Scheimpflug and Optical Coherence Tomography Analysis of Posterior Keratoconus

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Purpose: To report a case series of 7 eyes of 6 patients with posterior keratoconus, evaluating corneal Scheimpflug tomographic changes and anterior-segment optical coherence tomography (OCT).

Methods: In our descriptive study, 6 patients were diagnosed with posterior keratoconus: 5 unilateral (7-, 33-, and 42-year-old males and 64- and 60-year-old female) and 1 bilateral (45-year-old female). Patients were diagnosed with slit-lamp examination, which revealed corneal opacity with an underlying posterior corneal depression. Additional analysis with anterior-segment OCT and Scheimpflug tomography evaluation was performed.

Results: Localized paracentral posterior keratoconus was diagnosed in 7 eyes. Scheimpflug images demonstrated posterior corneal depression. Clinical findings were examined by OCT. Genetic analysis revealed no alterations or associated syndromes. All patients were amblyopic in the affected eye, and no surgery was offered to improve their visual acuity.

Conclusions: Posterior keratoconus is a rare noninflammatory condition usually present at birth and sometimes related to developmental abnormalities. Posterior keratoconus is usually unilateral and can present as a generalized or localized change in posterior corneal curvature.

Key Words: posterior keratoconus, Scheimpflug tomography

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Posterior keratoconus was first described by Butler in 1930.[1] It is a rare corneal noninflammatory disorder, usually unilateral and nonprogressive, characterized by posterior corneal depression. It can be secondary to a developmental defect, and it is also considered to be part of anterior-segment dysgenesis.[2] Acquired cases because of corneal trauma have also been reported.[3,4]

Duke-Elder classified posterior keratoconus as keratoconus *posticus generalis* and keratoconus *posticus circumscriptus*.[5,6] In the *generalis* group, the entire posterior corneal surface is affected, and in the *circumscriptus* group, the defect is localized. Both can be associated with corneal thinning, stromal opacity, and a Fleischer ring surrounding the affected area.[2] Generally, the anterior corneal surface is not compromised and does not progress to anterior keratoconus, although changes in the anterior surface have been observed.[2,6,7]

Systemic anomalies associated with posterior keratoconus are growth retardation, cleft lip, brachydactyly, and broad nose. Also, ophthalmic alterations associated with this entity are upward displacement of the lateral canthi, retinal sclerosis, aniridia, ectropion uvea, hyperteleorism, ectopia lentis, and glaucoma.[2,8]

MATERIALS AND METHODS

This is a descriptive study of 6 patients diagnosed with posterior keratoconus. Slit-lamp examination, anterior-segment optical coherence tomography (OCT), Scheimpflug tomography, and genetic evaluation were performed. The study was approved by the Institutional Review Board and Ethics Committee. All subjects signed an informed consent.

RESULTS

Case 1

A 33-year-old male presented with decreased visual acuity in the left eye. He had a history of penetrating keratoplasty in his right eye because of corneal opacity of unknown origin and strabismus surgery 15 years ago with secondary amblyopia and nystagmus. There was no history of intrauterine infections, trauma, or systemic diseases. Best-corrected visual acuity (BCVA) was hand motion in the right eye and 20/50 in the left eye with +3.50 −4.00 × 30 refraction, respectively. Slit-lamp examination of the left eye revealed a corneal anomaly (Fig. 1A), which was confirmed by OCT images (Fig. 2A). Corneal tomography showed no steepening of the anterior corneal curvature; instead, flat keratometries (37.0/37.9 × 103 degrees) were observed. He was managed conservatively with a contact lens.

