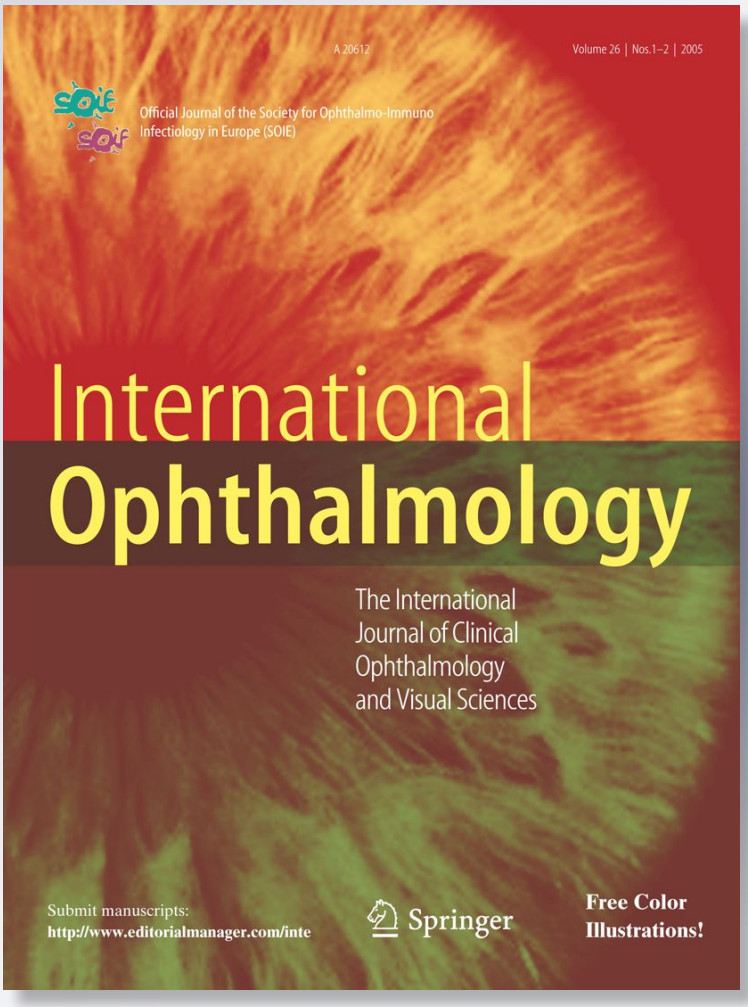
Cornea plana associated with open-angle glaucoma: a case report

Bilge Ozturk Sahin, Goktug Seymenoglu & Esin F. Baser

International Ophthalmology
The International Journal of Clinical Ophthalmology and Visual Sciences

ISSN 0165-5701
Volume 31
Number 6

Int Ophthalmol (2012) 31:505-508
DOI 10.1007/s10792-011-9490-4



Springer

Int Ophthalmol (2011) 31:505–508
DOI 10.1007/s10792-011-9490-4

CASE REPORT

Cornea plana associated with open-angle glaucoma: a case report

Bilge Ozturk Sahin · Goktug Seymenoglu · Esin F. Baser

Received: 20 October 2011 / Accepted: 22 November 2011 / Published online: 11 December 2011

Abstract Cornea plana is a rare disease in which the cornea is flattened with a low refractive power. In addition to these features, hypermetropia, deep central corneal opacities, hazy corneal limbus, peripheral scleralization of the cornea and early arcus senilis can also be seen. Closed-angle glaucoma may occur as a result of shallow anterior chamber and narrow angle; however, open-angle glaucoma has also been reported in these patients. Measuring the real intraocular pressure (IOP) value of such eyes is difficult since tonometers are affected by corneal curvature. Therefore, the diagnosis of glaucoma may be delayed for a long time. In this case report we aimed to present a case of cornea plana with early open-angle glaucoma and to investigate which tonometer was appropriate for measuring the correct IOP value in such eyes.

Keywords Cornea plana · Preperimetric glaucoma · Ocular response analyzer

B. O. Sahin (✉)
Department of Ophthalmology, Akhisar State Hospital, 45010 Manisa, Turkey
e-mail: bilge@doctor.com

G. Seymenoglu · E. F. Baser
Department of Ophthalmology, Faculty of Medicine, Celal Bayar University, Manisa, Turkey

Introduction

Cornea plana is a rare congenital disease characterized by a flattened corneal curvature and low refractive power. It can be inherited as an autosomal dominant or autosomal recessive trait. The gene for both forms of cornea plana was mapped on the long arm of chromosome 12 [1, 2]. Mutations in keratocan gene (*KERA*) have been shown to be responsible for autosomal recessive cornea plana [3].

