American Academy of Optometry: Case Report 2
Non-Exudative Age-Related Macular Degeneration and Low Vision Rehabilitation

Alexa Moy
2300 Jefferson Ave NE #C114
Renton, WA 98056
(858) 945-7212
moyalexa@gmail.com

Abstract
Age-related macular degeneration is a retinal disease which can result in severe visual impairments. The structural deterioration can be seen both clinically and with imaging. Microperimetry is a useful tool to compare the structural and functional loss related to the disease. The functional loss may impair the patient's ability to perform certain activities like reading and driving. However, low vision devices and rehabilitation may allow patients to regain the ability to perform these tasks. This case presents a patient with advanced non-exudative macular degeneration who wanted to improve his ability to complete daily activities. Low vision rehabilitation techniques such as visual training, adaptation, and low vision devices were utilized to make his daily activities easier.

Keywords: macular degeneration, low vision rehabilitation, low vision devices, activities of daily living

Introduction

Age-related macular degeneration (ARMD) is a progressive retinal disease that primarily affects central vision. Patients may notice distortion, faded colors, and increased difficulty seeing in the dark[1,2]. Several imaging modalities such as optical coherence tomography (OCT), fundus photography, and microperimetry can be used as not only clinical tools, but also as educational tools to demonstrate to the patient how ocular health relates to visual symptoms[3,4]. Imaging improves the ability to monitor and manage patients, which ideally reduces vision change progression through patient compliance with treatment and follow up appointments. For example, microperimetry images allow the provider and patient to view the exact location of the macular degeneration that is affecting visual function. Comparison of all images over time allows us to follow the advancement of the disease. When patients use this visual tool to see their disease progression, they may become more complaint with management.

Visual symptoms can impair activities of daily living. For example, driving may become increasingly difficult hindering the ability of affected patients to transport themselves to different locations independently[5]. The lack of independence can then reduce quality of life. Although some patients may legally meet visual driving requirements, they may still be a danger to themselves and others due to reduced vision. For this reason, it is necessary to explain the situation to the patient with due diligence and include alternative forms of transportation.

Case Report

A 70 year old Caucasian male presented at the blind rehabilitation optometry clinic for a low vision exam on February 14, 2019 with the complaint of difficulty reading small labels and seeing in the dark. He also inquired if he should drive due to trouble seeing centrally. The patient was referred by the primary care clinic for low vision devices to help with near work. His last comprehensive eye examination was on December 14, 2018.

The patient's ocular history included age-related non-exudative macular degeneration, cataracts, and dry eye syndrome in both eyes. Ocular surgery included bilateral blepharoplasty in the 1980s. No ocular trauma was reported. Medical history consisted of insomnia, clonic seizure, postherpetic neuralgia, gastroesophageal reflux disease, hypertension, depression, arthritis, coronary artery disease, hypothyroidism, sleep apnea, and post-traumatic stress disorder. Medications included carbamazepine, gabapentin, levothyroxine, melatonin, omeprazole, prazosin, simvastatin, AREDS multivitamins, and artificial tears. Allergies included bee stings and penicillin. Family medical and ocular history were unremarkable. He was alert and oriented to time, place, and person.

Examination:

The patient's entering distance eccentric fixation visual acuities with spectacle correction were 20/40+2 right eye tilting his head to the left and 20/40-2 left eye tilting his head down. Manifest refraction was +2.00-1.25x088 with 20/32+2 right eye and +2.00-1.00x114 with 20/40-2 left eye. Near visual acuity with +2.50 ADD over the manifest refraction was 1.25M at 30 cm. Confrontation fields with no eccentric viewing were full peripherally at all quadrants in both eyes. Tests not performed during this low vision

exam included extraocular muscles, cover test, pupils, anterior and posterior segment because the patient had an eye examination two months prior. Findings from the last visit were unremarkable except for his longstanding bilateral advanced non-exudative macular degeneration with geographic atrophy and cataracts in both eyes.

Although the patient's calculated magnification demand to see 1.0M was 4.2 diopters, the lowest power hand magnifiers available started at 10 diopters. Magnification demand was calculated by multiplying 1.25M by the inverse of 0.30 meters (3.33 diopters). Since the patient reported trouble seeing at near especially in low light settings, the Eschenbach mobilux 3.5x10D hand magnifier with built in lighting was first trialed. He adapted well to the device and achieved 0.6M. Since the device may be too bulky to bring outside the house, a 10 diopter Mattingly Donegan swing-out pocket magnifier was also trialed with the ability to achieve 1.0M which the patient felt would be very helpful as he could easily carry the device to the grocery store to look at labels. Additionally, a Luxo 5 diopter Lighter duty magnifier lamp was trialed. The patient appreciated that he could work at near with extra lighting with both hands available.

