

IRIDOCORNEAL ENDOTHELIAL SYNDROME

Rakshya Panta Sitoula

Introduction:

The iridocorneal endothelial (ICE) syndrome is characterized by abnormalities of the corneal endothelium, iris, and anterior segment leading to secondary angle closure glaucoma. ICE syndrome is usually unilateral, nonhereditary and most common in white women in the third to fifth decades.

Three histological variants are Chandler syndrome, progressive (essential) iris atrophy, and Cogan-Reese (iris-nevus) syndrome¹. Dysfunctional endothelial cells migrate over the trabecular meshwork and anterior portion of the iris, resulting in peripheral anterior synechiae, ectropion uvea, corectopia, and iris hole formation².

Thirty nine years female was referred with the diagnosis of Glaucoma due to the elevated IOP. She gave a history of pressure pain on and off in right eye and blurred vision.

She had no other significant past history or family history. On examination her VA was 20/20 OU. She had normal iris contour, deep AC with IOP of 24, 8 mm in OD and OS respectively. On gonioscopy (Figure 1) there were areas of pigmentation extending from 11:30 to 2 and 7 to 10 o'clock and PAS formation nasally and temporally in the right eye. Gonioscopy on the left eye was normal. Vertical cup disc ratio was 0.8 with inferior thinning OD and normal in OS. VF OD showed paracentral arcuate scotoma and was WNL in OS.



Fig 1

She was started on Lumigan with the differential diagnosis of Ring melanoma, Trabecular neavus, ICE syndrome and Secondary glaucoma. Specular microscopy (Figure2) OD revealed polymegathism, polymorphism and cell density of 1500/mm² and was normal in OS.

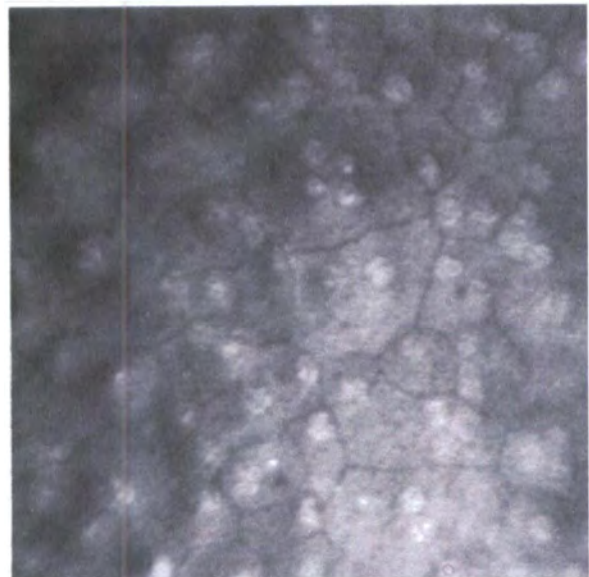


Fig 2

Despite treating her with timolol, dorzolamide, latanoprost and brimonidine her IOP was constantly high hence was planned for Trabeculectomy with MMC. She underwent Trabeculectomy with Mitomycin C (0.4mg/mL) OD. She underwent multiple TCNR (transconjunctival needle revision) after the surgery for the elevated IOP. Barvealt tube was implanted, for uncontrolled IOP and progressive worsening of visual fields. Since the tube surgery IOP has been under control and she is following up in the office every 3 months.

Initially when the patient presented to us she had no other features suggestive of ICE syndrome only sign evident was trabecular hyperpigmentation on gonioscopy. At presentation, significant trabecular hyperpigmentation and PAS formation with elevated IOP favoured the diagnosis of iris melanocytosis, ring melanoma or trabecular

nevus rather than ICE syndrome. Hammered silver appearance of the posterior corneal surface was not evident in our case in early stage of the disease³. This is an unusual case of ICE syndrome with trabecular As ICE syndrome can present.

With a wide spectrum of signs, clinicians should be vigilant and carefully look for signs of ICE syndrome.

References:

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2. **D.G. Campbell, M.B. Shields, T.R. Smith** The corneal endothelium and the spectrum of essential iris atrophy *Am J Ophthalmol*, 86 (1978), pp. 317–324
3. **Wilson MC, Shields MB:** A comparison of the clinical variations of the iridocorneal endothelial syndrome, *Arch Ophthalmol* 107:1465, 1989

Corresponding Address: Dr. Rakshya Pant Sitoula, Biratnagar Eye Hospital, Atithi marg, Rani 17, E-mail: rakshyasitoula@gmail.com