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Descemetocele and Corneal Neovascularization as Unusual Complication of Blepharokeratoconjunctivitis in Pediatric Patient : A Case Report

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A B S T R A C T
Introduction: Blepharokeratoconjunctivitis is a chronic and recurrent inflammatory eyelid disorder that often leads to secondary conjunctival and corneal complications, and frequently underdiagnosed in children.
Case Presentation: A 5-year-old girl presented with a blackish lump in her cornea, whitish spot, eye redness, and excessive tearing for 1 year. Her medical history included intermittent right eyelid swelling and eye redness over 3 years, which improved with eye drops, as well as a dust mite allergy. Visual acuity in the right eye was limited to hand movements. Examination showed eyelid telangiectasia, conjunctival inflammation, extensive corneal blood vessel growth, corneal thinning, and scarring. The patient received an anti-VEGF injection and amniotic membrane transplantation. At the two-month follow-up, her visual acuity improved to counting fingers at two meters and there was a notable reduction in corneal neovascularization.
Discussion: This case illustrates that severe blepharokeratoconjunctivitis can lead to serious complications such as corneal neovascularization and descemetocele. In this patient, the use of anti-VEGF injections was aimed at inhibiting further neovascularization in the cornea, while amniotic membrane transplantation was performed to manage the descemetocele and promote corneal healing.
Conclusion: Pediatric blepharokeratoconjunctivitis, along with its associated complications can pose a significant threat to vision. Early recognition and appropriate management of this condition are crucial in reducing disease progression and risk of long-term visual impairment.

Keywords: *Blepharokeratoconjunctivitis, descemetocele, pediatric, corneal neovascularization*

INTRODUCTION

Blepharokeratoconjunctivitis (BKC) is a chronic and recurrent inflammatory disorder that affects the eyelids, frequently extending to involve the conjunctiva and cornea. It is a common yet often underdiagnosed condition in children, typically manifesting with a range of clinical features such as anterior or posterior lid margin disease, recurrent episodes of conjunctivitis, and keratopathy, which can lead to complications like punctate erosions, phlyctenules, marginal keratitis, and corneal ulceration (1).
The onset of BKC usually occurs between the ages of 3.2 and 6.2 years, but the diagnosis is often delayed until around 6.9 to 9.3 years of age. The condition is particularly prevalent among South-Asian and Middle-Eastern populations, though it also affects Caucasians, with up to 30% of those patients potentially continuing to experience severe disease into adulthood (2).
Managing BKC is particularly challenging due to the absence of a definitive cure, making treatment primarily focused on disease control to prevent serious complications such as amblyopia, corneal scarring, thinning, and perforation. This case report details the presentation of a 5-year-old girl who developed a black lump on the right cornea accompanied by corneal neovascularization as a result of recurrent BKC. It emphasizes the importance of carefully identifying the primary cause and tailoring treatment to improve her visual prognosis

and prevent long-term complications.(2,3)

CASE PRESENTATION

A 5-year-old child presented in the ophthalmology clinic at Dr. Soetomo General Hospital with a one-month history of a blackish lump in the right eye, preceded by a whitish spot that had been present for a year along with redness, glare, and tearing. Despite previous treatment with ofloxacin, gentamycin, and artificial tears the whitish spot persisted. The patient had a history of intermittent eyelid lumps and redness since early 2019, which improved with eye drops. She also had a known allergy to house dust mites, managed with weekly immunotherapy injections and a family history of allergy.

