Article ▸ Small Spot, Big Problem: Low Vision Management of Central Neurological Visual Field Defects

Noah Merhar, OD, VA Central Western Massachusetts Healthcare System, Leeds, Massachuesetts
Theresa Zerilli, OD, VA Connecticut Healthcare System, West Haven, Connecticut
Nancy Shenouda-Awad, OD, VA Connecticut Healthcare System, West Haven, Connecticut

ABSTRACT

Background: Neurological visual field defects can occur after cerebrovascular accidents. Depending on the size and location, visual field defects can impair a patient's mobility and their ability to perform activities of daily living independently, including reading. Patients with impaired activities of daily living can often benefit from low vision assessment and visual rehabilitation. This case report describes the low vision work-up and management of an uncommon presentation of neurological visual field loss.

Case Report: A 70-year-old male presented with reading complaints secondary to a small central neurological scotoma following a cerebrovascular accident. Standard perimetry with 24-2 and 10-2 Humphrey Visual Field testing did not reveal central defects that would correlate with the patient's symptoms. Amsler grid testing revealed homonymous central scotomas less than 1 degree in size. Low vision intervention included compensation strategies to reduce the patient's awareness of the scotomas and to support the patient's goal of fluent reading.

Discussion: In rare cases, standardized automated perimetry may miss very small but symptomatic central neurological visual field defects. In these cases, multiple types of visual field testing may need to be used in order to assess central visual field function and to determine rehabilitation options. Low vision intervention should still be considered in patients with small neurological visual field deficits despite good visual acuity and relatively full visual fields, especially if activities of daily living are affected.

Keywords: cerebrovascular accident, low vision rehabilitation, neurological scotoma, visual field deficits

Introduction

Cerebrovascular accident (CVA), or stroke affects almost 800,000 people in the United States every year. The majority of CVAs are caused by an embolus occluding blood flow to part of the brain. They can also be caused by a hemorrhage from a leaking aneurysm or trauma.[1] Strokes can have many possible sequelae, including cognitive changes, paresis, seizures, and visual deficits, many of which can be permanent.[2] Visual consequences of CVA can include visual field (VF) loss, diplopia, visual neglect, and higher-order visual processing difficulties.[3] Up to 60% of patients note some visual impairment following a stroke, with 20% having persistent visual impairment after 3 months.[4]

Visual field loss following CVA has been reported to occur in at least 8% of patients,[3,5] although the prevalence increases to almost 50% in patients with suspected visual impairment.[6,7] Depending on the location and severity of the infarction, the VF loss can vary. The classic neurological VF defect is a homonymous hemianopsia, in which one entire side of the VF is affected in both eyes.[3] If less tissue is involved, the defect may involve less than the entire half of the VF, while still respecting the vertical midline. Complete homonymous hemianopsia has been found to account for 29-54% of VF loss following CVA.[3,6] These defects can either involve central vision/fixation or spare it.[8] While it is possible to have more focal infarctions that cause very small areas of VF loss, unless the defects are in the patient's central vision, they will likely neither be noticeable to the patient nor be identified on standard VF testing. Central neurological VF defects in the absence of a larger peripheral VF defect are also possible, although they are not frequently reported.[8,9]

This paper describes a case study of a small but bothersome central neurological VF defect not detected on standard perimetry. It highlights the need to employ other types of VF testing to aid in diagnosis and emphasizes the low vision rehabilitation strategies utilized.

Case Report

History

TK, a 70-year-old white male, was referred by ophthalmology to the Low Vision Clinic for evaluation following a CVA 2 weeks prior. He complained of difficulty reading since his stroke. He described small spots in his vision that moved with fixation and "danced around the focal point." The patient reported that the spots appeared at around the time that he was diagnosed with his latest stroke. He felt that he was having difficulty reading because the spots blocked letters.

