

Keratoconus and Macular Corneal Dystrophy Simultaneous Presentation with an Exacerbation After an Upper Respiratory Infection of COVID19 and Viral Conjunctivitis

Tariq Abdulmohsen Alanazi, MD* Abdulrahman Albuainain, MD** Hamda Mohammed AIKhalidi, MD*** Rawan Anwar Al-Abdulqader, MD**** Nasser Hijab Aldosari, MBBS*****

ABSTRACT

Case reporting of a patient with a rapid progression of keratoconus (KC) associated with Macular corneal dystrophy (MCD) after being infected with COVID 19 upper respiratory tract infection (URTI) and viral conjunctivitis. We open the floor to study COVID's implication over ectatic corneal diseases in either accelerating the sub clinical and mild clinical to severe clinical presentations, or a direct causation between COVID-19 and corneal ectasia, additionally reporting a coexistence of the keratoconus and macular stromal corneal dystrophy simultaneously.

INTRODUCTION

We aim to describe a case of a patient with rapid progression of keratoconus (KC) associated with Macular corneal dystrophy (MCD) after being infected with COVID 19 upper respiratory tract infection (URTI) and viral conjunctivitis.

CASE REPORT

A 17-year-old female who is medically free. Had history of COVID 19 infection with viral conjunctivitis at the same disease course 6 months before her presentation to our hospital. she complained of a decrease in vision, equally and gradually. Denied eye trauma nor surgeries or disease before this presentation including keratoconus or dystrophies. Not following with ophthalmologist. Has positive family history of keratoconus. Reported a long history of mildly decreased vision since her childhood even before her last complaint but didn't seek medical help. Her visual acuity was counting fingers, intraocular pressure was, pupils were round, reactive, no relative afferent pupillary defect, good redreflex, full extraocular muscle movement. external examination showed positive Munson's sign, quite conjunctivas, confluent corneal grayish opacities at the level of the stroma without clear spaces reaching the limbus, positive Vogt's stria and Flisher's ring. Healthy optic disc and macula. Figures 1 and 2 showed her anterior slit lamp photos, figure 3 and 4 showed her pentacam photos, figure 5 positive PCR result for covid 19.

Penetrating keratoplasty with pathological studying for the patient's cornea was done showed: Corneal tissue with subepithelial colloidal iron positive deposits disrupting Bowman's membrane. Alcian blue stains highlight the deposits to lesser degrees. Similar material is also seen in stromal keratocytes and endothelial cells. Masson trichrome was negative. Endothelium was relatively intact. The patient's slit lamp photos shown in figure 6 and reached 20/80 right eye vision by Snellen chart.

DISCUSSION

Keratoconus is a common corneal disorder where cornea undergoes a progressive thinning and steepening causing irregular astigmatism. The most common cause of keratoplasty in the Kingdom of Saudi Arabia (KSA)^{1,2}. Prevalence of KC is 1.38/1000 world widely. Moreover, risk factors for KC include eye rubbing the main independent risk factor reported in the literature. Along with family history of KC, allergy, asthma, and eczema^{2,3}. sleep apnea⁴. connective tissue disorders⁵. floppy eyelid⁶. In addition, Retinitis pigmentosa was reported to have poor prognosis if present with KC⁷. Furthermore, the prevalence of KC in Down syndrome patients is relatively high as compared to the population, although the progression was same in both groups⁸. KC is histopathologically defined as breaks in or complete absence of Bowman's layer, in addition to collagen disorganization, scarring, and thinning. Pathology slides showing keratoconus changes figure 7.

* Ophthalmology Department and Residency
King Saud University
Riyadh, Saudi Arabia.
E-mail: Tariqabdulmohsen@gmail.com

** Ophthalmology department
Eye and Laser Center
Bahrain Defense Force Hospital
Royal Medical Services
Military Hospital
Kingdom Of Bahrain.

