

Pure Homonymous Hemiachromatopsia

Findings with Neuro-ophthalmologic Examination and Imaging Procedures

Hans W. Kölmel

Department of Neurology, Freie Universität Berlin, Spandauer Damm 130, D-1000 Berlin 19, Germany

Summary. The study describes neuro-ophthalmologic findings in two patients with brain infarction who developed homonymous hemiachromatopsia with resolution to pure homonymous achromatopsia in an upper quadrant. All other visual parameters were normal; only color perimetry was capable of demonstrating the visual disorder. The results are presented with special emphasis on the macular region. Computed tomography studies and magnetic resonance imaging revealed lesions in the caudal and medial occipitotemporal gyri as well as in adjacent cortical regions. The lesions were secondary to disordered circulation in a proximal occipitotemporal branch of the posterior cerebral artery. The anatomical findings and functional relations of color vision in man are discussed in the light of animal findings.

Key words: Homonymous hemiachromatopsia – Medial occipitotemporal gyrus – Color perimetry

Introduction

Pure homonymous hemiachromatopsia is rare among the visual disorders of central origin. Most studies of homonymous hemiachromatopsia in the literature report a rather large homonymous scotoma for white light (Albert et al. 1975; Green and Lessell 1977; Damasio et al. 1980), color agnosia (Meadows 1974), or color anomia (Zihl and von Cramon 1980). The current study presents clinical findings including extensive testing of color vision and results of imaging techniques in two patients with brain infarction and homonymous quadrant achromatopsia without any other visual impairment. The fundamental anatomical and functional relations of central color vision in man are discussed in the light of animal findings (Zeki 1969; van Essen and Zeki 1978; Michael 1978).

Methods

Both patients were physicians who gave informed consent to the numerous tests required and who were highly cooperative throughout the course of the study. Their participation and ability to concentrate allowed time-consuming testing. Follow-up examinations were performed several times during the first 3 weeks, at 6 and 12 weeks, at 6 months, and at 1 and 2 years.

Visual acuity was tested with a Snellen chart. Visual fields were documented with dynamic Goldmann perimetry using at least three isopters. Small targets with low luminosity were preferred. Visual fields were determined by computer-assisted static perimetry using the Octopus. Approximately 400 measurements were made at 73 points within 30° of the fixation point, giving a resolution of 6° in this visual field. Finally profile perimetry was made with a Humphrey field analyzer (distance between stimuli 1° on both eyes in the 45°/225° and 135°/315° meridians).

The technique described by Zihl and von Cramon (1980) was employed to determine critical flicker fusion fields for 15 and 25 Hz (stimulus: 116 min arc diameter, 320 cd/m² luminance). Object recognition (circles and squares) was tested in each hemifield as far as possible using the Tübinger perimeter.

Monocular dark adaptation was tested with the Goldmann-Weekers adaptometer. Stereoscopic vision was tested with the stereoeidometer designed by Monie (1948) and with Titmus tables. Visual neglect was demonstrated with the tests designed by Luria (1960), Albert (1973), and by Heilman and Valenstein (1979). Patients were tested for pattern recognition with the Benton Facial Recognition Test (1968). Finger pursuit movement of the eyes and optokinetic nystagmus using a rotating drum with vertical stripes were tested.

Examination of Color Vision

The patients were asked to identify and sort wood blocks painted with the four primary colors (Hering 1920) and to identify and sort Holmgren's colored wool strands (containing 30 pairs of thick colored wool strands). Ishihara-Color-Plates and the Farnsworth-Munsell 100-hue test (Farnsworth 1943) were also employed.

