Corneal perforation during scleral indentation in a patient with pellucid marginal degeneration

Karl Mercieca, Aruna Dharmasena, Charles Hopley

An observational case report of corneal perforation following scleral indentation in a patient with previously undiagnosed pellucid marginal degeneration is presented. Clinical examination, investigations, and subsequent management of this unwarranted and rare complication are described and discussed. The case highlights the need for thorough anterior segment examination before indirect ophthalmoscopy particularly in the presence of ectatic corneal pathology in which case scleral indentation should be avoided.

Key words: Cornea, pellucid marginal degeneration, perforation, retinal detachment, scleral indentation

Pellucid marginal degeneration (PMD) is a bilateral progressive ectatic disorder which commonly affects the inferior corneal periphery. It differs from other corneal ectasias in its characteristic inferior location and lack of inflammatory signs. We report a case of corneal perforation resulting from retinal examination using indirect ophthalmoscopy with scleral indentation in a patient with previously undiagnosed PMD.

Case Report

A 33-year-old Afro-Caribbean woman presented with a 12-month history of gradual onset bilateral visual loss, with particular worsening of the superior altitudinal field in the right eye over the previous 3 months. On examination, best-corrected visual acuity (VA) was "count fingers" on the right and 20/60 on the left with a refractive error in the left eye of + 3.75D/−7.00D at 100. Refractive status of the right eye was not available. Anterior segment examination revealed bilateral inferior corneal ectasia with stromal scarring and inferotemporal thinning. Fundus biomicroscopy showed a chronic inferior macula "off" retinal detachment with an inferior retinal hole and pigmented demarcation lines.

Bilateral indirect ophthalmoscopy was performed with scleral indentation to exclude any further retinal breaks.

Access this article online	
Quick Response Code:	Website: www.ijo.in
	DOI: 10.4103/0301-4738.181750

Manchester Royal Eye Unit, Manchester Royal Eye Hospital, Manchester, United Kingdom

Correspondence to: Dr. Karl Mercieca, Manchester Royal Eye Hospital, Oxford Road, Manchester, M13 9WL, United Kingdom. E-mail: doctormercieca@yahoo.com

Manuscript received: 23.10.14; Revision accepted: 06.02.16

Subsequently the patient started complaining of a watery sore right eye. Slit lamp examination revealed a very shallow anterior chamber (AC) with peripheral iridocorneal touch and an inferotemporal corneal perforation [Fig. 1].

The patient underwent urgent corneal application of cyanoacrylate glue and placement of a bandage contact lens with consequent deepening of the AC. However, on the 3rd postoperative day, the AC was shallower with iris incarceration at the point of original perforation. Tectonic full-thickness keratoplasty was performed 1 week postapplication of corneal glue. The graft remained clear and attached up to 2 months postoperatively with VA improving to count fingers.

Over this period, significant cataract developed making fundal view impossible. B-scan ultrasonography confirmed a persistent inferior retinal detachment. The patient eventually underwent a right combined phaco-vitrectomy, internal search, retinopexy, and SF_6 gas. Over the following 3 months, VA improved to 20/200 with + 6.50 DS correction and the retina remained attached.

Discussion

PMD is a progressive, noninflammatory corneal ectasia which commonly affects the inferior periphery of the cornea. It typically presents with crescentic corneal thinning from the 4 to 8 o'clock position, 1–2 mm from the limbus. It differs from other corneal ectatic disorders in its characteristic inferior thinning below the apex of the cone and typically severe against-the-rule astigmatism.

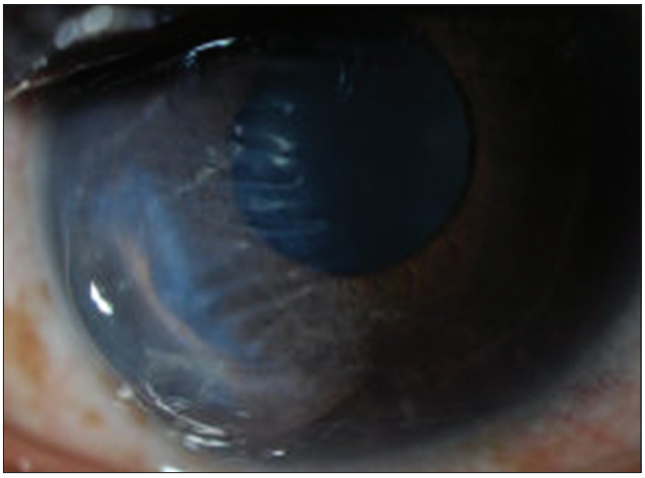

Figure 1: Right eye anterior segment photograph showing an inferotemporal corneal perforation with a shallow anterior chamber and peripheral iridocorneal touch

Cite this article as: Mercieca K, Dharmasena A, Hopley C. Corneal perforation during scleral indentation in a patient with pellucid marginal degeneration. Indian J Ophthalmol 2016;64:233-4.