Visual aids that were ordered which would take 4-6 weeks to receive included the Eschenbach mobilux, pocket magnifier, magnifier lamp, bifocal glasses, and prescription sunglasses. Since the patient's ocular health included macular degeneration and cataracts, a fundus examination and microperimetry were ordered to explore macular function and rule out additional retinal etiologies of night vision difficulties.

Differential Diagnosis:

The differential diagnoses regarding reduced night time vision included:

- Retinitis pigmentosa
- Glaucoma
- Choroideremia
- Age-related macular degeneration
- Cataracts

Retinitis pigmentosa is a genetic disorder causing degeneration of the photoreceptor cells in the retina. Symptoms include decreased night vision, gradual loss of peripheral vision and in advanced cases reduced central vision. Characteristic signs of the disease include optic nerve head pallor, attenuated retinal vessels, and bone spicules which were not reported in the patient's previous posterior segment evaluation. Additionally, the patient reported no family history of the condition and no peripheral vision loss had been detected.

Glaucoma results in progressive damage to the optic nerve which may cause difficulty with night vision and peripheral field loss. However, the patient's eye pressures were normal and no glaucomatous defects of the optic nerve head were noted in previous charts.

Choroideremia is an inherited retinal disease with gradual atrophy of the retinal pigment epithelium, photoreceptors, and choroid. The first symptom is typically night blindness in early childhood followed by tunneling field loss and reduced vision. Signs of the condition, which were absent in the patient, include widespread retinal pigment epithelium clumping, visible sclera, and large choroidal vessels. Again, the patient had an unremarkable family ocular history and no report of visual symptoms as a child.

Age-related macular degeneration is the deterioration of the macular region leading to reduced central vision and color vision, metamorphopsia, and loss in contrast sensitivity. Characteristic signs noted in the patient's previous posterior segment examination included drusen, geographic atrophy, and retinal pigment epithelial changes distinctive of advanced non-exudative macular degeneration. The patient had been diagnosed with the condition since at least 2010.

Cataracts result in the clouding of the crystalline lens with symptoms consisting of blurry vision, glare, poor night vision, and reduced color vision. Previous chart notes reported cortical and nuclear sclerotic cataracts which contributed to the patient's visual difficulties including night time driving, although the majority of visual symptoms most likely stemmed from macular degeneration.

Follow Up #1

The patient returned to the low vision rehabilitation clinic on February 21, 2019 for microperimetry and fundus examination. He reported stable vision since his last visit. Spectacle corrected distance visual acuity with head tilt was 20/40 right eye and 20/40-2 left eye. Pupils were equal, round, and reactive to light without afferent pupillary defect. Extraocular muscles were full without restrictions in all gazes. Cover test at distance with correction was orthophoric. Confrontation fields were stable and full peripherally at all quadrants in both eyes. Intraocular pressures were 17 mmHg right eye and 16 mmHg left eye with Goldman applanation tonometry at 12:55 pm.

Anterior segment evaluation with slit lamp biomicroscopy included meibomian gland dysfunction and dermatochalasis in both eyes. Lashes, conjunctiva, and cornea were clear. The irises were brown and flat. Anterior chamber was deep and quiet without cells or flare. Angles were deep with anterior chamber angles of 4/4 via Von Herrick method.

The patient was dilated with 1.0% tropicamide and 2.5% phenylephrine at 1:00pm. Posterior segment evaluation with slit lamp biomicroscopy and binocular indirect ophthalmoscope indicated symmetrical 1+ cortical and 1+ nuclear sclerotic cataracts in both eyes. Vitreous was clear without posterior vitreous detachment. Optic nerves were pink and flat with distinct margins. Cup-to-disc ratio of right eye was 0.25 round and left eye was 0.20 round. Right eye macula had geographic atrophy about 2 disc diameters centered nasal to the fovea, drusen temporal to the atrophic area, and deep retinal pigment disruption throughout the posterior pole. Left eye macula had geographic atrophy about 1.5 disc diameters centered nasal to the fovea, drusen at the edges of the atrophic area, and deep retinal pigment disruption throughout the posterior pole. Vessels were attenuated in both eyes. Right eye peripheral retina had drusen throughout greater superior-nasal and RPE clumping inferior and inferior-nasal. Left eye peripheral retina had superior drusen.

