Madeleine Jones
BVisSci MOptom
Eyewear on Pako, Victoria

An atypical case of multiple evanescent white dot syndrome

Multiple evanescent white dot syndrome (MEWDS) is an acute inflammatory disorder characterised by painless monocular vision loss and numerous, small (100-200µm), white-grey spots extending across the retina, with the fovea being spared. The rare condition is more likely to occur in young females, and myopia is an associated risk factor.[1]

Up to 50% of MEWDS cases may be preceded by a flu-like viral prodrome several weeks before the onset of symptoms.[2] Common symptoms include monocular reduction in vision, an enlarged blindspot, photopsia and floaters. In addition to the extrafoveal white dots, foveal granularity is also considered characteristic of MEWDS. Other signs may include mild vitritis and mild disc oedema. OCT scans may demonstrate disruption to the ellipsoid zone and either dome-shaped or vertically linear hyper-reflectivity at the level of the outer retina.[3] Typically, MEWDS resolves spontaneously without treatment over a period of weeks to months. However, recurrence and secondary choroidal neovascularisation have been documented in rare cases.[1]

Case report

A 65-year-old Caucasian female presented with a three-day history of blurred vision and floaters in the left eye. Visual acuities were R 6/6 and L 6/6= with habitual correction of R -4.25/-1.25x85 and L -5.75/-1.25x105. Aside from a medical history of asthma and breast cancer, which was treated with local radiotherapy, good general health was reported. The patient denied experiencing any flu-like symptoms in the weeks preceding presentation.

The patient has a longstanding posterior staphyloma at the left optic nerve (**Figure 1**). Asymmetric intraocular pressures have also been monitored since 2021.

Dilated fundus examination demonstrated scattered multifocal hypopigmented lesions in the left eye. The macula was spared of the retinal lesions, and no foveal granularity was observed. The appearance of the white dots was more prominent in coloured fundus photography than with fundoscopy. Mild vitritis was also observed in the left eye. There was no presence of optic nerve oedema and the left posterior staphyloma remained unchanged. Examination of the right eye was unremarkable.

Fundus autofluorescence demonstrated hyperfluorescence of the lesions (**Figure 2**). OCT scans revealed disruption to the ellipsoid zone (IS/OS layer) in areas corresponding to the white dots (**Figure 3**).

The patient attended an ophthalmology appointment 3 days later, at which time the left acuity had dropped from 6/6= to 6/9.5=. Blood tests, including syphilis serology, and a chest X-ray were completed to exclude masquerading chorioretinopathies. These returned negative results, so a diagnosis of multiple evanescent white dot syndrome was presumed. Maxidex was

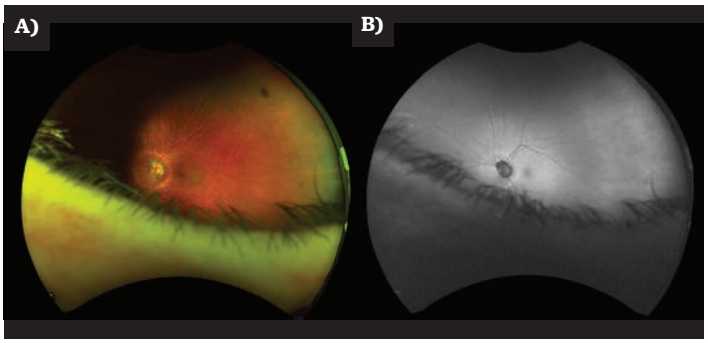

Figure 1.

A) Widefield colour fundus photo and **B)** fundus auto fluorescence of the left eye, taken five months prior to the onset of MEWDS. A longstanding peripapillary staphyloma is present.

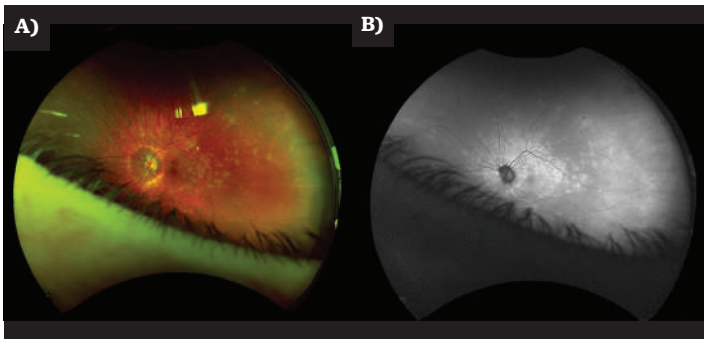

Figure 2.

A) Widefield colour fundus photograph and **B)** fundus auto fluorescence demonstrate multiple, light-coloured lesions that particularly surround the superior arcade. The fovea is spared of any lesions.

prescribed to treat the vitritis and the patient was monitored for self-resolution of the condition. By 8 months, the white dots had fully resolved and the patient was discharged from ophthalmology care. When the patient was reviewed in the optometry setting 9 months post initial presentation, visual acuities had returned to R 6/6 and L 6/6=. Fundus photographs at this visit demonstrated increased hypoautofluorescence and pigmentary change surrounding the left optic nerve (**Figure 4**).

Discussion

MEWDS was first described as a form of posterior uveitis in 1984.[4] Whilst the pathogenesis of MEWDS is not fully understood, it is believed that inflammation of the choriocapillaris leads to choriocapillaris hypo- or non-perfusion.[5] This disease mechanism is shared by other conditions that together are termed as

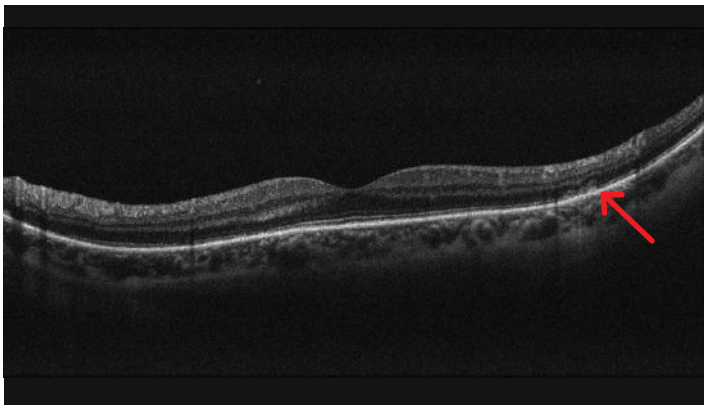

Figure 3.

OCT scan demonstrating disruption to the ellipsoid zone (IS/OS junction) and hyper reflective debris corresponding to deep retinal white dots.

'primary inflammatory choriocapillaropathies' (**Table 1**).[6] Previously, these conditions were included under the misnomer of 'white dot syndromes'. Since the advent of multimodal imaging, it has been determined that other conditions in this group have different underlying disease mechanisms.[2]

Unlike MEWDS, which is considered benign and reversible, the other primary inflammatory choriocapillaropathies tend to affect the larger choriocapillaris vessels or precapillary vessels, leading more severe disease and permanent scarring.[6] Additionally, MEWDS is also the only condition in this group that tends to occur unilaterally.

Multiple evanescent white dot syndrome
Punctate inner choroidopathy
Multifocal choroiditis
Acute posterior multifocal placoid pigment epitheliopathy
Serpinginous choroiditis

Table 1:

Primary inflammatory choriocapillaropathies

In 2021, the Standardisation of Uveitis Nomenclature (SUN) working group outlined the key classification criteria for MEWDS.[3] These were:

1. Multifocal grey-white chorioretinal spots with foveal granularity;
2. Characteristic fluorescein angiography 'wreath-like' hyperfluorescence on fluorescein angiography, and/or OCT scans demonstrating domed or vertically linear hyper-reflective lesions affecting the ellipsoid zone;
3. Absent-mild anterior chamber and vitreous inflammation.[3]

Exclusion criteria included positive serology for syphilis and evidence of sarcoidosis. Bilateral disease was also considered as an exclusion criterion, given that 96% of MEWDS cases are unilateral.[3]

In the case presented, the classification criteria outlined by the SUN working group were met; with the exception of foveal granularity, which was not observed at the patient's initial presentation. It is not known whether foveal granularity was noted at subsequent ophthalmology reviews. Foveal ganularity has been reported in 70-94% of MEWDS cases.[2] It is also well

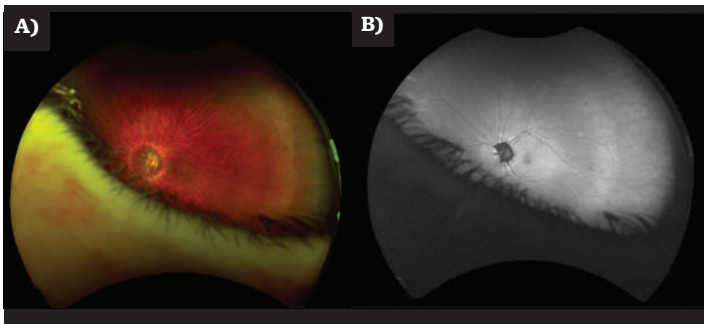

Figure 4.

A) Widefield colour fundus photograph and **B)** fundus auto fluorescence, taken 9 months after the onset of MEWDS, depict resolution of the white dots. There is an enlarged area of peripapillary atrophy and corresponding hypoautofluorescence when compared to baseline fundus photographs (**Figure 1**).

documented that in cases where initial presentation is delayed, foveal granularity may be the only sign of MEWDS, given the short lasting or 'evanescent' nature of the white dots.[7] This highlights that foveal granularity and multifocal grey-white chorioretinal spots may not present concurrently.

Also uncharacteristic in this MEWDS case was the age at which the patient was affected. MEWDS is most likely to occur in individuals aged 20 to 40 years, and cases outside this age group are atypical. However, at least 2 other cases of patients in their seventh decade having MEWDS have been reported.[8] It has been suggested that primary intraocular large cell lymphoma should be considered as a differential diagnosis, especially in patients older than 50 years, as early disease has been documented to mimic the appearance of MEWDS.[9]

Conclusion

Ultimately, MEWDS is considered as a form of choriocapillaritis, where full visual recovery and reversal of the condition is expected without treatment. The classification criteria outlined by the SUN working group form a sound framework to determine the presence of MEWDS disease, but exceptional cases do occur. Despite its self-resolving nature, any case of MEWDS requires referral to ophthalmology for systemic investigation to rule out any possible masquerading conditions. This is especially true in older patients presenting with MEWDS-like disease, as there is a greater need to exclude intraocular lymphoma. •

1. Marsiglia M, Gallego-Pinazo R, De Souza EC, Munk MR, Yu S, Mrejen S, Cunningham Jr ET, Lujan BJ, Goldberg NR, Albini TA. Expanded clinical spectrum of multiple evanescent white dot syndrome with multimodal imaging. Retina 2016; 36: 64-74.
2. Papasavvas I, Mantovani A, Tugal-Tutkun I, Herbort CP. Multiple evanescent white dot syndrome (MEWDS): update on practical appraisal, diagnosis and clinicopathology; a review and an alternative comprehensive perspective. Journal of Ophthalmic Inflammation and Infection 2021; 11: 45.
3. Classification Criteria For Multiple Evanescent White Dot Syndrome. American Journal of Ophthalmology 2021; 228: 198-204.
4. Jampol LM, Sieving PA, Pugh D, Fishman GA, Gilbert H. Multiple Evanescent White Dot Syndrome: I. Clinical Findings. Archives of Ophthalmology 1984; 102: 671-674.
5. Kuznetcova T, Jeannin B, Herbort CP. A case of overlapping choriocapillaritis syndromes: multimodal imaging appraisal. Journal of ophthalmic & vision research 2012; 7: 67-75.
6. Papasavvas I, Herbort CP. Diagnosis and Treatment of Primary Inflammatory Choriocapillaropathies (PICCPs): A Comprehensive Overview. Medicina 2022; 58: 165.
7. Mantovani A, Invernizzi A, Staurenghi G, Herbort CP. Multiple Evanescent White Dot Syndrome: A Multimodal Imaging Study of Foveal Granularity. Ocular Immunology and Inflammation 2019; 27: 141-147.
8. Lim JI, Kokame GT, Douglas JP. Multiple evanescent white dot syndrome in older patients. Am J Ophthalmol 1999; 127: 725-728.
9. Shah GK, Kleiner RC, Augsburger JJ, Gill MK, Jampol LM. Primary Intraocular Lymphoma Seen With Transient White Fundus Lesions Simulating the Multiple Evanescent White Dot Syndrome. Archives of Ophthalmology 2001; 119: 617-620

Pauline Xu
BOptom (Hons) MOptom GradCertOcTher
Lead Clinician – Retinal Dystrophies
Centre for Eye Health, Sydney

Managing retinal disease in the COVID era

The SARS-CoV-2 pandemic has brought unprecedented challenges to healthcare service[1] and the eyecare discipline is no exception. Patients who are highly vulnerable to permanent vision loss due to retinal disease such as neovascular age-related macular degeneration (AMD), retinal detachment, or proliferative diabetic retinopathy, are even more at risk during the pandemic.[2] In this case report, we describe a patient whose care was interrupted by the pandemic, demonstrating how effective and timely assessment and collaboration between optometry and ophthalmology is critical in optimising patients' visual outcomes.

A 66-year-old Caucasian male was referred to Centre for Eye Health (CFEH), University of New South Wales, by her optometrist for management of macular disease in the left eye. Relevant medical history includes well-controlled hypertension and heavy smoking until about 16 years ago. There was no family history of AMD. He was previously under the care of public hospital ophthalmology with a possible diagnosis of early AMD. However, his follow-up appointment was cancelled due to the COVID-19 pandemic.

Clinical examination results and multimodal imaging of the macula are provided in **Table 1** and **Figure 1** respectively. In the right eye, colour fundus photography (CFP) showed a few scattered drusen in the posterior pole and hypopigmentary abnormality in the right inferonasal macula (**Figure 1a**); spectral domain optical coherence tomography (OCT) showed focal outer retinal loss and thinning of retinal pigment epithelium (RPE) corresponding to the hypopigmentary disturbance (**Figure 1b**). In the left eye, CFP showed a hypopigmented lesion in the central macula (**Figure 1c**) and OCT showed a pigment epithelial detachment (PED) with overlying subretinal and intraretinal fluid, together with an epiretinal membrane (ERM) (**Figure 1d**). OCT angiography (OCTA) of the left macula (6 mm x 6 mm) showed hyper-intense flow signal at the 'RPE–RPE fit' en face slab (**Figure 1e**) with corresponding B-scan showing flow within the PED and below the RPE (**Figure 1f**). The optic nerve heads and the peripheral retina were unremarkable in both eyes.

	RIGHT EYE	LEFT EYE
Refraction and visual acuity	+3.00 (6/6-2)	+3.50 (6/19+1)
Pinhole acuity	No improvement	No improvement
Amsler grid	Clear	Metamorphopsia nasal and temporal to fixation
Intraocular pressures (icare tonometry) @3:50pm	16mmHg	16mmHg
Slit lamp examination	Open angles, early nuclear sclerosis both eyes	

Table 1.
Relevant clinical results from a 66-year-old patient with reduced vision in the left eye.

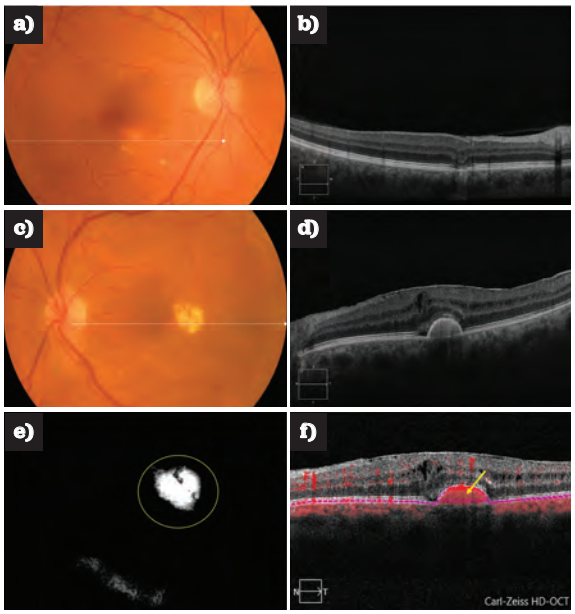

Figure 1.
Multimodal imaging of the macula from a 66-year-old patient with reduced vision in the left eye. Right eye **a)** colour fundus photography and **b)** OCT B-scan, showing macular drusen and outer retina thinning and RPE disturbance corresponding to the hypopigmented lesion. Left eye **c)** colour fundus photography and **d)** OCT B-scan showing epiretinal membrane, intra- and subretinal fluid and a pigment epithelium detachment. Note the hypertensive flow signal **(white circle)** on **e)** OCTA *en face* RPE-RPE fit slab and **f)** corresponding B-scan overlay showing flow underneath the RPE **(yellow arrow)**. *Red* shading = vascular flow signal.

The suspicious OCTA finding together with the reduced vision, disproportionate to the severity of ERM and nuclear cataract, suggested possible type 1 (sub-RPE) choroidal neovascularisation (CNV). The patient was recommended to see a retinal ophthalmologist within one week. He elected to return to public hospital ophthalmology due to financial concerns, however, we were unable to secure an immediate appointment due to the significant backlog of delayed appointments. Subsequently, as a failsafe plan, we scheduled a 3-month review at the CFEH while the patient waited for the hospital to arrange an ophthalmology appointment.

The patient returned to the CFEH after 3 months with no further contact from the hospital. The right eye was stable, however, best-corrected vision in the left eye reduced by one line to 6/24+1. OCT showed diffuse thickening of the macula (**Figure 2**, top panels) with increased intraretinal fluid suggesting active exudation (**Figure 2**, bottom panels). Due to significant changes in the left macular integrity, the patient was referred promptly to a retinal ophthalmology clinic with available bulk-billed anti-VEGF treatment.

A diagnosis of type 1 (sub-RPE or occult) CNV was confirmed with fluorescein angiography (FA), while polypoidal choroidal vasculopathy (PCV) was excluded via indocyanine green angiography (ICGA). Although type 1 CNV associated with AMD is typically associated with patients over 80 years old,[3] the ophthalmologist proposed that the patient's history of heavy smoking may be related to earlier onset. The ophthalmologist proceeded with intravitreal Lucentis at the time of the visit, with two repeat treatments organised in the next 2 months.

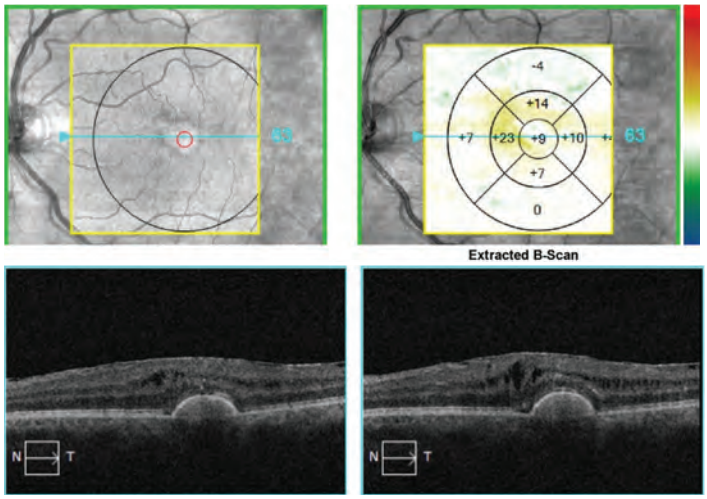

Figure 2.

OCT macular change analysis showing thickening of the retina **(top right panel)** and enlargement of the intraretinal cystic spaces **(bottom right panel)** at three-month follow-up compared to baseline. **(two left panels)**

Discussion

The COVID-19 pandemic imposed a massive barrier to retinal service delivery due to many reasons, including patients' reluctance to present for face-to-face healthcare, and reduced availability of healthcare providers.

Remote or electronic triaging and a decentralised care model have been proposed as potential solutions, although infrastructure is lacking, and institutional and organisational hurdles remain.[2]

The CFEH offers advanced diagnostic imaging and plays an important role in monitoring patients with chronic retinal diseases to improve triage accuracy and timeliness of referral, as demonstrated in this case.

Two major diagnostic dilemmas were present in this case. Firstly, there was equivocal evidence of CNV. The smooth, dome-shaped PED with homogeneous hyperreflective material was more consistent with a non-vascularised drusenoid PED, rather than the shallow, irregularly-shaped PED with heterogeneous hyperreflective material typically seen with neovascular PED. The cystic spaces were also diagnostically ambiguous, as these may have been due to ERM-related traction causing intraretinal hyporeflective pseudo-cysts, or may be a transudative rather than exudative process as seen at the edges of AMD-related PED.[7,8] There was also a lack of lacy-wheel, glomerular, sea fan, or dead tree tangled vessels[9] on en face OCTA, which are usually associated with active neovascularisation. On the other hand, the abnormal flow signal on OCTA B-scan overlay, the abnormally fast progression of intraretinal cystic spaces, and the suboptimal VA were highly suggestive of CNV and thus warranted immediate ophthalmological referral for FA confirmation.

The second dilemma relates to determining the cause of CNV. Pathological myopia and trauma were excluded based upon patient history, while other causes such as PCV, macular telangiectasia, inflammatory retinochoroidopathy, neoplasia, macular dystrophy were excluded due to discordance with clinical and multimodal imaging findings. Thus, AMD was rendered as the diagnosis of exclusion, particularly as it is the most common cause of CNV in the ageing population.[10] What is atypical is that drusen – the hallmark of AMD – did not have a pronounced presentation in this case, and the age of onset was earlier than expected for neovascular AMD.[3]

The management of this case demonstrates the value of optometrists in triaging chronic retinal diseases to reduce burden upon the public hospital system, thus improving patient accessibility to eyecare and visual outcomes. **Figure 3** illustrates the timeline of this patient's journey and showcases that effective and timely management, and collaboration between optometry and ophthalmology can be sight saving. Such a collaborative care model could be further explored to optimise service delivery in our increasingly ageing population, during and beyond the pandemic.

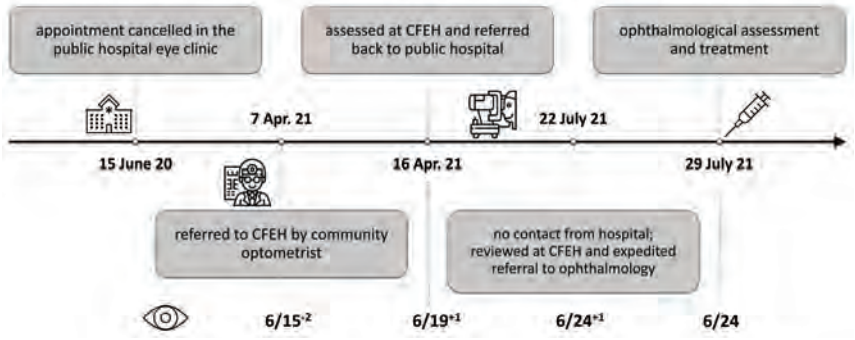

Figure 3.

Patient timeline showing how accessibility to eyecare and appropriate, timely management is key in vision preservation. Accessibility to eyecare and patients' visual outcomes can benefit from ongoing collaborative care between the primary optometrist, CFEH and ophthalmology.

