International Journal of Science and Research (IJSR)
ISSN: 2319-7064
Impact Factor 2024: 7.101

Uncommon Bilateral Involvement in Iridocorneal Endothelial Syndrome: A Case Report

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Abstract: ***Iridocorneal endothelial (ICE) syndrome is an uncommon ocular condition caused by abnormal behaviour of corneal endothelial cells, resulting in progressive iris atrophy, corectopia, peripheral anterior synechiae, and secondary angle involvement. The disease typically affects one eye, while bilateral presentation is rare and may delay diagnosis. This case report highlights a 41-year-old male presented with gradual, painless blurring of vision in both eyes. On examination revealed inferonasal corectopia, diffuse iris atrophy, extensive peripheral anterior synechiae, adherent leucoma, and an irregular anterior chamber depth. Specular microscopy demonstrated endothelial pleomorphism, loss of hexagonality, and a characteristic dark–light reversal pattern, supporting the diagnosis of ICE syndrome. This case highlights a rare bilateral presentation of ICE syndrome without secondary glaucoma and underscores the importance of endothelial evaluation and long-term follow-up.***

Keywords: Iridocorneal endothelial syndrome; Corectopia; Peripheral anterior synechiae; Specular microscopy

1. Introduction

Iridocorneal endothelial (ICE) syndrome comprises a spectrum of disorders characterized by abnormal proliferation and migration of corneal endothelial cells [1]. The three clinical variants include Chandler syndrome, essential iris atrophy, and Cogan–Reese syndrome [1,2]. These abnormalities lead to progressive iris distortion, formation of peripheral anterior synechiae, and secondary angle closure glaucoma [3]. ICE syndrome usually presents unilaterally; bilateral involvement is rare and sparsely reported in literature [2]. We report a rare case of bilateral ICE syndrome presenting with characteristic anterior segment findings.

2. Case Report

A 41-year-old male presented with complaints of gradual blurring of vision in both eyes. There was no associated pain, redness, photophobia, watering, or history of trauma. Systemic examination and vital parameters were within normal limits.

On ocular examination, the right eye visual acuity was 6/60, improving to 6/24 with pinhole, and BCVA was 6/24 with −2.75 diopters cylinder at 170 degrees. Slit-lamp examination revealed inferonasal displacement of the pupil (corectopia), diffuse iris atrophy, extensive peripheral anterior synechiae, adherent leucoma, and an irregular anterior chamber depth with inferior shallowing. Fundus examination was not possible due to a small slit-like pupil. B-scan ultrasonography showed evidence of chronic vitritis.

The left eye had a visual acuity of 6/24. Anterior segment examination showed grade 2 pterygium, irregular pupil, peripheral anterior synechiae, adherent leucoma, and nuclear sclerosis grade 1. Fundus examination revealed a cup–disc ratio of 0.4 with a healthy neuroretinal rim and normal macula and peripheral retina.

Intraocular pressure measured by applanation tonometry was 10 mmHg in the right eye and 12 mmHg in the left eye. Gonioscopy using a four-mirror Zeiss lens demonstrated peripheral anterior synechiae extending up to Schwalbe's line in the nasal, inferior, and temporal quadrants in both eyes. Specular microscopy revealed loss of normal hexagonal endothelial cell pattern with pleomorphism and a characteristic dark–light reversal pattern, confirming the diagnosis of ICE syndrome [2].

