

Case Report

Role of Anterior Segment Optical Coherence Tomography (AS-OCT) in the Diagnosis and Follow-Up of Irido-Corneal-Endothelial (ICE) Syndrome: A Case Report

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Received: October 14, 2025

Accepted: November 07, 2025

Published: November 10, 2025

Abstract

Irido-Corneal-Endothelial (ICE) syndrome is a rare, progressive, unilateral disorder characterized by abnormal proliferation and migration of corneal endothelial cells that form a fibrocellular membrane over the posterior cornea and anterior iris. We report the case of a 48-year-old woman presenting with progressive blurred vision in her left eye. Slit-lamp examination revealed a translucent posterior corneal membrane, shallow inferior anterior chamber, and iris nodules. Gonioscopy showed peripheral anterior synechiae, and AS-OCT demonstrated a hyper-reflective membrane involving the posterior cornea and iridocorneal angle. Specular microscopy confirmed a reduced endothelial-cell density compared with the fellow eye. AS-OCT proved to be a rapid, non-invasive, and highly informative imaging tool that facilitates early diagnosis and follow-up of ICE syndrome by visualizing subtle corneal and angle abnormalities and aiding timely management.

Keywords: Irido-Corneal-Endothelial Syndrome; AS-OCT; Endothelial Proliferation; Peripheral Anterior Synechiae; Chandler Syndrome; Corneal Endothelium; Secondary Glaucoma

Introduction

Irido-Corneal-Endothelial (ICE) syndrome is a rare, typically unilateral disorder affecting the corneal endothelium, iris, and iridocorneal angle. It is characterized by the proliferation and migration of abnormal endothelial cells forming a fibrocellular membrane on the posterior corneal surface and anterior iris. This process leads to peripheral anterior synechiae, iris atrophy, corectopia, and frequently secondary glaucoma or corneal decompensation. Anterior Segment Optical Coherence Tomography (AS-OCT) is a non-contact, high-resolution imaging technique based on low-coherence interferometry. It allows detailed visualization of the corneal layers, anterior chamber, and angle configuration, making it an important diagnostic and monitoring tool in ICE syndrome.

Case Presentation

A 48-year-old female presented with gradual decrease in visual acuity of the left eye despite stable optical correction since 2013. Past Medical History: Hypertension controlled by a calcium-channel blocker; no ocular surgery or trauma.

Ocular Examination

- **Refraction:** OD +2.25 ($-0.75 \times 104^\circ$); OS +2.25 ($-1.25 \times 56^\circ$)
- **Best-Corrected Visual Acuity:** 10/10 in both eyes
- **Intraocular Pressure:** 15 mmHg in both eyes
- **Slit-Lamp Findings:**

- **OD:** clear cornea, deep anterior chamber, normal iris and lens.
- **OS:** translucent membrane on the posterior peripheral cornea inferiorly, shallower inferior chamber, and small iris nodules.
- **Gonioscopy:** inferior peripheral anterior synechiae with a thin veil-like membrane on the anterior iris surface (OS); normal (OD).

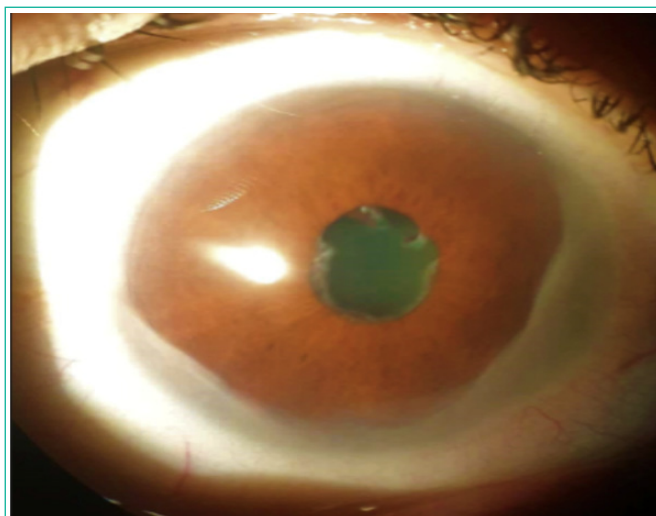


Figure 1: Biomicroscopic appearance of the left eye.

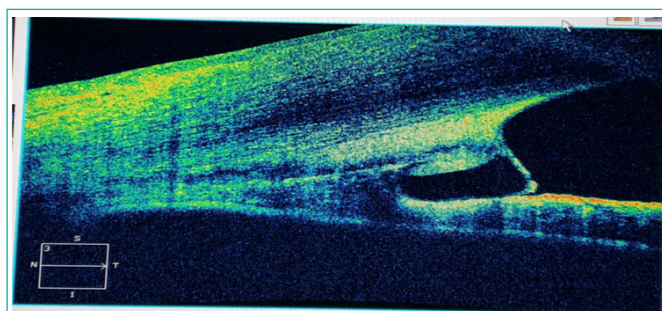


Figure 2: Anterior Segment OCT image along the 7 o'clock meridian (Cirrus HD-5000, Carl Zeiss Meditec).

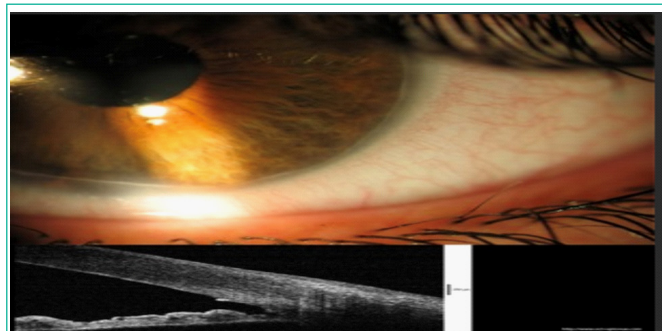


Figure 3: Combined biomicroscopic and AS-OCT image of the iridocorneal angle showing a posterior embryotoxon.

- **Fundus:** normal in both eyes
- **Specular Microscopy:** endothelial-cell density 2200 cells/mm² (OD) and 1700 cells/mm² (OS) with enlarged cells; pachymetry 500 μm in both eyes

• **Visual Field and Optic-Nerve OCT:** normal.

Diagnosis: Irido-Corneal-Endothelial (ICE) Syndrome, likely Chandler variant.

Differential Diagnosis: Axenfeld–Rieger syndrome, excluded due to lack of systemic or developmental anomalies.

Discussion

ICE syndrome represents a continuum of acquired proliferative

endothelial disorders, comprising essential iris atrophy, Chandler's syndrome, and Cogan-Reese (iris naevus) syndrome. The hall mark feature is migration of abnormal endothelial cells that form a contractile membrane, progressive involving the angle and iris, resulting in synechiae, corectopia, and glaucoma. AS-OCT is particularly useful because it allows non-contact, high-resolution imaging of the anterior segment. It can reveal the fibrocellular membrane and peripheral anterior synechiae even when gonioscopy is obscured. It also assists in evaluating corneal thickness and chamber depth. Furthermore, AS-OCT provides quantitative data for long-term monitoring, enabling the detection of progressive angle closure or corneal decompensation. When combined with optic-nerve and ganglion-cell OCT, it contributes to early identification of glaucomatous progression.

Conclusion

ICE syndrome is a rare, progressive endothelial disorder that may lead to significant visual morbidity from glaucoma or corneal decompensation. AS-OCT serves as an indispensable, reproducible imaging modality for diagnosis and follow-up. Its integration into the evaluation of unilateral anterior-segment abnormalities is strongly recommended.

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