CASE REPORT

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Unravelling Posner-Schlossman syndrome: Managing acute attacks and long-term control

Introduction

Within the realm of ocular disorders, Posner-Schlossman syndrome (PSS), or glaucomatocyclitic crisis, emerges as a rare yet significant conundrum blending glaucoma and ocular inflammation, thus warranting attention due to its potential impact on vision.

PSS typically manifests unilaterally with abrupt surges in intraocular pressure, accompanied by mild anterior chamber inflammation, causing symptoms like mild eye discomfort, visual blurring and the perception of halos around lights.[1] These intermittent spikes in pressure pose a risk to the optic nerve and can lead to irreversible vision impairment if not promptly managed.[1,2] While the precise aetiology remains elusive, it is conjectured that immunological factors, possibly triggered by viral infections or autoimmune responses, underlie the inflammatory cascade within the eye.[3,4] Comprehending this syndrome holds pivotal importance in preserving visual acuity and broadening our understanding of ocular pathologies. Thus, delving into its complexities and unlocking its mysteries becomes paramount for advancing ocular science and effective management strategies.

Case details

A 70-year-old male presented to the clinic with a complaint of blurry vision in his left eye for the past few weeks, along with mild eye ache. He reported using latanaprost 0.005% + timolol 0.5% (Xalacom) eye drops once nightly, and brinzolamide 1% (Azopt) eye drops twice daily for the left eye, with good compliance. The patient had medically controlled systemic hypertension and hypercholesterolemia, and there was no relevant family history of glaucoma.

Clinical presentation

The patient's visual acuity and refraction measurements were recorded as follows: R +0.75 / -3.50 x 112 (6/6) and L +1.50 / -2.25 x 65 (6/7.5). Intraocular pressure (IOP) readings demonstrated a significant discrepancy between the eyes, with the right eye at 15 mmHg and the left eye at 31 mmHg. Anterior eye examination identified small corneal keratic precipitates and grade 1 anterior chamber cells in the left eye, while the right eye appeared unremarkable. The posterior eye examination showed substantial optic nerve cupping (0.8 cup-to-disc ratio) and neuro-retinal rim thinning in the left eye.

Diagnosis and differential diagnosis

Differential diagnoses included acute-angle closure glaucoma, acute viral uveitic glaucoma, and open-angle glaucoma. However, clinical signs and the progression of the case were consistent with inflammatory glaucoma, specifically PSS.

Management

Initial treatment involved dexamethasone 0.1% (Maxidex) eye drops, one drop every 4 hours for the first week, followed by a tapering regimen of 4 times a day for 2 weeks, then reduced to 2 times a day for 2 weeks to manage the inflammation, alongside the continued use of Xalacom and Azopt. The choice of dexamethasone 0.1% for initial treatment was necessitated by the non-availability of prednisolone in Australia at the time. It is worth noting that both dexamethasone and prednisolone are effective anti-inflammatory choices. Nevertheless, prednisolone is generally preferred for anterior chamber (AC) reactions due to the risk of steroid-induced pressure spikes associated with dexamethasone.[5,6]

A prompt referral to an ophthalmologist was initiated. At the first follow-up, the patient was on dexamethasone 0.1% twice a day. Examination revealed controlled inflammation and reduced IOP of 14 mmHg. His ophthalmologist advised discontinuing anti-inflammatory drops while maintaining his glaucoma medications.

However, during the second follow-up a few months after the first attack, one month after tapering the steroid, the left eye's IOP increased to 29 mmHg. The patient experienced mild discomfort with fresh keratic precipitates and minimal AC reaction. Additional treatment involved reinitiating dexamethasone 0.1% 4 times a day, which was tapered over a month to 3 times a day for a week and then 2 times a day for a week. Unlike the initial treatment, the steroid was not discontinued this time. The patient continued on dexamethasone 0.1% once a day for the next 6 months, successfully controlling inflammation. However, the IOP remained variable, ranging from mid to high teens, prompting the decision for trabeculectomy surgery.

Surgical intervention and follow-ups

Trabeculectomy with mitomycin C was performed, resulting in the cessation of all eye drops. Post-surgery, IOP was well-controlled at 7 mmHg in the left eye. The anterior examination exhibited a well-formed, slightly elevated diffuse bleb with ideal vascularity. Subsequent reviews with an ophthalmologist in 2023 showed maintained visual acuity (OU 6/6), quiet eyes and an IOP of 5 mmHg in the left eye. →

Discussion

Glaucomatocyclitic crisis, known as Posner-Schlossman syndrome (PSS), is a distinctive recurrent unilateral ocular ailment characterised by acute non-granulomatous mild anterior uveitis and elevated IOP. Posner and Schlossman first documented this syndrome in 1948, emphasising its recurrent nature and potential for chronic optic nerve damage.[1]

The clinical course of PSS often involves frequent attacks with symptom-free intervals, leading to potential misdiagnoses during periods of normal IOP.[2] Although the pathophysiology remains elusive, hypotheses range from autoimmunity and autonomic dysregulation to vascular abnormalities. Notably, an infectious theory implicates cytomegalovirus (CMV) and herpes.[3,4]

Studies have linked CMV-positive PSS cases to increased retinal nerve fibre layer thinning, large cup–disc ratio, and corneal endothelial cell loss.[7] Trabecular meshwork inflammation is considered a potential cause for elevated IOP, contributing to progressive glaucomatous optic nerve damage.[8]

Differential diagnoses require careful consideration, including acute angle closure attack, primary open-angle glaucoma and Fuch's iridocyclitis. Gonioscopy is essential for ruling out synechiae, and distinctive characteristics such as iris heterochromia or recurrent spikes in pressures aid in discerning PSS.[9]

Although early reports suggested a positive prognosis with rapid IOP normalisation and minimal visual impact, extended or recurrent cases have revealed irreversible glaucomatous optic nerve damage (GOND). Recent research indicates a 20% incidence of GOND among PSS patients, challenging previous optimism about prognosis. The transient alterations in the optic papilla during acute phases are reversible, but recurrent attacks, prolonged inflammation, and sustained high IOP may contribute to trabecular impairment and secondary glaucoma. The study by Wang et al[10] indicates that PSS patients with glaucomatous damage have a longer disease course, a higher probability of iris depigmentation, elevated interferon gamma (IFN-γ) concentrations and CMV DNA copy numbers in the aqueous humor and reduced endothelial cell density.[10]

Therapeutic interventions focus on reducing IOP and inflammation. Topical anti-glaucoma medications, excluding prostaglandins and pilocarpine, are recommended to prevent exacerbation of inflammation. Topical steroids play a crucial role in inflammation control, with polymerase chain reaction tests on aqueous humor recommended for viral detection, particularly CMV.[3] Ganciclovir, both topically and systemically, has demonstrated efficacy in CMV-positive PSS cases.[3] Despite medical efforts, a considerable number of patients may necessitate glaucoma surgery.[11]

Trabeculectomy with mitomycin C, particularly with releasable sutures, has proven effective in reducing and stabilising IOP, minimising fibrosis-related complications and enhancing postoperative outcomes.[10] This surgical approach not only reduces PSS recurrence but also improves patients' quality of life and eliminating compliance issues associated with multiple eye drops.

Factors influencing patient outcomes in PSS underscore the need for heightened awareness among optometrists. The absence of overt symptoms may lead to care delays, emphasising the importance of optometrists' vigilance for timely diagnosis and management. Patient education on the recurrent nature of PSS is paramount, encouraging them to seek prompt care even for mild symptoms. Regular monitoring is essential, particularly for IOP, treatment response and potential glaucoma progression. Challenges such as cost and travel may hinder patients from returning to ophthalmologists; hence, educating patients about the significance of monitoring with local optometrists is crucial. Optometrists, in collaboration with general practitioners and pharmacists, play a pivotal role in ensuring compliance with topical medications, especially for patients with more recurrent attacks.

Conclusion

PSS presents diagnostic challenges due to its varied clinical course. Differential diagnoses, meticulous investigations and tailored therapeutic interventions are vital for accurately identifying and managing PSS. Awareness of specific clinical characteristics in PSS patients with secondary glaucoma guides clinicians in providing timely and appropriate interventions, emphasising the significance of close monitoring for preserving vision and improving overall outcomes. •

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CASE REPORT

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Re-thinking acute posterior vitreous detachments: Typical findings and risk factors for complications

Introduction

Occupying the posterior segment, the vitreous is a homogenous hydrogel primarily comprised of water, along with collagen fibres, hyaluronic acid and other small molecules.[1] The vitreous protects the retina from oxidative stress through nutrient management and can protect the lens and retina from mechanical damage through its non-Newtonian properties.[1] The vitreous also forms part of the ocular media, allowing efficient light transmission to the retina. During foetal development, it helps regulate the development of the anterior eye.[1]

Age-related changes to the vitreous at a molecular level involve separating the solid and liquid components, termed synchesis and syneresis, but often collectively termed syneresis.[1] This alters the viscoelastic properties of the vitreous, and the outcome is the separation of the posterior vitreous face from the inner limiting membrane of the retina, called posterior vitreous detachment (PVD).[1]

PVD is a normal age-related process and usually has no significant complication and no consequence on long-term visual function.[2] The more serious complications of PVD are retinal tears and rhegmatogenous retinal detachment,[2] which are sight threatening if not detected and managed within an appropriate timeframe.

