

Concurrence of iridocorneal endothelial syndrome in a patient with glaucomatocyclitic crisis

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Dear Sir,

I am Jin A Choi, from the Department of Ophthalmology of St. Vincent Hospital of Suwon, Kyungki-do, South Korea. I write to present a case report of a young female treated with the diagnosis of Posner-Schlossman syndrome for several years, in which she was ultimately proved to have iridocorneal endothelial (ICE) syndrome.

Posner-Schlossman syndrome (PS syndrome) is characterized by recurrent attacks of mild anterior uveitis with marked elevation of intraocular pressure (IOP)^[1]. The exact etiology of this syndrome is not fully established yet. However, the possible role of herpes simplex virus (HSV) is suggested by a study that revealed DNA evidence of the virus in all aqueous specimens of patients during acute attacks in PS syndrome^[1,2]. ICE is a spectrum of disease that displays common features of corneal endothelial abnormality leading to peripheral anterior synechiae, iris atrophy or nodules^[3]. Using the polymerase chain reaction, HSV DNA was reported to be present within the endothelium of ICE syndrome patients, and HSV-mediated infection of the corneal endothelium was found to transform the endothelial cells to lose contact inhibition and to transform into an epithelial-like cells^[4,5].

A 27 year-old female visited our clinic complaining of discomfort of her left eye (LE) due to acutely raised IOP (36 mm Hg). She did not report any systemic disorders, and familial diseases such as glaucoma or corneal dystrophy

were denied. Her best corrected visual acuity (BCVA) was 20/20 in both eyes. Slit lamp examination showed 1+ anterior chamber cells with some fine keratic precipitates (KPs) in the LE. No specific iris abnormalities were noted, and the posterior segment was also unremarkable. Gonioscopy revealed open-angle without any peripheral anterior synechiae. Standard achromatic automated perimetry (SAP) using a Humphrey visual field analyzer (Carl Zeiss, CA, USA) showed no field defect. There was no evidence of optic disc abnormality indicating glaucoma by fundoscopy (0.5:1 cup to disc ratio with a normal neuroretinal rim). Topical dexamethasone and timolol/dorzolamide were applied, and after 9d, her IOP became normalized and no anterior chamber reaction was noted. She was diagnosed as PS syndrome, and she had 3 to 4 times of relapse/year since then. Two years later, the frequency of relapse had increased to once a month, and anti-glaucoma medications (timolol/dorzolamide, latanoprost, brimonidine) were needed during the quiescent phase.

Four years later after the initial visit, she presented with complaints of blurred vision in her LE. Her BCVA was 20/20, and her IOP was 22 mm Hg. No definite evidence of glaucomatous changes by optical coherence tomography (OCT) 3.0 (Zeiss-Humphrey, CA, USA) was noted. SAP of the LE showed generalized reduction of sensitivity with MD of -4.85 dB ($P < 0.005$).

On slit-lamp examination, there were no changes in the cornea on low magnification. However, on high magnification, fine hammered silver- appearance of entire corneal endothelium was seen (Figure 1). Specular microscopy of both eyes demonstrated ICE cells-a sign of dark-light reversal as well as polymegathism and pleomorphism, which was more prominent in her LE (Figure 2A, 2B), with significantly decreased endothelial cell counts (543 cells/mm²) compared to the right eye (RE) (2114 cells/mm²). The pachymetry of the LE was increased (617 μ m) compared to the RE (568 μ m).

After 6mo, her BCVA was decreased to 20/25. On specular microscopy of the LE, the endothelial cell count was unmeasurable due to corneal edema and more prominent dark-light reversal cells were found (Figure 2C, 2D). The cell counts of the RE was maintained as 2252 cells/mm². She is on the waitlist of penetrating keratoplasty of the LE.

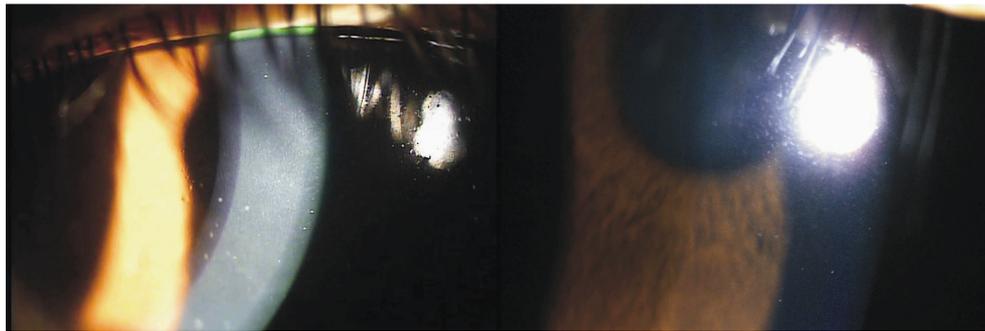


Figure 1 Corneal photography using tangentially applied slit lamp beam On low magnification slit lamp examination, the cornea showed some fine keratic precipitates. On high magnification slit lamp examination, fine hammered silver appearance of entire corneal endothelium was seen.

