condition, depending on its severity. Early treatment proves favourable in restoring visual acuity to the patient; thus, although rare, this diagnosis should remain on the differential of transient vision loss.

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

Thomas A. Vo, BS, Amy Patel, MD, Chantal J. Boisvert, MD
Gavin Herbert Eye Institute, University of California–Irvine, Irvine, CA.

Correspondence to:
Chantal J. Boisvert, MD, Gavin Herbert Eye Institute, University of California–Irvine, 850 Health Sciences Road, Irvine, CA 92697; cboisver@uci.edu: cboisver@uci.edu

References

1. Stammberger HR, Kennedy DW. Paranasal sinuses: anatomic terminology and nomenclature. *Ann Otol Rhinol Laryngol (Suppl).* 1995;167:7-16.
2. Kitagawa K, Hayasaka S, Shimizu K, et al. Optic neuropathy produced by a compressed mucocele in an Onodi cell. *Am J Ophthalmol.* 2003;135:253-4.
3. Ogata Y, Okinaka Y, Takahashi M. Optic neuropathy caused by an isolated mucocele in an Onodi cell. *ORL J Otorhinolaryngol Relat Spec.* 1998;60:349-52.
4. Klink T, Pahnke J, Hoppe F, et al. Acute visual loss by an Onodi cell. *Br J Ophthalmol.* 2000;84:801-2.
5. Fukada H, Fukumitus R, Andoh M, et al. Small Onodi cell mucocele causing chronic optic neuropathy. *Neurol Med Chir (Tokyo).* 2010;50:953-5.

Can J Ophthalmol 2018;53:e117–e120

https://doi.org/10.1016/j.jcjo.2017.09.015

Conjunctival-limbal allografts in graft-versus-host disease using same HLA-identical bone marrow transplantation donor

Hematopoietic stem cell transplantation (HSCT) allows for the transplantation of multipotent hematopoietic stem cells for treatment of hematologic, immunologic, metabolic, and neoplastic diseases. Graft-versus-host disease (GVHD) is a unique complication of allogeneic HSCT in which the donor cells mount an immune response against the host cells.[1] Ocular GVHD affects 60%–90% of patients with chronic systemic GVHD.[2]

Limbal stem cell deficiency (LSCD) is an uncommon complication of GVHD.[3–6] The pathogenesis of GVHD-associated LSCD is largely unknown but may be related to alloreactivity to recipient tissues or repetitive frictional microtrauma to the limbal stem cells in an already inflamed environment.[5,6] There are limited reports regarding the surgical management of LSCD in GVHD.[5,6] We report our ocular surface stem cell transplantation (OSST) experience with living-related conjunctival limbal allografts (lr-CLAL) in 2 GVHD patients. The details of this procedure have been described previously, and the surgical technique implemented here does not differ significantly.[7]

Case 1

A 41-year-old female with a history of HSCT for chronic myelogenous leukemia (CML) 8 years prior and subsequent GVHD (biopsy positive) presented with decreased vision and pain from recurrent erosions/persistent epithelial defects. Ocular history included prior bilateral herpes simplex virus (HSV) keratitis and a 10-year history of soft contact lens wear (switched to Boston scleral contact lens). Her best-corrected visual acuity (BCVA) was 20/125 OD and 20/20 OS. Both eyes demonstrated palpebral conjunctival subepithelial fibrosis. There was superior conjunctivalization involving 7 (Fig. 1A) and 4 clock hours with subepithelial haze extending to the visual axis OD and OS, respectively. Despite starting topical vitamin A ointment, loteprednol, and cyclosporine OD, there was progression of the conjunctivalization, and BCVA worsened to 20/300 OD. The decision was made to perform a lr-CLAL OD. Because the patient's brother was an HLA-identical match, he served as the lr-CLAL donor.