Flashes and floaters are the most common symptoms of PVD[2] and are a routine presentation to primary optometric care. Despite this routine nature, the clinician must have a thorough understanding of the process of PVD, risk factors and likely time course of severe complications, and the appropriate management based on these risk factors. A minimum standard of skills and equipment for examination is also crucial. This case report discusses the examination findings and management of a patient with a complicated acute PVD.

First visit

A 68-year-old male presented for optometric assessment due to an abrupt onset of floaters and reduced vision in his left eye 2 days prior to presentation. His right eye was not affected. Since onset there had been a constant central visual obstruction; not self-improving. The patient denied the presence of photopsia or a curtain-like obstruction in this affected eye. The patient also denied symptoms of photophobia, ocular pain or redness. The patient has a background of cataracts with myopic shifts and a right PVD 2 years prior. The patient has a strong family history of glaucoma but no known family history of macular degeneration or retinal detachment. His medical history was significant for borderline hypertension and dyslipidaemia, not requiring medical therapy.

Unaided vision was R 6/9.5++ and L 6/60, with subjective refraction giving the best corrected visual acuities of R -1.25 D 6/6 and L -5.00 / -1.00 x 90 6/15= (NIPH). This represents a myopic shift of approximately 2 dioptres since the patient's last spectacles were prescribed 18 months prior. Pupil examination found normal direct, consensual and near responses, with no anisocoria and no relative afferent pupillary defect. Intraocular pressures (IOP) were R 17 mmHg and L 16 mmHg by rebound tonometry at 11:27 am.

Anterior slit lamp biomicroscopy found clear corneas in all layers, white and quiet ocular surface, normal ocular adnexa, normal flat irises and deep, clear anterior chambers.

The presence of worsening nuclear sclerotic cataract – left worse than right – was noted. Pupils were dilated with tropicamide 1.0% and phenylephrine hydrochloride 2.5%.

The vitreous cavity was examined under maximum pupil dilation with slit lamp biomicroscopy. A complete PVD was observed in the right eye, which was previously known. In the left eye, a partial to complete PVD was observed with an unremarkable degree of vitreous degeneration and no evidence of pigment or blood in the vitreous chamber. A dilated fundal examination was undertaken using binocular indirect ophthalmoscopy with scleral indentation. The posterior poles were unremarkable, with healthy optic nerves, maculae and retinal vasculature. No abnormality of the macula reflex or vitreoretinal interface was observed in either eye. A dense vitreous opacity anterior to the posterior pole was observed in the left eye. It was occasionally obscuring view of fundal details but it was not haemorrhagic in appearance. There were no other vitreous opacities observed in the left eye. Examination of the retinal peripheries, with ultra-widefield retinal imaging used as an adjunct (Daytona Plus, Optos, California), found no retinal breaks, tears or detachments (see **Figure 1**). There were some small peripheral retinal haemorrhages in the superior and temporal periphery of the left eye.

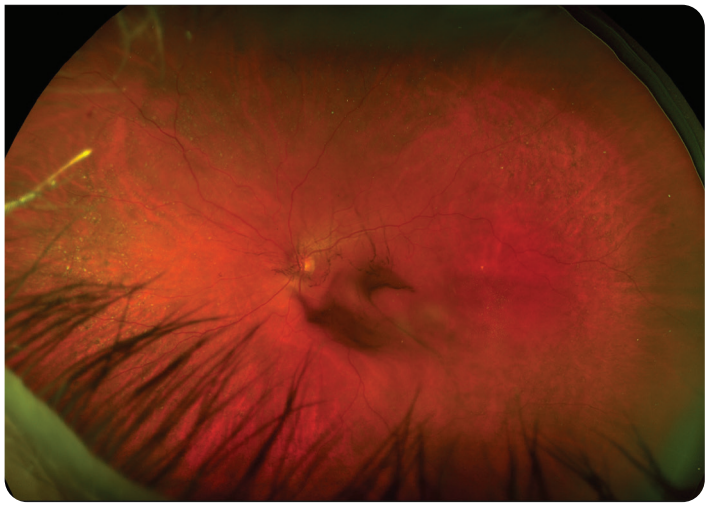

Figure 1. Ultra-widefield digital retinal image of the patient's left eye taken with Optos Daytona Plus at the initial visit. Note the significant vitreous opacity obstructing fixation, the peripheral retinal haemorrhages superiorly and temporally, but absent retinal breaks, tears or detachment.

The patient was diagnosed with an acute left PVD, complicated with a significant central vitreous opacity and minor peripheral retinal haemorrhaging but with no other complication. The secondary finding was the progression of their senile cataracts with a further myopic shift. This primary diagnosis was reached due to the visualisation of the posterior vitreous face on dilated examination of the intermediate segment and the complication of the significant central vitreous opacity in the context of the patient's acute symptoms of floaters and reduced visual acuity. Other complications to rule out in this acute period are retinal breaks, tears or detachment, vitreoretinal tufts, vitreous haemorrhage, vitreomacular traction syndrome, intermediate or posterior uveitis and uveal malignancy. The presence of new retinal haemorrhages is important to note as they can increase the risk of more serious complications both in the acute and delayed timeframes.[2]

It is important to note that delayed retinal tears, detachment or vitreous haemorrhage may still need to be differentially diagnosed for the next few weeks following the initial PVD. Because of the time frame from onset of symptoms to presentation, detection of retinal haemorrhages and significant reduction of visual acuity, the patient was scheduled for a follow-up dilated fundus examination in 6 weeks, but counselled to return as a matter of urgency if they notice a worsening of their floaters, the presence of new photopsia and especially a curtain or large scotoma in their vision.

Second visit (6 weeks later)

The patient presented for review, reporting some improvement in their central acuity, with the vitreous obstruction in vis axis, occasionally moving close to fixation on eye and head movement. There was no worsening of the initial presenting symptoms or other new symptoms.

Visual acuities with the refraction from the previous visit improved to R 6/6 and L 6/9.5=. Re-examination of the vitreous cavity using slit lamp biomicroscopy now showed clear PVD in both eyes with absent of vitreous pigment or haemorrhage. Dilated fundal examination with binocular indirect ophthalmoscopy and ultra-widefield digital retinal imaging was repeated (**Figure 2**).

Once again, no retinal breaks, tears or detachment were observed, nor any other new peripheral retinal abnormality or new vitreous opacity. The retinal haemorrhages remained identical in appearance and location to the previous examination. The vitreous opacity was unchanged in appearance but had moved inferior to fixation. →

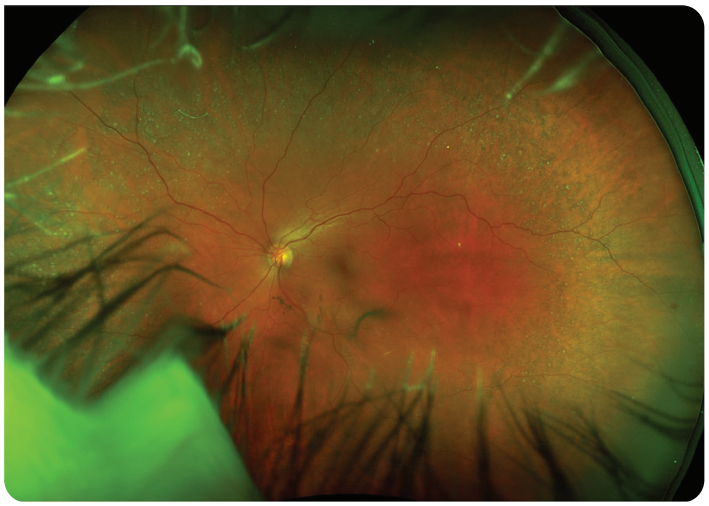

Figure 2. Ultra-widefield digital retinal imaging of the patient's left eye at the follow-up visit. Note the inferior displacement of the vitreous opacities, unchanged retinal haemorrhages and absent retinal breaks, tears or detachment.

Management

The patient was counselled once again about having an acute PVD in his left eye. It was explained that the occasional movement of the vitreous opacity close to the patient's visual axis is normal. However, most patients become increasingly less symptomatic of floaters over the coming weeks. Referral for consideration of pars plana vitrectomy combined with or without phacoemulsification cataract surgery was discussed but was not considered as an option at this stage.

The patient was again reminded to return for re-examination as a matter of urgency if a worsening of flashes and floaters or a curtain obstruction or scotoma becomes apparent. Regarding the patient's cataracts, as the patient retains reasonably good spectacle visual acuity and has no impact on the patient's daily living or quality of life, the patient declined a referral to an ophthalmic surgeon to consider cataract surgeries. A recall was set for 12 months for routine optometric review.

