A case report: Keratoconus and thyroid eye disease

Abstract

This paper describes the selection and design of scleral contact lens parameters and the lens fitting process based on the unique physical features of a patient presenting with both keratoconus and thyroid eye disease.

Case History and Examination

In August 2010, MP, a 34-year-old Asian American female was examined in the Stein Family Cornea and Contact Lens Service of the Eye Care Center in Fullerton, CA. She was pursuing a contact lens fitting for previously diagnosed keratoconus. Her chief complaint focused on a history of unsuccessful rigid gas permeable (GP) contact lens wear for the past several years. She also reported that her eyes are very dry and irritated. She recently lost her right rigid gas permeable corneal contact lens and has been functioning with her glasses. Significant ocular and medical history includes keratoconus in each eye, diagnosed fifteen years prior, and Grave’s Disease for over 10 years. Thyroid hormones are currently under good control with the use of Levothyroid medication at a low dosage.

MP presented with spectacle corrected visual acuities of 20/70 OD and OS. Physical examination revealed large bilateral palpebral fissure widths of both eyes, due to significant proptosis and lid retraction. The fissure widths were measured at 13 mm in the right eye and 12 mm in the left eye. Slit lamp examination revealed small and compressed superior and inferior fornices, dense stromal Vogt’s striae and apical scarring in both corneas, with the right eye worse than the left. At the time of examination, there was minimal inferior superficial punctate keratitis (SPK) suggestive of incomplete blinking or lagophthalmos due to her proptosis. The simulated keratometry readings from the Medmont Keratometer were unreadable in the right eye and the left eye measured at 58.00D x 71.40D@ 019. Manifest refraction further corrected her left eye to 20/40, while the right eye remained at 20/70.

Differential Diagnosis

Differential diagnoses for MP primarily included keratoconus or other keratoectasias, such as pellucid marginal degeneration or keratoglobus based on chief complaints. Thyroid eye disease was also a primary diagnosis based on physical appearance and previous diagnosis of hyperthyroidism. Other differential diagnoses, based on appearance, could include craniofacial abnormalities resulting in proptosis and lid retraction due to shallow orbits.

Diagnosis

A diagnosis of bilateral keratoconus was made based on the topography presentation, Vogt’s striae, apical scarring and a history of uncorrectable vision with glasses. Grave’s Disease was diagnosed by MP’s primary care physician, based on a laboratory work up suggestive of the condition. The protrusion of both eyes is consistent with the diagnosis of thyroid eye disease, also termed Grave’s ophthalmopathy.
Discussion

MP presents a unique case requiring careful management of two conditions, both equally challenging to any eye care provider. Both keratoconus and thyroid eye disease require thorough care of ocular health, comfort and vision.

Keratoconus

Keratoconus is a bilateral, progressive and asymmetric corneal degeneration leading to stromal thinning and eventual protrusion of the thinned cornea.\textsuperscript{1,2} As the most common primary corneal ectasia and it affects all ethnicities with a prevalence of about 5.4/10,000 people.\textsuperscript{1,3} There is recent debate over gender predilection, with recent studies finding males more affected than females. The condition typically presents in late puberty and later stabilizes around the fourth decade of life.\textsuperscript{1,2}

The corneal changes in keratoconus are responsible for increasingly blurry vision with and without spectacles, high myopia and irregular astigmatism.\textsuperscript{1} Early or mild stages of keratoconus present with increasing against-the-rule astigmatism and difficulty achieving best corrected spectacle acuities of 20/20.\textsuperscript{1} Later stages result in corneal thinning, iron deposits at the base of the cone (Fleischer’s ring), Vogt’s striae in the corneal stroma, and eventually apical scarring.\textsuperscript{1,2}

