

Bilateral Idiopathic Corneal Opacity: A Report of Ascher Ring and a Review of the Literature

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

Abstract

We present a case of a 70-year-old male with idiopathic bilateral circular mid-peripheral corneal stromal rings found incidentally on examination. After completion of extensive history, examination, imaging analyses, and laboratory studies, we arrived at a diagnosis of Ascher corneal ring, an extremely rare entity.

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 propranolol. 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 (Figure 1a and 1b). Corneal sensation was intact and no vascularization, fibrosis, thinning or fluorescein staining were appreciated. 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 the corneal stroma (Figure 1c). The patient proceeded with cataract surgery OD without complications.

Discussion

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. 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 chlorpromazine¹ and perifosine.² Topical use of brimonidine has also been associated with deep stromal opacities.³ 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 lecithin:cholesterol acyltransferase deficiency and Tangier disease present as a dense arcus lipoides.^{1,4} However, significant abnormalities in the lipid panel would be noted. Multiple myeloma and its precursor, monoclonal gammopathy of undetermined significance, present rarely with diffuse gray white crystalline corneal deposits.⁵ Posterior embryotoxon, or an anteriorly displaced and thickened Schwalbe's line,¹ 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.⁶ Infectious processes¹ such as herpes keratitis and acanthamoeba 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 literature⁷⁻¹⁸ (Table 1). Ascher rings are described as idiopathic intrastromal mid-peripheral corneal rings. It is characteristically bilateral, not visually significant, and without any associated inheritance patterns or lab abnormalities, 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,⁷ entire stromal involvement¹¹⁻¹² 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^{11,14} while reflective extracellular deposits are found on confocal microscopy.¹⁴ Initially, these opacification were believed to be a result of an immunologic reaction¹² 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}

To our knowledge, we present the 15th case of Ascher corneal ring reported in the literature. 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.

References

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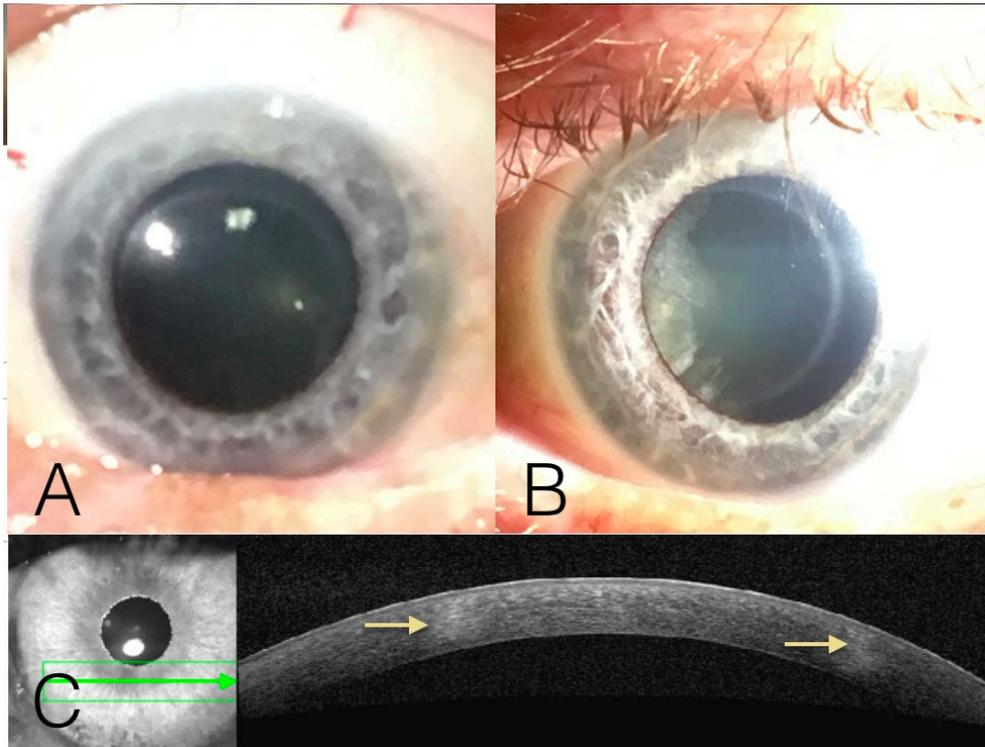


Figure 1. Slit lamp photograph showing the Ascher ring (A and B) and anterior-segment OCT showing highly reflective material throughout the entire stroma (arrows, C).