The author would like to thank Mr Matt Trinh (PhD candidate, School of Optometry and Vision Science, UNSW) for reviewing the manuscript.

1. Kuehn BM. Despite Improvements, COVID-19's Health Care Disruptions Persist. JAMA 2021; 325: 2335-2335.
2. Faes L, Rosenblatt A, Schwartz R et al. Overcoming barriers of retinal care delivery during a pandemic—attitudes and drivers for the implementation of digital health: a global expert survey. British Journal of Ophthalmology 2021; 105: 1738-1743.
3. Jung JJ, Chen CY, Mrejen S et al. The incidence of neovascular subtypes in newly diagnosed neovascular age-related macular degeneration. Am J Ophthalmol 2014; 158: 769-779 e762.
4. Tan ACS, Simhaee D, Balaratnasingam C et al. A Perspective on the Nature and Frequency of Pigment Epithelial Detachments. Am J Ophthalmol 2016; 172: 13-27.
5. Cheung CMG, Lai TYY, Teo K et al. Polypoidal Choroidal Vasculopathy: Consensus Nomenclature and Non-Indocyanine Green Angiograph Diagnostic Criteria from the Asia-Pacific Ocular Imaging Society PCV Workgroup. Ophthalmology 2021; 128: 443-452.
6. Schmidt-Erfurth U, Waldstein SM. A paradigm shift in imaging biomarkers in neovascular age-related macular degeneration. Prog Retin Eye Res 2016; 50: 1-24.
7. Hilely A, Au A, Freund KB et al. Non-neovascular age-related macular degeneration with subretinal fluid. Br J Ophthalmol 2021; 105: 1415-1420.
8. Lek JJ, Caruso E, Baglin EK et al. Interpretation of Subretinal Fluid Using OCT in Intermediate Age-Related Macular Degeneration. Ophthalmol Retina 2018; 2: 792-802.
9. Coscas GJ, Lupidi M, Coscas F et al. Optical coherence tomography angiography versus traditional multimodal imaging in assessing the activity of exudative age-related macular degeneration: a new diagnostic challenge. Retina 2015; 35: 2219-2228.
10. Lim LS, Mitchell P, Seddon JM et al. Age-related macular degeneration. Lancet 2012; 379: 1728-1738.

Simi Sarin
BMedSc(VisSc), MOpt OcTher
OptoMed Norwood, Adelaide

A rare retinal finding

Clinical approach for recognising and managing retinal arteriovenous malformations.

Retinal arteriovenous malformations (AVMs) are rare, congenital vascular anomalies characterised by direct and abnormal blood flow between an artery and vein without an intervening capillary network.[1] The exact aetiology of AVMs is unknown but is believed to involve abnormal embryogenic development of the vascular mesoderm.[2] Females are more commonly affected than males, and in both cases the diagnosis is predominantly made before the age of 30.[2] Three main variants of AVMs have been described and these are distinguished by their differing severity and associated ocular manifestations (Table 1).[1]

AVMs may be an incidental finding on routine examination where they pose no threat to vision, although severe cases have been associated with ocular complications such as central retinal vein occlusion, vitreous haemorrhage, and retinal haemorrhage.[3] They may also be associated with systemic AVMs in the midbrain, mandible, or orbit as part of the Wyburn-Mason syndrome.[3] This case discusses an unusual AVM of the peripheral retina presenting as an incidental finding in a relatively healthy individual.

Case report

A 64-year-old Caucasian female presented for the first time at the practice for her annual eye exam. She did not report any visual concern, however, did note recent dizzy spells when lying in bed. There were no relevant issues identified in ocular, medical, or family history. General health review was unremarkable.

On general inspection she appeared well, and best corrected visual acuity was 6/6 in both eyes. Pupils were normal, motilities were full and smooth and there was no visible proptosis. Anterior eye examination was unremarkable. Fundus examination revealed an unusual vascular lesion in the inferotemporal left retina (Figures 1 and 2).

After dilated fundus examination, a cluster of aneurysmal retinal vessels surrounding an arteriovenous anastomosis was visible. There were exudates surrounding the lesion from the decompensated retinal vessels. An epiretinal membrane was also present in the same eye. No other retinal abnormalities were noted in either eye.

The patient was referred to a retinal specialist for fluorescein angiography and appropriate neuroimaging given the recent dizzy spells. A diagnosis of a type 2 retinal AVM was made. Neuroimaging of the head and orbit did not show any associated pathology. Given the lack of systemic AVMs, yearly observation was recommended for the patient.

Discussion

Retinal AVMs were first described by Magnus in 1874.[4] Since then, reports of retinal AVMs have been frequent and quite varied in presentation, ranging from localised lesions to lesions across the entire retina.[5] Localised AVMs have a predisposition for the superotemporal retina (41 per cent), papillomacular bundle (34 per cent) and nasal retina, in the perpipapillary region (four per cent).[5] The inferotemporal location of the AVM presented in this case seems to be a rare occurrence.[5]

Archer et al.[1] classified congenital AVMs based on their severity and relative prognosis (Table 1).

Both type 1 and 2 follow a stable and benign clinical course, and are rarely associated with systemic complications. Type 3 AVMs tend to involve the entire retina. These patients are at a higher risk of ipsilateral AVMs in the cerebrum and face as part of a

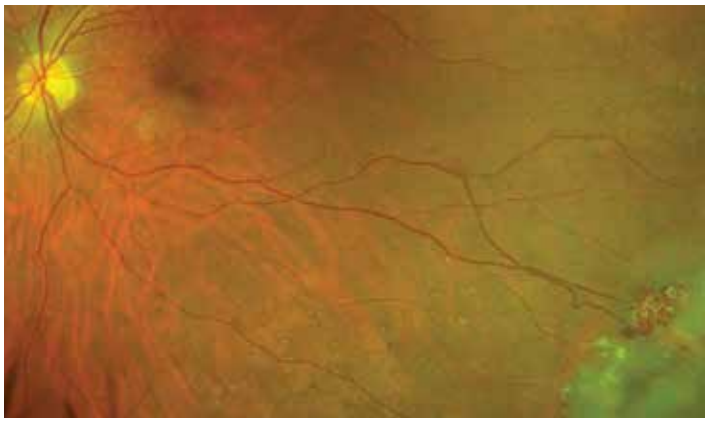

Figure 1.
Ultra-wide imaging of the vascular anomaly

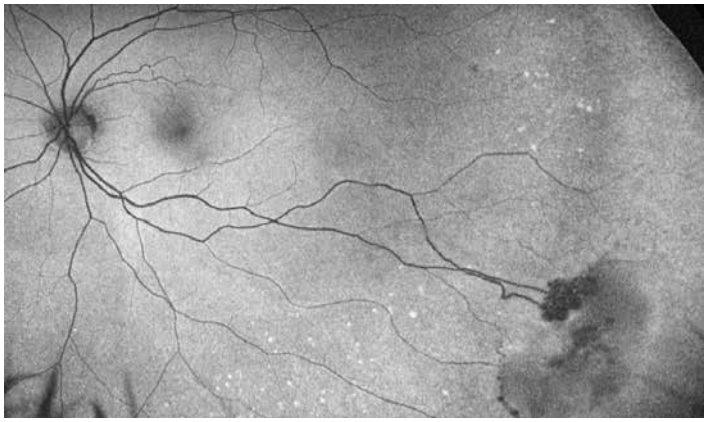

Figure 2.
Fundus autofluorescence imaging of the vascular anomaly

condition known as Wyburn-Mason syndrome.[6]

Wyburn-Mason syndrome is a rare disorder involving AVMs extending from the retina to the brain.[7] Other possible AVM locations include the skin, bones, kidney, and gastrointestinal tract.[7] The AVMs in Wyburn-Mason syndrome have minimal neoplastic potential; however, they can haemorrhage and result in significant morbidity.[7] The incidence of intracranial AVMs in patients with type 3 retinal AVMs is approximately 30 per cent.[7] However, changes in retinal AVMs do not hold any predictive value in the formation or progression of intracranial AVMs.[7]

Retinal AVMs can remain stable for years, slowly regress, or progress to affect other areas of the retina.[8] They are generally diagnosed as incidental findings or as the cause of visual impairment in low vision patients.[2] Severe cases can be associated with symptoms such as flashes, floaters, or sudden vision loss secondary to retinal complications.[2] These complications can include retinal or vitreous haemorrhages, retinal vein occlusion, and rarely neovascular glaucoma.[3]

Retinal complications secondary to AVMs are proposed to involve retinal ischemia stemming from three possible mechanisms. AVMs can result in increased venous flow and decreased retinal perfusion, causing the nearby retina/choroid to become ischemic from a 'steal phenomenon.'[8] In addition, abnormal vascular connections can increase hydrostatic venous pressure and damage vessel walls ultimately leading to thrombus formation.[8] Lastly, direct venous compression can cause an occlusive event.[8]

An optometrist may be the first to diagnose a retinal AVM or Wyburn-Mason syndrome. As per the classification system, not all patients with these lesions harbour cerebrovascular consequences. In severe forms of AVMs, extensive imaging including a CT scan, MRI, and carotid/vertebral angiography is recommended due to the increased risk of systemic complications.[9] Nonetheless, if a patient with a retinal AVM presents with suspicious symptoms (such as dizzy spells in this case), then neuro-radiological imaging is advised - regardless of the type of AVM.[9]

Asymptomatic AVMs do not require any treatment and routine observation should suffice.[9] In cases with associated retinal complications, management can include intravitreal anti-vascular endothelial growth factor (VEGF) agents, periocular steroids or photocoagulation.[9] Severe fundus lesions, as in Wyburn-Mason syndrome, unfortunately do not have any appropriate treatment due to the extent of retina involved.[10]

This case represents a rare presentation of an isolated type 2 retinal AVM. It is important for optometrists to be aware of the varying presentations of this disorder in order to appropriately manage their patients.

Table 1.
Variants of Retinal Arteriovenous Malformations[1]

Type	Characteristics	Retinal Signs	Visual Prognosis	Associations
I	Intervening abnormal capillary or arteriolar plexus between communicating artery and vein	- Involves one sector/quadrant of the retina - Appear as small complexes of dilating retinal vessels - Rarely decompensate nor cause structural abnormalities - Not associated with other retinal abnormalities	- Good - Visual acuity unaffected - No treatment required and can be observed	- Ocular or systemic associations are unusual
II	Direct arteriovenous communication between a branch artery or vein without an intervening capillary plexus	- End-to-end junctions or multiple anastomosis with intermediate (100-200 µm) or large (200-300 µm) channels - Afferent blood vessels may show beading with multiple fusiform dilations and occasional aneurysm formation - Capillary network may be dilated with small microaneurysms - Can lead to breakdown of blood retinal barrier causing leakage, retinal oedema, intraretinal exudates or haemorrhages	- Generally good but visual acuity can be affected if there is macular involvement - Tend to remain stable over time with rare instance of regression and remodeling - Ocular or systemic associations are unusual. No treatment required but photocoagulation can be used to reduce afferent artery calibre	- Usually not associated with intracranial vascular malformation - Carotid or cerebral angiography not necessary unless adequate clinical suspicion exists
III	Arteriovenous connections that are complex and extensive causing severe retinal complications and vision loss	- Cannot separate the arterial/venous components in these AVMs - Anastomoses involves large (500-600 µm) channels involving most of the fundus - Leads to widespread exudation, retinal oedema, and retinal cystic degeneration.	- Poor - Involves loss of vision at an early age - Requires neurosurgical therapy for cerebral complications	- Intracranial involvement very likely - Associated with Wyburn-Mason syndrome

1. Archer DB, Deutman A, Ernest JT et al. Arteriovenous communications of the retina. *Am J Ophthalmol* 1973; 75: 224-241.

2. Laey De J.J., Hanssens M. Vascular Tumors and Malformations of the Ocular Fundus, 1st ed. Ghent, Belgium: Springer Publishers, 1990.

3. Mansour AM, Wells CG, Jampol LM et al. Ocular Complications of Arteriovenous Communications of the Retina. *Arch Ophthalmol* 1989; 107: 232–236.

4. Reck SD, Zacks DN, Eibschitz-Tsimhoni M. Retinal and intracranial arteriovenous malformations: Wyburn-Mason syndrome. *J Neuroophthalmol* 2005; 25: 205-208.

5. Liu W, Sharma S. Ophthaproblem. Retinal arteriovenous malformations (type 2). *Can Fam Physician* 2005; 51: 203–209.

6. Stepanov A, Studnicka J, Dusová J et al. Vitreous hemorrhage due to a retinal arteriovenous malformation – case report. *Wulfenia* 2016; 23: 271-278.

7. Dayani PN, Sadun AA. A case report of Wyburn-Mason syndrome and review of the literature. *Neuroradiology* 2007; 49: 445-456.

8. Accou G, Nerinckx F, Leroy B et al. Vitreous Hemorrhage as Presenting Sign of Retinal Arteriovenous Malformation. *Case Rep Ophthalmol Med* 2020; 4: 1-4.

9. Knutsson KA, De Benedetto U, Querques G et al. Primitive retinal vascular abnormalities: tumors and telangiectasias. *Ophthalmologica* 2012; 228: 67-77.

10. Heimann H, Damato B. Congenital vascular malformations of the retina and choroid. *Eye (Lond)* 2010; 24: 459-467.

Dr Alex Hui
OD, PhD, GradCertOcTher, FAAO
School of Optometry and Vision Science, New South Wales

Herpes zoster case report

Herpes zoster (HZ) represents the reactivation of the varicella zoster virus after a previous, primary infection which established viral latency in nerve ganglia.[1] Unlike primary infections with the virus in children, which results in relatively mild symptoms of chicken pox, reactivation of the virus in adults typically leads to a painful, blistering rash limited to the dermatome of the nerve in which the virus was latent.[2]

When this reactivation occurs within the ophthalmic division of the trigeminal nerve, the presentation is known as herpes zoster ophthalmicus (HZO). In addition to a painful rash on the skin around the eye or forehead, HZO presentations can also affect numerous structures of the eye, including the cornea, conjunctiva, uvea, trabecular meshwork and retina.[2] Thus, HZO should typically be considered as part of a differential diagnosis and workup for numerous ocular conditions. HZO represents between 10 and 25% of herpes zoster presentations, and are of particular concern because they are more frequently associated with complications, particularly in the absence of treatment.[2]

Case study and examination

A 58-year-old woman presented with symptoms of itching, burning and tingling sensation on the skin of her right forehead and part of the skin around her right eye since the previous morning.

Corrected visual acuity was 6/6 in each eye, with normal pupillary reflexes and no restrictions of extraocular movements. There were no abnormalities observed on slit lamp evaluation of the anterior or posterior segments of the eye with pupillary dilation, with no signs of pseudodendrites on the cornea. The intraocular pressure in both eyes via Goldmann tonometry was 15 mmHg. Gross examination of the skin on her right forehead showed three distinct raised areas.

Directed questioning elicited a positive history of chickenpox infection in childhood, and otherwise no general health concerns or history of systemic diseases or medications which may cause immunosuppression. She also reported no history of allergy or use of any particular substances in the area around her forehead or eyes which may have caused an allergic reaction.

Diagnosis and management

Considering the symptoms of localised burning and the multiple raised areas only on the right side of the forehead, and an absence of any other history suggestive of causing discomfort, a working diagnosis of herpes zoster ophthalmicus was made. A discussion with the patient followed regarding treatment with oral antiviral medications, including the ideal timing of initiating therapy. She was informed of the greatest effectiveness of the medication in HZO treatment occurring if given within 72 hours of the rash onset, but at this stage the skin presentation was not conclusive. After further discussion regarding the relative safety of oral antiviral medications and expressing a desire to shorten the disease course and effects, the patient requested to be referred to her GP for a prescription of antiviral medications.

The patient was prescribed oral valaciclovir 1000 mg three times a day for seven days. She was also advised to manage any development of pain promptly with paracetamol and if insufficient to return to the GP for assessment. At follow up the next day her forehead took on the more characteristic HZ blistering rash appearance, however she fortunately did not develop any significant ocular complications.

Discussion

Herpes zoster management is centred on managing the acute episode, including the infection and associated pain, as well as mitigating long term risk of developing post herpetic neuralgia (PHN). In the eye, HZO presentations necessitate additional therapeutic goals of preventing or treating any associated inflammatory or infectious complications within the eye. This includes, among other conditions, HZO associated conjunctivitis, blepharitis, keratitis, uveitis, trabeculitis and retinitis. Diagnosis of HZO is typically made clinically, with the characteristic rash appearance often used as a definitive clinical feature, however this may not be obvious in the prodromal phases of the condition. Rarely, HZ can present without a rash at all (zoster sine herpete).[3]

Contemporary management of HZO utilizes systemic antivirals, as their use has been reported to decrease the rate of ocular complications from 50-60% down to 20-30%.[4] Available oral antiviral medications for HZ in Australia include aciclovir, valaciclovir and famciclovir. They all work through inhibition of viral DNA synthesis, and require viral thymidine kinase enzymes to become activated and thus are selective to only virally infected cells.[4] Incorporation of these molecules into the replicating viral DNA strand prevents further elongation.[4] Aciclovir has a relatively poor oral bioavailability of only 15-30%, thus requiring frequent (five times a day) and relatively high doses (800 mg each dose) to treat HZO.[4] In contrast, valaciclovir is a prodrug of aciclovir, which greatly improves bioavailability and results in higher peak circulating concentrations and a comparatively decreased dosage schedule (standard dosage for

HZO of 1000 mg three times a day).[4] Famciclovir is a prodrug of penciclovir, which is similar in structure to aciclovir and works through similar mechanisms. Famciclovir was developed to improve penciclovir's poor oral absorption.[4] The dosage for famciclovir to treat HZO in immunocompetent patients is 500 mg three times a day.[5] These drugs all have a favourable safety profile and are generally well tolerated by most individuals without any complications.[6] Some caution should be made for individuals who may have renal impairment as this drug is primarily excreted by the kidneys, and excessive concentrations may lead to neurotoxicity and hallucination, although this is reportedly rare.[2,4]

The majority of the literature and clinical trials have suggested that oral antiviral treatment for herpes zoster is most effective when given within 72 hours of the rash presenting

The majority of the literature and clinical trials have suggested that oral antiviral treatment for herpes zoster is most effective when given within 72 hours of the rash presenting.[4] For patients where the rash has begun to crust over, they are unlikely to gain benefit from antiviral use, however if there is evidence of continued rash development then treatment can be considered even if beyond the 72 hour window.[4] Given the high rate of complications, patients with HZO should be given oral antivirals even if outside of the 72 hour window in hopes of reducing associated ocular conditions.[2]

There is some controversy within the literature regarding whether antiviral use decreases the risk of PHN, primarily due to different definitions and measurement of PHN amongst different clinical trials.[2,4,6] Pain in the acute phases of the condition should be managed aggressively to reduce the risk of PHN developing later, with recommendations of using oral paracetamol alone or in combination with weak opioids as first line therapy before moving on to more aggressive treatments.[6] Involvement of a pain clinic or pain specialist early in the management of PHN is recommended.[6]

A live attenuated herpes zoster vaccine is available in Australia for patients 50 years or older. Up until 31st October 2021 it was available free of charge to patients aged 70-79 to encourage this group to be vaccinated, as they are expected to have higher rates of HZ as well as higher risk of developing PHN.[7] Timing of when the vaccine should be given is subject to debate within the literature, as the efficacy of the vaccine is expected to wane over time and at this stage booster shots are not recommended.[6] The current Australian Government Department of Health recommendation is for the vaccine to be given to those 50 to 59 years only if they live with someone with a weakened immune system.[8] The live, attenuated vaccine is not recommended in patients who are immunocompromised. A recombinant shingles vaccine using only part of the virus is also available in other jurisdictions such as the United States, and is thought to potentially provide longer term protection against HZ.[9] In a retrospective study, the recombinant vaccine was 89% effective in preventing HZO in patients 50 years of age or older over a two year period.[10]

Due to its potential for diverse presentations, HZO should be considered in presentations of inflammatory and infectious eye disease. If HZO is suspected or diagnosed, prompt management with oral antiviral medications should be initiated in coordination with the patient's GP, and pain managed aggressively to lower the risk of developing PHN and further ocular complications. •

1. Sauerbrei A. Varicella-zoster virus infections - antiviral therapy and diagnosis. GMS Infect Dis 2016; 4.

2. Wehrhahn MC, Dwyer DE. Herpes zoster: Epidemiology, clinical features, treatment and prevention. Australian Prescriber 2012; 35: 143-147.

3. Zhou J, Li J, Ma L et al. Zoster sine herpete: a review. Korean J Pain 2020; 33: 208-215.

4. Gnann JW. Antiviral therapy of varicella-zoster virus infections. In. Human Herpesviruses: Cambridge University Press, 2007. p 1175-1191.

5. Tyring S, Engst R, Corriveau C et al. Famciclovir for ophthalmic zoster: a randomised aciclovir controlled study. The British journal of ophthalmology 2001; 85: 576-581.

6. Cunningham AL, Breuer J, Dwyer DE et al. The prevention and management of herpes zoster. Med J Aust 2008; 188: 171-176.

7. Jayasinghe S, Sheridan S, Macartney K. Herpes zoster vaccination in Australia: what's available and who benefits? Aust Prescr 2020; 43: 2-6.

8. Department of Health Australian Government. Shingles (herpes zoster) immunisation service. https://www.health.gov.au/health-topics/immunisation/immunisation-services/shingles-herpes-zoster-immunisation-service-0. Published 2020. Accessed 26/07/2021.

9. Davis AR, Sheppard J. Herpes Zoster Ophthalmicus Review and Prevention. Eye Contact Lens 2019; 45: 286-291.

10. Lu A, Sun Y, Porco TC et al. Effectiveness of the Recombinant Zoster Vaccine for Herpes Zoster Ophthalmicus in the United States. Ophthalmology 2021.

This original case report was submitted by Optometry Australia member Hayley Birch in response to our call for papers.

Hayley Birch
BVis Sci, MOptom
OPSM Waurn Ponds, Victoria

Acute visual field loss:

Could a cotton wool spot cause that?