The autosomal dominant form of the disease is characterized by little loss of visual acuity and the corneal refractive power varies between 38 and 42 diopters. The autosomal recessive form is clinically more severe, results in a significant loss of visual acuity and the refractive power of the cornea is reduced to between 25 and 35 diopters [4, 5]. In addition to bilateral flattened corneal surface, other features such as hypermetropia, deep central corneal opacities, hazy corneal limbus, peripheral scleralization of the cornea, early arcus senilis, and shallow anterior chamber may also be seen, especially in the autosomal recessive form [4]. As a result of the shallow anterior chamber and narrow angle, closed-angle glaucoma may occur in these patients [1, 4, 6]. Open-angle glaucoma has also been reported in cornea plana patients [4, 7]; however, it is difficult to measure the real intraocular pressure (IOP) values of such eyes since tonometers are affected by extreme corneal curvatures. Therefore, glaucoma may not be diagnosed for a long time.

In this case report we aimed to present a case of cornea plana with early open-angle glaucoma and to investigate which tonometer was appropriate for measuring the correct IOP value in such eyes.

Case report

A 16-year-old female patient was referred to our clinic with suspicion of glaucoma. The referring physician reported that after routine cycloplegic examination, her IOP was 40 mmHg in both eyes with the non-contact tonometer (NCT).

We made a baseline examination including determination of visual acuity, manifest and cycloplegic refraction, slit-lamp examination and fundus examination. Best-corrected Snellen visual acuity was 7/10 with +3.50 (+1.00 × 25°) refraction in the right eye and 10/10 with +3.00 (+1.25 × 140°) refraction in the left eye. Cyloplegic refraction was +8.75 (+0.75 × 25°) diopters in the right eye and +6.50 (+1.00 × 135°) diopters in the left eye. Keratometric readings were 10.35 × 32°/10.05 × 15° mm in the right eye and 10.57 × 12°/10.30 × 102° mm in the left eye. Slit-lamp examination showed central corneal opacity and flattened corneal curvature in both eyes (Figs. 1 and 2). Ophthalmoscopic examination revealed that the cup-to-disc ratio was 0.4 in the right eye and 0.6 in the left eye. Goldmann applanation tonometer (GAT) readings were 20 mmHg bilaterally. Central corneal thickness (CCT) with ultrasonic pachymeter was 493 μm in the right eye and 494 μm in the left eye. Gonioscopy of the angle revealed open angles (Shaffer grade 3) in every quadrant of both eyes. Axial length was determined as 23.10 and 23.78 mm, and anterior chamber depth was 2.35 and 2.44 mm for the right and left eyes, respectively.

IOP measurements with other tonometers available in our unit were attempted. With the NCT, IOP readings were 25 mmHg in the right eye and 21 mmHg in the left eye. Attempts at measuring IOP with the Pascal dynamic contour tonometer (PDCT) and Tonopen were unsuccessful; no reliable readings could be obtained with these tonometers. With the ocular response analyzer (ORA) (Reichert) IOP measurements were as follows: corneal compensated IOP (IOPcc) was 24.5 mmHg in the right eye and 23.2 mmHg in the left eye, Goldmann-correlated IOP (IOPg) was 23.5 mmHg in the right eye and 23.7 mmHg in the left eye. With the ORA, corneal hysteresis and corneal resistance factor were 8.8 and 8.4 mmHg in the right eye, respectively, and 8.1 and 8.2 mmHg in the left eye, respectively.