Postoperatively, the patient was placed on prophylactic oral amoxicillin and acyclovir as well as topical prednisolone acetate, cyclosporine, and moxifloxacin. Oral cyclosporine (for GVHD) was tapered 1 month after surgery, and the topical corticosteroid and cyclosporine were slowly tapered. She attained 20/20 BCVA OD within 4 months after the lr-CLAL. Secondary to increased conjunctivalization and more frequent erosions OS 4 years after her first lr-CLAL, the decision was made to perform an lr-CLAL OS, which yielded successful epithelization. On last follow-up, over 7 years after her first lr-CLAL, both eyes maintained a clear, stable ocular surface (Fig. 1B–D) with 20/20 BCVA without topical or systemic immunosuppression (SI). She has continued topical artificial tears and oral acyclovir 400 mg BID for HSV prophylaxis.

Case 2

A 48-year-old male with a history of HSCT for CML 16 years prior (repeat HSCT 5 years after) and subsequent GVHD presented for decreased vision OS. Ocular history was also significant for severe dry eyes, soft contact lens

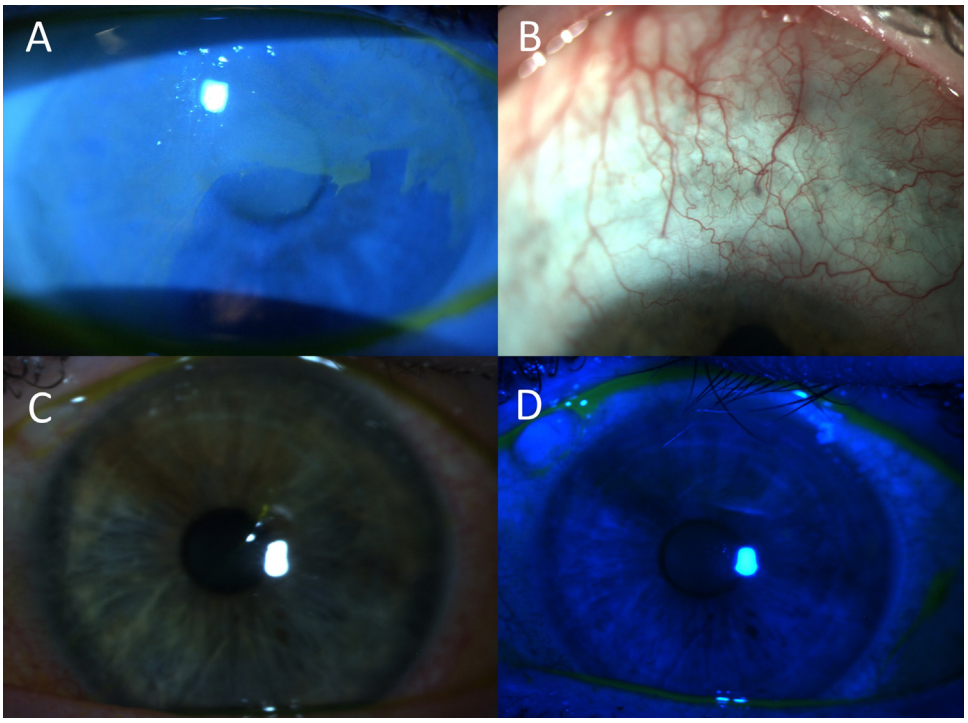

Fig. 1—Representative preoperative slit-lamp photograph of case 1 demonstrating conjunctivalization and significant late fluorescein staining involving the visual axis (A). Postoperative slit lamp from case 1 demonstrating the healed superior living-related conjunctival limbal allograft (lr-CLAL) site (B), a stable corneal surface (C), and no late fluorescein staining (D) >6.5 years after surgery.

