Visante in Atypical Posterior Keratoconus

Mohammad Ali Zare, MD[1] • Hadi Z-Mehrjardi, MD, MPH[2]
Fateme Zare, MD[3] • Jafar Oskoie, MD[1]

Abstract

***Purpose*:** To report a case of posterior keratoconus with atypical ocular findings

***Case report*:** An 8-year-old boy was referred to our clinic with the chief complaint of blurred vision in his right eye. The patient had no history prior ocular surgery of trauma. Her family history was also unremarkable. Comprehensive ocular examination was performed.

***Results*:** The patient's best spectacle corrected visual acuity (BSCVA) was $^{3}/_{10}$ in the right eye and $^{8}/_{10}$ in the left eye with refraction of +11.5-0.5×75 (OD) and +4-0.25×25 (OS). On slit-lamp examination of the right eye, normal anterior corneal surface with central posterior corneal depression, pigment deposition and diffuse stromal edema was noticed. Fundoscopic examination and intraocular pressure (IOP) measurements were normal in both eyes. Specular microscopy gave normal corneal endothelial cell count values. Axial topography revealed central flattening of the cornea that corresponded to the area of posterior keratoconus with peripheral steeping in the right eye and asymmetric bow-tie astigmatism in the left eye. Measurement of central corneal thickness with ultra-sound pachymetry showed corneal thickness in the right eye (OD=616 µm and OS=559 µm). Orbscan II demonstrated anterior and posterior Diff of 17 µm and 48 µm respectively, which were within normal limits. Simulated keratometry showed corneal flattening (OD and OS: 37×23 and 36.5×113, respectively). Also, Visante revealed normal anterior surface but the irregularity of posterior corneal surface was typically compatible with the diagnosis of posterior keratoconus.

***Conclusion*:** Atypical forms of posterior keratoconus can be presented with thick cornea and central corneal flattening. Although both Orbscan and Visante can reveal posterior corneal surface abnormalities, the latter is suggested for studying corneal architecture and excavation.

***Keywords*:** Posterior Keratoconus, Visante, Posterior Corneal Flattening

Iranian Journal of Ophthalmology 2011;23(4):61-64 © 2011 by the Iranian Society of Ophthalmology

1. Associate Professor of Ophthalmology, Eye Research Center, Farabi Eye Hospital, Tehran University of Medical Sciences, Tehran, Iran
2. Students' Scientific Research Center, Tehran University of Medical Sciences, Tehran, Iran
3. Ali-Asghar Pediatric Hospital, Tehran University of Medical Sciences, Tehran, Iran

Received: August 28, 2011
Accepted: November 1, 2011

Correspondence to: *Mohammad Ali Zare, MD*
Associate Professor of Ophthalmology, Eye Research Center, Farabi Eye Hospital, Tehran University of Medical Sciences, Tehran, Iran, Tel:+98 21 55414941-6, Email: zaremehrjardy@yahoo.com

Published by Otagh-e-Chap Inc.

Introduction

Posterior keratoconus is a rare, non-inflammatory disorder that is usually considered as a developmental anomaly. The disorder is characterized by abnormal corneal curvature that affects the entire (keratoconus posticus totalis) or a localized portion of posterior cornea (keratoconus posticus circumscriptus). Clinical manifestations usually include unilateral depression of posterior corneal surface with stromal thinning and opacification compatible to the affected area. Corneal abnormalities related to the disorder are usually unilateral, stationary, non-inflammatory and central.[1,2] However, cases with similar clinical findings have been found after ocular trauma.[4]

Since 1930 that Butler[1-3] described posterior keratoconus, several authors have described the disorder. Recently, advent of computer assisted imaging systems had improved the process of diagnosis and classification of the corneal pathologies such as posterior keratoconus. Topography (Orbscan II)[5,6] and tomography (anterior segment OCT; Visante) are two imaging methods for mapping the corneal surface, of which the latter is preferred to be used in patients with posterior keratoconus. To our knowledge it is the first time that Visante is applied in diagnosis of posterior keratoconus.

