Unilateral corneal edema in young: A diagnostic dilemma

Dewang Angmo, Harathy Selvan, Aswini Kumar Behera, Pramod Kumar Suman

Access this article online

Quick Response Code:

Website: www.ijo.in

DOI: 10.4103/ijo.IJO_564_18

Dr. Rajendra Prasad Centre for Ophthalmic Sciences, All India Institute of Medical Sciences, New Delhi, India

Correspondence to: Dr. Dewang Angmo, Room No. 374, Third Floor, Dr. Rajendra Prasad Centre for Ophthalmic Sciences, All India Institute of Medical Sciences, New Delhi, India. E-mail: dewang45@gmail.com

Manuscript received: 19.04.18; Revision accepted: 16.08.18

Key words: Congenital hereditary corneal dystrophy, corneal edema, Harboyan syndrome, polymegathism, polymorphism, specular microscopy

A 27-year-old male was referred to us as a case of unilateral glaucoma with corneal haze. Elaborate history taking revealed painless diminution in vision OS for past 18 months and presence of hearing impairment since childhood. Family history was not contributory [Fig. 1].

The best-corrected visual acuity OD was 6/6 and OS 1/60. Intraocular pressure (IOP) was OD: 12 and OS: 18 mmHg (on two topical medications) and both eyes had normal corneal sensations. Slit lamp showed OD to be apparently normal while OS demonstrated diffuse corneal haze [Fig. 2]. There

Cite this article as: Angmo D, Selvan H, Behera AK, Suman PK. Unilateral corneal edema in young: A diagnostic dilemma. Indian J Ophthalmol 2018;66:1612-4.

were no keratic precipitates or corneal vascularization, and further clinical evaluation was not possible due to media haze. Due to diagnostic dilemma, we re-evaluated the fellow eye to look for any clue. To our surprise, on specular reflection, the apparently normal fellow eye showed enlarged endothelial cells [Fig. 3]. Specular microscopy of OD showed 650 cells/mm^2 and co-efficient of variation 38. Hexagonality% could not be picked up, but the picture was suggestive of increased polymorphism [Fig. 3]. The specular count of OS was not capturable due to corneal edema. Anterior segment OCT of both eyes showed increased pachymetry, OD: 690 µ and OS: 850 µ (corneal edema) [Fig. 4]. Confoscan of the fellow normal eye confirmed polymegathism [Fig. 3].

Being a bilateral endothelial disease at this young age, a diagnosis of congenital hereditary endothelial dystrophy (CHED)[1] was made. Pure tone audiometry was performed which showed moderate sensorineural hearing loss, implying the diagnosis of the "Harboyan syndrome".[2]

The patient was then advised to stop the topical glaucoma medications one by one under monitoring, even after which the IOP remained within normal range. The patient was started on topical hypertonic saline and registered for endothelial keratoplasty.

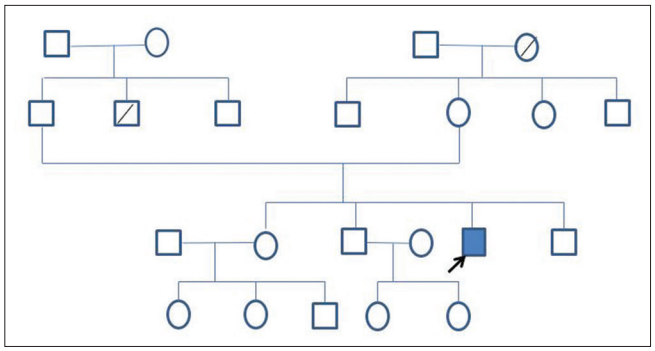

Figure 1: A four generation pedigree chart computed based on the elicited family history

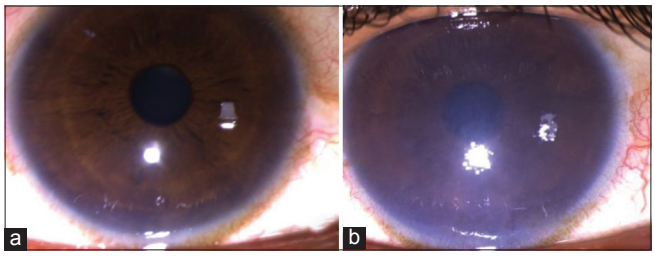

Figure 2: Slit-lamp biomicroscopic photography under diffuse illumination, (a) OD which appears apparently normal. (b) OS with diffuse corneal edema

