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(CASE REPORT)

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Corneal hydrops in advanced keratoconus

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Abstract

We report a rare case of late unilateral corneal hydrops in a 45-year-old woman with advanced keratoconus and a history of failed penetrating keratoplasty in the contralateral eye. The patient, monocular and diabetic, presented with a five-year history of progressively declining vision in her left eye. Examination revealed severe corneal protrusion, diffuse stromal edema, Munson sign, corneal neovascularization, and a dense cataract. Anterior segment optical coherence tomography (AS-OCT) showed stromal thinning and fibrosis with a Descemet's membrane tear with rolled edges, confirming chronic corneal hydrops. A penetrating keratoplasty was proposed.

This case highlights the importance of early diagnosis, vigilant follow-up, and appropriate management of keratoconus to prevent severe complications such as corneal hydrops, which may ultimately require corneal transplantation, a non-conservative therapeutic option. Awareness of such rare late presentations is essential to optimize patient outcomes and preserve visual function.

Keywords: Corneal hydrops; Keratoconus; Corneal edema; Anterior segment optical coherence tomography; Penetrating keratoplasty

1. Introduction

Keratoconus is a progressive, non-inflammatory corneal ectasia that may be complicated by corneal hydrops, a rare but severe complication resulting from a tear in Descemet's membrane and subsequent stromal edema due to aqueous humor influx. Corneal hydrops typically occurs in younger patients and presents with sudden vision loss, corneal swelling, and ocular discomfort. Late presentations in adulthood are uncommon, particularly in patients with a history of previous keratoplasty or long-standing keratoconus. Early recognition and close follow-up are crucial to prevent chronic corneal damage and the potential need for corneal transplantation [2,4,8,9,10].

2. Case Presentation

We report a rare case of late unilateral presentation of corneal hydrops in a 45-year-old woman. The patient was diabetic and monocular, with a history of advanced keratoconus and a failed penetrating keratoplasty (PKP) in the right eye performed ten years earlier.

She presented with a five-year history of gradually progressive visual loss in her left eye. Visual acuity was limited to light perception. Slit-lamp examination revealed a marked Munson's sign, severe conical corneal protrusion with diffuse stromal edema, associated with corneal neovascularization indicating the long-standing nature of the edema. The Seidel test was negative, and a dense cataract was also noted (Figure 1A–C). The right eye was in phthisis, with loose corneal

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sutures from the previous keratoplasty. Anterior segment optical coherence tomography (AS-OCT) showed stromal thinning and hyperreflective fibrosis, absence of intrastromal cysts or subepithelial bubbles, and the presence of a Descemet's membrane tear with rolled edges (**Figure 2**), confirming the diagnosis of chronic corneal hydrops. A penetrating keratoplasty was proposed.

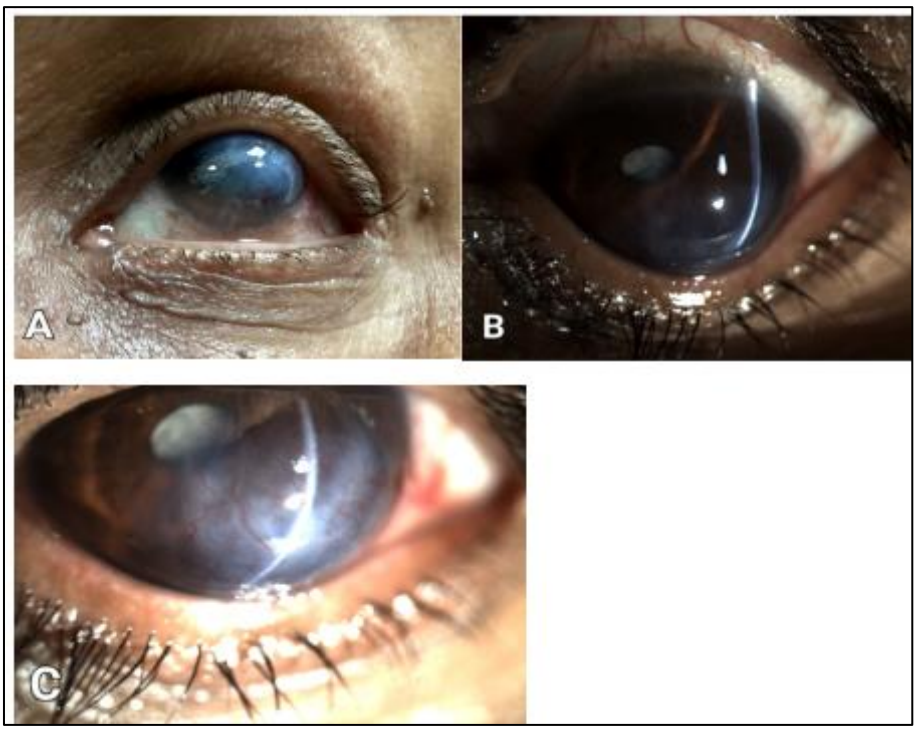

Figure 1 (A-B-C) Slit-lamp examination showing: 1.A: Chronic corneal hydrops, 1.B: Positive Munson's sign, 1.C: Corneal neovascularization

