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Case Report

Persistent Eye Contact by a Patient with Temporary Blindsight Followed By Visually Conscious Prosopagnosia: A Case Report

Richard G. Coss*

Department of Psychology, University of California, Davis, California, USA

***Corresponding Author:** Richard G. Coss, Department of Psychology, University of California, Davis, California, USA

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Abstract

A single case study of a female patient with temporary blindsight is reported. The patient experienced traumatic brain injury from a fall and experienced blindsight for approximately 61 hours. While hospitalized, the patient exhibited unusually persistent eye contact with hospital staff and visitors that was maintained at varying distances. Follow-up interviews during partial recovery of conscious vision revealed a complete failure to recognize familiar faces. Without individual recognition, facial Gestalts were perceived only by progressive scanning of facial features. Later CT-scans show bilateral scaring in the optic-radiation fibers projecting from the lateral geniculate nucleus to the primary visual cortex that includes scaring in grey matter. A perimetry test showed degraded vision in the upper visual fields and vision tests showed intact foveal perception sufficient for projected chart-letter recognition. The ability to see the eyes of other people using blindsight, as characterized by intense visual fixation of the eyes of others, suggests continued functionality of the superior colliculus, an evolutionarily ancient neural structure capable of recognizing the schema of two-facing eyes.

Introduction

The well-documented ability to know the locations of spatially distributed objects and navigate around them without conscious awareness is termed "blindsight" [1]. Typically the result of brain injury, it is a residual capacity to identify visual information without the ability to describe what visual features are perceived. To navigate around objects with blindsight, the outer contours of objects must be roughly perceived with finer detail detectable, but indescribable [2]. Affective blindsight is the ability to discriminate emotional facial expressions via functional magnetic resonance imaging (fMRI) without the subjective ability to describe what emotions are expressed [3,4].

Although not explicitly a facial expression, the orientation of a person's gaze conveys information about their direction of attention and interest. Many species, including humans, have experienced an evolutional history shaped by natural selection for detecting an agent's direction of gaze as a component of face-recognition. Such behavior is essential for adjusting social interactions [5] and for evading predators [6]. For example, the ability to discriminate a facing and averted gaze in facial photographs using blindsight is evident from fMRI brain scanning of a patient who experienced loss of his primary visual cortex [7]. The following case report describes a patient who experienced temporary blindsight from a fall and exhibited an atypical pattern of sustained eye contact without the ability to see two-facing eyes consciously.

Case Presentation

Patient JC is a 73-year-old well-educated right-handed women with Parkinson's disease who suffered a traumatic brain injury when, as witnessed by her husband, she abruptly lost consciousness and collapsed from a standing height onto a hardwood floor after complaining of intense pain. Her left forehead exhibited a small contusion and, while attempting to lift herself up from the floor aided by her husband, she stated that the well-illuminated room was dark with no consciously perceived images. An ECG conducted by paramedics indicated paroxysmal atrial fibrillation that vacillated with a sinus rhythm in the ED, requiring amiodarone loading to convert. Computer Tomography (CT) imaging showed no evidence of acute head injury and the CTA of head and neck with IV contrast was normal. However, CT imaging a year later showed substantial bilateral scaring of the optic-radiation fibers emerging from the lateral geniculate nucleus with scaring also appearing in the primary visual cortex (Figure 1). This assessment is based on aligning these brain images with published high-spatial-resolution diffusion tensor tractography of the visual pathways [8].

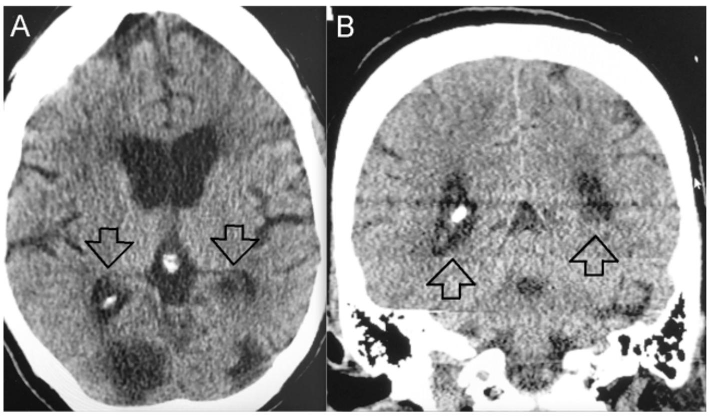

Figure 1: Computer Tomography brain scan showing bilateral scarring (arrows) of optic-radiation fibers projecting from the lateral geniculate nucleus to the primary visual cortex. **(A)** Coronal section also shows bilateral scarring of grey matter in primary visual cortex. **(B)** middle-posterior section.

