CASE REPORT

Vernal keratoconjunctivitis: culmination of management using immunosuppression, surgical and prosthetic therapy over quarter century

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SUMMARY

A 22-year-old male patient presented in 1988 with active vernal keratoconjunctivitis. He was treated with topical mast cell stabilisers and corticosteroids. Chronic inflammation despite topical treatment necessitated oral immunosuppressants. Active disease came under control with this; however, the patient gradually developed limbal stem cell deficiency. He underwent bilateral pannus resection with amniotic membrane transplantation that resulted in improved ocular surface. In 2007, patient was found to have significant bilateral posterior subcapsular cataracts and underwent bilateral cataract surgery with intraocular lens implantation with good visual outcome. In 2016, he was provided with scleral lens prosthetic device, which further improved vision. At last follow-up, more than 25 years after his initial visit, his visual acuity was 20/25 in both eyes with a stable surface. With a comprehensive approach using immunosuppression, surgical therapy and scleral lens prosthetic device, chronic vernal keratoconjunctivitis can be well managed as illustrated in this case.

treatment modification based on clinical presentation. This report describes long-term holistic management of VKC in a patient for more than a quarter century using a combination of medical, surgical and prosthetic therapy.

CASE PRESENTATION

A 22-year-old male patient presented in 1988 with itching, watering, redness and photophobia in both eyes, which were present since 10 years. The symptoms would subside on topical medication only to recur after discontinuation of medication. His unaided distant visual acuity was 20/20 (Snellen chart in feet) in both eyes. Slit lamp examination revealed multiple papillae in palpebral conjunctiva, gelatinous limbal thickening and hyperpigmentation and superficial punctate keratitis. Rest of the examination was normal. The patient was diagnosed with active VKC having mixed disease, predominantly limbal.

TREATMENT

The patient was treated with topical mast cell stabilisers (cromolyn sodium 2% eye drops), lubricants (carboxymethylcellulose sodium 0.5% eye drops), decongestants (naphazoline hydrochloride 0.05% eye drops) and corticosteroids in tapering doses (betamethasone 0.1% or prednisolone acetate 1% eye drops). The intraocular pressure (IOP) and optic disc cupping was regularly monitored. In 1994, patient had IOP elevation secondary to steroid response, recorded as 30 mm Hg and 40 mm Hg in the right eye and the left eyes, respectively. This was managed with timolol 0.5% eye drops twice daily for 2 months and tapering down of topical steroid drops dosage. Thereafter he maintained stable IOP and stable cup-disc ratio of 0.3 in both eyes until last follow-up. Over next 15 years, he was on continuous topical medication, the dosage being titrated based on clinical profile with addition of oral astemizole 10 mg/day for some duration. However, there was chronic inflammation with multiple exacerbations, hence systemic immunosuppressants (tablet prednisolone 40 mg/day gradually tapered to 5 mg/day and tablet methotrexate 7.5 mg weekly) were added in 2003 with regular monitoring of blood counts and systemic side effects. Active disease was controlled with this; however, the patient's visual acuity reduced due to corneal scarring and irregular astigmatism. There was loss of normal limbal architecture, peripheral corneal vascularisation and pannus

BACKGROUND

Allergic eye diseases are an entire spectrum of disorders affecting the conjunctiva, cornea and lids in different forms. Of them, vernal keratoconjunctivitis (VKC) is a florid condition characterised by papillae over tarsus or limbus, Horner-Trantas' spots, superficial punctate keratopathy and perilimbal pigmentary changes.¹ The various complications that can occur in VKC include corneal scarring, keratoconus, shield ulcers, cataract, glaucoma and limbal stem cell deficiency (LSCD).² This immunological disease usually subsides after puberty, but not in all cases. In patients where the disease persists after puberty, there is long-standing chronic inflammation which is likely to cause ocular surface damage.³

LSCD though rare, is the result of severe limbal disease occurring in VKC. Chronic limbal inflammation affects the microenvironment of stem cells due to direct damage caused by the toxic products of eosinophils and other inflammatory cells.⁴ Such changes in the ocular surface can cause permanent visual impairment.

Hence management of VKC involves both control of active inflammation and treatment of each of the associated complications with a combination of medical therapy, surgical therapy and/or visual prosthetic devices. When chronic in nature, it requires long-term follow-up of patients and

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formation suggestive of LSCD (figure 1A, B). The patient underwent pannus resection with amniotic membrane transplantation (AMT) in 2004 in both eyes.

OUTCOME AND FOLLOW-UP

This resulted in best corrected visual acuity (BCVA) of 20/50 in both eyes and there was improvement in ocular surface. On subsequent follow-ups, systemic immunosuppression was gradually discontinued as the disease was under control; the patient was continued on topical medication. In 2007, the patient reported with symptoms of gradual diminution of vision, with BCVA of 20/80 and counting fingers at 2 m in right and left eyes respectively, on examination. He had significant posterior subcapsular cataracts. He underwent phacoemulsification and intraocular lens implantation (left eye followed by right eye) after which the vision improved to 20/50 in both eyes (figure 2A, B). In 2016, he underwent scleral contact lens prosthetic device trial (prosthetic replacement of ocular surface ecosystem (PROSE) lens), which provided further visual clarity and comfort. At his last

Figure 1 Peripheral corneal vascularisation, superficial corneal scarring and pannus formation suggestive of limbal stem cell deficiency in right (A) and left eye (B) of patient with vernal keratoconjunctivitis.