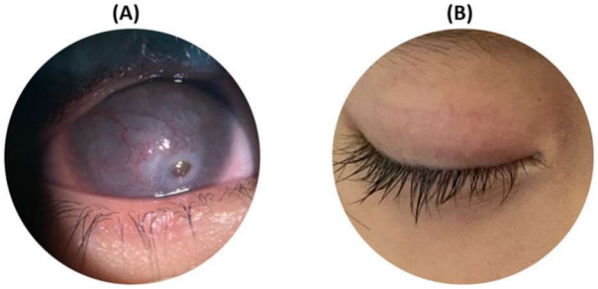

Figure 1. Right eye (A) and eyelid (B) examination

Ophthalmologic examination revealed right eye visual acuity of hand movement, with significant anterior segment findings including telangiectasis of the palpebra, hyperemic conjunctiva, extensive corneal neovascularization, a 2x2 mm descemetocele at 5 o'clock position, leucoma, and corneal thinning (**Figure 1**). The left eye had palpebral telangiectasis but was otherwise normal (**Figure 2**). Skin prick testing confirmed hypersensitivity to house dust mites, freshwater fish, and histamine.

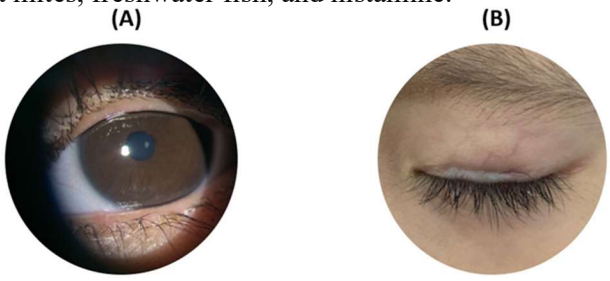

Figure 2. Left eye (A) and eyelid (B) examination

From anamnesis and physical examination, we diagnosed this patient as blepharokeratoconjunctivitis with descemetocele and corneal neovascularization. The patient was given lubrication therapy with artificial tears non preservative eye drop every 2 hour, sodium hyaluronate 0,1% eye drop every 6 hours. We also planned to perform amnion membrane transplantation with anti vascular endothelial growth factor (VEGF) subconjunctival injection.

From the surgery, we found papillary and follicle hypertrophy from both eyes and also subepithelial fibrosis from the right eye (**Figure 3**). Anti-VEGF subconjunctival 1.25 mg in 0.05 ml balanced salt solution (BSS) was injected at upper part of the cornea. After that we implanted amnion 2 layers inlay followed by suturing the amnion to corneoscleral with Polyglactin 910 8.0 suture then we added 2 more layers of amnion inlay and did the suture like before. At last, we added 4 amnion overlay and sutured the amnion to episcleral 1 mm from the limbus with nylon 10.0.

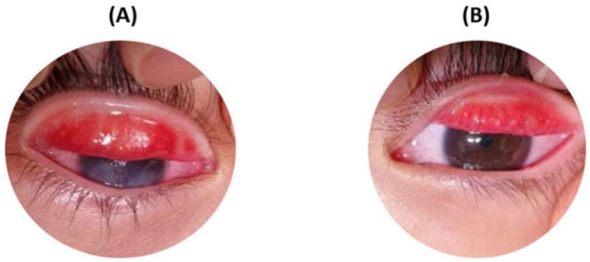

Figure 3. Right eye (A) and Left eye (B)

On day 1 post-surgery, the patient reported minimal pain in the right eye, with unchanged visual acuity. Examination revealed conjunctival hyperemia, subconjunctival bleeding, corneal neovascularization, an intact amnion, and a contact lens, though further anterior segment evaluation was challenging (**Figure 4**). Two months post-surgery, the right eye's visual acuity improved to counting fingers at 2 meters with reduced corneal neovascularization and corneal clarity was improved (**Figure 5**).

