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Case report

# Bilateral idiopathic corneal opacity: A report of Ascher ring and a review of the literature



M. Megalla\*, E. Li, P. Branden, J. Chow

Yale Department of Ophthalmology, New Haven, CT, USA

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ABSTRACT

Purpose: To present a case of a rare entity of Ascher ring, a bilateral corneal stromal opacification. Observation: A 70-year-old male with no ocular history who presented for cataract evaluation was found to have idiopathic bilateral circular stromal corneal rings.

Conclusions: After completion of extensive history, examination, imaging analyses, and laboratory studies for workup of corneal opacities, we arrived at a diagnosis of Ascher corneal ring, an extremely rare entity. Importance: A rare entity should be considered after excluding other etiologies.

#### 1. Introduction

Corneal opacities have a broad differential, including dystrophies, drug-induced deposits or sequelae of use, metabolic and crystalline depositions, posterior embryotoxon, and infectious immune processes. We present a case of a very rare entity of corneal opacification, Ascher ring, which only carries a handful of case reports in the literature.

#### 2. Case report

A 70-year-old male with medical history significant for diabetes mellitus, hypertension, and prostate cancer presented to the ophthalmology clinic for cataract evaluation of the right eye (OD). He denied photophobia, eye pain, discharge, contact lens use, ocular trauma, history of ocular procedures, and personal or family history of metabolic disorders or corneal dystrophies. Medications included atorvastatin, chlorthalidone, clopidogrel, metformin, primidone, topiramate, tamsulosin, and propanolol. Ocular history was significant for refractive error. On examination, best-corrected visual acuity was 20/40 OD and 20/30 left eye (OS). Pupillary reactions and intraocular pressures were normal in both eyes (OU). Corneal pachymetry OD was 549 µm and 681 µm centrally and peripherally, respectively. Corneal pachymetry OS was 547 µm and 673 µm centrally and peripherally, respectively. Anterior segment examination was significant for symmetric bilateral circular mid-peripheral full-thickness stromal white rings with a diameter of 7-8mm (Fig. 1a and b). Corneal sensation was intact, and no vascularization, fibrosis, thinning or fluorescein staining was appreciated.

#### 3. Discussion

Stromal dystrophies often appear as fleck like opacities and are inherited in an autosomal dominant fashion except for macular corneal dystrophy, which is inherited autosomal recessively.1 The stromal opacities in our patient were circular and symmetric without any family history of similar findings. Iron, gold and silver can develop stromal deposits as can some drugs such as chlorpromazine<sup>1</sup> and perifosine.<sup>2</sup> Topical use of brimonidine has also been associated with deep stromal opacities.<sup>3</sup> However, our patient denied past history of trauma and hyphema, or use of drugs with known adverse events of corneal stromal deposition. Metabolic disorders such as lecithinin-cholesterol acyltransferase deficiency (LCAT) and Tangier disease present with corneal clouding and stromal opacities.<sup>1,4</sup> However, significant abnormalities in the lipid panel would be noted. Multiple myeloma presents rarely with diffuse gray white crystalline corneal deposits. It's precursor, monoclonal gammopathy of undetermined significance (MGUS), can present

Gonioscopy revealed angles open to ciliary body without a thickened Schwalbe's line or iris processes OU. Posterior segment examination was unremarkable OU. Laboratory studies, which included basic metabolic panel, complete blood count, lactate dehydrogenase, beta-2 microglobulin, and serum protein electrophoresis, were within normal limits. His lipid profile showed slightly reduced high-density cholesterol levels. Anterior segment optical coherence tomography showed highly reflective material throughout the entire corneal stroma (Fig. 1c). The patient proceeded with cataract surgery OD without complications.

<sup>\*</sup> Corresponding author. 40 Temple St Suite 3D, New Haven, CT, 06511, USA. E-mail address: marez.megalla@yale.edu (M. Megalla).

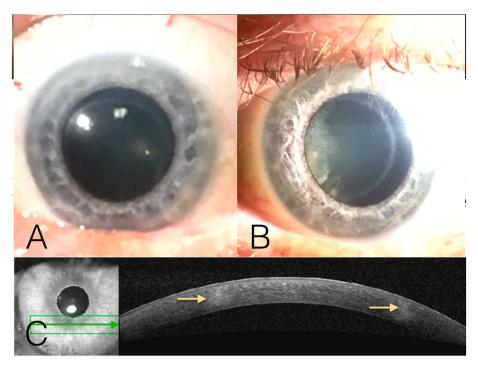


Fig. 1. Slit lamp photograph showing the Ascher ring (A and B) and anterior-segment OCT showing highly reflective material throughout the entire stroma (arrows, C).

with "chameleon-like" corneal opacities resembling numerous corneal entities including verticillata, lattice, and granular dystrophies. Posterior embryotoxon, or an anteriorly displaced and thickened Schwalbe's line, <sup>1</sup> is a congenital finding that would be noted on gonioscopy. Ring-shaped stromal opacities have also been described after corneal cross-linking without causing visual disturbance, but our patient denied any history of ocular procedures. Infectious processes such as herpes keratitis and acanthomeba are associated with corneal infiltrates however would present symptomatically and with other corneal findings, neither of which our patient had.