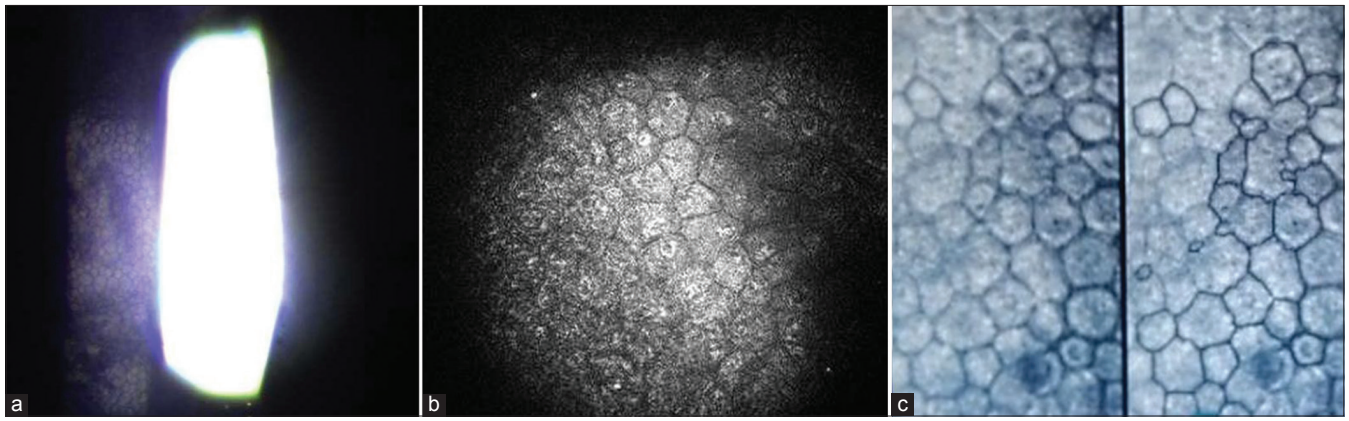

Figure 3: Images of OD, (a) slit-lamp biomicroscopic specular reflection showing enlarged polymorphic endothelial cells. (b) Confocal microscopic section of the endothelium showing the same. (c) Specular microscopy showing, cell count 650/mm^2, and co-efficient of variation 38%. The frame shows spread out, enlarged, and polymorphic endothelial cells

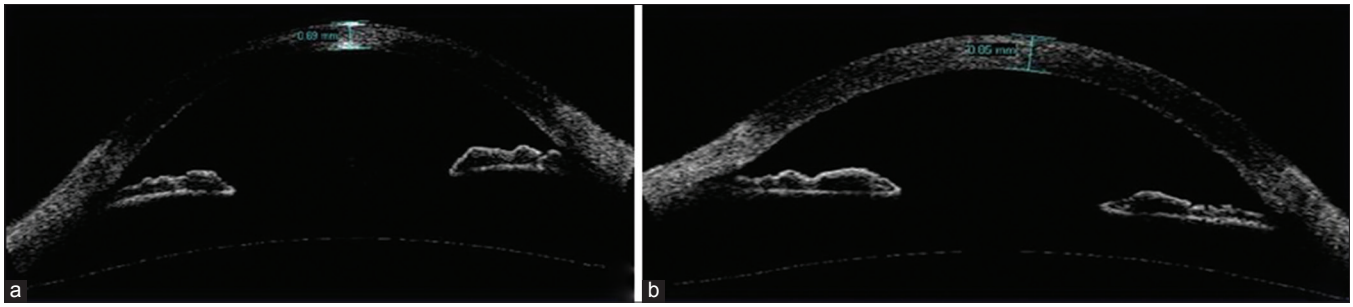

Figure 4: Anterior segment optical coherence tomography images, (a) of the right eye with pachymetry 690µ (b) of the left eye showing diffuse stromal edema (850 µ)

Conclusion

A comprehensive work-up of both eyes is essential before prematurely concluding at the diagnosis, to avoid redundant patient mismanagement.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Weiss JS, Møller HU, Aldave AJ, Seitz B, Bredrup C, Kivelä T, *et al.* IC3D classification of corneal dystrophies – edition 2. Cornea 2015;34:117-59.
2. Desir J, Abramowicz M. Congenital hereditary endothelial dystrophy with progressive sensorineural deafness (Harboyan syndrome). Orphanet J Rare Dis 2008;3:28.