

Case Report

Iris Nevus (Cogan-Reese) Syndrome Presenting with Zonular Dehiscence during Cataract Extraction

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Keywords

Iridocorneal endothelial syndrome · Cataract · Zonular dehiscence · Cogan-Reese syndrome

Abstract

The aim of this study was to report a novel presentation of Cogan-Reese syndrome presenting with zonular dehiscence during cataract extraction with intraocular lens placement. A 49-year-old woman presented with worsening vision over 2 years. The examination was significant for bilateral pupil miosis, visually significant cataracts, and unilateral glaucoma. No iris nodules or corneal endothelial disease was observed through slit-lamp examination and specular microscopy bilaterally. Cataract extraction on the left eye was complicated by significant zonular dehiscence. An iris sample taken during the procedure demonstrated histopathologic findings consistent with an iris nodule composed of melanocytic nevoid cells. Cogan-Reese, or iris nevus syndrome, is a subset of iridocorneal endothelial syndrome that usually presents with iridic stromal matting and stromal loss, nodule formation, and secondary unilateral angle-closure glaucoma. Here, we describe a presentation of Cogan-Reese syndrome that presented with pupillary miosis and glaucoma preoperatively, and zonular dehiscence during cataract extraction with no underlying corneal pathology.

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Introduction

Iridocorneal endothelial syndrome (ICE) involves abnormalities in the corneal endothelium, angle structures, and iris tissue to varying degrees. Patients with ICE usually present with unilateral disease and can have a variety of signs including atrophic iris tissue, glaucoma,

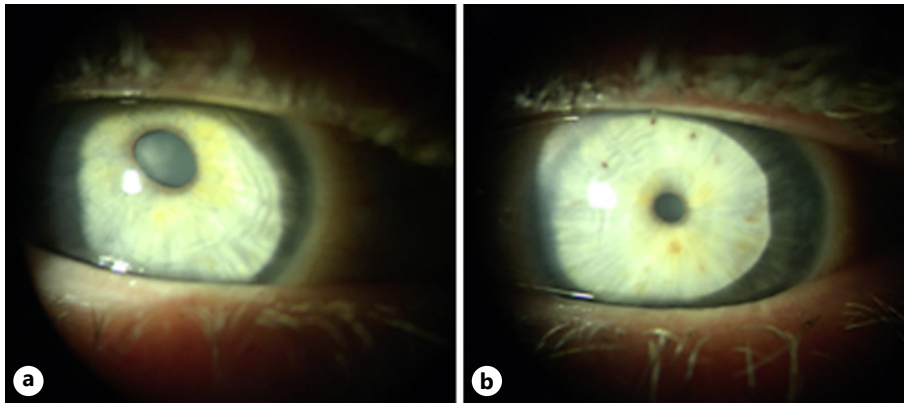


Fig. 1. Pre-operative slit-lamp photographs: right eye (a) and left eye (b). Both eyes were on atropine drops for 1 month prior to taking the photos.

and corneal endothelial failure. Herein we present an atypical case of bilateral ICE with thickened iris tissue resulting in severe bilateral pupillary miosis, endothelial irregularity, and diffuse zonulopathy.

Case Report/Case Presentation

A 49-year-old woman presented for evaluation of worsening vision over the past 2 years. Ocular history was positive for glaucoma in the left eye, which was well controlled with travoprost. She denied any other past medical or ocular conditions, including a history of herpetic eye disease. She was found to have visually significant cataracts bilaterally, as well as <1 mm pupils with very poor response to mydriatic drops.

The patient was started on atropine 1% OU after her initial exam due to poor dilation and extremely small pupils, affecting her total vision. A repeat examination on atropine 1 month later was still difficult due to significant miosis bilaterally (1.5 mm OD, 1 mm OS) but showed a clear cornea, slightly shallow but quiet anterior chamber, miotic pupil with a matted appearance and wrinkling/redundancy of the mid-peripheral iris tissue, and moderate nuclear sclerotic cataracts bilaterally (Fig. 1a, b). A gonioscopic exam revealed occludable angles without any iridocorneal adhesions. She had no prior history of glaucoma. During our exam, visualization or imaging of the optic nerve or fundus was not possible through her extremely small pupils. Her best spectacle-corrected visual acuity was 20/30 OD and 20/50 OS. She was a previous soft toric contact lens wearer. Her intraocular pressures by Goldman applanation tonometry were 18 mmHg bilaterally. Corneal topography revealed with-the-rule regular astigmatism of 2.8 D cylinder OD and 3.3 D cylinder OS. Central corneal thickness was 544 μm OD (Fig. 2a) and 554 μm OS (Fig. 2b), and specular microscopy of the corneal endothelium showed high pleomorphism and rounded morphology of the cells (Fig. 2).

