CASE REPORT

Superior Keratoconus: A Case Report and Review of Literature

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Received on: 28 June 2023; *Accepted on:* 05 September 2023; *Published on:* 23 April 2024

Abstract

Superior keratoconus (SKC) is a rare subtype of KC characterized by corneal steepening in the superior region, which may mimic other ocular surface irregularities. These irregularities can arise from contact lens warpage, upper lid ptosis, superficial corneal scars, Terrien's marginal degeneration (TMD), and dry eyes. Accurate diagnosis of superior KC can be challenging, but newer tomographic modalities can aid in differentiating it from other conditions. This review highlights the clinical features of superior KC and discusses the role of advanced imaging techniques in improving diagnostic accuracy.

Keywords: Acute hydrops, Allergic conjunctivitis, Case report, Superior keratoconus.

International Journal of Keratoconus and Ectatic Corneal Diseases (2023): 10.5005/jp-journals-10025-1194

Introduction

Keratoconus (KC) is an ectatic corneal disease characterized by progressive corneal thinning, protrusion, and irregular astigmatism with a reduction in uncorrected visual acuity (UCVA) and best spectacle-corrected visual acuity (BSCVA). Keratoconus is diagnosed using corneal topography, with newer topographic modalities being more sensitive to detecting early and subclinical stages of KC.[1–3] Corneal topography typically shows central or inferior paracentral steepening with corresponding corneal thinning in KC. There is a rare subtype of KC known as superior keratoconus (SKC), which is less commonly reported in the literature and may present with different clinical features. Here we report on a case of SKC.

Case Report

We report on a 40-year-old female who presented with deterioration in vision in both eyes over a few months. She denies eye rubbing and spectacle correction gave suboptimal vision, in addition, she was contact lens intolerant.

Her UCVA and best spectacle-corrected visual acuity (BSCVA) were 2-meter counting fingers (CF) improving to 20/60 and 3-meters CF improving to 20/30 in the right eye (RE) and left eye (LE) respectively. The refraction was plano, –14.0 diopter of cylinder (DC) × 50° in the RE and plano, –6.0 DC × 130° in the LE. Scissoring and oil droplet signs were detected in both eyes on retinoscopy. The cornea was clear with no evidence of corneal scarring, Vogt striae, or Fleischer ring, and the rest of the anterior and dilated posterior ocular examination was unremarkable. She had no sign of dry cornea, ptosis, or subtarsal eyelid pathology.

Corneal topography demonstrated superotemporal corneal steepening with corresponding thinning, and corneal tomography mapping showed superotemporal anterior and posterior elevation with epithelial thinning in the corresponding area in the RE Kmax – 65.7. K1 61.7, K2 55.8 D, thinnest point pachymetry – 345 micrometers. Similar findings were observed in the inferotemporal cornea of the LE Kmax – 56.2. K1 53.3, K2 50.9 D, thinnest point pachymetry – 378 micrometers.

The finding supported the diagnosis of SKC in the RE and inferotemporal KC in the LE. The ocular response analyzer (ORA) readings showed reduced corneal hysteresis and resistance factor and the ORA graphs displayed characteristic patterns consistent with KC.

Literature Review

A literature search was done on PubMed using keywords including SKC and superior corneal steepening (SCS). The literature was reviewed and reported to identify true cases of SKC. Table 1 summarizes the results of the PubMed literature search.

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How to cite this article: Barbara R, Berkowitz E, Tiosano B, *et al*. Superior Keratoconus: A Case Report and Review of Literature. Int J Kerat Ect Cor Dis 2023;10(1–2):32–34.

Source of support: Nil

Conflict of interest: Dr Ramez Barbara and Dr Adel Barbara are associated as the Editorial board members of this journal and this manuscript was subjected to this journal's standard review procedures, with this peer review handled independently of these Editorial board members and their research group.

Patient consent statement: The author(s) have obtained written informed consent from the patient for publication of the case report details.

Table 1: Summary table of available literature discussing superior corneal steepening (SCS) and presumed superior keratoconus. Data presented demonstrate year of report, unilateral/bilateral presentation and imaging modality used to diagnose keratoconus

Patient	*Ref*	*Year*	*Unilateral/ Bilateral*	*Imaging modality*	*Thinnest point*
1	Eiferman et al.	1993	Bilateral	Axial mapping	NA
2			Bilateral	Axial mapping	NA
3	Prisant et al.	1997	Bilateral	TMS-1	NA
4	Weed et al.	2005	Unilateral	TMS-2	NA
5			Unilateral	TMS-2	NA
6	Chiang et al.	2006	Bilateral	Orbscan	Inferior
7	Tananuvat et al.	2008	Bilateral	Orbscan II	Central
8	Rogers et al.	2014	Bilateral	Pentacam	Superior
9			Bilateral	Pentacam	Superior
10	Özalp et al.	2019	Bilateral	Pentacam and AS-OCT	Superior
11	Mounir et al.	2020	Bilateral	Sirius	Superior
12	Galperín et al.	2021	Bilateral	Pentacam	One eye Superior, other central

NA, not available; Ref, reference

Superior keratoconus is a unique form of keratoconus characterized by corneal thinning and steepening predominantly in the superior cornea. Several studies have reported on the clinical features, diagnostic modalities, and management of SKC.

