
Localization of Hemiachromatopsia

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Abstract

Impaired color perception with relative preservation of form vision (achromatopsia) caused by cerebral lesions was first described over a century ago. However, for many years some researchers questioned the existence of an area of cerebral cortex apart from the primary visual cortex specialized in color processing. The development of sophisticated structural and functional neuroimaging techniques has allowed verification of the cortical structures important in color perception. We describe a case of a patient with impaired color perception in one hemifield of vision (hemiachromatopsia) and compare the localization of the lesion with previous cases described in the literature. These cases show that lesions of the ventromedial occipital cortex can impair color perception and leave form vision intact.

Introduction

Central achromatopsia is a disorder of impaired color perception caused by an acquired cerebral lesion (Tranel, 1998). In 1888, Verrey reported a case of hemiachromatopsia that on post-mortem examination had a lesion located in the fusiform and lingual gyri of the contralateral inferior occipital cortex implicating these cerebral areas as specialized in color processing (Zeki, 1990). Even though several cases of central achromatopsia were reported over the ensuing decades, many researchers did not accept that an area of cerebral cortex specialized in color processing existed outside of the primary visual (striate) cortex. The reasons for questioning the concept of central achromatopsia included lack of standardized color testing techniques and lack of pathological data in some cases. Some authors also believed that tests of color perception were simply a more sensitive method of detecting visual field deficits (or impairments of form vision). Therefore, the lesions responsible for the loss of color vision were likely to be in the same areas associated with visual field defects, i.e. the primary visual cortex or optic radiations. However, in 1974, Meadows resurrected the hypothesis that the inferior occipital cortex is particularly specialized in color perception, based on a review of reported cases of central achromatopsia. All of the cases had visual field defects involving the upper visual quadrants but never exclusively the lower visual quadrants (Meadows, 1974). If the color vision center was in close proximity to the inferior striate cortex or inferior optic radiations then this could explain why lesions causing achromatopsia also often cause upper visual quadrant defects. A year previously, Zeki had demonstrated that certain cortical areas of the monkey may be preferentially responsive to color stimuli by showing pre-striate area V4 contained regions

that responded differentially to various wavelengths of light (Zeki, 1973, 1977). However, area V4 seemed to be located much more superiorly in the occipital cortex than the lingual and fusiform gyri and thus, the homologous area in humans, if it existed, still needed confirmation.

The advent of improved structural and functional neuroimaging techniques in the last two decades has allowed further definition of the cortical areas involved in color processing in humans. These techniques seem to confirm that the lingual and fusiform gyri in humans contain the homologue of area V4 in monkeys. We will review the neuroimaging of the previously reported cases of achromatopsia and our own case to localize further the cortical areas in humans most important in color perception.

Case

A 68-year-old right-handed woman was referred for evaluation of memory. She had been diagnosed several weeks prior with a stroke presumed to be cardioembolic from atrial fibrillation. Her only complaint was that she could not read and had no other neurological symptoms except for occasional mild headaches. She was a retired office worker with a high school education and she had been an avid reader prior to her stroke. She had a remote history of nephrectomy for kidney cancer and hysterectomy and oophorectomy.

On neurological examination she scored 24/30 on the Mini-Mental State Examination (MMSE) missing four points on orientation and two on delayed recall. Her reading was slow and she had to resort to spelling out many words,

