

Case report

# Total necrosis of cornea, iris and crystalline lens with exposure of vitreous hyaloid face in the context of recalcitrant acanthamoeba keratitis

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## Abstract

**Purpose:** To report a rare and complicated case of acanthamoeba keratitis (AK) presented with total necrosis and dislodgment of cornea, iris, and crystalline lens with exposure of vitreous hyaloids face.

**Methods:** Case report of 28-year-old female referred to the Farabi Eye Hospital with a history of known left eye AK since 4 months earlier. She also had a history of soft contact lens wear for two years and topical steroid use before proper diagnosis. Slit-lamp examination of the left eye revealed ring infiltration and stromal edema with haziness. The patient was prescribed anti-acanthamoeba treatment. She returned after 2 weeks with increasing ring infiltration and slight vision loss. Slit-lamp examination showed spontaneous total necrosis of cornea, iris, and crystalline lens with vitreous exposure to the air.

**Results:** The patient underwent an urgent operation consisting of total debridement of necrotic tissues including a 1 mm rim of the sclera, anterior vitrectomy, tectonic penetrating keratoplasty, and amniotic membrane transplantation (AMT) with temporary lateral tarsorrhaphy. The graft was clear within the 4 years of follow-up. At the last examination, the left eye was phthisic due to ciliary shut down and visual acuity remained light perception.

**Conclusion:** Early suspicion to AK, especially in contact lens wearers, and applying diagnostic modalities like confocal microscopy and early appropriate management with cysticide agents such as polyhexamethylene biguanide may prevent these untoward complications.

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**Keywords:** Acanthamoeba keratitis; Contact lens; Corneal melting

## Introduction

Acanthamoeba keratitis (AK) is a rare, visual-threatening condition which has increased in recent years due to wearing soft contact lens.<sup>1</sup> Acanthamoeba is a protozoa found in soil, fresh and sea water, and in contact lens cleaning solutions.<sup>2</sup> A strong association has been established between AK and

contact lens use.<sup>3</sup> The major risk factors for developing AK comprise epithelial microtrauma, contact lens overuse, improper contact lens maintenance, contact lens wear in contaminated water such as the swimming pool, and exposure to contaminated water.<sup>4</sup>

The diagnosis of AK is a challenging issue. The clinical appearance varies and may be misdiagnoses as other diseases, in particular herpes simplex virus keratitis. Clinical symptoms and signs are comprised of painful eye (sometimes out of proportion to the findings), photophobia, epithelial irregularities, stromal infiltrations (which may progress to ring infiltration), stromal opacities, and radial keratoneuritis.<sup>5</sup> Late complications reported in the literature include corneal melting, cataract, secondary glaucoma, dacryoadenitis,

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anterior chamber hypopyon, reactive ischemic retinitis<sup>2</sup> and iris atrophy.<sup>6</sup>

Here, we describe a patient with AK presented with spontaneous total necrosis and dislodgment of cornea, iris, and crystalline lens with exposure of vitreous hyaloids face.

### Case report

A 28-year-old female who wore soft contact lenses was presented to the emergency department with a history of known AK since 4 months earlier. Previously, she had been diagnosed with AK according to the confoscan biomicroscopy and prescribed anti-acanthamoeba topical medications including brolene (Propamidine isethionate) and Neosporin (Neo-bac-polym) drops for 3 months without any improvement. Prior to the diagnosis of AK, she was treated with topical corticosteroids 4 times a day for 2 weeks. At first examination in our hospital, visual acuity was 0.1 and 1.6 logMAR in the right eye and left eye, respectively. Slit-lamp examination of the left eye revealed severe ciliary injection, bulbar conjunctival congestion, ring infiltration, and stromal edema with haziness (Fig. 1). Confocal scan of the left cornea was performed showing bilayered cyst-like and irregular wedge-shaped structures within the epithelium and sub-epithelial region suggestive of active AK (Fig. 2). The patient was prescribed anti-AK treatment including polyhexamethylene biguanide (PHMB) (0.02% solution of Lavasept<sup>®</sup>

concentrate) and Chlorhexidine gluconate (0.02% solution of Betasept concentrate), and both drops were used every 2 hours. Oral fluconazole and cycloplegic eye drops were also added. Corneal scraping was also sent for smear and cultures. One week later, she did not show any improvement, and the depth of infiltration had increased. Culture results were positive for acanthamoeba. The patient returned after 2 weeks with increasing ring infiltration with slight loss of her vision. Vision loss seemed to decrease very slightly from 1.6 to 2 logMAR after two weeks. Examination showed spontaneous total necrosis of cornea, iris, and crystalline lens with vitreous exposure to the air (Fig. 3). The patient underwent an urgent operation consisting of total debridement of necrotic tissues including a 1 mm rim of the sclera, anterior vitrectomy, tectonic penetrating keratoplasty (PK), and amniotic membrane transplantation (AMT) with temporary lateral tarsorrhaphy (Fig. 4).