*** Ophthalmology Department
Imam Abdulrahman Alfaisal University
Aldammam, Saudi Arabia.

**** King Faisal University
AlAhsa, Saudi Arabia.

***** Almaarefa University
Riyadh, Saudi Arabia.



Figure 1: Anterior segment slit lamp photos show the patient's corneal changes



Figure 2: Anterior segment slit lamp photos show the patient's corneal changes

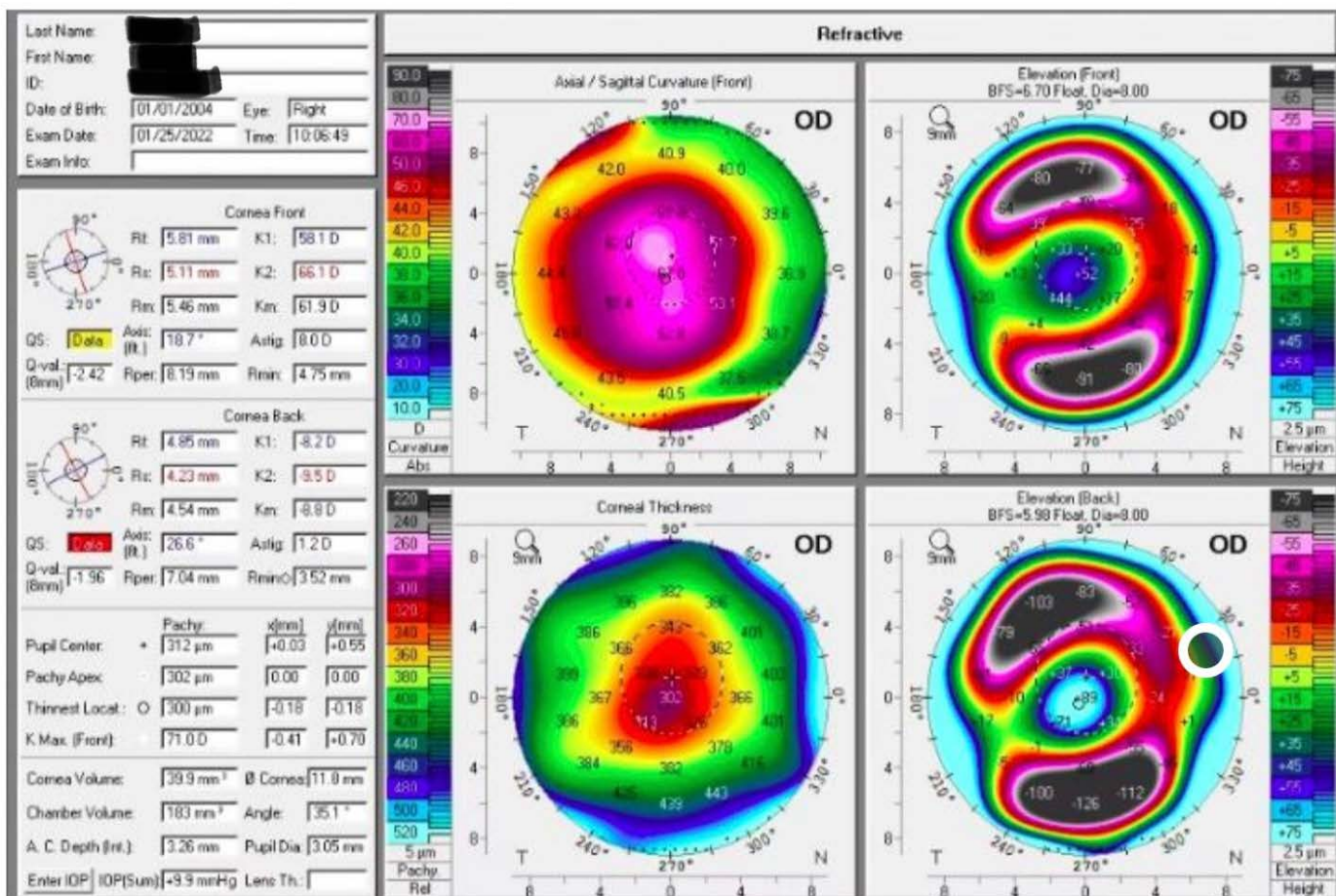


Figure 3: This is the best Pentacam result given the test is done on the patient many times with no improvement of QS values. In the right eye, Higher than normal K1 and K2 readings along with K max in the corneal front box, with a high amount of astigmatism measuring 8.0 D. Thinnest location was 300 um with displacement of the x and y -0.18 for both. The axial / Sagitta curvature has a round shaped steepening with S/I disparity more than 2.0 and I/S more