Discussion

The healthy vitreous can maintain constant contact and attachment with the retinal inner limiting membrane through higher concentrations of collagen fibres and proteoglycans in the peripheral vitreous, creating the anterior and posterior hyaloid membranes,[3] and maintaining a homogenous viscosity of the vitreous humor.[1] With aging, the vitreous undergoes a liquefication process called syneresis, whereby collagen fibrils are redistributed to be more concentrated within the central vitreous, and less concentrated in the periphery.[1] This causes retraction of the vitreous' gel-like structure and causes pockets of fluid in the peripheral vitreous called lacunae.[1] The expansion of lacunae eventually leads to PVD.[1] This is a slow process, occurring over months to years and, in many cases, is entirely asymptomatic until the process is complete, with anterior displacement of the entire vitreous body.[4] Sequential bilaterality of PVD is expected as this is an age-related process.[5]

Symptoms of flashes and floaters are classic symptoms of acute PVD. Flashes or photopsia are caused by vitreoretinal traction as the posterior vitreous face separates from the inner limiting membrane of the retina.[3] While floaters are a vitreous media opacity, variably obstructing some areas of the visual axis.[3] Floaters can be due to vitreous haemorrhage, precipitation of vitreous collagen, or torn glial tissue from the optic nerve.[4] The diagnosis of acute PVD is made with the clinical history of an acute onset of flashes and/or floaters, with or without symptoms of the other potential complications, and the visualisation of separation of the posterior vitreous face from the inner limiting membrane of the retina.[3] This visualisation can be achieved using optical coherence tomography or even more easily using slit lamp biomicroscopy of the vitreous chamber through a dilated pupil. **Figure 3** depicts 2 examples of PVD.

Alternatively, the diagnosis can be made using these symptoms plus the visualisation of a round precipitation of vitreous and glial tissue in the posterior vitreous adjacent to the optic nerve.[3,4] This is called a 'Weiss ring' and has been thought to be pathognomonic of PVD as the vitreous is firmly adherent to the retina at the optic nerve, and the final stage of posterior vitreous detachment is often at the optic nerve.[4] However, as found in this case, the visualisation of a definitive Weiss ring is not always possible. Therefore, visualisation of the hyaloid membrane separation as diagnostic criteria is more stringent[3] and provides other qualitative information such as the presence or absence of vitreous blood or pigment. Additionally, optical coherence tomography provides further diagnostic value,[3,4] including further qualitative information on the stage of PVD and other complications, including vitreomacular traction or macula hole.[4]

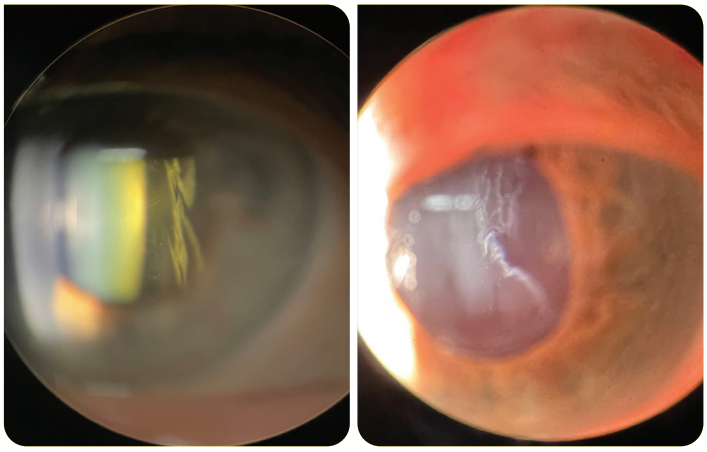

Figure 3. Examples of visualisation of the posterior hyaloid membrane through dilated pupils using slit lamp biomicroscopy of the vitreous chamber. The image on the left is a phakic patient, and the image on the right is a pseudophakic patient. In both images, note the defocus of the iris and lens (or lens implant).

PVD is observed in around 50% of patients over age 70.[2,4,6] Changes to connective tissue caused by menopause, including the collagen distribution in the vitreous, means postmenopausal female patients have a higher incidence of PVD in age-matched groups.[2, 4, 6] Increasing axial length creates additional stress on the vitreoretinal attachment interface and causes liquefication and posterior vitreous detachment to occur sooner in myopic patients.[6] In most cases, posterior vitreous detachment concludes with no complication;[2] however, vitreous haemorrhage, retinal tears and/or retinal detachment represent the most common sight-threatening complications.[2] With a lesser incidence, vitreomacular traction (VMT) syndrome, including an idiopathic macula hole, can also manifest as a sight-threatening complication of posterior vitreous detachment.[4] Flashes and floaters may be present in the patient presenting with VMT as their posterior vitreous detachment process concludes. However, the chief complaint of these patients is more often reduced central vision and metamorphopsia.[4,6] Because PVD begins at the macula, the formation of a persistent vitreomacular adhesion causing traction has often occurred long before the later stages of PVD, with much less clear time course of pathogenesis.[4,6] These complications are far less likely in the absence of PVD.[2]

On initial presentation, around 5% of patients who present with acute PVD have a retinal tear, and 4% have rhegmatogenous retinal detachment.[2] Patient history-based risk factors for these complications include any myopia (particularly high myopia), symptoms of blurred vision, male sex, family history of retinal detachment, age less than 60 and previous keratorefractive or cataract surgery.[2] Patients with a short duration from initial symptoms onset to presentation for care also have a higher risk of complications.[2] This may be due to more dramatic vitreous opacities and reduced vision, which can indicate significant vitreoretinal trauma and potential for serious complications. Flashes or photopsia, while quite specific to PVD,[3] do not increase the risk of complications.[2] Flashes, in fact, seem to be protective, as is hyperopia.[2]

Examination findings that increase the risk of complication do so significantly with a very high odds ratio and include vitreous pigment (i.e., Shafer's sign or 'tobacco dust'), vitreous haemorrhage, retinal lattice degeneration, retinal haemorrhage and visual acuity of 6/12 or worse.[2] The median time to late complications is around 3 weeks from initial presentation, with around 70% of late complications being detected within 6 weeks from initial presentation.[2] Risk factors for a late complication include vitreous haemorrhage, retinal lattice degeneration, history of retinal tear or detachment in the fellow eye, male gender, previous cataract surgery and age under 60.[2] Of the patients with these risk factors that are initially diagnosed with uncomplicated PVD, around 12% of these patients develop a late retinal tear or detachment.[2] Delayed incidence of complications is rare without these risk factors, representing less than 1% of patients who present with acute symptomatic PVD.[2] This highlights the importance of counselling patients not to ignore symptoms in the other eye, especially those with previous complications.[2,5]

Macula-on rhegmatogenous retinal detachment is an ophthalmic emergency, and macula-off detachments and retinal tears are pathologies that require urgent management.[7] Adequate ophthalmic examination to identify these serious complications in patients with acute PVD is crucial to prevent or limit vision loss. Dilated fundal examination with scleral indentation has traditionally been these patients' core/fundamental examination.[7] This allows stereoscopic examination up to the ora serata to best visualise vitreous opacities, retinal elevations from tears or detachment and also allows dynamic examination of sub-retinal fluid if present.[7] This procedure is often uncomfortable for patients and requires significant cooperation, which is sometimes impossible.[7]

Ultra-widefield digital retinal imaging is not a new technology but has had significant adoption in optometry and has been used as an adjunct to assist the examination of patients with acute PVD. Interestingly, studies show that this imaging modality is equivalent, if not superior, to traditional examination in identifying and evaluating the extent of rhegmatogenous retinal detachments.[7-10] There are also many advantages to ultra-widefield digital imaging, which often does not require complete, if any, pupillary dilation and is much faster at acquiring images covering a very large area of retina.[7,8,10] Images can also be stored for use in an office, for education or medicolegal purposes.[7,8] For identification of retinal breaks or tears however, ultra-widefield digital retinal imaging is limited in its efficacy.[7-10] The technology has good sensitivity for tears posterior to the sagittal equator, but for anterior retinal tears, it has been found to be inferior to traditional dilated fundus examination.[7-10] Identification of all breaks is crucial as they are a leading cause of failed surgery for retinal detachment.[7] Studies comparing ultra-widefield imaging generally use equipment similar to an Optos Daytona, and primary gaze imaging only. Detection rate of anterior retinal tears would undoubtedly be higher if steering functions were further utilised in studies, as they often are in clinical practice.[8-10]

Conclusion

Triaging, history-taking, examination utilising adequate techniques and equipment and making an appropriate review and management of the patient with an acute PVD requires a thorough understanding of the risk factors for complications in the immediate and delayed timeframes. Patients with symptoms suggestive of a PVD are typical in optometric practice; therefore, triaging policies must be implemented to allow patients access to timely examination to identify potential complications before vision loss occurs. The examination of the patient with a PVD must be performed at an exemplary level as the potential complications are sight threatening. Flashes and floaters and the presence or absence of tears or detachments at initial presentation may not show the whole story, often requiring a follow-up examination in the coming weeks. Patient follow up, where required, should be guided by history and examination findings, which identify the relevant risk factors for late complications. Patient counselling is also essential to aid in the prompt detection of delayed complications after the initial examination, and even in the fellow eye, which is likely to undergo PVD not long after. •

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CASE REPORT

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Ptosis crutch: An eye-opening approach to literacy in a disability setting

Patients with intellectual disability are often overlooked for interventions that may improve quality of life and access to education. This is the story of how a ptosis crutch has the potential to facilitate literacy for an older adult with an intellectual disability.

