
Hemiachromatopsia of Unilateral Occipitotemporal Infarcts

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Two patients developed unilateral occipitotemporal infarcts that produced inferior quadrantic achromatopsia and an accompanying superior quadrantanopia. Magnetic resonance imaging and single-photon emission computed tomographic studies of both patients supported the current view that color vision is encoded in the lingual and fusiform gyri. Although the quadrantic defect in color processing was profound, neither patient was aware of it. Simple bedside testing of patients with superior quadrantanopia may disclose an unrecognized quadrantic achromatopsia.

MORE THAN A CENTURY AGO, Verrey¹ described hemiachromatopsia, the loss of color vision in one hemifield, and claimed that a functionally separate cortex existed for color processing. However, it is only in the past 20 years that the notion of a neuroanatomically distinct region for color vision has been accepted.² Primate and human studies have designated a region of the visual association cortex known as V4 to be critical for color processing.³⁻⁹ In humans the V4 homolog resides in the ventromedial occipitotemporal cortex. The close proximity of this region to the inferior striate cortex suggests that hemiachromatopsia might commonly occur in patients with superior quadrant field defects. Using both single-photon emission computed tomography and magnetic resonance imaging, we studied the neuro-ophthalmologic and neuroanatomic findings in two patients in whom unilateral occipitotemporal infarcts produced inferior quadrantic achromatopsia and a superior quadrantanopia.

Accepted for publication March 18, 1994.

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Subjects and Methods

Single-photon emission computed tomography provides a measure of regional cerebral blood flow, corresponding to the uptake of an injected radiopharmaceutical. With both patients described in this study, the single-photon emission computed tomographic scan was initiated 20 to 30 minutes after the injection of 20 mCi of ^{99m}Tc-hexamethyl propylene amine oxime and was performed with a three-headed rotating single-photon emission computed tomography camera (Picker Prism; Picker International, Cleveland, Ohio) for 40 minutes by using high-resolution fanbeam collimators. Projection images were obtained at 3-degree angles on a 128 × 128 matrix over 360 degrees, and reconstructed in the transaxial, coronal, and sagittal planes (slice thickness, 10.7 mm). The images were generated by normalizing the maximal pixel counts to the maximal intensity of a signal density calibration bar, and thus represent a measurement of relative blood flow.

Case Reports

Case 1

In June 1992, a 56-year-old, right-handed man was hospitalized because of the onset of atrial fibrillation and sudden left visual field loss. He denied other neurologic symptoms, including weakness, sensory change, gait, or speech disturbance. Neurologic examination disclosed an isolated left homonymous superior quadrantanopia. Computed tomography of the brain and results of laboratory tests were unremarkable except for mildly increased cholesterol levels. A transthoracic echocardiogram did not disclose a thrombus. Because of the presumed diagnosis of a right occipital infarct that was cardioembolic in origin, he was discharged on warfarin therapy.

