

Case Series

Spontaneous corneal perforation associated with atopic keratoconjunctivitis: a case series and literature review

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ABSTRACT.

Purpose: To describe spontaneous corneal perforations in patients with atopic keratoconjunctivitis.

Methods: Records of patients with atopic keratoconjunctivitis and spontaneous corneal perforation seen by the authors were reviewed. A literature search of corneal complications in atopic keratoconjunctivitis and predisposing factors for spontaneous corneal perforations in corneal ectasias was undertaken.

Results: Three patients with seven incidents of corneal perforation were identified and presented. Corneal thinning and keratoconus are observed at a higher rate in patients with atopic keratoconjunctivitis, and severe ocular allergy has been reported as a risk factor for corneal hydrops.

Conclusions: Atopic keratoconjunctivitis appears to be a risk factor for spontaneous sterile corneal perforation. Corneal thinning and keratoconus in addition to inflammatory changes of the cornea in these patients probably predispose to the corneal melt in these perforations. A close observation of the fellow eye of affected patients is important and systemic immunosuppressive therapy should be considered.

Key words: atopic keratoconjunctivitis – atopy – corneal ectasia – corneal perforation – keratoconus

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Introduction

In atopic keratoconjunctivitis (AKC), irreversible visual loss is mainly attributed to corneal pathology. The corneal involvement often starts with superficial punctate keratitis, but with repeated flare-ups and persistence of the inflammation, there is a risk for long-standing epithelial defects followed by scarring and neovascularization. AKC patients are also predisposed to infectious keratitis with

scar formation as a consequence. Changes in corneal curvature, corneal thinning and frank keratoconus (KC) are reported at a high incidence in AKC patients (Garrity & Liesegang 1983; Foster & Calonge 1990; Tuft et al. 1991; Power et al. 1998; Jain et al. 2010). Spontaneous corneal perforation, however, has previously rarely been recognized as associated to AKC. Three cases only are found in the literature. Garrity & Liesegang briefly described a case of spontaneous

central corneal perforation necessitating evisceration (1983), Foster & Calonge reported a case with KC and advanced AKC with recurrent corneal perforation who stabilized after repeated penetrating keratoplasty (PKP) (1990), and Symes et al. reported of a patient with pellucid marginal degeneration (PMD) and perforation managed with a lamellar keratoplasty (2007).

At St. Erik's Eye Hospital in Stockholm, Sweden, a referral centre for severe corneal conditions in the Stockholm county (roughly 2 000 000 inhabitants), the authors have managed three AKC patients with spontaneous corneal perforations over a period of 23 years, 1990–2013. The diagnosis of AKC was based on atopic eczema in combination with eyelid eczema and chronic keratoconjunctivitis. Cases of corneal perforation in infectious keratitis were not considered spontaneous and hence not included. These three patients, two with bilateral and one with a unilateral perforation, are here presented. Risk factors for spontaneous corneal perforation, treatment and follow-up are discussed. The pictures are published with permission of the patients.

Case 1

A 70-year-old man with a history of AKC and long-standing eyelid treatment with steroid ointment presented at the emergency department due to decreased vision without pain in the

left eye. Two years previously, a hard contact lens had been fitted in the left eye due to KC. Right eye vision was decreased due to glaucoma. After a few months of contact lens wear, a keratitis caused by *Streptococcus group A* and *Staphylococcus aureus* had developed. The ulcer healed with antibiotic treatment and steroids drops. After a couple of months, contact lens wear was successfully resumed with best-corrected visual acuity (BCVA) 6/6.

On admission, a descemetocele had formed and perforated in the area of the old scar. The anterior chamber was shallow (Fig. 1). A bandage lens was applied and treatment with cephalosporin and quinolone eye drops was initiated. The patient was observed overnight and the anterior chamber reformed. After 3 weeks, the bandage lens was removed and eye was stable. A couple of months later, contact lens wear was again attempted and on latest follow-up BCVA was 6/9. This patient has since passed away.

Case 2

A 52-year-old woman with an amblyopic right eye and severe AKC with bilateral corneal scarring was admitted due to intensive pain and redness of the right eye since the night before. The AKC had shown poor response to steroid treatment, and two years previously, oral cyclosporine was given with good effect on the dermatitis but with insignificant effect on the eye condition. Instead, topical cyclosporine (2%) had been started for better control of the ocular surface inflammation.