Background

The United Nations Convention on the Rights of Persons with Disabilities (CRPD) defines disabilities as long-term physical, mental, intellectual or sensory impairments, which, in interaction with various barriers, may hinder an individual's full and effective participation in society on an equal basis with others. The purpose of the CRPD is to promote, protect and ensure the full and equal enjoyment of all human rights and fundamental freedoms by all persons with disabilities, and to promote respect for their inherent dignity.[1]

In 2023, 4 years after it was formed, the Disability Royal Commission published a series of reports detailing the nature and extent of violence, abuse, neglect and exploitation of people with disability, and recommendations to transform Australia into a more inclusive society that supports the independence of people with disability. The Royal Commission heard of many failures to provide an environment for each person to maximise their potential, including in the health and education systems.[2]

The number of Australian people living with disability is around 18% of the total population[3] and of those, about one in 10 are thought to have an intellectual disability.[4] People with intellectual disability face significant barriers to healthcare and are subject to systemic neglect in the Australian health system[5] and, as such, the Royal Commission has recommended that improved health practitioner education should be implemented at all stages of training.[6] It is recognised that when reasonable adjustments are made when delivering healthcare, health outcomes are greatly improved.[4]

The Australian College of Optometry (ACO) runs a specialist disability service with a wide range of diagnostic equipment. This service is staffed by experienced optometrists to provide optometry eye care to patients who may struggle to find appropriate care in commercial practice due to their disability.

Case report

A 68-year-old patient with an intellectual disability was referred to the ACO Disability Clinic for consideration of fitting with a ptosis crutch due to severe bilateral congenital ptosis. He had adopted a chin-up head posture for many years to manage the condition; however, this inhibited many visual tasks, including reading. His new support worker had observed his passion for word puzzles and encouraged him to learn to read and write for the first time. Nevertheless, this was proving very challenging – in primary gaze and downgaze, the right eye was almost completely closed. →

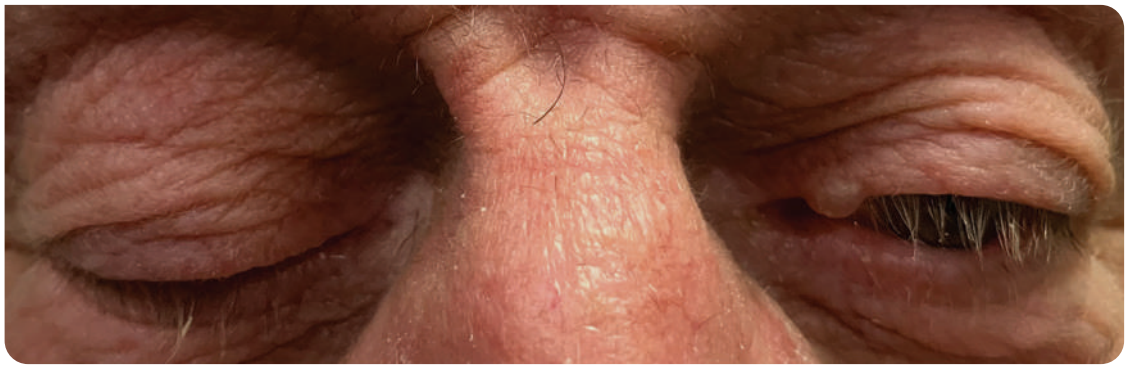

Figure 1. Primary gaze showing severe bilateral ptosis.

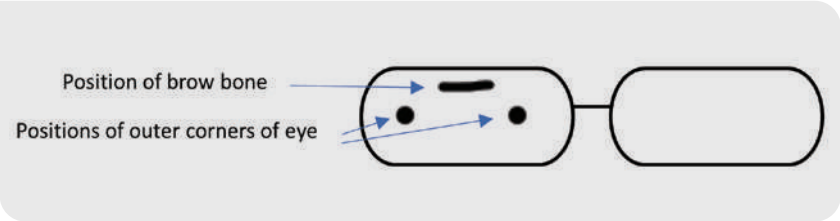

Figure 2. Marking up the frame.

Figure 3. Ptosis crutch attached to lens.

Searching for a solution, he had seen an ophthalmologist but had been advised to try a ptosis crutch before considering any ptosis surgery. In December 2022, the patient presented to the ACO Disability Clinic to determine suitability for ptosis crutch spectacles. His unaided vision was R 6/30++ and L 6/19-.

Refractive results were R +1.25 / -0.50 x 95 (6/12=) and L +1.75 / -1.50 x 170 (6/12-), Add +2.50 (N6). He was observed to have significant bilateral ptosis (right more than left [**Figure 1**]) and moderate cataracts, though his treating ophthalmologist had noted there was no plan for cataract surgery in the near future.

At this initial visit, photographs of the patient's face were taken, and a frame was chosen that fitted his face well. Consideration was given to:

- the size of the frame (larger is better)
- material of the frame (plastic is better as it hides the wire of the crutch)
- the fit against the brow (upper rim needs to sit well above the eye so pupils are approximately on datum)
- material of the lenses (stock polycarbonate lenses work best).

Measurements were taken of the position of the outer canthus of the eyes and the brow bone relative to the frame and the distance between the back of the lens and the brow bone (**Figure 2**).

It was decided to prescribe a pair of single-vision near spectacles with bilateral ptosis crutches using stock polycarbonate lenses. The ACO dispensing lab staff made the ptosis crutches using metal wire coated in plastic. This was attached to the lenses by drilling 2 holes on each side of the frame for the wire to go through (**Figure 3**).

The patient returned to the clinic with his 94-year-old mother and his support worker for fitting his ptosis crutch spectacles (**Figure 4**). The dispensing lab staff made minor adjustments, and he and his support worker were taught how to put the glasses on so that the crutches successfully propped up the lids.

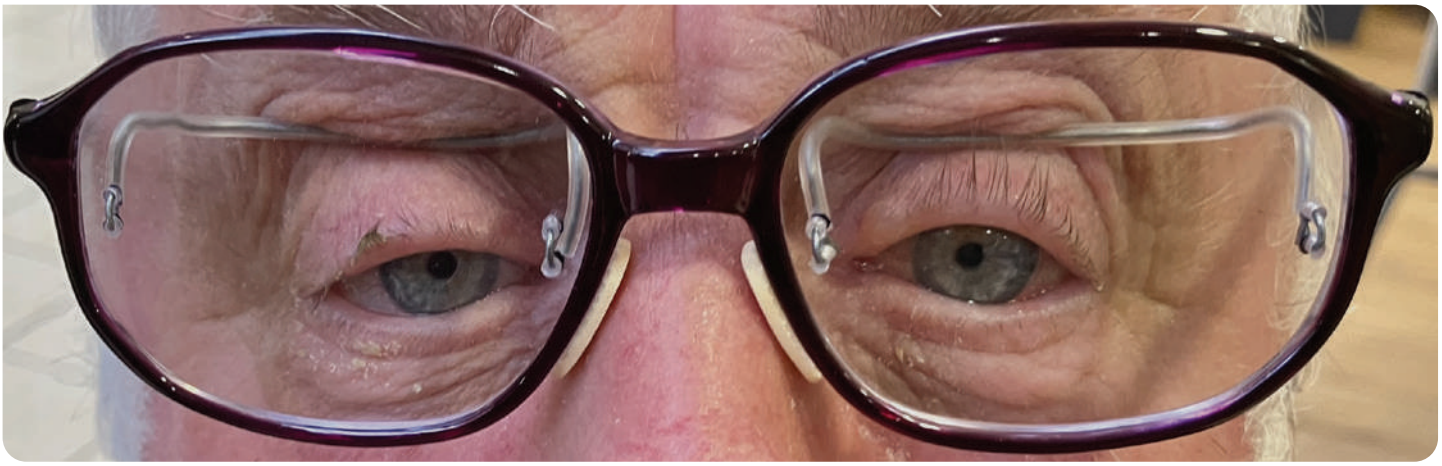

Figure 4. Ptosis crutch fitting.

While the patient was only able to wear the glasses for short periods due to discomfort (ocular lubricants provided only minimal relief), he was able to achieve significant improvement in his ptosis while wearing them, which enabled clearer near vision and access to literacy for the first time at the age of 68. Twelve months on, he is considering whether a surgical option would be a better long-term solution.

Discussion

For a patient with significant congenital ptosis, surgical intervention would be the most common treatment. However, for a patient with intellectual disability, the decision to operate is more complex. People with intellectual disability have a higher risk of anaesthesia-related and postoperative complications,[7] as well as communication and other barriers[8] that may complicate the consent process.

