Forgetting How to See: A Variant of Alzheimer’s Disease Affecting Vision

Abstract

A middle aged man shows progressively worsening vision, including reduced acuity, tracking, and visual spatial abilities. Posterior cortical atrophy, a variant of Alzheimer’s, causes these decreases in visual function while preserving cognitive abilities, as reported.

Case History

A 64 year old white male was first seen at the Michigan College of Optometry in 2013. He presented with reading and tracking issues, as well as visual perceptual deficits. Significant photophobia was also noted. At this point, he had been diagnosed with neurological impairments secondary to repeat traumatic brain injuries (TBIs) earlier in life, in addition to anxiety. This patient had a medical history positive for prostate cancer which had been treated by prostectomy 10 years prior. Presenting acuities were 20/40 OD, 20/30 OS. Phoric posture was noted as 2 exophoria at distance, and 3-6 exophoria at near. Near vergence recovery values were reduced, at X/15/4 BI and X/10/6 BO. DEM was reduced for vertical time, horizontal time, and ratio. Refraction and health findings were otherwise insignificant. Vision therapy was suggested for the saccadic dysfunction, but not pursued at this time.

This patient returned to MCO in early 2014. He was again complaining of issues with reading, as well as being more forgetful than in the past. No changes to systemic health had been noted since his last visit. Presenting acuities were 20/25 OD and OS at this visit, though he noted that the letters got “mixed up” on the chart as he read them. Distance cover test revealed a 1.5 prism diopter right hypertropia. Near cover test showed a 10 prism diopter alternating exotropia with 2-4 prism diopter right hypertropia. DEM again showed marked reduction of saccadic function, and 20 errors were made during the test. Vision therapy was prescribed, and the patient elected to begin therapy. He attended 5 sessions of vision therapy, during which his saccadic function showed improvements, though was still significantly reduced. Beyond session five, he was lost to follow up for two years.

In August 2016, now 67 years old, the patient presented to MCO with his caregiver, looking to improve his visual function. He had been experiencing a decrease in vision over the last several years, which he described as “slowly getting worse”. Since he was last in 2 years ago, this patient had been diagnosed with posterior cortical atrophy, which is also known as a visual variant of Alzheimer’s disease. This condition affects visual function, while leaving cognitive function relatively untouched. His medication list contained risperidone, lisinopril, trazodone, lansoprazole, venlafaxine, lorazepam, and docusate sodium. The patient was using a white cane to aid in mobility, though neither he nor his caregiver were
sure how long he had been using it for, or whether he had been formally trained on its use. His goals were to be able to use his phone, TV remote, and coffee maker, all of which he struggled to use, but were part of his daily routine.

Pertinent Findings

When walking from the waiting room to the exam room, it was observed that this patient had unsteady gait, and would drift to the left. Visual acuity was unable to be obtained monocularly, but binocular distance acuity was measured to be 20/400 with single letters. Binocular near acuity was measured at 20/500. Testing showed no defects in pupil function, and no relative afferent pupillary defect. EOM testing revealed no gross restrictions, though pursuits were impaired. Cover test was unable to be performed, as the patient was unable to fixate an object. Ocular health examination revealed a healthy anterior segment, with no corneal abnormalities that would account for decreased acuities. The right eye was pseudophakic, and the left eye showed only mild nuclear sclerosis. Posterior segment views were fleeting, but no retinal or optic nerve pathology was discovered. Both of the optic nerve heads had normal tissue distribution, and no pallor. When asked to point out a space halfway between the examiner’s hands, the patient consistently indicated this point to be to the left of the actual midline.

Differential Diagnosis

Previous medical records were obtained for this patient, revealing that posterior cortical atrophy had been diagnosed via PET/CT scan in early 2015. While a history of repeated TBI was present, and could explain this patient’s earlier complaints, it was unlikely that prior TBIs would cause a decrease in visual function of this magnitude. While the diagnosis in this case was straightforward due to the patient’s previous records, other differential diagnoses for reduced vision of this severity should always be considered. Bilateral retrobulbar optic neuritis, previous retinal artery occlusions, or ischemic or hemorrhagic cortical infarcts are other diagnoses that would not necessarily be visible on ophthalmic examination, but could have caused the decrease in vision over the past two years. Because posterior cortical atrophy was already diagnosed radiologically, it was not necessary to order further testing in order to rule these out.