Cotton wool spots (CWSs) are grey-white retinal lesions with fluffy margins of 0.1-1 mm in diameter[1] and are found at the level of the innermost retinal layer. CWSs are composed of localised accumulations of axoplasmic debris within adjacent bundles of unmyelinated ganglion cell axons.[2] They occur after arterial occlusion at the borders of large ischemic areas but should not be regarded as a retinal nerve fibre layer infarct.[2] A discussion by McLeod argues that CWSs are better defined as cotton wool 'sentinels' or indicators of vascular insufficiency to a particular area within the retina, rather than retinal infarcts.[2]

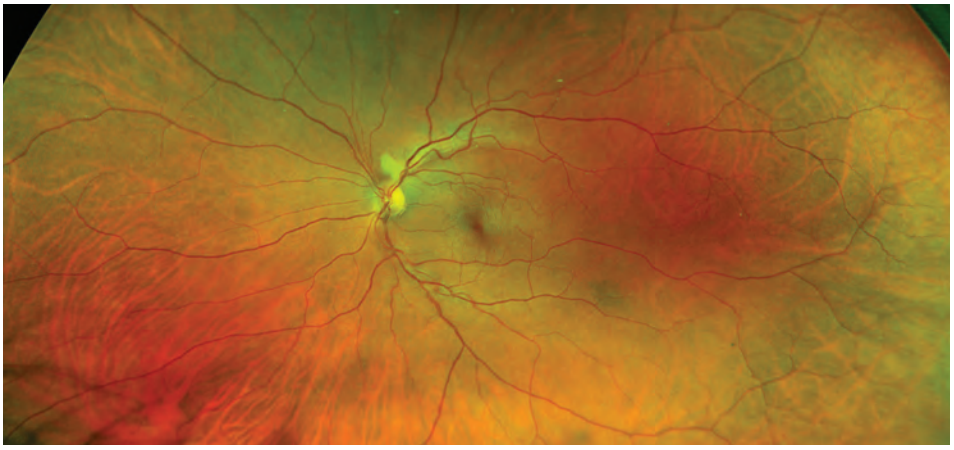

Figure 1.
CWS superior to the left optic nerve head

CWSs are uncommon in healthy patients, and therefore should be considered a 'red flag' for further investigation of systemic health. They are often seen in patients with diabetes, hypertension, retinal artery or vein occlusion and ischaemic optic neuropathy and often coexist with other features of retinopathy such as haemorrhages, exudates or oedema.[3,4] However, the presence of CWSs has also been reported in cases of systemic infection such as human immunodeficiency virus (HIV), immune and collagen vascular disorders, embolisms and malignancy.[3]

One study attempted to differentiate types of CWSs by their aetiology with a focus on CWSs caused by HIV compared to those seen in hypertension, diabetes and central retinal vein occlusions (CRVO). The study found that CWSs are smaller in size in HIV patients and the number of CWSs was higher in patients with CRVO.[3] There were no other significant differences reported.

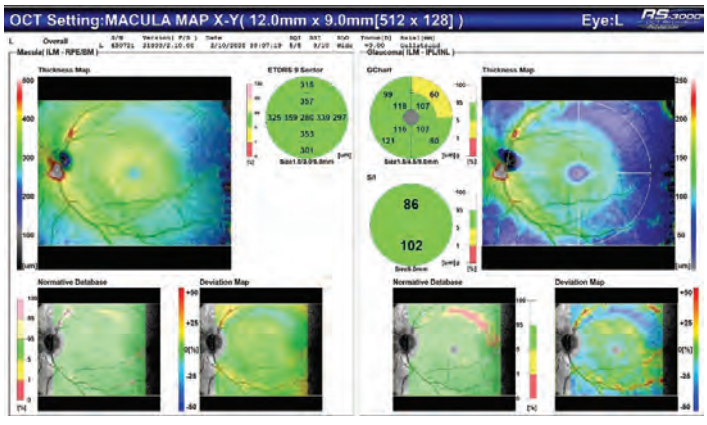

Figure 2.
OCT shows thickening of the inner retina at the location of the CWS

Due to the number of associated conditions, and the difficulty identifying a specific aetiology, both systemic and ocular differentials must be considered when CWSs are found on ophthalmoscopy. Ocular differentials for CWSs include any retinal lesion that appears white or yellow-white such as myelinated nerve fibre layer, commotio retinae, hard exudate, astrocytic hamartoma, retinal necrosis, active posterior uveitis, retinitis or vasculitis.[4]

Most patients with CWSs will be asymptomatic unless the cotton wool spot involves the central retina or fovea.[5] Reported symptoms include relative or absolute arcuate scotoma and/or blurred vision.[6] Following ophthalmoscopic resolution of the CWS, visual function does not always recover.[7] Differential diagnoses for these symptoms of CWSs include, but are not limited to, amaurosis fugax, migraine with aura, retinal detachment, optic neuritis and glaucoma.

Case report

Initial appointment

A 47-year-old male presented urgently to the practice reporting sudden, painless loss of his inferior visual field in his left eye. He did not have any recent trauma, photophobia or blurred vision. His prior ocular history showed a recent diabetic eye examination at the same practice a few months ago which showed no diabetic retinopathy of the left eye but a small isolated CWS in the right eye. At that time, the attending optometrist advised he quit smoking which the patient was successful in doing.

Vision was 6/6 in both eyes. Undilated fundus biomicroscopy showed resolution of the CWS in the right eye, but the presence of a new, larger CWS superior to the left optic nerve head (Figure 1).

Optical coherence tomography (OCT) demonstrated thickening of the inner retina at the location of the CWS. This is consistent with

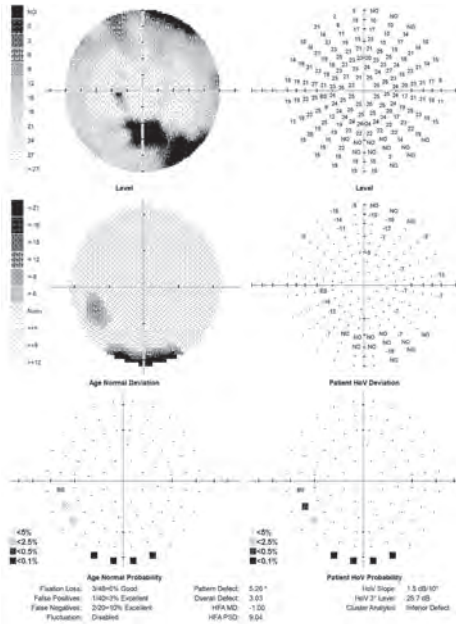

Figure 3.
inferior arcuate defect shown

the theory of accumulation of axoplasmic debris at the level of retinal ganglion cell axons,[2] visible with fundus biomicroscopy, as a CWS. There was also an adjacent area of arcuate shaped retinal thinning evident on the retinal thickness map (Figure 2). This could also be appreciated with the red-free filter on both slitlamp and on the ultra-widefield retinal image.

Medmont monocular full field testing showed an inferior arcuate defect in his left eye (Figure 3).

The patient was reassured that vision loss was related to the presence of the CWS with expectation of improvement over the next few weeks. A six-week review appointment was made and the patient was referred to see his general practitioner for a cardiovascular work-up (blood pressure measurement) and full blood test to rule out other underlying systemic causes including both vascular (hypertension, diabetes, hyperlipidaemia) and infectious causes of CWSs.

Review appointment

The patient returned for a review after six weeks reporting return of his left inferior visual field after approximately one week. Vision remained excellent at 6/6, undilated fundus biomicroscopy showed 90 per cent resolution of the CWS in the left eye (Figure 4). OCT was consistent showing reduced inner retinal thickening at the location of the CWS and no further thinning of the arcuate retinal defect. Medmont monocular full field testing showed significant improvement.

The patient reported he was undergoing further testing of his systemic health with his general practitioner. A three-month review was scheduled to check for complete resolution of the CWS.

Discussion

In patients with CWSs and no known medical history of diabetes, 20 per cent are found to have elevated blood glucose level and 50 per cent have an elevated blood pressure measurement (diastolic blood pressure of 90 mmHg or greater).[5]

Expected resolution of CWSs is relatively dependent on the underlying cause and size of the CWSs, as well as age of the patient. Most CWSs disappear within 4-12 weeks, however some studies have found that CWSs in diabetic patients can persist up to 8.1 months in those < 40 years old, and 17.2 months in those > 40 years old.[8] One study found that the small CWSs associated with HIV have a significantly shorter resolution time within an average of 6.9 weeks.[9] CWSs in hypertensive patients show similar resolution times to those seen in HIV patients. Overall, CWSs associated with hypertension or HIV resolve quicker than those associated with diabetes.

This case highlights the usefulness of OCT in giving optometrists not only a better understanding of the full extent of retinal pathology, but also enabling them to predict a visual outcome. OCT in the context of CWS will demonstrate retinal thickening at the location of the CWS on a retinal thickness map and generalised retinal thinning adjacent to the CWS. OCT radial scans show the location of the thinning extending from the ganglion cell layer to the outer plexiform layer.[7,10]

In this case, we saw a CWS with adjacent superior arcuate retinal thinning with a corresponding inferior arcuate visual field defect. We can use OCT technology in this same way to also predict visual field loss based on correlating retinal thinning in other cases such as glaucoma or stroke.[11]

Every CWS will leave a permanent inner retinal defect most easily appreciated with OCT due to the permanent change to the innermost retinal layers. Symptomatic vision loss due to CWSs is most likely caused by a CWS in the location of the posterior pole or fovea with the most common aetiology being underlying diabetes or hypertension.[5]

1. Dollery CT. Microcirculatory changes and the Cotton-wool Spot. *Proc Royal Soc Med* 1969; 62: 1267-1269

2. McLeod D. Why cotton wool spots should not be regarded as retinal nerve fibre layer infarcts. *Br J Ophthalmol.* 2005; 89: 229-237.

3. Mansour AM, Jampol LM, Logani S, et al. Cotton-wool spots in acquired immunodeficiency syndrome compared with diabetes mellitus, systemic hypertension, and central retinal vein occlusion. *Arch Ophthalmol* 1988; 106: 1074-1077.

4. Zimmerman J, Propes BD. Management of cotton wool spots in the retina. 2013, Oct. Accessed 10/10/20. <https://www.retinalphysician.com/issues/2013/october-2013/management-of-cotton-wool-spots-in-retina>

5. Purnima S. Patel, SriniVas R. Sadda. Retinal Artery Obstructions. In Retina (Fifth Edition), by Stephen J. Ryan, 1012-1025. Elsevier Inc., 2012.

6. Bek T, Lund-Andersen H. Cotton-wool spots and retinal light sensitivity in diabetic retinopathy. *Br J Ophthalmol* 1991; 75: 13–17.

7. Ioannides A, Georgakarakos ND, Elaroud I, Andreou P. Isolated cotton-wool spots of unknown etiology: management and sequential spectral domain optical coherence tomography documentation. *Clin Ophthalmol* 2011; 5: 1431-1433.

8. Eva M Kohner, C T Dollery, C J Bulpitt. Cotton-wool Spots in Diabetic Retinopathy. *Diabetes* 1969; 18: 691-704.

9. Mansour AM, Rodenko G, Dutt R. Half-life of cotton-wool spots in the acquired immunodeficiency syndrome. *Int J STD AIDS* 1990; 1: 132-133.

10. Zhang L, Xu L, Zhang JS, et al, Jonas J. Cotton-wool spot and optical coherence tomography of a retinal nerve fiber layer defect. *Arch Ophthalmol* 2012; 130: 913.

11. Alasil T, Wang K, Yu F, et al. Correlation of retinal nerve fiber layer thickness and visual fields in glaucoma: a broken stick model. *Am J Ophthalmol* 2014; 157: 953-959. doi:10.1016/j.ajo.2014.01.014

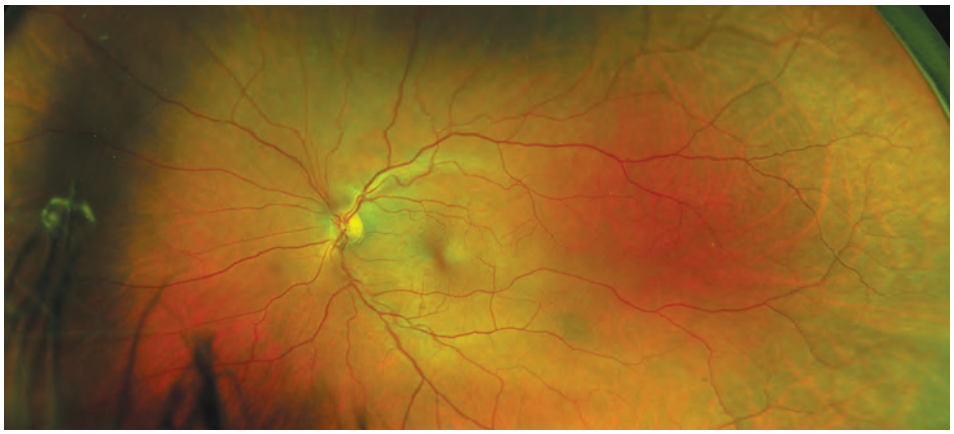

Figure 4.
A 90 per cent resolution of the CWS in the left eye

Adenoviral conjunctivitis

A challenging condition in a challenging time

Ka-Yee Lian
BOptom PGDipAdvClinOptom PGCertOcTher FACO

EyeQ Optometrists
Camberwell Victoria

Elaine Chong
MBBS MEpi PhD FAMS FRANZCO

Director of Ophthalmology Services
Royal Melbourne Hospital

Human adenovirus is the most common cause of infection in the conjunctiva, accounting for up to 75 per cent of all conjunctivitis cases, affecting people of all ages and demographics worldwide.[1] The most frequent presentation of adenovirus conjunctivitis is epidemic keratoconjunctivitis (EKC), typically caused by serotypes 8, 9, 19, 37, 53 and 54, followed by pharyngoconjunctival fever (PCF), usually caused by serotypes 1-5, 7 and 11.[1-3] Non-specific follicular conjunctivitis is another manifestation, primarily due to serotypes 1-11 and 19.[1,2]

The prevalence and incidence of adenoviral conjunctivitis is unknown as many affected people do not seek medical care.[1] Fortunately, most infections are mild and self-limiting, however there can be serious repercussions if complications are not recognised.

Patients with EKC usually present with a red, watery eye with possible foreign body sensation and photophobia. There may be ocular or periorbital pain and decreased vision in more severe cases. They may report flu-like symptoms, such as fever, malaise, myalgia and respiratory symptoms or a recent history of a family member being affected.

The common ocular signs include bulbar conjunctival redness, chemosis of the eyelid and conjunctiva, tarsal follicular reaction and petechiae.[1,4]

Pseudomembranes and true conjunctival membranes may form in EKC, ultimately causing subepithelial fibrosis and the formation of a symblepharon and punctal occlusion, which can lead to diplopia and ongoing epiphora.[1,4-6]

Corneal involvement distinguishes EKC from other adenoviral infections. Multifocal subepithelial infiltrates (SEIs) may typically develop seven to 10 days after the initial signs of infection, possibly reducing acuity and may persist for weeks to years.[1,4]

CASE REPORT

A 35-year-old Caucasian male presented to the Royal Victorian Eye and Ear Hospital with a unilateral red eye. He reported the conjunctiva in his left eye had been red accompanied by mild watery discharge for the previous three days.

He denied experiencing any cold or flu-like symptoms and was not in close contact with any other person with conjunctivitis.

He was not a contact lens wearer, denied taking any medication currently and was not aware of having any allergies.

On examination, visual acuities were R 6/6 and L 6/6. The bulbar conjunctiva was moderately hyperemic with prominent follicles present on the inferior palpebral conjunctiva of the left eye (Figure 1). His right eye was normal. Intraocular pressures were measured as R and L 13 mmHg with an iCare tonometer. All other ocular findings were unremarkable.

A diagnosis of possible adenoviral conjunctivitis was made and

Continued page 18

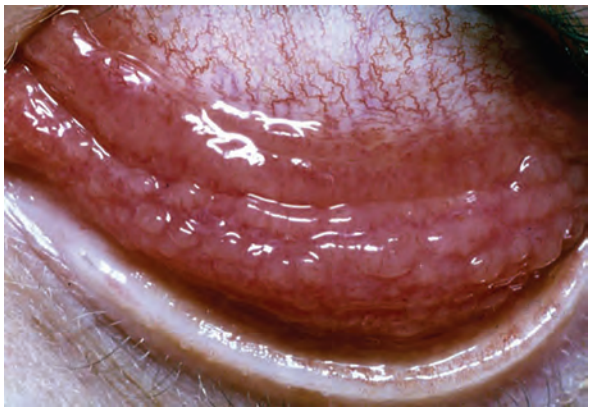

Figure 1. Inferior palpebral conjunctiva showing prominent follicles. Image: Royal Victoria Eye and Ear Hospital.

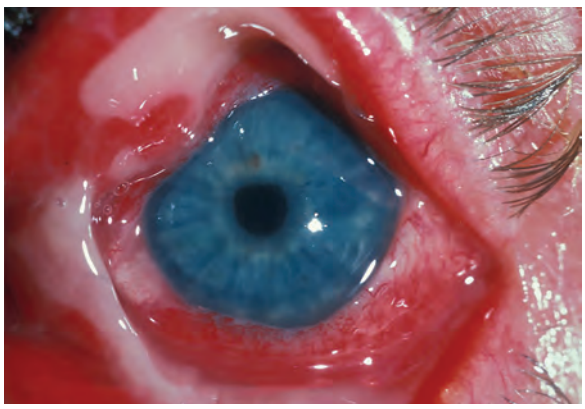

Figure 2. Inferior and superior palpebral conjunctivae with pseudomembranes. Image: Royal Victorian Eye and Ear Hospital.

Adenoviral conjunctivitiis

From page 17

the patient was advised to use preservative-free lubricants every two hours for the next two weeks or until symptoms resolve. He was also cautioned about being careful not to spread the infection to his right eye and to other members of his family. He was advised to take time off work as he was likely infectious. He was told to return if his left eye worsened, in particular to monitor for declining visual acuity and increasing discomfort over the next week.

Ten days later, the patient returned with bilateral swollen eyes complaining of increasing stringy and watery discharge. He had noticed that his symptoms were worsening.

On examination, pseudomembranes were present in the superior and inferior palpebral conjunctivae of the left eye, with marked conjunctival hyperemia and inflammation (Figure 2). There was an area of symblepharon which fortunately had not affected his eye movements, with no diplopia reported. His right eye had developed follicles only and there was no corneal involvement in either eye.

Visual acuity in the left eye dropped to 6/12, mainly due to the discomfort and discharge.

Debridement of the pseudomembranes was painstakingly performed, every two days, with Flarex cover to control the inflammation, until no further psuedomembranes formed. Careful use of steroid drops aims to limit the development of further symblepharon. Lubricants were continued regularly for relief of symptoms.

The patient's conjunctivitis resolved two weeks later, but the symblepharon remained permanently as an undesirable complication.

Discussion

The diagnosis of an adenovirus infection is typically made based on the history, symptoms and clinical findings. Laboratory diagnostic testing with polymerase chain reaction (PCR) is usually not performed due to the costs and time delay. The rapid antigen detection immunoassay may be a better alternative, carried out in-office with a result in 10 minutes.[1-4] Testing is done if there is uncertainty with the diagnosis as it is vital that the correct diagnosis is made before deciding on the management. Differential diagnoses of the more common forms of conjunctivitis with their key features are outlined in Table 1.[1,7,8]

Another differential diagnosis to consider, especially during the current novel coronavirus pandemic (COVID-19), is the SARS-CoV-2 virus.

There have been reported cases of acute viral conjunctivitis in patients with confirmed SARS-CoV-2 infection, often describing a sore throat, foreign body sensation, conjunctival redness, watery discharge, with the presence of palpebral conjunctival follicles and pre-auricular lymphadenopathy.[9] All patients with this presentation, seen during the pandemic, should be referred for COVID-19 PCR nasopharyngeal and throat swabs.

The symptoms and duration of adenoviral conjunctivitis can vary widely, with most resolving completely within three weeks. Supportive treatment, such as preservative-free artificial lubricants and cool compresses, can provide satisfactory symptomatic relief.[1,2,4,7]

Steroids should be restricted to cases of EKC with complications involving pseudomembranes or persistent subepithelial infiltrates (SEI) (Figure 3) which may reduce vision.[1,4,11] Ophthalmological referral may be necessary in these cases. Steroids reduce conjunctival and corneal inflammation, but may actually enhance adenoviral replication and increase the period of viral shedding, prolonging the entire clinical course of EKC.[4,9,12]

Before prescribing a steroid, it is important to rule out acute herpes simplex virus (HSV) conjunctivitis. Without accompanying skin or corneal involvement, herpetic clinical presentation may be very similar to adenoviral conjunctivitis,[2] comparative symptoms and signs shown in Table 1. Similarly, steroids drops in bacterial or Acanthamoeba infections can result in rapid deterioration, with severe corneal injury including corneal melts and perforation possible.[2]

Povidone-iodine (PVI), commercially available as Betadine, is a broad-spectrum antiseptic ophthalmic solution which has been used for many years to prophylactically reduce microbial flora prior to ocular surgery. PVI has been reported to

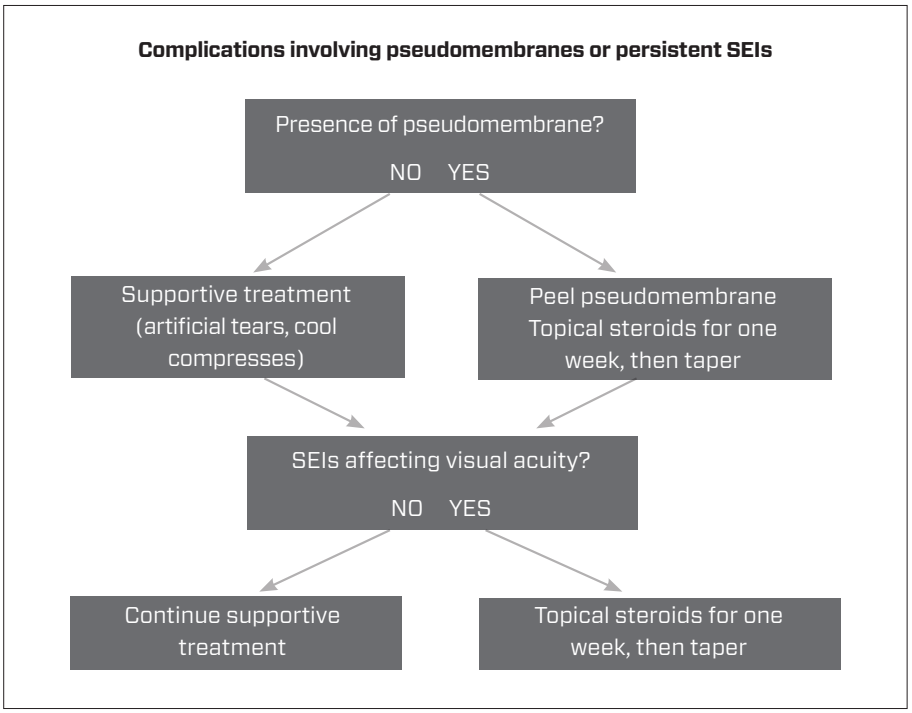

Figure 3. Treatment options for adenoviral conjunctivitis (Adapted from Pihos[4])

	Adenoviral	Herpetic	Bacterial	Allergic	Chlamydial
Unilateral/Bilateral	Begins unilateral, may become bilateral	Unilateral	Unilateral	Bilateral	Unilateral
Hyperemia	Generalised	Generalised	Generalised	Generalised	Generalised
Discharge	Watery	Thin, watery	Mucopurulent	Watery	Mucopurulent
Itching	Minimal	None	Minimal	Severe	Minimal
Palpebral involvement	Follicles Pseudomembranes	Follicles	Papillae	Papillae	Follicles
Pre-auricular lymphadenopathy	Common	Common	Uncommon	None	Common
Sore throat/fever	Occasional	Occasional	Occasional	Never	Never

Table 1. Differential diagnoses and key features of conjunctivitis

reduce the viral load and severity of symptoms *in vitro* and *in vivo* studies for the treatment of EKC, but there are no controlled trials supporting this treatment option,[11] so its use currently remains off-label.[12] After topical anaesthetic, a pre-irrigation non-steroidal anti-inflammatory drug (NSAID) drop is instilled followed by five drops of 5% povidone-iodine for 60 seconds. The lid margins are swabbed with 5% povidone-iodine and the ocular surface rinsed with sterile normal saline.[12] (That is: saline solution with 0.9% sodium chloride, as opposed to hypotonic or hypertonic saline solution).