than 1.4. The Elevation (front) and (back) maps showed more than the normal values. Corneal thickness showed central cone with abnormal values. These findings are concluding the diagnosis of keratoconus disease in the right eye

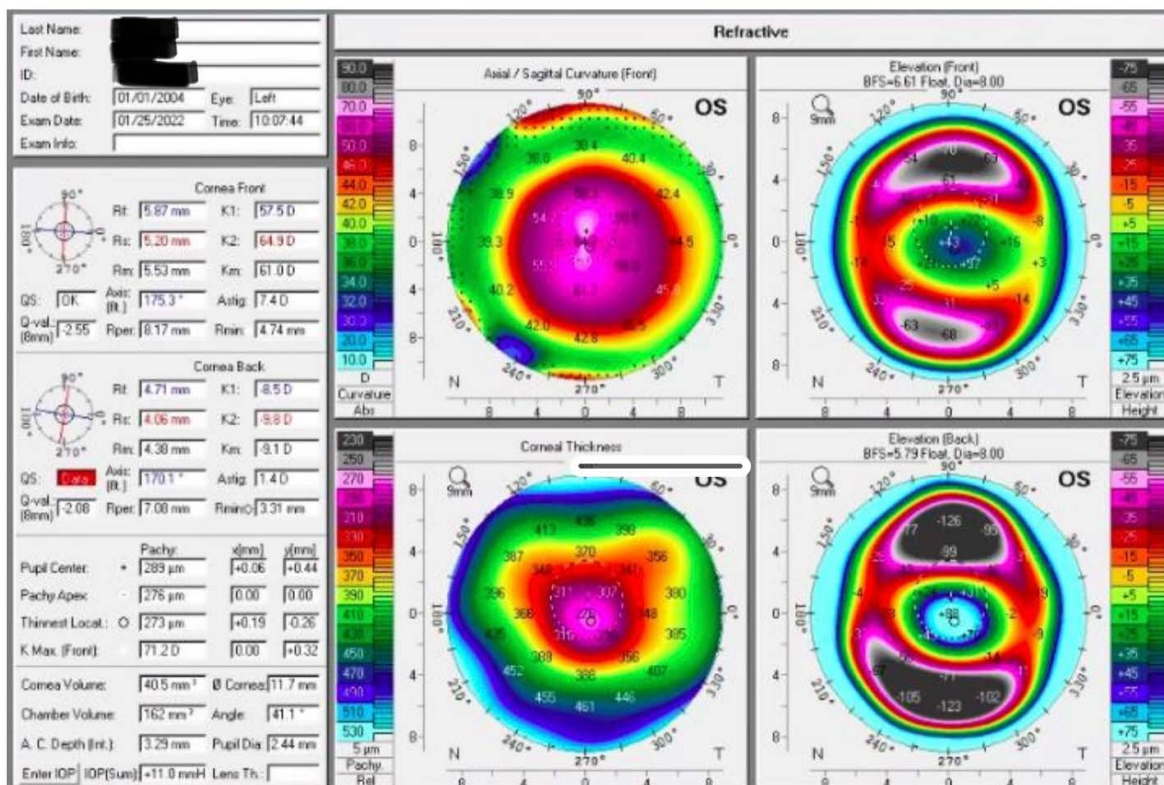


Figure 4: In the left eye, Higher than normal K1 and K2 readings along with K max in the corneal front box, with a high amount of astigmatism measuring 7.4 D. Thinnest location was 273 um with displacement of the x and y -0.19 and -0.26 respectively. The axial / Sagitta curvature has a round shaped steepening with S/I disparity more than 2.0 and I/S more than 1.4. The Elevation (front) and (back) maps showed more than the normal values. Corneal thickness showed central cone with abnormal values. These findings are concluding the diagnosis of keratoconus disease in the left eye