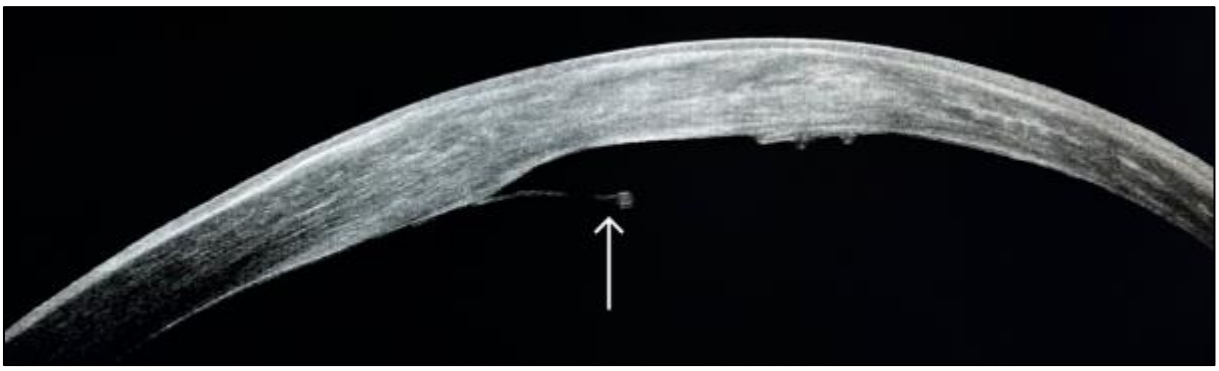

Figure 2 Anterior segment optical coherence tomography (AS-OCT): AS-OCT image of left eye showing stromal thinning and hyperreflective fibrosis, absence of intrastromal cysts or subepithelial bubbles, and the presence of a Descemet's membrane tear with rolled edges

3. Discussion

Keratoconus is a non-inflammatory, progressive ectatic disease that typically affects both eyes. It is characterized by corneal thinning and protrusion, and may lead to irregular astigmatism, myopia and corneal scarring, resulting in significant visual impairment. Keratoconus affects approximately 0.24% of the global population, with a pooled incidence of about four new cases per 100,000 person-years. Prevalence varies geographically, with higher rates reported in the Middle East and South Asia, and lower rates in Europe and North America. It is most commonly diagnosed in young adults aged 20–29 years [1,2,3].

Corneal hydrops is a rare but potentially vision-threatening complication of corneal ectatic disorders, particularly advanced keratoconus. It results from a tear in Descemet's membrane, allowing aqueous humor to leak into the corneal stroma, causing stromal edema and fluid-filled cystic spaces. Clinically, corneal hydrops may be asymptomatic or present with severe visual loss, corneal clouding, pain, and photophobia. Risk factors include vigorous eye rubbing and severe allergic eye disease [4, 5].

Corneal hydrops is diagnosed clinically and confirmed by anterior segment optical coherence tomography (AS-OCT). In the acute stage, AS-OCT shows a Descemet's membrane rupture with intrastromal fluid and stromal clefts. In chronic or resolving hydrops, AS-OCT reveals Descemet's membrane scarring, persistent stromal hyperreflectivity, residual corneal thickening, and posterior corneal irregularity, reflecting fibrotic remodeling rather than active fluid leakage. AS-OCT therefore plays a key role not only in confirming the diagnosis but also in differentiating active from chronic stages of corneal hydrops [6,7,8].

Management is usually conservative in the acute phase and may include topical hyperosmotic agents, cycloplegics, prophylactic antibiotics, and anti-inflammatory therapy to reduce edema and alleviate symptoms. However, conservative treatment may be insufficient in selected cases, which may require surgical interventions such as pneumatic descemetopexy, corneal compression sutures, endothelial keratoplasty, or penetrating keratoplasty. In late-presenting or advanced cases, as seen in our patient, persistent stromal fibrosis, neovascularization, and rolled Descemet's membrane often compromise visual recovery. In such situations, penetrating keratoplasty is usually required to restore corneal clarity and optimize vision. This highlights that while conservative management is appropriate for early acute corneal hydrops, late-stage hydrops frequently necessitates corneal transplantation [9,10,11].

4. Conclusion

Corneal hydrops is a rare but serious complication of advanced keratoconus that can lead to significant visual impairment and may ultimately require corneal transplantation if not managed appropriately. This case demonstrates that corneal hydrops can occur later than usual in adult patients, particularly those with long-standing keratoconus or previous corneal surgery. Recognizing such atypical cases is crucial for timely management and the prevention of further complications.

Compliance with ethical standards

Disclosure of conflict of interest

The authors declare no conflict of interest.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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