The Cirrus macular optical coherence tomography (OCT) indicated severe disruption of the outer retinal layer with areas of atrophy and no subretinal fluid in both eyes (Fig 1). The patient had difficulty fixating, so retinal nerve fiber layer OCT was unreliable. Microperimetry was performed and demonstrated reduced sensitivity in areas nasal and superior nasal to the fovea associated with geographic atrophy locations in both eyes (Fig 2 and 3). The greatest macular function in the right eye was temporal to the fovea. In the left eye it was inferior-temporal to the fovea.

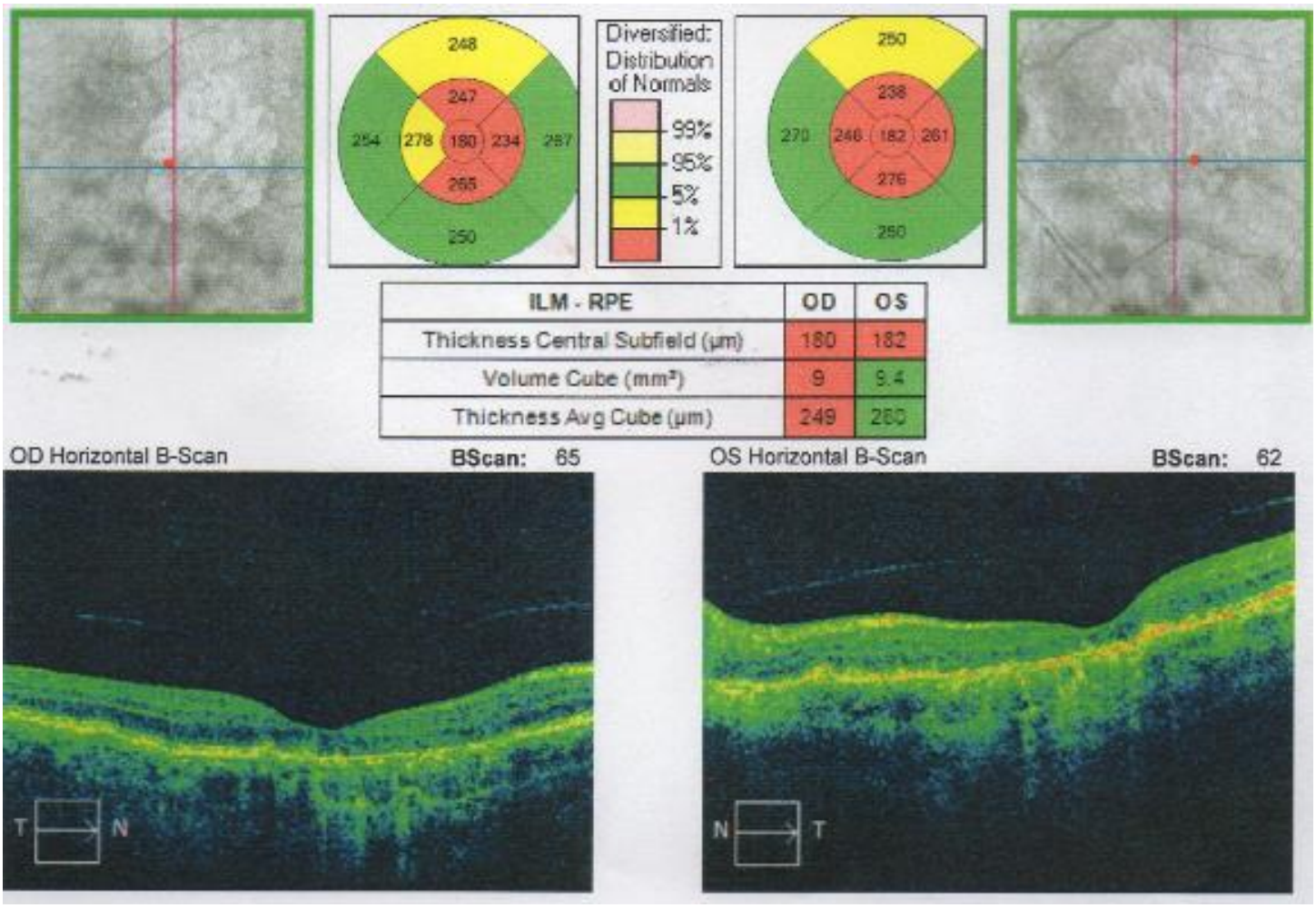

Figure 1. Macular optical coherence tomography demonstrated thinning with geographic atrophy and scattered drusen in both eyes.