Standardized color perimetry was performed on each eye with the Tübinger perimeter. Blue (480 nm)

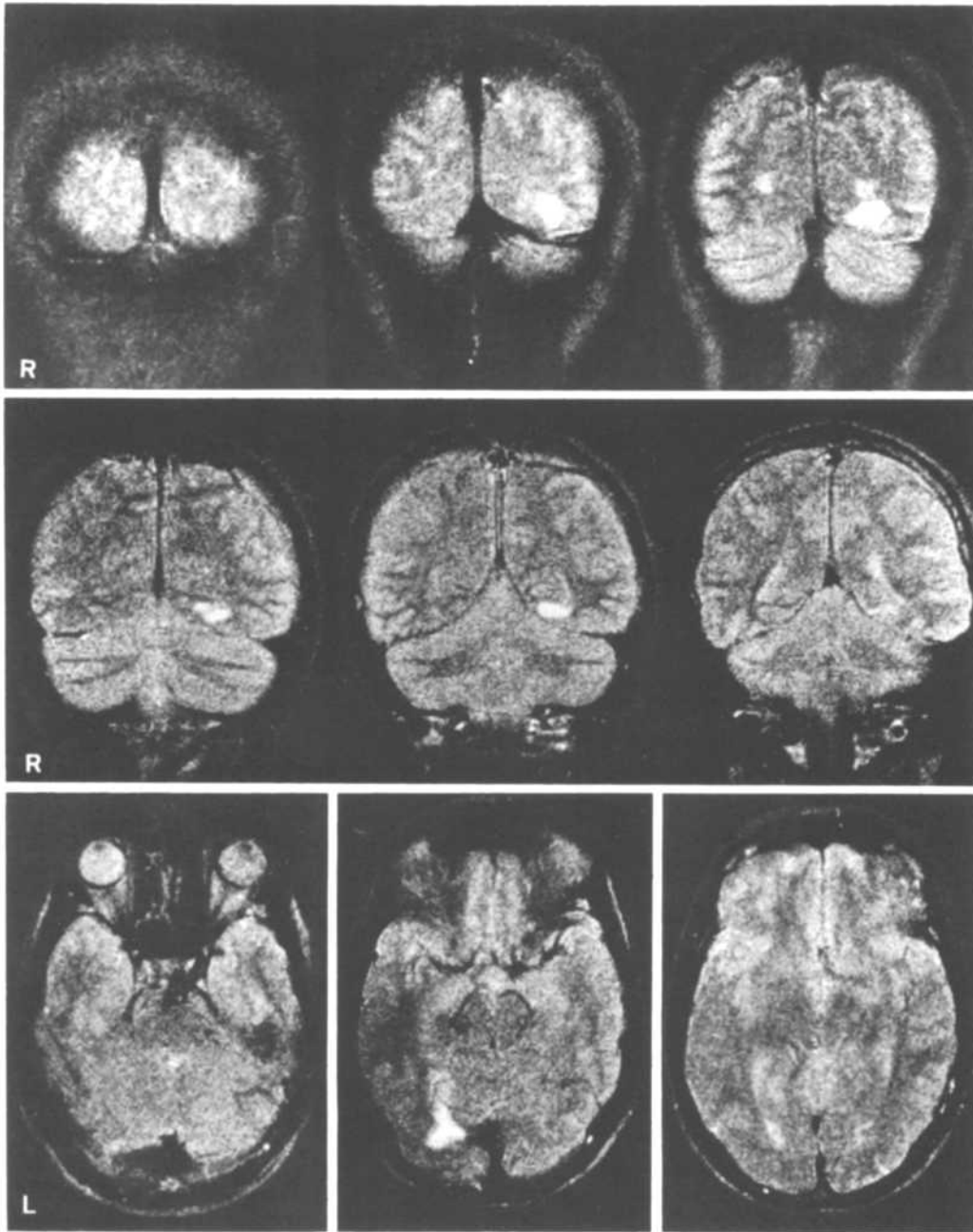


Fig. 1. Patient 1: Magnetic resonance imaging in the T2 mode. Increased signal intensity in the left occipitotemporal region corresponding to the medial occipitotemporal (MOT) gyrus

and red (656 nm) targets (116 min arc diameter, 320 cd/m² luminance) were used.

In a further test the patients sat at 2 m from a tangent screen, fixed the center of the screen, and attempted to identify the colors of wool strands presented in the periphery of the visual quadrants at 20°.

With the help of a second examiner color anomia was detected by simultaneous presentation of colored targets at the center and in the periphery of the visual field. In addition, the patients were asked to respond to the examiner's suggestions for sorting the colored

strands. All tests were conducted under identical ambient light conditions. Constant fixation of the center of the screen was verified.

In a subsequent examination isopters for white and colored targets were tested in the macular region.

