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Pellucid Marginal Degeneration and Bilateral Corneal Perforation: Case Report and Review of the Literature

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Abstract

Purpose—To review corneal perforation cases in pellucid marginal degeneration and report a case of bilateral spontaneous perforation at presentation in rigid gas permeable contact lens wear and pellucid marginal degeneration.

Methods—Case report and literature review.

Results—The presentation, clinical course, surgical intervention, pathologic analysis, and postoperative outcome of a case of bilateral spontaneous corneal perforation in pellucid marginal degeneration are detailed. All spontaneous corneal perforation cases associated with pellucid marginal degeneration are reviewed.

Conclusions—Pellucid marginal degeneration can lead to spontaneous corneal perforation resulting in the need for urgent therapeutic intervention. A potential for severe ocular morbidity should be considered in all patients with this disorder despite a seemingly stable disease and appropriate gas permeable contact lens wear. Unilateral perforation cases should be watched closely for development of complications in the fellow eye.

Keywords

Pellucid marginal degeneration; Corneal perforation; Keratoplasty; Hydrops

Pellucid marginal degeneration is a relatively uncommon corneal ectatic disorder characterized by inferior peripheral corneal thinning in the absence of infiltration or inflammation. The hallmark of this disorder includes an area of corneal protrusion above the point of maximal thinning rather than within the area of maximal thinning as with keratoconus. A 1 to 2 mm zone of uninvolved normal cornea separates the region of thinning from the peripheral limbus. A characteristic pattern of inferior corneal steepening and subsequent against-the-rule astigmatism is found on corneal topography. A variant of this condition, superior pellucid marginal degeneration, causes similar findings in the superior cornea.

Since Schlaeppli¹ first coined the phrase in 1957, “marginale inférieure pellucide de la corneé,” a number of reports of corneal edema and hydrops seen concurrently with pellucid marginal degeneration have been documented in the literature,^{2–7} yet a limited number of reports presenting with coexistent spontaneous corneal perforation and pellucid marginal degeneration exist.^{8–14} Herein, we describe an unusual presentation of a bilateral corneal perforation in a patient with previously stable pellucid marginal degeneration in the setting of bilateral rigid

gas permeable contact lens wear. To the best of our knowledge, this represents the first report of bilateral spontaneous perforation with pellucid marginal degeneration at presentation. All cases of spontaneous perforation reported in the literature are reviewed for similarities and possible predictors of this potentially devastating and rare complication of pellucid marginal degeneration. The intent of this report is to raise awareness to eye care professionals regarding the potential for severe vision-threatening complications associated with pellucid marginal degeneration despite contact lens wear and a previously documented stable examination.

CASE REPORT

A 40-year-old female presented by means of referral with progressive vision loss in both eyes during a 24-hr period in association with pain, tearing, and total vision loss during the last 4 hr, first in the right eye followed by the left eye. Symptoms began after awakening in the right eye and as the day progressed, symptoms worsened and developed in the left eye. Ocular history included a diagnosis of pellucid marginal degeneration made 7 years previously with successful treatment using rigid gas permeable contact lenses during the last 4 years. A review of records from examination 1.5 years before presentation found best-corrected visual acuity of 20/30 OD and 20/25 OS with contact lens wear. Refraction measured $-8.50 + 6.50 \times 150$ OD and $-7.50 + 3.50 \times 035$ OS at that time. Corneal topography demonstrated classic findings consistent with pellucid marginal degeneration with against-the-rule astigmatism and inferior corneal steepening in a butterfly pattern, worse in the right eye (Fig. 1A,B). The patient denied a history of allergies, systemic disorders, or systemic medication use. History revealed the only potential trauma was a 4-year history of daily bilateral rigid gas-permeable lens wear and occasional eye rubbing after lens removal.

Examination at presentation revealed best-corrected visual acuity of hand motions OD and count fingers OS. Contact lens wear was discontinued on the day before presentation because of ocular discomfort. The right eye demonstrated a flat anterior chamber with a clear superior and central cornea and an inferior crescentic area of subepithelial/anterior stromal haze and edema measuring 3 mm vertically and 4.5 mm horizontally in greatest dimension with a 1.5 mm zone of normal cornea extending from below the region of thinning to the inferior limbus. A central arcuate area of perforation and iris prolapse was found within the center of the corneal thinning underlying a thin flap of corneal stroma. The arcuate-shaped perforation site measured 1 mm $\times$ 2 mm with no signs of corneal neovascularization or infiltration and no significant anterior chamber reaction. The left eye showed a formed but moderately shallow anterior chamber with severe inferior peripheral cornea thinning and central descemetocoe formation, but only minimal surrounding stromal edema in an arcuate region measuring 2.5 mm $\times$ 3.5 mm in greatest dimensions. The thinned portion of cornea was Seidel negative with no corneal neovascularization or infiltration. A 1.5 mm zone of normal cornea was present below the corneal melt extending to the adjacent limbus. No Fleischer ring was noted in either eye, but vertical striae were present in the left eye within the region of minimal edema just outside the descemetocoe. The anterior chamber, iris, and lens appeared normal in the left eye with no iris prolapse. Corneal sensation was symmetric and intact in both eyes. Gram stain and cultures for infectious agents were negative in both eyes.