Following the surgery, she was prescribed chloramphenicol, brolene, and PHMB drops every 3 h as well as oral acetazolamide and fluconazole every 6 h. Thereafter, she was discharged after seven days with visual acuity of light perception in the left eye. The patient was lost to regular follow-up. The next examination was performed 9 months later showing light perception visual acuity in the left eye. The graft was clear within the 4 years of post-operation.

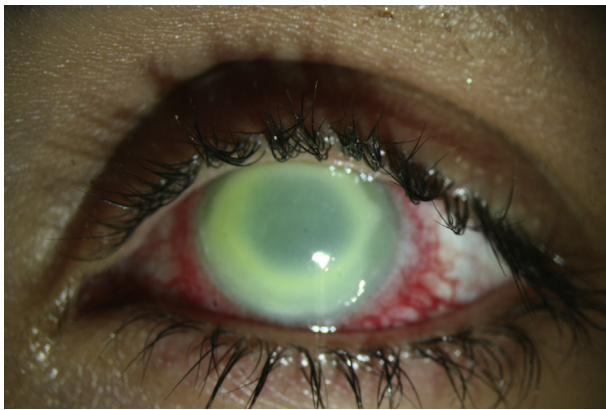


Fig. 1. Ring infiltrations of the cornea showing deep stromal invasion.

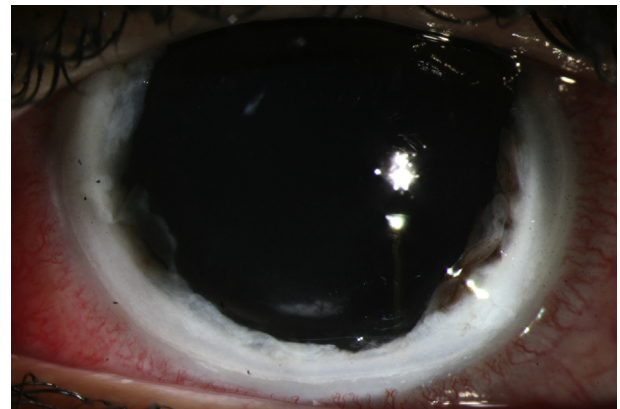


Fig. 3. Necrosis of the cornea, iris, and crystalline lens with exposure of hyaloids face of vitreous to the air.

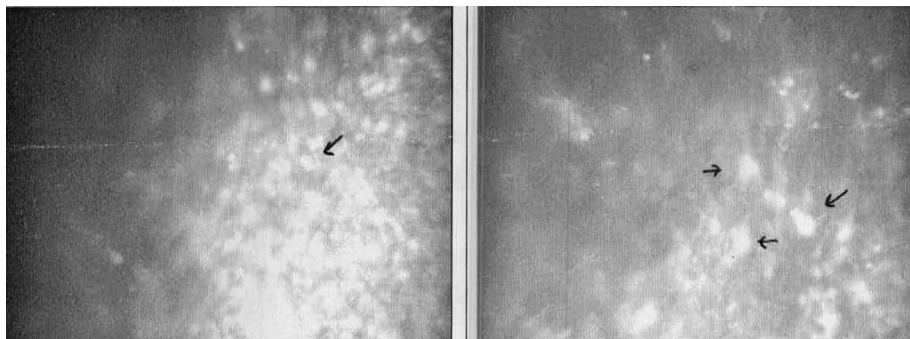


Fig. 2. Confoscan image showing bilayered, cyst-like and irregular wedge-shaped structures.

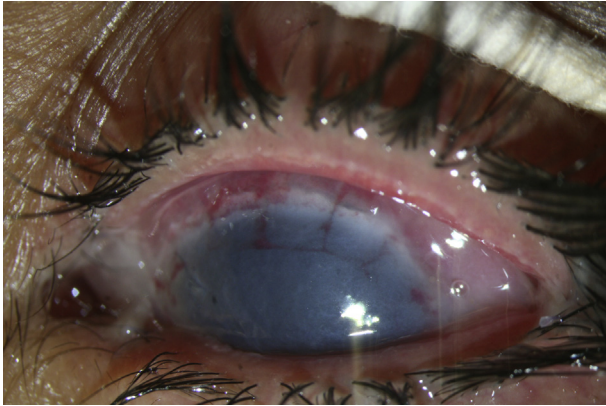


Fig. 4. Photo slit showing postoperative amniotic membrane transplantation (AMT) over large corneal graft.

At the last examination, the left eye was phthisic due to ciliary shut down and visual acuity remained light perception.

