Clinical Reasoning: An 80-Year-Old Woman With a Homonymous Hemianopsia: Clinical Reasoning

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Abstract
An 80-year-old woman who was experiencing visual symptoms for two years was found to have a left homonymous hemianopsia. On further evaluation the following month, she was noted to have simultanagnosia and alexia. Magnetic resonance imaging (MRI) of the brain did not reveal a structural etiology for the symptoms. [18 30 F]-fluorodeoxyglucose positron emission tomography-computed tomography (PET-CT) was performed to aid in the diagnostic investigation. This case highlights the differential diagnosis of a homonymous hemianopsia in the absence of a structural lesion on MRI and the role of PET-CT imaging in such patients.
An 80-year-old woman with hypertension, hyperlipidemia, and type 2 diabetes mellitus reported two years of progressive difficulty reading and loss of depth perception. More recently, she stopped driving and was running into objects while walking. Initial evaluation by a local ophthalmologist identified bilateral cataracts resulting in surgical extraction, which did not improve the visual symptoms. Automated visual field (AVF) testing demonstrated a left homonymous hemianopsia (HH). Routine laboratory evaluation showed normal blood counts and chemistries. Patient was referred to a neurologist and a normal neurological examination was noted. A brain magnetic resonance imaging (MRI) with contrast showed normal age-related changes but specifically there was no evidence of a prior stroke, restricted diffusion on the diffusion weighted images, tumor, abnormal posterior cortical asymmetry, or enhancement. Patient was referred to neuro-ophthalmology for further evaluation.

On our evaluation, visual acuity was 20/30 in each eye. She identified only 1 of 13 plates in each eye from the Ishihara pseudochromatic book. Confrontation testing was inconsistent to finger counting but there was a nasal defect in the right eye and a temporal defect in the left eye. Pupils were equal with normal light reflexes and no relative afferent pupillary defect. Slit-lamp examination showed centered posterior chamber intraocular lenses with normal intraocular pressure. The optic disc, macula and retina appeared normal in each eye. Repeat AVF testing confirmed the left HH (Figure 1A).
Questions for consideration

Where is the lesion responsible for the visual field defect?

What is the cause of the decreased color vision?

What is the differential diagnosis for a homonymous hemianopsia with a negative MRI?

SECTION 2

The pattern of a visual field defect can often determine the anatomical location of the lesion (Figure 2). In general, the following characteristics of a visual field defect should be noted: monocular or binocular, homonymous or heteronymous, complete or incomplete, incongruous or congruous. Historically a congruent HH was considered to originate from a lesion involving the occipital lobe, but this paradigm has been shown not to be a reliable localizing sign. Therefore, in our patient the presence of a left HH localizes the site of the lesion to anywhere along the right retrochiasmal visual pathway (i.e. right optic tract, lateral geniculate nucleus, optic radiations, or occipital lobe).

AVF is designed to detect visual field defects by projecting lights of different brightness, size, and location while the patient fixates at the center of a hemispherical bowl. Typically, the central 24-30° of vision is tested with a target size of III (4 mm²) stimulus. These stimuli are algorithmically planned to decrease patient anticipation and increase reliability. For the test to be valid, the patient must be able to reliably participate and complete the test. Patient factors such as familiarity with the test (i.e. learning curve), age, cognition, and physical abilities affect
reliability. Fixation losses, false positive responses (a positive response but there is no target displayed) and false negative responses (failure to signal at a target that was previously seen in the same location) are metrics measured and displayed on the printout that determine the reliability of the test. Fixation losses should be less than 20%, and false negative errors and false positive errors less than 20%-30% for the test to be reliable (Figure 1B).

Any visual field defect should be interpreted within the context of other clinical signs and symptoms. The Ishihara pseudochromatic color plates are often used clinically to screen for dyschromatopsia by displaying a symbol (number or geometric figure) composed of colored dots against a background of differently colored dots; however, they can also be utilized as a screening tool for simultanagnosia. Patients with simultanagnosia are unable to perceive more than a single image at a time, and often have trouble depicting the numbers from the background without actual impairment in color vision due to the inherent nature of the test. Further testing was notable for alexia and inability to name single objects in overlapping figures. Our patient’s difficulty with the Ishihara color plates was likely due to simultanagnosia as she did not have a history of congenital colorblindness and there was no evidence of optic neuropathy or retinopathy on examination. She scored 27 out of 38 on the Kokmen short test of mental status, by obtaining 8 of 8 points in Orientation, 6 of 7 in Attention, 4 of 4 in Registration, 0 of 4 in Calculation, 2 of 3 in Similarities, 1 of 4 in Construction, 4 of 4 in Knowledge, and 2 of 4 in Recall.
A congruent HH without a structural lesion on brain MRI often localizes to the occipital cortex and has a short but important differential diagnosis that includes the Heidenhain variant of Creutzfeldt-Jacob disease (CJD), posterior cortical atrophy (PCA) syndrome, non-ketotic hyperglycemia, seizures, subtle occipital ischemia or a functional visual disorder. In our patient, the duration of symptoms and lack of diffusion restriction on the brain MRI made CJD unlikely. Normal laboratory evaluation and the progressive nature of the symptoms made hyperglycemia or a vascular etiology unlikely. The history and examination findings were not consistent with seizures or a functional visual disorder.