His medical history was remarkable for chronic lymphocytic leukemia, atrial fibrillation, hypertension, hyperlipidemia, cerebral infarction, and osteoarthritis. His medications

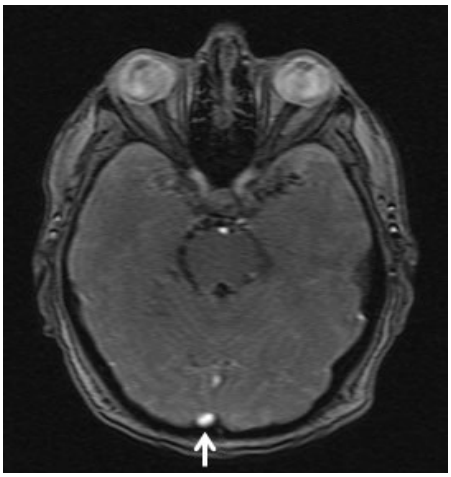

Figure 1. *Brain MRI from 7/1/16 showing a focal infarction to the tip of the right occipital lobe*

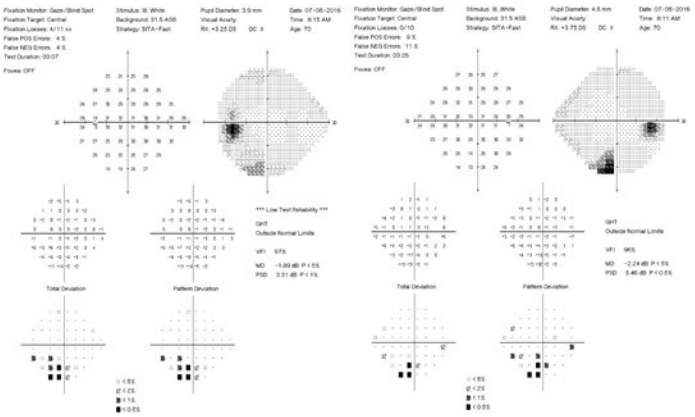

Figure 2. *24-2 visual field test from 7/5/16 showing a small inferior left homonymous defect*

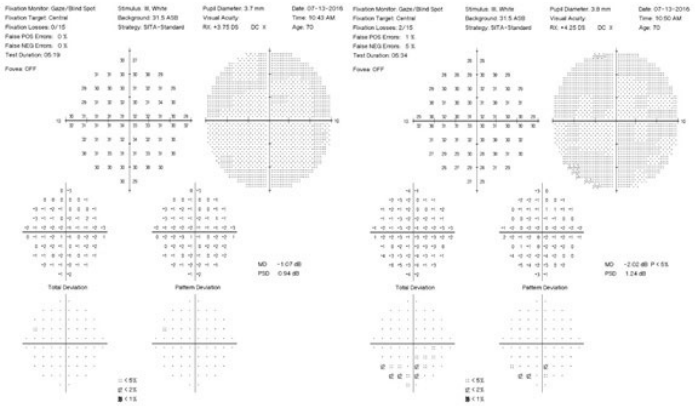

Figure 3. *10-2 visual field testing revealing no central defects in either eye*

included rosuvastatin, metoprolol, venetoclax, docusate/sennosides, dabigatran, allopurinol, acetaminophen prn, and tramadol prn. The patient had a history of multiple embolic CVAs and a subarachnoid hemorrhage 2 months prior; he was subsequently started on anticoagulation therapy. He was then hospitalized for a left leg hematoma that was believed to be secondary to the anticoagulation regimen. Consequently, his anticoagulants were discontinued. During his hospital stay, he reported a mild headache and central visual blur OU. An MRI was performed that revealed new punctate areas of restricted diffusion and mild enhancement in the right frontal and right occipital lobes (Figure 1), which was consistent with late acute vs. early subacute infarction.

Four days later, the patient was referred to the Ophthalmology Clinic for blurry vision OU. His visual acuity was found to be 20/20-1 OD and 20/25+1 OS. Pupil responses were normal, without an afferent pupillary defect. The 24-2 VF testing (Figure 2) revealed a small inferior left defect OU. His intraocular pressures were recorded as 20 mm Hg OD and 17 mm Hg OS. The anterior segment was unremarkable except for mild cataracts OU. The dilated fundus examination revealed moderate cupping with healthy rim tissue OU. He had no optic disc pallor or edema OU. The maculae were flat and clear OU. The retinal vasculature was normal, and the peripheral retina was flat and intact OU. He was diagnosed with cataracts OU and an inferior left homonymous quadrantanopic visual field defect secondary to the CVA. Despite the VF defects involving far less than a full quadrant, they were felt to be neurological in origin as they were congruous and respected the vertical midline in a patient with a recent occipital lobe infarction. The management plan involved monitoring his risk factors for additional CVAs and referring to the Low Vision Clinic to address the VF loss.