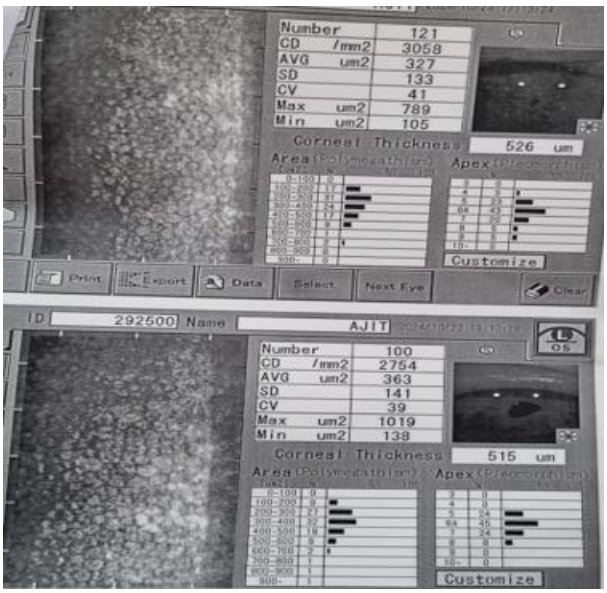

Image 1: Specular microscopy both eye

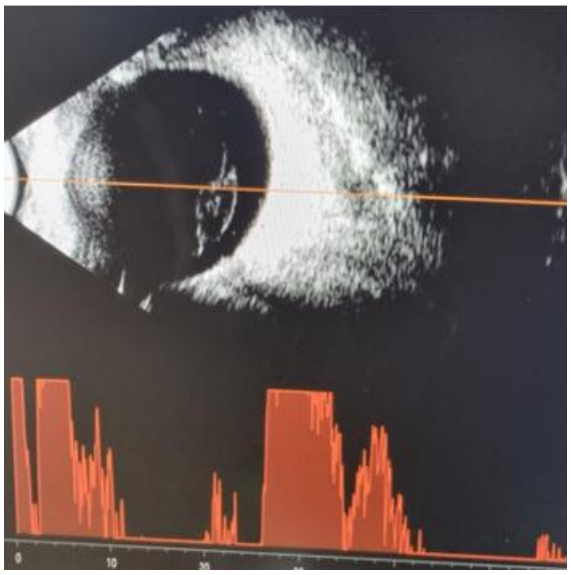

Image 2: B- scan image of right eye

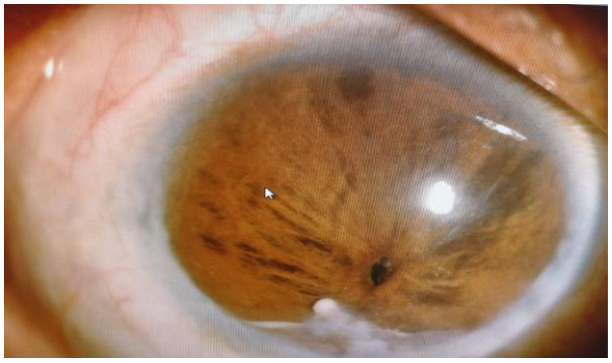

Image 3: Right eye slit lamp photograph

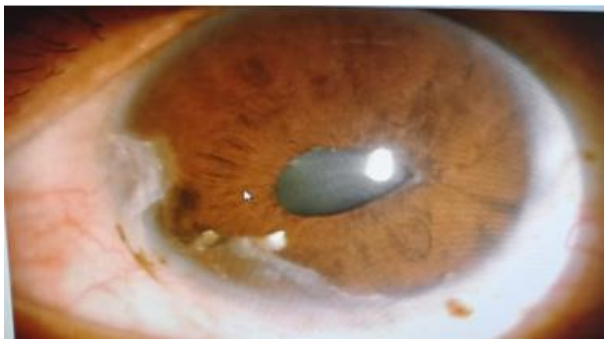

Image 4: Left eye slit lamp photograph

3. Discussion

ICE syndrome is a rare disorder typically affecting middle-aged adults and is characterized by abnormal endothelial cells that acquire epithelial-like properties [1,2]. These cells migrate across the trabecular meshwork and iris surface, forming a contractile membrane that leads to iris distortion, corectopia, and progressive angle closure [2]. Specular microscopy is a valuable diagnostic tool, demonstrating endothelial pleomorphism and the characteristic dark–light reversal pattern [2].

Secondary glaucoma is a common complication of ICE syndrome due to progressive synechial angle closure; however, patients may initially present with normal intraocular pressure and optic disc appearance, as seen in this case [3,4]. Bilateral involvement, although rare, necessitates careful and long-term follow-up to detect disease progression and prevent vision-threatening complications [2].

4. Conclusion

Bilateral presentation of ICE syndrome is rare and may occur without secondary glaucoma in the early stages. Detailed anterior segment evaluation, specular microscopy, and regular follow-up are essential for early detection of complications and optimal patient management [1,3].

Declaration of patient consent
The authors certify that they have obtained all appropriate patients consent forms. In the form, the patient has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initial will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship: Nil

Conflicts of interest: There are no conflict of interest.

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