

Salvaging the sight of a 6-year-old keratoconic kid with vernal keratoconjunctivitis

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A 6-year-old boy presented with hand-movement vision in the right eye and counting finger 2 m vision in the left eye. Clinical examination revealed hydrops in the right eye, apical scar in the left eye, and pseudogerontoxon in both eyes. Descemetopexy was planned for the right eye, but after 4 days, he came with severe pain and perception of light vision in that eye. Clinical examination, scraping, and culture revealed *Pseudomonas* corneal ulcer. After scarring, optical penetrating keratoplasty was done for the right eye, resulting in best spectacle-corrected visual acuity of 6/12. The left eye also developed hydrops, which scarred with topical steroid only, resulting in best-corrected visual acuity 6/9 with miniscleral lenses.

Key words: Corneal ulcer, hydrops, miniscleral lenses, optical PK, pseudogerontoxon, vernal keratoconjunctivitis

A 6-year-old male child with a history of severe itching, photophobia, and intense eye rubbing presented with dimness of vision in both eyes for 6 months. The parents complained of abrupt dimness of vision in the right eye for last 2 days.

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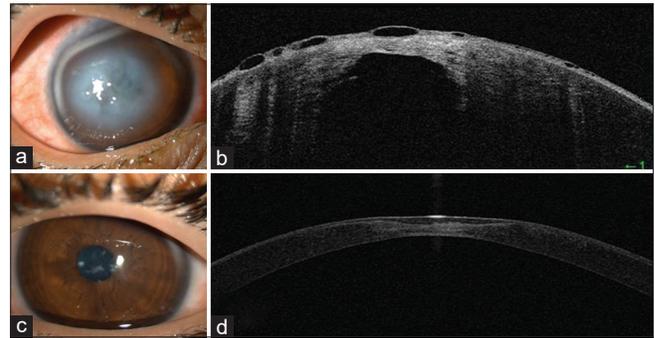


Figure 1: Slit-lamp photographs of the anterior segment: (a) right eye hydrops and pseudogerontoxon, (c) left eye apical scar and pseudogerontoxon. ASOCT pictures of (b) right eye stromal edema, (d) left eye stromal scarring. ASOCT = anterior segment optical coherence tomography

On examination, best-corrected visual acuity was hand movement in the right eye and finger counting 2 m in the left eye. Slit-lamp examination revealed acute hydrops in the right eye and Vogt striae with apical scarring in the left eye [Fig. 1a and c]. Both eyes had pseudogerontoxon [Fig. 1a] and upper tarsal papillae less than 1 mm in diameter. Right eye ultrasonography (USG) and left eye fundus examination were normal. Left eye Pentacam revealed thinnest pachymetry of 154 μm with global thinning [Fig. 2]. Anterior segment optical coherence tomography (ASOCT) in the right eye showed huge stromal edema [Fig. 1b] and left eye stromal scarring [Fig. 1d]. There was no systemic association like asthma, rhinitis, or eczema. The child was advised for right eye descemetopexy under general anesthesia (GA). Prednisolone acetate eyedrop six times/day was given till he got GA fitness. He came back after 4 days with severe redness, pain, and lid swelling with perception of light (PL) vision in the right eye. Slit-lamp examination revealed central corneal ulcer with 1 mm hypopyon in that eye [Fig. 3a]. Right eye USG was repeated and found to be normal. The ulcer was scraped and sent for Gram staining, potassium hydroxide (KOH)

