Discussion

Peters anomaly is a congenital central corneal opacity that causes a sensory deprivation amblyopia. Until now, surgery has been limited to penetrating keratoplasty with a final visual acuity of worse than 6/60 in 38% of patients.[2] This case report adds a new less invasive treatment option for patients with mild type 1 disease.

Shah et al.[4] first described descemetorhexis without endothelial keratoplasty in 2012 for a patient with Fuchs endothelial corneal and posterior polymorphous corneal dystrophy, with larger series on descemetorhexis without endothelial keratoplasty published subsequently.[5–7] This procedure is based on the premise that there is an abnormal area of endothelium/Descemet's membrane, which if removed becomes replaced with normal peripheral endothelium. In Peters anomaly the posterior endothelium and Descemet's membrane are attenuated or absent with posterior stromal opacity.[3] In mild cases it would make sense that there could be an improvement if the abnormal endothelium/Descemet's membrane is removed and the normal peripheral endothelium migrates to cover the defect. The caveat being that the stromal opacity is not too dense. This can be difficult to assess clinically; however, OCT allows us to assess the depth of the opacity with greater ease. Indeed, Soh and Mehta[8] recently described the selective removal of only endothelium in mild type 1 Peters anomaly with improvement of visual acuity from 20/960 to 20/30. In their case, OCT did not show the thickening of Descemet's membrane as seen in our case, highlighting the preoperative benefit of OCT.

Ni et al.[3] reported a "multiple-layer" Descemet's membrane on histopathology in a patient who underwent a penetrating keratoplasty for type 1 Peters anomaly. This was present at the edge of the corneal opacity. This is the only case report documenting this finding, but it would be interesting to know if this is the main cause of the opacity in mild disease. Being able to identify this on OCT would allow the clinician to plan their surgery and allow for improved prognostication. Our patient showed significant thickening of the Descemet's membrane on OCT, rather than the typical attenuation (Fig. 1). This could explain the improvement in visual acuity from 6/36 to 6/20 with only a descemetorhexis.

Nodules were noted at the superior border of the rhexis. This has also been noted in the series by Moloney et al.[5] who hypothesized a traumatic reaction to the Descemet's stripping. In our patient the nodules disappeared over time with only pigment remaining on the endothelium at the last follow-up, 16 weeks after surgery.

Peters anomaly causes a sensory deprivation amblyopia from a central corneal opacity. The opacity is caused by the changes in the endothelium, Descemet's membrane, and posterior stroma. Until now, surgery has been limited to penetrating keratoplasty; however, we report on the success of primary descemetorhexis in a patient with mild type 1 Peters anomaly. Visual recovery is limited by both the amblyopia and residual corneal opacity; therefore, early surgery may result in better visual outcomes. Careful patient selection is recommended and further clinical validation is necessary.

Disclosure: The authors have no proprietary or commercial interest in any materials discussed in this article.

Roland Höllhumer,[*,†] **Dirk Booysen**[‡]
[*]University of the Witwatersrand, Johannesburg, South Africa;
[†]The Cornea Foundation, Johannesburg, South Africa;
[‡]University of Johannesburg, Johannesburg, South Africa

Correspondence to:
Dr. R. Höllhumer; hollhumer@gmail.com.