Due to the visually significant nature of the central nuclear sclerotic cataract, a decision was made to move forward with cataract extraction in the left eye. After discussing risks, benefits, and alternatives, cataract surgery in the left eye with placement of a toric lens for astigmatism correction was scheduled. During surgery, an iris expansion ring was used to aid in the dilation of the pupil. It was observed that there was significant zonulopathy as early as during the capsulorhexis with wrinkling of the capsule and movement of the entire lens/capsule complex. The zonulopathy was not secondary to surgical manipulation as the nuclear

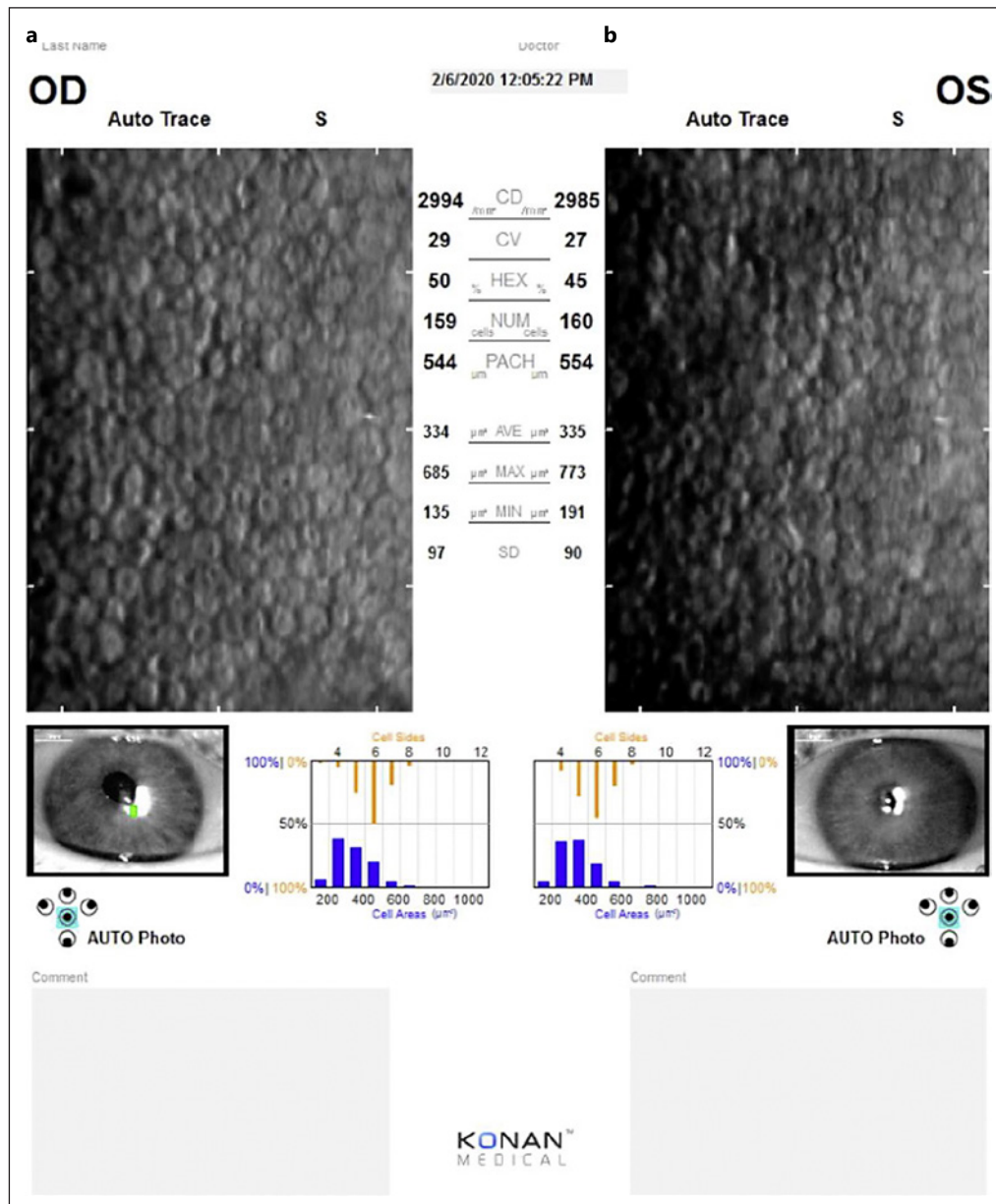


Fig. 2. Central corneal thickness was 544 μm OD (**a**) and 554 μm OS (**b**), and specular microscopy of the corneal endothelium showed high pleomorphism and rounded morphology of the cells.

