Cerebral Polyopia With Extrastriate Quadrantanopia: Report of a Case With Magnetic Resonance Documentation of V2/V3 Cortical Infarction

Matthew R. Jones, M.D., Rebecca Waggoner, and William F. Hoyt, M.D.

This is a case report of the occurrence of cerebral diplopia with right-side superior homonymous quadrantanopia in a young woman after chiropractic neck manipulation. Magnetic resonance imaging confirmed an infarct in the left inferior V2/V3 (extrastriate) cortex. The characteristics of the diplopia are illustrated with the patient's drawings, and persisting abnormalities in perception are described in the area of the initial field defect after static (computed) visual field testing yielded normal results.

Key Words: Cerebral diplopia—Chiropractic—Extrastriate—Polyopia—Quadrantanopia.

Diplopia of cerebral origin, or polyopia, is a rare manifestation of occipital lobe lesions. For almost a century, neurologists have speculated about the level of processing of visual information at which a disorder could cause perception of multiple images (1-10). We record and discuss our findings in a case of cerebral diplopia with extrastriate quadrantanopia occurring in a woman with an infarct in the inferior bank of the V2/V3 cortex in her dominant left occipital lobe.

CASE REPORT

A healthy 32-year-old commercial artist with chronic lower back pain had been treated by a chiropractor. During one of her visits, rapid rotational neck manipulation produced severe head and neck pain followed by bilateral blindness. The blindness began to clear in 1 to 2 hours. She was admitted to our hospital where she reported double vision and difficulty seeing in the right upper visual field. Visual acuity was 20/20 in each eye. There was no afferent pupillary defect. Both eyes were straight and moved conjugately without evidence of esophoria, exophoria, or nystagmus. Slit lamp and ophthalmoscopic examination revealed no abnormalities. Confrontation visual field testing showed right upper homonymous quadrantanopia to hand motion.

The patient reported diplopia that was present during occlusion of either eye and during binocular viewing. Looking through a pinhole aperture did not eliminate the diplopia. When a pencil was held upright, she reported a second vertically oriented image displaced slightly to the right. When the pencil was held horizontally, she reported seeing a second horizontal image slightly above. The images were separated by an estimated 1° when measured at 1, 3, and 6 feet from her face.

Magnetic resonance imaging showed areas of increased signal on T2 sequencing, consistent with embolic cortical infarction in the left occipital lobe, adjacent and inferior to the primary visual cortex (V1) (Fig. 1). A small area of increased signal was also present in the right cerebellum. The lesion extended along the gyrus anteriorly into the parietal lobe and posteriorly just short of the tip of the occipital lobe. White matter tracts appeared to be spared.

Vertebral angiogram showed dissection of the right vertebral artery with formation of an intraluminal thrombus. She received heparin and warfarin. Two days later, Humphrey 30-2 visual field testing showed dense right upper homonymous quadrantanopia that precisely respected the horizontal meridian (Fig. 2).

Ten days later, hand movements could be detected in the quadrantopic field, but the patient was unable to count fingers in the affected quadrant. There was 3° of macular sparing, and diplopia remained. Color vision was normal when tested with the Hardy-Rand-Rittler plates, yet she was totally color-blind within the borders of the field defect.

When given a series of figures, she was instructed to draw what she saw, while identifying the figure and the secondary image (Fig. 3). She did not instantly see the second image; it appeared after a latency of 2 to 12 seconds. The images were consistently 1° apart, whether tested at 1, 3, or 6 feet. The images were of similar quality and size, and there was no perseveration of the image; it disappeared as soon as the figure was removed. She could usually distinguish the image she was fixating
on as the "true" image. However, at times she had difficulty distinguishing the true image from the "false" one, given their similar qualities. Motion of the fixation figure across the visual field horizontally or vertically did not elicit the diplopia, nor did smooth-pursuit tracking of a pen tip that was brought across the figure horizontally or vertically. Responses were similar in monocular, binocular, and pinhole viewing.

Two months later, the patient's diplopia remained. She reported exacerbation of the diplopia with attempted reading or writing, when she would see doubling of the written letters and of her hand holding the pen. Familiar surroundings were less likely to elicit the diplopia than were novel or "complex" surroundings. For example, when she looked at items in the produce department of a supermarket, the items would become jumbled "like a jar full of jelly beans" (Fig. 4). It had become less difficult for her to distinguish the true from the false image: Attempted fixation on the false image to the right would result in fixation on an unintended target (to the right of the intended target) with doubling of the unintended target, whereas fixation on the true image would result in fixation and doubling of the intended target. Humphrey 30-2 visual field testing showed asymmetric resolution of the quadrantanopia (Fig. 5). Testing at the tangent...
CEREBRAL POLYOPA

shown

seen

primary image

secondary image

primary image

secondary image

primary image

secondary image

primary image

FIG. 3. Patient's drawing of her image perceptions. The figures on the left represent the visual stimulus shown to the patient, and the figures on the right represent what she saw. It should be noted that both images were of similar size and quality. Note the consistent displacement into the quadrantanopic field.

primary image

secondary image

primary image

FIG. 3. Patient's drawing of her image perceptions. The figures on the left represent the visual stimulus shown to the patient, and the figures on the right represent what she saw. It should be noted that both images were of similar size and quality. Note the consistent displacement into the quadrantanopic field.

screen, however, showed relative homonymous defects, especially for form recognition. For example, the examiner's hand appeared indistinct when presented in the affected quadrant, as if it were a "fuzzy mitt."

One month later, the patient reported that the diplopia was gradually becoming less noticeable, especially in familiar surroundings such as her home. Humphrey 30-2 computed visual field and Goldmann field testing showed no defects (Fig. 6).