| | | |
|---------------------------------------|---|---------------------------|
| Date of Birth | 2004-10-11 | تاريخ الميلاد |
| Nationality | المملكة العربية السعودية Kingdom of Saudi Arabia | الجنسية |
| Blood Type | - | مجموعة الدم |
| COVID-19's Vaccination Details | | التفاصيل لقاح كورونا |
| First Dose | 2021-08-16 Batch Number - رقم اللقاح: F12026 أفزر بيو تكنولوجي Pfizer-BioNTech (COMBINATYB) | الجرعة الأولى |
| Second Dose | 2021-09-20 Batch Number - رقم اللقاح: F12026 أفزر بيو تكنولوجي Pfizer-BioNTech (COMBINATYB) | الجرعة الثانية |
| Covid-19's Related Details | | التفاصيل متعلقة بكوفيد 19 |
| Recovery Date | 2020-10-09 | تاريخ التعافي |
| PCR | | فحص كورونا |
| Latest PCR Result | - | النتيجة لفحص كورونا |
| Medical Insurance | | التأمين الطبي |
| Insurance Status | - | حالة التأمين |

Figure 5: The picture listed below from the National Saudi Portal of Tawakalna proving the patient had the infection of COVID 19 before the presentation to the ophthalmology emergency at King Abdulaziz University Hospital