On admission, the patient had medicated with topical cyclosporine 2% for 6 weeks. BCVA was 6/54 and a small perforation was seen in the lower part of the cornea. A bandage lens was

applied but did not restore the anterior chamber, and after six days, an amniotic membrane transplant (AMT) was successfully performed (Fig. 2). The cyclosporine drops were discontinued as an association with the perforation could not be ruled out. Three weeks later on follow-up of the right eye, examination of the left eye revealed a descemetocele with threatening perforation even in this eye and an AMT was performed the following day. The right amblyopic eye recovered well (Fig. 3), while healing in the left eye was complicated by a *Staphylococcus aureus* keratitis causing corneal scarring. After careful consideration, a penetrating keratoplasty (PKP) was carried out in the left eye. BCVA on latest check-up, six years after the PKP, was 6/36 in the right eye and 6/18 in the left eye.

Case 3

A 47-year-old man with atopic disease and KC had worn hard contact lenses for one year only when he presented at the emergency department with decreased vision in the right eye since 4 days. There was no history of pain or secretion. Examination revealed a corneal perforation in the lower temporal part of the cornea with prolapsed iris tissue. The patient was admitted and on the following day a Gundersen conjunctival flap was performed. After one year and despite vascularization of the scar (Fig. 4), BCVA was 6/6 with a scleral contact lens. Two years later, the patient was once more admitted with a perforation in the same eye in the adjacent corneal area; an AMT was performed twice before a lamellar keratoplasty successfully stabilized the cornea. Three years had passed when the patient again visited the emergency

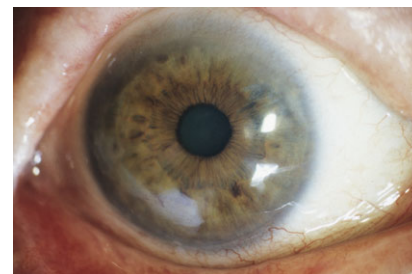


Fig. 3. Corneal healing following amniotic membrane transplant in AKC patient with spontaneous corneal perforation in the right eye, Case 2.

department due to decreased vision, now in the left eye and with pain and tearing. BCVA was 6/30. There was intensive injection, cornea was greyish in the lower part with limbal corneal thinning, and a slight flare of the anterior chamber was seen. Cultures were secured and topical antibiotics were initiated. Due to impending perforation, a lamellar patch keratoplasty was performed on the following day. Cultures revealed coagulase-negative staphylococci, but the melt was considered inflammatory rather than infectious. Cornea healed well and BCVA was 6/6 in both eyes with refitted scleral contact lenses.

Five years after the first corneal perforation, the patient moved to southern Sweden. One year after his relocation, he presented at the ophthalmic department of the University Hospital of Lund, and a paracentral corneal perforation was found in the left eye. The perforation was initially sealed by tissue glue and the eye treated with topical antibiotics. A couple of days later, the tissue glue was replaced with a lamellar corneal patch transplant. The patient was treated with oral prednisolone (40 mg) together with methotrexate (12.5 mg/week) and



Fig. 1. AKC patient with spontaneous corneal perforation of the left eye, anterior chamber shallowing, Case 1.

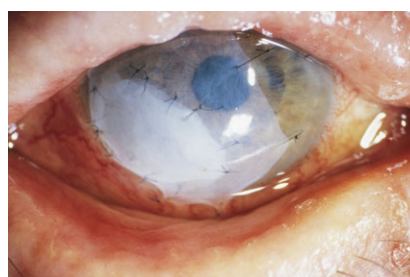


Fig. 2. Amniotic membrane transplant in AKC patient with spontaneous corneal perforation in the right eye, Case 2.

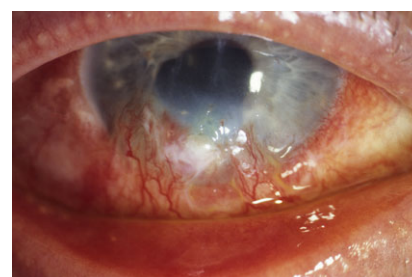


Fig. 4. Corneal scarring following conjunctival patching in AKC patient with spontaneous corneal perforation in the right eye, Case 3.

cornea healed. The prednisolone was tapered and stopped, while the methotrexate therapy was continued for little more than one year. In the left eye also corneal cross-linking was performed as a preventive procedure. On the latest check-up, 4 years after the last perforation event, visual acuity was 6/15 on the right eye and 6/30 on the left eye, rigid contact lenses could no longer be tolerated, but the eyes were stable.