To evaluate color and form recognition, we constructed 3-cm testing figures of various shapes and colors and used these to perform kinetic perimetry on a black tangent screen (Fig. 7). The patient had relative quadrantanopia with macular sparing of varying degrees, depending on the specific color or shape of the test object (Fig. 8).

DISCUSSION

Polyopia of cerebral origin has been associated with lesions of the occipital cortex or central visual pathways. Infarction, tumors, multiple sclerosis, trauma, encephalitis, seizures, and migraine have been reported as causative factors (1-7,11-17). The characteristics of polyopia vary in different reports. The number of images seen ranges from one and one half to hundreds (1,8). Typically, patients report similarity between the true and false images and may even have difficulty distinguishing between them (1,5,8,11,12,15). Some patients, however, report that the false images differ in intensity, color, or size (1-4,11,12). Ocular fixation is often the precipitating activity (2,3,5,8,11-13), although head movement or movement of the target stimulus has also been described (1,4). Polyopia typically manifests several seconds after fixation; however, this latent period may be less than 1 second (1,2,11). The false images may be fleeting (2,4), lasting only as long as the stimulus is fixated on (1,15) or may persist after the stimulus has been removed (pali-nopia) (3,11,12). When more than two images are present, they are typically parallel and evenly spaced (1,5).

Perhaps the most interesting association is that of polyopia with homonymous visual field defects. In the first report of cerebral polyopia in 1908, Mingazzini (7) described homonymous hemianopia and polyopia resulting from an occipital lesion. In Bender's landmark work in 1945 (1), the first description of polyopia in the English language, he presented four cases, three with homonymous visual field defects. Subsequent reports have confirmed that homonymous defects, extending within several degrees of fixation are the rule (2-4,11-13). Rarely, a migrainous scotoma, transient or permanent, may combine with polyopia (6,14,15). When the location of the polyopic images is mentioned, it is usually on the side of the homonymous field defect within the area of macular sparing (2-4,11,12,15).

Our patient had a homonymous visual field defect, exhibited a false image in the quadrantanopic field, and had many other features consistent with previous reports of cerebral polyopia (fixation as a precipitating activity, latency of image appearance, similarity in quality of false and true image). Her drawings were remarkably similar to those of one of Bender's patients (1). However, our case was unique because we could, for the first time, localize the occipital lesion perimetrically (18) and by magnetic resonance imaging to the extrastriate (V2/V3) cortex.

Magnetic resonance imaging in our patient revealed a cortical lesion in gyri adjacent and inferior to the primary visual (striate) cortex, with sparing of the primary visual cortex and underlying white matter, effectively placing the lesion in the extrastriate cortex. Horton and Hoyt (18) showed that lesions bordering striate cortex (V1), involving extrastriate cortex (V2/V3) produce quadrantic defects that sharply respect the horizontal meridian, and proposed that such quadrantanopia is the hallmark of extrastriate lesions. In our patient, the visual field precisely respected the horizontal meridian, providing additional evidence for an extrastriate injury.

Extrastriate areas (V2, V3, V4, V5) have been shown to contain cells especially sensitive to motion, orientation, direction, and color, reflecting the importance of the extrastriate cortex in shape, pattern, and color recognition (19). This may provide some insight into the dis-
crepancy between our patient's Humphrey and Goldmann fields and her form-color fields. Although our color targets were not contrast matched, our patient's normal quadrants served as control quadrants with which to compare the pathologic quadrant. We postulate that recognition of shapes and colors versus discrete, nondenot points of light requires, to some degree, functioning extrastriate cortex. As such, our patient's relative defect with such testing figures was revealed in the face of normal Humphrey and Goldmann visual fields.

The cause of polyopia remains elusive. Bender postulated that lesions of the occipital lobe and homonymous visual field defects cause faulty fixation (1). "New maculas" result, and the stimulation of the true and false maculas causes polyopia. Kinsbourne and Warrington (5) proposed that posterior cerebral lesions disrupt normal inhibition of some components of sensory input, resulting in prolongation of after-sensations and producing visual perseveration. Jacobs (3) presented an interesting case of unilateral seizure leading to polyopia and palinopia. Noting that visual perseveration often occurs in association with incomplete homonymous hemianopia, and using evidence that partial deafferented visual cortex tends to retain visual images (3,20,21), he subsequently argued that palinopia is responsible for many instances of polyopia, with the extra images abnormally preserved in a defective but not blind visual field (16).

Gottlieb (4) thought that incongruity in the processing of visuospatial information between afferent (through retinal projections) and efferent (through collaterals of efferent impulses to the oculomotor system) sources results in poor visuospatial localization in the hemianopic field,
CEREBRAL POLYPIA

monkey visual cortex that responded maximally to stimuli of noncorresponding retinal points. These latter two reports provide evidence that discrete areas in the visual cortex subserve multiple areas of the visual field, and lesions to these areas may result in faulty visuospatial processing and polyopia.

Our patient's extrastriate injury emphasizes the importance of V2 and V3 in visuospatial processing. Her difficulty in detecting color and form in the affected quadrant highlights the role of extrastriate cortex in these processes. As the representation of the visual field in V1 is duplicated in V2 and V3, there are connections between primary visual cortex and extrastriate areas suberving analogous points in the visual field. In addition, within the extrastriate cortices, certain areas are responsible for integrating information from adjacent areas within the visual field, giving rise to end-stopped, center-surround, or orientation-selective cell groups. Disruption of these connections between V1 and V2/V3, or of the integration of topographic information within the extrastriate cortex itself may result in incomplete, delayed, or otherwise faulty transmission and processing of visuospatial information and thus the perception of multiple images. Although the mechanism of polyopia remains a mystery, the location of our patient's lesion focuses attention on the pathophysiologic processes within the extrastriate cortex.

REFERENCES