On her first day of hospitalization, JC reported that could not see any objects and people from her hospital bed, but could see their movement that facilitated face-to-face interactions. Most importantly, JC engaged in unusually persistent eye contact with visitors and hospital staff from her hospital bed that was verbally acknowledged by her husband, three visitors, and the attending physician. Although eye contact is a salient, socially significant phenomenon, sustained eye contact by a patient with blindsight was mentioned briefly by Solcà and colleagues [2], and this observation inspired their further enquiry into the unconscious properties of face perception. This case report documents JC's temporary blindsight from her traumatic brain injury and partial recovery of conscious vision. A review of the literature on blindsight is discussed that provides insight into the subcortical properties mediating sustained eye contact and the lack of intermittent gaze aversion that typically occurs during social interactions [5].

Behavior using blindsight

On initial observation of JC's persistent eye contact, this investigator immediately recognized the importance of this observation and documented JC's progressive changes in face-perception ability during hospitalization followed by repeated questioning of this ability during follow-up interviews spanning three months. JC exhibited blindsight during the first and second day of hospitalization (approximately 61 h) in which she could visually track the movement of hospital staff and visitors while reporting that she could not see them. Her hospital room appeared completely dark despite its fluorescent illumination and large window. JC was able to maneuver around her hospital bed with her mobile IV stand, avoiding collision with visitor chairs. With respect to eating, she could visually grasp the black plastic utensils on her bed table despite saying that could not see them. This visuomotor capability while partially reclined was well illustrated by her precision gripping of the handle of a black fork held vertically in front of her by this investigator at ~20 cm distance.

As mentioned above, JC engaged in persistent eye contact with all visitors and hospital staff, presumably aided by the direction of their voices, movement, and facing orientation. With blindsight thought to characterize functional uncoupling to the striate cortex and possibly extrastriate cortex from subcortical visual areas [9], JC's ability to answer questions during her transition from unconscious to conscious vision sheds important light on the interactions and functional constraints of these perceptual systems. It must be noted here that JC's exhibited impaired short-term memory that impacted her long-term memory consolidation two days before her concussion and four days afterward. Nevertheless while experiencing blindsight, she was capable of engaging in coherent conversations about the limitations of her perceptual abilities.

JC greeted visitors and hospital staff with effusive smiles and rapidly stared directly at their eyes without averting her gaze as if signaling her need for assistance. This investigator tested JC's sustained eye contact by varying his distance from her during social discourse when he approached closely to within 20 cm and by withdrawing more than 5 m while maintaining eye contact. Based on JC's comments that people's movement and direction of voices facilitated her tracking their location, this investigator initiated an episode of continuous eye contact as he moved slowly and silently around the hospital bed in a 180° arc at a 3-m distance from JC's face. Together, JC's maintenance of eye contact during these exploratory actions implied that her recognition of the facing-eye schema was size-invariant in the absence of conscious vision.

Recognition of two-facing eyes without consciously seeing them was tested further by requesting JC to point to each eye of this investigator, an action that she did correctly by aligning her right forefinger toward each pupil within a 10 cm distance. This investigator's next query involved JC's ability to see his mouth at ~30 cm distance. Other than his two-facing eyes, no other facial features were unconsciously apparent, even when this investigator opened his mouth and waved his tongue sideways for several seconds to increase mouth conspicuousness. Moreover, JC's eye contact was never broken during this test despite this investigator's tongue movements. This absence of animate mouth perception during JC's period of blindsight is especially surprising in light of her ability to detect the movement of nearby people. During a test of JC's blindsight vision by the attending physician, JC could identify correctly whether his right and left arms were waving up and down, an ability indicating course recognition of an upright human body without conscious awareness.

