Clinical and imaging clues to posterior cortical atrophy

Victoria S. Pelak, MD

Christopher M. Filley, MD

A 54-year-old man complained of progressive vision loss over 3 years. Several eye specialists were consulted and no ocular cause for his visual complaints was identified, but Ishihara color testing was noted to be abnormal. After an unsuccessful trial of prism lenses for depth perception complaints, an MRI of the brain was obtained and interpreted as normal. He was referred for neuro-ophthalmic consultation after he developed difficulty driving and impaired work performance as an electrician. Neurologic examination revealed a best-corrected visual acuity of 20/25 in both eyes, Ishihara color testing deficits, impaired clock drawing (figure 1), ideomotor apraxia, ocular motor apraxia, and optic ataxia, without other abnormalities. Neuropsychological testing disclosed deficits in visuospatial function, processing speed, and executive function, with preserved memory and language. Hypometabolism in occipitoparietal and occipitotemporal regions was observed on [18 F] fluorodeoxyglucose PET-CT and found to correspond with focal atrophy on the original

Figure 1 Performance on clock drawing after instruction to insert the hands of the clock to indicate 10 after 11

Departments of Neurology (VSP, CMF), Ophthalmology (VSP), and Psychiatry (CMF), University of Colorado School of Medicine, Aurora; and Denver Veterans Affairs Medical Center (CMF), CO.

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Correspondence to: victoria.pelak@ucdenver.edu

Figure 2 T1 axial MRI (A–D) and [^{18}F] fluorodeoxyglucose PET-CT images (E–H)

Focal cortical atrophy on MRI corresponds with regions of hypometabolism on PET-CT in occipitotemporal regions (B and F) and at the occipitoparietal junction (C and G), noted by arrows. At the level of the cerebellum (A and E) and rostral frontoparietal regions (D and H), there is no evidence of selective atrophy with hypometabolism.

MRI (figure 2). The patient was diagnosed with posterior cortical atrophy (PCA),^{1,2} a neurodegenerative disorder most commonly associated with Alzheimer disease pathology. A delay in diagnosis of PCA is common owing to a prolonged search for ocular causes, and cortical disease may be neglected as a possible etiology when a brain MRI is interpreted as normal. If the nature of visual complaints remains elusive, clinically useful signs for the consideration of PCA include unexplained Ishihara color defects and impaired clock drawing, which should prompt special attention to the posterior cortical regions on MRI and consideration of PET-CT imaging to support the diagnosis.²

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