Discussion

Spontaneous corneal perforation is a rare condition. In systemic disorders such as Rheumatoid arthritis (RA), Sjogren's syndrome and graft-versus-host disease (GVHD), spontaneous melting and perforation are reported (Bernauer et al. 1995; Hick et al. 2005; Inagaki et al. 2011). The same complications are found also in ocular rosacea (Akpek et al. 1997; Al Arfaj & Al Zamil 2010) and rarely in KC and PMD as shown in Table 1. There is no previous report specifically addressing AKC and spontaneous corneal perforation. This report presents 3 patients with AKC, one with unilateral and 2 with bilateral sudden, spontaneous corneal perforation, and patient characteristics are summarized in Table 1.

It is well known that there is an association between keratoconus and atopic disease (Spencer & Fisher 1959; Gasset et al. 1978; Harrison et al. 1989; Bawazeer et al. 2000; McMonnies & Boneham 2003). Also, a difference with thinner corneas in atopic versus non-atopic KC subjects has been demonstrated (Kerr Muir et al. 1987; Kaya et al. 2007). In our experience as well

as in several studies, a higher incidence of ectatic corneal disorders, KC and PMD, is observed in patients with severe ocular allergy, see Table 2. Searching the literature for reports of spontaneous corneal perforation in KC and PMD proves perforation to be very rare. We found 20 case reports describing 27 patients as presented in Table 3. In 6 cases, atopy or AKC was reported and in the remaining 21 cases, allergic disease was denied. In all these case studies, perforation was preceded by hydrops, and in studies evaluating risk factors for hydrops, a statistically higher risk was found in patients with chronic allergic disease (Tuft et al. 1994; Grewal et al. 1999; Sharma et al. 2009). Patients with AKC do not always have ongoing allergic symptoms or eczema, but a careful patient history might reveal childhood eczema, asthma or a strong family history suggesting an atopic genotype. All in all, ectatic corneal disorders do appear to be more common in patients with severe ocular allergy, and these patients might have a higher risk for complications than KC patients in general.

Two of the patients in this report had KC and the third patient had corneal scarring and advanced astigmatism, case 1 had steroid ointment for eyelid eczema, case 2 was on topical cyclosporine drops, while case 3 had no topical medication and the perforations were sudden at all occasions. In other sterile melting disorders, a non-healing ulcer usually precedes perforation. Corneal remodelling and breakdown is at least in part related to matrix metalloproteinase (MMP) activity (Symes et al. 2007). An over expression

of MMPs or decrease in MMP inhibitors has been proposed in KC pathogenesis (Mackiewicz et al. 2006; Balasubramanian et al. 2012) and increased levels of several MMPs have been demonstrated in corneal melting in systemic disorders (Brejchova et al. 2010). In vernal keratoconjunctivitis (VKC), MMP-2 and -9 have been demonstrated in tear fluid (Kumagai et al. 2002) and the activity of MMP-9 has further been correlated with corneal involvement in VKC (Aldave et al. 2003). One could speculate that an imbalance in these mediators led to perforation in our cases.

The sealing of the perforations was varied on a case-by-case basis at these seven incidents of perforation but successful healing of the cornea was achieved at all events. The management of a perforation in an AKC patient may however require special vigilance concerning also the non-affected eye. Perforation in one eye appears to be a risk factor for perforation also in the fellow eye. In case 2 and 3, perforation occurred later in the contra lateral eye and in case 3, there were repeated perforations. Bilateral and recurrent perforation is seen also in RA, GVHD and rosacea and oral anti-inflammatory drugs constitute the main treatment of severe ocular manifestations in these disorders. Case reports of systemic treatment in resilient AKC are encouraging (Cornish et al. 2010; Lee et al. 2008; Stumpf et al. 2006; Hoang-Xuan et al. 1997), but there are no controlled studies and no standard regimen. Systemic immunosuppressive therapy in joint effort with dermatologist specialists should

Table 1. Patient characteristics.