References

1. Bhandari R, Ferri S, Whittaker B, Liu M, Lazzaro DR. Peters anomaly: review of the literature. *Cornea*. 2011;30:939–44.
2. Zaidman GW, Flanagan JK, Furey CC. Long-term visual prognosis in children after corneal transplant surgery for Peters anomaly type I. *Am J Ophthalmol*. 2007;144:104–8.
3. Ni W, Wang W, Hong J, Zhang P, Liu C. A novel histopathologic finding in the Descemet's membrane of a patient with Peters anomaly: a case-report and literature review. *BMC Ophthalmol*. 2015;15:139.
4. Shah RD, Randleman JB, Grossniklaus HE. Spontaneous corneal clearing after Descemet's stripping without endothelial replacement. *Ophthalmology*. 2012;119:256–60.
5. Moloney G, Petsoglou C, Ball M, et al. Descemetorhexis without grafting for Fuchs endothelial dystrophy–supplementation with topical ripasudil. *Cornea*. 2017;36:642–8.
6. Iovieno A, Neri A, Soldani AM, Adani C, Fontana L. Descemetorhexis without graft placement for the treatment of Fuchs endothelial dystrophy: preliminary results and review of the literature. *Cornea*. 2017;36:637–41.
7. Borkar DS, Veldman P, Colby KA. Treatment of Fuchs endothelial dystrophy by Descemet stripping without endothelial keratoplasty. *Cornea*. 2016;35:1267–73.
8. Soh YQ, Mehta JS. Selective endothelial removal for Peters anomaly. *Cornea*. 2018;37:382–5.

Can J Ophthalmol 2019;54:e52–e54
0008-4182/17/$-see front matter
https://doi.org/10.1016/j.jcjo.2018.05.014

Simple limbal epithelial transplantation to treat recurring kissing pterygium

Pterygium is a common ocular surface disorder characterized by chronic proliferative conjunctival fibrovascular tissue growth over the cornea.[1] Numerous pterygium excision techniques have been tried with a wide range of recurrence rates, including bare sclera excision (30%–70%),[2] amniotic membrane (6.4%–42.3%), and conjunctival autograft with or without glue (0%–16.7%).[3,4] A recent Cochrane study found a 47% reduced risk of recurrence with conjunctival autograft at 6 months compared with amniotic membrane for

pterygium excision.[5] The use of fibrin tissue adhesive in comparison to sutures further reduces recurrence rates.[6] In cases of recurrent pterygium, adjunctive use of mitomycin C has been used with positive results (3.5% recurrence rate),[7] When recurrences do happen, there is a 50% chance the recurrence is detected within the first 4 months postoperatively and a 97% chance within the first year.[8]

Recent studies have suggested that limbal stem cell deficiency (LSCD) can contribute to the corneal conjunctivalization seen in pterygium.[9] As such, replenishing the local stem cell population is a logical approach to manage pterygium. Simple limbal epithelial transplantation (SLET) is a novel surgery to address unilateral LSCD[10] that has been used in cases of chemical burn and ocular surface squamous neoplasia. It has also been described as a technique in the treatment of primary pterygium excision.[11]

In this article, we describe using SLET to treat an aggressive case of recurrent, large, kissing pterygiums. A pterygium excision was performed and intraoperative mitomycin C was used to reduce recurrence.[7] Amniotic membrane and limbal stem cells were placed on the denuded cornea. Our approach combined the antiangiogenic property of amniotic membrane, antifibroblast proliferation effect of mitomycin C, and autologous limbal epithelial transplantation to repopulate stem cells.

Case report

A 37-year-old male experienced LSCD in both eyes, more severe in the left than the right. The most likely etiology of the LSCD was severe atopy and chronic blepharitis. His left eye presented with a densely vascularized recurrent double-headed "kissing" pterygium that covered most of the cornea. The left cornea also showed signs of LSCD, such as scarring, late staining at 12 and 6 o'clock, extensive pannus, and an early tear breakup time. There was only a small island of relatively clear limbus at 1−2 o'clock, with less late staining, from which the autograft would be taken (Fig. 1). His right eye had a pterygium extending nasally (2.5 × 3.5 mm) as well as a mild 360 degree pannus.

The patient's past ocular history included removal of a large double-headed pterygium from the left eye in 2011. The primary surgery involved pterygium excision with conjunctival autograft and the use of mitomycin C 0.02%. Preoperative visual acuity in the left eye was counting fingers at 1 foot. The patient agreed to undergo repeat pterygium excision as long as his right eye, which had a visual acuity of 20/30, was not involved in the surgery.