Cyanoacrylate tissue adhesive was applied to the inferior peripheral cornea of the both eyes followed by bilateral bandage soft contact lens placement (Focus Night & Day, Ciba Vision, Duluth, GA.). Topical gatifloxacin 0.3% (Zymar) was started every 2 hr for infection prophylaxis and the patient was admitted for urgent repair of the right eye using a lamellar keratoplasty (Fig. 2). The left eye was repaired with a large diameter inferiorly decentered therapeutic penetrating keratoplasty 5 days later because of the leakage through the glue bandage from enlargement of the descemetocoe (Fig. 3). Corneal histopathology confirmed no signs of bacteria, amoeba, or fungal elements within the corneal tissue (Fig. 4A,B).

Six months after the initial surgery, visual acuity remained limited OD by the interface scar and combined regular and irregular astigmatism, thus an optical penetrating keratoplasty was performed with preoperative best-corrected visual acuity of 20/100 OD and 20/25 OS (Fig. 5). Selective suture removal to minimize astigmatism as well as subsequent bilateral cataract surgery using phacoemulsification and posterior chamber lens implantation and simultaneous relaxing incisions were ultimately performed in both eyes. Postoperative examination at 30 months after the initial presentation showed best-corrected visual acuity measured 20/15 OD with a refraction of plano +1.00 $\times$ 175 and 20/20 OS with a refraction of -2.50 + 4.50 $\times$ 075. Simulated keratometry values on corneal topography measured 41.75/43.30 at 170 OD and 41.12/47.62 at 070 OS.

DISCUSSION

Acute corneal hydrops has been estimated to occur in the setting of keratoconus and pellucid marginal degeneration in 2.5% to 6% of cases.^{3,5} Although acute corneal hydrops remains a rare complication of pellucid marginal degeneration, spontaneous corneal perforation remains an even more exiguous finding. In fact, in the largest review of patients with pellucid marginal degeneration to date, no cases of spontaneous perforation were noted in 116 eyes of 58 patients.⁵ A separate review of 31 eyes of 20 patients likewise found no cases of spontaneous corneal perforation.⁴ A meager 8 patients (10 eyes) with spontaneous perforation and pellucid marginal degeneration are identified through a literature search of the United States National Library of Congress and National Institutes of Health (Table 1). Indeed, classic cornea teaching suggests corneal perforation with corneal ectatic disorders “seldom, if ever, occurs” or “rarely occurs,” yet cases have now begun to surface which dispute these teachings.^{15,16}

Lucarelli et al.⁸ were the first to report spontaneous perforation in pellucid marginal degeneration. Jeng et al.¹² published the first report of sequential bilateral spontaneous corneal perforation occurring 20 months apart between eyes after the patient's first eye was reported by Aldave et al.^{11,13} Forooghian et al.¹³ reported the second case of sequential spontaneous corneal perforation in a patient with pellucid marginal degeneration with corneal perforation occurring 1 year apart between the affected eyes. Our case is unique in that the corneal perforation occurred acutely in both eyes at presentation in the absence of trauma, with the exception of daily rigid gas permeable contact lens wear and occasional eye rubbing.

A literature review was undertaken in hopes of finding a potential link between cases with spontaneous perforation. Including the 2 eyes in this report, the total number of reported cases with coexisting pellucid marginal degeneration and spontaneous corneal perforation totaled 12 eyes in 9 patients. Of these 9 patients, 7 were men and 2 were women. The 3 bilateral perforation cases included 2 men and 1 woman. Ages at presentation of all eyes ranged from 29 to 74 years old (mean 53), matching the ages of cases reported by Krachmer.² The mean age was significantly higher than additional case series showing an average age of 43 and 34, respectively.^{4,5} Risk factors for perforation included 5 of 12 eyes (41.7%) with a history of contact lens wear before perforation and an additional patient with preexisting atopic keratoconjunctivitis, a condition synonymous with eye rubbing. Thus, 5 patients (6 eyes) had some degree of potential trauma to the corneal surface as a risk factor to perforation. Given that most of patients with pellucid marginal degeneration are treated with contact lenses, other factors must exist in these patients, otherwise, larger numbers of perforation cases would occur if trauma was the only inciting factor.