Case	1	3	2
Gender	Male	Female	Male
Age at first perforation	70	52	47
Systemic disease	AD, asthma, hay fever	AD, asthma	AD, hay fever
Ocular disease except for AKC	Keratoconus. Bacterial keratitis two years earlier.	Amblyopic right eye. High astigmatism from corneal scarring.	Keratoconus
Contact lens	Yes	No	Yes
Ocular treatment at time of perforation	Steroid ointment for the eyelids	Cyclosporine eye drops	None
Perforation	Once left eye	Once right and once left eye	Twice right eye, twice left eye
Management	Bandage lens	AMT	Conjunctival grafting, AMT, glue and lamellar transplant
PKP	No	Yes, left eye	No
BCVA at last follow-up	6/9 with hard contact lens	6/36 right eye, 6/18 left eye	6/15 right eye, 6/30 left eye

AD = atopic dermatitis, AKC = atopic keratoconjunctivitis, AMT = amniotic membrane transplantation, PKP = penetrating keratoplasty, BCVA = best-corrected visual acuity.

Table 2. Reports of keratoconus and pellucid marginal degeneration in case studies and series of patients with chronic ocular allergy.

Reference, chronologic order	Patient group	Number of patients	Patients with KC or PMD	Percentage %
Anzaar et al. (2008)	Severe AKC cases	6	3	50%
Stumpf et al. (2006)	Severe AKC cases	3	1	33%
Totan et al. (2001)	KC study in VKC	82	22	26,8%
Tabbara (1999)	Corneal complications in VKC	53	3	6%
Hoang-Xuan et al. (1997)	Severe AKC cases	4	0	0
Rehany & Rumelt (1995)	VKC, hydrops as presenting sign	3	3	100%
Foster & Calonge (1990)	AKC at referral centre	45	3	7%
Cameron et al. 1989	VKC with corneal ectasia	61	61	100%
Tuft et al. (1988)	AKC at referral centre	37	7	19%
Garrity & Liesegang (1983)	AD patients with ocular affection	85	3	4%

AD = atopic dermatitis, AKC = atopic keratoconjunctivitis, KC = keratoconus, PMD = pellucid marginal degeneration, VKC = vernal keratoconjunctivitis.

Table 3. Atopic disease was reported in 6 of 27 patients in studies of spontaneous corneal perforation in keratoconus and pellucid marginal degeneration.

Reference	Diagnosis	Age/gender	Nb of eyes	Risk factor except KC/PMD
Chan et al. (2013)	PMD	61/F	2	Eczema
	PMD	62/M	1	Previous bulb kontusion
	PMD	35/F	1	Atopy
Tsaousis et al. (2012)	KC	53/M	1	Rheumatoid arthritis
Lam et al. (2011)	KC	34/F	1	None
Mostafavi & Chu (2010)	KC	55/F	1	None
	KC	58/F	1	None
	PMD	M	1	None
Gharebaghi et al. (2009)	PMD	55/M	1	Atopy
Millar & Maloof (2008)	PMD	41/F	2	None
Anzaar et al. (2008)	PMD	66/M	1	None
Page-McCaw et al. (2007)	PMD	54/K	1	AKC
	PMD	38/M	2	None
Forooghian et al. (2006)	KC	63/M	2	Eye rubbing
Dantas & Nishiwaki-Dantas (2004)	KC	55/M	1	None
	PMD	56/M	2	None
	PMD	29/M	1	None
Stoiber et al. (2003)	KC	59/F	1	Down
Akpek et al. (2001)	PMD	56/M	1	None
Nicoli et al. (1999)	KC	21/M	1	Atopic disease
Orlin & Sulewski (1998)	PMD	74/M	1	None
Lucarelli et al. (1997)	PMD	56/F	1	None
Ingraham et al. (1991)	KC	23/M	1	None
	KC	19/F	1	None
Rubsamen & McLeish (1991)	KC	35/M	1	Eye rubbing
Lahoud et al. (1987)	KC	24/F	1	Pregnancy, topical steroid
Musco & Aquavella (1987)	KC	31/F	1	Atopic disease

KC = keratoconus, PMD = pellucid marginal degeneration.

be considered in similar cases. In patients with progressive KC and PMD with corneal thickness $>400\ \mu\text{m}$, collagen cross-linking is an option to stabilize cornea. Given the experience presented above, with sequential perforation bilaterally, cross-linking may be considered for the fellow eye even without documented progress if the inflammation is under control.

In summary, this report illustrates that AKC may predispose to sterile corneal perforation. Identification of AKC as a background factor, closure

of the perforation and anti-inflammatory treatment as well as close monitoring also of the fellow eye are essential parts of optimal management.

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