A retrobulbar anesthetic injection of 4 cc (2 cc Marcaine and 2 cc xylocaine 2% without epinephrine) was administered to the patient, as well as a regional Van Lint block (4 cc). The eye was then prepared and draped in the usual sterile fashion. A wire lid speculum was placed. A superficial peripheral lamellar dissection to isolate an area of relatively normal limbal stem cells (LSCs) was performed at the 1−2 o'clock region of the left eye. These LSCs were preserved in a Petri dish with balanced salt solution and covered in viscoelastic. Next, a 360 degree conjunctival peritomy was performed. A WECK-CEL (Beaver-Visitec International, Waltham, Mass.) was then cut into 4 pieces and soaked in mitomycin C 0.02%. The wedges of mitomycin C-soaked WECK-CEL were placed in the 4 quadrants and left in place for 3 minutes. The ocular surface was irrigated with 2 bottles of balanced salt solution to ensure no mitomycin C remained on the eye. Next, the pterygium was carefully demarcated and dissected to obtain a good plane, and the excess scar tissue was removed. Amniotic membrane was subsequently placed over the de-epithelialized cornea and bare sclera and tucked under the conjunctiva. The amniotic membrane was held in place by 4 interrupted 10-0 Vicryl sutures and tissue glue. The donor LSCs were cut into 12 pieces and placed on the corneal surface paracentrally; tissue glue was applied to secure the LSC pieces in place. A second piece of amniotic membrane was then superimposed to cover the donor LSCs, fixed in position with tissue glue. Lastly, a 16 mm diameter Kontur bandage lens (Kontur Kontact Lens Co, Hercules, Calif.) was placed over the operated eye.

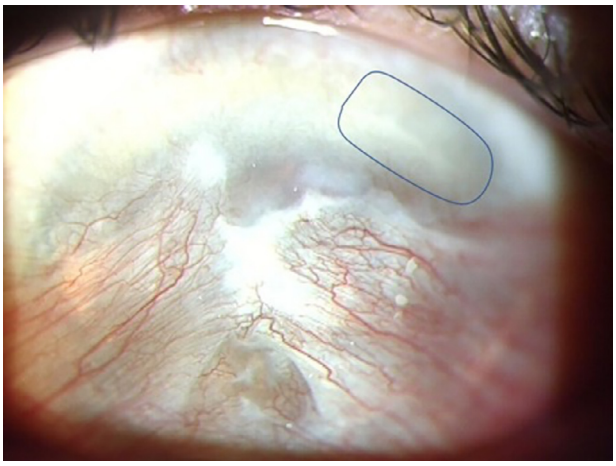

Fig. 1—Left eye with massive double-headed kissing recurrent pterygium. There is marked corneal scarring with late staining at 12 and 6 o'clock and a small island of relatively normal appearing limbus at 1–2 o'clock (delineated area).

The left eye's visual acuity subsequently improved to 20/200 at 1 month post-SLET, to $20/80^{-1}$ at 2.5 months and then to $20/50^{-3}$ (pin hole: $20/40^{-1}$) at 4 months. At his 8-month postoperative visit, his best corrected visual acuity was $20/40^{-2}$. The cornea was fully epithelialized, avascular, and with no signs suggestive of recurrence. A small central corneal scar was observed (Fig. 2).