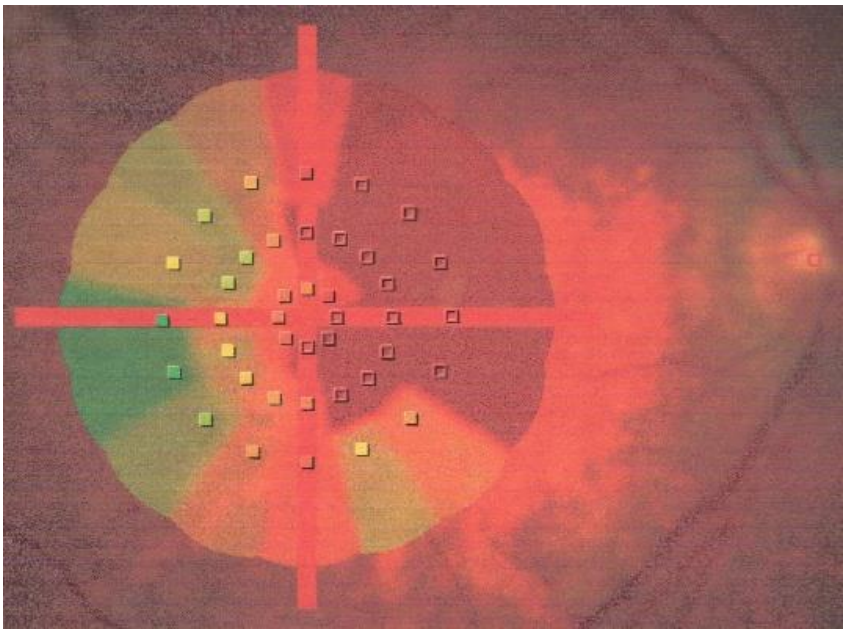

Figure 2. Microperimetry of the right eye indicated areas of atrophy associated with reduced macular function nasal and superior-nasal to the fovea and highest sensitivity temporal to the fovea.

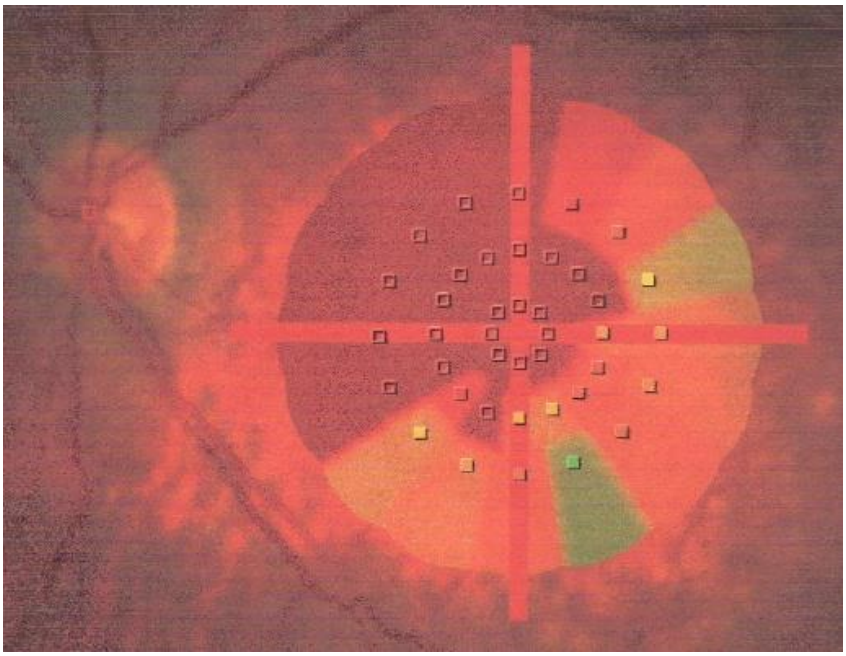

Figure 3. Microperimetry of the left eye indicated areas of atrophy associated with reduced macular function nasal and superior-nasal to the fovea and highest sensitivity inferior-temporal to the fovea.

Using the OCT and microperimetry images, the patient was educated about the location and size of his macular degeneration. OCT (Fig 1) was used to show him the drusen and atrophy. The importance of his continued use of the AREDS multivitamins twice a day was emphasized to delay disease progression.

Microperimetry images (Fig 2 and 3) were helpful to show the patient the specific areas of atrophy which reduced his vision. Areas marked red indicated reduced visual sensitivity and related atrophy. In contrast, areas marked green were locations the patient could see more clearly with less degenerative defects. Microperimetry helped the patient understand why the recommended head tilt and eye positioning techniques would give him the best eccentric viewing.