Case Reports and Results

Case 1. The patient, a 34-year-old right-handed radiologist had observed occasional flickering scotomas in the right lower quadrant associated with fatigue for the past 7 years. There

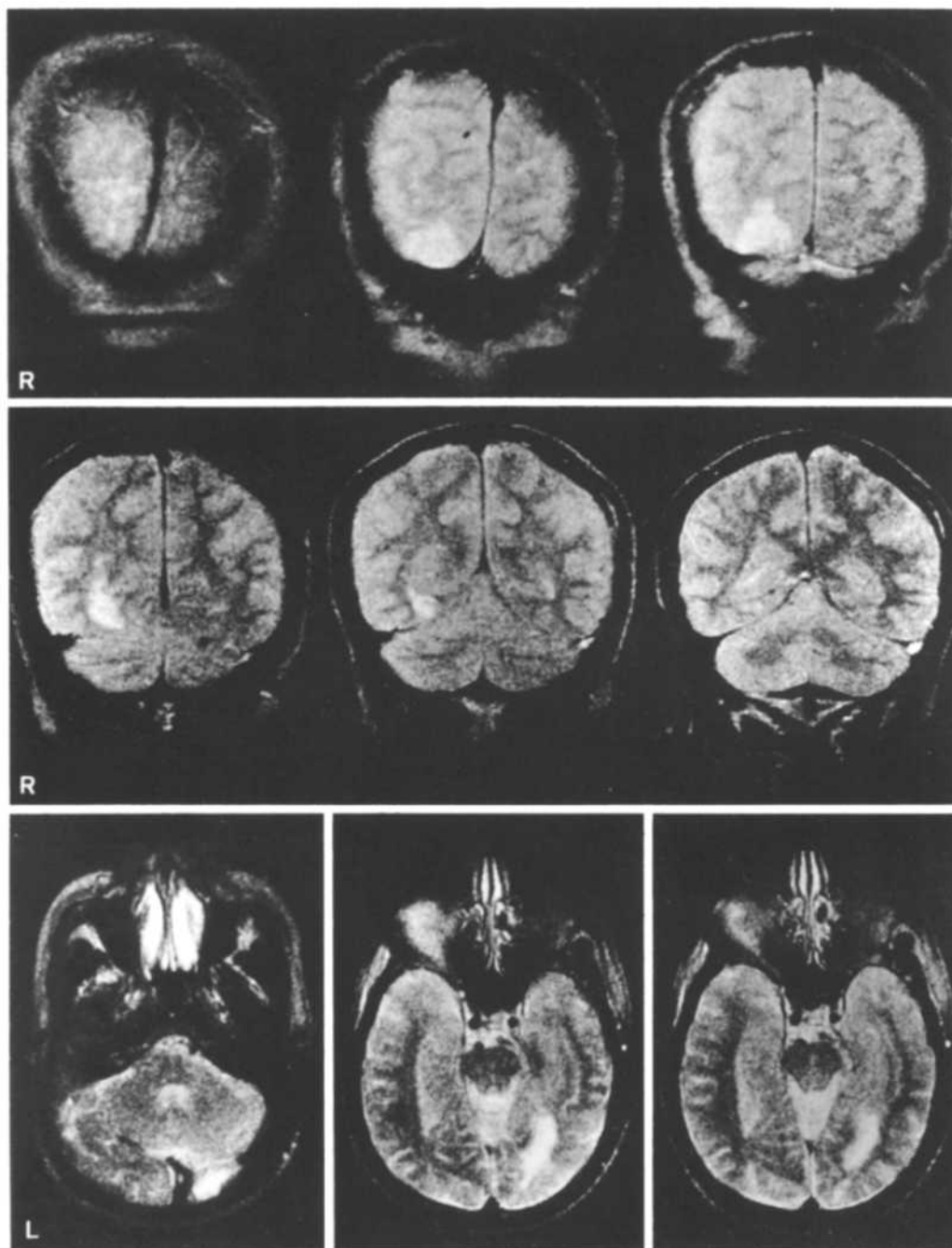


Fig. 2. Patient 2: Magnetic resonance imaging in the T2 mode. Same findings as in patient 1. Involvement of the rostral portion of the right lingual gyrus and the MOT gyrus

was no history of headache. One morning he suddenly felt pressure behind the left eye, followed by paraesthesia in the right arm and an unsteady gait. He examined himself in a mirror, noted that vision was limited to the left hemifield and made the diagnosis of right homonymous hemianopia.

