

CASE REPORT

Pregnancy-associated New-onset Progressive Keratoconus with Horizontally Aligned Vogt's Striae in a 36-year-old Female

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ABSTRACT

Aim: The aim of the study is to report a case of pregnancy-associated progressive keratoconus with atypical horizontally aligned Vogt's striae.

Background: Hormonal changes during pregnancy have been proposed as a risk factor for progressive keratoconus. There have been only a few cases of progressive keratoconus diagnosed with progression after pregnancy, without either an existing disease or an attributable cause.

Case description: A 36-year-old New Zealand European woman presented with progressive myopic astigmatism following her first pregnancy. Stigmata of keratoconus were present on clinical examination and included horizontally aligned Vogt's striae. Tomography measurements confirmed the diagnosis with characteristic inferior corneal steepening and ectasia bilaterally. The patient was advised to abstain from eye rubbing, commenced topical olopatadine, and underwent corneal collagen cross-linking following delivery to prevent further keratoconus progression.

Conclusion: Pregnancy-associated progressive keratoconus in a 36-year-old woman was documented after the pregnancy.

Clinical significance: Atypical presentation features that include horizontally aligned Vogt's striae, advanced age at diagnosis, and rapid pregnancy-associated progression in a previously stable patient.

Keywords: Horizontal Vogt's striae, Keratoconus, Pregnancy-associated progression.

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BACKGROUND

Keratoconus (KC) is associated with bilateral progressive corneal thinning and ectasia. Patients typically present in adolescence and early adulthood with blurred vision, myopic shift, and irregular astigmatism. The underlying etiology is not fully understood; however, there are recognized associated risk factors, such as eye rubbing, allergic eye disease, and contact lens wear.¹ Sex hormones may also be a risk factor for the progression of KC.² New-onset, progressive, pregnancy-associated KC has been reported in a small number of cases, excluding patients with preexisting KC or other attributable causes (i.e., refractive surgery or hormone therapy).^{3,4}

CASE DESCRIPTION

We report a case of a 36-year-old European female with a new diagnosis of KC and documented progression associated with her first pregnancy.

She first presented to her optometrist in August 2018, complaining of progressively blurred vision with onset during the pregnancy of her first child five months earlier (March 2018). A significant increase in astigmatism of 2D right and 1.25D left was noted on examination. Refractive history (Table 1) included relatively stable regular astigmatism over the previous four years, measuring 0.0/−2.0 × 80 and +0.25/−1.5 × 82.5 in the right and left eye prior to pregnancy. The patient's medical history included asthma, allergic rhinitis, and eye rubbing. She was not on any medications and there was no family history of corneal ectasia disorders.

Best-corrected visual acuity on presentation was 6/18 (right) and 6/5+1 (left). Slit-lamp examination revealed the stigmata of KC,

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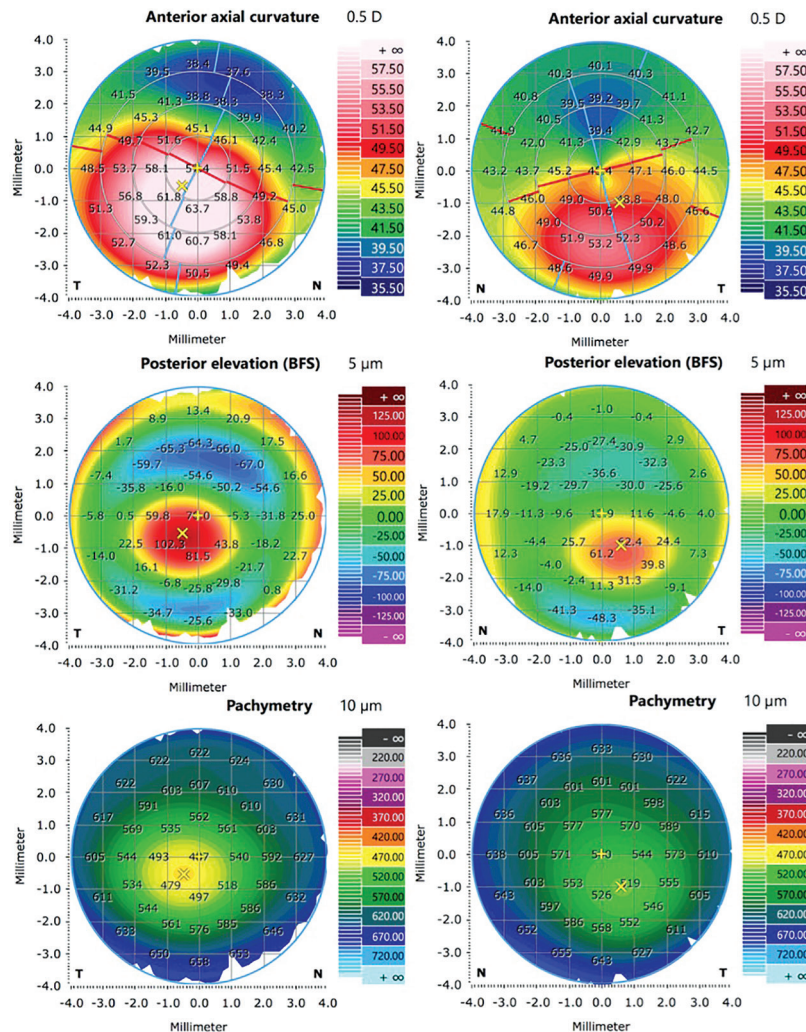
including Munson's sign, scissoring red reflex on retinoscopy, and Vogt's striae (VS) with horizontal alignment.

