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Posterior Cortical Atrophy – Case Report and Retrospective Review

Jennifer J. Lowe1, Julie Falardeau2, Robert A. Egan3, William L. Hills2

1Oregon Health & Science University, School of Medicine, Portland, OR, USA, 2Oregon Health & Science University, Casey Eye Institute, Portland, OR, USA, 3St. Helena Hospital, Neuro-ophthalmology, St. Helens, CA, USA

Introduction:
Posterior cortical atrophy (PCA) is a rare progressive neurodegenerative disease with prominent cortical visual dysfunction, first described by Benson in 1988. Studies have shown visual field defects on formal testing, increased phosphorylated-tau protein in the CSF, and hypometabolism in the parieto-occipital regions on PET imaging. We describe one patient with PCA whose symptoms are classic but whose progression has been atypically slow. To further investigate this disease entity, we retrospectively reviewed 8 additional charts and report the commonalities in all 9 cases.

Methods:
We reviewed the index subject’s symptoms and findings on presentation and follow up. We reviewed 8 additional cases of PCA evaluated by Neuro-Ophthalmology. We analyzed the signs and symptoms in, as well as the demographics of, these cases to look for common themes at presentation.

Results:
A 58 year old woman presents with a progressive 10 year history of difficulty processing spatial information, reading handwriting, and recognizing faces. She described visual hallucinations, photophobia, poor depth perception, and visual recall. Visual field testing found an incomplete hemianopsia. CSF analysis detected reduced $\beta_{(1-42)}$ Tau Index and elevated phosphorylated-tau. Poor visuospatial skills were found on neuropsychological testing and occipito-parietal hypometabolism on PET imaging. Furthermore, 7/9 patients reported trouble reading and 4/8 depth perception difficulty, which were the most recurrent complaints. Recurrent signs included abnormal color plates (9/9), simultanagnosia (6/8), hemianopsia (3/7), decreased stereopsis (4/4), and abnormal Amsler’s grid (4/5).

Conclusions:
Posterior cortical atrophy is an uncommon atypical variant of Alzheimer’s disease, sometimes termed “Benson’s syndrome”. Patients commonly report difficulty reading despite normal visual acuities. Our observations and previous studies describe visual signs and symptoms attributable to the parieto-occipital cortex. More recently, CSF phosphorylated-tau has been utilized as a bio-marker. A systematic approach using clinical, laboratory, and radiographic data can aid in diagnosis of PCA. This may help clinicians with similar diagnostic dilemmas.

References:


Keywords: Posterior cortical atrophy, Neurodegenerative, Cortical visual dysfunction, Diagnosis, Bio-marker

Financial Disclosures: The authors had no disclosures.