Diagnosis and Discussion

Posterior cortical atrophy is recognized as a variant of Alzheimer’s disease that disproportionately affects visual function over cognitive function. Neuroimaging shows that the occipital lobe, and to a lesser extent the parietal lobe, of affected individuals experience a reduction in grey matter when compared to control patients¹. This shows stark contrast to typical Alzheimer’s grey matter loss, which
occurs primarily in the medial temporal lobes. Our patient’s lesions on neuro imaging were congruous with posterior cortical atrophy findings.

Epidemiologically, the incidence of this condition is unknown, as it is fairly uncommon, and has not been studied nearly as intently as typical Alzheimer’s disease. Globally, the prevalence of Alzheimer’s disease is approximately 24 million. Analysis of Alzheimer’s treatment centers suggests that about 5% of patients being treated for Alzheimer’s are afflicted by this visual variant. This condition is distinct from typical Alzheimer’s disease primarily in that visual function is severely impacted, while cognitive function is left relatively intact. Patients with posterior cortical atrophy are also afflicted at a younger age than typical Alzheimer’s patients, with an age of onset in the range of 50 to 65 years. These characteristics are consistent with our patient. He began experiencing related symptoms in his mid-60s, and was very cognitively aware, despite reduced visual function. His depression and anxiety were also likely due, at least in part, to his decline in visual abilities.

Posterior cortical atrophy causes gradual decrease in reading skills and visual spatial abilities, as well as causing visual agnosias. Based on this, our patient’s visual acuity may not have been as reduced as was measured. Rather, visual agnosia may have prevented the patient from recognizing targets accurately. Saccadic issues have also been seen in these patients. Patients with posterior cortical atrophy are typically very aware of their symptoms, and often experience depression as a consequence. These patients often show better memory function than typical Alzheimer’s patients.

Patients with posterior cortical atrophy typically first present with vague visual complaints, such as difficulties reading, driving, or using stairs. Light sensitivity and difficulties recognizing objects are also common complaints. Ophthalmological examination will often show no ocular abnormalities in these cases, which can prompt a neurology referral. Initially, diagnosis can be difficult, and the patient may be misdiagnosed as simply having anxiety until symptoms worsen. Because our patient had a history of repeated TBIs, his visual symptoms may have overlapped, or were misattributed to those.

**Treatment and Management**

Treatments for posterior cortical atrophy have not been extensively researched. Acetylcholinesterase inhibitors are often used, as these are a commonly employed treatment for typical Alzheimer’s disease, and anecdotally show modest benefit in some patients. Low vision and psychological services are perhaps more appropriate for these patients, as they tend to have relatively preserved cognitive function. Yoked prisms were trialed in an attempt to improve this patient’s gait. With 8 prism diopter base right yoked prism, our patient demonstrated a steadier, straighter gait. 8 prism diopter base right Fresnel prisms were added to the patient’s habitual spectacles for full time use. A clothespin drop activity was also trialed in office, and our patient demonstrated the ability to accurately drop a large
clothespin into a large-mouthed coffee container. This activity was prescribed for daily home use, in hopes that this may slow further loss of visual motor skills.

Because this patient was having trouble using his telephone, coffee machine, and TV remote, modifications to these objects were discussed. For the telephone, it was suggested that a large sticker with tactile elements could be placed on the “5” button, which would allow our patient to dial the correct numbers based on using this sticker as a starting place and moving his finger from there. A similar suggestion for modifications was discussed for the TV remote, along with potentially covering unused buttons with black tape and using high contrast stickers for high-use buttons. For the coffee machine, high contrast stickers of varying colors were suggested to allow our patient to find the buttons that he needs to press to start brewing coffee.

**Conclusion**

While uncommon, posterior cortical atrophy is a clinical entity that optometrists should be cognizant of. One should make special note of middle aged patients reporting vague decreases in visual function, especially if these do not match up to any exam findings, or if these complaints are consistent over the course of several exams. While prompt diagnosis of this condition does not necessarily affect prognosis, it can allow for much better management and patient education. Optometrists should be capable of providing low vision services to patients with posterior cortical atrophy, including referrals to appropriate community resources. Providing such services is an invaluable part of optometry, and has the potential to positively impact patients going through stressful and debilitating life changes.

**Bibliography**