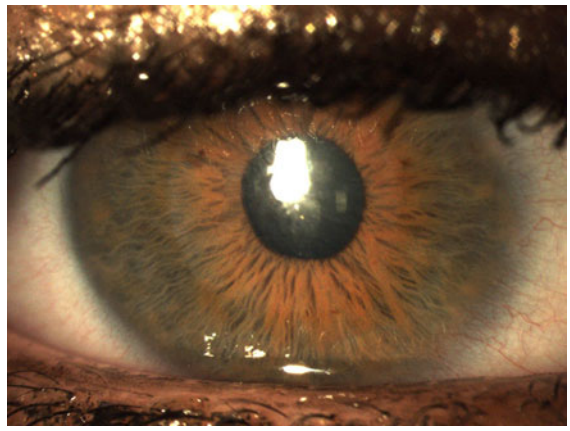


Fig. 1 Central corneal opacity

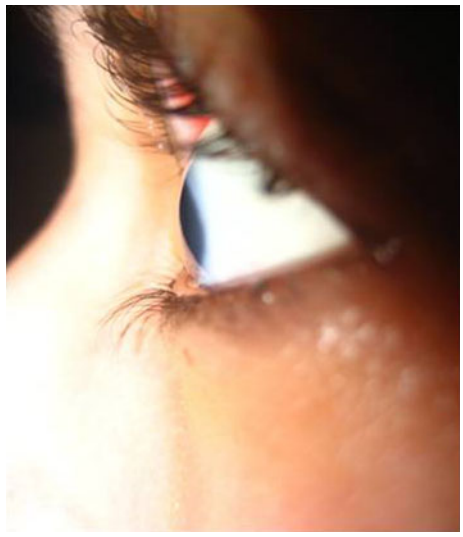


Fig. 2 Flattened corneal curvature

Bilateral standard automated perimetry and blue-on-yellow perimetry with the Humphrey perimeter were normal. Examination with the Heidelberg Retina Tomography III (HRT-III) showed that Moorfields regression analysis was outside normal limits and the glaucoma probability score was outside normal limits in both eyes. Retinal nerve fiber layer thickness analysis with optical coherence tomography (OCT) displayed localized thinning in both eyes.

With the above findings the patient was diagnosed as cornea plana associated with preperimetric glaucoma. We started antiglaucoma medication (timolol maleate 0.5%) to both eyes.

Discussion

Cornea plana is a rare corneal disease which can be inherited as either autosomal dominant or autosomal recessive. These two forms of the disease can usually be distinguished based on phenotype since the recessive form is a more severe disease than the dominant form. It has been shown that *KERA* mutations are responsible for the recessive form. This mutation results in lack of keratocan which is a small leucine-rich proteoglycan. This molecule is thought to be important in developing and maintaining corneal transparency. Keratocan knockout mice have been showed to have clear corneas with a thin corneal stroma, narrower angle between the cornea and iris, abnormal collagen fibril spacing and larger stromal fibril diameters [8].

Our patient had bilateral flat corneas, low corneal refractive power and central corneal opacity which are typical of recessive cornea plana. She had been referred to us for suspicion of glaucoma. Glaucoma associated with cornea plana has been described in several previous publications [4, 7]. Risk of increased narrow-angle glaucoma is plausible as a result of shallow anterior chamber and narrow angle. Forsius et al. found glaucoma in 4 eyes of 78 cornea plana patients, with the chamber angle open in most of the patients [4]; three of these patients were blind due to glaucoma. Thus, there is an association between cornea plana and glaucoma, either the narrow-angle or open-angle type.

Since tonometric measurements may be affected by extreme values of corneal curvature, we aimed to search for the most reliable tonometer for measuring the IOP in our patient. If IOP is erroneously measured lower than the actual value, glaucoma may not be diagnosed for a long time, i.e., until optic nerve head changes occur. Hafner et al. reported a 66-year-old male with cornea plana and IOP readings consistently below 21 mmHg with GAT where glaucoma was not suspected until typical glaucomatous optic nerve head changes appeared [7]. They converted the GAT values according to the flat corneal power (addition of 1 mm Hg to the applanatory values per 3 diopters decreased corneal power), and suggested using a mathematical correction of the applanatory IOP in order to diagnose glaucoma early enough.

Today GAT is still the golden standard tonometer for measuring IOP. Some studies have investigated whether corneal properties such as CCT, corneal curvature and astigmatism affect the GAT measurements [9, 10]. It has been reported that corneal curvature affects GAT measurements by approximately 0.18–0.34 mmHg/diopter (1.14–2.0 mmHg/mm radius) [11, 12]. On the other hand, others have suggested that curvature has no effect on GAT readings [13–15]. Broman et al. [16] investigated the influence of some corneal characteristics on the measurement of IOP with 3 different tonometers (GAT, TonoPen and the ORA) and found that the ORA was least affected by differences in corneal curvature.

Besides having flat corneal curvature, our patient had a thin CCT—493 and 494 μm in the right and left eyes, respectively. This can also possibly affect the GAT measurement. There are many published studies indicating that the CCT affects IOP measurements [10, 11, 17, 18]. According to these studies a 10-μm increase in CCT results in 0.23–0.27 mmHg increase in IOP [11, 17, 18]. In other studies this effect is up to 0.40–0.78 mmHg per 10 μm [9, 19].

Besides the NCT and GAT, we also attempted to measure the IOP of our patient with the PDCT and the Tonopen unsuccessfully. The patient's flat corneal curvature was beyond the measurement capability of both devices.

Assuming that the low corneal curvature and the thin CCT in our patient had additive effects, we speculated that we could be measuring our patient's IOP about 5–8 mmHg lower than the actual value with the GAT. Since we had concerns about the influence of the CCT and corneal curvature on GAT measurements, we used the newly defined tonometer ORA which is claimed to measure IOP independent of corneal structure. The ORA measurements in our patient consistently displayed values above 21 mmHg. She had no visual field defects on both standard achromatic perimetry and blue-on-yellow perimetry; however, the HRT-III and the OCT findings indicated early glaucoma. Thus, we concluded that our patient had preperimetric glaucoma and started antiglaucoma medication (timolol maleate

0.5%). 1 month later, the IOPcc was 17.1 mmHg in the right eye and 16.2 mmHg in the left eye, and the IOPg was 16.2 mmHg in the right eye and 17.6 mmHg in the left with the ORA. She has been under follow-up for over a year without any visual field defects and with stable HRT-III and OCT findings.