In this report, we discussed application of Visante in verification of the diagnosis in a patient with atypical posterior keratoconus.

Case report

An 8-year-old boy was referred to our clinic with the chief complaint of blurred vision in his right eye. His parents gave no history of ocular trauma and/or surgery. His family history was unremarkable for any other ocular diseases. Comprehensive ocular examination was performed.

The patient's best spectacle corrected visual acuity (BSCVA) was $^{3}/_{10}$ and $^{8}/_{10}$ with refraction of +11.5-0.5×75 and +4-0.25×25 in his right and left eye, respectively. On slit-lamp examination of the right eye, normal anterior corneal surface with central posterior corneal depression, pigment deposition and diffuse stromal edema was noticed (Figure 1). Fundoscopic examination and intraocular pressure (IOP) measurement were normal in both eyes. Specular microscopy showed normal corneal endothelial cell count.

Ultra-sound pachymetry of central corneal thickness was 616 µm (OD) and 559 µm (OS).

Axial topography showed central corneal flattening that corresponded to the area of posterior keratoconus with peripheral steeping of the right eye and asymmetric bow-tie astigmatism in the left eye. In axial topography Simk was 36.24×90 and 37.53×180 OD and 44.66×4 and 45.37×94 OS (Figure 2).

In the right eye, Orbscan II showed anterior and posterior Diff of 17 µm and 48 µm. But Simk was calculated as 37×23 and 36.5×113 that was very flat. Corneal thickness was 719 µm (Figure 3).

Visante showed normal anterior surface but the irregularity of posterior corneal surface was typically compatible with the diagnosis of posterior keratoconus (Figure 4).

The patient has been treated for amblyopia that resulted in BSCVA increase up to $^{8}/_{10}$ in the right eye. Unfortunately, no therapeutic modality was effective on decreasing corneal thickness.

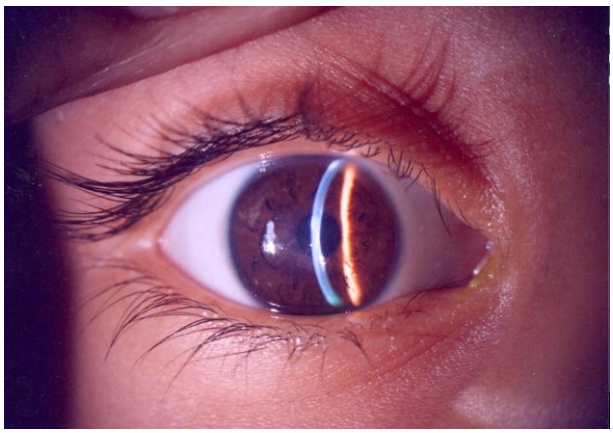

Figure 1. Slit-lamp diffuse illumination of the right eye showed a central annular opacification with corneal edema

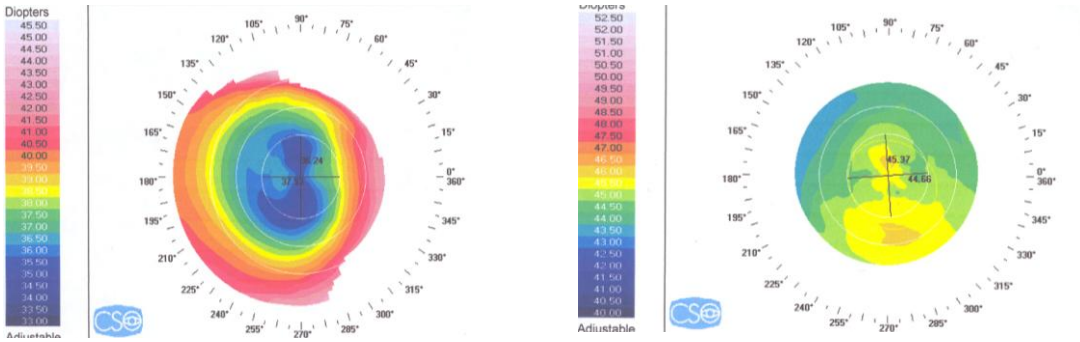

Figure 2. OD: Axial topography showed an abrupt localized central flatness with corneal keratometry of 36.67. OS: Corneal keratometry was 46.04.

