regarding stroke management, particularly in acute bilateral occipital infarcts without visual anosognosia.

VULNERABILITY

The subject in this research was a patient of the secondary investigator.

Nevertheless, vulnerability was reduced when the primary investigator

obtained consent from the patient due to the decreased visual acuity. Since the patient is considered to be part of the vulnerable population, the principal investigator obtained informed consent from the patient as witnessed by his son.

PRIVACY AND CONFIDENTIALITY

Patients' confidentiality is protected by removing patient identifiers in the case report, with full compliance with the Data Privacy Act of 2012 and its implementing rules and regulations in 2016. There are no identifiers about the patient, which minimizes the risk of breach of privacy and confidentiality.

ACKNOWLEDGEMENTS

None

CONFLICT OF INTEREST

None

REFERENCES

1. Maddula M, Lutton S, Keegan B. Anton's syndrome due to cerebrovascular disease: a case report. Journal of Medical Case Reports. 2009 Sep 9;3(1):9028.
2. Kakaletsis N, Tziomalos K, Spilioti M, Diavati O, Chryssogonidis I, Savopoulo C. Bilateral cortical blindness due to bilateral occipital infarcts without anosognosia. J Neurol Stroke. 2016;4(1):00120.
3. Kaufman DM, Geyer HL, Milstein MJ, Rosengard J. Kaufman's Clinical Neurology for Psychiatrists-E-Book. Elsevier Health Sciences; 2022 Jan 14.
4. sen Tanrikulu C, Hocagil H, Kaya U, Hocagil AC. Acute bilateral vision loss in emergency department: A case report. Turkish Journal of Emergency Medicine. 2016 Mar 1;16(1):38-40.
5. Chang KM, Subrayan V, Patel DK. Bilateral occipital infarction: an uncommon cause of bilateral visual loss. Journal of Emergency Medicine. 2013 Mar 1;44(3):668-9.
6. Ziaul YH, Mittal J, Afroze T, Kumar V. Anton-Babinski syndrome: A visual anosognosia. Cureus. 2024 Mar 6;16(3).
7. Kondziella D, Frahm-Falkenberg S. Anton's syndrome and eugenics. Journal of clinical neurology (Seoul, Korea). 2011 Jun 28;7(2):96.
8. Atallah O, Badary A, Almealawy YF, Farooq M, Hammoud Z, Alrubaye SN, Alwan AM, Awuah WA, Abdul-Rahman T, Sanker V, Chaurasia B. Insights into Anton Syndrome: When the brain denies blindness. Journal of Clinical Neuroscience. 2024 Feb 1;120:181-90.
9. Amin HP, Madsen TE, Bravata DM, Wira CR, Johnston SC, Ashcraft S, Burrus TM, Panagos PD, Wintermark M, Esenwa C. Diagnosis, workup, risk reduction of transient ischemic attack in the emergency department setting: a scientific statement from the American Heart Association. Stroke. 2023.
10. Satija L, Singh AP, Khanna SK, Baijal V, Verma BS. Rare cause of complete blindness: Bilateral occipital cortical infarction: A Report on Three Cases. Medical Journal Armed Forces India. 1999 Apr 1;55(2):149-50.
11. Maddula M, Lutton S, Keegan B. Anton's syndrome due to cerebrovascular disease: a case report. Journal of Medical Case Reports. 2009 Sep 9;3(1):9028.
12. Andiappan K. Post-traumatic Anton's syndrome and its rehabilitation challenges: a case report. Iranian Rehabilitation Journal. 2021 Sep 10;19(3):331-6.
13. Tharaldsen AR, Kurz KD, Kurz MW, Midelfart A, Sand KM, Dalen I, Hoff JM. Volume and Visual Field Defects in Occipital Stroke: The NOR-OCCIP Study.Acta Neurologica Scandinavica. 2023;2023(1):3564863.
14. Markus HS, Michel P. Treatment of posterior circulation stroke: Acute management and secondary prevention. International Journal of Stroke. 2022 Aug;17(7):723-32.