An unusual hydrops in keratoconus

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A young 19-year-old male was referred for the management of acute hydrops in the left eye. During follow-up, other eye cornea developed edematous bands as if hydrops was developing at two parallel sites. Edema progressed very slowly and new edema bands continued to develop. This creeping type of hydrops took 4 months to develop into typical hydrops. To the best of our knowledge, hydrops has always been reported to develop in an acute manner and at a single site. Our case is unusual because hydrops progressed very slowly and developed at multiple sites in a creeping manner.

Key words: Anterior segment optical coherence tomography, hydrops, keratoconus, optical coherence tomography

Acute corneal hydrops is a term used for sudden onset of severe corneal edema due to a tear in stretched Descemet's membrane.[1] Corneal ectasias such as keratoconus,[1] keratoglobus,[2] and pellucid marginal degeneration[3] are associated with hydrops. Young males with advanced keratoconus at presentation and allergic eye disease are more likely to develop acute hydrops.[4] The incidence of hydrops in keratoconus is approximately 3%.[1,5] The term "acute hydrops" was coined by Rychener and Kirby[6] in 1940, which appropriately describes the acute onset of this complication. Since its onset is unpredictable and rapid, there is no literature available to show the evolution of hydrops. There is no clinical or instrument-based marker to predict the onset of hydrops. Recently, Fuentes *et al*.[7] have described optical coherence tomography (OCT) features of impending hydrops.

We are reporting a case of hydrops in keratoconus, in which one eye presented with acute hydrops but other eye had an unusual presentation and course of hydrops. Unlike reported literature, hydrops development was extremely slow and it appeared at multiple sites. Even OCT features were unlike those reported earlier.[7]

Case Report

A 19-year-old Indian male presented with complaints of sudden onset of watering, photophobia, and diminished vision in the left eye. His unaided visual acuity was 20/200 in the right eye and counting finger 2 m in the left eye. His intraocular pressure was 16 mmHg in the right eye with Goldmann applanation tonometer, while it was digitally normal in the left eye. On slit-lamp examination, right eye revealed subtle central two parallel linear lesions at Descemet's level, resembling Haab's striae. The upper linear lesion was just below the center of pupil and the lower ran parallel to it at a distance of 1.5 mm. Lesions were 7 mm long. These were too subtle that we were not able to capture on our photograph slit lamp. Left eye had severe central corneal edema with a cystic space [Fig. 1a]. Edema involved central area, 4 mm in height and 7 mm in width. Rest of the anterior segment examination was normal in both the eyes. He denied any trauma, forceps delivery, or previous hydrops in either eye. He had no history of atopy or vernal keratoconjunctivitis. Young males with these two allergies are more prone to develop keratoconus and hydrops. His right eye topography (Tomey; TMS-5) revealed central steep cornea with maximum k reading of 66.9 D [Fig. 2a]. Specular microscopy was planned in the right eye for endothelial linear lesions, once he was comfortable in the left eye. Hence, with the clinical diagnosis of both eye keratoconus and left eye acute hydrops, conservative medical management comprising hypertonic saline drops (sodium chloride 5%), and prophylactic antibiotic (moxifloxacin 0.3% drops) was started. Option of C3F8 descemetopexy with its pros and cons[5] was discussed, but he opted for conservative management. He was kept under regular follow-up. The cystic space resolved gradually, but severe edema persisted.

After 1 month, he complained of diminished vision in the right eye. Diffuse examination of the right eye revealed two parallel bands of moderate corneal edema with a small strip of clear cornea in between the two bands [Fig. 3a and b]. These bands were at the same location where linear horizontal lesions were noticed on the first visit. Those linear lesions were still visible at the edges of edematous bands [Fig. 3c].On slit examination of the edematous area, anterior two-third stroma had mild haze, but posterior one-third was densely opaque.

