

## ICE SYNDROME – CASE REPORT

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### Abstract

Iridocorneal endothelial (ICE) syndrome encompasses a group of rare ocular pathologies with unilateral involvement, frequently affecting young women. The disease complex includes essential iris atrophy, Chandler's syndrome, and Cogan-Reese syndrome.

In the following article, we present a case of Iridocorneal endothelial syndrome in which a late diagnosis was made and who underwent surgery for advanced glaucoma.

**Keywords:** Essential iris atrophy, glaucoma, trabeculectomy

Iridocorneal Endothelial Syndrome (ICE) syndrome is a unique ophthalmic disorder that involves an abnormal corneal endothelium that leads to varying degrees of corneal edema, iris atrophy, and secondary angle-closure glaucoma [1]. This syndrome, which typically affects young women unilaterally with no family history [2], encompasses three clinical variants: Chandler Syndrome, Essential (Progressive) Iris Atrophy, Cogan-Reese Syndrome (Iris Nevus Syndrome).

The true etiology of ICE syndrome is unclear. Alvarado et al. have proposed a viral cause for the disease, based on a history of inflammation in certain cases and on the presence of inflammatory cells on histological analysis [3]. Further exploring this hypothesis, the same author revealed Herpes Simplex DNA in the pathological corneas by using the PCR (Polymerase Chain Reaction) technique [4].

The pathological elements observed in the endothelium are the most important clinical findings seen in ICE syndrome, namely "the ICE cell" which is pathognomonic. These cells are abnormally large and show increased pleomorphism [5]. Desmosomes, tonofilaments and numerous microvilli (100 versus 10 in a

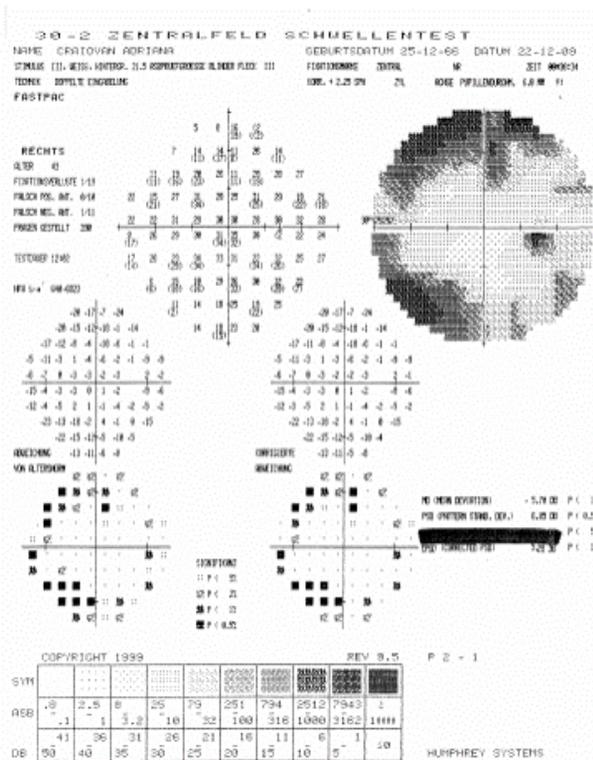
normal endothelial cell) [3], have all been identified by means of electronic microscopy, proving that the ICE cell shows epithelial-like characteristics [6]. The abnormal endothelial cells may migrate posteriorly, forming a membrane that covers the adjacent structures, iris and trabecular meshwork [7]. The contraction of this membrane leads to characteristic iris changes, iridotrabecular synechiae, corectopia with the pupil being drawn towards the area where the synechiae are most prevalent and to secondary angle-closure glaucoma [5]. Glaucoma may appear in the absence of synechiae, because of the membrane migration phenomenon that can functionally close the angle, but still allow for an open angle on gonioscopy [2]. Hence, the degree of angle closure is not associated with the IOP level [8].

### History

A 43-year-old female, with no relevant familial history, was admitted to our clinic for blurred vision in her left eye, reevaluation and treatment. The patient had been admitted to a

clinic in Vienna six weeks prior for blurred vision in her left eye, nausea drowsiness and vomiting.

Upon examination in Vienna, the patient had BCVA 0,2 OS, IOP OS 80mmHg. After managing the acute phase, clinical examination showed temporal pupil traction, pigment dispersion on the endothelium and cup-disk ratio 0,9. The patient was investigated (**Fig. 1** Humphrey Visual Field) and released with the diagnosis of OS Rieger Anomaly. Secondary Glaucoma and maximal glaucoma topical medication ( $\beta$ -blocker, CAI, prostaglandin analogue,  $\alpha_2$ -agonist).



On gonioscopy, the angle was completely closed and the cup-disk ratio was 0,9 with nasal shifting of central vessels and peri-papillary atrophy (Fig. 4).

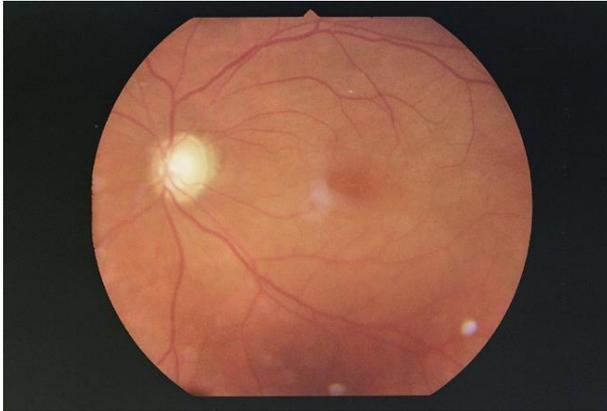


Fig. 4 Posterior pole OS

A clinical diagnosis of OS ICE syndrome with secondary glaucoma was made.

