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Disconnection between parietal and temporal areas without simultanagnosia: a case study of prosopagnosia.

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Abstract

This study presents a comprehensive neuropsychological evaluation of a unique case of prosopagnosia (patient EP) with atypical lesion patterns, characterized by intact face-specific nodes but significant damage to the Vertical Occipital Fasciculus (VOF). Given the presumed interruption of ventral-parietal connectivity, we focused on assessing the potential presence of simultanagnosia and its relationship to face recognition deficits. Our extensive neuropsychological battery included tests of global and local processing, scene perception, and face recognition. Results revealed intact global processing abilities and no evidence of simultanagnosia, despite the patient's prosopagnosia. These findings suggest that EP's face recognition impairment is likely attributable to disrupted connectivity within the face processing network rather than a general deficit in global visual processing. This case highlights the importance of comprehensive neuropsychological assessments in atypical presentations of prosopagnosia and contributes to our understanding of the complex relationship between white matter integrity and face recognition abilities.

Keywords: Prosopagnosia, Neuropsychological Assessment, Face Recognition, Global Processing, Simultanagnosia.

Introduction

Face recognition is a complex cognitive process that relies on the intricate interplay of various brain regions and neural networks. Prosopagnosia, a selective impairment in face recognition, has traditionally been associated with lesions in face-selective areas such as the fusiform face area (FFA) and occipital face area (OFA) (Corrow et al., 2016), or to damage of white matter tracts, particularly the inferior longitudinal fasciculus (ILF) and inferior fronto-occipital fasciculus (IFOF), which connect nodes of the face processing circuitry (Valdés-Sosa et al., 2011). However, recent research has highlighted the importance of considering broader visual processing deficits that may contribute to face recognition impairments, particularly in cases with atypical lesion patterns (Cohen et al., 2019; Fox et al., 2008; Grill-Spector et al., 2017).

Simultanagnosia, a neurological disorder characterized by the inability to perceive multiple elements of a visual scene simultaneously, is one such deficit that can potentially impact face recognition (Sakurai et al., 2016). While typically associated with bilateral parietal lobe damage, simultanagnosia can also result from lesions in other brain regions, leading to complex presentations that challenge our understanding of visual processing disorders (Cui et al., 2022).

The co-occurrence of prosopagnosia and simultanagnosia has been observed in various neurological conditions, most notably in posterior cortical atrophy (PCA) (Cui et al., 2022). PCA is a neurodegenerative syndrome characterized by progressive visual dysfunction, often presenting a combination of visual agnosias, including prosopagnosia and simultanagnosia (Cui et al., 2022). This co-occurrence raises important questions about the potential interactions between these two disorders and their combined impact on visual processing, particularly in face recognition.

In this study, we present a unique case of a prosopagnosia, patient EP, with lesions outside the typical face-selective areas, specifically the Fusiform Face Area (FFA) and Occipital Face Area (OFA). Additionally, the patient has damage to the white matter tracts, including the Inferior Longitudinal Fasciculus (ILF) and the Vertical Occipital Fasciculus (VOF). As a result of the VOF interruption, the patient exhibits diminished connectivity between the ventral-temporal cortex and dorsal visual areas. This disruption raises the possibility that other visual processing deficits may be contributing to the observed face recognition deficits.

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The primary objective of this research is to conduct a comprehensive neuropsychological evaluation to assess the potential presence of simultanagnosia in our prosopagnosic patient. This assessment is crucial as it allows for a thorough examination of visual processing in a case with atypical lesion patterns and connectivity issues, helps determine whether other visual processing deficits beyond damage to face-selective areas contribute to the observed face recognition deficits, and provides an opportunity to investigate how disrupted connectivity between ventral-temporal and dorsal visual areas might impact face recognition processes.