density of the cataract was moderate and nuclear removal was not complex. A simple quick chop technique was used to remove the nuclear fragments, which was completed without incident. A capsular tension ring was then placed to reform and support the capsular bag prior to cortical removal. After placement of the capsular tension ring and after the safe removal of the cortical material, there was still significant decentration and instability of the capsular bag complex with diffuse zonular weakness. A decision was made to place a scleral-fixated Ahmed capsular tension segment oriented in the area of greatest zonular loss inferiorly. The toric intraocular lens was safely inserted within the now-centered capsular bag. A small sample of iris tissue was then excised from the pupillary margin and sent to pathology.

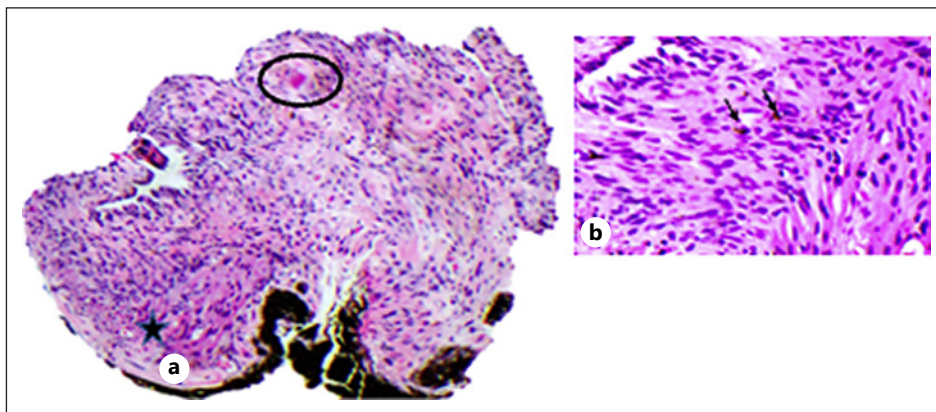


Fig. 3. (a) Iris nodule adjacent to the pupillary margin (asterisk) revealing a focal dystrophic calcification (encircled). H&E original magnification $\times 100$. (b) Inset: spindle-shaped nevoid cells with scattered intracytoplasmic melanin pigment granules (arrow). H&E original magnification $\times 400$. H&E, hematoxylin-eosin stain.

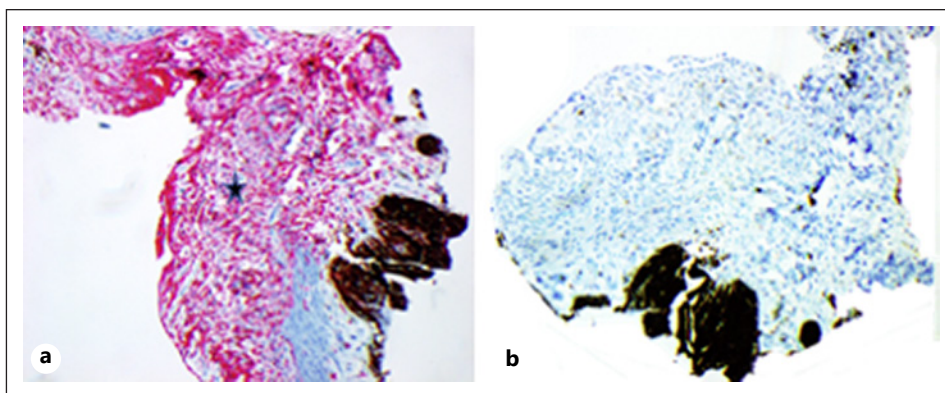


Fig. 4. (a) Immunohistochemical study, melanoma antigen recognized by T cells (melan-A) stain was performed confirming the melanocytic nature of the lesion. (b) The proliferation index, Ki-67, was positive in less than 1% of the cells.