**Questions for consideration**

*What further testing would you perform?*

**SECTION 3**

\[^{18}F\]-fluorodeoxyglucose positron emission tomography-computed tomography (FDG PET-CT) showed asymmetric moderate hypometabolism involving the right posterior cerebral hemisphere in a pattern consistent with PCA (Figure e-1). Studies have shown FDG PET-CT, which measures functionality, is more sensitive and specific when evaluating patients with mild cognitive impairment compared to MRI, which is a structural measurement. She was referred to neurology for management but declined further neuropsychiatric testing or initiation of treatment.

**DISCUSSION**
PCA is a rare, progressive, neurodegenerative syndrome that affects the posterior cortices resulting in visual dysfunction early in the disease\textsuperscript{9}. The disease process may initially affect parietal, occipital or occipita-temporal regions therefore patients can present with a wide range of visual symptoms affecting higher-order visual function, while sparing anterograde memory, language, executive functions and personality\textsuperscript{10,11}. PCA typically presents in the fifth and sixth decade of life, and as the disease progresses patients develop dysfunction of other cognitive domains. The most common etiology of PCA syndrome is Alzheimer disease pathology, however, it can also be seen with other underlying pathologies such as dementia with Lewy body, corticobasal degeneration, prion disease and subcortical gliosis\textsuperscript{9,12-14}.

The initial symptoms of PCA can be visuospatial-visuoperceptual impairments such as, alexia, homonymous visual field deficits, features of Balint’s syndrome (simultanagnosia, oculomotor apraxia, optic ataxia, environmental agnosia) or Gerstmann syndrome (acalculia, agraphia, finger agnosia, left/right disorientation)\textsuperscript{15}. Patients with PCA syndrome commonly present with reading difficulties, while simultanagnosia and homonymous visual field defects are also common findings in the initial evaluation. Homonymous visual field defects have been reported to be present in up to 60\% of patients, highlighting the importance of AVF testing and cognitive assessment in suspected cases\textsuperscript{13}.

Our patient with HH, alexia and simultanagnosia met diagnostic criteria for PCA syndrome\textsuperscript{12}. This case highlights the importance of considering neurodegenerative disease when a patient over 45 years of age presents with a reproducible HH and negative findings on MRI.
REFERENCES


FIGURE LEGENDS

Figure 1: Interpretation of Humphrey visual field test printout

A. 24-2 Humphrey analyzer (Carl Zeiss Meditec AG, Dublin, CA). There is a left homonymous hemianopsia. Annotations highlight the various metrics needed to interpret the results.

B. Approach to automated perimetry interpretation.
Identify patient:
- Patient name
- Birthdate (incorrect entry results in comparison with the wrong age group)
- Visual acuity
- Pupil size (pupil smaller than 2 mm or larger than 6 mm may affect outcomes)

Identify testing parameters:
- Stimulus size (size III stimulus [4 mm²] used in patients with visual acuity of 20/200 or better)
- Testing algorithm (10-2, 24-2, etc.) (i.e., central 24-2 indicates the central 24 degrees of visual field were tested and "2" indicates that the recorded values straddle the vertical and horizontal meridians)

Acceptability reliability indices:
- Fixation losses: Patient reports seeing a stimulus in the expected physiologic blind spot (>20% could result in unreliable test)
- False positive errors: Patient presses button when no stimulus was present “trigger happy” (>20–30% could result in unreliable test)
- False negative errors: Patient fails to see a brighter stimulus at a previously seen location (>20–30% could result in unreliable test)

Interpret results:
- Numerical total deviation map compares patient’s visual sensitivity to age-matched controls (positive values represent increased sensitivity than normal, and negative values represent decreased sensitivity than normal)
- Numerical pattern deviation map extrapolates localized areas of sensitivity loss from a field that is diffusely depressed (i.e., depicts focal field depression in a patient with decreased threshold across the entire field due to factors such as dense cataracts)
- Grayscale probability plots of total deviation and pattern deviation maps are useful visual representation of the statistical significance of field defects (should be interpreted in conjunction with the numerical maps)
- Mean deviation (MD) represents the average difference from the expected age-matched normative database
- Pattern standard deviation (PSD) provides information about focal defects (higher values represent more focal losses, and lower values represent either no loss or diffuse loss)
- Gaze tracker monitors patient’s eye movements during testing: upward bar indicates fixation disparity, and downward bar represents tracking failure. The length of the upward bar indicates the magnitude of the disparity, while a long downward bar represent eyelid closure.
Figure 2: Visual pathway anatomy with localization of visual field defects

Illustration of the anterior and posterior visual pathway with various locations of a lesion resulting in their corresponding visual field defect. With permission from the Mayo Foundation for Medical Education and Research.
Figure e-1: Visual pathway anatomy with localization of visual field defects

[18F]-fluorodeoxyglucose positron emission tomography-computed tomography showing asymmetric moderate hypometabolism of the right posterior cerebral hemisphere.
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