Initial Low Vision Evaluation

Two weeks following the CVA, the patient presented for his initial low vision exam. His entering distance acuities with his habitual prescription of OD +0.75-0.75x080 and OS +1.50-0.75x075 were recorded as 20/20-1 OD and OS.

As the patient was complaining of central defects not appreciated on his previous 24-2 VF testing, a 10-2 VF was also performed (Figure 3). It showed no defects in either eye. However, with Amsler grid testing, the patient reported a single scotoma of less than 1 degree located inferior left of but adjacent to fixation OU (Figure 4). The small defect on Amsler grid testing was thought to be secondary to his recent occipital lobe CVA as it was bilateral, homonymous, and respected the vertical midline. The patient reported that this central spot was what he noticed every time he tried to read. Due to the recent nature of his CVA and the possibility of improvement during the immediate recovery period, the patient was asked to return in 2 to 3 months to repeat visual field testing. At this follow up visit, his symptoms would be reassessed, and the need for assistive devices would be evaluated.

Low Vision Follow-up Visit #1

The patient returned 2 months later for a progress evaluation. He reported that the central spot in his vision was still present and had not improved since his last visit, although he was not aware of it all of the time, just during certain activities. He reported that reading was still difficult as he still missed small words frequently because of the scotoma.

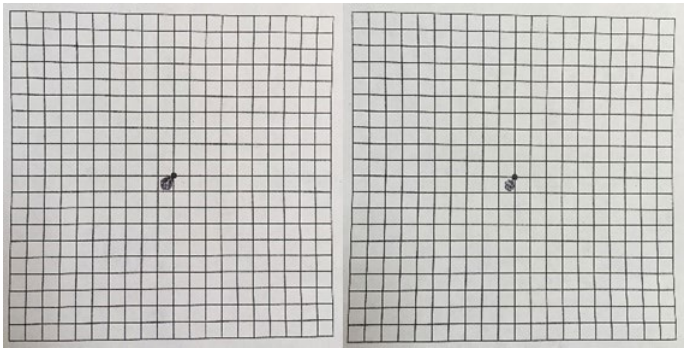

Figure 4. *Amsler grid of the right eye and left eye showing a small central scotoma in both eyes adjacent to fixation*

His visual acuities were stable at 20/20 OD and 20/20-OS. On standard Amsler grid testing with a black grid on a white background, he reported a stable central scotoma OU that was less than 1 degree in size and within 1 degree of fixation. He felt that the scotoma appeared a little less dense than it had previously, but it was still the same size. As the patient reported that he mostly noticed the scotoma when looking at a white surface such as a page of text, we decided to repeat the Amsler grid testing with reversed contrast. When the patient viewed the white grid on a black background, he did not notice the scotoma at all.

As the patient continued to report difficulty reading, it was determined that he would benefit from low vision evaluation in order to maximize his central visual function. Contrast sensitivity on the Pelli-Robson chart was near normal for his age in each eye. On tint evaluation, he preferred a 90% transmission plum tint, which objectively and subjectively improved his contrast sensitively slightly. He also appreciated a 70% transmission plum tint for reading. The patient preferred a +2.75 add over his habitual +2.50 add. He was able to read 20/20 at near. He had excellent fluency while reading out loud from the 1M (20/50) Sloan card; however, he subjectively felt that his reading was still slower and more laborious than it was prior to his stroke. It was hypothesized that increasing the magnification for reading might make the scotoma less prominent and increase his reading fluency. Low-powered hand-held magnifiers were demonstrated for spotting small print, but he did not feel that he had a need for them at that time. He was also shown a 2D Optivisor with a light. He liked the Optivisor, a head-mounted binocular magnifier, for assisting with household repairs.