Ptosis crutch is an uncommon intervention – at the ACO, only one or 2 patients per year are referred for consideration of this option, which is not suitable for all of these patients. Ptosis crutch glasses physically hold the eyelids open, which inhibits regular blink action, so it would not be recommended as a treatment for patients with severe ocular surface disease. Further contraindications for patients with intellectual disability may include concerns about compliance, but clinicians should be careful not to make assumptions about the appropriateness of spectacle correction based on the presenting behaviour.[9]

For this patient, it was decided that ptosis crutch was a simple, low-risk and inexpensive option that could improve his literacy without putting him at undue risk. He may still undergo lid surgery in the future. However, in the interim, he has a solution prioritising his human rights to health and education.

With the recent Disability Royal Commission findings, this is a timely reminder that while delivery of healthcare for patients with intellectual disability can be challenging, with some creativity and compassion, all optometrists can contribute to a more inclusive and just society.

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The Australian College of Optometry's (ACO) Carlton clinic operates a specialist disability service, ensuring that patients who may struggle to find appropriate care in community practice receive comprehensive optometry care. The clinic is specifically equipped to meet the diverse needs of patients, including wheelchair users, and is led by optometrists experienced in disability services.

Referrals are not required to attend the ACO's disability service but are welcome/can ensure the patient's needs are accommodated.

Patients who may benefit from the service include those impacted by physical disabilities, intellectual disabilities, Down syndrome, acquired brain injury, Alzheimer's disease and dementia. The ACO's Outreach service also operates a visiting disability service to community residential units and day facilities.

For more information, or to refer to ACO's disability service, follow the link **https://www.aco.org.au/disability-services/** or scan the **QR code**

CASE REPORT

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A rare clinical presentation of superior ophthalmic vein thrombosis

Introduction

Superior ophthalmic vein thrombosis is a rare, sight and life-threatening disease requiring urgent systemic investigation and management. Characterised by an enlarged superior ophthalmic vein and orbital congestion secondary to impaired venous drainage, it has a reported incidence of 3-4 cases per million per year, with 93 documented cases worldwide.[1,2]

Superior ophthalmic vein thrombosis may arise after infective or non-infective aetiologies, with the typical clinical presentation being acute in nature with painful proptosis, orbital oedema, ptosis, ophthalmoplegia, conjunctival injection and chemosis.[3] Uncommonly, concurrent reduced visual acuity or diplopia is present. It has unilateral predilection.

Clinically, superior ophthalmic vein thrombosis may mimic cavernous sinus thrombosis or orbital cellulitis. Therefore, definitive diagnosis is confirmed via neuroimaging modalities, computed tomography venography or magnetic resonance venography with enhanced contrast.[2] In the majority of cases, superior ophthalmic vein thrombosis resolves without significant sequelae following prompt systemic intervention, though instances of longstanding reduced visual acuity and death have been reported.[1]

Case report

This case report illustrates the importance of timely referral and triaging, as well as a multidisciplinary collaborative care approach involving optometrists both in a primary care and private ophthalmology setting, as well as a team of specialists, including an ophthalmologist, neuro-ophthalmologist and cardiologist.

A 72-year-old Caucasian female was referred urgently by her primary care optometrist for ophthalmological assessment due to symptoms of sudden, painful vision loss in the right eye with redness, headache, diplopia on lateral gaze, dyspnoea and tinnitus. She was triaged by TF, the clinical optometrist working at the ophthalmology clinic. They were pseudophakic and had previously undergone right vitrectomy with internal limiting membrane peel and gas to repair a full-thickness macular hole 7 years prior. Medical history included hypertension, dyslipidaemia, asthma, rosacea and neuropathic pain. Trauma was denied.

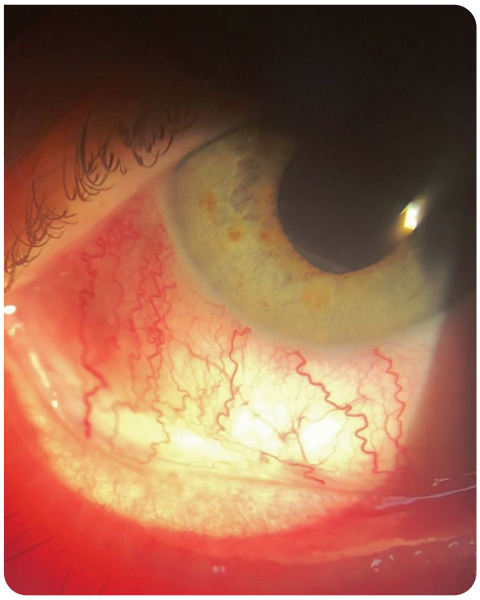

Figure 1. Anterior segment photograph demonstrating right episcleral vascular engorgement due to orbital congestion secondary to impaired venous drainage via the superior ophthalmic vein.

An assessment revealed visual acuities of R 6/9 (PHNI) and L 6/6, with normal pupillary function, motility and monocular colour vision. Intraocular pressures measured R 21 mmHg and L 13 mmHg via Goldmann tonometry. Temporal arteries were non-tender and pulsatile, with right proptosis of 3 mm via Hertel exophthalmometry, superior lid oedema, episcleral vascular engorgement and chemosis (**Figure 1**). The anterior chamber was deep and quiet and the posterior segment was unremarkable.

Following urgent consultation with a neuro-ophthalmologist and cardiologist, a conjunctival swab was performed, after which the patient underwent a thrombophilia screen to assess deficiencies in anticoagulants and commenced oral anticoagulant apixaban 5 mg twice daily. Given that clinical signs suggested a potential carotid cavernous fistula, contrast-enhanced computed tomography venography and magnetic resonance venography of the head and orbits were ordered. Furthermore, a computed tomography pulmonary angiogram was ordered to screen for the presence of pulmonary emboli.

Subsequent neuroimaging demonstrated an acute right superior ophthalmic venous thrombus, given evidence of a longitudinal filling defect within the anterior portion of the right superior ophthalmic vein at the superior aspect of the right orbit in addition to superior ophthalmic vein engorgement (**Figure 2**). Absence of sinus expansion ruled out cavernous sinus thrombosis, and no pulmonary emboli were detected via the computed tomography pulmonary angiogram. Thrombophilia screen and conjunctival swab, which assessed for viral or bacterial cause, were each negative. Therefore, the patient was diagnosed with unilateral isolated superior ophthalmic vein thrombosis.

The 6-week review saw the complete resolution of ocular symptoms with visual acuities returning to 6/6 monocularly. Repeat neuroimaging revealed thrombus resolution in addition to residual perivascular adipose tissue stranding of the superior orbital vein (**Figure 3**). Underlying aetiology was suspected to be secondary to systemic risk factors alongside potential association with recent COVID-19 infection, where ongoing monitoring and investigations continue.

Discussion

The valveless superior ophthalmic vein arises from the union of the angular and supraorbital veins within the orbit, having a mean diameter of 2 mm.[4,5] As the largest vein of the orbit, it is responsible for the majority of orbital venous drainage with termination directly into the cavernous sinus, where dilation is determined when vessel diameter exceeds 2.5 mm. Such cases are positively correlated with intracranial pressure among other sequelae.[4,5]

Superior ophthalmic vein thrombosis may arise secondary to hypercoagulable states, blood stasis or vascular trauma, which impede venous blood flow and drainage, resulting in orbital congestion and symptom onset subsequent to the regurgitation of blood flow.[6,7,8] Such risk factors from Virchow's triad also point to symptom onset on patient awakening, where venous stasis secondary to a supine position may be an exacerbating factor.[7,8]

Underlying aetiologies may be infective or non-infective, with orbital cellulitis responsible for most non-infective cases (**Table 1**).[1,6] Infective causes are most commonly linked to *Staphylococcus aureus*; therefore, broad-spectrum antibiotics should be considered in first-line treatment of cases of suspected infectious aetiology.

Given its similar presentation to cavernous sinus thrombosis and orbital cellulitis, superior opthalmic vein thrombosis can be difficult to distinguish clinically. Further, superior opthalmic vein thrombosis, cavernous sinus thrombosis and orbital cellulitis may present concurrently as a result of orbital adipose tissue and/or cranial nerve involvement within the cavernous sinus, leading to significant intracranial sequelae. Differentiation in such instances is critical given that superior ophthalmic vein thrombosis is a precursing symptom of cavernous sinus thrombosis, which, if not appropriately diagnosed and treated, could lead to pituitary insufficiency, hemiparesis and death.[9]

Enhanced contrast computed tomography venography or magnetic resonance venography enables the definitive diagnosis of isolated superior ophthalmic vein thrombosis or superior ophthalmic vein thrombosis with concurrent cavernous sinus thrombosis and orbital cellulitis.[2,9] In superior ophthalmic vein thrombosis, computed tomography venography demonstrates an enlarged superior ophthalmic vein with intraluminal filling defects, which may be present in conjunction with perivascular