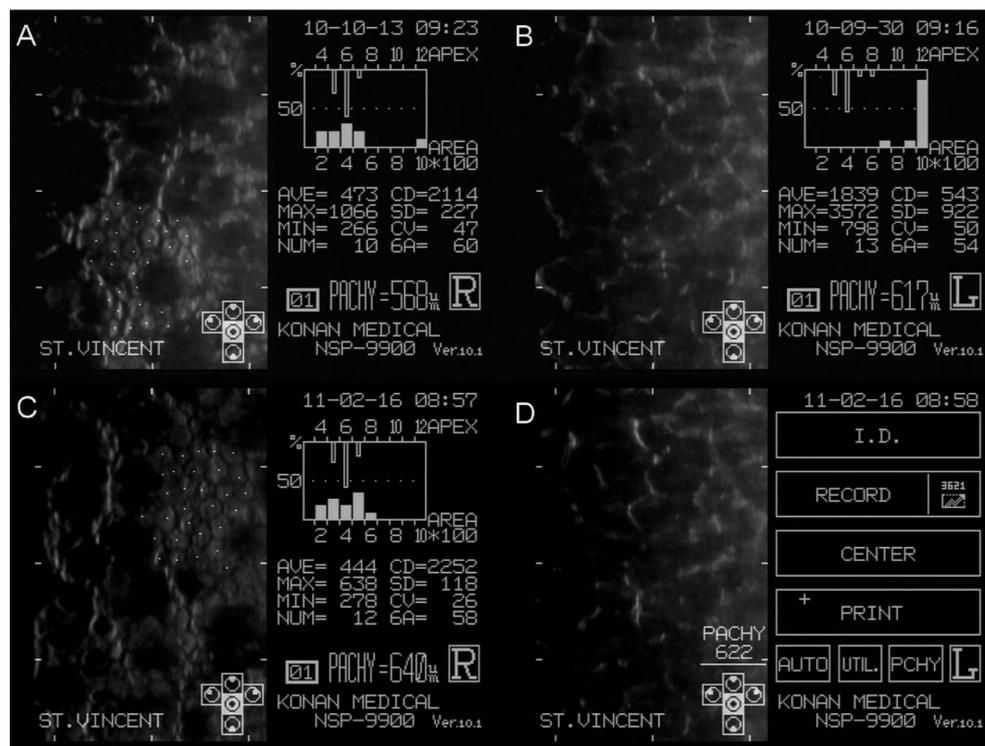


Figure 2 Specular microscopic appearance of the patient in the right eye (A) and the left eye (B) and follow-up examination 6 months later in the right eye (C) and the left eye (D) Specular microscopy of both eyes demonstrated sign of dark-light reversal as well as polymegathism and pleomorphism, which was more prominent in her left eye. On the follow-up examination, more prominent dark-light reversal cells compared to the previous examination were found.

In the study of Setälä and Vannas^[6] who investigated corneal endothelial cells in the PS syndrome, there were no morphological changes in the endothelium except a decrease in endothelial cell density in the PS syndrome. In our case patient, the follow-up specular microscopy showed progressive endothelial cell loss with increased ICE cells, high pleomorphism, a decrease in the percentage of hexagonal cells, and progressively increased corneal thickness. Also, in process of time, IOP was not controlled well even in the quiescent phase, which gives more weight to the diagnosis of ICE syndrome.

The disease complex, which includes essential iris atrophy, Chandler's syndrome, and iris nevus syndrome, is usually unilateral, non-familial, and typically occurs in females

during young adulthood^[3]. However, it has been reported that subclinical abnormalities of the corneal endothelium in the fellow eye are common^[7]. The patient in this case seemed to have Chandler's syndrome among the three clinical variants of ICE syndrome, because she reported no familial inheritance of the ophthalmic disease, and no iris abnormalities such as nodules or atrophy were found. The RE of the patient had demonstrated similar ICE cells, which suggest subclinical involvement of ICE syndrome in the RE. The ICE syndrome should be distinguished from posterior polymorphous dystrophy and Fuchs' endothelial dystrophy. Particularly, Fuchs' endothelial dystrophy may also present a hammered silver endothelial appearance. Although the case patient is not typical ICE syndrome, two former diseases can

be distinguished from ICE syndrome by their familial origin, bilaterality, and lack of typical ICE cells. For the diagnosis in this case, we used the specular microscopy, which is an invaluable tool for confirmatory diagnosis in the ICE syndrome. However, in cases of corneal decompensation, confocal microscopy can provide a useful tool for the diagnosis and differential diagnosis of the ICE syndrome.

As far as we know, this is the first case report that showed the concurrence of PS syndrome and ICE syndrome in the same eye. HSV is suggested to play an important part in the etiology of the diseases as mentioned above [1-2,4-5]. Furthermore, Köhler^[8] had been suggested that PS syndrome, heterochromic cyclitis, anterior-chamber cleavage syndrome and ICE syndrome often produce similar effects at the Descemet membrane, the anterior chamber angle and the iris. Because of the same mesodermal origin of these tissues, the diseases may be merely different clinical expressions of one main disease, which may explain the concurrence of PS syndrome and ICE syndrome in this case.

Although these two distinct clinical manifestations have similar pathogenesis, the prognosis is quite different: ICE syndrome is progressive anterior segment disease that is quite challenging to manage, whereas in PS syndrome, most cases can be controlled well with corticosteroids and anti-glaucoma agent. Therefore, the possibility of the concurrence of the two diseases needs to be considered when IOP is not controlled well with conventional anti-glaucomatic medications in PS syndrome. Furthermore,

detailed corneal evaluation should be performed even in patients with typical features of PS syndrome.

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