While the exact etiology of keratoconus is poorly understood, there are proposed theories. Advances in corneal topography support a genetic link to the condition. Prior to the widespread use of corneal topography, only 6% to 8% of patient had a close relative with keratoconus. The Collaborative Longitudinal Evaluation of Keratoconus Study (CLEK) found about 13% of patients with keratoconus have a close relative with the condition.\textsuperscript{4} The inheritance pattern has not been clearly defined, but autosomal dominance with variable expression\textsuperscript{1} or incomplete penetrance\textsuperscript{2} has been suggested. There are also physical differences between normal corneas and those with keratoconus. Keratoconic corneas have been found to have a different type of collagen in the corneal tissue, increased levels of critical enzymes and interleukin-I. The modified levels of these corneal components result in poor wound healing and an increase in keratocyte apoptosis.\textsuperscript{1}

Keratoconus can be classified into several groups: nipple cones, oval cones, keratoglobus and “D” shaped cones.\textsuperscript{1} Nipple cones are small areas of thinning, typically less than five millimeters in diameter. The cones are centrally or inferior nasally located, and typically contact lenses are very successful for this group. Oval cones are larger, greater than five millimeters in diameter and are located paracentrally or peripherally in the cornea. Fitting rigid contact lenses on corneas in this group is difficult, but generally successful. Keratoglobus cones present with corneal thinning in 75% or more of the cornea. Contact lenses are an extremely difficult fit in this case. Lastly, the “D” cones are a recently recognized group. These patients are those that typically present with keratoectasia post LASIK surgery.\textsuperscript{1}

Keratoconus can also be categorized into groups based on severity. There are several accepted methods when staging keratoconus. Ophthalmology journals generally categorize the severity of keratoconus based on the Amsler staging system, which classifies patients based on keratometry mire quality, corneal thinning and the presence of apical opacities. In optometry journals, the severity of keratoconus is based on the dioptic values measured by keratometry, as previously grouped by the CLEK study.
Based on slit lamp examination, topography and keratometry values of the left eye, MP could be classified as a severe oval cone. There is corneal scarring and dense Vogt’s striae in both eyes. The corneal apex of the left eye is located just inferior to the visual axis and extends peripherally to the temporal limbus, affecting a large area of the cornea.

Thyroid Eye Disease

Thyroid eye disease (TED) affects 25%-50% of patients with hyperthyroidism, which is also termed Grave’s Disease.²,⁵ For many patients, TED is only a cosmetic problem, but can be severe enough to cause blindness from exposure keratopathy or optic neuropathy.² Hyperthyroidism is an autoimmune condition, generally presenting in the third or fourth decade of life.² Usually affecting women more than men, signs of TED are eyelid retraction, proptosis of one or both eyes, periorbital edema and constriction of the extraocular muscles.²,⁵ Symptoms of the condition include dry eyes, diplopia and potential vision loss if the condition is severe enough to affect the optic nerve.²,⁵

Thyroid eye disease occurs alongside Grave’s disease, and is independent the severity of the hyperthyroidism.⁶ The process begins when lymphocytes infiltrate the extraocular muscles, and glucosaminoglycans accumulate in orbital fat.²,⁶ These processes lead to a swelling of the extraocular muscles and therefore pressing on the globe and optic nerve head.² The course of TED is independent of the severity of the hyperthyroidism. The extraocular muscles can swell up to eight times their normal size² and an increase in four milliliters of volume can produce six millimeters of proptosis.⁶

There are five major clinical manifestations of thyroid eye disease, and two developmental stages.⁶ The clinical manifestations seen are soft tissue involvement, lid retraction, proptosis, optic neuropathy and restrictive myopathy.² The disease course is also grouped into two stages: a congestive, inflammatory stage and an inactive, fibrotic stage. The active stage usually lasts about three years and the patient’s eyes will be red and irritated. After the active stage has passed, there may be a painless ocular motility abnormality, but otherwise the eyes are white and quiet.²

In MP’s case, the active stage has passed. She has proptosis, lid retraction, and dry eye, but no diplopia. She is currently taking a low dose of thyroid medication to maintain hormone levels.