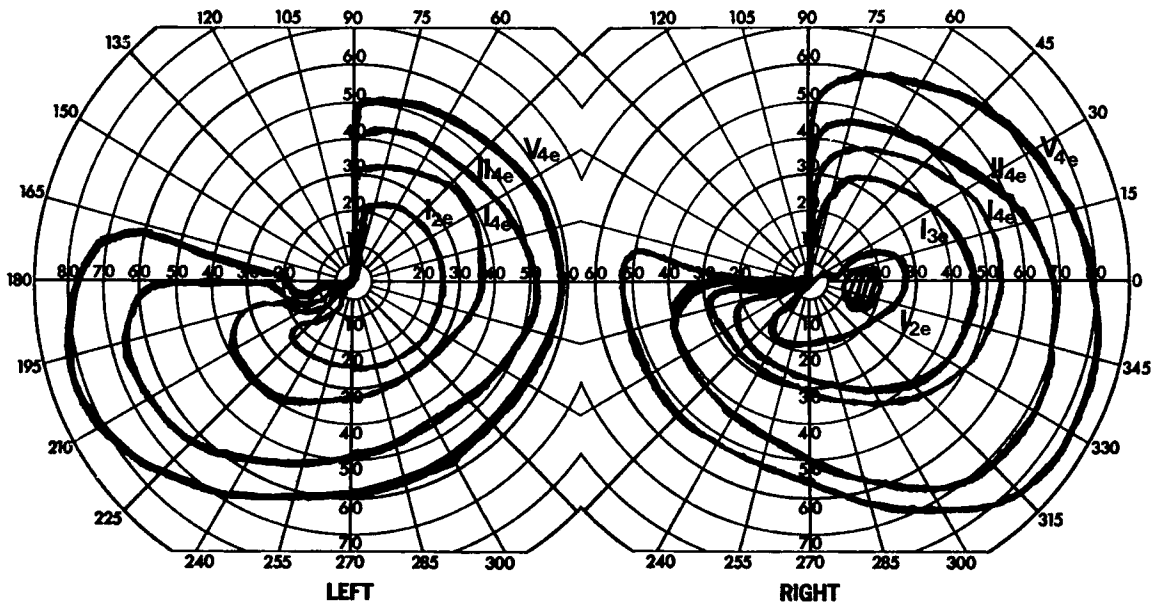


Fig. 1 (Paulson and associates). Goldmann perimetry in Patient 1, two months after his infarct, demonstrates a left homonymous superior quadrantanopia with an intact inferior quadrant.

When first seen by us two months later, he reported that his visual field loss had not changed. Neurologic examination disclosed a congruous left superior quadrantanopia, confirmed by Goldmann perimetry (Fig. 1). Results of the rest of the neuro-ophthalmologic examination were normal.

When he was seen again seven and ten months after the infarct, his superior quadrantanopia persisted and was confirmed by repeat Goldmann perimetry. Light detection and form vision were normal in the left inferior quadrant, but color vision in this quadrant was profoundly disturbed. When vision was fixed at a central point on a white background, the patient could not sort colored threads or identify the color of objects in the left inferior visual quadrant. In this quadrant, objects and threads were most frequently described as "washed out" or various hues of gray. When colored threads or objects crossed the midline, he immediately identified their color. Only one of 25 colored threads (orange) ranging from red to indigo was correctly named, and only one of 15 attempts to sort pairs of isoluminescent colored threads was successful in the left inferior field. Even when he was made aware of this marked disturbance in color processing, he could not retrospectively identify any problem with color vision. Results of the rest of the neurologic examination were normal. Specifically, there

was no neglect, prosopagnosia, object agnosia, or akinetopsia.

Magnetic resonance imaging six months after the onset of his visual disturbance demonstrated an infarct in the inferomedial right temporal lobe centered in the fusiform gyrus, partially involving the lingual gyrus, and sparing the occipital pole (Fig. 2). A lacunar infarct in the right thalamus was also seen.

Seven months after the infarct, ^{99m}Tc -hexamethyl propylene amine oxime single-photon emission computed tomographic scanning was obtained to determine the extent of the perfusion defect corresponding to the infarct. The single-photon emission computed tomographic scan showed a prominent perfusion defect spanning the right ventromedial occipital and inferomedial temporal lobes (Fig. 2).

Case 2

In January 1993, a 76-year-old, right-handed woman with a history of hypertension and diabetes mellitus developed a right temporal headache and a feeling that her vision was "not right." Three days later she came to the hospital because of dizziness, transient left-sided clumsiness, and persistent visual disturbance. A head computed tomography revealed a subacute right occipitotemporal infarct, and an electrocardiogram showed atrial fibrillation.