The patient was informed that UV light can harm the melanin in the macula and worsen his macular degeneration. The melanin protects the macula from oxidative stress. Thus, UV protection sunglasses were recommended. The patient was informed AREDS and UV protection may delay progression of his dry ARMD to the wet form. Follow-up exams were necessary to monitor any changes. If the ARMD progressed to the wet form, he would be referred to an ophthalmologist for an evaluation and possible ocular injections.

The patient understood that macular degeneration and cataracts were most likely the components causing poor night vision. Although cataract surgery was a possible option to reduce night vision problems, his condition was mild and the patient did not want surgery in the near future. Additionally, it was recommended to use artificial tears up to four times a day or as needed in both eyes for dryness and to have a better quality tear film for clearer vision. Since the patient was curious if he met the visual driving requirements, a kinetic visual field was scheduled for his next visit.

Follow Up #3

The patient returned to the low vision rehabilitation clinic on February 28, 2019 for a kinetic visual field to test for driving eligibility. The patient reported he had given up driving long distances, night driving, and driving in bad weather. Central spectacle corrected distance visual acuity without eccentric fixation and no head tilt was 20/64 right eye and 20/160 left eye.

Kinetic visual field was performed with right eye normal isopter at all fields of view and left eye mild superior constriction possibly secondary to dermatochalasis (Fig 4).

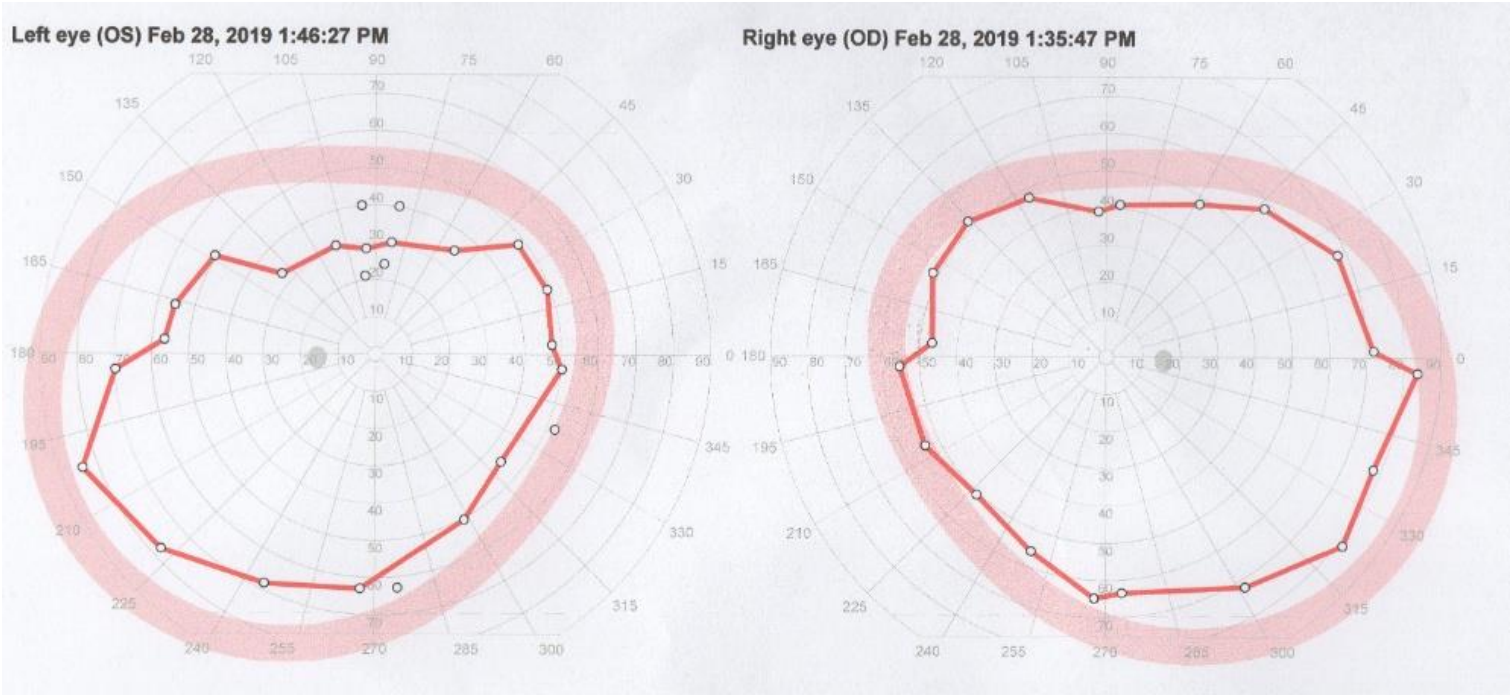

Figure 4. Kinetic visual field indicated full peripheral field in the right eye and mild superior constriction left eye.