wear, and prior cataract surgery OU. He had been maintained on chronic prednisone 2.5 mg daily. BCVA was 20/60 OD and 20/50 OS. Both eyes demonstrated symblephara, OS worse than OD. There was 360° conjunctivalization OS encroaching the visual axis. OD demonstrated mild peripheral conjunctivalization with diffuse punctate staining. The patient was started on topical vitamin A ointment and loteprednol OS; contact lens wear was also stopped OS. On follow-up 1 month later, BCVA OS decreased to 20/200 from conjunctivalization now involving the visual axis. The decision was made to perform an lr-CLAL using the patient's HSCT donor (sister), who was an HLA-identical match, as the lr-CLAL donor. Postoperatively, the patient was placed on topical prednisolone acetate and moxifloxacin (discontinued after 3 weeks). Oral prednisone was increased to 20 mg daily postoperatively and tapered back to the baseline dose over 5 months. On the last follow-up, 8.5 years after OSST, his BCVA OS was 20/40 with mild corneal scarring and a stable ocular surface. His dry eyes have required frequent administration of preservative-free artificial tears and gel drops and a recent trial of lifitegrast (poor response to topical cyclosporine). He is not on any SI medications.

Discussion

Although keratoconjunctivitis sicca and cicatricial conjunctivitis are common manifestations, LSCD is rare in chronic ocular GVHD.[5,6] Because both of our patients had a history of contact lens wear, it is possible that the etiology of LSCD was multifactorial, with ocular GVHD contributing a significant role. Sivaraman et al. hypothesized that superior limbic keratoconjunctivitis (SLK)-like inflammation may play an intermediary step, leading to frank LSCD in GVHD.[6] In their study of 26 GVHD eyes with SLK-like inflammation, all eyes demonstrated superior LSCD with superior corneal epithelial staining in a wave/whorl-like pattern, whereas 3 eyes developed frank LSCD.[6] Impression cytology of the whorl-like staining area without frank conjunctivalization confirmed goblet cells, suggesting that subclinical LSCD may exist in such patients.[6] Both eyes from our first case demonstrated LSCD starting superiorly.

Reports of the surgical management of GVHD-related LSCD have included superficial keratectomy with amniotic membrane transplantation,[6] allogeneic cultivated corneal epithelial transplantation (requiring betamethasone, cyclophosphamide, and cyclosporine SI),[4] and modified cultivated limbal epithelium transplantation (CLET) using a HLA-identical living-related allogeneic donor (same HSCT donor without SI).[5] This last report was a single case with shorter follow-up than our 3 eyes and developed corneal thinning that led to perforation (treated with a tectonic keratoplasty).[5]

The kidney transplantation literature has demonstrated success in inducing allograft tolerance (stable allograft function in the absence of any immunosuppression) by

combining HSCT with subsequent kidney transplantation using the same HLA-matched donor.[8,9] After HSCT, the immune system reconstituted by the HLA-identical donor's cells should not view other transplanted tissue as foreign because it is from the same donor. In our 3 eyes, long-term successful allogenic OSST without immunosuppression was achieved. Because SI could be avoided, the associated risks for the recipient were theoretically more akin to a conjunctival limbal autograft.

With long-term follow-up (>7 years), treatment with lr-CLAL has maintained a stable ocular surface. By providing conjunctival tissue with additional goblet cells, a lr-CLAL also helps to treat the GVHD-related keratoconjunctivitis sicca. This may be a benefit of using a lr-CLAL in these cases over a procedure such as CLET.

Although GVHD-related LSCD is uncommon, lr-CLAL can successfully restore the ocular surface. In the setting of GVHD-related LSCD after HSCT, there is a unique opportunity to provide a lr-CLAL from the same HLA-identical donor as the HSCT, allowing for true long-term allograft tolerance and avoidance of SI.

Disclosure: E.J.H. has consulted for Alcon Laboratories, Allergan, Bausch & Lomb, Kala Pharmaceuticals, Mati Pharmaceuticals, Omeros, PRN, Senju Pharmaceuticals, Shire, TearLab, and TearScience. A.Y.C., B.M.G., N.J.A., E.S., and A.G. do not have any disclosures to report.

Albert Y. Cheung, MD,[*,†] Brad M. Genereux, OD,[*] Nicholas J. Auteri, BS,[*] Enrica Sarnicola, MD,[*,‡] Amit Govil, MD,[§] Edward J. Holland, MD[*,†]

[*]Cincinnati Eye Institute, Cincinnati, OH; [†]Department of Ophthalmology, University of Cincinnati, Cincinnati, OH; [‡]Department of Medicine Surgery and Neuroscience, University of Siena, Siena, Italy; [§]Division of Nephrology and Hypertension, University of Cincinnati, Cincinnati, OH.