Numerous trials have been underway worldwide to develop a safe and effective antiviral drug for ocular adenoviral infections,[3,12] including a topical treatment aimed at reducing symptom duration, currently recruiting at the Royal Victorian Eye and Ear Hospital.

EKC is highly contagious and easily transmitted through hand to eye contact or respiratory droplets and commonly from exposure to infective ophthalmic clinics or family members.[2,14] Adenoviruses, in their desiccated form, may remain viable and can be recovered at a clinically infectious concentration up to 28 days on dried plastic or metal surfaces.[15] Patient education is vital to minimise spread of the infection particularly in the two week period from when symptoms begin. Contact lens wearers should dispose of their lenses as the virus can survive in both chemical and hydrogen peroxide disinfection systems.[16]

Eye-care practitioners should also take additional precautions when examining patients with known or suspected adenoviral infections. Single-use instruments and equipment should be employed, such as disposable gloves, single-dose eye drops and disposable tonometer prisms or shields.[17,18] Adenoviruses can be resistant to many disinfectants, with recent data suggesting that 70 per cent isopropyl alcohol is ineffective.[19] Surfaces in the consulting and waiting rooms as well as frequently touched objects, such as door knobs and handrails, must be cleaned and disinfected regularly with a bleach-based solution.[19]

Although adenoviral conjunctivitis can be a common condition, its presentation and treatment may be quite variable and challenging to manage, with particular vigilance required during the COVID-19 pandemic. ▲

The authors would like to acknowledge the RVEEH for providing the photographic images.

1. Jhani V, Chan TC, Li EY et al. Adenoviral keratoconjunctivitis. *Surv Ophthalmol* 2015; 60: 435-443.
2. Kaufman HE. Adenovirus advances: new diagnostic and therapeutic options. *Curr Opin Ophthalmol* 2011; 22: 290-293.
3. Chigbu DI, Labib BA. Pathogenesis and management of adenoviral keratoconjunctivitis. *Infect Drug Resist* 2018; 11: 981-993.
4. Pihos AM. Epidemic keratoconjunctivitis: A review of current concepts in management. *J Optom* 2013; 6: 69-74.
5. Akkaya S, Ozkurt YB. Persistent symblepharon in an infant following epidemic keratoconjunctivitis. Med Hypothesis *Discov Innov Ophthalmol* 2016; 5: 74-77.
6. Chintakuntlawar AV, Chodosh J. Cellular and tissue architecture of conjunctival membranes in epidemic keratoconjunctivitis. *Ocul Immunol and Inflam* 2010; 18: 341-345.
7. Rhee DJ, Pyfer MF. The Wills Eye Manual: Office and Emergency Room Diagnosis and Treatment of Eye Disease, 3rd ed. Philadelphia: Lippincott Williams & Wilkins, 1999.
8. Azari AA, Barney NP. Conjunctivitis: A systematic review of diagnosis and treatment. *JAMA* 2013; 310: 1712-1729.
9. Chen L, Liu M, Zhang Z et al. Ocular manifestations of a hospitalised patient with confirmed 2019 novel coronavirus disease. *Br J Ophthalmol* 2020; 0: 1-4 (Epub ahead of print).
10. Romanowksi EG, Yates KA, Gordon YJ. Short-term treatment with potent topical corticosteroid of an acute ocular adenoviral infection in the New Zealand white rabbit. *Cornea* 2001; 20: 657-660.
11. Asena L, Singar Ozdemir E, Burcu A et al. Comparison of clinical outcome with different treatment regimens in acute adenoviral keratoconjunctivitis. *Eye* 2017; 31:781-787.
12. Labib BA, Minhas BK, Chigbu DI. Management of adenoviral keratoconjunctivitis: Challenges and solutions. *Clin Ophthalmol* 2020; 14: 837-852.
13. Pepose JS, Ahuja A, Liu W et al. Randomized, controlled phase 2 trial of povidone-iodine/dexamethasone ophthalmic suspension for treatment of adenoviral conjunctivitis. *Am J Ophthalmol* 2018; 194: 7-15.
14. Muller MP, Siddiqui N, Ivancic R et al. Adenovirus-related epidemic keratoconjunctivitis outbreak at a hospital-affiliated ophthalmology clinic. *Am J Infect Control* 2018; 46: 581-583.
15. Gordon YJ, Gordon RY, Romanowski E et al. Prolonged recovery of desiccated adenoviral serotypes 5, 8, and 19 from plastic and metal surfaces in vitro. *Ophthalmol* 1993; 100: 1835-1839.
16. Kowalski RP, Sundar-Raj CV, Romanowski EG et al. The disinfection of contact lenses contaminated with adenovirus. *Am J Ophthalmol* 2001; 132: 777-779.
17. Lian KY, Napper G, Stapleton FJ et al. Infection control guidelines for optometrists 2016. *Clin Exp Optom* 2017; 100: 341-356.
18. Fernandes P, Oyong K, Terashita D. Understanding infection prevention practices in optometry clinics. *Optom Vis Sci* 2020; 97: 24-27.
19. Centers for Disease Control and Prevention. Prevent Epidemic Keratoconjunctivitis (EKC), March 2019 [cited 17 May 2020]. Available from: https://www.cdc.gov/conjunctivitis/clinical/prevent-ekc.html

What lies beneath

The role of systemic conditions in normal tension glaucoma

Dr Steven Lam
BVisSci MOptom BHSci MOrth

Optiplex Eyecare
Melbourne VIC

This original case report was submitted by Optometry Australia member Steven Lam in response to our ongoing call for member papers.

Normal tension glaucoma (NTG) is a sub-category of glaucoma, a term used to describe a group of diseases that causes optic neuropathy. Glaucoma is characterised by the progressive loss of the retinal nerve fibre layer (RNFL), ganglion cell layer (GCL) and functional visual field.[1] Although the exact cause is unknown, common risk factors identified are people of Japanese ethnicity, an increase in age, raised intra-ocular pressure (IOP), decreased diastolic ocular perfusion pressure (DOPP) and obstructive sleep apnoea (OSA).[2,3] Glaucoma can be classified as 'open angle' which includes primary open angle glaucoma (POAG) where IOP is > 21mmHg or NTG with IOP < 21mmHg.[3,4] Other classifications include narrow angle and secondary glaucoma.[5]

Depending on the severity, patient symptoms can range from being asymptomatic to severe paracentral vision loss. Visual acuity (VA) can also decrease if macula fibres are affected.[4] The rate of peripheral vision loss in untreated NTG can vary from -0.2 decibel (dB) to > -2.0dB per year on a total deviation plot of a standard automated perimeter (SAP).[6] Clinically, NTG may present with optic disc haemorrhages, optic nerve rim thinning accompanied by peripapillary atrophy and IOP less than 21 mmHg. Asymmetry of IOP can also be seen with the more severely affected eye usually 1-2 mmHg greater than the other eye.[4] Medical imaging with optical coherence tomography (OCT) can show RNFL thinning at the optic nerve (ON) and GCL thinning at the macula region. Visual field (VF) examinations usually show arcuate defects respecting the horizontal mid line and will correspond to the region of RNFL/GCL loss.[4,7]

Management of NTG is targeted at lowering IOP to slow the progression of VF and RNFL/GCL loss. Current therapies include topical medication, selective laser trabeculoplasty (SLT), stent insertions and trabeculectomy. The decision to start management will depend on the rate of glaucoma progression and potential impact on the patient's quality of life.[4]

CASE REPORT

Mrs X, a 50-year-old female with a family history of glaucoma, presented for examination reporting gradual blurred vision over the last year.

Her best corrected distance VA was 6/6 OU and near vision was N5. Anterior chamber angle assessment revealed an open angle and no signs of secondary glaucoma. Assessment of the optic nerve through a Volk super 66 lens revealed large optic discs of R 1.9 mm and L 1.8 mm. The neural retinal rim (NRR) of the RE showed superior nerve excavation and an inferior notch (Figure 1) was found in the LE. Both NRRs had relative concentric thinning with a cup-to-disc ratio of R 0.7 and L 0.8. The patient's IOP was 21 mmHg bilaterally measured with a Perkins tonometer at 12:33pm and central corneal thickness was R 505 µm and L 499 µm.

Medical imaging results with an OCT confirmed that there was RNFL thinning of the superior rim in the RE and inferior rim in the LE. Ganglion cell complex analysis showed bilateral inferior thinning. Further investigation with a Zeiss Humphrey Field Analyzer 3 (24-2) showed a normal RE and central defects in the LE. Analysis with a 10-2 (Figure 2) revealed significant superior arcuate defects

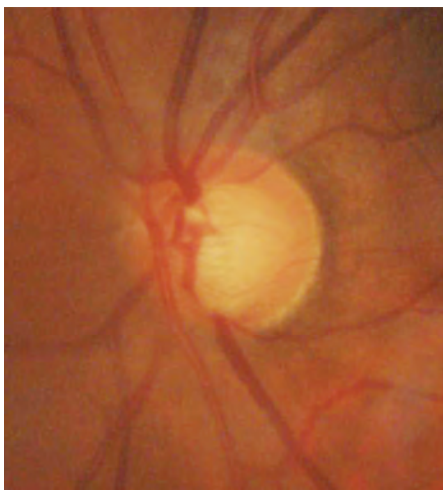

Figure 1. Inferior notch seen on left optic nerve on initial presentation

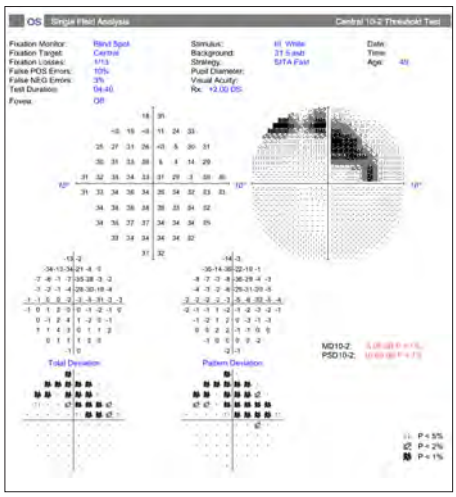

Figure 2. HVFA3 (10-2) showing a superior arcuate defect on initial presentation

in the LE only. These results were successfully repeated one week later.

Mrs X was diagnosed with NTG and a treatment plan was developed to reduce her IOP by 30 per cent. After a diurnal IOP was established at 21 mmHg with a maximum of 22 mmHg, a target pressure of 15 mmHg was set. She was prescribed Xalatan eye drops once a night in the LE, the RE was used as a control. After three weeks, target pressure was achieved and bilateral treatment initiated. She was scheduled to be reviewed in three months and was also referred to the local ophthalmologist for confirmation of the treatment plan and consideration for SLT. The ophthalmologist confirmed the plan and Mrs X was happy to continue Xalatan rather than SLT.

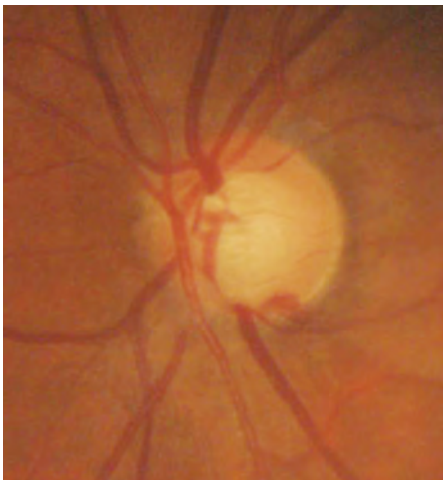

Figure 3. Left optic nerve showing an inferior Drance haemorrhage seen after treatment initiated

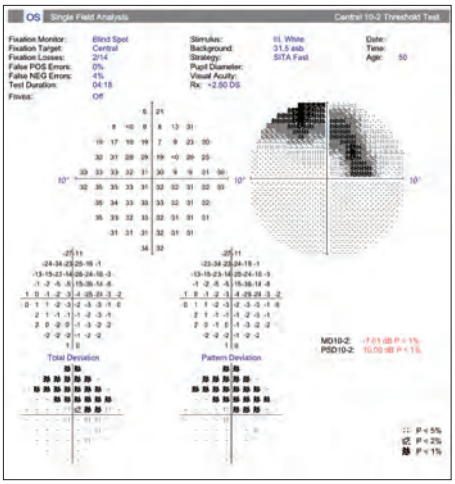

Figure 4. HVFA3 (10-2) showing progression of VF defect in the LE three months after initial treatment

At her three month review, an inferior Drance haemorrhage (Figure 3) was seen on the L ON. VF (10-2) results revealed significant progression of the superior defects of the LE (Figure 4). A new target pressure of < 12 mmHg was set and timolol was added to her treatment with a three-week review scheduled. During this review, Mrs X also stated that she was tired, felt dizzy when sitting up and normally found it hard to sleep as she would wake up in the middle of the night. Our optometrist then referred her to her general practitioner (GP) for investigations into low blood pressure (BP) and OSA.

Mrs X's BP was confirmed to be low at 90/50 mmHg and she was diagnosed with moderate OSA. She was then referred to her pharmacist for a continuous positive airway pressure machine (CPAP) and a dentist to create a mandibular advancement device (MAD) for treatment of OSA.

Her IOP at the three-week review was R 10 mmHg L 11 mmHg, reaching target pressures, and no progression was seen three months later.

Discussion

The interactions between BP and IOP have been shown to influence glaucoma as it can affect DOPP (Diastolic BP – IOP).[3] Low BP can occur spontaneously or secondary to anti-hypertensive medication. Dips in BP have been shown to occur more commonly at night and patients can be classified as normal-dippers (< 10 per cent decline) or extreme dippers (> 20 per cent decline).[2,3] Diurnal variation in IOP is influenced by body position, with IOP likely to rise nocturnally when patients are sleeping in the supine position.[8] Low BP and increased IOP can decrease DOPP resulting in ischemic damage to the ON. A DOPP of < 55 mmHg has shown to increase the risk of glaucoma by 3.2 times and in regards to Mrs X, her DOPP before treatment was 30 mmHg (50 mmHg-20 mmHg).[8]

OSA is a significant risk factor in developing glaucoma. It is characterised by repetitive episodes of complete or partial obstruction to the upper airway during sleep leading to apnoea, hypopnea and hypoxia. This causes a reduction in oxygenation to the ON which can eventually lead to glaucoma.[7,9,10] Clinical symptoms include loud snoring, nocturnal gasping, lack of energy, reduced concentration, memory impairment, dry throat upon waking and morning headaches. Systemic implications of OSA include impeded neurocognitive behaviour and cardiovascular disease. First line therapy is usually with a CPAP machine which has shown positive effects on glaucoma by preventing a drop in arterial oxygen.[7,10] In regards to Mrs X, her symptoms matched that of OSA and her diagnosis was confirmed by a sleep specialist. ▲

Conclusion

Mrs X's risk for glaucoma was increased by her low DOPP and OSA. Although there has been some evidence that managing OSA may slow glaucoma progression, there is currently no research to show that CPAP therapy alone is enough to consider withdrawal of glaucoma medication. This case highlights the importance of collaborative health care and for optometrists to consider the patient's systemic status in NTG. ▲

1. Furlanetto R, Moraes CG, Teng C et al. Risk Factors for Optic Disc Hemorrhage in the Low-Pressure Glaucoma Treatment Study. *Am J Ophthalmol* 2014; 157: 945-952
2. Leeman M, Kestelyn P. Glaucoma and Blood Pressure. *Hypertension* 2019; 73: 944-950
3. Raman P, Suliman N, Zahari M et al. Low nocturnal diastolic ocular perfusion pressure as a risk factor for NTG progression: a 5 years prospective study. *Eye* 2018; 32: 1183-1189
4. Song B, Capriolo J. New directions in the treatment of normal tension glaucoma. *Ind J Ophthalmol* 2014; 62: 529-537
5. Emanuel M, Gedde S. Indications for a systemic work up in glaucoma. *Can J Ophthalmol* 2014; 49: 506-511
6. Hejil A, Bengtsson B, Hyman L et al. Natural history of open-angle glaucoma. *Ophthalmology* 2009: 116: 2271-2276
7. Chaitanya A, Pai V, Mohapatra A et al. Glaucoma and its association with obstructive sleep apnoea. *Oman J Ophthal* 2016; 9: 125
8. Melgarejo J, Lee J, Petitto M et al. Glaucomatous optic neuropathy associated with nocturnal dip in blood pressure. *Am Acad Oph* 2018; 125: 807-814
9. Morsy N, Amani B, Magda A et al. Prevalence and predictors of ocular complications in obstructive sleep apnea patients. *Open Res Med J* 2019: 13; 19-30
10. Yamada E, Himori N, Kunikata H et al. The relationship between increased oxidative stress and visual field defect progression in glaucoma patients with sleep apnoea syndrome. *Act Ophthal* 2018; 96: 479-484

Thygeson's superficial punctate keratopathy

Anna Delmadoros
MOptom BOptom(Hons)
Grad Cert Oc Ther

School of Optometry and Vision Science, UNSW Sydney

This original case report was submitted by fellow Optometry Australia member Anna Delmadoros in response to our nation-wide call for papers.

Thygeson's superficial punctuate keratitis (TSPK), first described by Phillips Thygeson in 1950,[1] is a relatively uncommon, recurrent and chronic non-infectious corneal condition that is typically bilateral and asymmetric.[1-5] It has no sex predilection and no age bias.[2,6] The prevalence of the condition is unknown and the underlying aetiology remains controversial.[1-5]

Diagnostic features of TSPK are multiple, mildly elevated, whitish/grey granular or stellate intraepithelial opacities, occurring predominantly in the central cornea, in the absence of accompanying oedema or conjunctival involvement.[1-4] Symptoms vary in degree and include photophobia, foreign body sensation and lacrimation.[2] Visual acuity may be slightly reduced depending on the density and location of the opacities.[2,3]

A hallmark feature of TSPK is the presence of active and quiescent phases, which vary in duration and frequency.[1-3] Symptomatic exacerbations lasting weeks to months appear to be sporadic and the triggers unknown.[1,2] During periods of remission (months to years), the cornea is essentially void of any signs and the patient is completely asymptomatic.[1,3] TSPK is a self-limiting condition that is reported to resolve spontaneously on average 3–7.5 years after the first presentation,[2-5] however course durations of up to 41 years have been reported.[3,5]

Therapeutic intervention with low-dose topical corticosteroids is the mainstay of treatment during the active phases of the condition for symptomatic relief.[2,6,7] Ocular lubricants, therapeutic soft contact lenses (SCLs) and cyclosporin A (CsA) have also been recommended in the literature.[2,6] There is no cure for TSPK.

While TSPK is an uncommon condition, it should be considered as a differential in cases of chronic non-specific ocular discomfort and is therefore an important condition for eye-care practitioners to recognise and manage. A case is presented where, after 11 months of misdiagnosis, the condition was eventually diagnosed as TSPK and successfully managed.

A 22-year-old Caucasian female first presented to the clinic reporting an 11-month history of recurrent episodes of moderate-to-severe light sensitivity, watery, stinging eyes and gritty sensation that lasted 5–7 days, occurring approximately every 4–5 weeks. The ocular discomfort on the day was subjectively graded as 3 out of 10, with reports of grade 8 discomfort in prior instances. She denied ocular redness, discharge and/or contact lens wear and medical history was unremarkable.

She reported consulting several eye-care practitioners, each with conflicting diagnoses. Various topical preparations were prescribed including ocular lubricants, Chloramphenicol 0.5% and Aciclovir 3% w/w ointment, all with minimal alleviation of symptoms despite strict compliance.

Best corrected acuities were 6/6+2 in each eye. Both corneas exhibited multiple, scattered whitish-grey round/oval and stellate lesions (twelve in the right and seven in the left), confined within the superficial epithelium (Figure 1) that stained with sodium fluorescein (Figure 2). Some lesions were essentially flat and others minimally elevated. The eyes were otherwise white and quiet, and corneal sensitivity intact.

A provisional diagnosis of TSPK was made, with herpes simplex keratitis (HSK) still a differential, although unlikely.

Management approach was conservative given the mild symptoms and corticosteroids possibly contraindicated; existing drops were discontinued, and non-preserved artificial tears were recommended every 1–2 hours and ointment before bed; advice was to return immediately if symptoms worsened. Review at 24 hours revealed an unchanged appearance of the corneal lesions.

Significant improvement in symptoms was reported at the follow-up visit three days later, although the patient reported some impracticalities in maintaining 1–2 hourly dosing and an aversion to the ointment. Acuities remained unchanged and besides a reduction in the number of lesions, eyes continued to be white and quiet. The marked corneal improvement and absence of any dendritic lesions solidified the diagnosis and led to management with preservative-free ocular lubricants three times a day and to return for review at the next flare up.

Three weeks later, the patient presented with symptoms identical to previous occurrences, this time with acuity mildly reduced to 6/6-2 in the right eye and 6/6-1 in the left. There were numerous grouped corneal intraepithelial lesions, indistinguishable in appearance to

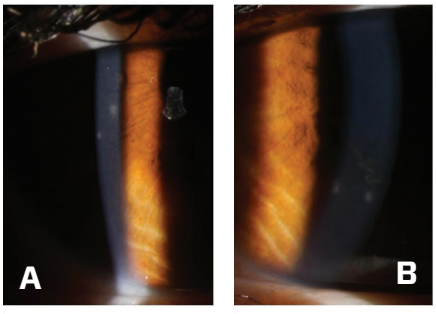

Figure 1. TSPK corneal lesions. A: right eye. B: left eye.

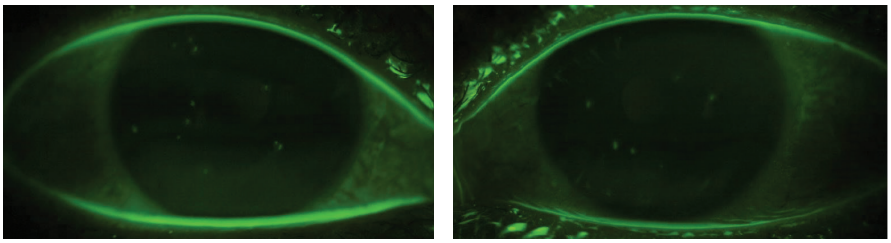

Figure 2. Fluorescein staining of corneal lesions. Figure A: fight eye. Figure B: left eye

previous, with greater than twenty lesions in each eye, worse in the right.

Active TSPK was diagnosed and patient commenced on fluorometholone (FML) 0.1% four times a day with continued use of 1–2 hourly non-preserved lubricating eye drops in between steroid dosing, at least 15 minutes either side of steroid use. There was a marked improvement in symptoms at the 24-hour follow-up visit.

Five days later, there was complete resolution of symptoms with all except one corneal lesion remaining in the right eye. The corneas did not stain with fluorescein; intraocular pressures were 12 mmHg in each eye. FML 0.1% use was tapered and a two-week review advised. An essentially identical treatment protocol was implemented for each of the subsequent flare ups.

The patient was unfortunately lost to follow-up due to a move overseas. Subsequent correspondence revealed she was self-managing flare ups with FML and ocular lubricants. Two years from onset, exacerbations were reportedly milder, less frequent (every 3–4 months) and shorter in duration (1–2 days). Despite the improvement, the importance of regular ophthalmic reviews was stressed with regards to the possible side effects of long-term topical corticosteroid use; and the opportunity to discuss and consider alternate management therapies with an enhanced safety profile.