Examination by a neurologist 1 day later documented normal findings with the exception of right homonymous hemianopia. The patient recognized his visual disorder and was fully oriented to time, place, and situation. Reading, writing and calculation were undisturbed.

The visual field defect quickly resolved to right upper quadrantanopia with entirely normal findings 6 days after the acute event. At 6 weeks Goldmann perimetry and computer-assisted perimetry with the Octopus showed normal findings. The patient complained of photophobia and easy fatigability during reading. Color perimetry revealed homonymous hemiachromatopsia in the right upper quadrant. The test results are given in Table 1 and Figs. 3–6. Initial EEGs documented slowing in the left temporal region. Computed tomography (CT) studies revealed that the lesion was located primarily in the

Table 1. Test results of the two patients with homonymous achromatopsia in an upper quadrant

	Patient 1 (6 weeks after onset of visual disorder)	Patient 2 (20 days after onset of visual disorder)
Visual acuity	1.2/1.2	1.0/1.0
Goldmann perimetry	Normal (Fig. 5a)	Normal (Fig. 6a)
Octopus perimetry	Normal	Normal
Profile perimetry	Normal*	Normal*
Color perimetry	Homonymous achromatopsia in the right upper quadrant (Fig. 5b), no change 2 years later	Homonymous achromatopsia in the left upper quadrant (Fig. 6b)**, no change 2 years later
Naming/separating mixed colors	No mistakes	No mistakes
Ishihara plates	No mistakes	No mistakes
Farnsworth-Munsell test	Slight to moderate disturbance (Fig. 3a), 2 years later: normal (Fig. 4a)	Slight to moderate disturbance (Fig. 3b), 1.5 years later: slight uncertainty in blue range (Fig. 4b)
Stereoscopy	Normal	Normal
Dark adaptation	Normal	Normal
Flicker fusion frequency	Equal on both sides	Equal on both sides
Visual neglect/prosopagnosia	No pathological findings	No pathological findings

* 2 years after onset of visual disorder

** 3 months after onset of visual disorder

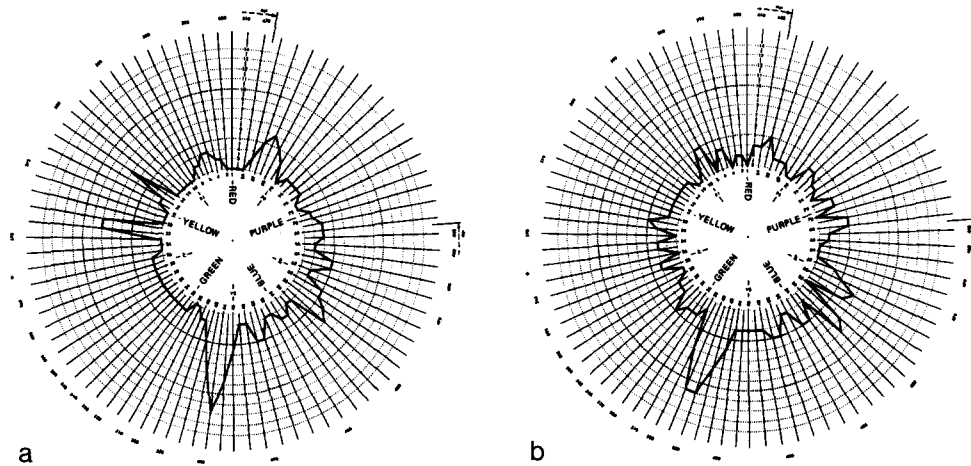


Fig. 3. **a** Results of the Farnsworth-Munsell test in patient 1 6 weeks after onset of the visual disorder. **b** Tests results in patient 2 20 days after onset of the visual disorder

occipital white matter. There was no evident involvement of area 17. The T2 weighted images in magnetic resonance imaging demonstrated increased signal intensity in the left occipitotemporal region corresponding to the location of the medial occipitotemporal (MOT) gyrus (Fig. 1). Increased signal intensity extended to the parahippocampal gyrus. Xenon clearance studies with single photon emission tomography showed reduced perfusion in the left occipitotemporal region.