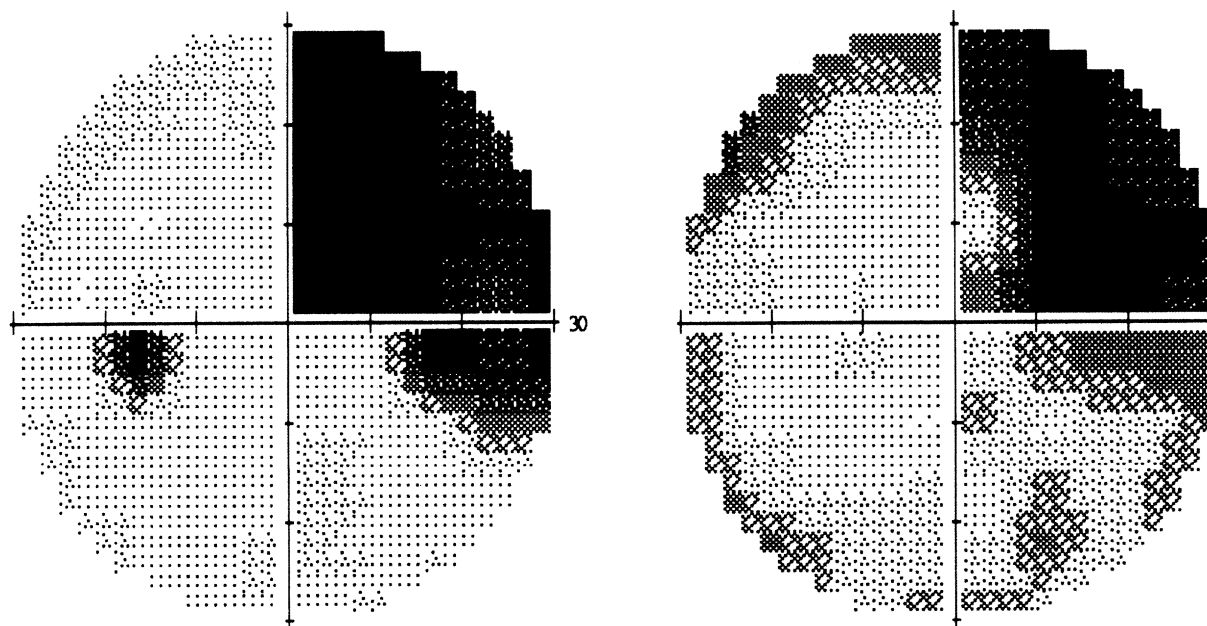


Fig. 1. Humphrey visual fields.

letter by letter. There was no finger agnosia, dysgraphia or acalculia. She had some mild difficulty in naming objects within the room, but otherwise had no aphasia. There was no visual field deficit to confrontation, nor did she have simultanagnosia, optic ataxia or optic apraxia. However, she described bright colors (red, green, blue, and yellow) presented in the right hemifield of each eye to be gray and could not identify the colors. She was surprised that the colors appeared gray as she had not noticed it prior to the examination. Thirty minutes later, when asked about visual problems, she acknowledged something was wrong with her vision but she did not know what it was. She could name colors correctly in the left hemifield. Motor and sensory examination, reflexes, coordination, gait and station were all normal. The color perception deficits on examination were reproduced 3 weeks later and 1 year later on follow-up visits, indicating a stable deficit.

On neuro-ophthalmological examination she had a right superior quadrantanopsia (Fig. 1). Visual acuity was 20/30 OD and 20/25 OS. She was able to identify nine of 10 Hardy-Rand-Rittler (HRR) color plates in all except the right superior quadrant. No other ophthalmological abnormalities were found except for mild bilateral nuclear sclerotic cataracts. The neuropsychological testing results are summarized in Table 1, which shows impairments in general intellect, memory and language. On the Wide Range Achievement Test Revision 3 (WRAT3) subtests of reading and spelling, she scored in the 30th and 68th percentiles, indicating more difficulty with reading than writing (i.e. partial alexia). The responses on the Boston Naming Test were circumlocutory without paraphasic errors and without much benefit from phonemic cues. The responses indicated intact knowledge about objects. Examples of her responses include: whistle—‘something you blow into’, when given

phonemic cue—‘not a whistle’; scissors—gestures correctly with hands and answers ‘not scissors but in the family’; broom—gestures sweeping motion and answers ‘what you would do to a floor’, answered correctly when given phonemic cue. Generative naming showed above average scores on letter fluency but impaired in category fluency. Her copy of the Rey-Osterrieth figure was accurate in the parts completed but she received a low score due to time limits. She did not have neglect on a letter cancellation test.