In regards to treatment of pellucid marginal degeneration, contact lens wear remains the mainstay of treatment, yet surgical intervention is required in nearly 10% to 15% of cases.^{3, 17-20} The reverse is true in cases of spontaneous corneal perforation in that most cases are ultimately treated with either a penetrating keratoplasty or lamellar keratoplasty after initial

attempts at glue application and therapeutic contact lens placement.¹⁷⁻²¹ Forooghian et al.¹³ conversely reported conservative management of both cases of spontaneous perforation using a therapeutic bandage lens and prophylactic topical antibiotic therapy. Although conservative management should be considered first-line in cases of hydrops alone, conservative treatment in perforation cases warrants close observation for worsening of the perforation size and potential infectious keratitis as documented by a number of prior reports depicting microbial keratitis and worsening of perforation size.^{3,22,23} A tissue adhesive such as cyanoacrylate glue followed by therapeutic bandage lens placement should be considered first line in the treatment of this condition with a low threshold to perform either an eccentric penetrating keratoplasty or lamellar keratoplasty if the glue dislodges or the perforation increases in size.

One question that remains unanswered in all the cases of spontaneous corneal perforation is the underlying cause. The small sample size of cases within the literature precludes a direct cause of which cases of pellucid marginal degeneration can progress to corneal perforation; however, several cases including the current report indicate that corneal perforation in one eye may be the most significant risk factor for a similar outcome in the fellow eye. It appears that 2 forms of pellucid marginal degeneration exist. The most common form referred to in classic teaching is most often treatable with contact lenses and less commonly requires corneal surgical treatment, whereas the second form represents a very rare, yet severe form, ultimately leading to spontaneous corneal perforation that is most commonly treated with acute surgical intervention.

The second question remains whether the corneal edema represents a true corneal hydrops or a secondary effect as a result of corneal rupture. In fact, each of the 12 eyes reported with perforation had documented corneal edema at presentation. Although the right eye of our patient had significant surrounding stromal edema, the patient's left eye had only mild edema, raising the question of whether hydrops represents the origin of the corneal edema. Orlin and Sulweski suggested that the edema was not a true hydrops but a secondary effect from the perforation, yet many authors describe the break in Descemet's membrane, as with acute hydrops, as the probable precipitating factor for corneal edema. If perforation develops as a result of hydrops, why do so few cases of perforation develop in the setting of corneal hydrops? One hypothesis of corneal thinning in ectatic disorders suggests an interleukin-1-dependent mechanism may account for stromal loss and perhaps a subset of pellucid marginal degeneration cases have an extremely overactive immune response that can potentiate perforation, leading to the second form of more severe pellucid marginal degeneration.²⁴ As more cases develop in the future, perhaps we can elucidate risk factors which better predict which patients to watch closely for potential perforation, thus better differentiating the 2 forms of pellucid marginal degeneration and preventing the need for emergent surgical interventions in the severe form of pellucid marginal degeneration cases.

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FIG. 1. Corneal topography consistent with pellucid marginal degeneration 1.5 years before perforation in the right eye (A) and the left eye (B).

FIG. 2.
Slitlamp photograph showing a lamellar patch graft after inferior corneal perforation from pellucid marginal degeneration in the right eye.

FIG. 3.
Slitlamp photograph showing an inferior eccentric penetrating keratoplasty after inferior corneal perforation in the left eye.

FIG. 4. Corneal histopathology of both corneas. A, A hematoxylineosin stain with no sign of fungi, amoeba, or bacteria within a specimen from the corneal perforation region and adjacent sclera of the right eye (hematoxylin-eosin stain; $\times 40$). B, A hematoxylin-eosin stain of a corneal section from the left eye with no signs of infectious keratitis or inflammatory infiltrate (hematoxylin-eosin stain; $\times 40$).

FIG. 5.
Slitlamp photograph of the right eye after optical penetrating keratoplasty.

TABLE 1

Cases of Pellucid Marginal Degeneration and Spontaneous Corneal Perforation

Study	Age	Gender	Eyes	Predisposing factors	Initial treatment	Final treatment
Lucarelli et al. ⁸	56	F	1	No	ThPK	PK
Orlin et al. ¹⁰	74	M	1	No	Glue, BCL	ThPK
Aldave et al. ¹¹	55 29	M M	1 1	CTL No	Glue, BCL ThPK	Repeat Glue ThPK
Jeng et al. ¹²	56	M	1 ^a	CTL	Glue, BCL	ThPK
Alpeke et al. ⁹	56	M	1	CTL	LKP	—
Forooghian et al. ¹³	38	M	2 ^b	?	Glue, BCL	—
Symes et al. ¹⁴	66 54	M F	1 1	? Eczyma (atopic)	Glue BCL	LKP LKP
Lee et al (present study)	41 X = 53	F M > F (8:4)	2 ^b 12	CTL OU 6/12	Glue, BCL OU	LKP, PK OD EPK OS

^aSecond eye of previous reported case.

^bBilateral case.

ThPK, therapeutic keratoplasty; PK, penetrating keratoplasty; EPK, eccentric penetrating keratoplasty; LKP, lamellar keratoplasty; CTL, contact lens wear; BCL, therapeutic bandage contact lens.