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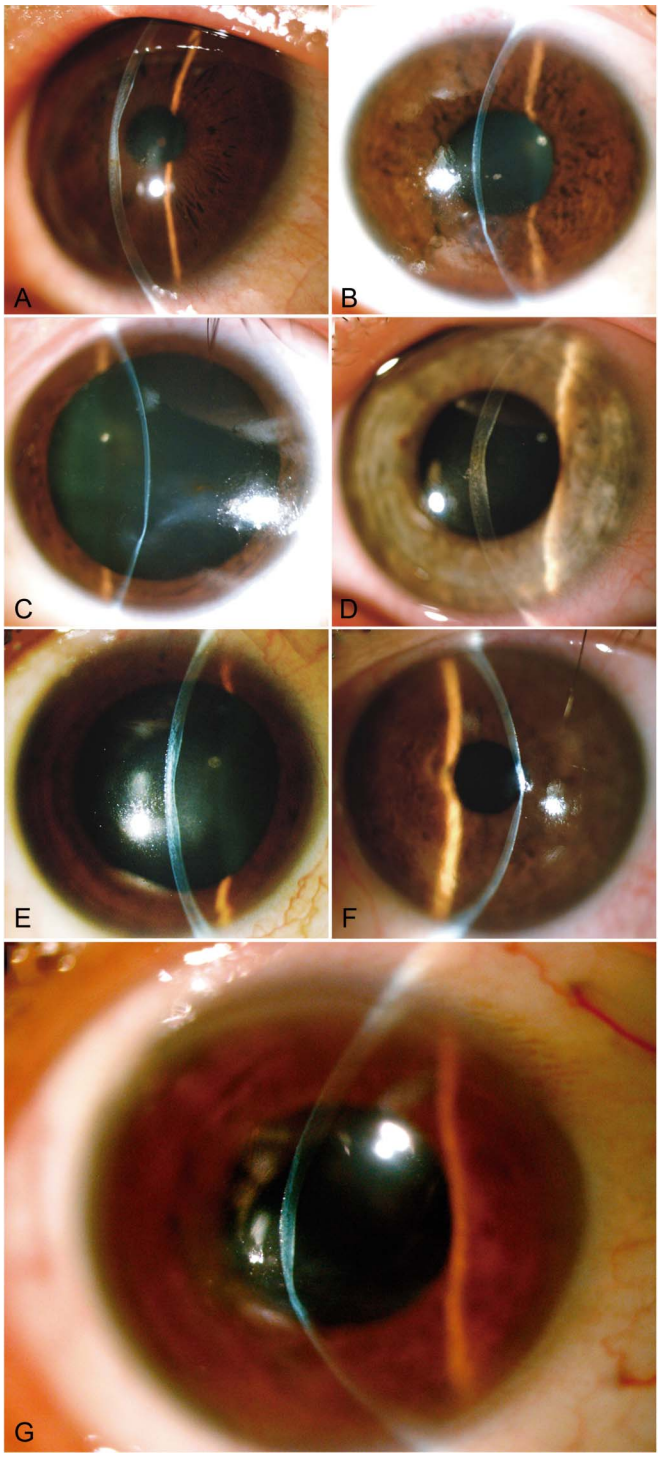

FIGURE 1. Slit-lamp examination revealed posterior corneal depression with corneal thinning corresponding to the area of corneal opacity in all cases (A–G).

Case 2

A 45-year-old female presented with decreased visual acuity in both eyes. The patient had a history of rheumatoid arthritis and used spectacles to correct high compound myopic astigmatism since she was 33 year old. BCVA was 20/400 with −18.25 −3.75 × 180 refraction and 20/300 with −10.75 −2.50 × 180, respectively. Bilateral slit-lamp examination revealed corneal opacity with posterior corneal depression (Figs. 1B, C) confirmed by OCT images (Figs. 2B, C). Scheimpflug corneal tomography reported bilateral anterior curvature steepening, with keratometries of 44.2/49 × 5 degrees in the right eye and 47.1/49.5 × 158 degrees in the left eye. Scheimpflug images of the left eye are shown in Figure 3.

Case 3

A 7-year-old boy was brought by his mother who noticed a corneal opacity in his left eye. He wore spectacles since he was 5 years old. He presented with BCVA of 20/20 with +0.50 −1.50 × 180 in his right eye, and 20/100 with +3.25 −2.00 × 160 refraction in his left eye. Normal slit-lamp examination was performed in the right eye, and central corneal opacity of 2 × 2 mm was associated with corneal thinning and posterior depression in his left eye (Fig. 1D). Anterior-segment OCT confirmed posterior corneal keratoconus (Fig. 2D).