Magnetic resonance imaging (MRI) with coronal cuts parallel to the plane of the face showed an area of ischemic infarction in the left medial occipital/temporal cortex (Fig. 2). Also present were areas of subcortical ischemia and a small area of infarction in the left sensory cortex. Routine laboratory evaluation, transesophageal echocardiogram and magnetic resonance angiography were unremarkable.

Mapping of the MRI lesion to coronal templates (90° angle to orbitomeatal line) in a previously described technique (Damasio and Damasio, 1989), revealed an area of infarction in nearly all of the fusiform gyrus and the lingual gyrus and smaller portions of the parahippocampus and hippocampus (Fig. 3). This corresponded to Brodmann areas 18, 19, 28, 36, and 37. Spared areas included the supracalcarine cortex, forceps major, and paraventricular area. The area of infarction correlated to the distribution of the lateral branch of the left posterior cerebral artery. Figure 4 is a photograph of the ventral occipital and temporal cortex of a gross pathological specimen with the lingual and fusiform gyri highlighted.

Discussion

Previous cases of achromatopsia reported with neuroimaging findings are summarized in Table 2 (Green and Lessell, 1977; Pearlman *et al.*, 1979; Damasio *et al.*, 1980; Kolmel, 1988;

Table 1. Neuropsychological scores

Parameter	Raw or standard score	Descriptor/percentile
Wechsler Adult Intelligence Scale – Revised ^a		
Verbal IQ	78	7%
Performance IQ	78	7%
Full-scale IQ	76	5%
Wide Range Achievement Test (Revision 3)		
Reading	92	30%
Spelling	68	25%
Arithmetic	85	16%
Rey–Osterrieth Complex Figure, copy	26	<1%
Random Letter Cancellation Task		
Right	29	Normal
Left	30	Normal
Boston Naming Test	8	<1%
Letter fluency ^b (C, F, L)	41	72–81%
Category fluency ^b (animals, fruits, vegetables)	22	3–5%
Trail Making Task ^b , part A	67 s	2%
Trail Making Task ^b , part B	>5 min	<1%
Wisconsin Card Sorting Test		
Perseverative responses	43	6%
Categories	3	>16%
Failure to maintain set	0	>16%
Wechsler Memory Scale–Revised ^b		
Logical memory I	6	Severely impaired
Logical memory II	0	Severely impaired
Auditory Verbal Learning Test ^b		
Delayed recall	0	Severely impaired
Brief Visual Memory–Revised		
Delayed recall	2	Severely impaired

^aAge-corrected standard scores (mean, 100 ± 15) for factor scores of Wechsler Adult Intelligence Scale using Mayo's Older Americans Normative Studies standard scores (Ivnik *et al.*, 1992a, b, c).

^bAge-corrected scaled scores (mean, 10 ± 3) using Mayo's Older Americans Normative Studies scaled scores (Ivnik *et al.*, 1996; Lucas *et al.*, 1998).

Victor *et al.*, 1989; Rizzo *et al.*, 1992, 1993; Paulson *et al.*, 1994). Alexia and prosopagnosia are common additional neurobehavioral abnormalities. All but two cases were caused by an ischemic infarction. The cause of the infarction was not reported in several of the studies, although both of Paulson's cases were presumed cardioembolic (Paulson *et al.*, 1994). Consistent with Meadows' review, visual field defects are common, and when present always involve the superior visual fields. The visual defects usually do not involve the entire quadrant or hemifield, but spare central fixation. Kolmel's two cases are particularly important as they demonstrate that achromatopsia can occur without visual field deficits (Kolmel, 1988).