Discussion

The clinical picture of TSPK is characterised by recurrent, bilateral epithelial keratitis, with an essentially white and quiet eye in an otherwise healthy patient. The keratitis is variable in its presentation and follows a relapsing course with remissions and exacerbations over several years, until spontaneous resolution. There is generally an excellent long-term visual outcome, although there have been reports of some patients developing permanent faint sub-epithelial opacities.[6,8]

TSPK is commonly confused with other conditions, and patients frequently report conflicting diagnoses from different practitioners and minimal success in treating their symptoms. Differential diagnoses and key differential features are outlined in Table 1.

Although there is no cure, therapeutic intervention is focused on symptomatic relief in the active phase of TSPK, as the presenting symptoms can be debilitating for some patients.

Treatment varies depending on the case severity. Tolerable symptoms in mild cases may be relieved by regular use of non-preserved ocular lubricants with an ointment at bedtime.[6] Otherwise the mainstay of treatment is a mild topical corticosteroid (FML 0.1%, Loteprednol or similar), although a more potent steroid such as fluorometholone acetate may be required in more severe cases, with minimal strength and dose to control symptoms.[2,3,5,6] A gradual taper of the corticosteroid is essential to prevent recurrence, however, it can prove challenging given the variable presentation of TSPK, with some patients requiring a tapering schedule over the course of weeks to months to avoid a recurrence.[6] Other treatment options such as therapeutic SCLs or topical CsA where available can be considered, the latter indicated in recalcitrant cases or where topical steroid treatment is contraindicated.[2,10,11]

The risks of extended corticosteroid use in an otherwise benign condition must be considered, bearing in mind that TSPK can take many years to resolve.[2,3,5] Although low potency steroids have less likelihood of side effects,[9] there are still concerns associated with long term topical corticosteroid use, including steroid-induced glaucoma, cataracts and

Continued page 12

Differential	Unilateral/bi-lateral	Lesion appearance, associated features	Vital staining	Additional features
Early herpes simplex keratitis	Usually unilateral	Fine/course SPK. Small bullous epithelial vesicles.	+ Fluorescein Sodium + Rose Bengal	SPK coalesce to dendritic lesions. Heaped margins and terminal bulbs. Reduced corneal sensitivity
TSPK	Typically bilateral	Multiple, mildly elevated, whitish central intraepithelial corneal opacities	+ Fluorescein Sodium	Recurrent keratitis in a white and quiet eye
Adenoviral keratopathy	Bilateral	Nummular sub-epithelial corneal opacities	-	Swollen lymph nodes. Follicles
Keratoconjunctivitis sicca (KCS)	Usually bilateral	Small, finer SPK	+Fluorescein Sodium + Lissamine Green	
Sterile sub-epithelial infiltrates	Usually unilateral	Usually peripheral corneal distribution	-	
Microcystic epithelial oedema	Usually unilateral	Small raised bullae. Corneal oedema	-	Blurred vision, halos
Acanthamoeba keratitis	Mostly bilateral	Raised subepithelial opacities	-	History of CL wear. Blurred vision. Symptoms disproportionate to signs.
Toxic epitheliopathy	Usually bilateral	Widespread superficial punctate epitheliopathy (SPE)	+ Fluorescein Sodium	Stinging, burning sensation. Conjunctival injection. History multiple/chronic use of topical drugs/CL and/or solution use
Neurotrophic keratopathy (mild/early)	Usually unilateral	SPE	+ Fluorescein Sodium	Red eye. Blurred vision. No irritation/pain.

Table 1. Differential diagnoses and key differential features

TSPK

From page 11

increased susceptibility to corneal infections; there's also speculation that corticosteroids may potentially prolong the chronic nature of the disease.[2,3,5,6,8]

Those receiving topical corticosteroid treatment should have their intraocular pressures monitored closely, especially if they are at risk of a steroid response.

Finally, the chronic and recurrent course of TSPK may lead patients to self-medicate, putting themselves at higher risk for steroid related ocular complications. It is therefore imperative that all patients be acutely aware of the potential side effects of topical steroid use and the importance of regular reviews. In addition, the practitioner should be astute in limiting the number of repeats when prescribing topical steroids. With compliance, patients can be reassured that TSPK usually resolves without any long-term effects on vision.

1. Thygeson P. Superficial Punctate Keratitis. *JAMA* 1950; 144: 1544-1549
2. Nagra PK, Rapuano CJ, Cohen EJ et al. Thygeson's superficial punctate keratitis. Ten years' experience. *Ophthalmol* 2004; 111: 34-7
3. Tabbara KF, Oster B, Dawson C et al. Thygeson's superficial punctate keratitis. *Ophthalmol* 1981; 88: 75-7
4. Darrel RW. Thygeson's SPK: a natural history and association with HLA-DK3. *Trans Am Ophthalmol Soc* 1981; 79: 486-516
5. Tanzer DJ, Smith RE. Superficial punctate keratitis of Thygeson's: the longest course on record? *Cornea* 1999; 18: 729-730
6. Duszak RS. Diagnosis and management of Thygeson's superficial punctate keratitis. *Journal Am Optom Assoc* 2007; 78: 333-338
7. Bruce A, Loughnan M. Anterior eye Disease and Therapeutics A-Z. Boston: Butterworth Heinemann, 2003
8. Chen LL, Young AL, Wong AK. In vivo confocal microscopy of Thygeson's superficial punctate keratitis. *Clin Exp Ophthalmol* 2004; 32: 325-340
9. Jaanus SD, Cheetham JK, Lesher GA. Anti-inflammatory drugs. In: Bartlett JD, Jaanus SD, editors. Clinical Ocular Pharmacology, 4th ed. Boston: Butterworth, 2001. p 273-276
10. Forstot SL, Binder PS. Treatment of Thygeson's superficial punctate keratitis with soft contact lenses. *Am J Ophthalmol* 1979; 88: 186-189
11. Reinhard T, Sundmacher R. Topical cyclosporine A in Thygeson's superficial punctate keratitis. *Graefes Arch Clin Exp Ophthalmol* 1999; 237: 109-112

Contact lenses: beyond

Dr Lyndon Jones
PhD DSc FCOptom FAAO

Deborah Jones
BSc FCOptom FAAO

Centre for Ocular Research and Education (CORE), School of Optometry & Vision Science, University of Waterloo, Ontario, Canada

Rebecca Jones
BSc

Michael G. DeGroote School of Medicine, McMaster University, Cairns Family Health and Bioscience Research Complex, Ontario, Canada

The use of contact lenses continues to grow, with an estimated 140 million wearers globally. While the majority of wearers use contact lenses for the correction of refractive error, there is growing interest in their use for 'non-standard' concepts. As we approach the magical year (for optics) of 2020, we ask: if we glimpse into the future, what will the contact lenses of (say) 2035 look like? What options will they provide to our patients that will differ from the lenses of today?[1]

Myopia control

Myopia is a significant public health problem. In 2010 it was estimated that 28 per cent of the world's population was myopic, but it is predicted that by 2050, 50 per cent of the global population could be myopic.[2] As discussed in the September issue of *Pharma*, the most rapid increase has been in East Asian countries, where it has already reached epidemic proportions, affecting over 90 per cent of adults in some regions such as Korea, Taiwan and Singapore.[3] Myopia is not merely an inconvenience resulting in the patient requiring an optical correction. The myopic eye, particularly those with high myopia of > 6.00 D, has an increased risk of developing ocular pathology that may lead to vision loss, in particular retinal detachment and myopic macular degeneration. Thus, methods to slow or prevent myopia progression (and the ocular axial elongation that accompanies it) are extremely important if myopia-induced pathologies are to be avoided. Recent estimates suggest that slowing myopia progression by one dioptre should reduce the likelihood of a patient developing myopic maculopathy by 40 per cent.[4]

Several treatment paradigms for slowing myopia progression have been evaluated in intervention studies, largely encompassing the use of either pharmacological means (most commonly through the use of the topical anti-muscarinic drug atropine) or various optical interventions.[5-13] These optical interventions include progressive addition spectacle lenses, bifocal spectacles, orthokeratology rigid contact lenses and multifocal soft contact lenses.[8,14-19]

To date, many of the contact lens methods used remain 'off-label,' meaning that while studies would suggest that such technologies do indeed appear to show a slowing of myopia progression, regulatory approvals for these products do not yet exist to support the product being used in such a manner.[20] One such example is orthokeratology, which is approved for use in the reduction of refractive error, but not approved for slowing the progression of myopia, despite many studies supporting this to be the case.

However, as the interest in using contact lenses for slowing myopia progression increases, more products will gain regulatory approval for this indication, resulting in this being a significant growth area for contact lenses over the next decade. Two commercially available daily disposable soft lens products that have gained regulatory approval for slowing the progression of myopia in various countries following successful clinical trials are MiSight (CooperVision)[21] and NaturalVue Multifocal (Visioneering Technologies).[22]

Drug delivery

One of the biggest opportunities for the development of 'specialised' contact lenses relates to their use as drug delivery devices. A lens that would

to download, and are free to order in hard copies within Victoria. We also have a range of translated video clips developed to communicate the importance of regular eye tests for people who do not speak English which we encourage all optometrists and health professionals to access.

When it comes to individual practitioners, the Victorian Refugee Health Network report recommends health professionals can work better with clients from CALD backgrounds by improving cultural competence, being welcoming and friendly, listening and being respectful, being patient and sensitive to people's difficult past experiences, taking time to develop the client's trust, and maintaining confidentiality.[3] Optometrists and all eye health practitioners can work to ensure our services are more accessible and inclusive of all Australians.

1. Australian Bureau of Statistics 2024.0 - Census of Population and Housing: Australia revealed, 2016. [cited 2020 Mar 20]. Canberra, Australia. Available from: https://www.abs.gov.au/ausstats/abs@.nsf/mf/2024.0.
2. Victorian State Government. Department of Health and Human Services. Delivering for Diversity, Cultural diversity plan 2016–2019. [cited 2020 Mar 19] Melbourne, Australia. Available from: https://www2.health.vic.gov.au/about/populations/cald-health.
3. Tyrrell L, Duell-Piening P, Morris M et al. Talking about health and experiences of using health services with people from refugee backgrounds, Final Report. Melbourne, Australia: Victorian Refugee Health Network; 2016 Sept.
4. Marmot M. The health gap: the challenge of an unequal world. Bloomsbury, London; 2015.
5. World Health Organization [Internet]. Social determinants of health, 2020 [cited 2020 Mar 19] Available from: https://www.who.int/social_determinants/sdh_definition/en/.
6. Australian Institute of Health and Welfare. Australia's health 2016 [Internet]. Canberra: Australian Institute of Health and Welfare, 2016 [cited 2020 Mar 19]. Available from: https://www.aihw.gov.au/reports/australias-health/australias-health-2016.
7. Centre for Eye Research Australia & Vision 2020 Australia. National Eye Health Survey, 2016. Melbourne, Australia: Vision 2020.
8. Nixon PJ, Li JC, Baker RJ et al. Barriers to vision correction for children in a disadvantaged school community. *Optometry* 2015; 16: 85-94.
9. World Health Organization [Internet]. Health promotion, 2020 [cited 2020 Mar 19]. Available from: https://www.who.int/health-topics/health-promotion#tab=tab_1 .

Caring for culturally and linguistically diverse (CALD) patients

Dr Mandy Truong
PhD MPH BOptom

Research Fellow, School of Nursing & Midwifery, Faculty of Medicine, Nursing & Health Sciences

Monash University

In Australia, 10.6 million people were either born overseas or have one or both parents who were born overseas.[1] Twenty-one per cent of Australians speak a language other than English at home and increasing proportions of migrants arriving in Australia are coming from China and India. Therefore, many optometrists are providing eye care services to culturally and linguistically diverse CALD) Australians. Many optometrists themselves are from CALD backgrounds (as recent migrants and first/second/third-generation Australians).

Culture is important in health care because it provides a framework through which individuals and communities interpret the world, negotiate their health behaviours and make decisions about their health care.

Culture generally refers to knowledge, beliefs, traditions, values, or the way of life of a particular people, society or nation.[2]

Within optometry, other health professions and our broader health system in Australia, health-related beliefs, practices and policies are centred on the biomedical model and traditional Anglo-Australian concepts of health and health care.

Therefore, when people from non-Western societies migrate to places such as Australia, they are confronted with a health system which may differ and conflict with their own health beliefs, practices and experiences.

We know that people from CALD backgrounds can find it challenging accessing health services for a variety of reasons, including: communication difficulties related to English-speaking proficiency, health literacy, knowledge and beliefs about eye health, lack of familiarity with the health system (Medicare billing, referrals for medical specialists and so on) and availability of culturally-appropriate services.

A pilot study found that optometrists can experience challenges when providing care to patients from different cultural backgrounds.[3] Optometrists interviewed in this study described language difficulties with patients that resulted in the need to adjust their management advice or spend more time explaining things to patients.

Some optometrists expressed frustration at times when encountering

Continued page 8

CALD care

From page 7

patients who prioritised their spectacle needs and ignored or downplayed advice given about their eye conditions (for example: cataracts or retinal detachment) which were seen as more important by the practitioner.[3]

But when the priorities, beliefs and attitudes of patients do not align with the optometrist, the treatment and management will need to be adapted and negotiated.

CASE REPORT

David, a 10 year-old male Chinese student who recently migrated to Australia with his parents attended an optometry practice reporting blurry distance vision. Both of his parents wear glasses and were quite concerned about his vision. The examination revealed early myopia: R -1.00 L -0.75. No other eye conditions were detected.

The consulting optometrist advised single vision distance glasses, however, the patient and his parents were very reluctant to buy glasses because the parents felt that from their experience, wearing glasses from a young age caused their vision to progressively worsen. They expressed this was a common perception among their community back in China.

There are several possible strategies that an optometrist can employ in a situation like this.

First, show understanding and empathy towards the patient and his family. It is a common misconception across many different cultures and communities that glasses weaken eye sight.

Next, provide an information sheet (in the requisite language or plain English) that explains what myopia is and that not correcting or under-correcting myopia will cause myopia to worsen. The optometrist can then explain that there are other therapies available that can potentially slow down myopia progression and that you are willing to help them if the need arises in future

Finally, if possible, offer to get an optometrist who speaks the patient's language to call the parents to explain the diagnosis and management in their preferred language and address any concerns/questions.

Discussion

The practice of competent optometry in Australia and New Zealand requires that we improve our ability to provide more culturally-responsive and culturally-aware care across our diverse community, which includes refugees, recent migrants, international students and First Nations Australians.

Recommendations[4-6]

Identify and address any language barriers and provide information in languages other than English.

Consider being more flexible with appointment systems. Avoid penalising patients for missing appointments or being late without trying to understand the circumstances which resulted in the late or missed appointments.

Be pro-active; respectfully engage with cultural/ethnic groups in your community. For example, attend cultural events and offer to do a talk about an eye-health issue at a community gathering.

Find out whether there are alternative perspectives and beliefs about eye health issues.

Employ and train optometrists and dispensing/support staff from diverse backgrounds.

Encourage all your staff to engage in cultural competency training so everyone in the practice is culturally-aware and responsive.

Conduct a review of how welcoming the environment is to people from diverse backgrounds, including Aboriginal and Torres Strait Islander peoples. Is diversity represented in pictures, posters and promotional material, for example?

Engage with your diverse patients in an open-minded and respectful manner – listen, ask and learn.

Avoid the following:

Using a 'checklist approach' (that is learning that Chinese people 'think and do X'; 'Greek people think and do X') that results in negatively stereotyping and generalising patients.

Providing good quality eye care to patients does not necessarily mean 'treating everyone the same' because each individual has a unique set of characteristics and experiences. Therefore, those in our community who are more disadvantaged may need additional support and consideration. At times we will need to adapt our practices and treatments in the best interests of each patient.

Finally, it's possible for optometrists to unintentionally provide care that is not culturally responsive. We should recognise and understand that our attitudes and practices, both conscious and unconscious, impact on how we provide care to our patients. Therefore, self-reflective practice is critical to ensuring we are providing high quality eye care to all patients in our community.

1. Australian Bureau of Statistics 2071.0 - Census of Population and Housing: Reflecting Australia - Stories from the Census, 2016. [cited 2020 April 7]. Canberra, Australia. Available from: https://www.abs.gov.au/AUSSTATS/abs@.nsf/Lookup/2071.0Main+Features100012016?OpenDocument
2. Smedley A, Smedley BD. Race as biology is fiction, racism as a social problem is real: Anthropological and historical perspectives on the social construction of race. *Am Psychol* 2005; 60: 16-26.
3. Truong M, Fuscaldo G. Optometrists' perspectives on cross-cultural encounters in clinical practice. *Clin Exp Optom* 2012; 95: 37-42.
4. Truong, M and Selig S. Advancing cultural competence in optometry. *Clin Exp Optom* 2017; 100: 385-387.
5. Centre for Culture, Ethnicity & Health [Internet]. Melbourne: Resources in Cultural Competence [cited 2020 April 7]. Available from: https://www.ceh.org.au/resource-hub/category/cultural-competence/
6. Migrant & Refugee Health Partnership [Internet]. Canberra: Competency Standards Framework: Culturally responsive clinical practice: Working with people from migrant and refugee backgrounds [cited 2020 April 7]. Available from: https://culturaldiversityhealth.org.au/competency-standards-framework

Christmas Eye

From page 20

Localised corneal thinning

I have seen three cases where there has been a localised area of stromal thinning. The corneal response appears as expected until the four-day mark where marked corneal inflammation and oedema seems to remain. As it subsides, the stroma appears to thin and the corneal topography changes to show an area of depression. There is associated vision disruption due to the altered topography.

Detailed anterior OCT scans confirm the stromal thickness loss as the epithelium regrows with normal thickness.

Visual recovery from this is very slow and can be incomplete. OCT scans and topography over the past three years show both a localised thickening of the overlying epithelium and an improvement in the corneal regularity.

Dense corneal scarring

Unfortunately, I have one patient who experienced the development of a disc-shape intrastromal corneal scar. His incident history was identical until the day-five mark, when he developed a disc shaped sub-epithelial lesion with marked corneal thickening. His corneal epithelium was nearly fully healed (Figure 6).

Over the next six months the cornea involved proceeded to flatten and thin. The density of the corneal scarring also reduced. The original size of the scar is indicated by the white outline in Figure 8. At its peak, the corneal thickness through the lesion was 721microns. Four weeks later, the thickness had reduced to 489 microns (Figures 7 and 8).

A year later, the lesion is still present but less dense. The topography and corneal thickness changes have stabilised. Fortunately, the lesion was off axis and is now only causing slight blur and mild flare at night while driving.

Discussion

Why there was a cluster of these adverse events in the summer of 2017–18 is unknown. Last summer (2018–19) I experienced no complications.

It may be the causative agent was slightly different, creating a greater inflammatory response or that these individuals happened to be more susceptible to corneal damage from inflammation.

The events have led me to remove the topical NSAIDS from the management plan. There have been literature reports of corneal damage related to topical NSAID use.[4] I am unsure why this would become an issue in one particular year and it may be completely unrelated.

These events appear to be individual reactions to the extreme corneal inflammation that occurs with Christmas Eye. They are a cautionary reminder that unusual events can occur when treating eye conditions.

Imaging

The development of imaging systems such as topography, anterior OCT and digital photography allows the practitioner to capture some of very strange images that can be associated with Christmas Eye. This technology allows us to monitor the effects on the eye as never before (Figures 9 and 10).

Conclusion

Christmas Eye can be a confronting condition for those who are unaware of its signs and symptoms. Any condition that causes extreme pain is stressful for both the practitioner and particularly the patient. Care, reassurance and pain control are the keys in managing this unusual condition.

1. Howsam G. The Albury – Wodonga syndrome. A tale of two cities. Aust NZ J Ophthalmol 1995; 23: 135–137
2. Farrow R. *Insects of South-Eastern Australia: An Ecological and Behavioural Guide*. Clayton South: CSIRO Publishing, 2016.
3. Mullen G, Durden L, editors. *Medical and Veterinary Entomology*, 2nd ed London: Academic Press, 2019.
4. Lin JC, Rapuano CJ, Laibson PR et al. Corneal melting associated with use of topical nonsteroidal anti-inflammatory drugs after ocular surgery. *Arch Ophth* 2000; 118: 1129–1132

TAU: Tattoo-

Cutaneous

Debra Gleeson
Assoc Dip Orth

Senior Orthoptist/Orthoptic Glaucoma Lead Royal Victorian Eye and Ear Hospital, Melbourne

As a cosmetic and decorative body art, tattooing has dramatically increased particularly among young adults. A survey of 1,013 Australians by market researcher McCrindle in April 2018[1] showed that the number of people getting tattooed had hit a record high with one in five people having one or more tattoos.

The majority have more than one tattoo (61 per cent) and around 14 per cent have six or more. Fifty-one per cent had obtained their first tattoo between the ages of 18 and 25, and thirty-six per cent at 26 or older. Australian women with tattoos (20 per cent) outnumber men (19 per cent).

Given these figures, we need to be aware of a possible increase in presentations of tattoo-associated uveitis (TAU).

CASE REPORT

A 25-year-old male presented to the emergency department in 2017 with decreased visual acuity, intermittent redness and a feeling of 'pressure' in both eyes (OU). These symptoms had been intermittent for approximately one year.

The patient's medical history was unremarkable; he denied any past ocular history but was diagnosed with allergic conjunctivitis in 2015. Old keratic precipitates (KP) OU were also noted at this time, indicating previous inflammation. His vision was 6/36 OU with no pinhole improvement. Bilateral anterior uveitis and posterior synechiae were noted, intraocular pressures (IOP) were RE 9 mmHg and LE 10 mmHg.

-associated uveitis

reactions and ocular problems

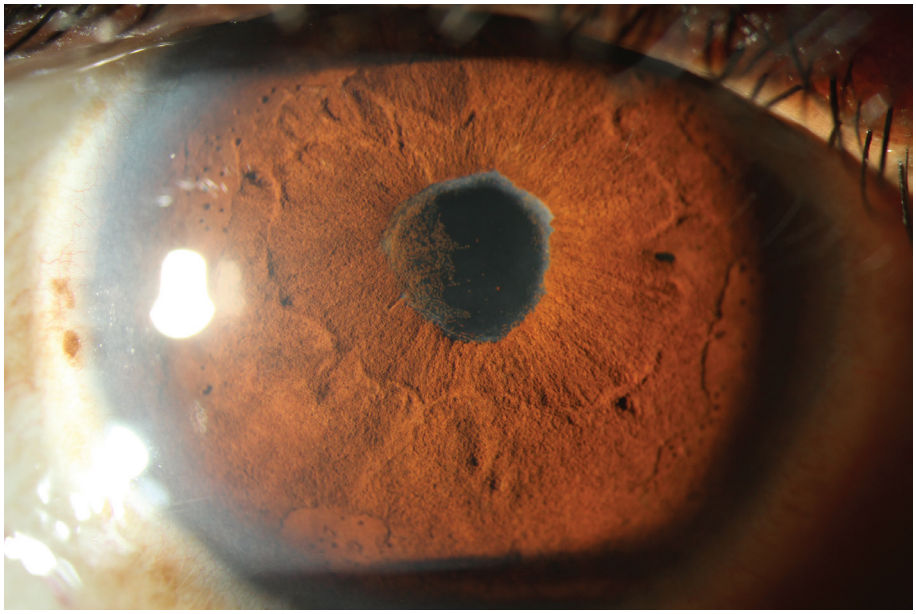

Figure 1. Posterior synechiae (adhesion of the iris to the capsule of the lens) due to inflammation.