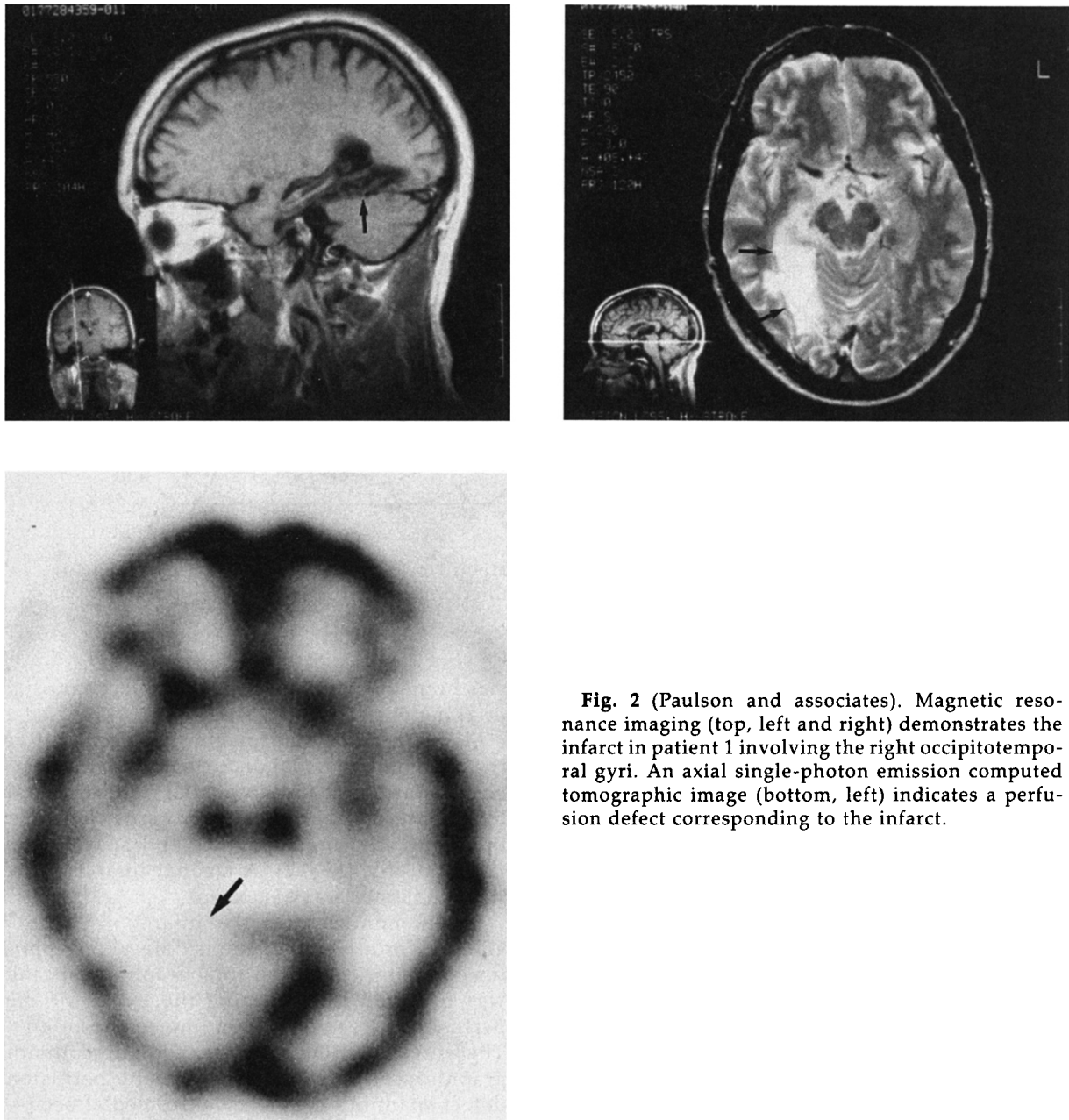


Fig. 2 (Paulson and associates). Magnetic resonance imaging (top, left and right) demonstrates the infarct in patient 1 involving the right occipitotemporal gyri. An axial single-photon emission computed tomographic image (bottom, left) indicates a perfusion defect corresponding to the infarct.

Although the patient recognized her son, she thought his face looked "foreign" to her. She had some difficulty recognizing and describing complex visual scenes, but named individual objects and people correctly. There was a congruous left superior quadrantanopia, confirmed by Goldmann perimetry. Light detection and form vision were normal in the left inferior quadrant, but color vision was markedly abnormal in this quadrant. She could not sort colors or name the color of objects in the left inferior

quadrant but immediately recognized colors once they crossed the midline. Our clinical testing made her aware of this color disturbance; however, she, like the first patient, could not retrospectively identify any noticeable change in color vision since the infarct. Ocular pursuit movements to the right were saccadic. There was a mild left sensory drift without neglect along with a moderate stocking-and-glove peripheral neuropathy.