Case 4

A 42-year-old male presented with poor visual acuity in his right eye since childhood. He had a history of exotropia and no other associated diseases. His BCVA was 20/400 with −4.25 −2.25 × 90 refraction in the right eye and 20/20 with −1.50 −0.50 × 180 refraction in the left eye. Examination of the right eye revealed central corneal opacity and thinning (Fig. 1E) with a posterior corneal depression confirmed by high-resolution OCT (Fig. 2F). Flat keratometries (34.1/37.6 × 86 degrees) and changes in posterior curvature were observed on Scheimpflug corneal tomography; examination of left eye was unremarkable.

Case 5

A 64-year-old female consulted for reduced visual acuity in her left eye. Her refraction was −2.25 spherical in both eyes. Her BCVA was 20/50 for the right eye and 20/200 for the left eye. Ophthalmological examination was normal in the right eye. However, a localized central posterior corneal depression with associated thinning and opacity was observed in her left eye (Fig. 1F) and was confirmed by OCT (Fig. 2E). Corneal Scheimpflug tomography reported steep keratometric values (52.1/55.1 × 145 degrees) with changes in posterior curvature map.

Case 6

A 60-year-old female consulted for reduced visual acuity in her right eye. She had a history of penetrating keratoplasty because of keratoconus in her left eye 20 years ago and advanced glaucoma. Right eye refraction was +1.50 −6.50 × 105 degrees with a BCVA of 20/60. Keratometric values in the right eye were 58.78/68.55 × 109 degrees by

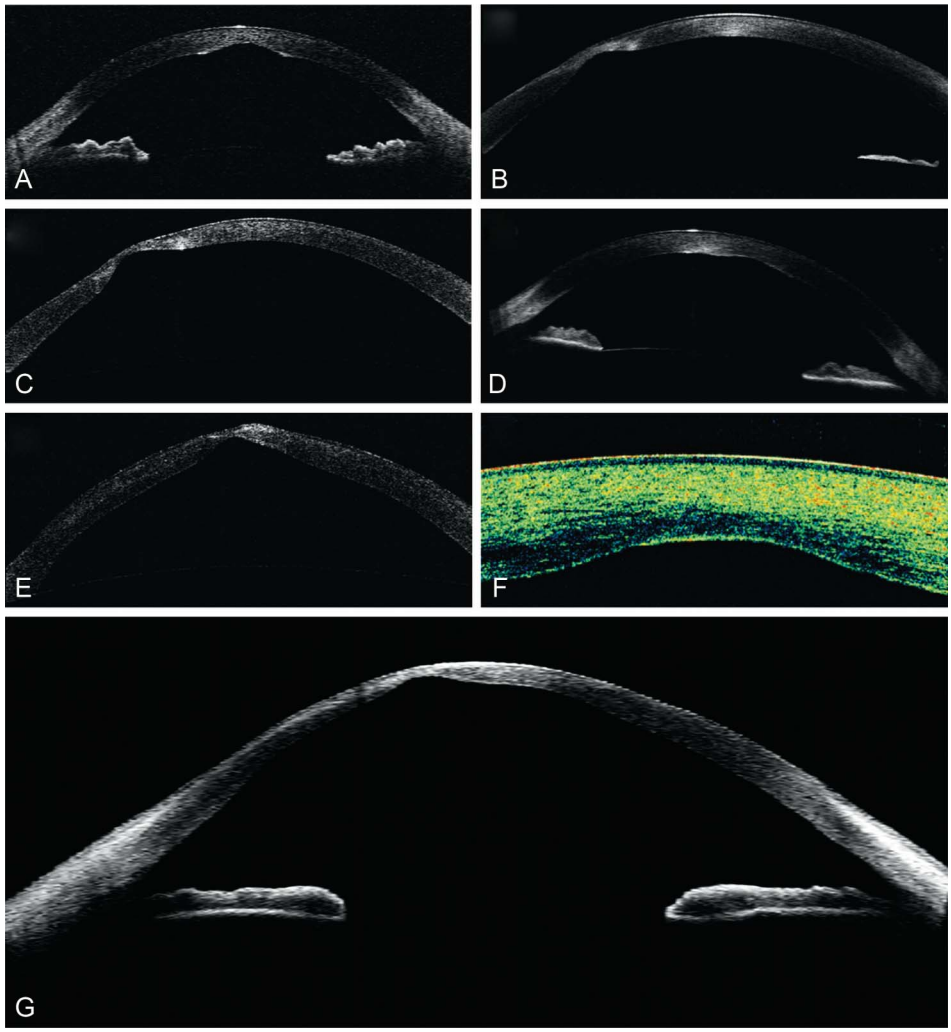

FIGURE 2. Anterior-segment OCT demonstrated central or paracentral posterior corneal depression with corneal thinning (A–E, G). High-resolution OCT revealed similar findings as the other cases (D).

