Recurrence of keratoconus after deep anterior lamellar keratoplasty following pregnancy

Zisis Gatzioufas¹, Georgios D. Panos², Evangelia Gkaragkani¹, Stylianos Georgoulas¹, Romesh Angunawela¹

¹Moorfields Eye Hospital, London EC1V 2PD, United Kingdom
²Department of Ophthalmology, Ipswich Hospital NHS Trust, Ipswich IP4 5PD, United Kingdom

Correspondence to: Georgios D. Panos. Department of Ophthalmology, The Ipswich Hospital NHS Trust, Heath Road, IP4 5PD, Ipswich IP4 5PD, Suffolk, United Kingdom

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Dear Editor,

Keratoconus is a progressive, non-inflammatory disease of the cornea, which is characterized by marked corneal steepening and thinning². It induces myopia and irregular astigmatism leading frequently to severe visual impairment³. Although several aetiological factors have been implicated in its pathophysiology, the exact mechanisms underlying keratoconus are not fully elucidated yet. Corneal crosslinking is the treatment of choice in order to inhibit the progression of keratoconus, whereas advanced cases require penetrating or lamellar keratoplasty for visual restoration.

Interestingly, sporadic literature reports indicate that keratoconus can recur in the recipient of a corneal transplant². There is a number of reports on recurrence of keratoconus following penetrating keratoplasty, including histopathological confirmation of the disease³⁴. Keratoconus can also recur after lamellar keratoplasty, as shown by other groups⁵⁶. However the complex phenomenon of keratoconus recurrence remains elusive, with some authors debating the true nature of recurrence and others suggesting potential mechanisms that could explain the re-emergence of keratoconus². Basically, it has been proposed that the re-emergence of keratoconus after a latency period is most likely due to migration of the disease from the host to donor cornea⁷, since keratoplasty involves only partial excision of the cornea, and recent research evidence strongly suggests the presence of the pathology in the peripheral host cornea⁸. Other factors such as vigorous eye rubbing and contact lens wear have also been involved in keratoconus recurrence². Nevertheless, the trigger elements that activate the recurrence phenomenon in certain patients, have not been identified yet.

In this case report (informed consent was obtained by the patient) we highlight a patient with keratoconus who underwent deep anterior lamellar keratoplasty and presented recurrence of the disease after pregnancy. Our clinical observation supports the hypothesis that hormonal changes during pregnancy, which are involved in post-laser corneal ectasia and may contribute to development of corneal hydrops⁹¹⁰, may also play a role in the recurrence of keratoconus after keratoplasty.

A 23-year old female with known keratoconus attended our clinic in December 2012. Corneal topography showed the presence of keratoconus stage 3-4 in the right eye (OD) and keratoconus stage 3 in the left eye (OS) (Figure 1). Her vision was 6/24 OD with rigid gas permeable contact lenses (RGPs) and 6/12 OS with RGPs. Kmax was 61 diopters (D) OD and 58 D OS. Corneal thinnest point was 344 microns OD and 355 microns OS. The patient had no history of vigorous eye rubbing and was otherwise healthy. After discussion we decided to list her for right deep anterior lamellar keratoplasty, which was successfully performed in January 2013. Her right vision in February 2014 and after corneal suture removal was 6/9 with RGPs. In April 2014 she underwent penetrating keratoplasty OS (conversion to penetrating keratoplasty after unsuccessful deep anterior lamellar keratoplasty). In January 2015 her vision was 6/9 OD with RGPs and 6/9 OS with RGPs. During the last month of her pregnancy the patient was experiencing gradual decrease of vision OD. She attended our clinic in December 2015 after delivery. On slit-lamp examination both corneal grafts were clear without any signs of rejection. However corneal topography revealed the presence of corneal ectasia in the right corneal graft (Figure 2), whereas the left corneal graft looked normal with mild increase of the keratometric readings. Her vision was 6/12 OD with RGPs and 6/9 OS with RGPs. Kmax was 54 D OD and 46 D OS. Corneal thinnest point was 422 μm OD and 508 μm OS. After discussion with the patient we decided to proceed with corneal crosslinking OD. Her visual acuity 6mo after the treatment was 6/9 OD with RGPs and 6/9 OS with RGPs. No signs of progression were identified until today.
Recurrence of keratoconus following penetrating or lamellar keratoplasty has been infrequently described in literature. Abelson et al. were first to report a histopathologically confirmed case of keratoconus recurrence following keratoplasty. It has been hypothesized that most recurrences of keratoconus resulted from incomplete cone excision, but further evidence confirmed that keratoconus can re-emerge due to migration of the pathology from host to donor cornea. Interestingly, the recurrence latency is considerably shorter after lamellar keratoplasty (average 3-4y) compared to penetrating keratoplasty (average 19y), supporting the clinical hypothesis that recurrences stem from the underlying pathology in the non-excised corneal tissue. However, recurrence of keratoconus could also happen in a reverse manner, by transplanting a donor cornea with keratoconus in a recipient requiring corneal transplantation for reasons other than keratoconus.

Our patient underwent deep lamellar keratoplasty OD in January 2013 and penetrating keratoplasty OS in April 2014. After her pregnancy, she was diagnosed with recurrence of keratoconus OD in December 2015. There is evidence in the literature that hormonal changes occurring during pregnancy may induce corneal ectasia after laser refractive surgery or exacerbate pre-existing keratoconus. Our patient was treated with corneal crosslinking OD and remained stable until today. Her vision during her last follow-up was 6/9 OD with RGP and 6/9 OS with RGP.

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Figure 1 A: Anterior corneal curvature map derived by Pentacam, showing keratoconus stage 3-4 in the right eye and stage 3 in the left eye; B: Corneal thickness map showing significant corneal thinning in both eyes.

Figure 2 Corneal topography A: Anterior corneal curvature map showing irregular astigmatism after right deep anterior lamellar keratoplasty OD in January 2013 and keratoconus stage 3 OS; B: Corneal curvature map showing regular astigmatism after left penetrating keratoplasty OS in April 2014 and irregular astigmatism OD; C: Corneal curvature map showing recurrence of ectasia in the right corneal graft OD in December 2015 and irregular astigmatism OS; D: Corneal thickness map showing decrease of corneal thickness in the ectatic right corneal graft OD and normal corneal thickness in the left corneal graft OS.

The early recurrence of keratoconus OD, manifesting in the last month of pregnancy, is leading us to the conclusion...
that pregnancy could have accelerated the re-emergence of keratoconus in the “vulnerable” right cornea. The left cornea showed non-significant increase of the keratometric readings, but no evident signs of corneal ectasia could be detected. The latter is in agreement with the clinical observation that corneal topographical and biomechanical variations could occur during pregnancy\[12\].

The exact pathophysiological mechanisms underlying the complex interactions between hormonal influences and corneal biomechanics are currently under investigation. However it has been suggested that high levels of oestrogen and relaxin hormones during pregnancy, as well as the frequently observed hypothyroxinaemia during gestation may affect corneal thickness, corneal topography and corneal elasticity, and thereby play a role in the development or progression of keratoconus\[12-13\].

This case report emphasizes on the potential risk for re-emergence of keratoconus during or after pregnancy particularly following lamellar keratoplasty. Physicians should be aware of this rare complication and counsel their patients accordingly.

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REFERENCES