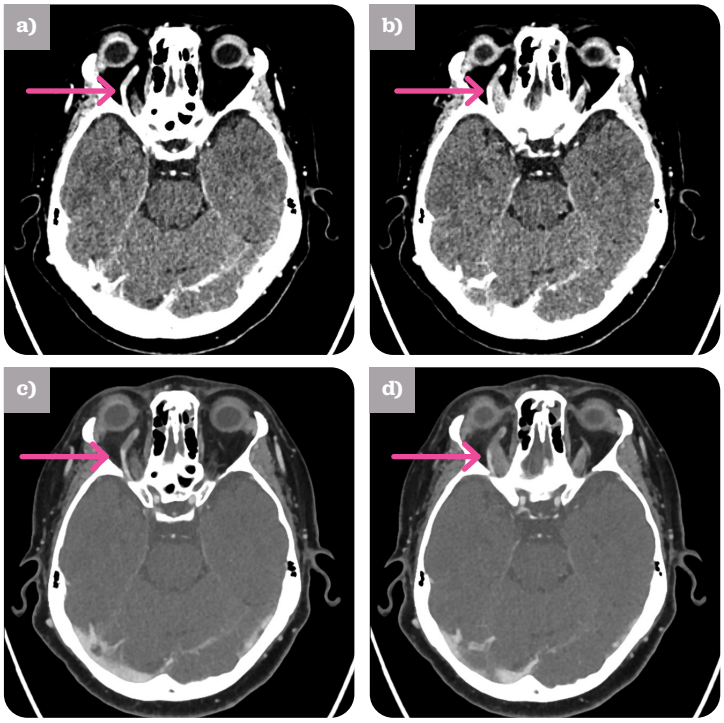

Figure 2. a & b) Enhanced contrast computed tomography venography and **c & d)** magnetic resonance venography axial plane demonstrating right superior ophthalmic vein engorgement and longitudinal filling defect within the anterior portion of the right superior ophthalmic vein at the superior aspect of the right orbit.

oedema, adjacent extraocular muscle enlargement or adipose tissue stranding.[2] Magnetic resonance venography, on the other hand, is more sensitive, demonstrating the enhancement of enlarged extraocular muscles and intraorbital adipose tissue with filling defects of the superior ophthalmic vein intraluminal space.[8,9] Proptosis may also be evident in computed tomography and magnetic resonance modalities.

In the majority of cases, superior ophthalmic vein thrombosis resolves without significant or longstanding sequelae, though permanent vision loss and death have been reported in chronic or infective diseases.[10,11] Critical to optimal prognosis are prompt systemic investigations to determine underlying aetiology, particularly those without infective symptoms. Comprehensive laboratory evaluation should include complete blood count, serum angiotensin-converting enzyme, rheumatoid factor, antineutrophil cytoplasmic antibody, antimicrosomal antibody, and thyroid function.[10] A hypercoagulability screen is also indicated, including factor V Leiden, prothrombin gene mutation, lupus anticoagulant and antithrombin III.[11] →

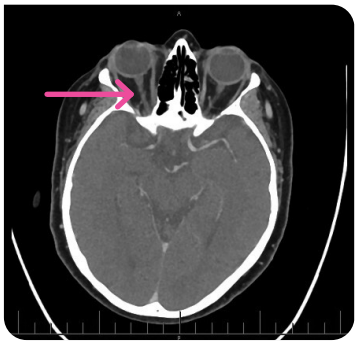

Figure 3. Enhanced contrast magnetic resonance venography axial plane demonstrating right superior ophthalmic vein thrombus resolution at 6-week review in addition to residual perivascular adipose tissue stranding of the right superior orbital vein.

NON-INFECTIVE		INFECTIVE	
Vascular malformation	Dural cavernous fistula Arteriovenous fistula Carotid cavernous fistula Ophthalmic vein varix	Sinusitis	Paranasal sinusitis Sphenoidal sinusitis Ethmoidal sinusitis Pansinusitis
Autoimmune	Graves' disease Systemic lupus erythematosus Ulcerative colitis	Orbital infection	Orbital cellulitis
Trauma	Facial trauma	Dental infection	Dental infection with orbital cellulitis or pansinusitis
Haematological	Hypercoagulable states Autoimmune haemolytic anaemia Antiphospholipid syndrome Sickle trait Hereditary haemorrhagic telangiectasia	Facial infection	Facial cellulitis Nasal furunculosis Masticular space abscess
Neoplasm	Adenoid carcinoma of the nasal mucosa Antifibrinolytic and thrombopoietin receptor agonists Cavernous sinus meningioma	Other	Lemierre syndrome Otomastoiditis
Hormonal	Hormone replacement therapy Tamoxifen (selective oestrogen receptor modulator) Oral contraceptive pill		
Other	Diabetes mellitus Idiopathic orbital inflammatory syndrome Tolosa-Hunt syndrome Idiopathic		

Table 1. Aetiologies of superior ophthalmic vein thrombosis. Adapted from Van der Poel et al.[1]

Following the determination of an underlying cause, targeted treatment can ensue. However, while awaiting systemic investigations, broad-spectrum oral antibiotics and anticoagulants should be initiated in the absence of contraindications.[6,7,12] While such an approach remains controversial within the limited literature, it is deemed to be clinically sound through documented case reports after considering the risks and benefits.[3,10,11] Oral and intravenous antibiotics or steroids may be employed once aetiology is determined, where surgical intervention may be required in infectious causes such as sinusitis and orbital cellulitis with the drainage of the infection source or orbital decompression in cases of Graves' disease.[11,12]

Conclusion

Isolated superior ophthalmic vein thrombosis is a rare clinical presentation and ophthalmic emergency that requires early detection and management to enhance prognosis and prevent life-threatening thromboembolic events. As primary care clinicians, it is important to recognise the clinical presentation of superior ophthalmic vein thrombosis and its differential diagnoses, particularly cases of sudden onset painful proptosis, which warrant urgent referral. It should be considered in all cases of sudden onset painful proptosis while being distinguished from cavernous sinus thrombosis and orbital cellulitis through neuroimaging. With timely triage and systemic treatment, prognosis is variable, though typically favourable, with complete recovery commonly reported; however, urgency is key to reducing the risk of progression, profound vision loss and mortality.

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Fundus albipunctatus: What the fleck is that?

Fundus albipunctatus is a form of congenital stationary night blindness and a rare and currently untreatable autosomal recessive retinal dystrophy. Due to the presentation of multiple fleck-like lesions scattered across the retina, fundus albipunctatus is categorised under flecked retina syndromes alongside more well-known genetic dystrophies like Stargardt's disease, familial drusen and retinitis punctate albescens.[1] Affected individuals have mutations in the RDH5 gene that encodes the 11-cis retinol dehydrogenase enzyme, thus impairing communication in the retinoid cycle.[2] The decreased 11-cis chromophore in the rhodopsin layer prolongs dark adaptation duration to 120 minutes.[1,2] The prevalence of fundus albipunctatus is unknown; there are only a few hundred cases reported in the literature thus far; hence, it may not present commonly in clinical practice. It is estimated that 38% of fundus albipunctatus cases involve cone dystrophy alongside the underlying rod abnormality.[3] The traditional view that fundus albipunctatus is a mild, nonprogressive condition is increasingly being contended as there are reports of cone dystrophy development associated with older patients.[4] The level of vision impairment from retinal dystrophies varies with individuals; however, maintaining transparency about the potential visual ramifications of fundus albipunctatus is key. Proper communication and education enable patients to make informed decisions about important aspects of their lives.

Case report

A 40-year-old Caucasian female presented with declining near vision and longstanding poor night vision. She reportedly experienced slow dark adaptation, which was detected during childhood and had remained stable over the past 3 decades. Her general practitioner had initially referred her upon concerns of having retinitis pigmentosa. There was no known family history of retinal dystrophies or degenerations.

Unaided vision was R 6/9.5 and L 6/9.5++. Refraction was R +1.25 / -1.50 x 160 (6/6++) and L +0.75 / -1.50 x 46 (6/6++) with a reading add of +0.75 in both eyes. During the vision screening process, it was noted that the patient required a minute to regain a reasonable level of visual acuity in the left eye after the occluder was removed.

Intraocular pressures measured with iCare tonometry were R 21 mmHg and L 23 mmHg. Anterior slit lamp biomicroscopy was unremarkable. Posterior fundus examination and ultra-widefield retinal photography revealed numerous white-yellow, fleck-like, punctate lesions radiating from the central arcade to the mid-peripheral retina, primarily concentrated in the superior, nasal and temporal quadrants (**Figures 1a & b**). No bone spicules were apparent. →