Figure 2 Post amniotic membrane transplantation and phacoemulsification with intraocular lens implantation (PCIOL) improved corneal surface, reduction in vascularisation and PCIOL in place in right (A) and left eye (B) of patient.

Figure 3 At last visit, scleral contact lens prosthetic device in place (prosthetic replacement of ocular surface ecosystem, PROSE), improving vision to 20/25 in right (A) and left eye (B).

visit, BCVA with PROSE lenses was 20/25 in both eyes with a stable surface (figure 3A, B). The patient was continued on once daily fluorometholone 0.1% eye drops, tacrolimus 0.03% eye ointment at bedtime and lubricants. Figure 4A, B show the patient's corneal topography scans performed in 2016.

DISCUSSION

This report on VKC features successful management of active disease as well as its sequelae over 25 years. With a comprehensive approach using immunosuppression, surgical and prosthetic therapy this severe disease can be managed well.

Limbal form of VKC that is more common in tropical countries is characterised by gelatinous limbal thickening, Horner-Trantas dots and perilimbal pigmentation in its active form. It requires prompt management in order to reduce symptoms as well as prevent irreversible sight-threatening damage to the limbal microenvironment. For active disease, as a first line of therapy topical medications are recommended. While antihistamines mast cell stabilisers and non-steroidal anti-inflammatory drugs have a role in mild cases, topical steroids are the most effective treatment for moderate-to-severe forms of VKC.⁵ Immunomodulators such as ciclosporin A and tacrolimus are other established alternatives in the treatment of VKC. Ciclosporin has been recommended in grade 3 and grade 4 VKC in conjunction with topical steroids and has also been proved effective in preventing seasonal recurrences.^{6 7} In refractory cases, systemic therapy is required. The use of oral steroids is restricted because of their systemic and ocular side effects, but severe cases warrant their use under regular monitoring.⁶

Topical decongestants and steroids (topical as well as systemic) can cause spikes in IOPs secondarily, which can lead to progressive glaucoma if not recognised and managed promptly. Other common ocular complications of long-term steroid use include cataract and secondary ocular infections.⁶ Long-term systemic steroid use can cause serious side effects like weight gain, muscle weakness, susceptibility to infections, osteoporosis, etc, and these need to be regularly watched out for. In case manageable side effects develop, it is preferable to continue steroid usage rather than lose eye sight to severe VKC. In the case of our patient, systemic immunosuppressants were given when topical therapy was inadequate to control the disease. Later they were gradually discontinued when the disease was under control. The cataract that probably developed secondary to steroid use was managed well with cataract surgery. There were no other side effects in our patient.

Long-term complications of VKC such as LSCD, corneal scarring, cataract, glaucoma, etc, may require surgical management. LSCD is a rare but sight-threatening complication of chronic VKC. To prevent visual deterioration secondary to corneal

Figure 4 Oculyzer scans of right (A) and left eye (B) showing varied corneal curvature, elevation and thickness at different points consistent with irregular corneas secondary to limbal stem cell deficiency and surface surgery (pannus resection with amniotic membrane grafting).

conjunctivalisation and scarring, AMT and limbal stem cell transplantation (LSCT) are used to manage LSCD.³ Amniotic membrane suppresses conjunctival fibrosis and inflammation, promotes corneal epithelialisation and differentiation.⁸ Several studies have shown that in eyes with partial LSCD, AMT promotes expansion of the remaining limbal epithelial stem cells thus resulting in improved ocular surface.^{9–11} An added advantage is that it is simple and it avoids the use of allogenic LSCT as a treatment for bilateral LSCD, that necessitates strong and at times unaffordable systemic immunosuppression for lifetime. AMT was performed in our patient with this in view and it resulted in a good outcome.

In VKC, associated irregular astigmatism reduces visual acuity necessitating wearing of contact lens. The fitting of regular corneal contact lenses is dependent on factors such as presence of active disease, corneal scars, concomitant dry eye and LSCD.¹² These patients are often symptomatic after exposure to non-specific stimuli and may be intolerant to regular corneal contact lenses. PROSE, which is a fluid-filled scleral contact lens prosthetic device, masks the corneal surface astigmatism thus improving vision. It also provides comfort with maintenance of ocular surface health in LSCD cases by acting as a liquid corneal bandage.¹³ Our patient had remarkable improvement in vision after using PROSE lenses with reduction in photophobia and increased comfort.

The aim of VKC treatment should be to combat active disease, prevent irreversible complications as well as to visually rehabilitate patients. While an aggressive but monitored immunosuppression controls disease activity, timely surgery for the management of complications and optical prosthetics can greatly enhance patients vision and comfort. In our patient active disease was managed with topical and systemic immunosuppression, surgery was performed for sequelae of LSCD and cataract, and scleral contact lens prosthetic devices were prescribed for visual refinement. This case highlights long-term comprehensive management of VKC resulting in well-controlled disease with good vision and a stable ocular surface.

Learning points

- Vernal keratoconjunctivitis (VKC) management requires a comprehensive approach that includes treatment of active disease, prevention of irreversible complications and visual rehabilitation of patients.
- Severe VKC needs aggressive but monitored immunosuppression in the form of topical and systemic steroids.
- Amniotic membrane transplantation followed by cataract surgery performed for limbal stem cell deficiency and cataract secondary to VKC can significantly improve vision.
- Optical prosthetics such as scleral contact lenses can greatly enhance patients vision and comfort, and should be an option considered in all cases with irregular corneas.

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Competing interests None declared.

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