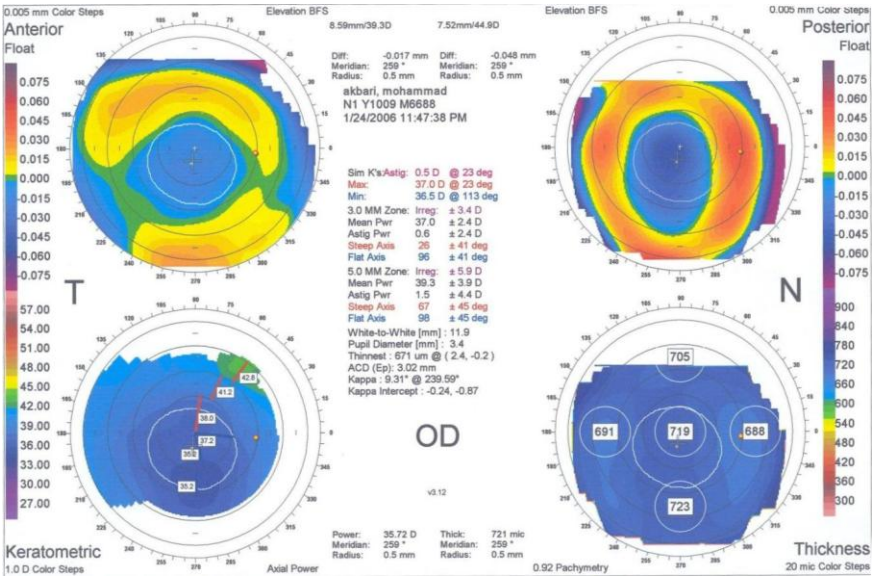

Figure 3. Orbscan II of the right eye showed a very flat and thick cornea

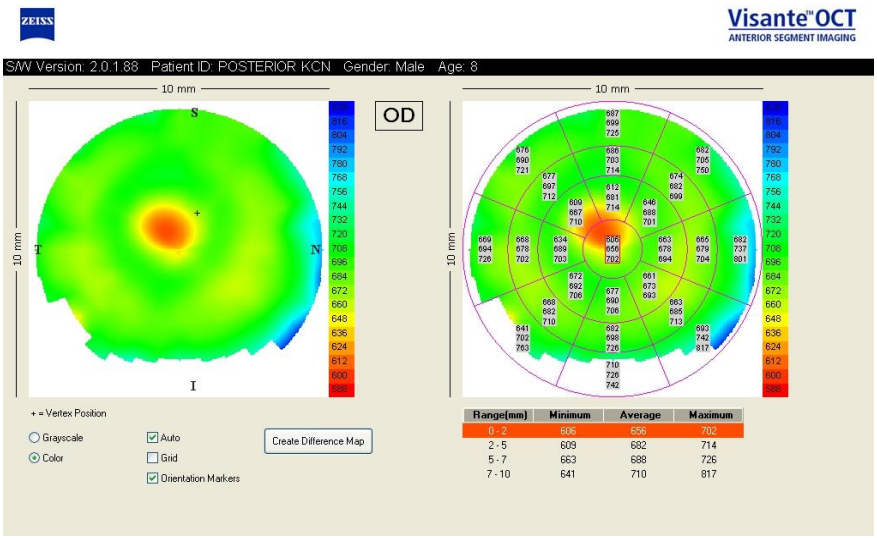

Figure 4. Mapping of corneal thickness. Visante demonstrated irregularity of posterior corneal surface