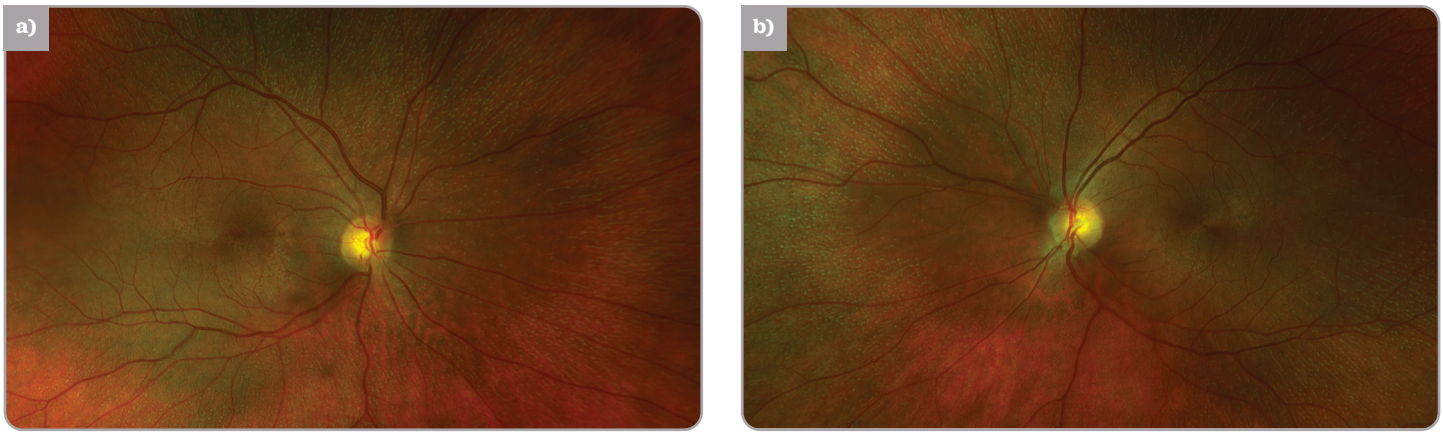

Figure 1. Ultra-widefield retinal photography revealing numerous white-yellow, fleck-like, punctate lesions radiating from the central arcade to the mid-peripheral retina, primarily concentrated in the superior, nasal and temporal quadrants. **a)** Fundus of the right eye. **b)** Fundus of the left eye.

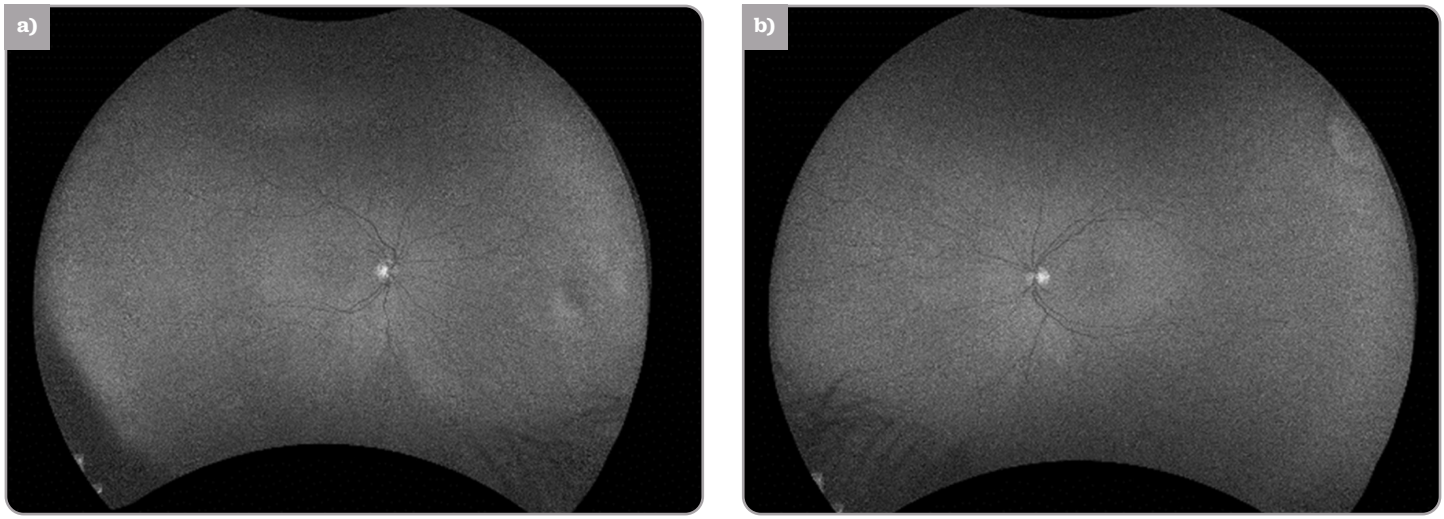

Figure 2. Fundus autofluorescence showed decreased background autofluorescence and poor, grainy image quality. **a)** Fundus autofluorescence of the right eye. **b)** Fundus autofluorescence of the left eye

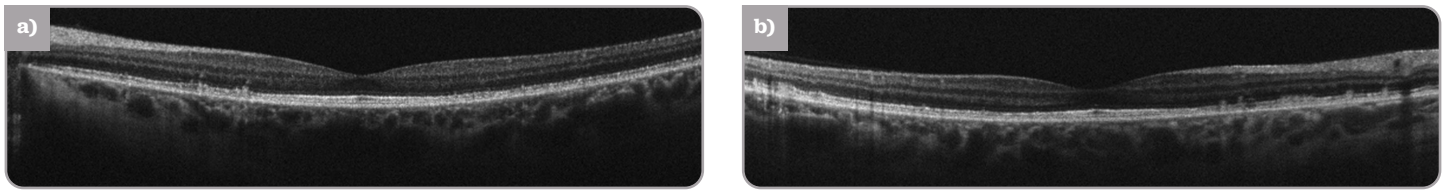

Figure 3. Optical coherence tomography (OCT) macula radial scans showed multiple, small disturbances in the photoreceptor layer corresponding to the lesions' locations. **a)** OCT macula radial scan of the right eye. **b)** OCT macula radial scan of the left eye.

A decreased background autofluorescence and poor, grainy image quality was salient on fundus autofluorescence (**Figures 2a & b**). Optical coherence tomography (OCT) macula radial scans showed multiple, small disturbances in the photoreceptor layer corresponding to where the lesions were located (**Figures 3a & b**). Visual fields were full to confrontation.

The patient was diagnosed with fundus albipunctatus, a rare retinal dystrophy involving congenital night blindness, which was deemed untreatable but likely nonprogressive.

At a subsequent visit, she undertook auxiliary colour vision testing. The Farnsworth D15 test established 2 Tritan crossings. Simultaneously, the City University Test revealed 3 Tritan errors, compounding suspicions of a tritanomalous defect.

The patient was referred to a tertiary centre for further investigation, including genetic testing and diagnosis confirmation via electroretinography (ERG) and Goldman perimetry. Additionally, her ophthalmologist recommended she return to primary optometry care for presbyopia correction.

Discussion

Diagnostic clinical pearls

Diagnosing fundus albipunctatus in a community care setting relies on visual recognition of the fundus signs and cues from the irregular autofluorescence. The low autofluorescence signal on the widefield scans is due to the lack of retinoid-derived fluorophores and the poor uptake of lipofuscin.[5] On OCT, half of the patients with fundus albipunctatus have a degree of macula involvement.[6] Foveal sparing involving no lesions in the central zone was present in 83.3% of affected cases, while 16.7% had macula disruptions and consequently decreased visual acuity.[6]

On electroretinogram (ERG), all patients with low autofluorescence signal on fundus autofluorescence have decreased rod responses, exhibited through the drop in b-wave amplitude in scotopic conditions when responding to a flash stimulus.[5,6] Concurrently, 33%–73.9% of patients' photopic ERG results are outside normal limits, with the poor cone responses equating to colour vision impairment.[5,6] Thus, colour vision screening, especially checking for blue-yellow confusion from S-cone impairments, is important for patients with retinal dystrophies. As there is a lack of treatment for colour vision defects, Chan et al. reinforced that it is in a patients' best interests to be counselled about career restrictions or disadvantages.[7]

Understanding the impact on vision – The patient's perspective

During a conversation, the patient revealed that, fortunately, fundus albipunctatus had limited impact on her quality of life. She experienced no difficulties with activities such as night driving and watching movies at cinemas, occurring in scotopic and mesotopic settings. The patient was amused to discover that she had a tritan colour vision defect. Recently, she hung 2 contrasting blue and green paintings adjacent to each other in the hallway. While she saw similar shades of aquamarine in harmony, guests with presumably normal colour vision often commented about the visually mismatched arrangement. She described ongoing disagreements with family members about colour perception and was enlightened to learn the cause of her unique artistic taste.

Management of retinal dystrophies: Clinical trials and novel treatment options

Options for slowing disease progression or reversing damage are being explored. Gene augmentation therapy has a growing potential as a treatment means for inherited retinal dystrophies like fundus albipunctatus.[8] The procedure relies on an adenoviral vector to transport a package of tailor-made, modified genetic material to replace the faulty RDH5 gene.[8] Gene therapy has an invasive delivery method. It is often performed surgically via vitrectomy alongside a subretinal injection or, in lesser cases, administered intravitreally.[9] Promisingly, 41% of gene therapy participants improved their best corrected visual acuity, while 51% improved in visual sensitivity (full light sensitivity, contrast sensitivity and visual fields).[9]

There are valid qualms about the safety of gene therapy. Adverse effects are prevalent within 30 days of the procedure, including ocular surface inflammation and external adnexa.[9,10] Anterior uveitis is the most common side effect of intravitreal administration of gene therapy.[10] The long-term implications are unknown, although perifoveal and retinal thinning are risks.[9] Clinical trials have been primarily performed in the United States, and there is no FDA-approved gene therapy treatment for fundus albipunctatus.