The patient was informed by the low vision specialist about the Washington state driving license visual requirements including central vision and peripheral field of view. He was shown the visual field results (Fig 4) which demonstrated a full peripheral field in the right eye and mild superior constriction and full field in other quadrants in the left eye. Although the patient met the peripheral vision driving requirements, his central vision reduction would call for restrictions to his driver's license. The patient was cautioned about driving due to borderline visual acuities, so he was given information about alternative modes of transportation including government funded vans and public transit. Since the patient stated he may want to drive again in the future, he was offered a referral to have a comprehensive driving evaluation with the Rehabilitation Driving program. The patient reported understanding, but wanted to wait to contact the Rehabilitation Driving program for a consult. The patient was to return to the primary care clinic as scheduled for his regular 3-4 month macular degeneration follow up exam.

Discussion

Age-related macular degeneration (ARMD) is a progressive retinal disease and leading cause of blindness in the developed world[1,2]. Prevalence increases with age with about 2.4% occurrence in those over 50 years old to 12.2% over 80 years old[1]. The disease prevalence also depends on ethnicities with highest frequency in Caucasian descent and lowest in African descent. Systemic risk factors include cardiovascular disease, high body mass index, hypertension, and high plasma fibrinogen. Modifiable risk factors include cigarette smoking, high salt and fat intake, and heavy alcohol consumption. Other risk factors include family history, exposure to ultraviolet light, light skin color, light iris color, and female predilection. The presence of complement factor H (CFH) and Age-Related Maculopathy Susceptibility Gene 2 (ARMS2) are additional genetic risk factors[1].

Macular degeneration is classified into two main types (non-exudative dry ARMD and exudative wet ARMD) and four categories[1]:

1. No ARMD: No drusen or few small drusen (<63 um)
2. Early ARMD: Multiple small drusen or few medium drusen (63-124 um) with mild retinal pigment epithelium changes (RPE)
3. Intermediate ARMD: Numerous medium drusen, at least one large druse (>125 um) or geographic atrophy not involving the center of the fovea
4. Late ARMD: Exudative ARMD or geographic atrophy from non-exudative ARMD involving the foveal center

Drusen are yellow or white buildup of extracellular material. They include a lipid and waste product called amyloid beta, which build up in retinal layers including Bruch's membrane and retinal pigment epithelium (RPE)[1,2]. Approximately 80% of people over 60 years old have some drusen which have no visual significance[1]. The two main types of drusen are hard and soft. Soft drusen are larger, not as well demarcated, and characteristic of higher risk of ARMD progression[1]. There is less risk with hard drusen due to the smaller size and hence less scar formation. Pigmentary changes include both hyper and hypo pigmentation. Hyperpigmentation is due to focal pigment clumping. Hypopigmentation is associated with loss of RPE cells, outer layers of the retina, and choriocapillaris leading to atrophy[1]. Retinal atrophy may present as isolated patches that merge around the fovea leading to geographic atrophy, ultimately encompassing the central macular region.

Drusen, pigmentary changes, and atrophy in dry ARMD can be seen clinically without imaging. However, several imaging modalities help with patient education, management, and treatment. Fundus autofluoresence and near-infrared reflectance imaging detect atrophic areas due to loss of RPE cells. In this case, autofluoresence and near-infrared reflectance imaging were not necessary to perform as the dense atrophy was visible with biomicroscopy. Optical coherence tomography (OCT) offers a cross-sectional view of the retinal layers to assess for drusen, atrophy, and subretinal fluid[3]. The patient's OCT indicated a fairly stable amount of atrophy without subretinal fluid. Fluorescein and indocyanine green angiography help detect ARMD neovascularization[3]. These images were not necessary as OCT and clinical findings indicated no exudation. Microperimetry quantitatively maps retinal sensitivity based on retinal degeneration[4]. The device is similar to a visual field test by measuring sensitivity threshold in decibels while also overlaying the information on a fundus image. This imaging modality is beneficial for those with retinal diseases like ARMD who may have difficulty focusing on the target due to eccentric

fixation by tracking retinal movement. This patient's microperimetry results showed reduced sensitivity in locations associated with atrophy in the macular region. The data also corresponded to the patient's concern of central vision difficulty.