He was diagnosed with bilateral acute anterior uveitis (AAU) and cystoid macular oedema (CMO), and commenced on topical medications: prednefrin forte (PF) hourly, and atropine twice daily in both eyes for pain management and to reduce further posterior synechiae formation.

Blood tests were taken to rule out sarcoidosis, syphilis, HLA-B27 positivity and various infectious and inflammatory aetiologies (repeated throughout his follow-up).

On review 10 days later, it has been noted that compliance had been poor with the topical medications. His vision had deteriorated further (6/60 OU), the uveitis persisted (+2 cells) and his optic nerves were swollen and hyperaemic.

He was commenced on systemic prednisolone (PNL) 50 mg daily. With improved compliance of all medications, his vision improved to 6/9 OU due to decreased CMO and inflammation. Intraocular pressures (IOP) were RE 19mmHg and LE 16 mm Hg. He was advised to reduce his PNL to 37.5 mg and atropine was replaced with tropicamide. Vision and IOP remained stable and further weaning of PNL was suggested.

He missed his next review, and on presentation seven weeks later there was a recurrence of bilateral uveitis (2+ cells, SUN grading), disc swelling and left CMO. Compliance was stressed but it was thought that a left orbital floor injection of triamcinolone may be needed in the future.

Having missed an appointment, he was reviewed five weeks later. He had lost his prescription and had only instilled topical medications on a few occasions. His left CMO had reduced, and an indurated red, raised left tricep tattoo was noted.

In retrospect, he felt that his ocular problems commenced around the time that he got the tattoo. In light of previous negative investigations and the presence of inflamed tattoos, a diagnosis of tattoo-associated uveitis (TAU) with CMO OU was made.

He was lost to follow-up for almost five months after which bilateral recurrent anterior uveitis (3+ cells) and CMO (LE > RE) had recurred. Bilateral posterior synechiae and a left iris nodule were noted. The retina could not be visualised. Due to severe eye-threatening uveitis, he was given a left orbital floor injection and was advised to reduce his PNL by half and continue with PF hourly OU.

At his next visit he complained that his tattoos were feeling 'lumpy' again. Control of his ocular inflammation was difficult, exacerbated by his poor attendance and treatment compliance, so he was commenced on a weekly dose of both immunosuppressant drug methotrexate (MTX) and folic acid. He was to continue with 5 mg PNL and PF four times each day. Monthly blood tests were initiated to monitor for dosage and side effects of the MTX.

Improvement was noted in his vision and ocular inflammation, however, a right IOP of 32 mm Hg was noted (left 12 mm Hg). His topical and systemic medications were reduced by half and he was sent to the Glaucoma Unit due to development of right ocular hypertension (OHT) caused by iris bombe (synechial closure R > L). He underwent a right Yag laser peripheral iridotomy and his IOP was noted to be 8 mmHg after this. His cup-to-disc ratios were 0.2 OU and his visual field tests were essentially normal.

When last seen, his vision was 6/9 OU; both eyes were quiescent and there was minimal CMO. IOPs were RE 8 mmHg and LE 8 mmHg. He had been weaned off the PF but was to continue with the immunosuppressant drug MTX to curtail the cutaneous reaction and reduce the risk of recurrent uveitis.

Discussion

There have been an increasing number of cases in literature[2] of TAU since Rorsman et al.[3] described three cases with light blue tattoo granuloma and anterior uveitis with no features of systemic disease in 1969. In 2014 Ostheimer et al.[4] submitted the largest study which followed seven patients with various sequelae of uveitis with simultaneous tattoo induration over 20 months.

TAU presents with bilateral recurrent/chronic uveitis, though in one case the second eye became involved one month later.[5] Cases cited have

Continued page 24

Tattoo uveitis

From page 23

ranged from having anterior uveitis (predominately non-granulomatous) to chronic pan uveitis and hypopyon.

Tattoo swelling has been said to precede uveitis for a week on recurrent episodes.[6]

The time frame of onset can occur from at least six months after tattoo placement and up to 13 years which possibly presents a specific granulomatous-delayed allergic response to ink containing metal compounds.[7] Obtaining a number of tattoos over a short period of time possibly increases the toxic load.[4] Most had extensive tattoos and were predominately male.

The majority of cases have been in the USA and many of the inks used were industrial grade colours suitable for printers and automobile ink. The induration appeared in more extensively tattooed areas that contained or consisted entirely of black ink. The black ink possibly contained toxic, mutagenic or carcinogenic compounds.[4] Two articles cite skin reaction to light blue[3] and red[8] ink.

In Australia, state and territory authorities are responsible for regulating the safety of tattoo inks including product labelling and restrictions on their use in tattooing. Chemicals used in tattoo and permanent makeup (PMU) inks are classified as industrial chemicals in Australia.[9] These regulations may not be adhered to, particularly if used outside of a registered tattoo parlour or in a country without a regulatory body.

The cutaneous reactions of erythema, pruritis, indurated papules or nodules can occur on the border or within the tattoo area (Figure 2). Similar cutaneous reactions may arise in patients with sarcoidosis (33 per cent), as can granulomatous uveitis (80 per cent).[9] There have been a number of cases where TAU has been the presenting feature of sarcoidosis. It has been postulated that sarcoidosis may be diagnosed in these cases in the long term[10] or that this entity is perhaps a subset of sarcoidosis.[7] It was noted that when there was complete excision of the affected tattoo, ocular symptoms completely resolved without medication,[4] an option not possible where large areas are affected.

Figure 2. Induration (cutaneous reaction) of a black chest tattoo.

The inflammatory response can be difficult to control and many patients have suffered potentially vision-threatening ocular complications such as seclusio pupillae, iris bombe, OHT (often refractory), uveitic glaucoma, pupillary membranes, severe CMO, elevated and hyperaemic optic nerves with papillomacular exudates and retinal detachment.[4]

As shown, in addition to topical, periocular and systemic steroids, systemic immunosuppressants may be required. Reactivation is common on tapering steroids. Regular ophthalmic review and blood tests to watch for serious side effects are required. Raised IOP due to uveitic mechanisms and/or topical steroid use may require topical glaucoma drops. Traditional glaucoma surgery can have poor outcomes in inflamed eyes. Drainage tubes have been required due to refractory OHT or secondary glaucoma.

The following questions need to be asked of anybody presenting with uveitis: do you have tattoos and if so, are they inflamed, red or lumpy? Those presenting with old KPs need to be asked in regard to tattoos and previous ocular/cutaneous reactions.

Tattoo parlours should make their customers aware of cutaneous reactions which may be a precursor to ocular problems and that an urgent eye check is required with symptoms such as ocular pain, photophobia, redness and blurred vision.

Immediate treatment may reduce the severity and sequelae of uveitis which may require laser, surgery and medications with possible serious side effects.

The author would like to acknowledge Dr Catrin Bertalot (Medical Retinal Fellow, RVEEH) for her advice and Matthew Ayres (Medical Photographer, RVEEH) for the photographic images.

1. McCrindle [Internet]. Tattoos in Australia: Perceptions, trends and regrets. c.2016 [Cited 2019 Sep 16] Available from: http://mccrindle.com.au/insights/blog/tattoos
2. Mansour AM. Tattoo-associated uveitis. *Am J Ophthalmol* 2015; 159: 408-409
3. Rorsman H, Brehmer-Andersson E, Dahlquist I et al. Tattoo granuloma uveitis. *Lancet* 1969; 294: 27-28
4. Ostheimer TA, Burkholder BM, Leung TG et al. Tattoo-associated uveitis. *Am J Ophthalmol* 2014; 158: 637-643
5. McElvanney AM, Sherrif SM. Uveitis and skin tattoos. *Eye (Lond)* 1994; 8: 602-603
6. Mansour AM, Chan CC. Recurrent uveitis preceded by swelling of skin tattoo. *Am J Ophthalmol* 1991; 111: 515-516
7. Jacobs J, Van Calster J.Skin tattoos and the development of uveitis. Acta Ophthalmologica 2013: 650-652
8. Barbarasi Z, Kiss E, Balaton G et al. Cutaneous granuloma and uveitis caused by a tattoo. Wien Klin Wochenschr 2008; 120: 18
9. Australian Government Department of Health [Internet]. Characterisation of tattoo inks used in Australia. c.2018 [Cited 2018 May 15]. Available from: https://www.nicnas.gov.au/chemical-information/Topics-of-interest2/subjects/tattoo-inks-used-in-Australia/Characterisation-of-tattoo-inks-used-in-Australia
10. Pandya VB, Hooper CY, Merani R et al. Tattoo-associated uveitis with choroidal granuloma: a rare presentation of systemic sarcoidosis. *Retin Cases Brief Rep* 2017; 11: 272-276

Paediatric optic nerve anomalies

Associate Professor Ann Webber
PhD MS BAppSc(Optom)(Hons) Grad Cert Oc Ther FAAO (Diplomate BVPPO)

School of Optometry and Vision Science, Queensland University of Technology

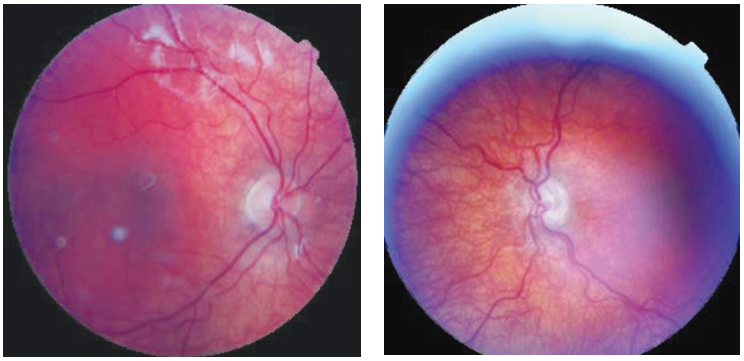

Figure 1. Retinal photographs of a two years, 11-month-old male. A: RE normal ONH and B: LE hypoplastic ONH.

The optic disc or optic nerve head is the portion of the visual pathway where the axons of the retinal ganglion cells collect together to form the optic nerve to exit the eyeball. Optometrists routinely employ direct and indirect ophthalmoscopy, slitlamp biomicroscopy, retinal photography and optical coherence tomography (OCT) to assess and evaluate the optic disc and neuro-retinal rim for the purpose of screening for health, disease and ability to function.[1]

In paediatric optometry practice, careful inspection of the optic nerve head is essential to identify congenital anomalies that might explain or predict findings of vision loss in children and, importantly, to identify children who may be at risk of accompanying neurological pathology.

Gathering sound clinical information in young children is particularly important as these patients have a vulnerable developing visual system, with any structural ocular abnormality that reduces visual acuity in infancy likely to lead to amblyopia. Further, an abnormal optic disc appearance may herald sight or life-threatening pathology that needs referral for further neuro-ophthalmic investigation.

A paediatric eye examination can be challenging, especially if the child is incapable of providing reliable responses for a measure of visual acuity. The clinician may need to rely on objective assessments to examine and record vision and ocular structures, with retinal photography and OCT highly beneficial to the ophthalmic exam, and particularly valuable to record for change in nerve head characteristics over time.

The two cases presented here are examples of when ocular photography and OCT scanning in the primary care optometry practice, both at baseline and at time of onset of new signs or symptoms, proved highly useful to diagnosis and monitoring of structural optic nerve head anomalies.

CASE REPORT 1

Optic nerve head hypoplasia

A Caucasian male aged two years and 11 months was referred for paediatric optometry assessment and amblyopia management advice. He was reported to have a recent onset left esotropia and reduced acuity in the left eye. He was born at full gestational term, of normal birth weight and had met developmental milestones at expected ages. General health was unremarkable with no prescribed medications or allergies.

Entering unaided acuities were RE 6/6 and LE 6/120 (tested with isolated LEA optotypes employing a matching card). Cover test showed 30 prism dioptre LE constant esotropia at both distance and near fixation. His parent reported that the strabismus was first noted approximately six months prior as an intermittent turn, but had become more frequent over the intervening period. Pupil reactions and extra ocular muscle motility were normal. Cycloplegic refraction determined refractive error of R +1.00 DS L +1.00/-2.00 x 180. Anterior segment examination was unremarkable. Posterior segment examination by binocular indirect ophthalmoscopy through dilated pupils indicated an anomalous disc appearance, with careful inspection limited by diminishing child cooperation.

Fundus photography was attempted. The child was asked to kneel on the examination chair so that the positioning of chin on retinal camera support was not limited by his small stature. The goal of obtaining a photo of the eye was explained to the child, with instructions that positioning onto the instrument 'was just like riding a motor-bike,' he was to hold onto the side of the chin support like holding bike handles and to rest against the forehead support as if that were his helmet. He was asked to 'look at the green GO light' as the photo was taken. While the resultant photos are unlikely to win recognition for their quality, they clearly document the normal optic disc appearance of the right eye, and the optic nerve head hypoplasia of the left eye (Figures 1A–B).

Optic nerve head hypoplasia is the most common optic disc anomaly reported in ophthalmic practice.[2] The disc appears as an abnormally small optic nerve head – pink, grey or pale in colour – and is surrounded by a yellowing, mottled peripapillary halo, bordered by a ring of increased or decreased pigmentation (the 'double

Continued page 18

Optic nerve

From page 17

ring' sign). The major retinal veins are often tortuous, with this finding helpful to establish diagnosis.

Visual acuity (VA) in optic nerve head hypoplasia is reported to range from 6/6 to no light perception, and can have accompanying visual field defects. Since VA depends on the integrity of papillo-macular nerve fibre bundle, VA loss does not always correlate with the size of the disc. There is a strong association between optic nerve head hypoplasia and astigmatism,[3] a finding that was present in this case. Optic nerve head hypoplasia is often associated with central nervous system (CNS) abnormalities and endocrine disorders, including isolated growth hormone, thyrotropin, corticotropin, or antidiuretic hormone deficiency.[3] Magnetic resonance imaging (MRI) is considered the optimal non-invasive neuro-imaging modality for delineating associated CNS malformations in patients with optic nerve hypoplasia.[2]

This patient was referred for neuro-ophthalmic MRI and endocrine function investigation, which fortunately returned findings that were unremarkable. Specific amblyopia treatment, such as correction of refractive error or patch penalisation, was not prescribed as significant vision recovery was considered unlikely with the evidence of substantial visual pathway structural anomaly. The esotropia was likely a sensory secondary strabismus and not the primary cause of reduced vision development, with little functional improvement deemed likely with strabismus surgery. The severe vision loss in the affected eye of this patient renders him essentially monocular, so his parent was counselled regarding the importance of eye protection for the fellow eye and periodic review of the fellow eye.

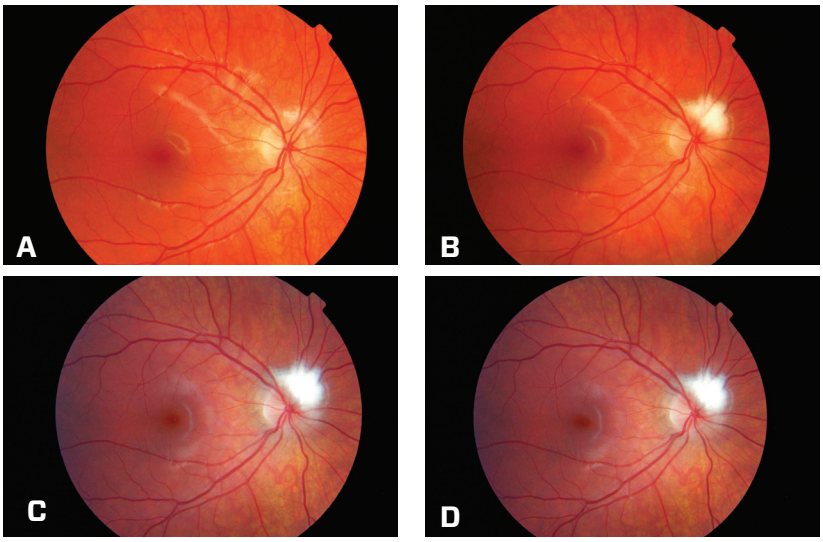

Figure 2. A: September 2012, age six years. B: March 2016, age 10 years. C: May 2017, age 11 years. D: May 2019 age 13 years.

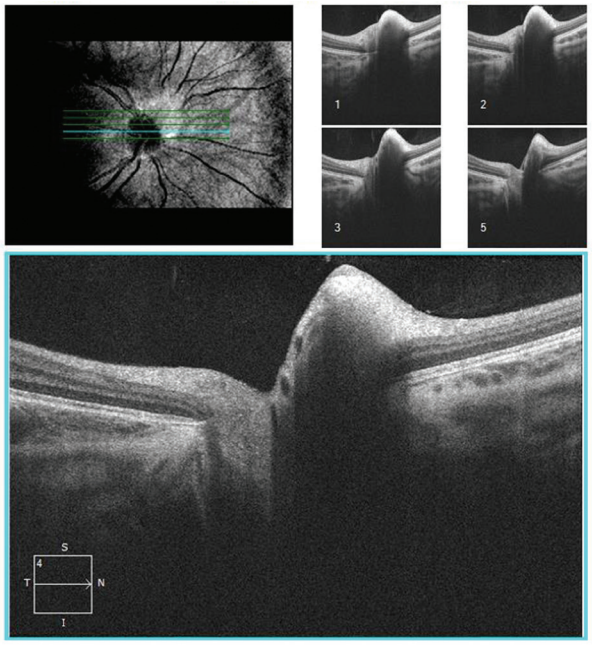

Figure 3. OCT scan May 2017, age 11 years

CASE REPORT 2

Acquired and progressive medullation of optic nerve head retinal nerve fibre

A Caucasian female aged six years presented in September 2012 for investigation of intermittent blurred vision. Entering unaided acuities were: RE 6/6+ LE 6/6++. High AC/A convergence excess esophoria at near was identified and managed conservatively with vision therapy. Baseline retinal images were taken (Figure 2A). The patient re-presented in March 2016, aged 10 years, with low myopic refractive error (RE -1.00 DS LE -1.00 DS) and high AC/A convergence excess esophoria, with VA correcting to RE 6/4.5 LE 6/4.5. Inspection of the optic nerve head showed white striations emanating from the superior temporal rim of the right optic nerve head (Figure 2B). While the appearance was consistent with the focal benign malformation myelination of retinal nerve fibre layer, this was not present in the prior photos, therefore the patient was referred for ophthalmological opinion to differentiate from juxtapapillary inflammation. Ophthalmic ultrasound and OCT scans were performed to rule out the presence of optic nerve oedema, which found no fluid distension of the optic nerve sheath. Continued monitoring with retinal photography and OCT shows increased thickening of the myelination (Figure 2C–D).

Mild thickening of myelination was seen in May 2017 at age 11, with high definition OCT imaging able to document the elevated dense RNFL (Figure 3). Review in May 2019 showed no further change.

Discussion

The prevalence of retinal nerve fibre myelination is nearly one per cent. Acquired and progressive myelination of nerve fibre is rare, however a number of cases are reported in the literature with associations with optic nerve head drusen or other optic nerve head trauma, suggesting that the oligodendrocyte-like cells are able to infiltrate the retina due to an acquired insult.[4]

Medullated nerve fibres are usually seen as white striated patches emanating from the superior and inferior aspects of the disc. Because they can elevate portions of the disc and obscurate blood vessels, mild presentations can be mistaken for papilloedema. The myelination of the afferent visual pathways commences at approximately five months gestation at the lateral geniculate body and terminates at the lamina cribrosa at about term. Oligodendrocytes, which are responsible for myelination of the CNS, are not usually present in the human retina, however they are found in areas of medullated nerve fibres while absent in other areas. Speculative pathogenic theories for how both congenital and acquired cases occur include a defect in the lamina cribrosa or late development of the lamina cribrosa that may allow oligodendrocytes to allow access and migration of these cells into the visible retina.

1. Kiely PM, Slater J. Optometry Australia Entry-level Competency Standards for Optometry 2014. *Clin Exp Optom* 2015; 98: 65-89.
2. Brodsky MC. 1994. Congenital optic disk anomalies. *Surv Ophthalmol* 1994: 39: 89-112
3. Brodsky MC. Congenital optic disk anomalies. In Hoyt CS, Taylor D. *Pediatric Ophthalmology and Strabismus*. New York: Elsevier; 2013. p 726-732.
4. Jean-Louis G, Katz BJ, Digre KB et al. Acquired and progressive retinal nerve fiber layer myelination in an adolescent. *Am J Ophthalmol* 2000: 130: 361-362.

The IMI Reports and Clinical Management Guidelines

The International Myopia Institute's clinical strategies for myopia

Dr Kate Gifford
PhD BAppSc (Optom) Hons GCOT FBCLA FIACLE FCCLSA FAAO

Clinical Optometrist, Gerry & Johnson Optometrists, Brisbane

Visiting Research Fellow, Queensland University of Technology

INTERNATIONAL MYOPIA INSTITUTE

The International Myopia Institute (IMI) White Paper Reports were published in the high-ranking journal *Investigative Ophthalmology and Visual Science* in February 2019. In a similar spirit to the Tear Film and Ocular Surface Society Dry Eye Workshop (TFOS DEWS and DEWS II) reports, the IMI Reports present a comprehensive peer consensus from over 85 participant authors on a wide scope of topics relating to research of myopia mechanisms, product research and development, clinical and industry best practice and the public health message. The IMI Reports are open-access and freely available, creating a clear picture of the current landscape of myopia research and practice, with an eye to the future.