Tomography imaging (Heidelberg Engineering Anterior®, Germany) confirmed the diagnosis of KC with inferior corneal steepening and ectasia noted bilaterally. KC severity was worse in the right eye with a maximum keratometry and BAD-D of 70.5 D, 16.52 and 54.1D, 6.11 in the right and left eyes, respectively. Tomographic maps are shown in Fig. 1, and the horizontally aligned VS are shown in Fig. 2.

The patient was advised to abstain from eye rubbing and commence treatment with topical olopatadine hydrochloride. Corneal cross-linking was performed in the right and left eyes in February and May 2020.

Table 1: Subjective refraction over time. Diagnosis of KC and progression is noted following the onset of pregnancy. Last menstrual period before pregnancy was August 7, 2017

	<i>Right eye</i>			<i>Left eye</i>		
	<i>Sphere</i>	<i>Cylinder</i>	<i>Axis</i>	<i>Sphere</i>	<i>Cylinder</i>	<i>Axis</i>
05-22-2010	+0.25	-1.50	84	+0.25	-1.25	83
09-27-2014	0.00	-2.00	80	+0.25	-1.50	82.5
08-07-2018	+1.50	-4.00	83	+1.50	-2.75	85
06-07-2019	+3.00	-7.00	70	+3.00	-4.75	95

**Fig. 1:** Tomography (Heidelberg Engineering Anterior®, Germany) demonstrating progressive astigmatism with inferior thinning and steepening

DISCUSSION

We document a newly diagnosed progressive keratoconus (KC) in a 36-year-old woman following pregnancy, with the atypical findings of advanced age of KC and horizontally aligned VS. The criteria for progression were provided by sequential refractions, showing increasing myopic astigmatism over a 10-month period.⁵

Pregnancy may be a risk factor for KC progression, with the onset after pregnancy reported in a few cases of patients without other risk factors.³ Pregnancy is associated with transient refractive

changes and progression of preexisting KC. Hormonal changes during pregnancy have been proposed as a potential explanation for these findings. Sex hormone receptors have been identified in the cornea, suggesting that this tissue may be responsive to changes in the sex hormone levels.⁶ There is increasing evidence that estrogen may have a role in the corneal biomechanics after estrogen was shown to have a stiffness-reducing effect on the cornea, predisposing KC development.⁷ Published case reports of treatments containing estrogen and estrogen-like compounds have

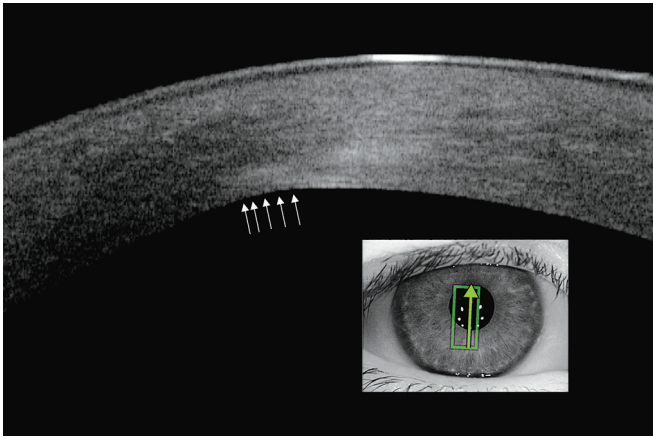


Fig. 2: Corneal optical coherence tomography imaging (Heidelberg Engineering Anterior®, Germany) of the right cornea demonstrating small horizontally aligned folds in the posterior stroma and Descemet's membrane located at the apex of the cone

been linked with KC progression.^{4,8} Estrogen may, in part, regulate the production of corneal extracellular matrix proteins by resident corneal cells, leading to reduced corneal stiffness.² Progesterone and androgen receptors are also present in the cornea; however, the significance of these hormones on corneal stability is not yet understood.² The current case experienced rapid progression associated with pregnancy, which is a period when estrogen levels are elevated then return rapidly to baseline at delivery.⁹

Age is an important prognostic indicator in KC. New-onset KC in older patients is typically less likely to demonstrate progression or require a corneal transplant.¹⁰ The diagnosis is usually made during adolescence, with the average age in a New Zealand setting being 20.9 years old.¹¹ However, progression is not uncommon above age 30, even in apparently stable patients.¹²

VS are vertically aligned striae located in the posterior stroma that can be seen at slit-lamp examination and characteristically disappear when external pressure is applied to the globe. Up to half of the keratoconic patients present with VS that are visible with slit-lamp microscopy.¹³ VS can also be detected using corneal imaging modalities.¹⁴ The striae pattern and microstructure of keratoconic corneas have a different microstructure to the striae of the normal and edematous corneas.¹⁴ It is postulated that the vertical orientation along the cone axis is due to the corneal conical shape and represents degradation of the corneal viscoelasticity and biomechanics.¹⁴ In KC patients, VS are associated with advanced KC, and deteriorating corneal and biomechanical status.^{15,16} To our knowledge, only two cases of horizontal VS are documented in the literature.^{17,18} This case is the third report of horizontal VS in the literature.

CONCLUSION

Our report documents the development and progression of KC following pregnancy, in combination with advanced maternal age and the finding of horizontal VS.

CLINICAL SIGNIFICANCE

Pregnancy is a risk factor for the development and progression of KC, and this occurs during and after pregnancy.

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