Case 2. The patient, a 66-year-old right-handed surgeon with a 10-year history of diabetes mellitus controlled by diet, reported occasional palpitations. After a party he noted unsteady gait and a reduction in visual acuity. On admission to hospital the patient was fully conscious and aware of his surroundings. Neurologic examination demonstrated left hemianopia and in-

creased deep tendon reflexes on the left. The patient reported a slight disorder of spatial orientation and difficulty in recognizing faces during the first few days. 16 days later Goldmann perimetry and computer-assisted static perimetry demonstrated normal findings but color perimetry showed complete left homonymous hemiachromatopsia. The patient complained of easy fatigability during reading and found that the letters appeared to move before his eyes after a short period. He also felt blinded by bright light. At this point the initial problems with spatial orientation and facial recognition had resolved completely. Neurologic findings at 6 months and 2 years after the acute event were normal, as were the visual fields. Left homonymous hemiachromatopsia had improved to upper left quadrant achromatopsia. The test results are given in Table 1 and in Fig. 3–6. The patient was not able to assess the extent of

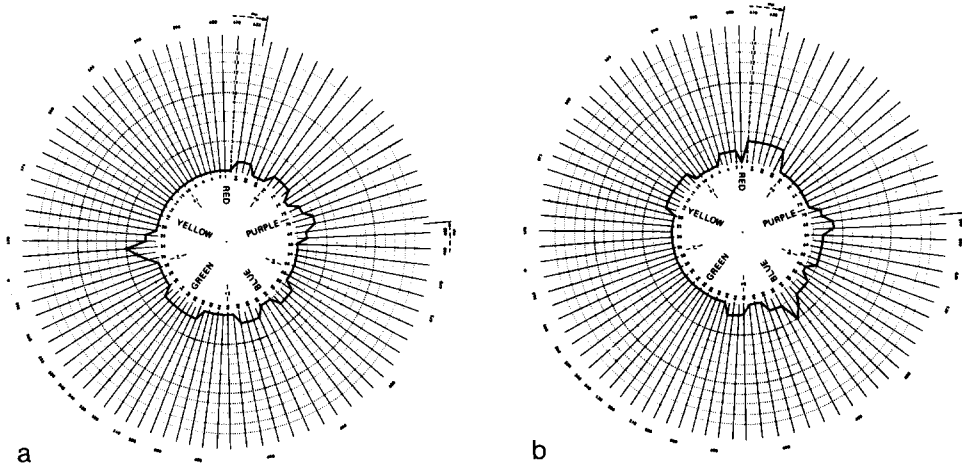


Fig. 4. **a** Farnsworth-Munsell test in patient 1 24 months after onset of the visual disorder: normal findings. **b** Farnsworth-Munsell test in patient 2 18 months after onset of the visual disorder: very slight uncertainty in the blue range