The IMI Reports have come at exactly the right time. Over the past few years there has been a dramatic increase in clinician awareness and product innovations by industry to match the research findings of a global increase in the prevalence of myopia, forecast to affect 50 per cent of the world's population by 2050.[1] The well-informed optometrist would benefit from reading any and all of the IMI Reports, however, if pressed for time, the place to start is the Myopia Control Reports Overview and Introduction, which details the background of risk factors for myopia onset and progression, along with providing an overview of each report to direct further learning. From there, essential practitioner reading includes the following Reports:

Defining and Classifying Myopia – Get clear on the definitions of pre-myopia, myopia, high myopia, and myopia complication with key references.[2]

Interventions for Myopia Onset and Progression – Understand the research behind optical, pharmacological, environmental (behavioural) and surgical interventions for myopia.[3]

Clinical Management Guidelines – Appreciate the scope of risk identification, parent and patient communication, informed consent, basic examination procedures, follow-up schedules, when to change and stop treatment, future treatments and additional resources for clinical practice.[4]

Continued page 20

Management of patients in a domiciliary setting

Susan F Kalff
BScOptom LOSc PGCertOcTher FACO

Katy Kalff
BOptom PGD Adv Clin Optom GCOT

Brighton, VIC

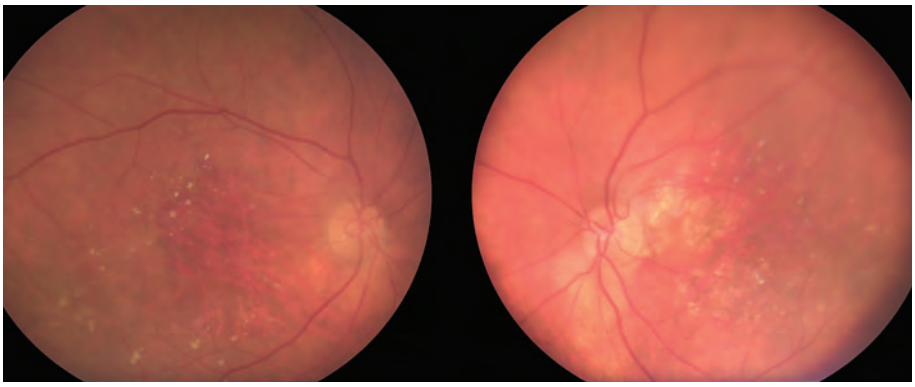

Figure 1. Patient NB: bilateral geographic macular degeneration at last visit May 2019

The incidence of eye disease increases with age, as does the incidence of co-morbidities (ocular and general) and increased frailty. With this comes increased difficulty with mobility, confinement to home or care facility and further confinement to a room or bed. This makes accessing standard desktop instrumentation difficult. Patients with chronic illness, and physical and mental disabilities–including those with challenging behaviours or who are too distressed when taken out of their familiar surrounding–are often also unable to attend an external practice.[1,2,5]

This increases the difficulty of accessing eye care as well other allied health care.

Optometric domiciliary visits may be the only way in which ocular health can be maintained, managed and treated, and refractive corrections kept up to date for the vulnerable patient.[3,4]

Optimum eye care for patients in the home or aged-care or other facilities should include communication with other relevant involved health-care providers, carers and family (if appropriate) with due care and respect for the privacy of the patient.

A full eye examination will involve full medical and ophthalmic history and medications taken. This may need to be done with a family member or carer present, particularly if there are behavioural issues or cognitive disability, followed by an accurate refraction and low vision assessment, dilated fundus examination, with provision of any necessary vision aids.[3,4]

A full eye report provided for each resident seen in a care facility will inform and complement their general health care. This is combined with instructions to carers about eye hygiene, correct application of eye drops and discussions about low vision and visual field loss to enhance patient care and help prevent and manage falls.

Referral for ophthalmological treatments can involve ambulance transport, so suitable wheelchair-access to ophthalmology clinics should be considered. Treatment must be balanced against multiple pathologies and priority of treatment for multiple conditions.[5]

Conditions which may require management in place are: dry eyes, low vision due to AMD, glaucoma, diabetic retinopathy and cataract (if the patient is too frail for surgery), and visual field loss due to stroke or glaucoma.

Glaucoma management is often difficult for a confined patient who may not have seen their ophthalmologist for a long time; has been lost to follow-up; or has inappropriate or discontinued treatment. Co-management with the patient's ophthalmologist may be required, or new treatment initiated by the optometrist.

The following case reports demonstrate the importance of communication between the providers of health care to the vulnerable patient (optometrist, ophthalmologist, general practitioners, nurses, family and personal carers) and the importance of being flexible and able to provide accessible health care options according to the needs of the patient. Sometimes the clinical diagnosis is straightforward and the management becomes complex due to physical and behavioural limitations.

CASE REPORT 1

Mrs NB, a 92-year-old female in 2016, presented for a routine eye examination at an aged-care facility a month after her admission. She presented with a complaint of dry eyes. Mrs NB reported a history of glaucoma, dry eyes, bilateral cataract surgery and left pterygium removal. Mrs NB reported that she was advised to cease glaucoma treatment after the cataract surgery.

Visual acuities were R 6/15 (+1.00/-0.50 x 40) and L 6/9.5-2 (+0.25/-1.00 x 60), n5 part. Intraocular pressures were R 11 mmHg and L 11 mmHg. Slitlamp assessment showed right eye with peripheral iridotomy, the left had two peripheral iridotomies and clear bilateral IOLS. Dilated fundus examination showed optic nerves with RE/LE 0.30, with the left showing a resolving haemorrhage. Both maculae had early hyper- and hypo-pigment

changes. Lubricants were prescribed.

At review three months later, eyes were much more comfortable with regular lubricants. Vision and intraocular pressures were stable. The haemorrhage at the left disc was almost gone. Three months later, intraocular pressures were stable, and vision slightly reduced RE 6/19+2 LE 6/12+. The disc haemorrhage had resolved. At review six months later, her visual status was stable. Annual reviews were recommended.

At annual review in July 2018, Mrs NB reported no visual changes. Vision was measured to be RE 6/30 LE 15-2, IOPS 11/11. The right IOL had some mild fibrosis on the nasal segment. The left IOL was clear. There were early geographic atrophic changes at the right macula and early pigment disturbance at the left macula. Referral was discussed but was declined. Visual acuities were unchanged over the next six months but there was evidence of developing confusion with glasses being mixed up and worn incorrectly.

She was next seen in May 2019 at the request of her family and her general practitioner. The family reported that Mrs NB had complained about 'darkness.'

Her medical practitioner had recently diagnosed Mrs NB with vascular dementia and noted that she had confused images in cognitive assessments. Mrs NB reported no visual complaints. Vision was now RE 6/48 +2 LE 6/36+2, N24 with best correction. Assessment of acuity and refraction was limited due to poor identification of letters associated with her dementia. Near vision assessment was easier as identification of words was better. IOPs were stable (RE 11 LE 12 mmHg). Anterior ocular segment was stable. Posterior ocular segment showed dry geographic macular degeneration more advanced in the left eye. However, visual acuities did not match the appearance of the maculae.

Her increasing vascular dementia and debility limited subjective refraction and accurate measurement of acuity.

Management options were discussed with her family. As there are no treatment options for dry AMD, Mrs NB is frail, has limited ability to leave her aged-care facility and has no insight into her condition, her family decided against referral to her ophthalmologist and to review on an annual basis.

CASE REPORT 2

MH, a 60-year-old female with a history of intellectual disability, tuberous sclerosis, schizophrenia and high anxiety was referred to her local optometrist as she had become hesitant when walking, unable to see steps or uneven surfaces and had lost interest in watching television. As she approached the optometrist's consulting rooms, she had become physically and verbally agitated and refused to co-operate with the examination. She was referred for a domiciliary visit to attempt an examination in her home (a special accommodation house).

MH was calmest in her own room with a trusted carer sitting with her and offering her a slow careful description of what was about to happen.

MH had bilateral, dense brunescent cataracts with no view of the fundi. There was a left exotropia which had been present since birth (noted in her past medical history). Intraocular pressures were R 21 mmHg and L 20 mmHg (iCare tonometer). Visual acuities were R 1/15 and L 1/30 with her existing spectacle correction of: R +4.25/-1.00 x 180 and L +3.00/-1.00 x 180.

MH was referred to a public hospital and was offered an appointment within a month but baulked at the hospital entrance and was unable to attend her appointment. The hospital was advised of the difficulties and rescheduled her appointment a few weeks later, which she attended successfully, mildly sedated and in the company of her sister and her carer.

Bilateral cataract surgery was performed the following month with general anaesthetic and overnight hospital stay. The patient was seen at her domicile two weeks, four weeks and three months post-operatively.

MH had bilateral posterior chamber IOLs, clear and in good position. Visual acuities were R 6/12+ and L 2/30 with insignificant residual refractive correction (Nidek Automated Refractor). Mobility has improved, she is confident and has regained interest in her surroundings.

Treatment of cataracts is generally straightforward and can mean significant improvement to quality of life[6] but appropriate management here was challenging due to the patient's complex issues.

Conclusion

Patients who are extremely frail or have multiple health issues whether physical, cognitive or emotional may be isolated in their home, aged-care or accommodation facility. They frequently have little or no access to eye care in a conventional office. Patients can easily become lost to follow-up when they have missed a few appointments and no longer receive recall letters.

Domiciliary visits by the optometrist to identify and, where possible, manage ocular problems–which may range from correcting a refractive error to managing a complex ocular health issue–can be a significant contributor to improved quality of life, even if the patient is close to end of life. Good communication with those involved in the patient's care and welfare is vital.

1. Kaneko M, Van Boven K, Takayanagi H et al. Multicentre descriptive cross-sectional study of Japanese home visit patients: reasons forencounter, health problems and multimorbidity. *Fam Pract* 2019; pii: cmz056. doi: 10.1093/fampra/cmz056. [Epub ahead of print]
2. Smith, SM, Ferede A, O'Dowd T. Multimorbidity in younger deprived patients: an exploratory study of research and service implications in general practice. *BMC Fam Pract* 2008; 9: 6.
Keel S, McGuiness MB, Foreman J et al. Prevalence, associations and characteristics of severe uncorrected refractive error in the Australian National Eye Health Surgery. *Clin Exp Ophthalmol* 2019. doi: 10.1111/ceo.12647 [Epub ahead of print]
3. Bowen M, Edgar DF, Hancock B et al. The Prevalence of Visual Impairment in People with Dementia (the PROVIDE study): a cross- sectional study of people aged 60-89 years with dementia and qualitative exploration of individual, carer and professional perspectives. Southampton (UK): NIHR Journals Library; 2016.
4. Rizza A, Kaplan V, Seen O et al. Age and gender related prevalence of multimorbidity in primary care: the Swiss FIRE project. *BMC Fam Pract* 2012; 13: 113
5. Owsley C, McGwin G Jr, Scilley K et al. Impact of cataract surgery on health-related quality of life in nursing home residents. *Br J Ophthalmol* 2007; 91: 1359-1363.
6. Owsley C, McGwin G Jr, Scilley K et al. Effect of refractive error correction on health-related quality of life and depression in older nursing home residents. *Arch Ophthalmol* 2007; 125: 1471-1477

Hiromi Yoshikawa
BAppSc (Optom), Grad Cert Oc Ther (QUT), ACO-ACG
Stuart Macfarlane Optometrist, Logan Central, Queensland

Iridoschisis

Iridoschisis is a rare and unusual iris atrophy characterised by separation of iris stroma, in which anterior iris stroma rupture from the posterior layers and the loose iris fibrils project into the aqueous humor (**Figure 1**).[1-4] It commonly affects the elderly between 60 and 70 years of age with a slight female predominance.[4,5] A genetic predisposition is controversial and inconclusive.[5,6] The condition is often bilateral, observed in the inferior iris and it can be progressive.[5] Although iridoschisis is mostly considered to be idiopathic and senile atrophic change of the iris, it is frequently associated with primary angle closure glaucoma.[2,3,6,7] Iridoschisis is also reported to coexist with other ocular pathologies including other types of glaucoma, lens subluxation, keratoconus and ocular blunt trauma.[2,5] The differential diagnosis includes iridocorneal endothelial (ICE) syndrome and Axenfeld-Rieger syndrome, which both share the characteristics of iris atrophy.[3,7] The age onset of ICE syndrome is earlier at 30 and 40 years of age with an ectopic pupil, corectopia and peripheral anterior synechiae (PAS).[3,7] Axenfeld-Rieger syndrome is a congenital disorder accompanied with dental, facial and musculoskeletal anomalies. There are bilateral and asymmetrical involvement of iris atrophy, corectopia and PAS as well as ectropion uvea and prominent Schwalbe's line.[3,7]

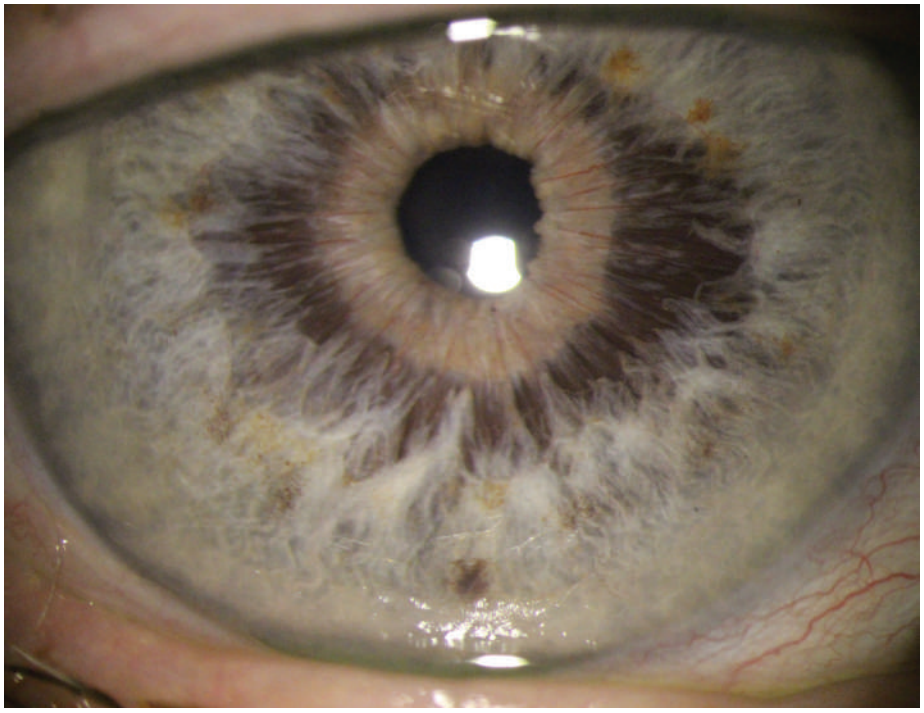

Figure 1.
Iridoschisis

A 77-year-old Caucasian female presented for an ocular review after recent cataract surgery. She mentioned that her unusual eye colour was often commented on. Subjective refraction was R plano (6/6) and L plano/-0.75x100 (6/6). Intraocular pressure was 14mmHg, measured with Goldmann applanation tonometer, and pachymetry of 519 microns, measured with auto kerato-refracto tonometer TRK-2P, in both eyes. Fundus biomicroscopy was unremarkable with symmetrical cup-to-disc ratios, healthy neural rims and no vascular abnormalities. Slit lamp showed numerous disintegrated iris strands floating in the anterior chamber, described as 'shredded' appearance in multiple literatures.[5] Anterior OCT confirmed the diagnosis of iridoschisis (**Figure 2**).

Otherwise, she had clear and well-centered IOL in both eyes with no corneal guttata, quiet anterior chamber and normal conjunctiva.

The patient was diagnosed with bilateral iridoschisis and advised to attend for an annual ocular review to exclude glaucoma and corneal decompensation. •

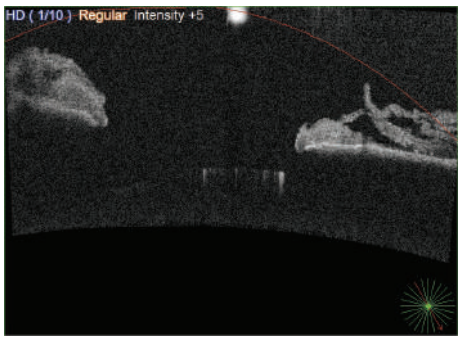

Figure 2.
Iridoschisis clearly visible in anterior OCT imagery

1. Loewenstein, A., and Foster, J., Iridoschisis with multipe rupture of stromal threads. The British Journal of Ophthalmology, 1945. 29: p. 277-282.
2. Mills, P.V., Iridoschisis. British Journal of Ophthalmology, 1967. 51: p. 158-164.
3. Schoneveld, P.G., and Pesudovs, K., Iridoschisis. Clinical and Experimental Optometry, 1999. 82(1): p. 29-33.
4. Pegu, J., Jain, K. and Dubey, S., Iridoschisis: Spectrum of Presentation. Middle East African Journal of Ophthalmology, 2021. 27(4): p. 224-227.
5. Pieklarz, B., Grochowski, E. T., Saeed, E., Sidorczuk, P., Mariak, Z. and Dmuchowska, D. A., Iridoschisis - A Symstematic Review. Journal of Clinical Medicine, 2020. 16(9): p. 3324-3344.
6. Danias, J., Aslanides, J. M., Eichenbaum, J. W., Silverman, R. H., Reinstein, D. Z., and Coleman, D. J., Iridoschisis: high frequency ultrasound imaging. Evidence for a genetic defect? British Journal of Ophthalmology, 1996. 80: p. 1063-1067.
7. Salmon, J.F., and Murray, A., The association of iridoschisis and primary angle-closure glaucoma. Eye, 1992. 6: p. 267-272.

These original clinical images were submitted by Optometry Australia member Minh Nguyen in response to our call for images

Minh Nguyen
BVisSc, MOptom
The Eyewear Shop, Camp Hill QLD

Optometry AUSTRALIA MEMBER

Lipaemia retinalis in a diabetic patient

A 31-year-old female (MI) presented for routine ocular review. She was on insulin, metformin, and lipitor for type 2 diabetes (since 2011) and hyperlipidemia. The patient did not monitor her blood-glucose levels and reported improving serum lipids levels from her last consultation with her general practitioner. Ocular examination revealed best corrected visual acuities of R 6/6-2, and L 6/6-1 with R -0.50/-0.50x33, and L -0.75/-0.50x140. Intraocular pressure with non-contact tonometry was 15mmHg in both eyes. Ocular motility was full and normal. Pupil reactions were normal with no signs of relative afferent pupillary defect. Dilated fundus examination revealed pale salmon appearance of the fundus and creamy white discoloration of blood vessels in both eyes (**Figures 1** and **2**).

The retina appeared significantly different in comparison to retinal imaging in 2018 in **Figures 3** and **4**. In the peripheral retinal there was moderate nonproliferative diabetic retinopathy in the form of blot haemorrhages and microaneurysms in all four quadrants of both eyes. An OCT scan revealed normal retinal nerve fibre layer thickness in both eyes. The macula contour appeared normal with marked hyperreflectivity from blood vessels. MI was diagnosed with lipaemia retinalis and referred to her general practitioner for targeted review of her triglyceride levels. She was also referred through the public health system to an ophthalmologist for further ophthalmic review to rule out other differential diagnosis and for further systemic health review.

Lipaemia retinalis (LR) is an ocular manifestation of elevated serum triglyceride levels in hyperlipidemia.[1, 2, 3] It is characterised by the milky discolouration of retinal blood vessels that reverts back to its original appearance when serum triglyceride levels return to normal range.[1,2] Hyperlipidemia as a primary disorder is related to conditions of elevated chylomicrons, a lipoprotein that transports triglycerides from the intestinal site or absorption to the systemic circulation.[5] Secondary causes of hyperlipidemia include systemic disorders such as diabetes mellitus, obesity, alcoholism, systemic lupus erythematosus, hypothyroidism, and certain medications.[3, 5]

LR occurs when serum triglyceride levels exceed 111.1mmol/L due to increased concentration of chylomicrons.[1, 4] Chylomicrons scatter light and at high concentrations gives blood vessels a characteristic milky white appearance as seen in LR.[5] This can make veins and arteries more difficult to distinguish in the retina.[1] The ophthalmic appearance of LR directly corresponds with serum triglyceride levels in hyperlipidemia. Peripheral retinal vessels are initially affected, appearing creamy and thin in early stages with triglyceride levels ranging from 138.9mmol/L to 194.4mmol/L.[1, 5] In moderate LR with triglyceride levels between 194.4mmol/L to 277.8mmol/L, central retinal vessels are affected. At a severe stage of LR where triglyceride levels exceed 277.8mmol/L, choroidal blood vessels become visibly affected giving the retina a salmon pink colour.[1, 5]

The patient in this report presents a severe case of LR where triglyceride levels likely exceed 277.8mmol/L. Plasma triglyceride levels below 1.7mmol/L are considered within normal healthy range and between 2 to 6mmol/L is considered high.[6] MI has hyperlipidemia secondary to poorly controlled type 2 diabetes. At a three-month review, MI's clinical retinal appearance was unchanged and she reported poorly controlled blood-glucose levels and hyperlipidemia. Close systemic review and lipid-lowering measures are vital to prevent complications of hyperlipidemia such as stroke, heart attack and cardiovascular disease such as atherosclerosis.[6] Measures such as a low-fat diet, regular exercise and maintaining blood-glucose levels to a normal range are necessary for lowering serum-triglyceride levels. Losing weight, consuming more omega 3 fatty acids, and including food with low glycemic index into the patient's diet are also recommended.[5, 6]

LR is a clinical presentation of elevated serum triglyceride levels in hyperlipidemia and optometrists should be aware of the implications of this serious but treatable metabolic disorder.[4] The condition LR does not require treatment but systemic hyperlipidemia requires treatment by lowering serum triglyceride levels with a low fat diet and lipid-lowering medication.[1, 3] Once triglyceride levels return to normal level, the clinical ocular appearance of LR resolves.[4] LR does not typically affect vision, but long-standing cases can lead to irreversible lipid exudation in the retina and loss of vision.[5] In the early stages of the condition LR presents in the peripheral retina and is potentially underdiagnosed. Thorough examination of the peripheral retina may serve as a good clinical indicator of patients with high chylomicron and triglyceride levels.[5] Diagnosis and ophthalmic evaluation are important indicators for systemic evaluation and targeted treatment of these patients.[1]