Understanding the potential contributions of various visual processing deficits to face recognition impairments is particularly relevant in cases where prosopagnosia presents with atypical lesion patterns. Through careful neuropsychological assessment and analysis, we hope to contribute to the growing body of knowledge on face recognition disorders and provide valuable insights for both researchers and clinicians working in this field. By examining the visual processing abilities in a case of prosopagnosia with intact face-selective areas but disrupted white matter connectivity, we may enhance our understanding of the complex neural networks underlying visual processing and face recognition. This case study highlights the importance of comprehensive assessments in atypical presentations of prosopagnosia and underscores the need for a nuanced approach in understanding the various factors that can contribute to face recognition deficits.

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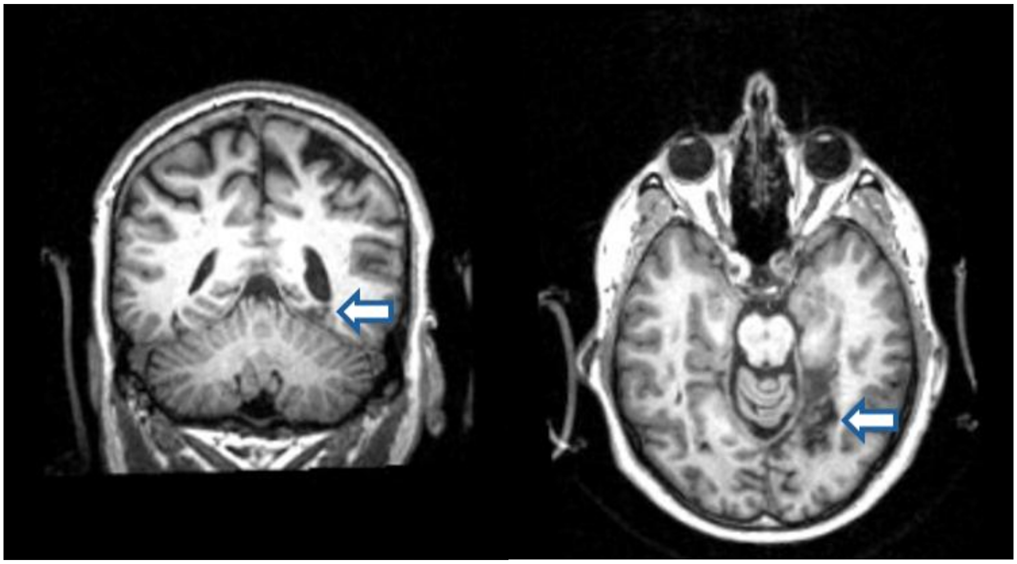
Materials and Methods

Patient Description and Previous Neuropsychological Assessment

Subject EP, currently 56 years old, initially presented for evaluation at age 46 following two consecutive strokes within a 1-day interval. The primary lesion was identified in the right hemisphere, affecting parts of the lingual gyrus, medial fusiform gyrus, and a small portion of the inferior temporal gyrus (see Figure 1). This lesion extended dorsally into white matter, slightly impacting the ventral part of the optic radiation. A secondary, smaller lesion was identified in the left hemisphere, confined to the middle and posterior fusiform gyrus. Further neuroimaging studies 4 and 8 years later and voxel-based morphometry (VBM) analysis, consistently confirmed the location of the lesions. Importantly, these analyses revealed that the lesions did not directly intersect the Fusiform Face Area (FFA) but did affect more medial portions of the ventral-temporal cortex within the broader face processing network (Bobes et al., 2021).

Figure 1.

T1-weighted structural image of patient EP's lesion. Arrows indicated the lesion on the right hemisphere described in the main text.



During his initial evaluation, EP exhibited significant impairments in facial configuration processing. He scored 46 out of 64 on the Facial Identity Matching Test, 10 out of 27 (37%) on the Benton Face Recognition Test, <50% in the Cambridge Face Memory Test, and was severely impaired in recognizing famous faces, with only 5 out of 14 celebrity faces being visually recognized. EP reported relying on non-facial features like hairstyle for recognition. Despite intact low-level visual perception, as revealed by average scores on orientation, length, and size matching tasks from the Birmingham Object Recognition Battery, he struggled with intermediate-level tasks dependent on configural information, such as face and house part-to-whole matching tasks (De Gelder et al., 2022).