Magnetic resonance imaging (Fig. 3) obtained

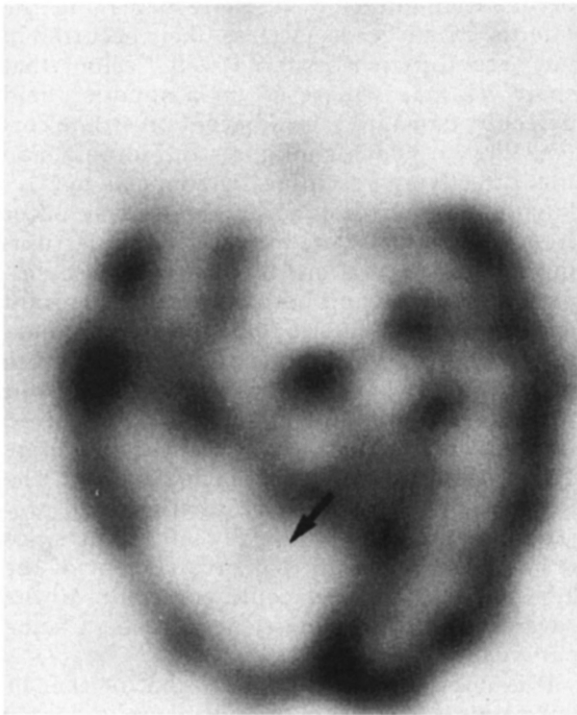
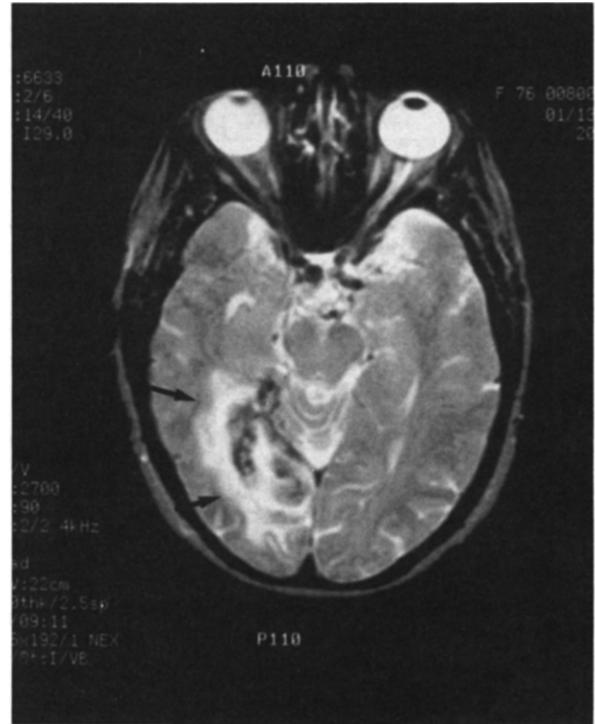
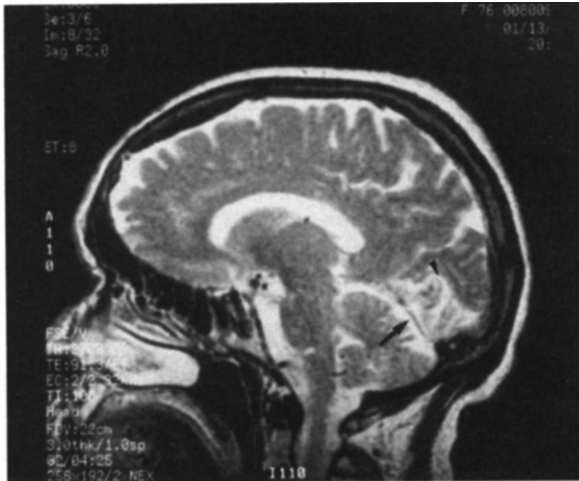


Fig. 3 (Paulson and associates). Magnetic resonance imaging (top, left and right) demonstrates the infarct in Patient 2, involving the right occipitotemporal gyri. The sagittal view demonstrates that the infarct extends posteriorly to include the lingual gyrus (arrow), but does not extend beyond the calcarine sulcus (arrowhead). An axial single-photon emission computed tomographic image (bottom, left) illustrates a perfusion defect corresponding to the infarct.

four days after admission showed a subacute hemorrhagic infarct involving the right inferomedial temporal and occipital lobes, extending dorsally to the calcarine fissure. The infarct involved the lingual and fusiform gyri as well as adjacent white matter. There were also several previously noted small areas of signal abnormality in the brain stem, left caudate nucleus, and periventricular and subcortical white matter, all consistent with prior small-vessel disease. $^{99\text{M}}\text{Tc}$ -hexamethyl propylene amine

oxine single-photon emission computed tomographic scan of the brain (Fig. 3) several days later disclosed a prominent perfusion defect in the right ventromedial occipital and inferomedial temporal lobes.

Laboratory studies disclosed increased glucose, the presence of sickle cell trait, and a microcytic anemia. Carotid and transcranial Doppler studies showed mild to moderate plaque in both carotid systems, and normal flow in the posterior circulation. A transesoph-

ageal echocardiogram disclosed a left atrial thrombus, supporting the clinical impression that her infarct was cardioembolic.

Discussion

As a result of unilateral ventromedial occipitotemporal infarcts, both of our patients developed contralateral inferior quadrantic achromatopsia and superior quadrantanopia. Achromatic vision was preserved in the inferior quadrant, indicating the defect in color processing was not caused by a lesion in the primary visual cortex, but rather by one in the prestriate visual association cortex. Although in both patients the quadrantic achromatopsia was profound, both patients were unaware of their problem with color vision until bedside testing revealed it to them. These two patients allow us to make several points about the manifestation and localization of color processing in the human central nervous system.