Although different anatomical terms are used in the previous reports, some comparisons can be made. The lingual gyrus and the posterior fusiform gyrus occupy the inferior and medial aspects of the occipital lobe below the calcarine fissure. The fusiform gyrus is sometimes referred to as the medial occipital temporal gyrus. Brodmann areas 18 and 19 are visual association areas above and below the calcarine cortex (area 17). Both the lingual and posterior parts of the fusiform gyrus contain areas 18 and 19. The common area of involvement in all of these cases is the ventromedial aspect of the occipital lobe, which comprises the lingual and posterior fusiform gyrus and areas 18 and 19. Although our

patient had a more extensive lesion in the fusiform gyrus extending anteriorly into the temporal lobe (area 37), based on previous cases, the area causing the hemiachromatopsia was located more posteriorly. All of the cases had some sparing of the primary visual cortex and the optic radiations, indicating that these areas are not responsible for the impairment in color perception. Therefore, our review implicates the lingual gyrus and occipital portion of the fusiform gyrus as the cortical areas most likely involved when there is impairment of color perception. This is consistent with the findings of previous authors that the caudal portion of the lingual gyrus is the area most commonly involved in central achromatopsia (Damasio *et al.*, 1991).

Functional imaging with positron emission tomography (PET) scanning provides further evidence that these areas are important in color processing. Lueck *et al.* (1989) showed the lingual and fusiform gyri in the occipital cortex to be selectively activated when individuals were shown abstract color (Mondrian) displays in comparison with black and white displays. McKeefry and Zeki (1997) have localized color-related activation even further using functional MRI (fMRI) studies and Mondrian displays. Their data show activation of the medial aspect of the posterior fusiform gyrus, which they termed human area V4. This area lies in the lateral bank of the collateral sulcus, which separates the