In further evaluations of face recognition conducted 4 and 8 years after the initial assessment, EP's performance remained impaired but showed some variability. Four years after the initial evaluation, EP scored 35 out of 54 (65%) on the Benton Facial Recognition

Test and 56% on the Cambridge Face Memory Test. In the assessment conducted 8 years post-initial evaluation, EP's scored 46% on the Cambridge Face Memory Test. These results indicate a persistent deficit in face recognition abilities, consistent with the diagnosis of prosopagnosia, while also revealing subtle fluctuations in performance over time.

Procedure

A comprehensive neuropsychological assessment was conducted to evaluate the potential presence of simultanagnosia and its contribution to the patient's face recognition deficits. The assessment was performed in a quiet, well-lit room. The assessment was conducted remotely, and considered guidelines for remote neuropsychological assessment (Bilder et al., 2020). Test instructions were presented verbally and written by screen sharing. Stimuli was presented via screen sharing, except for the Navon classical task that was executed directly on the patient’s computer. The patient was accompanied by a family member who was proficient in using audiovisual technology for the assessment. All researchers who participated in the remote assessment had also been part of previous in-person assessments of the patient. The patient does not wear corrective lenses for reading, he only has a prescription for larger distances.

Neuropsychological Tests

The following tests were administered to assess various aspects of visual processing, with a focus on evaluating simultanagnosia and its potential impact on face recognition.

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1. Integration of Complex Scenes. The patient was asked to describe as accurately and thoroughly as possible what he saw that was going on in two pictures. The first picture was the "Cookie theft" from the Boston Diagnostic Aphasia Examination (Goodglass & Kaplan, 1983), and the second one was the scene from The Birthday Party test (BPT; de Vries et al., 2022). The patient was encouraged to give more information when he was not giving further responses for some time. Scoring was done following Croisile et al. (1996) guide for the first picture, and the scoring aid of the BPT.

2. Global/Local Processing. The patient was asked to describe as thoroughly as possible a set of 11 figures with different elements forming local and global features. The first two figures were composed of geometric figures (squares and triangles), the next two were schematic figures (cup, umbrellas, fish), and the fifth figure was a classic Navon figure of a global H composed of smaller F's (Navon, 1977). The remaining figures in the set were paintings by Giuseppe Arcimboldo and Octavio Ocampo that had different figures at local and global hierarchies. This task was not time limited.

Furthermore, the patient was administered a full classical Navon task designed in PsychoPy (Peirce et al., 2019). The task consisted of four blocks (half local letters, and half attending global letters) of hierarchical letters. Each trial consisted of a hierarchical letter (e.g. a big letter 'H' composed of small letters, either 'H' or 'S') that could be congruent (same letter on both levels) or incongruent. The patient was asked to respond which letter was represented in the local level in half of blocks, and in the local level in the other half, as fast as possible without sacrificing accuracy. There was a practice block for each level at the beginning of the task that ended when the patient had at least 80% of accuracy. At a distance of 60 cm, the size of large letters was 3.31° x 5.23° of visual angle, and the small letters were 0.47° x 0.66°. The patient remained at a constant distance from the screen throughout the task. For each trial, there was a blank screen for 500ms, then 500ms of fixation cross, followed by 180 ms of the hierarchical letter, that was substituted by a mask made out of dots. This mask remained on-screen until participant gave a response. The classical Navon effect consists of slower reaction times for incongruent letters only when attending the local level. The task was administered via Pavlovia, and the size of stimuli was standardized by adjusting it according to the size of a credit card compared at the beginning of the task.

