

A Photo Essay

Extremely Steep Keratoconic Cornea without Corneal Hydrops; How Far Can Corneal Thinning Progress?

Zisis Gatzoufas, MD, PhD; Housam Haidar, MD; Panagiotis Georgoudis, MD; Mohamed Elalfy, MD
Samer Hamada, MD

Department of Ophthalmology, Corneo-Plastic Unit, Queen Victoria Hospital NHS Trust, East Grinstead, UK

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PRESENTATION

We report a 51-year old male patient with advanced keratoconus who presented to our clinic. He was diagnosed with keratoconus in 1986 and registered severely sight-impaired in 2006. His medical history included hay fever and asthma. He had no family history of keratoconus and rubbed his eyes excessively for many years. The patient was wearing scleral contact lenses and managing well. His contact lens-corrected visual acuity was 6/30 in the right eye (OD) and 6/30 in the left eye (OS). On slit-lamp examination, we observed advanced keratoconus (OD>>OS) with extreme corneal thinning and marked Munson's sign OD [Figure 1]. Corneal topography found advanced keratoconus bilaterally with a maximum keratometry of 112.7 diopters OD and 70.9 diopters OS [Figure 2a]. The thinnest point corneal thickness was 184 microns OD and 280 microns OS, as measured using ocular coherence tomography-assisted corneal pachymetry [Figure 2b]. Despite the extremely advanced keratoconus, particularly OD, there was no evident corneal scarring and the patient had no episodes of acute corneal hydrops (ACH). The patient refused corneal

transplantation because he was managing well with his scleral contact lenses.

DISCUSSION

ACH is a rare complication of keratoconus, caused by breaks in the Descemet membrane, allowing aqueous humor to enter the corneal stroma.^[1,2] This condition is clinically characterized by marked corneal edema, resulting in a reduction in visual acuity, and usually associated with significant discomfort, photophobia, and epiphora.^[1,2] ACH is usually self-limiting, leading to formation of sight impairing corneal scarring and usually requires corneal transplantation for visual recovery.^[1,2]

Atopy, use of steroids, and eye rubbing related corneal tissue micro-trauma are suggested as risk factors for the development of ACH.^[1-3] The role of intraocular pressure (IOP) is also important, since coughing, sneezing, or nose blowing, all related to IOP elevation, have been implicated in the pathogenesis of ACH in patients with keratoconus.^[1]

Empirically, ACH is known to occur in patients with advanced keratoconus; however, there has been no evidence for this widely accepted clinical dogma. Recently, Fuentes et al provided evidence that marked corneal thinning is a major risk factor for the development of ACH, confirming the clinical theory that the more advanced the corneal ectasia, the higher risk of ACH.^[4] Nevertheless, it is unknown whether this general rule applies to all cases

Correspondence to:

Zisis Gatzoufas, MD, PhD. Queen Victoria Hospital NHS Trust, Holtye Road, RH19 3DZ, East Grinstead RH19 3DZ, United Kingdom.

E-mail: zisisg@hotmail.com

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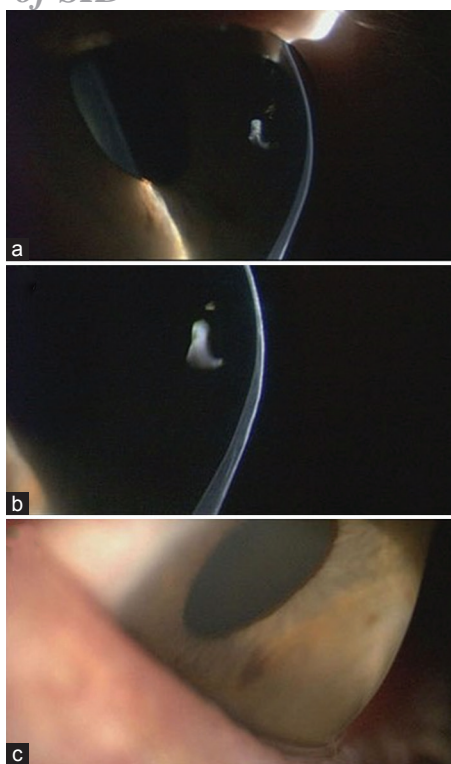


Figure 1. Slit-lamp examination of the right eye showing advanced keratoconus with extreme corneal thinning (a and b) and marked Munson's sign (c).

with keratoconus and whether there is a minimum corneal thickness threshold for the development of ACH.

Our patient presented with the steepest and thinnest keratoconic cornea that has ever been described without ACH. This case questions the validity of the clinical theory that extremely thin keratoconic corneas necessarily develop ACH, as well as the potential existence of a minimum corneal thickness threshold. ACH pathogenesis appears more complex than previously thought, expanding beyond the purely anatomical and geometrical characteristics of the cornea. Therefore, physicians should not be reluctant to monitor keratoconus patients with very advanced disease, who manage well with scleral contact lenses, since their condition could maintain long-term stability.

Declaration of Patient Consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/

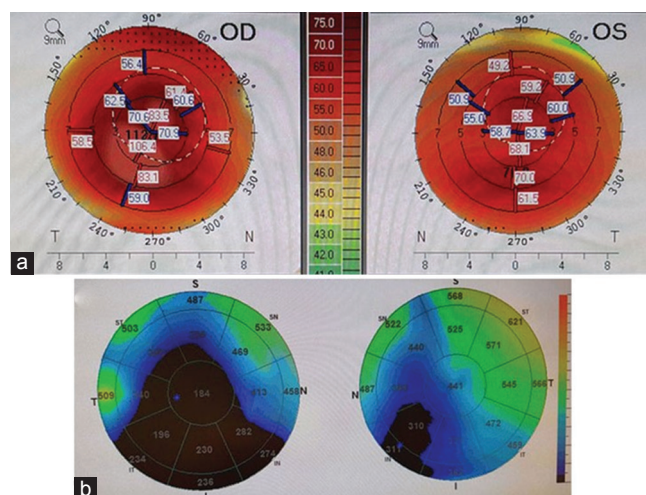


Figure 2. Corneal topography showing advanced keratoconus with a maximum keratometry of 112.7 diopters OD and 70.9 diopters OS (a). Ocular coherence tomography-assisted corneal pachymetry finding the thinnest point corneal thickness of 184 microns OD and 280 microns OS (b).

have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of Interest

There are no conflicts of interest.

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