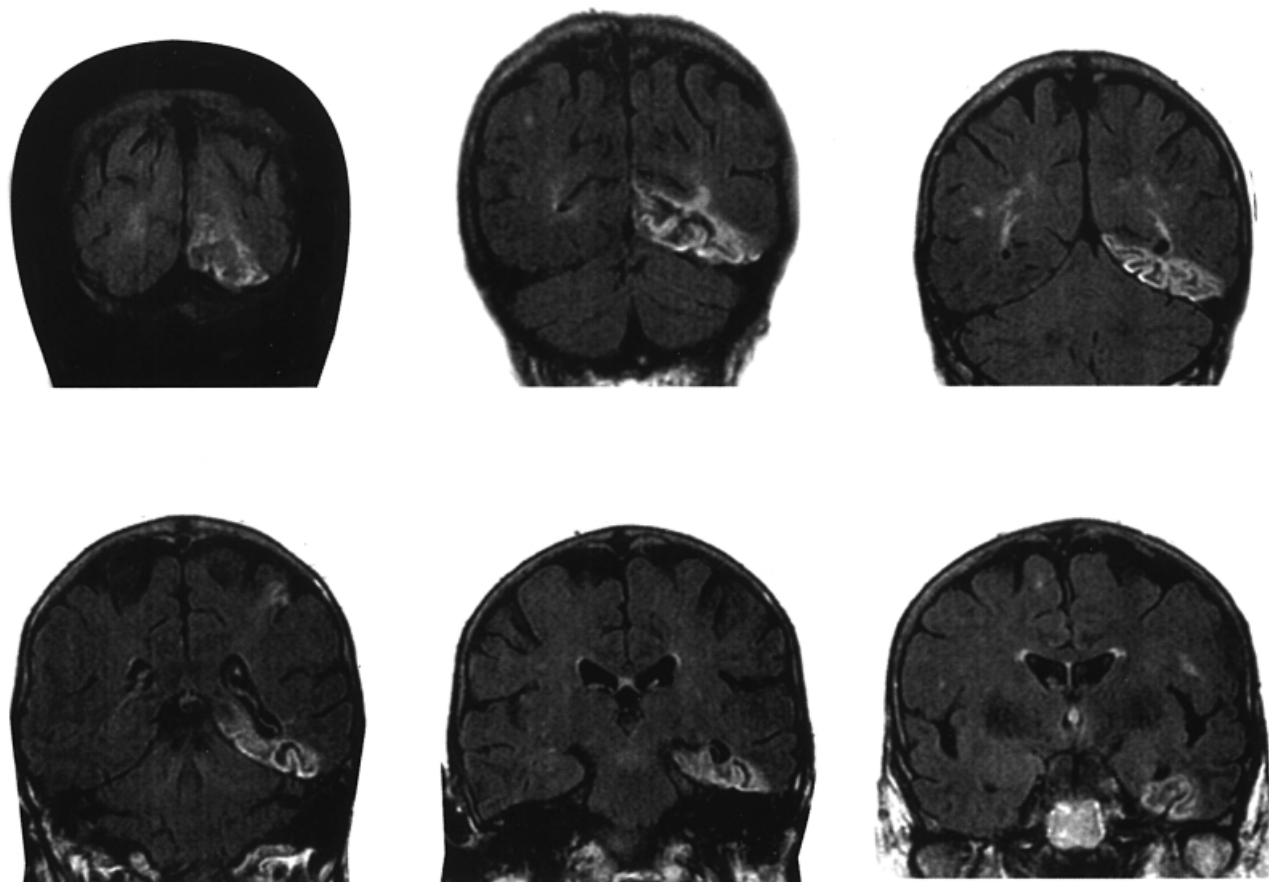


Fig. 2. Fluid attenuated inversion recovery (FLAIR) magnetic resonance imaging (MRI) images, coronal cuts.



Fig. 3. Templates corresponding to images in Fig. 2.

lingual and fusiform gyri. However, Beauchamp *et al.* (1999) have shown that more anterior areas in the fusiform gyrus and collateral sulcus are also activated on fMRI using passive viewing of the Farnsworth-Munsell 100-Hue test. Activation of these color-selective areas increases more than 3-fold when active sequencing of colors is required, suggesting an attention component. Beauchamp *et al.* (2000) have also reported similar testing on a patient with an incomplete achromatopsia who was able to name viewed colors but was impaired on the Farnsworth-Munsell 100-Hue test. On MRI the patient had ischemic lesions in the bilateral ventral occipitotemporal cortex with some preservation of more posterior areas on the left. These posterior areas were still activated in fMRI imaging, providing a nice correlate to the fMRI studies on normals. The results of both studies emphasize the importance of the posterior ventral occipito-

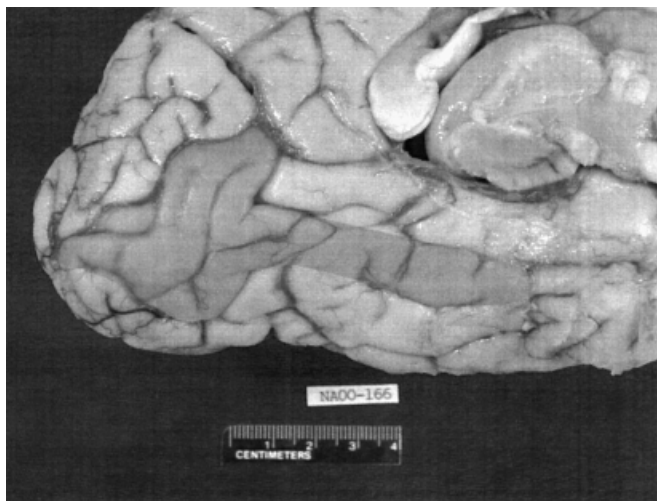


Fig. 4. Medial aspect of occipital and temporal cortex. The highlight shows the lingual gyrus and the fusiform gyrus.

temporal cortex in simple color perception tasks and the more anterior areas in more complex color perception tasks.

A few other characteristics of our case deserve mention. Neither of Paulson's two patients was aware of the color deficits and both had left hemiachromatopsia and superior quadrantanopia with lesions involving the right lingual and fusiform gyri. The authors postulated that this could be a form of anosagnosia related to a right hemisphere lesion (Paulson *et al.*, 1994). Our patient also had lack of awareness of the impaired color perception even after the deficit had been demonstrated to her, suggesting that this is not specific to right hemispheric lesions. This lack of awareness does not seem to be caused by visual neglect because she performed normally on the line cancellation test.