3. Dot counting task. The patient was asked to count the number of dots in a square, without pointing to the screen. This task was adapted from the Addenbrooke's Cognitive Examination-III (ACE-III, Hsieh et al., 2013). The dot counting task is one of the most sensitive task for ventral simultanagnosia (Sakurai et al., 2016).

4. Subjective Exploration. A semi-structured interview about day-to-day difficulties with objects or spatial abilities. Patient was asked: "Have you or anyone close to you noticed difficulties recognizing or telling apart some things in your day-to-day activities? Can be at home, work, or other activities", "How would you qualify your vision in the last 3 years? Has it improved, worsened, or stayed the same?", and "When you need to find something, either at your house or at your work or else, for example, missing keys, how do you usually manage? Do you find it easy or difficult? Have you needed help in situations like these?".

5. Visual Imagery. We adapted the Vividness of Visual Imagery Questionnaire (Blomkvist & Marks, 2023) by administering the first four items ("Think of some relative or friend whom you frequently see") and a new set of four items that more directly asked the patient to imagine the face of that same relative or friend ("Think of the face of some relative or friend whom you frequently see") asking about the clarity of the imagined face features ("overall image of the face, eyes, mouth, nose", "expressions of the face", "face smiling", and "different colors and textures of the skin"). Responses were given in a 5-point scale (from "Perfectly clear and vivid as real seeing" to "No image at all, you only 'know' that you are thinking of the object"), in two conditions: eyes open and eyes closed.

6. Object Closure. The first task was the incomplete letters from the ACE-III (Hsieh et al., 2013), where the patient is asked to identify letters that are missing some parts. The second task was an adaptation of the Mooney Face Test (Mooney, 1957). The patient was asked to select which of three images shown in a slide was a face. Stimuli remained on the screen for a maximum of three seconds. There were 20 items and a practice trial. Stimuli was one Mooney face per trial and two distractors (scrambled Mooney stimuli) selected from the set published by Schwiedrzik et al. (2018).

7. Facial Feature Processing. The patient was asked to classify a set of 20 neutral faces by their age (young vs old) and gender (male vs female) (Tranel et al., 1988). There were 20 photos of males and 20 photos of females. Half of each set was young (19-31 years old) and half older (69-80 years old). Stimuli remained on the screen for a maximum of three seconds. Images were selected from the FACES database (Ebner et al., 2010).

Data Analysis

Performance on each task was evaluated against normative data where available. Qualitative analysis of response patterns was conducted to identify signs of simultanagnosia, such as piecemeal processing, difficulty with global integration, and impaired perception of multiple elements simultaneously. The patient's performance on face-specific tasks was compared to their performance on non-face visual processing tasks to assess the specificity of deficits. In the classical Navon task, we estimated accuracy for all trials (excluding practice trials), and response times averages were estimated only on correct trials and response times above 200ms.

Ethical Considerations

The study was conducted in accordance with the Declaration of Helsinki and approved by the institutional ethics committee. Informed consent was obtained from the patient prior to the assessment.

Results

The patient underwent a comprehensive neuropsychological assessment to evaluate potential simultanagnosia and its contribution to face recognition deficits. The results are summarized below.

Integration of Complex Scenes and hierarchical figures

For the Cookie Theft picture, the patient's performance was within the normal range compared to healthy subjects (de Vries et al., 2022), scoring 18 out of 22 overall (Mean healthy subjects = 18.61, SD = 2.82). In the Birthday Party Test (BPT), the patient performed well, scoring 28 out of 40 total (Mean healthy subjects = 26.49, SD = 5.71) (de Vries et al., 2022). Notably, the patient identified all persons/animals and performed above average in actions/relations (see Table 1).

In the Hierarchical Figures test, the patient successfully reported both global and local levels for all 11 figures, although they took longer to recognize the global level in the 11th figure.