The Dundee University Scottish Keratoconus Study (DUSKS) reported on 200 patients with keratoconus diagnosed using computerized video-keratography (CVK) TMS-2 (Bausch & Lomb), unilateral SKC was present in 2 patients (1%).[4] In one case, Fleischer's ring was evidence clinically with significant corneal thinning. Both cases were managed successfully using RGP contact lenses. Prisant et al.[5] and Eiferman et al.[6] reported on two cases of bilateral asymmetrical SKC diagnosed using axial mapping only as corneal tomography was not available at the time. Chiang et al.[7] reported on a case of bilateral superior corneal protrusion and inferior paracentral corneal thinning detected by Orbscan ® (Bausch and Lomb, Inc, Rochester, NY, USA). Tananuvat et al.[8] reported on a patient with SKC and corneal hydrops detected clinically without previous corneal imaging in one eye. The second eye had an asymmetric subclinical KC with superior steepening and central thinning detected by the Orbscan II Topography System (Bausch & Lomb, Orbtek Inc., Salt Lake City, UT, USA). Rogers et al.[9] reported on two cases of bilateral asymmetric SKC with evidence of previous hydrops in one eye. The diagnosis was supported by Pentacam (OCULUS Optikgeräte GmbH, Wetzlar, Germany) demonstrating corresponding SCS and thinning. Özalp et al.[10] reported on bilateral SKC with corneal hydrops in one eye. The diagnosis was confirmed by Pentacam, anterior segment optical coherence tomography, and epithelial thickness map, while the difference in the corneal thicknesses between the central and the superior zones was only 5 μm, the difference in the epithelial thicknesses between the respective zones was 24 μm. Galperín et al.[11] reported on a 16-year-old male with asymmetric SKC in both eyes diagnosed using Pentacam. His worst eye demonstrated superior corneal thinning while the other eye had central corneal thinning.

Discussion

Superior keratoconus is rare and certain pathologies may mimic SKC and present with SCS; these conditions include contact lens warpage, ptosis, superficial corneal scars, Terrien's marginal degeneration (TMD), and dry eyes. When the inciting factor of these conditions is treated, the SCS may therefore resolve. Kim et al., reported on blepharoptosis-induced corneal steepening that resolved after ptosis repair was performed.[12] Superior pellucid marginal degeneration (PMD) were reported in several case reports, and it may mimic SKC, however, in PMD there is paralimbal thinning and protrusion of the peripheral cornea with crab claw shape corneal topography.[13–16] Terrien's marginal degeneration may also mimic SKC however, SKC lacks the characteristic paralimbal thinning, lipid deposition, and pannus seen in TMD.[17]

Most SKC-reported cases in the literature were not supported by modern tomographic modalities. In some, there was no correspondence between corneal steepening and thinning. This may raise doubts as to whether some of these reported SCS was truly SKC.

This is the first case report on SKC in one eye and inferotemporal KC in the second eye of the same patient, without any known inciting factors for SCS. To our knowledge, this combination has not been reported previously.

The treatment approach in such cases would follow that of other subtypes of KC. Mounir et al.[18] reported on the early results of combined accelerated corneal collagen crosslinking (CXL) and intrastromal Kerarings implantation by femtosecond laser in a case of advanced SKC diagnosed by Sirius Scheimpflug corneal tomography (CSO, Florence, Italy). Following the treatment, there was significant improvement in visual acuity, keratometric readings, and anterior and posterior elevation maps.

In our case, due to unsatisfactory BSCVA and the patient's intolerance to contact lenses, intrastromal corneal ring segment (ICRS) surgery was proposed as a treatment option. However, the patient did not undergo the procedure due to economic constraints, as ICRS surgery is not provided by the public healthcare system in Israel. Additionally, corneal cross-linking (CXL) was not offered due to the patient's age and the absence of a clear history of disease progression. Instead, a follow-up was recommended.

In conclusion, the diagnosis of SKC requires careful exclusion of other conditions that may mimic SKC, some of which may be reversible. Meticulous history, examination, and assessment using corneal tomographic modalities may aid diagnosis and optimize treatment options. The case report and review presented provide valuable insights into the diagnosis of SKC and raise awareness among clinicians about other potential causes of SCS. By enhancing our understanding of SKC and its differential diagnosis, clinicians can provide more precise and effective management strategies for patients with this unique form of keratoconus.

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