Bilateral Chandler's syndrome: Uncommon entity diagnosed by ultrasound biomicroscopy and confocal microscopy

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A 22-year-old female presented with bilateral, progressive diminution of vision. Slit-lamp examination revealed bilateral sectoral corneal edema. Gonioscopy showed broad-based peripheral anterior synechiae and a membrane obscuring angle structure in both the eyes. On ultrasound biomicroscopy (UBM), a membrane extending from corneal endothelium to anterior iris surface causing traction was seen. Confocal microscopy showed an "epithelium-like" transformation of the corneal endothelium. This case demonstrates a bilateral Chandler variant of the iridocorneal endothelial (ICE) syndrome where the diagnosis of Chandler's disease was confirmed by confocal microscopy, after the mechanism of secondary angle closure was demonstrated by the UBM.

Key words: Chandler's syndrome, confocal microscopy, iridocorneal endothelial cells, ultrasound biomicroscopy

Case Report

A 22-year-old Asian Indian female presented with a history of bilateral, painless, progressive diminution of vision. On examination, best-corrected visual acuity was 20/200(OD) and 20/30(OS) and intraocular pressure was 16 mmHg in both the eyes. Slit-lamp examination of the right eye revealed sectoral corneal edema in the nasal quadrant associated with peripheral anterior synechiae (PAS) [Fig. 1a and b, arrow], corectopia, and minimal iris stromal atrophy temporally [Fig. 1c]. The left eye examination revealed sectoral stromal edema accompanied by subepithelial calcareous deposits along with multiple PAS [Fig. 2a-c]. Gonioscopy of both the eyes revealed broad-based PAS and a membrane obscuring the angle structures. Central corneal thickness in the right and left eye were 565 microns and 556 microns, respectively.

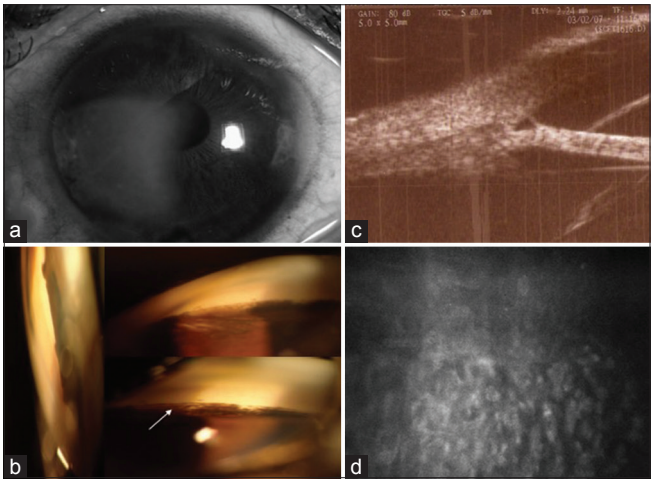

Figure 1: (a) Diffuse illumination photograph of the right eye showing sectoral corneal edema in the nasal quadrant associated with peripheral anterior synechiae and corectopia. There was also minimal iris stromal atrophy temporally. (b) Gonioscopy picture of the right eye showing presence of peripheral anterior synechiae (arrow) and a membrane obscuring angle structures. (c) Ultrasound biomicroscopy scan of the angle showing tenting of iris. (d) Confocal micrograph showing a distinct transition between ICE cells and normal-appearing endothelial cells. The ICE cells were characterized by light-dark reversal, hyper-reflective nuclei, and a highly irregular cellular arrangement

Ultrasound biomicroscopy (UBM Model 840, Paradigm Medical Industries Inc.) of the angle in both the eyes revealed a membrane-like structure extending from corneal endothelium to anterior iris surface. *In-vivo* confocal microscopy (Confoscan 2, Nidek, Japan) clearly showed an "epithelium-like" transformation of the corneal endothelium with polymegathism and hyper-reflective nuclei and surrounding areas with relatively normal-appearing cells. The ICE cells so imaged established the diagnosis of Chandler's syndrome [Figs. 1d and 2d].

Discussion

A review of scientific literature shows sparse evidence.[1-6] Huna *et al*. have reported two different clinical variants in the same subject.[4] In a study from south India, bilaterality was seen in 10% of cases among 223 ICE syndrome patients. Among the eyes with bilateral involvement, six patients had

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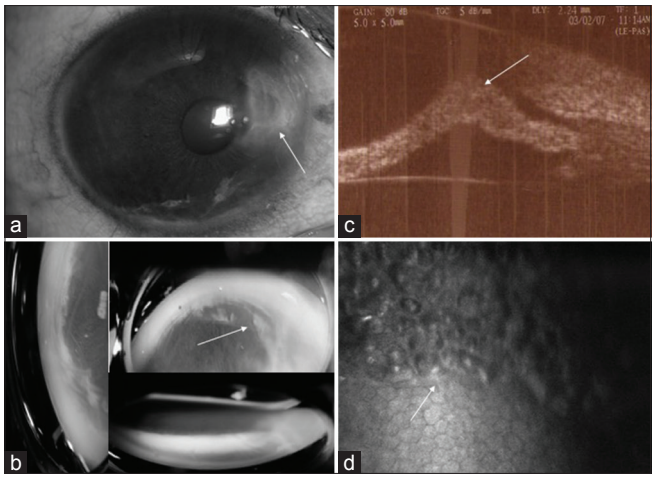

Figure 2: (a) Diffuse illumination photograph of the left eye showing similar findings. The sectoral stromal edema was accompanied by subepithelial calcareous deposits along with multiple peripheral anterior synechiae (b) Gonioscopy picture of the left eye showing the presence of peripheral anterior synechiae obscuring angle structures. Note the heavy pigmentation. (c) Ultrasound biomicroscopy scan of the angle showing a membrane overlying the angle. (d) Confocal micrograph showing large, irregular cells with hyper-reflective nuclei typical of ICE cells. Normal endothelial cells were absent

Chandler's syndrome in both the eyes while five patients had bilateral progressive iris atrophy.[7] Malhotra has reported a case with apparent heterogeneity that may represent two different stages of the disease process at the level of the corneal endothelium.[8] In a recent study by Malhotra and coworkers, out of 21 patients with ICE syndrome, only two patients had bilateral and asymmetric Chandler's disease.[9] These studies suggest that bilaterality of this entity may be unrecognized, rather than as uncommon as was believed. The poor vision in patients with Chandler's syndrome can be explained by corneal endothelial dysfunction (even in subclinical forms).

Zhang and colleagues have used UBM in cases with Chandler's syndrome and noted the presence of marked corneal edema with Descemet's folds while PAS were less evident in their series.[10] Most case reports in literature use only one modality, i.e., confocal microscopy and do not describe the use of UBM to document the angle changes resulting in secondary glaucoma.

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Conflicts of interest

There are no conflicts of interest.

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