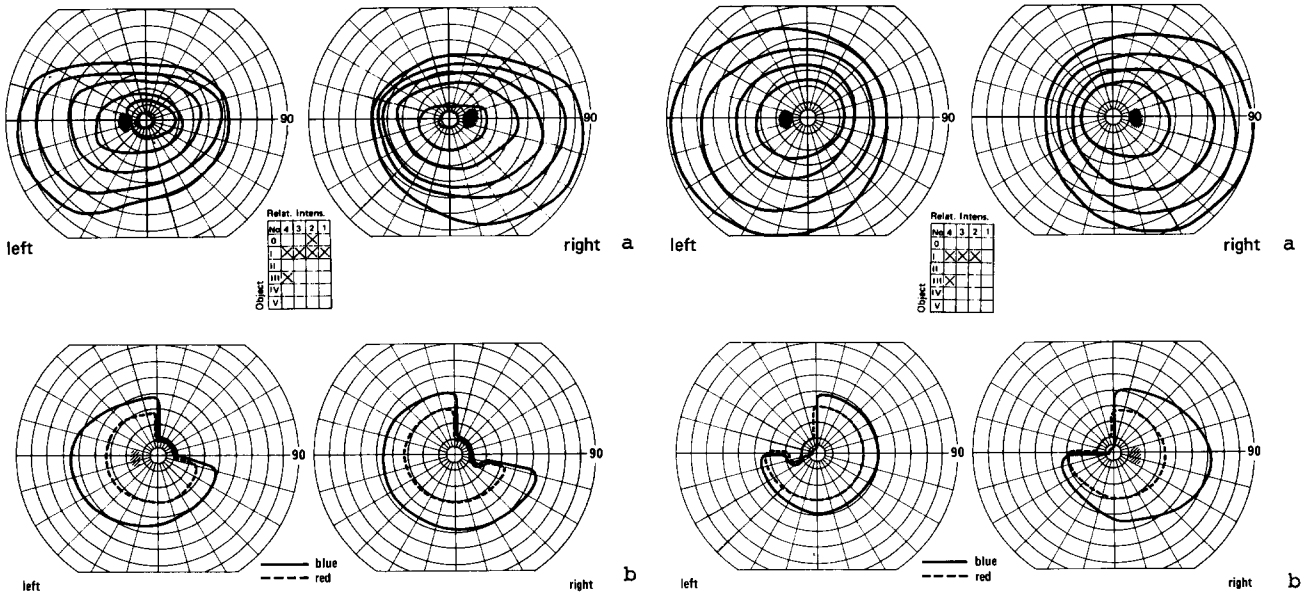


Fig. 5. (Patient 1) **a** Goldmann perimetry 6 weeks after onset of the illness: normal visual fields. **b** Color perimetry. Blue (480 nm), red (656 nm). Size of targets 116 min arc diameter. Luminance 320 cd/m². Achromatopsia of the right upper quadrant

Fig. 6. (Patient 2) **a** Goldmann perimetry 20 days after onset of the visual disorder: normal visual fields. **b** Color perimetry. Achromatopsia of the left upper quadrant with splitting of the macula. No change 30 months later

his visual disorder. He noticed a deficiency in the left visual field but was not aware of the disturbance of color vision. He reported that reading for long periods of time tended to tire him, but he returned to his professional activity as a surgeon 6 months after brain infarction.

The EEG documented temporary slowing in the right temporoparietal region. The CT studies revealed an oblong hypodense zone that appeared to be limited to the white matter in the right occipitotemporal region. Magnetic resonance imaging 6 months after the acute event demonstrated increased signal intensity in the right occipital lobe – white and grey matter – without apparent involvement of area 17 (Fig. 2). The lesion extended to the temporal region and corresponded roughly to the MOT gyrus. Xenon clearance studies showed reduced perfusion in this area.

Discussion

Reports from a number of experienced neuro-ophthalmologists may have contributed to the limited recognition of color perimetry. Wilbrand and Saenger (1917) reported a slight but constant reduction in light perception in hemiachromatopsias and suggested that conventional perimetry with white light would be sufficient in such cases. Brouwer (1936) and Critchley (1967) were in general agreement with this evaluation. Teuber et al. (1960) reported additional visual disorders in all patients with hemiachromatop-

sia. These included a reduction in flicker fusion frequency and disorders of dark adaptation. Harrington (1981) claimed that disorders affecting central color vision could be demonstrated with white light if the targets used were small enough.

However, many reports have shown that color perimetry is the best technique for demonstrating the presence and extent of unilateral disorders of visual perception. In some cases tests with white light and small targets showed normal visual fields, while color perimetry documented homonymous hemiachromatopsia or quadrantanopia. Bjerrum (1881) described a patient with a brain abscess in the temporobasal region who had normal visual fields and acuity but homonymous hemiachromatopsia. In the same year Samelsohn (1881) reported on a patient with homonymous left hemiachromatopsia due to a stroke with normal visual fields determined by white light and small targets. Bender and Kanzer (1939) described incomplete hemiachromatopsia as the only sequela of subarachnoid hemorrhage in a 23-year-old woman.

Color perimetry demonstrated total loss of color vision in an upper quadrant in both our patients. All other examinations produced normal findings, which underscores the particular value of color perimetry. The patients made no errors in sorting and naming colors when the undisturbed visual fields were tested. Color anomia such as that described by Geschwind and Fusillo (1965) was not present, nor was there color anomia in a half field such as that found by Zihl and von Cramon (1980) in their patient.