Discussion

Posterior keratoconus was first described by Butler in 1930. Since then, several reports have focused on giving more complete description of the anomaly and its classification. Posterior keratoconus is mostly unilateral and centrally localized with corresponding posterior stromal opacity. It is rare and can be associated with other ocular diseases such as glaucoma, iris atrophy and lens opacity[7] and/or other systemic abnormalities.[8] Various theories have been proposed as the etiology of posterior keratoconus like fetal development arrest, delayed separation of lens from cornea and mesodermal wave migration.[9] In addition, familial[10] and secondary traumatic cases of this anomaly have been reported. Visual loss is not progressive and is mainly moderate. Generally posterior corneal curvature is focally steeper than normal. However, subtle abnormalities of the posterior corneal surface may be undetected. Posterior corneal curvature changes may have a limited effect on visual acuity and therefore posterior keratoconus is usually considered a harmless corneal disease with no significant changes in refraction.

Previously reported cases of posterior keratoconus were mostly presented with steepening and thinning of the central cornea. But, as follows, we found different ocular features in our case:

1- The right eye was highly hyperopic (refraction OD: +11.5-0.5×75 and OS: +4-0.25×25).
2- Thick cornea (corneal thickness OD= 616 µm and OS= 559 µm, with normal cell count by specular microscopy OD= 3250 and OS= 3450)
3- Central Flatness of cornea with peripheral steepening (Orbscan II showed normal anterior corneal surface, posterior corneal surface flatness and thickened cornea).

Previous studies used Orbscan II for detecting abnormalities of the cornea in patients with posterior keratoconus. A new imaging modality -Visante- have demonstrated its' higher ability in detecting subtle irregularities of anterior and posterior corneal surfaces and architecture. In our case Visante clearly showed normal anterior surface and abnormal posterior surface which typically was compatible with posterior keratoconus. In this regard, Visante seems to be more applicable in detecting the architecture of cornea especially its' posterior surface abnormalities.

Conclusion

We believe that applying Orbscan II and Visante imaging systems are useful for the analysis of anterior and posterior corneal surfaces and its architecture but as it is Orbscan slightly over estimates the corneal thickness. It can be used not only to detect abnormalities in corneal shape such as posterior keratoconus, but it can be used also to detect the subtle corneal irregularities before and after refractive surgeries.

References

1. Butler TH. Keratoconus posticus. Trans Ophthalmol Soc UK 1930;50:551.
2. Mannis MJ, Lightman J, Plotnik RD. Corneal topography of posterior keratoconus. Cornea 1992;11(4):351-4.
3. Côté MA, Gaster RN. Keratohematoma leading to acquired posterior keratoconus. Cornea 1994;13(6):534-8.
4. Jacobs HB. Traumatic keratoconus posticus. Br J Ophthalmol 1957;41(1):40-1.
5. Rao SK, Padmanabhan P. Posterior keratoconus. An expanded classification scheme based on corneal topography. Ophthalmology 1998;105(7):1206-12.
6. Charles N, Charles M, Croxatto JO, et al. Surface and Oorbscan II slit-scanning elevation topography in circumscribed posterior keratoconus. J Cataract Refract Surg 2005;31(3):636-9.
7. Krachmer JH, Rodrigues MM. Posterior keratoconus. Arch Ophthalmol 1978;96(10):1867-73.
8. Streeten BW, Karpik AG, Spitzer KH. Posterior keratoconus associated with systemic abnormalities. Arch Ophthalmol 1983;101(4):616-22.
9. Greene PB. Keratoconus posticus circumscriptus; report of a case. Arch Ophthal 1945;34(5):432.
10. Haney WP, Falls HF. The occurrence of congenital keratoconus posticus circumscriptus in two siblings presenting a previously unrecognized syndrome. Am J Ophthalmol l961;52:53-7.