Figure 6: Showing the post operative corneal pictures

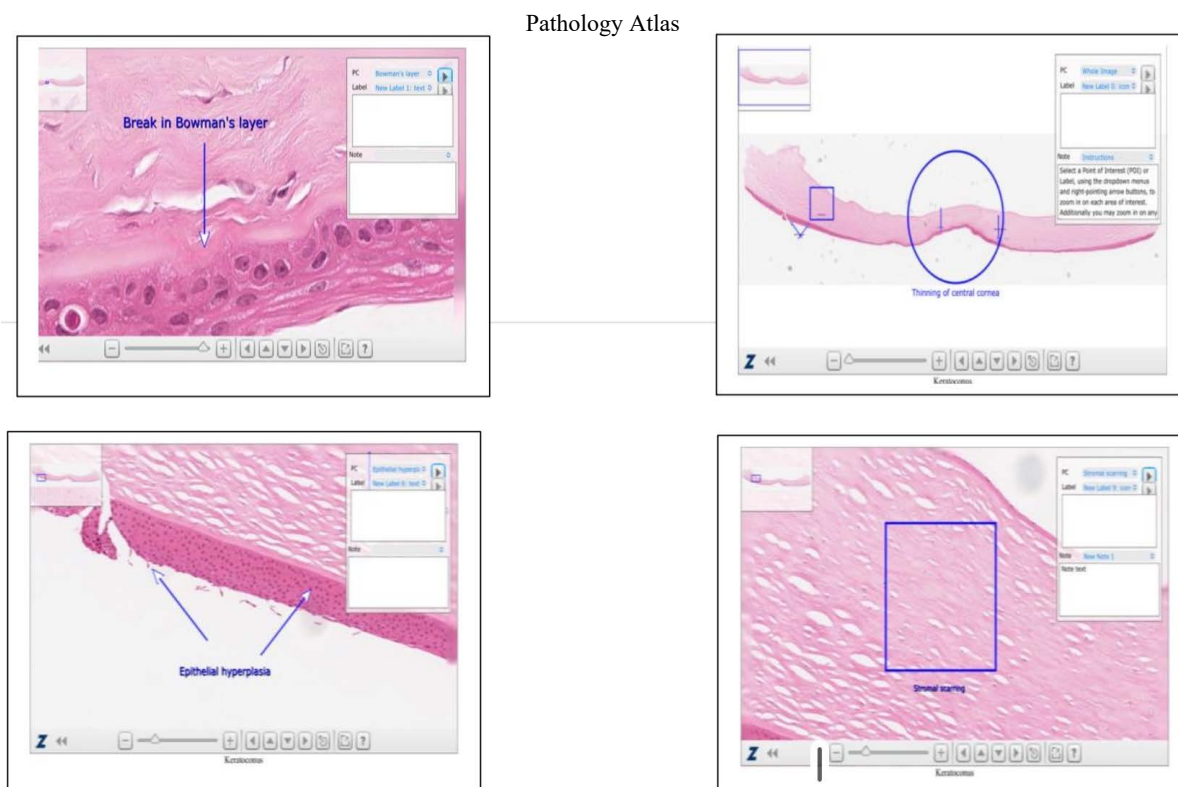


Figure 7: Histopathological pictures are from American Academy of Ophthalmology's

Although it is not an inflammatory disease, inflammatory mediators are found to play a role in association with KC progression. A study conducted by Lema et al; studied tears of 30 eyes already diagnosed as keratoconus and 30 eyes of sub clinical KC and 20 eyes of control subjects, found that IL-6 and TNF-alpha are overexposed in the tears of subclinical and KC eyes, Increased MMP-9 levels were found only in the KC eye in which that can be used as evidence of the association of KC with chronic inflammatory processes in the pathophysiology⁹. The diagnosis of KC is mainly clinically and the clinical signs are constellating the diagnosis criteria. The clinical signs of KC include hemosiderin arc line (Fleischer's ring), Vog't stria which is the vertical lines at the descemet's membrane, V shaped deformation of the lower eyelid (Munson's sign), Rizzuti's sign¹⁰.

Treatment of KC is variable with many strategies and modalities depending on severity¹⁰.

Macular corneal dystrophy (MCD) along with the corneal stromal dystrophies Lattice and Granular corneal dystrophy considered most

common corneal dystrophies in KSA. Macular corneal dystrophy is the commonest¹¹. It was first noticed and described by Oskar Fehr, in 1904. Histopathologically, Dr. Jones and Dr. Zimmerman studied MCD and found that there was an abnormal accumulation of glycosaminoglycans in the stroma, bowman's and Descemet's membrane. Inherited as an Autosomal recessive as per Bucklers¹². Onset in the first decade of life and then progresses causing severe visual impairment 20/40 to 20/200. Presentation includes, photophobia, pain, irritation, decreased corneal sensitivity, recurrent corneal erosions, with diffused haziness which is whitish in color due to deposition of glycosaminoglycans in anterior and posterior stroma of cornea, poorly demarcated and extending from limbus to limbus. Pearls of treatment are Penetrating keratoplasty, Deep anterior Lamellar keratoplasty, Femtosecond laser-assisted lamellar keratectomy, Phototherapeutic Keratectomy and mitomycin C. A promising Gene therapy¹².

Era of COVID 19 is deferent than previous outbreaks of infectious disease ever passed on humanity's history, where it impacted the whole world in all aspects. Nevertheless, health systems worldwide were

impacted deeply with this pandemic. Health care systems collapsed in many countries due to loss of precious personnel, and shortage of equipment¹³. In ophthalmology; the progression of KC in association with COVID-19 was noticed. In which there was a delay of care provided to the patients which resulted in a progression of the disease and worsening of the best corrected visual acuity¹⁴. Concomitant presentation of KC and MCD was reported from different countries world widely. Nevertheless, this is the first case reported in KSA Riyadh^{17,18}. Concomitant presentation of KC and MCD post COVID 19 respiratory infection and viral conjunctivitis to our knowledge was not reported yet. nonetheless, we now know that there are inflammatory mediators playing rule in both COVID 19 and KC progression. Such as IL6 and TNF a. these two mediators were the strongest 2 markers in predicting severity and survival of COVID 19 infected patients¹⁹.

CONCLUSION

Opening the floor to study COVID's implication over ectatic corneal diseases in either accelerating sub-clinical and mild clinical to severe clinical presentations, or a direct causation between COVID-19 and corneal ectasia, additionally reporting a coexistence of the keratoconus and macular stromal corneal dystrophy simultaneously.