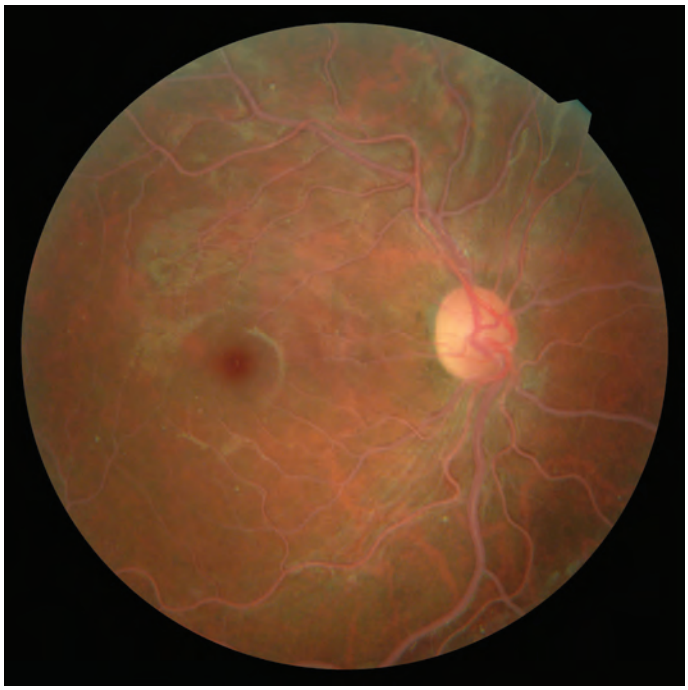

Figure 1.
Right eye retinal fundus photograph with lipemia retinalis 2021

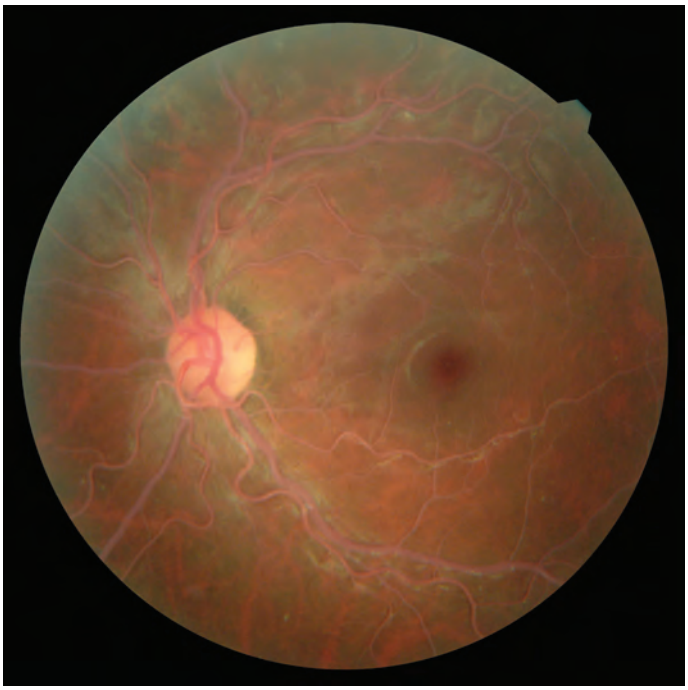

Figure 2.
Left eye retinal fundus photograph with lipemia retinalis 2021

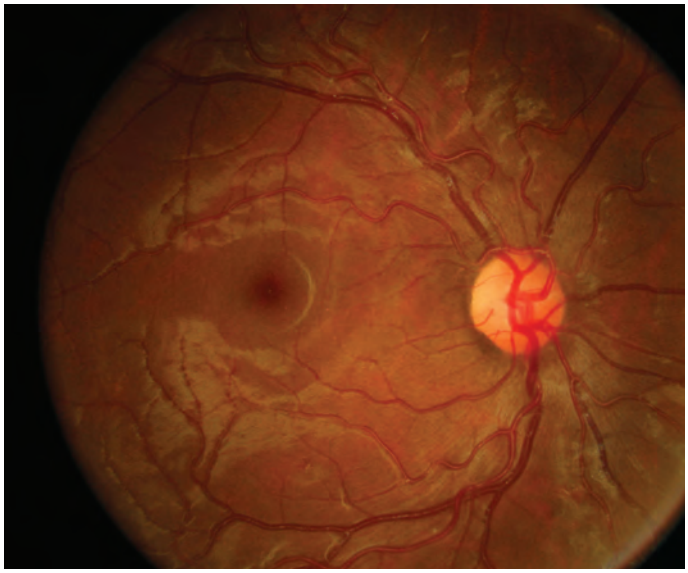

Figure 3.
Right eye digital retinal photography 2018

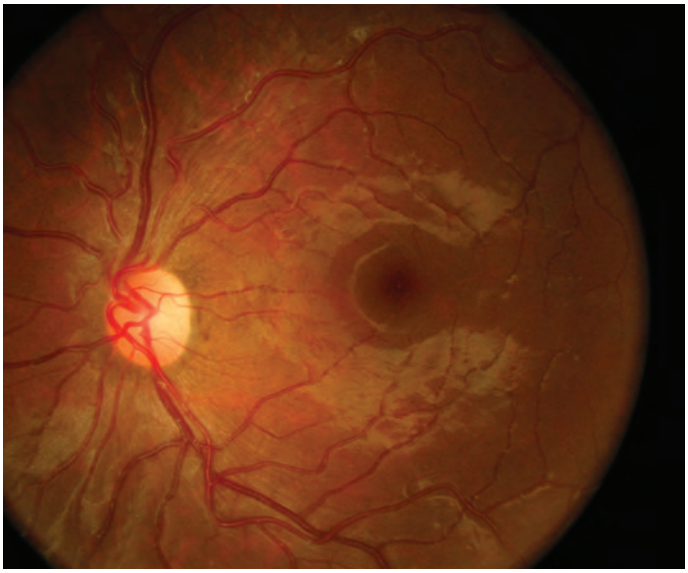

Figure 4.
Left eye digital retinal photography 2018

1. Mishra C, Tripathy K. Lipemia Retinalis. [Updated 2021 Feb 14]. StatPearls [serial on the Internet]. Treasure Island FL: StatPearls Publishing; 2021 [cited 1 Aug 2021]. Available from: https://www.ncbi.nlm.nih.gov/books/NBK555977/

2. Zahavi A, Snir M, Kella YR. Lipemia retinalis: case report and review of the literature. J AAPOS. 2013; 17(1):110-1.

3. Augusto F, Ayhara T, Cypel M, Ishida N, Manzano R. Lipemia retinalis in a 35-day-old infant with hyperliproproteinemia: case report. Arguivos Brasileiros de Oftalmologia 2000; 71(2):254-6

4. Chaudhury D, Meenakshi R, Ramakrishnan R, Mitra A, Raju S. Lipaemia Retinalis.Delhi Journal of Ophthalmology 2015; 26:107-110

5. Rayner, S., Lee, N., Leslie, D., & Thompson, G. Lipaemia retinalis: A question of chylomicrons? Eye 1996; 10(5), 603–608. Available from: https://doi.org/10.1038/eye.1996.138

6. Victoria State Government Department of Health [Internet]. Melbourne: Triglycerides. Better Health Channel; 2020 [cited 1 Aug 2021] Available from: https://www.betterhealth.vic.gov.au/

Hiromi Yoshikawa
BAppSc (Optom), Grad Cert Oc Ther (QUT), ACO-ACG

Optometry AUSTRALIA MEMBER

Coats disease

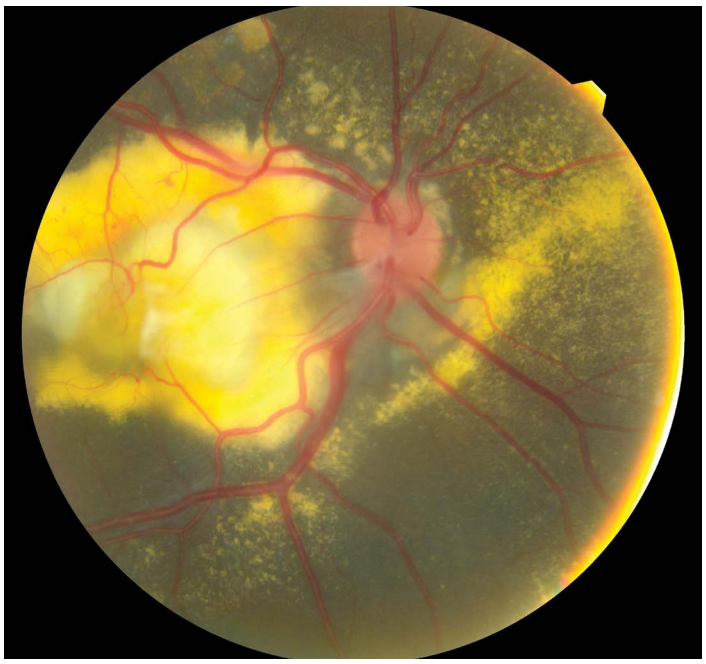

Figure 1.
Diagnosis: Coats disease

A 5-year-old boy presented for his first eye examination, referred from a school vision screening program. Newly acquired strabismus was reported with increasing headaches over the past few months. General health was unremarkable with negative family ocular history.

Unaided vision was R 6/21 with a left head turn and L 6/21 in the primary head position. He had uneven pupil reflexes with right partial relative afferent pupillary defect. Cover test showed right exotropia. Ocular motility was full and smooth. Cycloplegic retinoscopy showed R -0.25/-1.75x22 (6/21) and L +0.50/-2.25x135 (6/12). Whilst the best corrected visual acuity improved to 6/12 in the left eye, there was no visual improvement in the right eye. Dilated fundus microscopy revealed a large area of yellow exudation in the right eye, located temporally to the optic disc with telangiectasias, hard exudates and disc pallor. His left retina was normal. The patient was urgently referred to Queensland Children's Hospital, for fundus fluorescein angiography and neuroimaging, and was subsequently diagnosed with Coats disease. He was treated with repeated intravitreal injections of Bevacizumab and laser photocoagulations over a period of 4 months. Close follow-up with repeated fluorescein angiography was recommended as the visual prognosis was guarded due to extensive foveal involvement. The patient was advised to wear spectacles to correct his left astigmatism.

Coats disease is a rare, idiopathic and non-hereditary retinal vascular anomaly which is characterised by retinal telangiectasia and intraretinal or subretinal exudation.[1,2] The disease is often unilateral and predominantly affects young males in their first or second decades of life. Common presentations are strabismus and xanthocoria or leukocoria.[1,3] The severity of Coats disease at presentation can be categorised into different stages to predict the visual prognoses, including secondary retinal detachment, secondary glaucoma and phthisis bulbi.[4,5] Poor visual outcome is demonstrated in the advanced stages, particularly with the involvement of macular fibrosis subsequent to foveal exudates and sub-foveal nodules.[4]

Retinoblastoma is a critical differential diagnosis of Coats disease, sharing common presentations of leukocoria and strabismus, as well as retinal presentations of vascular dilatation and exudative retinal detachment.[1] In comparison to Coats disease, retinoblastoma is often bilateral and presented at an earlier stage in life with mean age of diagnosis at 1.5 years old. There is no gender predisposition and positive family history may be relevant.[6] In addition, the lipid exudates in Coats disease are typically more yellow compared to white retinal mass in retinoblastoma.[1,6]

1. Wan, M.J., Ma, J., Chiu, H. H. and Lam, WC., A Quick Guide to Pediatric Retina, W.a.L.W. Wu, Editor. 2021, Springer: Singapore. p. 243-251.
2. Yang, X., Wang, C. and Su, G., Recent advances in the diagnosis and treatment of Coats disease. International Ophthalmology, 2019. 39: p. 957-970.
3. Sen, M., Shields, C. L., Honavar, S. G. and Shields, J. A., Coats disease: An overview of classification, management and outcomes. Indian Journal of Ophthalmology, 2019. 67(6): p. 763-771.
4. Daruich, A.L., Moulin, P., Tran, H. V., matet, A. and Munier, F. L., Subfoveal Nodule in Coats Disease: Towards an Updated Classification Predicting Visual Prognosis. Retina, 2017. 37(8): p. 1591-1598.
5. Ong, S.S.a.V., L., Retinal Vascular Disease, A.a.F. Sheyman, A., Editor. 2020, Springer: Singapore. p. 67-75.
6. Aaberg, T.M.a.S., L., Clinical Ophthalmic Oncology, A.a.D. Singh, B., Editor. 2019, Springer, Cham.

Optometry Australia and New Zealand Association of Optometrists would like to recognise the valuable contribution of members, like Hiromi Yoshikawa, who take the time and effort to share their learning experiences with us all. Therefore, we are pleased to announce this article is eligible to be nominated for the ***Optometry Connection™*** **Case Report and Clinical Image Award** for 2022.

The Award is an AUD$1,000 cash prize to be made to the author(s) of the most meritorious case study or clinical image published in *Optometry Connection™* in 2022. Authors must be members of Optometry Australia or New Zealand Association of Optometrists to be eligible. To find out more about the Award and how to submit your case report or clinical image please visit https://www.optometry.org.au/institute-of-excellence/publications/optometry-connection

These original clinical images were submitted by Optometry Australia member Geraldine Bendell in response to our call for images

Geraldine Bendell
BAppSc (Optom) Grad Cert (Oc Ther)
Stuart Macfarlane Optometrist, Brisbane, QLD

Tuberous Sclerosis

Corneal and retinal involvement

Tuberous Sclerosis Complex (TSC) is one of the Phakomatoses. It is a rare multi-system genetic disease that most commonly causes benign tumours in the brain, lungs, skin, renal and cardiac structures.[1] Ocular signs are present in about 50% of people with TSC and include optic nerve harmatomas, elevated intracranial pressure, cranial nerve palsies, cortical visual impairment, and visual field defects.[2] The most common ocular signs are retinal harmatomas which are also found with neurofibromatosis. The lesions can be smooth surfaced noncalcified harmatomas or the classical calcified mulberry harmatoma. Hypopigmented retinal areas are sometimes present. Generally, these retinal lesions are stable and rarely affect vision, with about 50% of retinal signs unilateral.[1]

A 51-year-old patient had been under biannual neurological and annual ocular review since 1997. Acuity was RE 6/6= and LE 6/7.5=. She had a superior field defect in the left eye which had been stable. Perimetry was sometimes difficult as it was a triggering factor for her photosensitive epilepsy. The right fundus was clear. In the left fundus she had an obvious superior harmatoma along with a parafoveal harmatoma which appeared as a hazy area (**see Figure 1 and 2**). These lesions had been stable since initial diagnosis in 1997. She also had a rare finding of a primary corneal harmatoma[3] in the left eye which extended over the inferior third of the cornea to just below the visual axis. The corneal lesion has been slowly extending the last two decades and may warrant a keratoplasty if her visual axis is invaded.

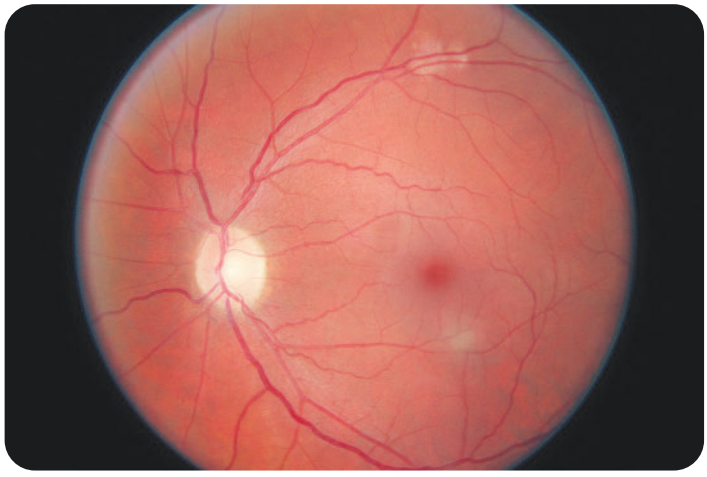

Figure 1.
Fundus image of the LE revealing a retinal harmatoma inferior to the macula and superior harmatoma.

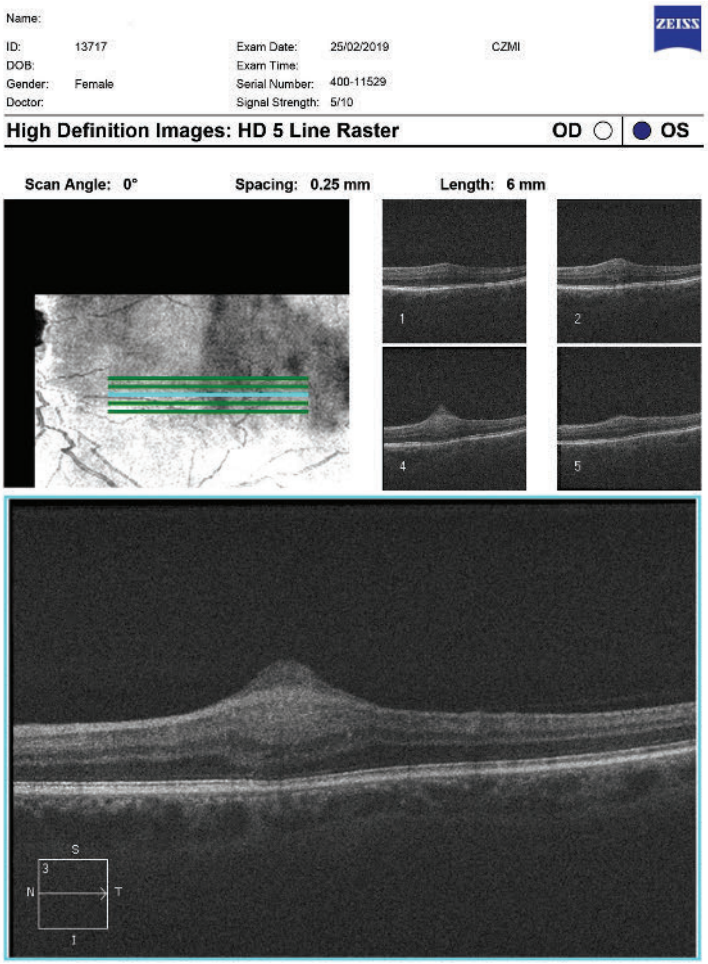

Figure 2.
OCT image of the LE retinal harmatoma inferior to the macula.

1. tsa.org.au [Internet]. Sydney: Tuberous Sclerosis Australia [cited 2021 Feb 2]. Available from: https://tsa.org.au/information/eyes/.

2. Rowley SA, O'Callaghan FJ, Osborne JP Ophthalmic manifestations of tuberous sclerosis: a population based study BJO 2001; 85: 420-423.

3. El Houda HA, Belghmaidi S, Bouda R et al Corneal hamartoma: a rare case report. Poster presented at: 24th European Society of Cataract & Refractive Surgeons Winter Meeting; 2020 Feb 21-23; Marrakech.

Minh Nguyen
BVisSc, MOptom
Stuart Macfarlane Optometrist, Logan Central, Queensland

Talc Retinopathy

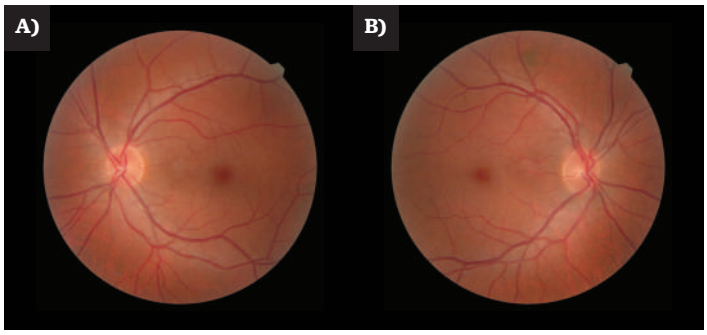

Figure 1.
Retinal photographs **A)** left eye and **B)** right eye

A 43-year-old male presented for an eye examination with chief complaints of postural vertigo and headaches. He reported a history of chronic obstructive pulmonary disease and medications included Ritalin, Sandimigraine, Spiralto, Somac, Champix, Maxalt, Ondasetron, panadol rapid, and occasional recreational marijuana use. Uncorrected visual acuity (VA) was R 6/9, and L 6/12. Best corrected VA was R 6/6+1 and L 6/4.5-2 with R plano/-1.25x105 and L plano/-2.50x80. Intra-ocular pressure measured with Perkins contact tonometer was R 11mmHg and L 13mmHg. Biomicroscopy showed normal anterior segment structures. Dilated fundus findings showed small, refractive crystalline deposits in the retina of both eyes, as shown in **Figure 1A** and **1B**. The optic nerve heads appeared healthy. Optical coherence tomography (OCT) scans and visual field tests (VFT) were performed. VFT with 24-2 testing modality on the Humphrey Field Analyser was normal for both eyes. OCT scan showed multiple hyper-reflective punctate deposits in the inner retinal layers corresponding to the areas of crystalline deposits seen in the retina, as shown in **Figure 2A** and **2B**.

Talc retinopathy is an established retinal finding in patients with a history of long term intravenous drug use of medications containing talc.[1,3,4,5] Talc or magnesium silicate is a mineral used as a filler for some oral medications, such as methylphenidate, and narcotics, such as heroin.[5] It is insoluble in blood and can therefore accumulate in blood vessels and capillaries in the body and the eye.[3,4] Talc retinopathy occurs when enough talc emboli accumulate within retinal vasculature and appear as hyperreflective refractive bodies on fundoscopy.[1] These talc emboli do not typically obstruct retinal vasculature and are usually found incidentally. In these cases, no treatment is required aside from annual retinal review and counselling regarding intravenous drug abuse.[1,3,4] In the rare event that talc emboli occlude retinal vessels, ischemic retinopathy in the form of cotton wool spots and exudates can occur, which may result in proliferative retinal disease. Proliferative retinal disease secondary to vessel occlusion is treated with laser photocoagulation therapy.[4]

Differential diagnoses to consider include other crystalline maculopathies such as Bietti crystalline dystrophy, macular telangiectasia type 2, tamoxifen retinopathy, and Sjogren-Larsson syndrome.[5] Upon further questioning, the patient admitted to previous history of long-term intravenous drug use of amphetamine, commonly referred to as 'speed' and thus this patient was diagnosed with talc retinopathy. The patient had ceased intravenous drug use for several years before his first presentation to our clinic and his retina has remained stable with annual optometric reviews of vision, fundoscopy, OCT, and digital retinal photography for the past 4 years. •

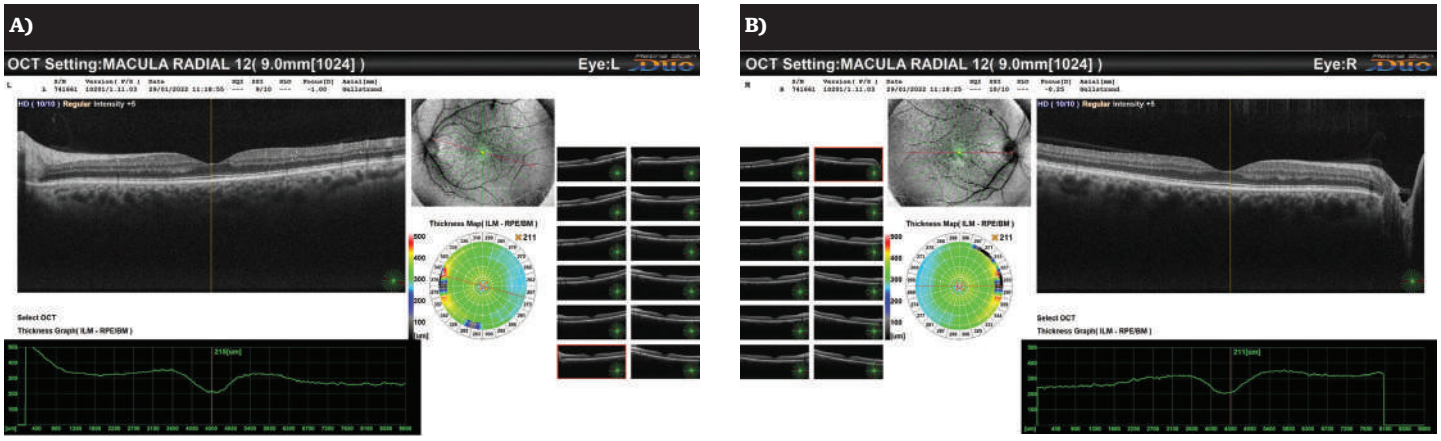

Figure 2.
Macula radial OCT **A)** left eye and **B)** right eye

1. Biousse V, Newman NJ, Peragallo J. Ocular manifestations of drug and alcohol abuse. Current Opinion in Ophthalmology; 2013 Nov [cited 14 Mar 2022]; 24(6): 566-573. Available from: www.ncbi.nlm.nih.gov
2. Garcia DP, Kovach JL. Crystalline retinopathy and optical coherence tomography angiography: new insights in pathogenesis. Ann of Eye Science 2020; 5:24. Available from: www.aes.amegroups.com
3. Gurwood AS. Chalk it up to drug use; c2014 [cited 2022 March 14]. Available from: www.reviewofoptometry.com
4. Muchnick, Bruce G.. Clinical Medicine in Optometric Practice. United Kingdom, Mosby/Elsevier, 2007.
5. Cassell M, Poulose A, Sabates NR, Shah VA. Talc Retinopathy. Ophthalmology, 2008 [cited 18 March 2022]; 115(4): 755-755. Available from: www.aaojournal.org