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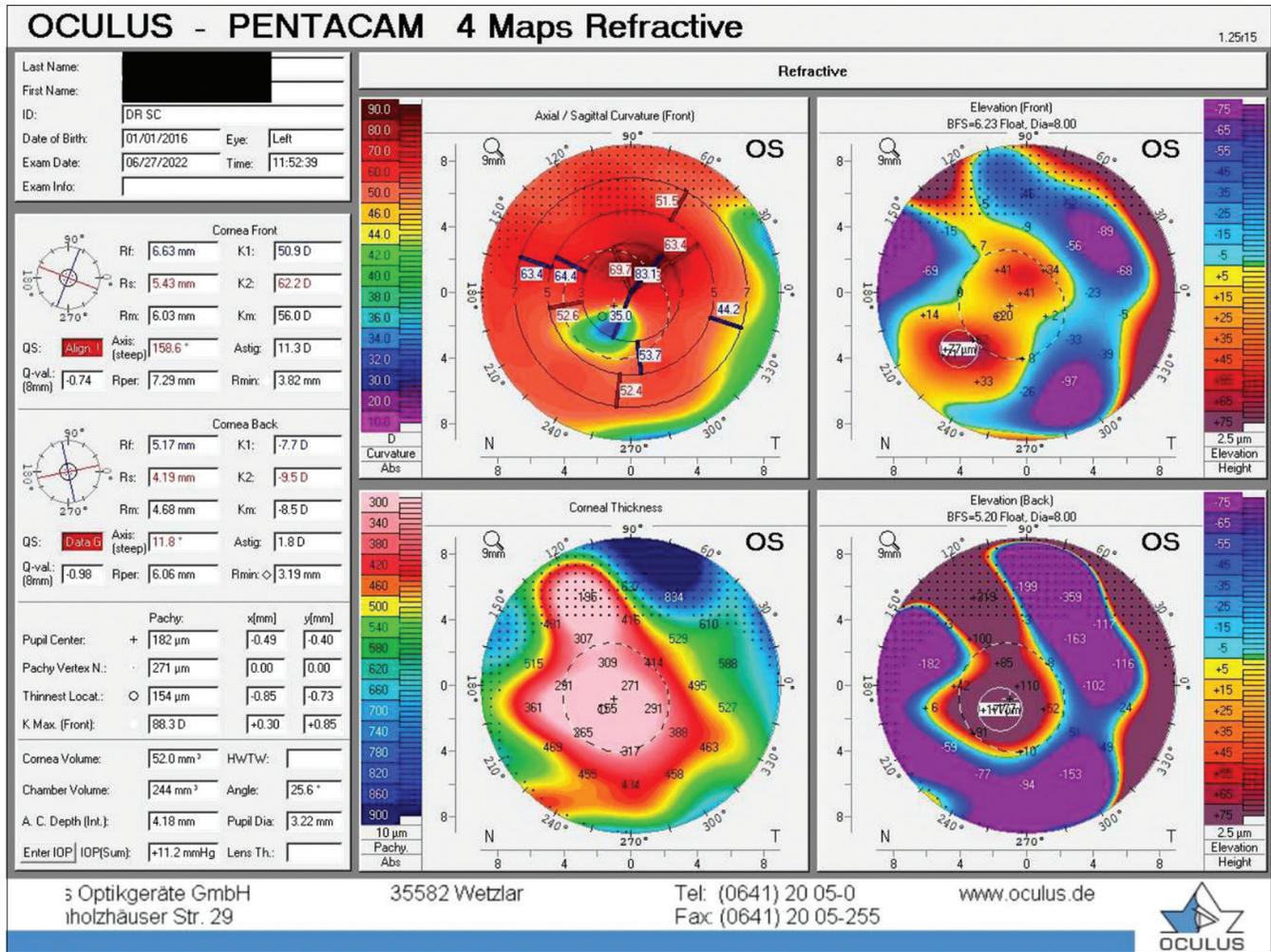


Figure 2: Pentacam showing global thinning in the left eye

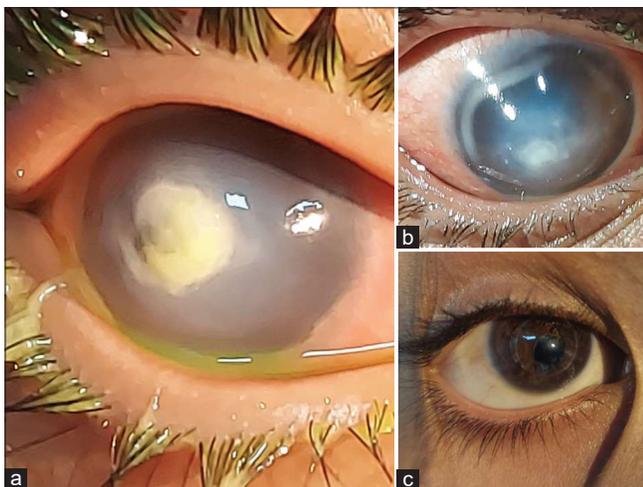


Figure 3: Slit-lamp photographs of the anterior segment: (a) *Pseudomonas* corneal ulcer of the right eye, (b) scarring of the ulcer with medical management, (c) post-optical PK clear graft. PK = penetrating keratoplasty