In the classical Navon task, the patient had good accuracy in general (86.9%), except on incongruent letters during local attention (67.5%) which had lower accuracy than congruent letters (88.8%). The reaction times on correct trials showed the classical congruency effect during the local attention condition (congruent mean RT=812ms, SD=194ms; incongruent RT=845ms, SD=177ms), and no congruency effect on the global condition which showed overall faster reaction times (congruent mean RT=582ms, SD=102ms; incongruent mean RT=585ms, SD=102ms). Though the congruency effect during local attention was small (incongruent trials were 33 ms slower on average), note the speed-accuracy trade-off with lower accuracy for incongruent trials.

Detection of Multiple Objects

The patient achieved a perfect score (4/4) on the dot counting task from the ACE-III.

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Table 1.
Patient EP scores in the battery (points scored/maximum), and conversion to T-scores based on means (SD) of controls.

Test	EP Raw score	Mean (SD) in controls	T	Interpretation
BFRT – Long form	35/54	45.4 (3.96)	-2.63	Severely impaired
Famous faces test	36/73	NA	NA	Impaired
CFMT (%)				
4 years after lesion	56/100	80.4 (11)	-2.22	Severely impaired
8 years after lesion	46/100	80.4 (11)	-3.13	Severely impaired
Hierarchical figures	11/11	At ceiling	NA	Normal range
Cookie theft				
Subjects	3/4	3.48 (0.56)	-0.86	Normal range
Objects	8/11	8.38 (2.02)	-0.19	Normal range
Places	2/2	1.69 (0.57)	0.54	Normal range
Actions/facts	5/7	5.06 (0.97)	-0.06	Normal range
Total	18/23	18.61 (2.82)	-0.22	Normal range
The Birthday Party (TBP)				
Persons/animal	9/9	7.78 (1.52)	0.80	Normal range
Objects	11/18	11.42 (3.99)	-0.11	Normal range
Actions/relations	8/13	7.29 (1.94)	0.37	Normal range
Total	28/40	26.49 (5.71)	0.26	Normal range
ACE-III				
Dot counting	4/4	At ceiling	NA	Normal range
Incomplete letters	4/4	At ceiling	NA	Normal range
Mooney faces	19/20	Near ceiling	NA	Normal range
Facial characteristics				
Categorize age	20/20	At ceiling	NA	Normal range
Categorize sex	19/20	Near ceiling	NA	Normal range
Navon, Accuracy (% correct)				
Global, consistent	98	98 (2.1)	-0.09	Normal range
Global, inconsistent	94	97 (3.3)	-0.85	Normal range
Local, consistent	89	97 (3.2)	-2.32	Impaired
Local, inconsistent	68	87 (18.1)	-0.99	Normal range
Navon, RT (milliseconds)				
Global, consistent	582	569 (136)	0.09	Normal range
Global, inconsistent	585	559 (132)	0.18	Normal range
Local, consistent	812	662 (134)	1.05	Normal range
Local, inconsistent	845	784 (194)	0.29	Normal range
Inverse Efficiency Scores (IES)				
Global, consistent	597	582 (139)	0.10	Normal range
Global, inconsistent	624	577 (141)	0.31	Normal range
Local, consistent	914	684 (151)	1.43	Normal range
Local, inconsistent	1252	905 (287)	1.13	Normal range
Global-Local bias	0.704	0.781 (.076)	-0.95	Normal range

Notes. NA=Not available. BFRT=Benton Face Recognition Test. CFMT= Cambridge Face Memory Test. ACE-III=Addenbrooke's Cognitive Examination-III. T=Crawford's t-values to compare single cases against a control group. RT=Response time.

Subjective Exploration

The patient reported that neither him nor other people have noticed other difficulties of visual recognition apart from their face recognition difficulties and the upper-left scotoma. Patient reports not having any change in their vision in the last 3 years, and does not report any further difficulties with vision, visual search (e.g. finding misplaced keys) or spatial navigation in their work, which involves high visuospatial skills.