Discussion

The current benchmark for treatment of primary or recurrent pterygium is excision with conjunctival autograft. If the defect is too large to be covered by an autograft, then amniotic membrane is used instead. The pterygium could be excised in

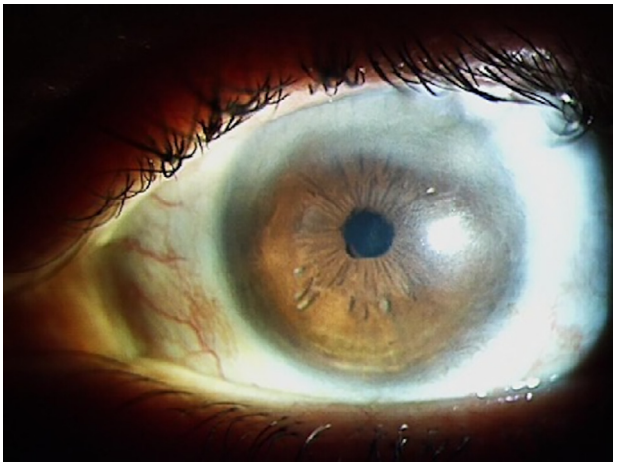

Fig. 2—Postoperative SLET 6 months: the surface was completely epithelialized, stable, avascular, and with no sign of recurrence of the pterygium.

a stepwise manner—removing the pterygium 1 side at a time and waiting a minimum of 3 months apart. This allows the conjunctiva to heal and gives the surgeon the chance to harvest the same region of conjunctiva for a second time. Mitomycin C is often used as an adjuvant therapy to further reduce the rate of recurrence.

In the present case, SLET was thought to be an appropriate alternative because (*i*) the patient experienced a recurrent, substantial pterygium that was resistant to the conventional conjunctival autograft and mitomycin C, and (*ii*) due to the lack of healthy conjunctiva available for an autograft. Since pterygium has been well characterized as a local stem cell disease,[12] re-establishing an LSC population is a logical step to preclude conjunctival invasion. Accordingly, SLET has been described as a novel technique to manage LSCD, including unilateral corneal burns, ocular surface squamous neoplasia, and pterygium.[10] Hernández-Bogantes et al. applied SLET to treat primary pterygium; among the 9 cases described, none showed recurrence at 8 months.[11]

In this case, the patient sustained bilateral pterygium with greater severity in the left eye. Obtaining donor tissue from the right eye was not an option as this might have exacerbated the existing LSCD in this better-seeing eye. Hence, we decided to obtain LSCs from a small, healthy limbal island on the left eye (Fig. 1). The success of this case highlighted the possibility of addressing treatment-resistant pterygia by restoring local stem cells via transplantation of a small amount of healthy tissue from the ipsilateral eye. Previous studies have shown that median time to pterygium recurrence after pterygium excision and conjunctival autograft is 2.64 months without mitomycin C and 3.70 months with mitomycin C.[3] In cases of recurrent neovascularization, supplemental subconjunctival Avastin injections could be used.

Looking forward, SLET represents a promising method to address extensive and recurrent pterygiums, but studies with longer follow-up periods and larger sample sizes are needed. Compared with keratolimbal allografting, the salient advantage of SLET lies in its tissue efficiency; only a minute piece of donor tissue is required, thereby reducing the risk of iatrogenic limbal stem cell deficiency. This is particularly advantageous in cases when the surgeon and patient are hesitant to operate on the better-seeing eye. Furthermore, the need and the associated adverse effects for systemic immunosuppression are obviated with SLET, as opposed to keratolimbal allografting.

In the future, the success rate of SLET might be further optimized when surgeons are able to identify distinct limbal tissues rich in healthy LSCs. An innovative optical coherence tomography device that accurately visualizes healthy LSC islands is currently being investigated by our group.

Disclosure: The authors have no proprietary or commercial interest in any materials discussed in this article.

Tanguy Boutin,* Zale Mednick,*
Tianwei Ellen Zhou, PhD,† Mahmood Showail,*
Adi Einan-Lifshitz,* Nir Sorkin,* Allan R. Slomovic*
*Department of Ophthalmology and Vision Sciences, University of Toronto, Toronto, Ont.; †Department of Medicine, McGill University, Montreal, Que.

Correspondence to:
Tanguy Boutin, MD; boutin.tanguy@gmail.com.