## Discussion

Contact lens wearing is the most common predisposing factor for development of AK.<sup>3,4</sup> Early diagnosis of AK enabling the successful treatment leading to the better visual outcome and also early therapeutic keratoplasty as an effective therapy for treating unresponsive AK cases should be considered.<sup>7</sup>

Diagnosis of AK is difficult due to wide variable clinical presentation. Early stage presenting symptoms include photophobia, painful eye (sometimes out of proportion to the findings), epithelial irregularities, stromal infiltrations (which may progress to advance stage including ring infiltration), stromal opacities, and radial keratoneuritis.<sup>5</sup> These findings are non-specific and may delay diagnosis of AK. Sun et al. have reported that a large number (90%) of cases are mistakenly diagnosed as viral, fungal, or bacterial keratitis.<sup>8</sup> Of particular interest is herpes simplex keratitis and presence of unilaterality, dendritiform epitheliopathy, positive response to antivirals, and decreased corneal sensation increase the suspicion for misdiagnosis as herpes simplex keratitis.<sup>9</sup> In terms of treatment delay, Claerhout et al. showed a time interval less than 18 days between onset of symptoms and start of treatment results in a better final visual acuity.<sup>10</sup> Current medications used against acanthamoeba are biguanides such as PHMB which is used at low concentration (0.02%) and chlorhexidine, which is effective against both amoebic and trophozoite forms. Chlorhexidine is often used in combination with aromatic diamidines such as propamidine isethionate (brolene) 0.1%, 0.15%, dibromopropamidine and neomycin. Corticosteroids may be used in persistent inflammation, however there are controversies regarding their use because they produce resistant conditions due to the suppression of immunologic response of the body and inhibition of the processes of encystment and encystation of acanthamoeba.<sup>1</sup> In the present case, the patient was treated with topical steroids before initiation of anti-AK treatment, probably worsening the course of disease.

Late diagnosis, low initial visual acuity, corneal neovascularization, large infiltrates, and preperforated infiltrates<sup>11</sup> were identified to need surgical interventions including PK and AMT. As in the present case, development of large infiltrates and low visual acuity (2 in logMAR system) as well as resistance to medications led to surgery in spite of active AK. In the report by Nguyen et al.,<sup>12</sup> none of the 9 patients had graft rejection after PK performed during active phase of AK. Similarly, Illingworth et al.<sup>13</sup> reported good outcome following PK. Awwad et al.<sup>14</sup> also reported good results and no rejections in 13 patients who underwent PK after resolution of the keratitis. In accordance with these studies, our patient did not show graft failure. AMT has also been reported as a safe and effective treatment which decreases pain, tissue destruction, and corneal neovascularization, particularly during the active inflammatory phase. It may also be effective in reepithelialization and permit PK to be delayed.<sup>15</sup> The most important factors affecting prognosis in AK are involvement of stromal tissue or the presence of a ring infiltrate and the time period between onset of symptoms and the initiation of therapy.<sup>16</sup> Special findings in the present patient are total melting of cornea, iris, and crystalline lens with exposure of vitreous to the air. Previous complications associated with AK include corneal melting and perforation, dacryoadenitis, anterior chamber hypopyon, reactive ischemic retinitis,<sup>2</sup> sympathetic ophthalmia,<sup>17</sup> glaucoma, cataracts, and iris atrophy.<sup>6</sup> Kelley et al. reported the development of secondary glaucoma in 30% of AK patients progressing to light perception or no light perception vision.<sup>18</sup> Ficker et al. reported cataracts in 50% of eyes that underwent PK after medical therapy and in 100% of eyes that underwent PK during the active AK.<sup>19</sup> Ehlers et al. reported mature cataract and iris atrophy in 2 patients with AK.<sup>6</sup>

Total necrosis of the cornea, iris, and crystalline lens with exposure of vitreous hyaloid face is a rare and devastating complication that has not previously been reported. Delayed diagnosis may lead to penetrance and encystment of acanthamoeba to the deep stroma. Apart from acanthamoeba pathogenicity and cytotoxic enzymes, the possibility of toxic effects of medications used for treatment of AK and other ocular conditions has also been proposed. Murthy et al.<sup>20</sup> ascribed the progressive ulcerative keratitis in a patient to the use of chlorhexidine. Additionally, Ehlers et al.<sup>6</sup> related the cataract and iris atrophy to the long-term use of chlorhexidine and brolene. Johns et al.<sup>21</sup> also reported the toxic effect of brolene on the cornea. Lee et al. evaluated the effect of PHMB and brolene on human keratocytes in in-vitro setting and observed the toxicity of them; however, PHMB is more toxic.<sup>22</sup> Therefore, anti-acanthamoeba medications may have an attribution to mentioned complications in the present case.

In conclusion, delayed diagnosis of AK may lead to catastrophic complications such as total necrosis of cornea, iris, and crystalline lens with exposure of vitreous hyaloid face which to the best of our knowledge has not yet been reported in the literature. Early suspicion of AK especially in contact lens wearers and applying diagnostic modalities like confocal microscopy and early appropriate management with cysticide

agents such as PHMB may prevent these untoward complications.

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