At the conclusion of the exam, 3 pairs of glasses were ordered for the patient: a pair of single-vision distance glasses with a 90% transmission plum tint, a pair of reading glasses with a 70% transmission plum tint, and a pair of distance-only sunglasses with a 15% transmission grey tint. The 2D Optivisor with a light was also ordered for him. He was asked to return in 2 months for another re-evaluation to monitor his reading symptoms with the new prescriptions.

Low Vision Follow-up Visit #2

The patient returned to the Low Vision clinic 2 months later. He reported that his vision was much clearer with his new glasses, but there was no improvement in his awareness of the scotoma. He felt like reading was getting a little easier, although he still found it frustrating.

At this visit, the patient reported an episode of amaurosis fugax 1 month prior. After finishing some housework, he sat down to watch television and noticed a black cloud come across the right side of his vision. He reported that it lasted 15 to 30 minutes and then resolved completely. The episode was not associated with any other neurological symptoms. He had reported the episode to his neurologist as well as his hematologist/oncologist, who had increased his aspirin from 81mg every other day to 81mg per day. No other episodes of amaurosis fugax occurred after this initial episode.

His visual acuity was stable at 20/20 OD and 20/20-OS. The location of the scotoma was also stable on Amsler grid testing. Since the patient was still not happy with his reading ability, a video magnifier was trialed. He found that both reverse contrast and enhanced contrast were helpful in reducing his awareness of the scotoma. Additionally, he found that enlarging the print slightly made the scotoma less bothersome. A video magnifier was ordered, and the patient was told to return for training on the device.

Due to the amaurosis symptoms, the patient was dilated. Anterior segment was unremarkable except for mild cataracts OU. His intraocular pressures were recorded as 15 mm Hg OD and 17 mm Hg OS. Dilated fundus exam was stable except for a refractile plaque in a small vessel inferior temporal to the optic nerve head OS.

A bilateral carotid ultrasound was ordered to rule out carotid stenosis as the cause of the embolus. The patient's other providers were alerted to this finding as well. The patient was educated to return immediately if he noticed any other episodes of amaurosis fugax. A retinal consult for fluorescein angiography was not indicated as the artery appeared well perfused with no signs of retinal ischemia and no new defects on Amsler grid testing.

The carotid ultrasound showed no hemodynamically significant stenosis on either side. As the patient was on aspirin and had resolution of his symptoms, his other providers determined that monitoring without additional testing was appropriate. The patient was scheduled for VF testing to monitor the homonymous visual field defect in 1 month and a dilated fundus exam to monitor the refractile plaque in 4 months.

Discussion

The visual pathway through the brain is susceptible to damage at any point along its path, resulting in different patterns of vision loss. After the optic nerve exits the orbit, it

courses medially to the optic chiasm, where the nasal fibers cross to join the contralateral optic tract.[3] The optic tracts travel posteriorly to the lateral geniculate nucleus, and then the optic radiations course through the temporal and parietal lobes before reaching the visual cortex in the occipital lobe. A number of different arteries supply this pathway and the visual cortex.[3]

Ischemia to the optic radiations will cause a complete homonymous hemianopsia, while a lesion to part of the optic radiations will cause an incomplete hemianopsia or a quadrantanopsia. A lesion to the visual cortex will also cause a homonymous hemianopsia, often with macular sparing.[3] Macular sparing is thought to occur because the posterior and middle cerebral arteries provide dual blood supply to the posterior occipital lobe, which may protect it from ischemia.[3] Isa et al.[8] proposed that posterior cerebral artery occlusion likely damages the anterior calcarine fissure and spares the tip of the occipital lobe where the central visual field is represented.

While homonymous hemianopsia VF loss is often seen in patients who have suffered a CVA, this case demonstrates a rare presentation of central VF loss. Automated perimetry demonstrated a small, more peripheral homonymous defect, which was not bothersome to the patient due to its size and distance from fixation. His central neurological scotoma was not seen on automated perimetry, including a 10-2 visual field, but it was evident on Amsler grid testing. As the defect seen on Amsler grid testing was bilateral, homonymous, and respected the vertical meridian in a patient with a known occipital lobe lesion, additional work-up was not warranted. In cases where the central VF loss is asymmetric or unilateral, fluorescein angiography is indicated. Additionally, neuroimaging is warranted in any patient without known cerebral lesion who presents with VF loss that appears neurological in origin.