References

1. Chui J, Di Girolamo N, Wakefield D, Coroneo MT. The pathogenesis of pterygium: current concepts and their therapeutic implications. *Ocul Surf*. 2008;6:24–43.
2. Youngson RM. Recurrence of pterygium after excision. *Br J Ophthalmol*. 1972;56:120–5.
3. Fernandes M, Sangwan VS, Bansal AK, et al. Outcome of pterygium surgery: analysis over 14 years. *Eye (Lond)*. 2005;19:1182–90.
4. Bahar I, Kaiserman I, Weisbrod M, McAllum P, Slomovic A. Extensive versus limited pterygium excision with conjunctival autograft: outcomes and recurrence rates. *Curr Eye Res*. 2008;33:435–40.
5. Clearfield E, Hawkins BS, Kuo IC. Conjunctival autograft versus amniotic membrane transplantation for treatment of pterygium: findings from a Cochrane Systematic Review. *Am J Ophthalmol*. 2017;182:8–17.
6. Romano V, Cruciani M, Conti L, Fontana L. Fibrin glue versus sutures for conjunctival autografting in primary pterygium surgery. *Cochrane Database Syst Rev*. 2016;12:CD011308.
7. Shehadeh-Mashor R, Srinivasan S, Boimer C, Lee K, Tomkins O, Slomovic AR. Management of recurrent pterygium with intraoperative mitomycin C and conjunctival autograft with fibrin glue. *Am J Ophthalmol*. 2011;152:730–2.
8. Hirst LW, Sebban A, Chant D. Pterygium recurrence time. *Ophthalmology*. 1994;101:755–8.
9. Ahmad S. Concise review: limbal stem cell deficiency, dysfunction, and distress. *Stem Cells Transl Med*. 2012;1:110–5.
10. Sangwan VS, Basu S, MacNeil S, Balasubramanian D. Simple limbal epithelial transplantation (SLET): a novel surgical technique for the treatment of unilateral limbal stem cell deficiency. *Br J Ophthalmol*. 2012;96:931–4.

11. Hernández-Bogantes E, Amescua G, Navas A, et al. Minor ipsilateral simple limbal epithelial transplantation (mini-SLET) for pterygium treatment. *Br J Ophthalmol*. 2015;99:1598–600.
12. Tseng SC. Staging of conjunctival squamous metaplasia by impression cytology. *Ophthalmology*. 1985;92:728–33.

Can J Ophthalmol 2019;54:e54–e57
0008-4182/17/$-see front matter
https://doi.org/10.1016/j.jcjo.2018.06.003

Irreversibility of transverse venous sinus stenosis and optic nerve edema post-lumbar puncture in idiopathic intracranial hypertension

A 34-year-old woman was referred to our centre with a complaint of daily headaches, pulsatile tinnitus, and bilateral optic nerve oedema on ophthalmological exam. She had history of hormonal therapy use for endometriosis and a recent weight gain of 30 pounds, with her current weight being 230 pounds, and a body mass index of 38.3 kg/m^2. She had no visual complaints. Her vitals were stable. Corrected visual acuity was 20/20 in both eyes. Confrontation visual fields and colour vision were normal. Her pupillary and extraocular exam revealed no abnormalities. Ocular examination revealed normal anterior segments and optic nerve head swelling in each eye (Fig. 1). A screening neurological exam was unremarkable.

She underwent a magnetic resonance imaging (MRI) scan of her brain, which was normal, but a venogram showed bilateral distal transverse sinus stenosis with a combined conduit score[1] of 3 (Fig. 2A). Following this, fluoroscopic-guided lumbar puncture (LP) revealed an opening pressure of 30 cm H_2O and a closing pressure of 14 cm H_2O. Cerebrospinal fluid (CSF) analysis was normal. She was diagnosed with idiopathic intracranial hypertension (IIH) based on the Modified Dandy Criteria.[2] MRI brain and venogram performed immediately after LP showed persistent stenosis of the distal transverse sinuses bilaterally, and the combined conduit score remained at 3 (Fig. 2B). In addition, peripapillary optic nerve optic coherence tomography performed 2 hours preceding LP, 2 hours following LP, and 5 days later showed persistent bilateral optic nerve edema. Optic nerve edema typically lasts for weeks following resolution of elevated intracranial pressure. Despite the immediate resolution of intracranial pressure, our patient's bilateral distal transverse venous stenosis persisted. At the writing of this report, this is the first case of IIH with immediate venous imaging after relief of intracranial pressure through LP.