Our patient's color deficit could not be demonstrated with HRR color plates. Color plates are designed to test vision within a central few degrees of fixation, which explains the difficulty in demonstrating the hemiachromatopsia with the standard color plates (Damasio *et al.*, 1991). Therefore, when testing for a partial central achromatopsia, using brightly colored objects and moving them from one visual quadrant to another is the preferred method (Damasio *et al.*, 1991) even though this relies on the patient's subjective perception of color.

Our patient also had a combination of an anomia, partial alexia and amnesia. Although she was not tested for prosopagnosia, her responses on the Boston Naming Test did not indicate a visual object agnosia because she was able to provide circumlocutory responses or gestures, indicating intact knowledge of the picture objects. However, as shown in the review table, visual agnosia, particularly prosopagnosia, often accompanies central achromatopsia. Pure alexia also often accompanies hemiachromatopsia and in fact in Dejerine's classic 1892 case of pure alexia there was mention of impaired color perception in the right visual field (Damasio and Damasio, 1983).

Table 2. Summary of cases with achromatopsia and neuroimaging

Author	Achromatopsia	Visual field deficit	Associated findings	Neuroimaging	Lesion location	Cause
Green and Lessell, 1977	Bilateral	Bilateral	Alexia, prosopagnosia	CT	B occipital	Tumor
Pearlman <i>et al.</i> , 1979	Bilateral	LSQ	Color anomia, prosopagnosia, topographagnosia	CT	B lingual and fusiform	Infarct
Damasio <i>et al.</i> , 1980	Left	Left upper scotoma	None	CT	Right ventromedial occipital (18, 19, 19, 19)	Infarct
Damasio <i>et al.</i> , 1980	Bilateral	Left upper scotoma	Prosopagnosia	CT	B ventromedial occipital white matter (21, 36, 37, 19, 18)	Infarct
Victor <i>et al.</i> , 1989	Left	Right hemianopia	Anomia, alexia, amnesia	MRI	Left extensive occipital, right ventromedial occipital	Infarct
Kolmel, 1988	Right	None	None	MRI	Left medial OTG	Infarct
Kolmel, 1988	Left	None	None	MRI	Right lingual and medial OTG	Infarct
Rizzo <i>et al.</i> , 1992	Bilateral	Left hemianopia	Prosopagnosia	MRI	Extensive right occipital, left inferior occipital (18, 19)	Trauma
Rizzo <i>et al.</i> , 1993	Bilateral	Left hemianopia	Alexia, prosopagnosia	MRI	Extensive left occipital, right fusiform and posterior lingual	Infarct
Paulson <i>et al.</i> , 1994	Left	RSQ	None	MRI	Right fusiform, partial lingual	Infarct
Paulson <i>et al.</i> , 1994	Left	RSQ	None	MRI	Right fusiform and lingual	Infarct

LSQ, left superior quadrant; RSQ, right superior quadrant; B, bilateral; OTG, occipitotemporal gyrus; MRI, magnetic resonance imaging; CT, computed tomography.

In summary, our case and review of the literature demonstrate that the ventromedial occipital cortex is crucial to color processing and that lesions in this area can cause central achromatopsia. Visual field deficits and various other neuro-behavioral deficits commonly occur with the achromatopsia. The most common cause of the lesion is probably embolic ischemic infarction in the lateral branch of the posterior cerebral artery. Patients can sometimes be unaware of the color deficit. Therefore, the clinician must check for color perception in patients with lesions in this area to identify the impairment.

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Journal

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O227

Primary diagnosis of interest

Hemiachromatopsia

Author's designation of case

A 68-year-old right-handed woman

Key theoretical issue

- Left ventral occipitotemporal lesions can cause hemiachromatopsia with lack of awareness of the colour perception deficit

Key words: hemiachromatopsia; colour perception; ventromedial occipital cortex

Scan, EEG and related measures

Magnetic resonance imaging

Standardized assessment

Neuropsychology, visual fields

Other assessment

Color perception

Lesion location

- Left ventral occipitotemporal

Lesion type

Ischaemic

Language

English