Small, central, neurological scotomas have not frequently been reported in the literature. While studies have shown the presence of homonymous macular defects[8,9] and central scotomas following CVA,[3] they are uncommon presentations of neurological VF loss. Isa et al.[8] looked at patients with infarctions of the posterior cerebral artery and found that 14% had homonymous macular defects (involving the central visual field without peripheral involvement). All of these defects, however, involved at least 10 degrees of the central vision, with the patients with infarction that did not extend beyond the most posterior portion of the calcarine cortex having the most focal defects.[8] As the central VF defect in this case was approximately 1 degree, only an extremely focal area of ischemia to the posterior tip of the occipital cortex could have resulted in such a small defect. Based on the patient's MRI results, we know that this was indeed the case. This patient had a history of multiple small embolic events, consistent with focal areas of ischemia. Such focal CVAs are likely underreported as they are more likely to be asymptomatic.

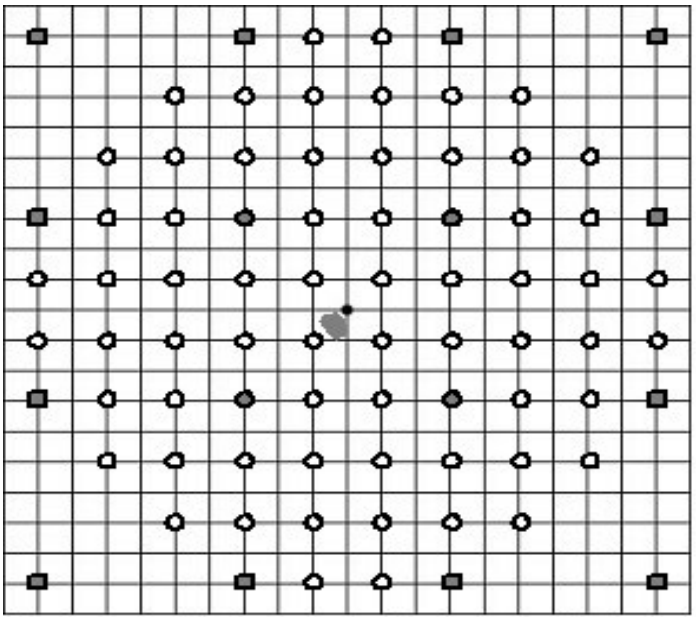

Figure 5. *Amsler Grid showing the patient's central scotoma with overlying diagram of points tested on a 10-2 VF (circles) and 24-2 VF (grey squares) to demonstrate how a small scotoma can be missed on standard automated perimetry.*

Standard testing for neurological VF loss usually involves automated central and peripheral perimetry in an outpatient setting and confrontation VF testing in an inpatient setting. Many smaller or more central defects are not picked up on confrontation testing.[8] Additionally, tangent screen VF testing may be beneficial, especially if a patient has difficulty with standard automated perimetry.

This case demonstrated that automated perimetry may miss defects if they are small. The standard stimulus size for both the 10-2 and 24-2 VF test is approximately 0.5° in diameter.10 In the 10-2 VF test, the central 4 points tested are each 1 degree from central fixation, with all test points being 2 degrees apart. In the 24-2 test, the central 4 points are 3 degrees from fixation, and all points tested are 6 degrees apart.[10] Therefore, a 10-2 VF test can miss scotomas that are less than 2 degrees in diameter, and a 24-2 VF test can miss scotomas that are less than 6 degrees in diameter. The patient had a defect less than 1 degree in extent and located less than 1 degree from fixation. Such a defect would not be picked up on a 10-2 and would only be shown with Amsler grid testing (Figure 5). For this case, Amsler grid testing allowed us to document the patient's scotoma and to monitor it over time.

Although small, the patient's central scotoma was very distracting to him as it was quite noticeable while he was reading. Our goal was to decrease his awareness of the scotoma so as to increase his reading fluency.