Scheimpflug corneal tomography. In her left eye, BCVA was 20/300 with contact lens. Clinical examination of the right eye revealed posterior corneal depression with associated thinning (Fig. 1G), confirmed by OCT (Fig. 2G).

DISCUSSION

Posterior keratoconus is a rare corneal disorder; although most frequently unilateral, bilateral cases have also been reported.[9] It is usually a nonprogressive disorder that can be sporadic, congenital, or associated with trauma.[2–4] This condition can be classified as generalized posterior keratoconus or circumscribed posterior keratoconus. In the former, there is an increased curvature of the entire posterior corneal surface, whereas in the circumscribed form, only a localized central or paracentral area is affected.[5,6] Posterior depression of the cornea is strongly associated with posterior keratoconus, in contrast with anterior keratoconus where these clinical manifestations scarcely ever occur. We report 1 case of circumscribed bilateral involvement and 5 cases of circumscribed unilateral compromise. Although, we cannot exclude the possibility of bilateral keratoconus in case 1, it is possible that the eye that underwent penetrating keratoplasty was secondary to posterior keratoconus.

Posterior keratoconus has earlier been described as having a normal anterior surface.[4] However, this concept has changed over the years. Mannis et al in 1992 reported anterior surface changes with an area of steepening corresponding to the posterior corneal protrusion observed in one patient using the Topographic Modeling System (TMS; Computed Anatomy Incorporated, New York, NY).[6] Rao et al in 1997 reported a case series of 11 patients with posterior keratoconus; he classified the cases according to different topographic patterns using the TMS system. The localized group was subdivided into central, paracentral, and peripheral types.[5] Topographic changes in generalized posterior keratoconus were a uniform steepening of the corneal surface. In the localized central or paracentral keratoconus, a steepening over the affected area was reported,[5] as observed in our second, fifth, and sixth cases.

Although subepithelial iron deposits are encountered more often in anterior keratoconus, this finding may also be present in posterior keratoconus attributed to an irregularity of the anterior surface.[2,5] Iron deposits surrounding the area of