Visual Imagery

On the Vividness of Visual Imagery Questionnaire (VVIQ), focusing on facial imagery, the patient's scores ranged from 3 to 5 (on a scale of 1-5), indicating moderate to high vividness of visual imagery for facial features and associated details. There was no difference between conditions with eyes closed (mean=4.1) and eyes open (mean=4.1). The lowest rated item was when the patient was asked to imagine the "overall face, eyes, mouth and nose" of the person, which the patient rated as 3 ("Moderately clear and lively").

Object Closure

The patient scored perfectly (4/4) on the Incomplete Letters task from the ACE-III. The patient demonstrated excellent performance on the Mooney Faces test, correctly identifying 19 out of 20 items. The error was made on item 17, but the patient self-corrected. This item had a difficulty higher than the average published stimuli, with a 72% of correct responses, compared to the average of 88.5% obtained by non-neurological participants (Schwiedrzik et al., 2018).

Facial Feature Processing

In the facial feature classification task, the patient scored perfectly (20/20) for age discrimination and nearly perfectly (19/20) for gender discrimination, with one initial error that was subsequently corrected.

Clinical Judgment

Based on the Birthday Party Test performance, our qualitative analyses concluded that: 1) the participant described the picture accurately based on both details and the whole, 2) some encouragement was needed regarding the whole picture description, in particular about the type of scene that it was (i.e. a birthday party), 3) the encouragement was very helpful in improving the description, and 4) the possibility of simultanagnosia was deemed "very unlikely."

Overall, the patient's performance across these tests suggests intact global processing abilities and no clear evidence of simultanagnosia. The patient demonstrated good performance in tasks requiring integration of complex scenes, object closure and recognition, and facial feature processing. The patient also showed the classical congruency effect while attending the local level of hierarchical letters, but not during attention to the global level. While some initial difficulties were noted in describing the whole scene in the BPT, these were readily overcome with minimal encouragement.

Discussion

This study aimed to evaluate the potential presence of simultanagnosia in a prosopagnosic patient with atypical lesion patterns and disrupted connectivity between

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visual and parietal areas. The comprehensive neuropsychological assessment revealed several key findings that shed light on the patient's visual processing abilities and the nature of their face recognition deficits.

Contrary to our initial hypothesis, the results strongly suggest that simultanagnosia is not a significant contributing factor to this patient's prosopagnosia. The patient demonstrated intact global processing abilities across various tasks with different levels of difficulty, particularly in the integration of complex scenes and hierarchical figure recognition. This finding is crucial, as it indicates that the face recognition deficits observed in this case are likely not due to a general impairment in processing multiple elements of a visual scene simultaneously, which is characteristic of simultanagnosia (Dalrymple et al., 2013).

The patient's performance on the Birthday Party Test (BPT) and the Cookie Theft picture was particularly informative. Despite initial hesitation in describing the whole scene in the BPT, the patient was able to overcome this with minimal encouragement, ultimately performing at or above the level of healthy controls. This suggests that while there might be a slight tendency towards local processing, the patient retains the ability to integrate information globally when prompted. This pattern is inconsistent with the severe global processing deficits typically seen in simultanagnosia (Dalrymple et al., 2013).

The excellent performance on tasks requiring facial feature processing, such as age and gender discrimination, is intriguing given the patient's prosopagnosia. This dissociation between intact facial feature processing and impaired face recognition

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supports the idea that prosopagnosia in this case may be more related to higher-level integration of facial features or accessing facial memories, rather than a fundamental deficit in perceiving facial elements (Barton & Corrow, 2016).

The patient's ability to recognize Mooney faces, which require holistic processing due to their high-contrast, two-tone nature, further argues against a global processing deficit. This finding aligns with research suggesting that some prosopagnosic patients retain the ability to perceive the global form of faces while still struggling with individual face recognition (Rossion, 2008).