mount, and bacterial and fungal culture. The reports revealed the organism to be *Pseudomonas aeruginosa*, and fortified ceftazidime hourly was started in the right eye, along

with homatropine eyedrop thrice a day. The ulcer started scarring [Fig. 3b] and hypopyon disappeared after 1 week, and the eye became quiet after 1 month. After 6 months, optical penetrating keratoplasty (PK) was done for the right eye. The child got back 6/12 vision with -2.00Dsph/-2.75Dcyl @135° spectacle correction after 3 months of PK [Fig. 3c]. He came back after 1 month complaining of sudden dimness of vision in the left eye for last 2 days. Slit-lamp examination and ASOCT revealed hydrops in the left eye [Fig. 4a]. Prednisolone acetate was started for the left eye six times a day as the parents were not ready for descemetopexy. The edema resolved over 3 months, and his vision improved to 6/9 with miniscleral lenses in the left eye [Fig. 4b]. He was prescribed cyclosporine eyedrops (0.05%) twice daily and tacrolimus eye ointment (0.03%) at night to be continued in both the eyes along with profuse lubrication to prevent flaring up of symptoms of vernal keratoconjunctivitis (VKC). The importance of not rubbing the eyes was explained to both the parents and the child repeatedly.

Discussion

The association of keratoconus with VKC is well known.^[1] In children with keratoconus, the percentage of patients with VKC ranges from 8.8% to 36%.^[2] Corneal hydrops



Figure 4: Slit-lamp photographs of the anterior segment: (a) hydrops of the left eye, (b) scleral lens fitting of the left eye after resolution of corneal edema

has been common and may be a presenting sign in keratoconus with VKC,^[3] like in our case. It is noted that allergic keratoconjunctivitis with eye rubbing may increase the incidence of corneal hydrops in children with keratoconus.^[4] The association with complicated corneal findings of VKC, such as pseudogerontoxon, shield ulcer, and punctate epithelial keratitis, showed a high incidence with keratoconus cases with VKC.^[5] In our case, the presence of pseudogerontoxon in both the eyes and history of vigorous eye rubbing as stated by the parents may explain the bilateral hydrops in such a young boy. The unique finding in our case is the occurrence of hydrops despite stromal scarring in the left eye. Increased epithelial thickening, stromal thinning at the keratoconus cone, anterior hyperreflective material at the Bowman's layer level, and the absence of stromal scarring are associated with a high risk of developing corneal hydrops, whereas the presence of corneal scarring is a preventive factor.^[6] In our case, the thinnest pachymetry was 154 μm , as evident from Pentacam [Fig. 2], and stromal scarring was present in ASOCT [Fig. 1d]. In pediatric patients, keratoconus is often more aggressive because of higher rate of corneal collagen remodeling.^[7] Whether vigorous rubbing negated the effect of scarring and resulted in hydrops is a point to ponder in our case. It is already known that a dose-dependent relationship between the length and force of eye rubbing and keratoconus severity exists.^[8] Eye rubbing increases the level of tear metalloproteinase-13, interleukin (IL)-6, and tumor necrosis factor (TNF)- α in normal and keratoconic eyes.^[9] On the other hand, the rapid resolution of edema without any surgical intervention may be due to the scarring itself in the left eye. Our case shows that timely and appropriate intervention helps in recovering the vision satisfactorily. As the age is vulnerable for developing amblyopia, rigorous teaching was given to the parents for the regular use of scleral contact lens and spectacles. VKC is a chronic and severe disease, so avoiding exposure to dust, using sunglasses, and using cold compress while itching, as it acts as a natural decongestant, and profuse use of lubricants, which help to dilute the allergens and inflammatory mediators, were stressed upon. The requirement of regular follow-up and

strict avoidance of eye rubbing was ensured. The calcineurin inhibitor cyclosporine A (CsA) 0.05% was prescribed for both the eyes as it has marked steroid-sparing effect with good symptomatic control. A randomized controlled 2-year crossover study demonstrated the safety and efficacy of CsA 0.05% for the long-term prevention of VKC relapses.^[10] Tacrolimus (0.03%) eye ointment, as seen clinically, also helps to reduce the symptoms of VKC. Both these drugs were prescribed topically to reduce the side effects associated with long-term use of topical steroid, the most detrimental being development of glaucoma. This case reinforces the importance of spreading awareness regarding the deleterious effects of eye rubbing in allergic conjunctivitis and the importance of seeking early medical help to salvage the vision.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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