Treatment and Management

After collecting clinical data, it was apparent that scleral lenses would be of great benefit for MP. Not only would the lenses provide her with optimal vision correction for severe keratoconus, they would also protect her corneas from any exposure keratopathy due to the proptosis from thyroid eye disease. Initially, a Jupiter Scleral rigid gas permeable contact lens with a diameter of 18.2 millimeters was selected for both eyes. The large overall diameter contact lenses were applied, but the compression on the lens by her inferior lid and orbital bone caused discomfort. She reported pinching and pressure between her eye and inferior orbit. The next lens diameter applied was the 15.6 millimeter overall diameter scleral lens. These medium sized gas permeable scleral lenses were chosen because the inferior and superior fornices of MP’s eye were very small and compressed due to her proptosis. After selecting base curves that successfully vaulted MP’s keratoconus, the scleral alignment was evaluated. The attempt was
unsuccessful because of MP’s large palpebral fissures. She was able to blink out the scleral lenses when changing gaze directions, especially in superior gaze. In order to successfully wear a scleral contact lens, the 18.2 millimeter overall diameter trial lens was revisited.

After several unsuccessful attempts at applying the larger scleral lens, the application technique was modified. The inferior edge of the lens was gently slid behind her lower lid, and then tucked deep into the inferior fornix. This allowed sufficient space for the superior edge of the lens to be applied to the sclera, with the upper lid pulled back. Her lids were then able to close over the entire contact lens. After several minutes of wear, she was comfortable wearing the larger diameter scleral lenses in both eyes. A fit evaluation and over refraction was performed and scleral lenses were ordered to provide a custom fit to MP’s eye. In order to provide and optimum fit, the parameter changes ordered (when compared to the trial lens from the fitting set) were as follows: steeping the third and fourth peripheral curves to increase comfort and decrease bubble formation in the corneal optic zone, and increasing the corneal chamber depth to allow sufficient limbal clearance.

At the first follow up visit, the new scleral lenses fit very well. In situ visual acuities were 20/60 OD, 20/50 OS and 20/30 OU. An over refraction yielded a small amount of plus power in the right eye, and a small amount of minus power in the left eye. On fit evaluation, minimal scleral vessel blanching was noted in the right eye and no blanching visible in the left eye. There was complete apical and limbal clearance of both lenses. MP reported excellent comfort of both lenses. New lenses were to be ordered with power changes to both lenses and a small amount of flattening to the third and fourth peripheral curves of the right lens. Application and removal of the scleral lenses was taught in office at the first follow up and she was able to wear the lenses home, with instruction to build up wearing time an hour each day, beginning with two hours of wear time on dispense day.

MP recently returned for her second follow up. She reported that she was able to wear the lenses with excellent comfort for ten to twelve hours per day. The lenses were easily applied and removed each day, with only occasional bubble formation in the optic zone. She noted that her vision became hazy after four to five hours of wear, but symptoms improved with the use of artificial tears. The new scleral lenses were applied in office. In situ visual acuities were 20/50 OD, 20/40 OS and 20/30 OU. Over refraction yielded less than a diopter of minus correction in both eyes, with best corrected acuities of 20/25 OD, 20/40 OS. Fit evaluation revealed slight apical touch on the right eye with complete limbal clearance and no peripheral blanching of the scleral vessels. The left lens completely vaulted the corneal apex and limbus, with no scleral blanching.

Treatment Discussion

The use of scleral contact lenses was initiated over a century ago by Kalt in 1888, and by Mueller and Fick around the same time period. Created from polymethylmethacrylate or glass, corneal edema was a major initial setback to scleral lens wearers, as the decreased oxygen levels reaching the cornea eventually forced these patients into other modes of visual correction. The original indication for scleral contact lenses was similar to those of modern scleral lenses; they were prescribed to protect the corneas of those patients with ocular surface diseases or improve vision in those with irregular astigmatism.