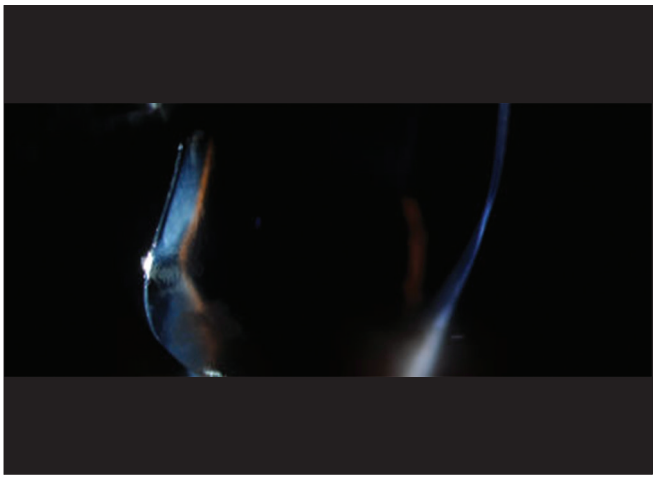

Figure 2: Slit lamp photography showing the glued inferior corneal perforation in the right eye (OD) and mid-peripheral inferior corneal stromal thinning with overlying intact epithelium and thicker perilimbal stroma in the left eye (OS)

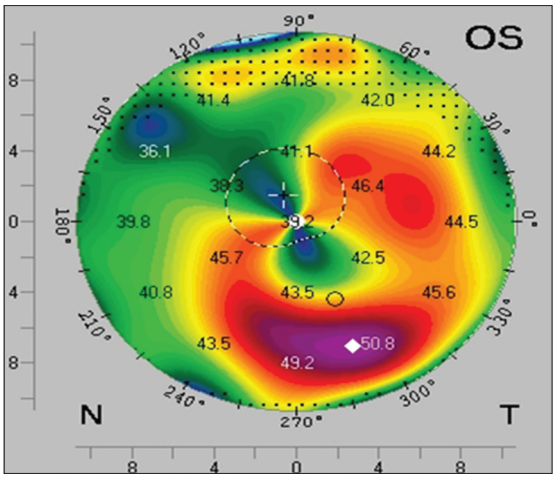

Figure 3: Corneal topography of the left eye showing the typical "kissing birds" or "crab claw" configuration

Our patient had a classical background presentation between the 4th and 5th decades of decreased VA due to high irregular against-the-rule astigmatism. The significant degree of astigmatism should have provided an early clue, but PMD was only considered after careful examination of the left eye following perforation in the right [Fig. 2]. The typical findings of inferior corneal thinning with intact epithelium and ectatic cornea superior to this area should have pointed to the diagnosis, particularly in the absence of scarring, vascularization, or lipid deposition. The retinal detachment had evidently diverted attention away from these anterior segment features of PMD.

The steepening of the inferior periphery with extension to the horizontal and oblique meridians is believed to be a topographic characteristic of PMD.[1,2] The "kissing birds" or "crab claw" appearance on corneal topography was important in eventually confirming the diagnosis in our patient [Fig. 3].

Spontaneous corneal perforation in PMD is uncommon and can rarely be bilateral.[3-5] To the best of our knowledge, corneal perforation during retinal examination with sclera indentation has not been previously reported in the literature. This case highlights the importance of meticulous anterior segment examination before posterior segment assessment and suggests that if ectatic pathology is discernible indirect ophthalmoscopy with indentation should be undertaken only if absolutely necessary and with appropriate caution.

Visual rehabilitation in cases of corneal perforation is difficult. Tectonic grafting causes a greater degree of astigmatism when compared to elective procedures such as decentered standard-sized penetrating keratoplasty, crescentic wedge excision, and crescentic lamellar keratoplasty.[5-7] The loss of AC stability during perforation and the application of corneal glue make the eye more prone to complications such as cataract, inflammation, and raised intraocular pressure. In the case described the visual prognosis was further limited by the presence of a chronic retinal detachment. Ultimately the patient had a relatively good outcome following her multiple procedures, and the possibility of an elective penetrating keratoplasty in the future has not been ruled out.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Karabatsas CH, Cook SD. Topographic analysis in pellucid marginal corneal degeneration and keratoglobus. Eye (Lond) 1996;10(Pt 4):451-5.
2. Maguire LJ, Klyce SD, McDonald MB, Kaufman HE. Corneal topography of pellucid marginal degeneration. Ophthalmology 1987;94:519-24.
3. Lucarelli MJ, Gendelman DS, Talamo JH. Hydrops and spontaneous perforation in pellucid marginal corneal degeneration. Cornea 1997;16:232-4.
4. Jeng BH, Aldave AJ, McLeod SD. Spontaneous corneal hydrops and perforation in both eyes of a patient with pellucid marginal degeneration. Cornea 2003;22:705-6.
5. Symes RJ, Catt CJ, Sa-ngiampornpanit T, Males JJ. Corneal perforation associated with pellucid marginal degeneration and treatment with crescentic lamellar keratoplasty: Two case reports. Cornea 2007;26:625-8.
6. Biswas S, Brahma A, Tromans C, Ridgway A. Management of pellucid marginal corneal degeneration. Eye (Lond) 2000;14(Pt 4):629-34.
7. Rasheed K, Rabinowitz YS. Surgical treatment of advanced pellucid marginal degeneration. Ophthalmology 2000;107:1836-40.