Table 1. Comparison and review of the reported cases of Ascher ring.

	Age/Sex	Laterality	Vision	Corneal Involvement	Ocular History	Family History	Ocular Testing	Laboratory Analyses
Ascher ⁷	39y M	Bilateral	Unknown	Anterior stroma 7-8mm diameter	Iritis	None	Unknown	Lipid profile normal
Reiger ⁸	65y M	Unilateral	Unknown	Anterior 2/3 stroma 7-8mm diameter	None	Unknown	Unknown	Lipid profile- elevated cholesterol
Rohrbach ⁹	63y M	Bilateral	20/20 OU	Full-thickness stroma 8mm diameter 0.25mm wide rings	None	None	Unknown	Lipid profile and uric acid- elevated cholesterol, LDL, and uric acid
Silvestri ¹⁰	82y M	Bilateral	20/30 OU	Unclear depth 5-6mm diameter	Blepharitis	None	Keratometry, topography, pachymetry, gonioscopy, specular-normal	High cholesterol and serum uric acid Protein electrophoresis- normal
Moshirfar ¹¹	63y M	Bilateral	20/20 OD 20/250 OS	Full-thickness stroma ~4mm diameter	Macular hole OS	None	Anterior segment OCT- hyper reflective opacities	Unknown
Bron ¹² case 1	59y M	Bilateral	6/5 OU	Full-thickness stroma	None	Unknown	Unknown	Serum protein and electrophoresis and cholesterol- normal
Bron ¹² case 2	66y F	Bilateral	6/9 OU	Full-thickness stroma ~7mm diameter	Crocodile Shagreen	Unknown	Unknown	Unknown
Bopp ¹³ case 1	80y F	Bilateral	Unknown	Anterior 1/2 stroma 7-8mm diameter	Cataract	None	Unknown	Lipid profile normal
Bopp ¹³ case 2	65y M	Bilateral	Unknown	Anterior 2/3 stroma 4-5mm diameter	Cataract	None	Unknown	Lipid profile normal
Nguyen ¹⁴	80y M	Bilateral	6/6 OU	Full-thickness storm 7.8mmx7.7mm diameter 0.1mm wide rings	None	None	Pachymetry and topography- normal Anterior segment OCT- hyper reflective opacities Confocal microscopy-reflective extracellular deposits	Lipid, protein, and autoimmune-negative
Melles ¹⁵	25y M	Bilateral	1.0 OU	Anterior to mid stroma ~8.0mm diameter 1/3mm wide rings	Trauma	None	Pachymetry, topography, gonioscopy- normal	Lipid, syphilis, sarcoid, Lyme and urinalysis-negative
Caroline ¹⁶	68y F	Bilateral	20/20 OU	Unclear depth 7.0mm x 8.0mm diameter 0.3mm wide rings	Contact lens use, strabismus surgery	Unknown	Pachymetry, topography, specular, keratometry, gonioscopy- normal	Unknown
Almeida ¹⁷	80y M	Bilateral	8/10 OD 3/10 OS	Unclear depth 7mmx6.3mm diameter	Cararact	Unknown	Topography, specular, gooniescopy- normal	Lipid, urinalysis, syphilis, lyme, autoimmune- negative
Khan ¹⁸	56y F	Bilateral	20/20 OU	Unclea depth 7-8mm diameter	Posterior vitreous detachment	None	Pachymetry, specular- normal	Autoantibodies and lipid panel- normal
Our patient	70y M	Bilateral	20/40 OD 20/30 OS	Full-thickness stroma 7-8mm diameter	Cataract	None	Pachymetry, gonioscopy- normal	SPEP negative Lipid- low HDL