Aside from gene therapy, oral intake of 9-cis-betacarotene based on a mouse model has preliminary experimental trials with mixed results in human participants. According to Rotenstreich, high doses of the medication for 90 days promote rod recovery, boosting ERG B-wave amplitude and visual field results.[11] By contrast, low levels of 9-cis-betacarotene administered in oral alga capsules for one year hold the undesirable effect of diminishing ERG responses.[12] The research available does not investigate the safety of plasma betacarotene levels, lacks placebo-control groups, and is based on low patient numbers, primarily due to the rarity of fundus albipunctatus.[11,12]

As fundus albipunctatus does not have a cure, genetic testing and counselling benefit affected individuals. As 70% of patients with inherited retinal dystrophies have parents of consanguineous marriages, genetic testing is recommended for immediate relatives.[13] Seeking genetic counselling before family planning can inform a couple of the likelihood of their offspring carrying mutated genes. Fundus albipunctatus is a non-syndromic disease, meaning the clinical abnormalities are limited to the eyes and visual system.[13] Fortunately, the expressions of the RDH5 gene mutation are not linked to classic syndromic issues such as polydactyly, intellectual disability, impaired speech and hearing.[13] Referring a patient for genetic testing offers an opportunity to discover the full extent of their condition and gain access to partake in clinical trials where available.

Conclusion

As our modern society continues to pursue breakthroughs in research and medicine, an incurable condition like fundus albipunctatus has hope for treatment. This case report illustrates the use of multi-modal imaging and the importance of recognising diagnostic features of fundus albipunctatus, understanding the anticipated long-term visual implications and recommending genetic counselling for patients.

Acknowledgements

The author would like to thank Dr Roland Bunting and colleagues for their insights relating to the case. Written patient consent was obtained for producing this case report. ●

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CASE REPORT

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Papillary puzzles: Navigating the CLPC maze with a steroid response twist

Introduction

Contact lens intolerance is a common presentation in optometry practice, prompting patients to seek professional advice for their discomfort. Contact lens-induced papillary conjunctivitis (CLPC), or giant papillary conjunctivitis, is a frequent cause of such intolerance.[1] The characteristic signs are the presence of giant papillae on the superior tarsal conjunctiva, accompanied by symptoms of itching, redness, eyelid swelling, burning or stinging, discomfort, watery secretions, foreign body sensation, white-stringy mucous secretions and contact lens intolerance.[1,2] CLPC aetiology is not fully understood but is likely multifactorial, with lens bio-deposition, lens material, poor hygiene practices and mechanical friction all contributing to the condition.[1,3,4] While the mainstay therapy for CLPC is the cessation of contact lens wear, additional medical therapy with topical antihistamines, mast cell stabilisers and corticosteroids are frequently employed to control inflammation and alleviate symptoms.[1]

CLPC management is often time-consuming, complex and particularly challenging in patients with coexisting ocular conditions.[5] This report presents a case of CLPC in a patient with known steroid responsiveness and pigment dispersion syndrome (PDS), highlighting the need for individualised treatment plans in such circumstances. The complex ocular history necessitated careful consideration to manage the potential risks and benefits of different treatment options.

Clinical findings

Mrs J, a 44-year-old female, presented with an 8-day history of bilateral red, watery eyes and foreign body sensation, exacerbated by contact lens wear. The discomfort began in her left eye (LE) and quickly after was felt in her right eye (RE). Removing the lenses provided temporary relief, but symptoms recurred immediately upon reinsertion a week later, prompting her to seek an eye examination before trying other treatments.

Her ocular history included approximately 20 years of SiHy (silicon hydrogel) monthly replacement contact lens wear following photorefractive keratectomy for moderate myopia. There was also a 10-year history of seasonal allergic conjunctivitis and hay fever, with one episode requiring topical corticosteroid treatment, resulting in a moderate steroid response with elevated intraocular pressures (IOP). Subsequent episodes were managed with a combination topical antihistamine/mast cell stabiliser (ketotifen 0.025%) and oral antihistamines. Steroid response history was significant as Mrs J had been diagnosed with PDS 5 years prior and was being monitored regularly for inferior retinal nerve fibre layer (RNFL) thinning in the LE, with no appreciable visual field or optic nerve head changes.

On examination, best-corrected visual acuities were RE 6/6 and LE 6/7.5, and a mild LE ptosis was evident. Anterior slit lamp examination showed flaky skin and telangiectasia on both upper eyelids, stringy discharge in the nasal canthi and Efron grade 3 conjunctival injection near the limbus, tapering to grade 2. Eyelid eversion revealed cobblestone papillae RE grade 4 and LE grade 3. Both inferior palpebral conjunctivae were inflamed, with a single follicle evident in the RE. Mild diffuse punctate keratitis was present on both corneas, with a sizeable coalescent patch centrally in the LE. Krukenberg spindle and spoke-like iris transillumination defects were evident in both eyes, and posterior segment findings were unchanged, with only inferior RNFL thinning of the LE. IOP was RE 15 mmHg and LE 19 mmHg with Perkins tonometry at 5:15 pm, with central corneal thickness of 534 µm in each eye.

Given the patient's history and examination findings, a diagnosis of CLPC was made. The patient was advised to discontinue all contact lens wear for the time being. Due to her desire to resume lens wear for upcoming plans, a cautious approach to medical management was adopted. For allergy control, ketotifen 0.025% was prescribed twice daily in each eye. Given past steroid responsiveness and PDS, fluorometholone alcohol 0.1% (a low penetrance corticosteroid) was prescribed 4 times daily, and a follow-up was scheduled a week later.

The following week, ocular discomfort had subjectively improved, but conjunctival injection persisted. Central superficial punctate keratitis and papillae in the LE had worsened, resulting in 6/9 corrected acuity, while the RE remained stable. IOP was unchanged. The patient reported trialling lens wear once and reducing the corticosteroid dosage over concerns of another steroid response. Discussion regarding medication adherence and following the prescribed treatment plan was well received, and a trial of daily disposable contact lenses was provided for short periods of wear during her holiday instead of the existing monthly lenses. →

Two weeks later all signs and symptoms had improved, with a return to 6/6 acuity in each eye. Both corneas were clear and palpebral hyperaemia had mostly resolved. The cobblestone papillae reduced to grade 3 in each eye and IOP remained stable with no evidence of steroid responsiveness. Six weeks after the initial presentation, steroid tapering was commenced, reducing to 3 drops per day for 1 week, 2 drops for another week, then 1 drop for another week, and finally a drop every second day in the final week. Weekly IOP checks were undertaken before altering the taper and complete resolution of the CLPC was seen, with no signs of steroid response. New SiHy monthly replacement lenses were fitted for financial reasons, and the storage solution was changed to hydrogen peroxide with digital cleaning before and after storage advised to reduce lens bio-deposition.

Discussion

This case report highlights the complexities of managing CLPC, particularly in patients with coexisting ocular conditions. The patient's history of steroid responsiveness and PDS presented a unique challenge.

Unfortunately, CLPC pathophysiology is still debated as it is not a true hypersensitivity reaction, meaning there are no set management guidelines.[3,6] The key to managing CLPC is removing the irritating factor: the contact lenses. When lens wear continues, providing a compromise like silicone hydrogel daily disposable lenses may enhance the overall outcome.[7] Fitting these lenses long term can address key risk factors for CLPC development such as lens material, inadequate replacement schedules, and poor lens hygiene by providing enhanced oxygen transmissibility, lower lens protein deposition and simplifying the hygiene requirements.[4,7,8] If the patient, such as with Mrs J, requires less frequent replacement schedules, a change in lens care routine is warranted. After much discussion, changing the regular multipurpose disinfecting solution to a hydrogen peroxide cleaning solution, combined with more frequent digital cleaning, was agreed upon to assist in removing lens protein deposits and reducing ocular inflammation.[4,9]

In addition to lens care, pharmaceutical management of the concomitant atopy needs consideration each allergy season.[1] While dual antihistamine and mast cell stabiliser drops are the consensus for allergy control, corticosteroids should be considered with CLPC. As corticosteroids come with side effects, such as steroid-induced IOP changes, patient-specific risk factors must be taken into consideration, as long-term therapy of many months is typical.[5,10]

While no studies could be found to determine the risk for Mrs J, increased pigment deposition from PDS and the structural changes to the trabecular meshwork that are thought to produce steroid-induced ocular hypertension had the potential to reduce aqueous outflow enough to increase IOP significantly and irreversibly damage the already thin RNFL.[11] The lower penetrance of fluorometholone alcohol 0.1% reduced penetrance into the anterior chamber, minimising the risk of steroid-induced ocular hypertension while providing sufficient therapeutic potency to control the inflammatory processes of the CLPC effectively.[10]

In the future, the availability of various therapeutic options, like loteprednol, will likely change this area again and revolutionise how we treat ocular allergy and hypersensitivity. Loteprednol has been proven to reduce inflammation effectively while decreasing the impact on IOP.[12] In the Australian setting, discontinuing fluorometholone acetate 0.1% leaves a gap in our therapeutic options. It reminds optometrists to consider patient-specific factors when considering the potency and penetrance of ocular therapeutics. ●

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