This pupilloplasty also served to enlarge the miotic pupil. Six months postoperatively, the patient's uncorrected visual acuity was 20/20 with a slightly decentered but stable toric zero-spherical aberration (Envista) intraocular lens. Postoperative exam revealed an optic nerve cup-to-disc ratio of 0.5 mm OU and an otherwise healthy fundus exam.

Histopathologically, the iridectomy specimen revealed a benign proliferation of uniform spindle-shaped cells with eosinophilic cytoplasm, scattered intracytoplasmic melanin granules, and focal dystrophic calcification (Fig. 3a, b). The histopathologic sections reveal a portion of smooth muscle of the presumed pupillary sphincter. The cells demonstrated nevus cell appearance arising from the iris stroma in close proximity to the pupillary margin. No stromal atrophy, endothelialization, or Descemet-like membrane was present. In this immunohistochemical study, melanoma antigen recognized by T cells (melan-A) stain was performed confirming the melanocytic nature of the lesion (Fig. 4a). The proliferation index, Ki-67, was positive in $<1\%$ of the cells (Fig. 4b). The overall histopathologic findings favor a benign iris nodule composed of melanocytic nevoid cells.

Discussion/Conclusion

ICE is a unique disorder that presents unilaterally with abnormal corneal endothelium, corneal edema, iris atrophy, and secondary angle closure glaucoma. Most commonly, the corneal endothelium in ICE syndrome is described as “hammered silver” or “beaten bronze” appearance when visualized with specular reflection [1]. ICE syndrome is considered to be an acquired disease, as polymerase chain reaction of patients has demonstrated herpes simplex viral DNA [2].

Cogan-Reese syndrome, otherwise known as iris nevus syndrome, is the least common variant of ICE syndrome [3]. In Cogan-Reese syndrome, iridic stromal matting and stromal loss, fine iris nodules, and unilateral angle-closure glaucoma often develop secondary to endothelial membrane formation on the anterior portion of the iris [4]. The presence of these nodules is often required for diagnosis, though it is important to note that they may develop late in the course of the disease [5]. The nodules that develop from Cogan-Reese do not usually cause marked pupillary displacement [6].

Most patients who present with Cogan-Reese present with unilateral reduced vision, pain, sectoral color changes in the iris, and possible visual field defects related to glaucoma. Bilateral presentation of the disease has been reported but is very uncommon [7, 8]. Neither the presence of ICE cells on specular microscopy nor iris nevus on slit-lamp examination was observed in this patient. Alternative differential diagnoses should be considered with the discovery of iris nodules including iris mammillations, neurofibromatosis, sarcoidosis, or melanoma of the iris, among others.

However, the constellation of anterior segment findings including the iris abnormalities and secondary angle-narrowing observed in this middle-aged woman raised the possibility of early stages of the clinical spectrum of the ICE syndrome. Although there was no frank endothelial dysfunction or membranes seen in the angle, specular microscopy did reveal rounded morphology and high pleomorphism of corneal endothelial cells not typical in normal hexagonal endothelial cell morphology. The diffuse zonular dehiscence observed during cataract surgery further points to a genetic abnormality of the anterior segment anatomy not regularly seen in the ICE spectrum of disorders. The bilateral nature of the findings in this case also makes it unique. To our knowledge, this is the first reported case of Cogan-Reese syndrome presenting with diffuse zonular dehiscence during cataract extraction and no corneal endothelial disease.

Statement of Ethics

Ethical approval is not required for this study in accordance with local or national guidelines and written informed consent was obtained from patient for publication of the details of their medical case and any accompanying images.

Conflict of Interest Statement

The authors have no financial or conflicts of interest to disclose.

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Author Contributions

Priyanka Chhadva, MD – Substantial contributions to the conception or design of the work; or the acquisition, analysis, or interpretation of data for the work; drafting the work or revising it critically for important intellectual content; final approval of the version to be published; and agreement to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. Maria Del Valle Estopinal, MD – Substantial contributions to the conception or design of the work; or the acquisition, analysis, or interpretation of data for the work; drafting the work or revising it critically for important intellectual content; final approval of the version to be published; and agreement to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. Marjan Farid, MD (corresponding author) – Substantial contributions to the conception or design of the work; or the acquisition, analysis, or interpretation of data for the work; drafting the work or revising it critically for important intellectual content; final approval of the version to be published; and agreement to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

Data Availability Statement

All data generated or analyzed during this study are included in this article. Further inquiries can be directed to the corresponding author.

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