In conclusion, cornea plana can be associated with open-angle glaucoma even in young patients. Since the flat corneal surface may lead to false IOP measurements with GAT, we suggest the use of ORA for the diagnosis and follow-up of such patients.

Acknowledgment The authors have no proprietary or financial interest in any materials discussed in this article.

References

1. Tahvanainen E, Forsius H, Karila E, Ranta S, Eerola M, Weissenbach J, Sistonen P, de la Chapelle A (1995) Cornea plana congenita gene assigned to the long arm of chromosome 12 by linkage analysis. Genomics 26:290–293
2. Tahvanainen E, Villanueva AS, Forsius H, Salo P, de la Chapelle A (1996) Dominantly and recessively inherited cornea plana congenita map to the same small region of chromosome 12. Genet Res 6:249–254
3. Pellegata NS, Dieguez-Lucena JL, Joensuu T et al (2000) Mutations in KERA, encoding keratocan, cause cornea plana. Nat Genet 25:91–95
4. Forsius H, Damsten M, Eriksson AW, Fellman J, Lindh S, Tahvanainen E (1998) Autosomal recessive cornea plana: a clinical and genetic study of 78 cases in Finland. Acta Ophthalmol Scand 76:196–203
5. Ebenezer ND, Patel CB, Hariprasad SM, Chen LL, Patel RJ, Hardcastle AJ, Allen RC (2005) Clinical and molecular characterization of a family with autosomal recessive cornea plana. Arch Ophthalmol 123:1248–1253
6. Sigler-Villanueva A, Tahvanainen E, Lindh S, Dieguez-Lucena J, Forsius H (1997) Autosomal dominant cornea plana: clinical findings in a Cuban family and a review of the literature. Ophthalmic Genet 18:55–62
7. Hafner A, Seitz B (2001) Primary open angle glaucoma in cornea plana masked by false normal applanation tonometry (Goldman)—a case report. Klin Monatsbl Augenheilkd 218:621–625
8. Liu CY, Birk DE, Hassell JR, Kane B, Kao WWY (2003) Keratocan-deficient mice display alterations in corneal structure. J Biol Chem 278:21672–21677
9. Whitacre MM, Stein R (1993) Sources of error with use of Goldmann-type tonometers. Surv Ophthalmol 38:1–30
10. Whitacre MM, Stein RA, Hassanein K (1993) The effect of corneal thickness on applanation tonometry. Am J Ophthalmol 115:592–596
11. Gunvant P, Baskaran M, Vijaya L, Joseph IS, Watkins RJ, Nallapothula M, Broadway DC, O'Leary DJ (2004) Effect of corneal parameters on measurements using the pulsatile ocular blood flow tonograph and Goldmann applanation tonometer. Br J Ophthalmol 88:518–522
12. Mark HH (1973) Corneal curvature in applanation tonometry. Am J Ophthalmol 76:223–224
13. Kaufmann C, Bachmann LM, Thiel MA (2004) Comparison of dynamic contour tonometry with goldmann applanation tonometry. Invest Ophthalmol Vis Sci 45:3118–3121
14. Rask G, Behndig A (2006) Effects of corneal thickness, curvature, astigmatism and direction of gaze on Goldmann applanation tonometry readings. Ophthalmic Res 38:49–55
15. Kohlhaas M, Boehm AG, Spoerl E, Pürsten A, Grein HJ, Pillunat LE (2006) Effect of central corneal thickness, corneal curvature, and axial length on applanation tonometry. Arch Ophthalmol 124:471–476
16. Broman AT, Congdon NG, Bandeen-Roche K, Quigley HA (2007) Influence of corneal structure, corneal responsiveness, and other ocular parameters on tonometric measurement of intraocular pressure. J Glaucoma 16:581–588
17. Bhan A, Browning AC, Shah S, Hamilton R, Dave D, Dua HS (2002) Effect of corneal thickness on intraocular pressure measurements with the pneumotonometer, Goldmann applanation tonometer, and Tono-Pen. Invest Ophthalmol Vis Sci 43:1389–1392
18. Ehlers N, Hansen FK, Aasved H (1975) Biometric correlations of corneal thickness. Acta Ophthalmol (Copenh) 53:652–659
19. Ehlers N, Bramsen T, Sperling S (1975) Applanation tonometry and central corneal thickness. Acta Ophthalmol (Copenh) 53:34–43