Authorship Contribution: All authors share equal effort contribution towards (1) substantial contributions to conception and design, acquisition, analysis and interpretation of data; (2) drafting the article and revising it critically for important intellectual content; and (3) final approval of the manuscript version to be published. Yes.

Consent: A written informed consent been signed by the patient and the family while registration to King Abdulaziz University Hospital, a copy will be provided to the editor upon request.

Potential Conflict of Interest: None.

Competing Interest: None.

Acceptance Date: 10 March 2023

REFERENCES

1. <https://eyewiki.aaopt.org/Keratoconus>.
2. Alabdulmunem MA. Keratoconus in Saudi Arabia: A Retrospective Study at a Tertiary Eye Care Institution (KKESH). *Invest Ophthalmol Vis Sci* 2009;50(13):3536.
3. Hashemi H, Heydarian S, Hooshmand E, et al. The Prevalence and Risk Factors for Keratoconus: A Systematic Review and Meta-Analysis. *Cornea* 2020;39(2):263-70.
4. Gupta PK, Stinnett SS, Carlson AN. Prevalence of sleep apnea in patients with keratoconus. *Cornea* 2012;31(6):595-9.
5. Maumenee IH. The cornea in connective tissue diseases. *Ophthalmology* 1978;85(10):1014-7.
6. Donnenfeld ED, Perry HD, Gibraltar RP, et al. Keratoconus associated with floppy eyelid syndrome. *Ophthalmology* 1991;98(11):1674-8.
7. Zemba M, Zaharia AC, Dumitrescu OM. Association of retinitis pigmentosa and advanced keratoconus in siblings. *Rom J Ophthalmol* 2020;64(3):313-21.
8. Hashemi H, Mirafteb M, Amanzadeh K, et al. Keratoconus detection by novel indices in patients with Down syndrome: a cohort population-based study. *J Ophthalmol* 2020;64(3):285-91.
9. Lema I, Sobrino T, Durán JA, et al. Subclinical keratoconus and inflammatory molecules from tears. *Br J Ophthalmol* 2009;93(6):820-4.
10. Romero-Jiménez M, Santodomingo-Rubido J, Wolffsohn JS. Keratoconus: a review. *Cont Lens Anterior Eye* 2010;33(4):157-66.
11. Alzuhairy S, Alkatan HM, Al-Rajhi AA. Prevalence and histopathological characteristics of corneal stromal dystrophies in Saudi Arabia. *Middle East Afr J Ophthalmol* 2015;22(2):179-85.
12. Aggarwal S, Peck T, Golen J, et al. Macular corneal dystrophy: A review. *Surv Ophthalmol* 2018;63(5):609-17.
13. Blumenthal D, Fowler EJ, Abrams M, et al. Covid-19 - Implications for the Health Care System. *N Engl J Med* 2020;383(15):1483-88.
14. Shah H, Pagano L, Vakharia A, et al. Impact of COVID19 on keratoconus patients waiting for corneal cross linking. *Eur J Ophthalmol* 2021;31(6):3490-3.
15. Sen M, Honavar SG, Sharma N, et al. COVID-19 and Eye: A Review of Ophthalmic Manifestations of COVID-19. *Indian J Ophthalmol* 2021;69(3):488-509.
16. Ozturker ZK. Conjunctivitis as sole symptom of COVID-19: A case report and review of literature. *Eur J Ophthalmol* 2021;31(2):NP161-6.
17. Al-Hamdan G, Al-Mutairi S, Al-Adwani E, et al. Bilateral coexistence of keratoconus and macular corneal dystrophy. *Oman J Ophthalmol* 2009;2(2):79-81.
18. Javadi MA, Rafee'i AB, Kamalian N, et al. Concomitant keratoconus and macular corneal dystrophy. *Cornea* 2004;23(5):508-12.
19. Del Valle DM, Kim-Schulze S, Huang HH, et al. An inflammatory cytokine signature predicts COVID-19 severity and survival. *Nat Med* 2020;26(10):1636-43.