With the introduction of gas permeable materials in the 1980s, the use of scleral contact lenses were reintroduced to the cornea and contact lens arena. Corneal metabolism and
edema levels were shown to be of satisfactory levels while using the new materials, and the use scleral contact lenses was revived.\textsuperscript{9,11}

The principle advantages of scleral rigid gas permeable contact lenses are due to the large overall diameter. The large diameter lens is designed to completely vault the corneal surface, producing a fluid filled reservoir that masks corneal irregularities.\textsuperscript{9} This quality of scleral contact lenses is primarily beneficial for those patients with keratoconus, penetrating keratoplasties and other corneal ectasias. The reservoir also serves to promote corneal surface healing in cases of ocular surface disease, Steven-Johnson’s Syndrome, recurrent erosions and ocular cicatrical pemphigoid.\textsuperscript{10} Other advantages of the large overall diameter is initial patient comfort, and lack of lens dislodging.\textsuperscript{9}

Disadvantages of gas permeable scleral contact lenses are minimal. Some patients report a feeling of bulkiness, or eye bulging. Others are intimidated by the large size of the lens. Few eye care practitioners continue to be concerned about sufficient oxygen reaching the cornea. The average center thickness of a modern scleral contact lens is between 0.5 and 0.8 millimeters in thickness. This mass is needed to maintain optical stability and prevent lens flexure. Studies on corneal metabolism while wearing scleral contact lenses have shown that there is no difference in corneal swelling from hypoxia when the lens thickness was reduced from 0.6 millimeters to 0.15 millimeters. In fact, the corneal swelling remained at an acceptable level until the center lens thickness was increased to 1.2 millimeters, which far exceeds even a high plus powered lens.\textsuperscript{11} Other concerns regarding scleral lenses are similar to those of any contact lens modality, including conjunctival hyperemia, epithelial edema, stromal edema, corneal infiltrates and vascularization. In a study done by Visser, et al (2007), slit lamp examination indicated no significant anterior segment findings in 26.4% of eyes after at least 3 months of full time daily wear. The most common side effect of scleral contact lenses was conjunctival hyperemia, 48.2% of eyes.\textsuperscript{10}

Scleral lenses are not only providing a large number of patients with excellent corneal health, but excellent performance in visual acuity, as well. The majority of post keratoplasty patients are reaching acuities of 6/6 or 6/9, and those with primary corneal ectasias are achieving acuities of 6/9 or 6/12.\textsuperscript{9}

Modern scleral lenses are currently being used for a wide range of corneal conditions with excellent success in patient satisfaction and comfort.\textsuperscript{12} In a satisfaction survey given to new scleral contact lens wearers, patients rated their comfort, visual quality and overall satisfaction at 98% after switching from previous modalities of contact lenses.\textsuperscript{12}

**Conclusion and Take Home Points:**

Rigid gas permeable scleral contact lenses are a special tool that every corneal or contact lens specialist should learn to utilize. While each lens has many parameters to specify, this also allows for each lens to be completely customized to each patient. Special customizations that can be made for patients are countless. Examples could include toric back surface curvatures to increase stability, front surface toricity for residual astigmatism, or a lens “notch” to fit around scleral abnormalities like pterygia or post-surgical conjunctival blebs.\textsuperscript{12} The ability to completely customize a contact lens for a patient leads to patient loyalty and an excellent referral base.
Another key idea to remember about scleral lenses is that they are not just for refractive purposes. While most patients currently fit with scleral lenses are those with irregular astigmatism, there is a large population of corneal disease that could benefit from proper scleral contact lens wear. There are many safe and practical ways to use scleral lenses, and should not be reserved for only refractive purposes.

In conclusion, scleral contact lenses are safe for full time wear, even in those patients that have previously failed in contact lens wear attempts. These lenses provide excellent vision and comfort for the mass majority of patients in need of specialty contact lenses.

References


11. Pullum K, Buckley R. Therapeutic and Ocular Surface Indications for Scleral Contact