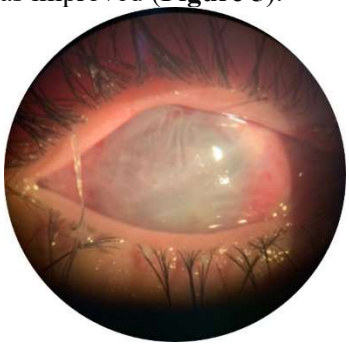

Figure 4. Right eye examination day 1 after surgery

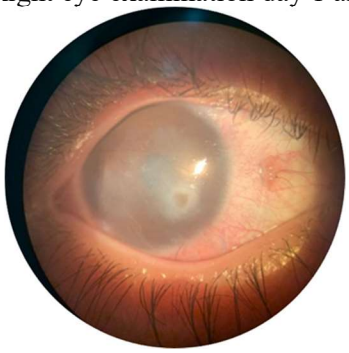

Figure 5. Right eye examination 2 months after surgery

DISCUSSION

Blepharokeratoconjunctivitis (BKC) in adult is well-characterized as an inflammatory eyelid margin disease with secondary involvement of the conjunctiva and cornea presenting with symptoms such as inflamed eyelids, telangiectasia, and crusting at the base of the cilia. However, BKC is less clearly defined in children but has gained recognition due to rising cases in Asian and Middle-Eastern populations (3,4). Clinically BKC in children involves eyelid margin scarring, blepharitis, meibomitis and chalazia or hordeolum, accompanied by corneal complications such as keratitis, scarring, vascularization and even perforation. Children usually present with symptoms such as red eye, photophobia, epiphora and recurrent chalazia with corneal involvement being common and often central or paracentral (2,5). The pathogenesis of BKC is thought to involve hypersensitivity reactions, particularly type IV against bacterial antigens, direct damage from staphylococcal exotoxins, and possibly the role of Demodex mites. Due to their immature immune response, children are more susceptible to severe corneal damage. This patient had history of recurrent chalazion, eye redness, epiphora and also history

of house dust mites allergy. From examination was found palpebral telangiectasia, papillary and follicular hypertrophy from the palpebral conjunctiva of both eyes (5–7).

Severe BKC can lead to complications such as corneal neovascularization and descemetocele formation, with management strategies depending on the maturity of the blood vessels. Treatments like laser therapy, steroid, anti-VEGF, fine needle diathermy, gene therapy, and amniotic membrane transplantation to maintain corneal transparency and visual function. In this patient was found extensive descemetocele with corneal thinning and also corneal neovascularization. These findings support our diagnosis as blepharokeratoconjunctivitis with corneal neovascularization and descemetocele as complications (8,9).

Descemetocele involves the herniation of Descemet's membrane through a defect in the corneal stroma. This condition requires urgent intervention to restore ocular integrity and prevent further complications due to a high risk of corneal perforation. Treatment strategies include the use of tissue adhesives like cyanoacrylate glue, amniotic membrane transplantation and various keratoplasty techniques, such as deep anterior lamellar keratoplasty (DALK) and penetrating keratoplasty (PK) (10). Since the patient has corneal neovascularization, there was higher risk of allograft rejection after corneal transplantation. Transplanting onto vascularized corneal beds is generally discouraged due to the high risk of immune rejection. A meta-analysis involving 24,000 corneal transplants showed that the rate of rejection is higher in patients with corneal neovascularization. The analysis demonstrates that pre-existing corneal neovascularization before surgery raises the chance of transplant failure by 30% and more than doubles the likelihood of graft rejection. This finding suggests a direct link between the extent of neovascularization and an increased risk of rejection. Addressing neovascularization is essential for reducing the immunoinflammatory response and enhancing graft survival. Therapeutic options include anti-VEGF agents such as bevacizumab, which are effective against active vessel growth but have limited efficacy on mature vessels (9,11–14).

Consequently, the patient received an anti-VEGF injection to reduce corneal neovascularization and underwent an amniotic membrane transplant to address the descemetocele, as this approach was deemed the most appropriate for the patient's condition. This combined management strategy yielded satisfactory results (15,16).

CONCLUSION

Blepharokeratoconjunctivitis in pediatric patient can pose a serious threat to vision, especially due to complications arising from its treatment. In the Asian population, the disease appears to manifest more severely, often necessitating prolonged treatment and leading to a higher frequency of corneal thinning and spontaneous corneal perforation, which may require surgical intervention. Prompt recognition and proper management of this condition are crucial in halting disease progression and reducing the risk of visual impairment.**REFERENCES**

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