The intact performance on visual imagery tasks, as assessed by the VVIQ, is noteworthy. Patients with posterior cortical atrophy (PCA) commonly show both deficits in visual imagery (Dietz et al., 2023) and simultanagnosia (Maia Da Silva et al., 2017). Nevertheless, lesions that cause simultanagnosia may co-occur with certain types of visual imagery deficits but not others, e.g. deficits in imagery for mental rotation or spatial processing, but not in imagery for color and shape (Foley et al., 2020). Our patient showed neither type of impairment, narrowing their deficit to a highly specific impairment for face perception, with possibly spared visual imagery for faces. This dissociation between face perception and face imagery further supports the complexity of face processing networks and the potential for selective impairments within this system.

Given these findings, we must consider the role of white matter tract damage in our patient's prosopagnosia. The lesions to both the ILF (23% of volume) and VOF (60% of volume) likely disrupt the connectivity within the face processing network (Bobes et al., 2021). While the ILF connects occipital visual areas with temporal regions and high-level visual areas within the occipito-temporal region, the VOF damage may impair communication between ventral and dorsal visual streams (Yeatman et al., 2014). This pattern of disconnection could explain the apperceptive nature of our patient's prosopagnosia, disrupting the integration of facial features and configural face processing.

The disrupted connectivity between visual and parietal areas observed in this patient might lead to face recognition deficits even in the absence of damage to core face-selective regions like the FFA and OFA (Avidan & Behrmann, 2009). Kay & Yeatman (2017) suggest that parietal areas may modulate the size of perceptual receptive fields when attending to faces in healthy subjects via the VOF. Our patient's deficit with faces may thus stem from impaired modulation of spatial processing specific to faces, rather than a general deficit in global visual processing.

The absence of simultanagnosia symptoms in our patient, despite the ventral-parietal disconnection caused by damage to the vertical occipital fasciculus (VOF), can be explained by several factors. One possibility is that simultanagnosia typically requires more extensive or bilateral damage to ventral-parietal connections, and the unilateral lesion in our patient may not be sufficient to produce detectable symptoms. This is supported by research showing that simultanagnosia is often associated with bilateral parietal lesions or diffuse cortical atrophy (Chechlacz et al., 2012). Furthermore, the preservation of other white matter pathways or cortical areas involved in visual attention and spatial integration could be mitigating the effects of the VOF disconnection. For instance, the intact superior longitudinal fasciculus (SLF) might be compensating for the

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VOF damage, as the SLF is known to play a crucial role in spatial attention and visual processing (de Schotten et al., 2011).

This dissociation between prosopagnosia and the absence of simultanagnosia underscores the complexity of neural networks involved in visual processing and highlights the need for further research on the relationship between brain connectivity and specific visual deficits. It suggests that the brain's visual processing system may have redundant pathways or compensatory mechanisms that can maintain global visual integration even in the face of specific white matter tract damage.

This case highlights the complexity of face recognition processes and the importance of considering network-level disruptions in addition to focal lesions when studying prosopagnosia. It also underscores the value of comprehensive neuropsychological assessments in differentiating between various visual processing deficits and understanding the specific nature of face recognition impairments in atypical cases.

Future research could benefit from combining detailed behavioral assessments with advanced neuroimaging techniques to further elucidate the relationship between structural connectivity, functional activation patterns, and face recognition abilities in prosopagnosic patients with atypical lesion patterns.

In conclusion, while our patient presents with prosopagnosia, the neuropsychological profile does not support the presence of simultanagnosia. The face recognition deficits in this case are more likely attributable to disrupted connectivity within the face processing network rather than a general impairment in global visual processing. This study contributes to our understanding of the diverse presentations of prosopagnosia and emphasizes the need for nuanced approaches in diagnosing and understanding face recognition disorders.

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Data availability: The data that support the findings of this study are available on request from the corresponding author, MM. The data are not publicly available due to confidential restrictions, e.g. their containing information that could compromise the privacy of research participants.

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