Inverse Anton syndrome: A case report

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Abstract

This is a case report on a rare case of denial of visual perception termed as inverse Anton syndrome. It is a rare extension of perception without awareness in which, specific brain lesions affected a patient’s visual abilities. A 66-year-old Malay gentleman presented with sudden onset of painless bilateral total loss of vision with expressive aphasia for 2 days. His visual acuity was 6/12 in the right eye and 6/48 in the left eye. However, he was insistent that he could not see, but did not request any form of assistance for his blindness. He also had neurological signs suggestive of parietal lobe syndrome. Brain imaging showed subacute left middle cerebral artery territorial infarct with no occipital lobe involvement. The denial of visual perception by this patient may be explained by a disconnection of parietal lobe attentional systems from visual perception.

Keywords: Inverse Anton Syndrome, visual perception, blindsight, prosopagnosia

INTRODUCTION

Inverse Anton syndrome is related to the disorders of visual perception. This is the opposite of Anton syndrome, in which patients deny their blindness despite objective evidence of visual loss, and moreover confabulate to support their stance. In this case report, the patient stated that he had no vision, but was able to comfortably navigate in unfamiliar surroundings without assistance. This illustrates a rare phenomenon, where a patient presented with denial of intact visual perception in the absence of conversion disorder, following a cerebrovascular event.

CASE REPORT

A 66-year-old Malay man with underlying hypertension, hyperlipidemia and gouty arthritis presented with sudden onset of painless bilateral total loss of vision of 2 days duration. The visual loss was associated with mild right-sided body weakness and difficulty in retrieving words to describe things. Otherwise, there was no other ocular symptom.

On presentation, the Glasgow Coma Scale score was normal with raised blood pressure of 175/100 mmHg. Initial visual acuity was perception to light bilaterally, however, on further evaluation, there was presence of unconscious processing of visual information, which correlated to at least 6/60 vision in both eyes. Pupillary reflexes were normal suggesting an intact anterior visual pathway. The significant ocular findings were bilateral immature cataracts and left fundus examination showed features of non-ischemic central retinal vein occlusion (CRVO) with mild macular oedema. Neurological examination revealed paresis of right extremities and, acalculia, expressive aphasia with left and right disorientation, which was suggestive of parietal lobe syndrome. On further systemic assessment, the patient was neither hysterical nor psychotic. Urgent computed tomography (CT) of the brain revealed ill-defined hypodensities at the left frontoparietal region suggestive of acute infarct (Figure 1). There was no evidence of occipital lobe lesions or hypodensity; hence cortical blindness and blindsight were excluded.

During his follow up a month later, his neurological deficit had resolved, however, his complaint of total loss of vision was still persistent. Thus, magnetic resonance imaging (MRI) of the brain was performed and showed a subacute left middle cerebral artery (MCA) territorial infarct involving left parietal lobe and precentral gyrus (Figure 2) with no evidence of occipital lobe infarct. The magnetic resonance
angiography (MRA) demonstrated thrombosis with atherosclerotic changes of M1 segments of bilateral MCA. Repeated visual acuity using Kay pictures showed best corrected visual acuity (BCV A) of 6/12 in the right eye and 6/48 in the left eye. On confrontation test using a finger counting task, the patient exhibited normal visual fields. His conduction aphasia was prominent; however, he was still able to name target objects and able to follow instructions to draw a cross in a circle. The most notable observation was that he was able to walk unassisted in the clinic without any difficulty despite insisting that he had complete visual loss.

Based on the clinical evaluation and radiological findings, the patient was diagnosed to have Inverse Anton syndrome following left sided cerebrovascular disease with concurrent CRVO of the left eye. Antiplatelet therapy was prescribed for prevention of recurrent stroke and antihypertensives were added to optimize his blood pressure. In view of the recent cerebrovascular disease, the macular oedema secondary to CRVO was treated conservatively.

According to his family, he was competently carrying out simple daily activities at home unaided, which was inconsistent with his self-report of an inability to see. Counselling was given to his family members to understand that his confabulation was due to cerebrovascular disease and not due to malingering or psychotic disorders. He was referred to the rehabilitation unit for speech and occupational therapy.

**DISCUSSION**

Inverse Anton syndrome is rare and inadequately detailed in description, and therefore does not clearly document the clinical existence of this syndrome. Structurally, it may be explained by a disconnection of parietal lobe attentional systems from visual perception. The latest reported case of Inverse Anton syndrome was in 1991, where the patient denied the ability to see despite having had small areas of well-documented visual capability in the middle of his visual field. To the best of our knowledge, there are no reports describing patients with clearly documented preserved vision, particularly to the extent displayed by our patient. There was no evidence of any secondary gain for his blindness, yet he maintained being totally unaware of his well-documented visual skills. It is connected to the neuropsychological syndromes in which visual perception and consciousness are dissociated as a result of brain damage, such as in blindsight and prosopagnosia.

Blindsight is a pattern of behavior that is displayed by individuals who are blind due to a damaged striate cortex. In blindsight, patients demonstrate primitive visual skills, such as light detection and orientation to a target within a perimetrically blind visual field. Prospagnosia, is a specific form of visual agnosia characterized...
by the inability to recognize familiar faces despite apparent recognition of most other stimuli.\textsuperscript{1,4,7}

Similar to the above-mentioned conditions, our patient also demonstrated the ability to assess and utilize visual information without knowing that the input modality was in fact visual. Clearly, this deficit was not in face recognition and superior to that indicated in blindsight, as he was able to correctly identify numerous types of visual information presented within the intact visual field and make appropriate responses. There were no occipital cortex lesions as evidenced radiographically, which excluded cortical blindness and blindsight.

The diagnosis of disorders of higher cortical visual function often poses a considerable clinical challenge.\textsuperscript{2} Routine evaluations of visual function and examination may not readily yield a diagnosis; a refined examination of visual function is necessary to identify these elusive syndromes.\textsuperscript{2} More recently, neuroimaging techniques have allowed further insights into complex structure-function relationships. However, typical lesions have both hodological and topological effects; thus, pure disorders of one or other class may be largely theoretical.\textsuperscript{2-4}

Management of these disorders is mainly focused on secondary prevention and rehabilitation to improve quality of life.\textsuperscript{5} Timely diagnosis and optimal management of these disorders are important challenges faced by the practicing ophthalmologist and neurologist.\textsuperscript{2,3} We hope that this case report will provide a fresh perspective to challenge and refine the neuroscience of visual perception and visual perceptual dysfunction.\textsuperscript{2,4}

DISCLOSURE

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Conflicts of interest: None

REFERENCES