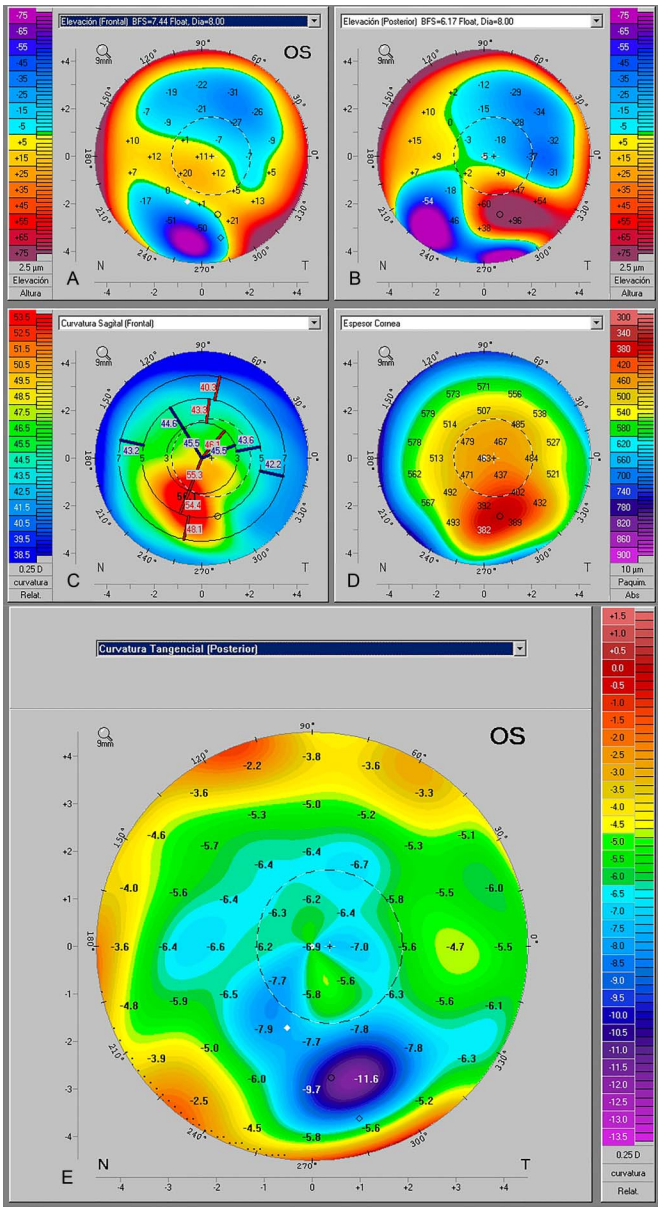

FIGURE 3. Scheimpflug corneal tomography of left eye. Elevation maps revealed inferior temporal areas of abnormal elevations (A, B) corresponding to an area of increased steepening and focal thinning in the anterior sagittal and pachymetry maps (C, D). Posterior corneal curvature is also altered focally (E).

posterior corneal depression were present in 3 cases. Changes on the posterior corneal surface have little effect on the refractive power, and most cases of paracentral or peripheral posterior keratoconus require no surgical treatment.[2] Visual impairment is more common in central, localized keratoconus associated with stromal scarring, which can lead to amblyopia in children. All patients developed amblyopia in the affected eye; thus, no surgical treatment was offered. Posterior keratoconus may be associated with systemic or ophthalmic manifestations.[2,8] However, all our patients underwent a genetic evaluation that revealed no alterations or associated syndromes. Park et al used Orbscan II and Pentacam to obtain the corneal power to calculate an accurate measurement for an intraocular lens.[10] More recently, Tamaoki et al reported a series of 4 cases with posterior keratoconus taking into account anterior and posterior corneal curvatures for intraocular lens power calculations by using partial coherence interferometry and anterior-segment OCT showing good clinical outcomes.[11]

In conclusion, posterior keratoconus is a rare, noninflammatory, nonprogressive disorder with posterior corneal curvature changes, which is the hallmark of this entity. Tomography derived from Scheimpflug and/or anterior-segment OCT imaging provides unique information about this pathology.

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REFERENCES

1. Butler TH. Keratoconus posticus. *Trans Ophthalmol Soc UK.* 1930;50:551–556.
2. Krachmer JH, Feder RS, Belin MW. Keratoconus and related noninflammatory corneal thinning disorders. *Surv Ophthalmol.* 1984;28:293–322.
3. Coté MA, Gaster RN. Keratohematoma leading to acquired posterior keratoconus. *Cornea*. 1994;13:534–538.
4. Williams R. Acquired posterior keratoconus. *Br J Ophthalmol.* 1987;71:16–17.
5. Rao SK, Padmanabhan P. Posterior keratoconus. An expanded classification scheme based on corneal topography. *Ophthalmology*. 1998;105:1206–1212.
6. Mannis MJ, Lightman J, Plotnik RD. Corneal topography of posterior keratoconus. *Cornea*. 1992;11:351–354.
7. Charles N, Charles M, Croxatto JO, et al. Surface and Orbscan II slit-scanning elevation topography in circumscribed posterior keratoconus. *J Cataract Refract Surg.* 2005;31:636–639.
8. Streeten BW, Karpik AG, Spitzer KH. Posterior keratoconus associated with systemic abnormalities. *Arch Ophthalmol.* 1983;101:616–622.
9. Rejdak R, Nowomiejska K, Haszcz D, et al. Bilateral circumscribed posterior keratoconus: visualization by ultrasound biomicroscopy and slit-scanning topography analysis. *J Ophthalmol.* 2012;2012:587075.
10. Park do Y, Lim DH, Chung TY, et al. Intraocular lens power calculations in a patient with posterior keratoconus. *Cornea*. 2013;32:708–711.
11. Tamaoki A, Kojima T, Hasegawa A, et al. Intraocular lens power calculation in cases with posterior keratoconus. *J Cataract Refract Surg.* 2015;41:2190–2195.