Unusual presentation of more common disease/injury

Acute corneal hydrops mimicking full thickness perforation

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Summary

A 26-year-old Caucasian female with keratoconus presented with an acutely painful and red left eye. Visual acuity on presentation was 3/60. Slit lamp examination revealed localised Descemet's membrane break with iris partially plugging it. There was a bulging stromal cyst which would intermittently flatten and reform. The appearance when the cyst was flattened mimicked a full thickness corneal perforation. However, no obvious overlying epithelial defect was detected and an intermittent leakage through micro-perforations in the corneal epithelium was the probable cause of the variable appearance. The anterior chamber reformed and iris plug freed following an insertion of a bandage contact lens and taped eyelid. On follow-up, the Descemet's membrane had healed with visual acuity improving to 6/18. Our case illustrates the importance of identifying corneal hydrops mimicking a full thickness perforation as conservative management has a greater chance of recovery.

BACKGROUND

In our case, the acute hydrops which led to acute overhydration of the corneal stroma and accumulation of lakes of fluid within the stroma was intermittently leaking probably through microperforations in the epithelium. It settled without requiring cyanoacrylate gluing. If a diagnosis of a full thickness corneal perforation was made and corneal gluing attempted, there was a risk of converting the corneal hydrops into an actual perforation especially when trying to reposition the presumed plugged iris.

CASE PRESENTATION

A 26-year-old Caucasian female presented with an acutely painful and red left eye and decreased visual acuity while driving. There was no history of any preceding trauma or eye rubbing. She did suffer from bronchial asthma, hay fever and eczema. She had a strong family history of

keratoconus in her mother and brother. Four months previously, the patient was diagnosed to have left keratoconus (and was fitted with a rigid gas permeable contact lens. But of late she had become intolerant to the lens and had stopped wearing it. Visual acuity was 3/60 on presentation. Slit lamp examination revealed a localised Descemet's membrane break inferotemporally with iris partially plugging it. There was stromal oedema overlying the area. On closer examination, there was a large bulging stromal cyst formation (figure 1). No overlying epithelial defect was detected and Seidel's test was negative.

DIFFERENTIAL DIAGNOSIS

An intermittent leak of the accumulated stromal fluid probably through epithelial microperforations, which would

Figure 1 Anterior segment photograph showing localised Descemet's membrane break with iris partially plugging it and stromal cyst formation.

Figure 2 A drawing explaining the alternating acute stromal cyst formation and flattening through microperforations.

Figure 3 Anterior segment photograph showing soft bandage contact lens with a reformed anterior chamber and freed iris plug.

then resealed, was likely responsible for the clinical picture which alternated between stromal cyst formation and later flattening of the same (figure 2). This mimicked a full thickness corneal perforation which would be associated with an epithelial defect.

TREATMENT

A large diameter (22 mm) soft bandage contact lens (BCL) was inserted and the eyelids taped shut for 1 h. After an hour, the anterior chamber reformed by itself and iris plug was freed (figure 3). The patient was discharged the same day with contact lens insitu, chloramphenicol eye drops four times a day and shield at night time.

OUTCOME AND FOLLOW-UP

Subsequent follow-up visits showed formed anterior chamber with stromal cleft formation, but no leak with BCL insitu. The BCL fell off after 3 weeks but the cornea had healed by that time. She was also given oral acetazolamide 250mg twice a day for 3 weeks to help with the healing of the Descemet's membrane. At the most recent clinic review, the Descemet's membrane had healed, visual acuity improved to 6/18 with pinhole. She was given olapatadine eye drops and shield at night time to prevent rubbing and accidental trauma.

DISCUSSION

Acute corneal hydrops is the development of stromal and epithelial oedema of the cornea caused by the rupture of Descemet's membrane.¹ It occurs in 2–3% of keratoconus patients.² Although corneal hydrops is commonly associated with keratoconus, it is exceedingly rare to have acute corneal hydrops with perforation according to classical textbooks.^{3 4} However, in the current literature, corneal perforation in keratoconus is possible contrary to the general assumption that it is very rare and there are about

12 cases which have reported spontaneous corneal perforation from an acute corneal hydrops.^{1 5–13} Among the documented risk factors are rapid progression of ectasia, eccentric cones, vernal keratoconjunctivitis, Down's syndrome, allergic disease, pregnancy, topical steroid treatment, raised intraocular pressure and eye rubbing.^{2 5}

Although most cases of acute corneal hydrops require penetrating keratoplasty as the definitive treatment, conservative managements such as soft bandage contact lens, pressure patching, aqueous suppressants, carbonic anhydrase inhibitors and anticholinergic medications are the treatment of choice in the acute stages. A careful assessment can distinguish an acute hydrops without a full thickness perforation from one with it and this has practical importance as the former has a higher chance of settling with a bandage contact lens and pressure patching. Furthermore, inappropriate surgical intervention would have resulted in a greater risk of complications and slower recovery.

Learning points

- ▶ Acute corneal hydrops perforation in keratoconic patients is rare.
- ▶ Careful assessment of an acute hydrops should be distinguished from a full thickness perforated cornea.
- ▶ Conservative management should be considered before attempting invasive methods such as glueing or surgery.

Competing interests None.

Patient consent Obtained.

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