On her second day of hospitalization, JC was able to discern the circular shape of a black ring (7.6 cm dia. with 13 mm width rim) on a cream-colored background denoting the speaker of her hospital-bed telephone. When handed to her, she repeatedly pressed the black circular rim to answer her phone rather than a nearby illuminated button while claiming that she did not see either. Together, these observations document JC's blindsight ability to maintain size-invariant eye contact at varying distances, point to each eye of this investigator, grasp the upright handle of a black fork, and differentiate the spatial aspects of arm waving. Such limited face-perception ability restricted to two-facing eyes contradicts experimental fMRI evidence of amygdala activation of healthy participants during unconscious perception of backward-masked facial images displaying parted lips [10,11]. In particular, Morris and colleagues [10] suggest that rapid activation of the colliculo-pulvinar-amygdala pathway might explain the ability to see a backward-masked face without conscious awareness.

Emergence of conscious vision

The first evidence of conscious face perception occurred on the third day of hospitalization when JC spontaneously commented that "light is coming out of people's eyes." Despite this visual anchoring of head position and orientation by the appearance to glowing eyes, the head contours of visitors and staff were only vaguely perceived.

On her fourth day of hospitalization, JC was discharged for testing by an ophthalmologist who reported no retinal damage except for a prior diagnosis of macular degeneration that did not compromised her previous driving ability. During this visit, JC was unable to read any projected chart letters resulting from obstructing visual distortions generated by her hallucination of a moving stream of letters on the projection screen analogous to a decade's-old scrolling luminous newspaper on a building facade. However, another optometrist's examination of chart-letter recognition 31 days after her concussion indicated only a slight degradation using exclusively foveal vision that was insufficient to warrant new eyeglasses. A standard automated perimetry test by a third ophthalmologist 40 days after brain injury revealed bilateral loss of vision mostly in JC's upper right visual fields and minor bilateral loss in the lower visual fields (Figure 2).

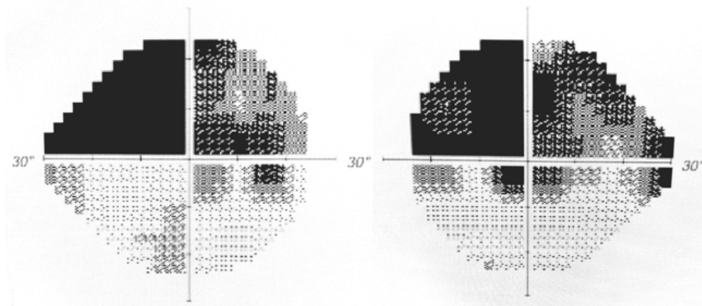

Figure 2: Degraded bilateral vision in the upper visual fields derived from a standard automated perimetry test.

The following review of JC's conscious face perception is restricted to the first three months following brain injury. As her conscious vision improved slowly during the first month after hospitalization, individual face recognition by JC was completely absent, a condition labeled as prosopagnosia [2]. Nevertheless, the basic shape of faces could be discerned by the appearance of two "greyish blobs" in the horizontal plane characterizing two-facing eyes within the boundary of indistinct head contours. Nevertheless, JC could still target the eyes with her concentrated gaze. With focused attention at ~30 cm distance, however, JC could discern only the dark pupil of an individual's left eye, but not the pupil of the right eye that appeared closed. Despite this limitation of seeing two-facing eyes, JC continued to make extensive eye contact with this investigator and other visitors that did not appear aberrant because of the natural episodes of visitor gaze aversion during social discourse.

There was minor improvement in which JC could see two-facing eyes more clearly during the second month after her concussion in which the cornea, sclera, and surrounding eyelids were more evident on the left sides of people's faces while their darker right sides continued to exhibit indistinct eyelid contours with the their eyelids appearing partially closed. Eyebrows, however, were not apparent and the lower regions of familiar faces were still indistinct, with only vague perception of smiling lips and teeth that could be discerned more clearly with sustained attention. When requested more than once to point to each eye of this investigator, her forefinger alignment toward each of his eyes was less precise than demonstrated during her period of blindsight; albeit, during her last test session of pointing, JC could align her finger appropriately with verbal feedback. As with number and text recognition described above, progressive scanning of facial features with focused attention over several seconds engendered the slow emergence of two vaguely perceived eyes and a mouth. Despite this level of perceptual integration yielding a coarse facial Gestalt, JC still failed to recognize the faces of her husband and other familiar people.