Recent studies show that transverse venous stenosis is a common finding in IIH patients,[1,3] suggesting a potential relationship between these entities. Several studies have shown resolution or an increase in transverse venous sinus diameter[4] after the resolution of intracranial pressure with LP, suggesting that the venous stenosis may have been the result of increased intracranial pressure. The theory is that the elevated intracranial pressure stretches the dura and begins to occlude the structurally susceptible transverse venous sinuses of individuals with IIH.[4] This causes obstruction in venous outflow and exacerbates intracranial pressure through reduced CSF reabsorption. CSF removal through cervical puncture, ventriculoperitoneal shunt, and lumboperitoneal shunts also appear to reverse the transverse venous sinus stenosis, lending support to this hypothesis.

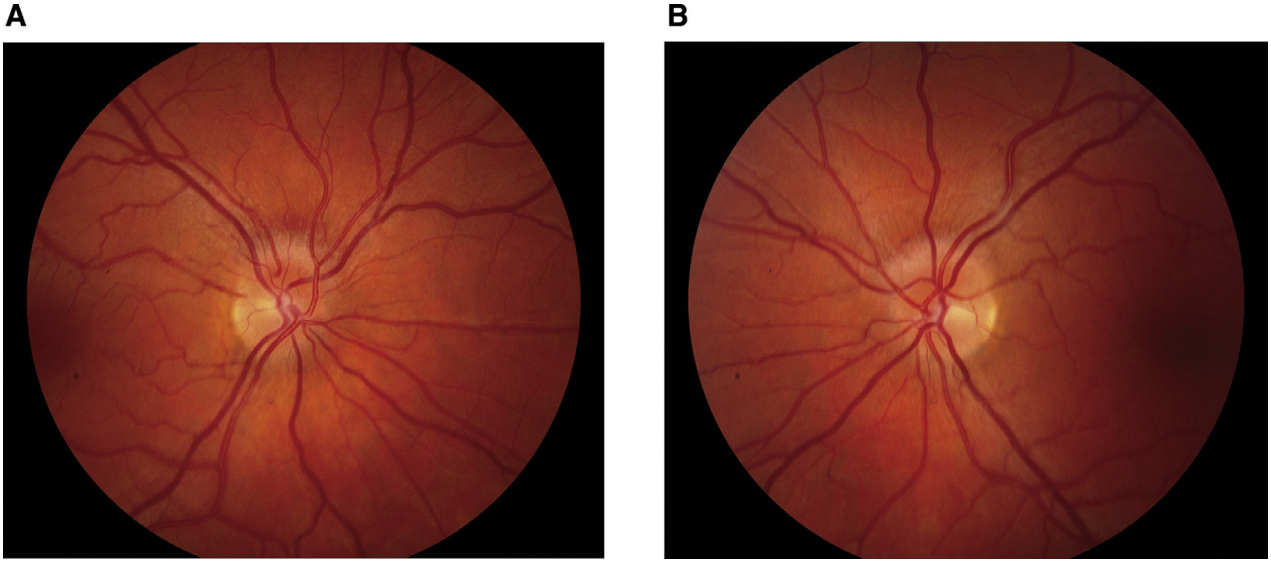

Fig. 1—A. Colour fundus photographs demonstrating optic disc edema in the right eye; B. Colour fundus photographs demonstrating optic disc edema in the left eye.