Unilateral corneal edema in young: A diagnostic dilemma

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Dear Sir,

We read with great interest the article titled "Unilateral corneal edema in young: A diagnostic dilemma" by Angmo *et al*.[1] We have few observations and suggestions to make. A delayed onset congenital hereditary endothelial dystrophy (CHED) has been seen, but only described in the first decade. The 2015 IC3D findings suggested that autosomal dominant CHED1 was insufficiently distinct to continue as a unique dystrophy and is included in posterior polymorphous dystrophy (PPCD).[2] It would be interesting to see slit-lamp photomicrograph of patient to highlight the endothelium in retroillumination against a dilated fundus to look for vesicles, bands, guttae, or diffuse opacities. PPCD classically starts in teens to early 20s with vesicles, bands, mild thickening, and edema of deep stroma. Broad based or fine peripheral anterior synechiae are present in 25% of eyes with PPCD with 15% presenting with raised intraocular pressure.[3] This could also be a case of early onset Fuchs' endothelial corneal dystrophy (FECD) described as category 1 by IC3D[1] with mutation in gene for alpha 2 chain of collagen VIII (*COL8A2*) on chromosome 1p34.3-p32. Corneal guttae in early onset FECD are small, rounded, and found in patchy distribution in comparison to coarse, distinct guttae of late onset FECD. Another clinical entity called non-guttae endothelial dystrophy has been described by Abbott *et al*.[4] He described phakic young patients with unilateral corneal edema with clinically normal contralateral eyes. There was decreased cell counts in the affected eye nut, no evidence of guttae either clinically or histologically and contralateral eye had pleomorphic endothelial cells on specular microscopy very similar to this case. Moreover *SLC4A11*, a sodium-borate transporter, has been associated with both CHED with progressive deafness (Harboyan syndrome: MIM 217400) and FECD. *LOXHD1* is the second FECD locus involved in both corneal dystrophy and deafness.[5]

It would be interesting to know if the authors send the stripped Descemet's membrane (DM) for histolopathological examination that would aid in differentiating PPCD (multi-layered endothelium with thickened DM), FECD (decreased endothelial cell with embedded guttae bodies), CHED (absence of endothelial cells, thickened DM 20–24 microns).

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Conflicts of interest

There are no conflicts of interest.

References

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