Two months after her concussion, JC's sharp visual acuity continued to be restricted to the foveal region, and she described her wider bilateral visual field as "mottled as if a painter had smeared translucent greyish globs of paint" on top of the objects and landscape scenes she viewed. Some improvement was evident three months after injury in which the sky continued to appear grey, but objects in the foreground had regained their natural color; although, visually fixated red and green traffic lights had white halos. Nevertheless, at the end of the third month following brain injury, JC continued to recognize small numbers and text by progressive foveal scanning, and the distinctiveness of larger shapes was hampered by the bilateral darkness of images in JC's right visual fields of both eyes. Such a hemifield defect in contrast perception is suggestive of greater injury to the right lobe of the striate (V1) cortex, consistent with the more extensive scaring of the right optic-radiation fibers and grey matter evident in the CT scans.

Discussion

The extensive interviews of patient JC during her hospitalization and recovery at home for a three-month period yielded substantive information useful for interpreting what brain functions mediated her prolonged eye contact during her episode of blindsight. Especially notable was the disorder of conscious face perception during JC's first week following hospitalization, specifically the appearance of a person's eyes as two ill-defined greyish blobs with only a vague definition of head contours and other facial features.

Following her recovery of conscious vision, JC could recognize the fine detail of images within her ~1° zone of foveal vision, a property that interfered with her seeing both eyes and adjacent facial detail of other nearby people simultaneously. To compensate, JC resorted to focused serial scanning of facial features without the ability to recognize individual faces. Such conscious visual impairment, contrasted by JC's ability to maintain eye contact at varying distances during her episode of blindsight, suggests that her concussion involved injury to striate (V1) and the extrastriate ventral stream (V2 to V4), presumably affecting the facial-feature integration of the fusiform face area [12,13]. However, JC's ability to correctly identify the left and right waving arms of the attending physician using blindsight suggests some degree of functional connectivity to the motion-sensitive STS (prestriate visual area V5/MT+) via the lateral geniculate nucleus, superior colliculus (SC) and pulvinar [14,15]. Nevertheless, it more likely that the SC afforded the arm-movement perception because the STS appears sensitive to small facial movement [13,16] and JC failed to detect this investigator's waving tongue with blindsight. In particular, JC's ability to establish prolonged eye contact at varying distances without visual awareness supports an argument that, with absent or degraded V1 functionality [17,18], subcortical visual perception by the SC that regulates gaze orientation and attention [19,20] remained sufficiently intact for eye-schema recognition.

To argue further for the role of the SC in JC's prolonged eye contact, research on macaques has shown that, within the SC, the superficial layers receive approximately 10% of the total projections from retina ganglion cells [21] and the SC is responsive to eyelike patterns [22].

Conclusion

The neural circuitry in the SC engendering recognition of two discoid shapes in the horizontal plane is phylogenetically ancient, as evident for the homologous optic tectum of some reptiles and teleost fish useful for evaluating the facing orientation and intentions of conspecifics and predators [23]. Apparently, recognition of two-facing eyes is very ancient phylogenetically in divergent reptilian and mammalian lineages, extending back in time to the Middle to Late Triassic period with the discovery of fossilized impressions of forewing eyespots on the wings of moth-like cicadas [24]. These paired eyespots resembling two-facing eyes, presumably acted as a Batesian mimicry device to deter insectivorous predators that would have relied on their optic tectum for visual-pattern discrimination.

With respect to JC's scaring of the optic-radiation fibers shown in CT scans (Figure 1) coupled with degraded vision in her upper visual fields (Figure 2), the most parsimonious explanation for JC's sudden loss of conscious vision when her head hit the hardwood floor was induced by the sudden deceleration/acceleration forces that stretched and compressed the optic-radiation fibers projecting to the primary visual cortex from the lateral geniculate nucleus. Such damage to the cortical visual stream yielded a form of retrogression into a more developmentally primitive state that still retained the ability to see two-facing eyes via the innate face-recognition properties of the SC [25]. It seems reasonable to argue that JC's persistent eye contact with facing individuals during her period of blindsight resulted from the absence of any other unconsciously perceived visual cues for directing gaze during social interactions. Within this context, the absence of gaze aversion that typically occurs following brief bouts of eye contact [5] suggest that the dynamic properties of gaze aversion are mediated exclusively by higher-order cortical processes.

Declarations

Patient consent for publication: The author received informed consent from the patient before proceeding with his examination.

Competing interests: The author declared no potential conflicts of interest with respect to the research or publication of this article.

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