Ascher ring, first described in 1964, is an extremely rare corneal entity described only a handful of times in the literature. 7-20 Ascher rings are described as idiopathic intrastromal mid-peripheral corneal rings that are characteristically bilateral and not visually significant. Although there is usually one concentric ring per eye, there is one report of multiple symmetric rings.<sup>20</sup> No associated inheritance patterns or lab abnormalities have been identified, although some patients have been found to have elevated serum cholesterol. 7-11 The extent of corneal stromal involvement varies, as some cases describe superficial involvement,<sup>7</sup> entire stromal involvement<sup>11,12</sup> or partial stromal involvement extending to one-half to two-thirds of the stromal depth.  $^{8,13}$  On imaging, OCT shows opacification of the corneal stroma<sup>11,14</sup> while reflective extracellular deposits are found on confocal microscopy.<sup>14</sup> Initially, these opacifications were believed to be a result of an immunologic reaction 2 as some patients with this entity had ocular history of unilateral idiopathic iritis. However, newer considerations suggest that the opacities are extracellular material likely deposited along a centripetal gradient. 10,14

### 4. Conclusions

Because of the broad differential diagnoses of corneal stromal opacities, including corneal manifestations of systemic illnesses, we recommend a diagnosis of Ascher corneal ring as a diagnosis of exclusion after a thorough history, ocular examination, and laboratory studies are completed. Once made, patients can be reassured of the good visual prognosis associated with Ascher corneal ring.

#### Patient consent

The patient consented to the publication of the case orally. This report does not contain any personal information that could lead to the identification of the patient.

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#### References

- Basic and clinical science course (BCSC). section 8: External Disease and Cornea. San Francisco (CA): American Academy of Ophthalmology (AAO); 2017.
- Keenan J, Fram N, McLeod S, et al. Perifosine-related rapidly progressive corneal ring infiltrate. Cornea. 2010;29(5):583–585.
- Tsujinaka A, Takai Y, Inoue Y, et al. A case of bilateral deep stromal corneal opacity and vascularization after use of multiple antiglaucoma medication including brimonidine tartrate ophthalmic solution. Acta Ophthalmol. 2019;97(6):e948–e949.
- Viestenz A, Schlotzer-Schrehardt U, Hofmann-Rummelt C, et al. Histopathology of corneal changes in lecithin-cholesterol acyltransferase deficiency. *Cornea*. 2002;21 (8):834–837.
- Lisch W, Wasielica-Poslednik J, Kivela T, Schlotzer-Schrehardt U, et al. The hematologic definition of monoclonal gammopathy of undetermined significance in relation to paraproteinemic keratopahty (an American ophthalmological society thesis). Trans Am Ophthalmol Soc. 2016;114(T7):1–21.
- Camesasca F, Vinciguerra P, Seiler R. Bilateral ring-shaped intrastromal opacities after corneal corsslinking. J Refract Surg. 2011;27(12):913–915.
- Ascher K. An unusual corneal ring. Ber Zusammenkunft Disch Ophthalmol Ges. 1964; 65:44–46.
- Reiger G. Primare, einseitige, ringförmige Trübung der Hornhaut. Fortschr Ophthalmol. 1987;84:242–244.
- Rohrbach J, Kleiser N, Kaufmann-Fechner J, Lisch W. Corneal ring opacity (Ascher ring)- a case report. Klin Monatsbl Augenheilkd. 2001;218(4):276–278.
- Silvestri A, Boisjoly H. Bilateral corneal ring opacity of unknown origin. Cornea. 2003;22(1):86–87.
- 11. Moshirfar M. Corneal surgical problem. *J Cataract Refract Surg.* 2015;41:895–896.
- 12. Bron A. Peripheral ring opacity of the cornea. Br J Ophthalmol. 1969;53:270–273.
- Bopp S, Laqua H. Corneal Ascher ring. A ring-shaped stromal corneal opacity. Klin Monatsbl Augenheilkd. 1991;198(3):201–204.
- Nguyen D, Quah S, Kumar N, et al. In-vivo scanning of Ascher intrastromal corneal ring opacity. Br J Ophthalmol. 2007:1710–1711.

- 15. Melles G, de Sera J, Eggink C, et al. Bilateral anterior stromal ring opacity of the cornea. Br J Ophthalmol. 1998;82:522-525.
- 16. Caroline P, Melles G. Two cases of bilateral, stromal ring opacity of the cornea. Cornea. 2001;20(2):237-238.
- 17. Almeida R, Ruao M, Almeida I, et al. Bilateral ring-shaped corneal opacity: case report and review of the literature. The Pan-American Journal of Ophthalmology. 2016;15:61–62.
- 18. Khan JC, Shuttleworth GN. Annular granular corneal opacity: a rare corneal stromal dystrophy or degeneration? *Br J Ophthalmol*. 2000;84(10):1205–1206.
  19. Winnick J. Bilateral corneal ring-opacity observed in long term contact lens wearer.
- Clin Eye Vis Care. 1999;10:165-172.
- 20. Uddaraju M, Macharenhas J, Das M, Prajna N. A case of bilateral, multiple, symmetric, concentric ring-shaped opacities in the cornea. JAMA Ophthalmology. 2015;133:483-484.