Correspondence to:
Edward J. Holland, MD, 580 South Loop Road, Suite 200, Edgewood, KY 41017; eholland@holprovision.com: eholland@holprovision.com

References

1. Ferrara JL, Levine JE, Reddy P, Holler E. Graft-versus-host disease. *Lancet*. 2009;373:1550-61.
2. Ple-plakon PA, Mian SI. Ocular graft-versus-host disease. In: Holland EJ, Mannis MJ, Lee WB, eds. *Ocular Surface Disease: Cornea, conjunctiva, and Tear Film.* New York: Elsevier; 2013, pp. 178-81.
3. Heinz C, Steuhl K-P, Meller D. Hornhautperforationen bei vitamin a-mangel. *Ophthalmologe*. 2004;101:614-7.
4. Nakamura T, Inatomi T, Sotozono C, et al. Transplantation of autologous serum-derived cultivated corneal epithelial equivalents for the treatment of severe ocular surface disease. *Ophthalmology*. 2006;113:1765-72.
5. Meller D, Fuchsluger T, Pauklin M, Steuhl KP. Ocular surface reconstruction in graft-versus-host disease with HLA-identical living-related allogeneic cultivated limbal epithelium after hematopoietic stem cell transplantation from the same donor. *Cornea*. 2009;28:233-6.
6. Sivaraman KR, Jivrajka RV, Soin K, et al. Superior limbic keratoconjunctivitis-like inflammation in patients with chronic graft-versus-host disease. *Ocul Surf*. 2016;14:393-400.
7. Holland EJ, Schwartz GS. The Paton lecture: ocular surface transplantation: 10 years' experience. *Cornea*. 2004;23:425-31.
8. Spitzer TR, Delmonico F, Tolkoff-Rubin N, et al. Combined histocompatibility leukocyte antigenmatched donor bone marrow and renal transplantation for multiple myeloma with end stage renal disease: the induction of allograft tolerance through mixed lympho-hematopoietic chimerism. *Transplantation*. 1999;68:480-4.
9. Scandling JD, Busque S, Dejbakhsh-Jones S, et al. Tolerance and chimerism after renal and hematopoietic-cell transplantation. *N Engl J Med*. 2008;358:362-8.

Can J Ophthalmol 2018;53:e120–e122
0008-4182/17/$-see front matter
https://doi.org/10.1016/j.jcjo.2017.09.004

Unusual ocular presentation in a patient with lichen planus

Case Report

Lichen planus is a relatively common mucocutaneous disorder of unknown etiology that is thought to be immunologically mediated. It is a chronic disorder characterized by periods of exacerbation and remission. Lichen planus has variable clinical manifestations, including the skin, oral mucosa, genital mucosa, nails, and scalp. The characteristic lesions have a well-established clinical appearance and histological features that help in establishing the diagnosis.[1] Ocular involvement may occur in patients with lichen planus. Although involvement of the eyelids, lacrimal ducts, conjunctiva, and cornea has been described in patients with lichen planus,[2] scleritis has not been reported in such patients. We report a case of scleritis diagnosed in a patient with lichen planus.

A 46-year-old female presented with redness, discomfort, and pain on ocular movement in the right eye. Review of systems revealed history of pruritic skin lesions of the elbows and knees. Ocular history revealed epiphora, for which she had irrigation of the nasolacrimal ducts, which revealed patent passages. She had been recently diagnosed with lichen planus by her dermatologist, for which she was given local steroid skin creams.

On eye examination, her uncorrected visual acuity was 20/25 OU. The Schirmer test without anaesthesia revealed 15 mm of wetting in the right eye and 13 mm of wetting in the left eye after 2 minutes. Slit-lamp biomicroscopy of the right eye revealed normal lids and conjunctiva. The sclera was injected temporally and inferiorly, with marked tenderness; the cornea was clear; the anterior chamber was quiet; the pupil was