Only the Farnsworth-Munsell test showed relatively inconsistent pathological findings. Color perimetry in patient 2 demonstrated that the disorder for blue extended very close to the fovea. We had suspected that more pronounced disorders in the blue-green range demonstrated in the Farnsworth-Munsell test might be related to the visual field defect. These results are similar to those reported by Zihl and Meyer (1981). The patients described by Meadows (1974) and by Zihl and von Cramon (1980) also had pathological results in the Farnsworth-Munsell test, but these patients also suffered from color agnosia or anomia. Follow-up studies of color vision with the Farnsworth-Munsell test at 2 years after brain infarction showed normal findings in both patients despite persistence of disturbed color vision in an upper quadrant. This would tend to limit the value of the Farnsworth-Munsell test and to underline the importance of color perimetry.

Our results and those reported by other researchers (Zihl and Meyer 1981) suggest that the upper quadrants are particularly susceptible to disorders of color vision. Homonymous hemiachromatopsia often

resolves to upper quadrant achromatopsia. Isolated homonymous achromatopsia in a lower quadrant has never been reported as far as we know. These findings show that the lower quadrants are especially resistant to impairment of color vision. Location and extent of the brain lesions detected in our patients were practically identical. The findings in patient 2 extended farther into the occipital region than did the lesion in patient 1. The topographic findings underline the fact that the hemispheres are equally important for color vision in the contralateral hemifield. Areas 18 and 19 and marginal portions of areas 37 and 20 of the Brodmann classification were affected in these patients. These involved the rostral portion of the lingual gyrus, the entire caudal and median portions of the MOT gyrus, and perhaps adjacent areas of the parahippocampal gyrus. The CT studies demonstrated that the white matter was primarily affected, but magnetic resonance imaging showed approximately equal involvement of the cortex and white matter.

There are a number of possible variations in blood flow to the occipital lobe through the posterior and medial cerebral arteries. The MOT gyrus is also served by several branches of the posterior cerebral artery. It may be assumed that a proximal branch of the posterior cerebral artery – the temporooccipital artery – without effective collaterals was involved in both of our cases.

The location of the lesions causing hemiachromatopsia was a matter of controversy for many years (Wilbrand and Saenger 1917; Critchley 1967; Holmes 1918; Lenz 1921a, b), although the significance of the MOT gyrus for color vision has been known for a long time. Verrey (1888) was the first to suggest this relationship in his clinical and pathological study. Mackay and Dunlop (1899) and Lenz (1921a, b) described identical topographic relations, and CT studies have demonstrated lesions in the same region in patients with hemiachromatopsia. Disorders in this gyrus are found in almost all cases with disturbances of color vision, including hemiachromatopsia, color agnosia and anomia. When the latter two disorders are found, additional lesions in connective visual pathways such as the splenium of the corpus callosum or in the contralateral MOT gyrus must be assumed (Green and Lessell 1977; Damasio et al. 1980; Meadows 1974; Zihl and von Cramon 1980; Pearlman et al. 1979). The importance of the MOT gyrus – a part of area 19 – for color vision in the contralateral hemifield suggests that specific visual functions are concentrated in a region relatively far from the striate area. This corresponds to findings in rhesus monkeys reported by Zeki (1969), who demonstrated a high concentration of color-coded

neurons in area V4, which is located far from the primary visual fields. Other investigators were unable to find a high concentration in area V4 (Schein et al. 1982), and definition of the topographic limits of this extrastriate area has encountered a number of problems (Ungerleider and Mishkin 1979). The region described by Zeki is located near the lunata sulcus, and it is not easy to find a relationship between this region in monkeys and the MOT gyrus in man. Meadows (1974) has already discussed this discrepancy. In order to resolve the problem, Damasio et al. (1980) postulated that color-selective neurons are located in a wide belt outside the striate area. However, this does not alter the fact that a circumscribed lesion in the MOT gyrus is capable of causing complete hemiachromatopsia. Therefore, we assume that this region corresponds to the area V4 described by Zeki (1969).

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