**Ancillary testing**

Visual field analysis, optic nerve head OCT (Fig. 5) and specular microscopy (Fig. 6) were performed.

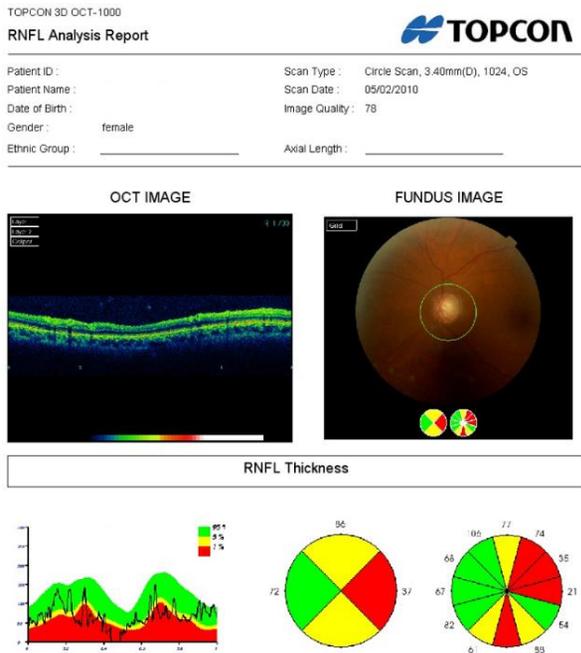


Fig. 5 The ONH OCT shows marked loss of RNFL fibers in the temporal, inferior, and superior sector (in this order)

**IMAGEnet 2000 Endothelial Cell Analysis**

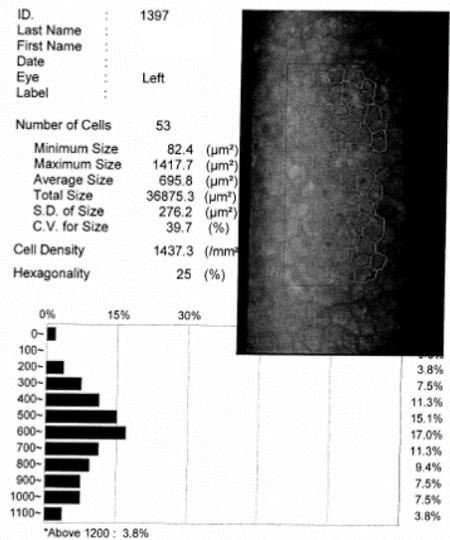


Fig. 6 Specular microscopy – endothelial cell count – shows polymegathism with a significant rise in the mean cell area (695.8µm<sup>2</sup>) compared to the normal value (537.0 ± 137.4 µm<sup>2</sup>) [9] and to the other eye (307.6 µm<sup>2</sup>)

A final diagnosis was made based on the epidemiological data (unilateral symptoms in a young female with no family history), patient history (sudden onset), clinical examination (ocular symptoms with no systemic manifestations), and ancillary tests (specular microscopy being useful).

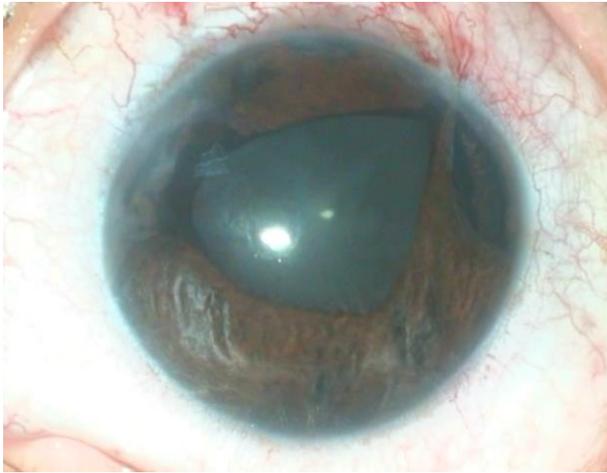
The final diagnosis was OS Essential Iris Atrophy (ICE Syndrome) with secondary glaucoma.

**Follow-up and management**

Surgery was recommended because of uncontrolled IOP in spite of maximal local treatment. A combined trabeculotomy-trabeculectomy with peripheral iridectomy was performed.

Surgery was uneventful and the following morning IOP OS was of 19mmHg with a medium anterior chamber depth. Glaucoma medication was stopped.

At the one month follow-up, BCVA OS was 0,6, IOP OS was 18mmHg without treatment, the ACD (anterior chamber depth) was medium and the filtering bleb was functional (Fig. 7).



**Fig. 7** Postoperative anterior segment OS

At the 4 months follow-up, BCVA OS was 0,6, IOP OS was 20mmHg without treatment, the ACD was medium and the filtering bleb was functional. A visual field was performed, which showed MD=-19.21dB and the patient was given topical medication (fixed combination dorzolamide-timolol).

## Discussion

Trabeculectomy is the surgery of choice for ICE syndrome. Shields et al. have reported a 69% success rate in a study conducted in 1978 on 33 eyes [10], while Yanoff reported a 64% success rate 1 year postoperatively and a 36% at 3 years [8]. When the trabeculectomy proves to be ineffective, the reason is usually excessive subconjunctival scarring [11] (a frequent occurrence in patients with ICE syndrome, given their young age). ICE-specific phenomena that lead to failure are bleb and/ or filtering ostium endothelialization [12] and PAS formation that obstruct the drainage pathway.

## Case particularity

This case stands out due to its complexity and the controversies associated with it (3 different diagnoses from 3 different clinics). A long-term follow-up is necessary because the disease itself is progressive in nature. Studies suggest follow-up at 2-3 months intervals when glaucoma is associated and depending on its severity. Serologic testing is also recommended (Epstein-Barr and Herpes Simplex viruses) [13].

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