The observation that the patient did not notice the scotoma against a black background was incorporated in our rehabilitation considerations. Prescribing a video magnifier with reverse contrast capabilities reduced scotoma awareness and allowed the patient to read more fluently. We also used other low vision rehabilitation strategies such

as magnification, tints to enhance contrast, and lighting to maximize his visual efficiency. Magnification enlarges the retinal image, which effectively decreases the linear extent of the scotoma.[11] While formal eccentric viewing training was not utilized in this case, the concept of eccentric viewing was introduced to assist with scanning. For instance, the patient was encouraged to move his fixation downwards slightly (towards the lower portion of a letter) if he felt that he was missing part of a word while he was reading.

There are many rehabilitation considerations for patients with neurological visual field loss. Training patients to scan into their blind field can be key for both mobility concerns as well as for reading.[12] Patients with complete homonymous hemianopsias or quadrantanopsias often benefit from mobility training. Yoked prism or sectoral Fresnel prism can also be used to increase a patient's awareness of their blind field by shifting images from the blind field into the seeing field.[12] Line guides can also be beneficial. Some patients may require low vision devices with optical character recognition capabilities in order to be able to read fluently.

Patients with neurological visual field loss should be monitored for stability of their visual field defects periodically. They also may need to be counseled on their eligibility for driving based on their visual field loss.

Conclusion

Although uncommon, small neurological central scotomas can greatly impair a patient's reading ability. Low vision strategies such as magnification, enhanced and reverse contrast, and tints can be beneficial in these cases. While standardized automated perimetry generally is ideal for detecting neurological visual field loss, it can miss very small scotomas. Other visual field testing, such as a 10-2 VF, Amsler grid, or a tangent screen should be considered when standard automated perimetry does not detect visual field defects coinciding with the patient's symptoms. Amsler grid testing can be more sensitive and beneficial in uncovering very small central VF loss.

References

1. Mozaffarian D, Benjamin EJ, Go AS, Arnett DK, et al.; on behalf of the American Heart Association Statistics Committee and Stroke Statistics Subcommittee. Heart disease and stroke statistics—2016 update: A report from the American Heart Association. Circulation 2016;133:e38-e360.
2. National Stroke Association. Post-stroke conditions. http://bit.ly/2M8Bqgm (Last accessed 27 April 2017).
3. Rowe FJ, Wright D, Brand D, Jackson C, et al. A prospective profile of visual field loss following stroke: Prevalence, type, rehabilitation, and outcome. BioMed Res Int 2013;2013:719096.
4. Ali M, Hazelton C, Lyden P, Pollock A, Brady M. Recovery from poststroke visual impairment: Evidence from a clinical trials resource. Neurorehabil Neural Repair 2013;27(2):133-41.
5. Gilhotra JS, Mitchell P, Healey PR, Cumming RC, Currie J. Homonymous visual field defects and stroke in an older population. Stroke 2002;33:2417-20.
6. Rowe F, Brand D, Jackson CA, Price A, et al. Visual impairment following stroke: do stroke patients require vision assessment? Age Ageing 2009;38:188-93.
7. Suchoff IB, Kapoor N, Ciuffreda KJ, Rutner D, et al. The frequency of occurrence, types, and characteristics of visual field defects in acquired brain injury: A retrospective analysis. Optometry 2008;79:259-65.
8. Gray LG, Galetta SL, Siegal T, Schatz NJ. The central visual field in homonymous hemianopia. Arch Neurol 1997;54:312-7.
9. Isa K, Miyashita K, Yanagimoto S, Nagatsuka K, Naritomi H. Homonymous defect of macular vision in ischemic stroke. European Neurol 2001;46:126-30.
10. Heijl A, Patella VM, Bengtsson B. The Field Analyzer Primer: Effective Perimetry, 4th edition. Dublin, California; Carl Zeiss Meditec, Inc., 2012.
11. Lovie-Kitchen, J. Low Vision. Optometry: Science Techniques and Clinical Management, 2nd Ed. Edinburgh: Butterworth Heinemann Elsevier, 2009: 475-96.
12. Luu S, Lee AW, Daly A, Chen CS. Visual field defects after stroke: A practical guide for GPs. Austral Fam Phys 2010;39(7):499-503.

Correspondence regarding this article should be emailed to Noah Merhar, OD, at noahmerhar@gmail.com.

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