Macular degeneration may cause visual deficits such as blurry central vision, difficulty seeing in low light levels, and visual distortions leading to decreased function in activities of daily living. In more advanced cases, as in with this patient, geographic atrophy results in worsening visual symptoms that increases difficulty with near work, driving, facial recognition, and mobility[5,6]. Sivaprasad *et al.* interviewed participants with geographic atrophy from macular degeneration. The study found patients have difficulty performing daily tasks, leisurely activities, and walking which causes them to become more prone to falling.[5] Such impact can cause emotional complications like fear of worsening vision or blindness, and frustration due to difficulty completing tasks as the patients' independence decreases.

Although there is no treatment for dry ARMD, close management of the condition is crucial. Nutritional supplements such as Age-Related Eye Disease Study (AREDS) multivitamins and UV protection help reduce the progression of the disease[2]. Currently, several studies are examining therapies that target pathogenic pathways of the disease. These include anti-inflammatories, neuroprotection, lipofuscin/visual cycle inhibitors, choroidal blood flow restoration, and stem cell-based therapies[7]. In this case, the patient reported good compliance with his AREDS multivitamins twice a day.

The Age-Related Eye Disease Study Research Group examined participants with varying stages of dry and wet forms of macular degeneration. They were given either a placebo, 80 mg zinc oxide, antioxidants (vitamin C 500 mg, vitamin E 400 IU, beta-carotene 15 mg), or a combination of zinc and antioxidants. Production of free radicals due to oxidative stress leads to cellular damage. In the retina, antioxidant agents include vitamins C and E as well as carotenoids. Vitamins C and E help protect the retina from blue light damage while carotenoids reduce lipid oxidation[2]. Carotenoids include beta-carotene which is also called provitamin A[2]. Zinc is the most abundant metal in the retina and is located in layers most affected by macular degeneration. The metal is a cofactor of many enzymes which help reduce inflammation in the retina which can give rise to the disease[2]. Results from the study indicated the combination of zinc and antioxidants significantly delayed progression from dry to wet macular degeneration compared to placebo[1].

In another study, the Age-Related Eye Disease Study Research Group found lutein and zeaxanthin supplements were more effective than beta-carotene to help delay progression of macular degeneration to more advanced stages[1].

Owsley *et al.* reported older adults with ARMD have greater difficulty driving compared to age-matched adults without the disease[8]. In the geriatric population, many tend to "self-regulate" their driving which includes when, how much, and under what circumstance they drive. For example, patients with ARMD report they use extreme caution like pulling over when another car passes, relying on memory of the location, using a "copilot" or passenger for help, and frequent scanning[8].

A study by Wood *et. al* assessed driving performance of those with ARMD compared to age-matched controls. The drivers with ARMD as a group made more errors observing the environment, finding relevant information on road signs, seeing road markings and other road users, planning ahead such as yielding, changing lanes, and pulling in or out of traffic[9]. Thus, it is important to fully assess driving ability of those with ARMD.

Driving requirements for a non-restricted Washington state license include binocular visual acuity of 20/40 or better with or without corrective lenses and a total visual field of 110 degrees or more in the horizontal meridian with both eyes[10]. A restricted driving license may be permitted if the patient passes the visual field requirement and can see between 20/40 and 20/70 binocularly[10]. A restricted license tends to limit where and when the person may drive. For example, most are unqualified for night time driving but are allowed to drive during the day. Patients using bioptics in Washington state are permitted to pass the non-restricted license test as long as visual acuity of 20/40 is met. A bioptic is a small telescope that is mounted on a corrected spectacle lens and used intermittently to spot distant objects with better detail.

A study by Bowers *et. al* reported that drivers with macular degeneration found bioptics enhanced quality of life[11]. These drivers reported less difficulty driving compared to those with ARMD who do not use bioptics[11]. Although the participants reported better driving ability with the device, it is valuable to make sure the patient is a good fit cognitively and physically for the visual aid. In this case, the patient was told about bioptics but due to the severity of his disease and his reporting that he might have adaptation difficulty, the bioptic device was deferred.

There are several other low vision devices and rehabilitation techniques useful for patients with macular degeneration. For example, this patient reported difficulty with near work, so hand magnifiers and lamp magnifiers were trialed with the patient noting improvement with reading and seeing finer detail. As the disease progresses, the type of devices are changed as needed. In more advanced cases, electronic magnifiers and CCTVs may be more beneficial. One rehabilitative technique includes identifying the patient's preferred retinal loci, and training the patient to use the specific areas of the eye with the best vision[12].