Although a designated area for color vision processing has recently been challenged,^{10,11} our patients confirm that a lesion inferior to the calcarine fissure produces a color vision deficit involving the entire hemifield. The location of our patients' infarcts suggests that the human homolog of V4 resides in the fusiform and lingual gyri. Even in Verrey's original 1888 report of a patient with hemiachromatopsia, the autopsy showed damage to these two gyri.¹ Most recently reported cases of central achromatopsia (both complete and hemiachromatopsia) have demonstrated bilateral cortical injury.^{3,4} There are surprisingly few high-resolution radiographic images of unilateral lesions causing hemiachromatopsia. The most compelling is Kolmel's description⁶ of two patients with pure hemiachromatopsia, in which magnetic resonance imaging disclosed discrete lesions within the fusiform and lingual gyri. Similarly, in our first patient the magnetic resonance imaging showed an infarct that involved the fusiform gyrus and, to a lesser degree, the lingual gyrus. Although our second patient's infarct was larger, it still encompassed the entire fusiform and lingual gyri as well as adjacent white matter. In both patients, brain single-photon emission computed tomographic scans corroborated the magnetic resonance imaging findings. The single-photon emission computed tomographic studies convincingly documented that the quadrantic achromatopsia was

associated with a unilateral perfusion defect confined to the ventral occipitotemporal region.

The superior quadrantanopia present in our two cases is common in patients with hemiachromatopsia or complete achromatopsia. For example, in Meadows's review³ of the literature on central achromatopsia, all 14 patients had a field defect in at least one superior quadrant, and frequently in both upper quadrants. This field defect associated with achromatopsia presumably occurs both because of the close anatomic relationship of the fusiform and lingual gyri to the inferior bank of striate cortex, and because of their shared vascular supply (an inferior branch of the posterior cerebral artery). An occipitotemporal lesion may cause a superior quadrantanopia by directly damaging the inferior striate cortex (V1), as likely occurred in our second patient. Alternatively, lesions that spare V1 may still result in a superior field defect by damaging the adjacent prestriate cortex, V2/V3, which contains a retinotopic map that directly and reciprocally connects to V1.¹² Finally, a superior quadrantanopia may occur from an isolated white matter lesion interrupting the optic radiations to V1.¹³ In most cases, an infarct producing both a superior quadrant field defect and inferior quadrantic achromatopsia will not be limited to cortex or white matter alone. This point is illustrated by our patients. In both, the lesions likely involved inferior optic radiations as well as prestriate visual association cortex, including V4. The involvement of both optic radiations and prestriate cortex may be clinically important, since it has been proposed that involvement of the adjacent white matter could contribute to the severe color processing deficits seen in some patients.¹⁰

It is important to emphasize that neither of our patients was aware of their dense quadrantic achromatopsia. They reliably experienced colors in the world around them because color vision was intact in their remaining hemifield. Yet even when told of their quadrantic defect, neither was able retrospectively to identify a change in color vision. It was only when attention was drawn to their color deficit that either patient "saw" colors as gray or dulled in the affected quadrant. Possibly this represents a form of anosognosia, or lack of awareness of the deficit. Although both patients' infarcts were in the nondominant right hemisphere, there were no other signs of neglect in either patient. A review of the literature suggests this lack of

awareness of a quadrantic achromatopsia might not be rare. For example, although all five of Green and Lessell's⁴ patients with achromatopsia had bilateral lesions, two of the patients initially had a superior quadrantanopia without obvious defects in color vision. In both cases a second contralateral lesion led to achromatopsia involving not one but both hemifields, which suggests that a unilateral defect in color processing went undetected after the first lesion. The same may apply to Pearlman, Birch, and Meadows's¹⁴ patient, who also initially had a superior quadrantanopia from a unilateral infarct, but developed achromatopsia in both hemifields only after a second contralateral lesion. Whether a lack of awareness of quadrantic achromatopsia occurs preferentially with right hemispheric lesions is unknown, and will require directly comparing the clinical findings in patients with left vs right hemispheric lesions.

There are many elaborate tests that may be used to define a deficit in color processing, including color perimetry and the Farnsworth-Munsell 100-Hue Test. However, the simple bedside tests we have used serve well to identify achromatopsia. An initial quick screen with colored objects will uncover a defect in color vision. A second test involving the sorting of isoluminescent colored threads or disks can identify a true achromatopsia. As long as the examiner documents that achromatic vision is well preserved in the affected quadrant and carefully monitors visual fixation when colors are presented to this quadrant, one can rapidly and easily detect a quadrantic achromatopsia.

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