Based on the microperimetry results of the patient in this case, he was informed where the best vision was located in each eye for eccentric viewing. Additionally, information about the Blind Rehabilitation Outpatient Specialist (BROS) program was given to improve orientation, mobility, and activities of daily living. Since this patient expressed concerns about driving and may qualify for a restricted driver's license, the Driver Rehabilitation for Veterans with Disabilities Program was suggested. This program evaluates patients who have limited physical and mental disabilities. If deemed qualified, the rehabilitation program helps veterans acquire the skills to drive safely as well as provide assistance in selecting an appropriate vehicle and equipment[13].

Conclusion

When macular degeneration becomes visually debilitating, activities of daily living and quality of life are diminished. During eye examinations, it is essential to assess residual vision and to note any changes in disease severity, which may continue to reduce ocular health and function. With the use of imaging modalities such as optical coherence tomography, disease progression may be followed over time. Microperimetry images allow providers and patients to see the structural changes of the retina which corresponds to functional changes. Many patients do not understand the extent of their disease and visual impairments until they see it on an image. The images serve as a powerful tool to encourage patient treatment compliance. Microperimetry shows providers the exact areas of retinal sensitivity in order to recommend specific viewing techniques. The takeaway in this case is that microperimetry

images were useful as an educational tool to show the patient the exact locations where he had the best vision. The patient then used this knowledge to adapt to proper head tilt and eye positioning techniques for eccentric viewing.

With low vision rehabilitation and aids, most patients are able to continue their hobbies with increased ease. However in cases where devices do not sufficiently help patients perform desired activities, low vision rehabilitation programs may be more beneficial. In this case, the patient reported concerns with driving. Because he was eligible for a restricted driver's license, he was given contact information about the Driver Rehabilitation for Veterans with Disabilities Program to evaluate if he is a safe driver and for assistance with driving skills. The patient was also presented with transportation alternatives.

Although visual function with macular degeneration does not improve, management with UV protection and AREDS can help delay the progression of the disease. Thus, patient education and treatment compliance along with follow up appointments are essential when monitoring ARMD.

References

1. Waugh, Norman, *et al*. Treatments for Dry Age-Related Macular Degeneration and Stargardt Disease: a Systematic Review. *Health Technology Assessment*. 2018; 22:1–168.
2. Carneiro, Ângela, and José Paulo Andrade. Nutritional and Lifestyle Interventions for Age-Related Macular Degeneration: A Review. *Oxidative Medicine and Cellular Longevity*. 2017; 2017:1–13.
3. Garrity, Sean T., *et al*. Multimodal Imaging of Nonneovascular Age-Related Macular Degeneration. *Investigative Ophthalmology& Visual Science*. 2018; 59(4).
4. Midena, E, and E Pilotto. Microperimetry in Age: Related Macular Degeneration. *Eye*. 2017;31:985–994.
5. Sivaprasad, Sobha, *et al*. Living with Geographic Atrophy: An Ethnographic Study. *Ophthalmology and Therapy*. 2019; 8:115–124.
6. Taylor, Deanna J, *et al*. How Does Age-Related Macular Degeneration Affect Real-World Visual Ability and Quality of Life? A Systematic Review. *BMJ Open*. 2016; 6(12).
7. Bandello, Francesco, *et al*. Recent Advances in the Management of Dry Age-Related Macular Degeneration: A Review. *F1000Research*. 2017; 6: 245.
8. Owsley, C and G McGwin. Driving and Age-Related Macular Degeneration. *Vis Impairment & Blindness*. 2019;102(10).
9. Wood, Joanne M., *et al*. Effects of Age-Related Macular Degeneration on Driving Performance. *Investigative Ophthalmology*. 2018; 59(1):273.
10. Getting a Driver License: Medical and Vision Screening. WA State Licensing (DOL) Official Site. Online: www.dol.wa.gov/driverslicense/medicalvision.html.
11. Bowers, Alex R., *et al*. Bioptic Telescope Use and Driving Patterns of Drivers with Age-Related Macular Degeneration. *Translational Vision Science*. 2016;5(5):1-18.
12. Hooper, Phil, *et al*. Age-Related Macular Degeneration and Low-Vision Rehabilitation: A Systematic Review. *Canadian Journal of Ophthalmology*. 2008;43(2):180–187.
13. VA Handbook 1173.16 - Driver Rehabilitation for Veterans with Disabilities Program. Online: helpdesk.vetsfirst.org/index.php?pg=kb.chapter&id=303.