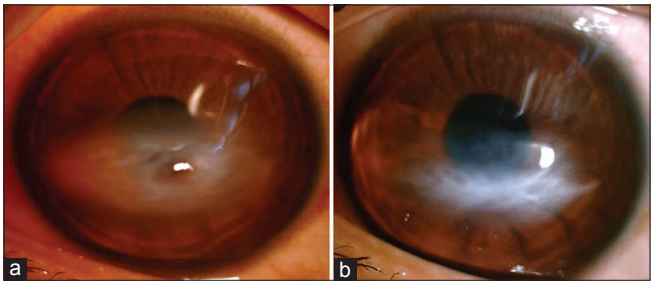

Figure 1: Left eye at presentation showing severe central corneal edema with large cystic space (a), which subsequently scarred after 3 months of conservative management (b)

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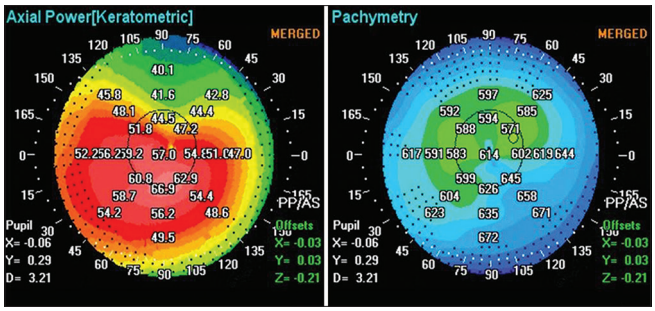

Figure 2: Tomography of the right eye showing very steep cornea on axial power (right) and pachymetry showing thick cornea in ecstatic area (left)

There was no visible Descemet's membrane tear. Left eye cornea showed decrease in edema with scarring at the edges.

Anterior segment OCT (RTVue; Optovue, Inc., Fremont, CA, USA) of the right eye showed findings corroborating with clinical picture, i.e., two areas of hyperreflectivity in deep stroma with intervening normal area. Descemet's membrane detachment was not apparent. Diagnosis of impending hydrops was made in the right eye and hypertonic saline (5%) was started in the right eye too. The edema patches progressed very slowly over next 2 months. New stromal edema bands developed adjacent to previous lesions, again with intervening clear area. Previous edema bands increased in size [Fig. 3d-f]. Meanwhile, corneal edema of the left eye decreased with conservative medical management and vision improved to 20/120 [Fig. 1b]. We expected that right eye too will improve with conservative management, but disease process kept on increasing over next 2 months. Right eye edema bands kept on increasing in width very slowly and merged till the picture resembled a classical hydrops [Fig. 3g]. This entire course took 4 months. This type of slow "creeping" hydrops has never been reported in literature. Since conservative course failed in the right eye, we went ahead with C3F8 descemetopexy under peribulbar anesthesia. After egressing a small volume of aqueous through paracentesis, nonexpansile 0.1 ml C3F8 was injected. Postoperatively, oral 250 mg acetazolamide twice a day was prescribed for 3 days. His edema showed substantial resolution after 3 weeks. After this visit, he was lost to follow up.

Discussion

Acute hydrops is a known complication in keratoconus. Hydrops develops suddenly hence the term "acute hydrops" is considered synonymous with hydrops. However, our case was unusual. First, its course was not acute. It took nearly 4 months for linear lesions on posterior cornea to develop into typical hydrops. Second, till now our understanding of hydrops meant a break in Descemet's. But, in our case, it appeared that hydrops started as endothelial dysfunction at multiple locations and then merged later in the follow-up period. The term endothelial dysfunction is used because edema was present without any apparent Descemet's tear clinically or on OCT. Pachymetry in this eye was also unusual [Fig. 2b]. In keratoconus, corneal ectasia corresponds to thinning, but in this case, thinnest pachymetry was 569 μm and lower ectatic part of cornea was thicker than adjacent cornea. This contradiction also indicates some endothelial dysfunction in ectatic cornea. Whether parallel band-like lesions represented an old resolved hydrops or posterior polymorphous membrane dystrophy, is not clear.

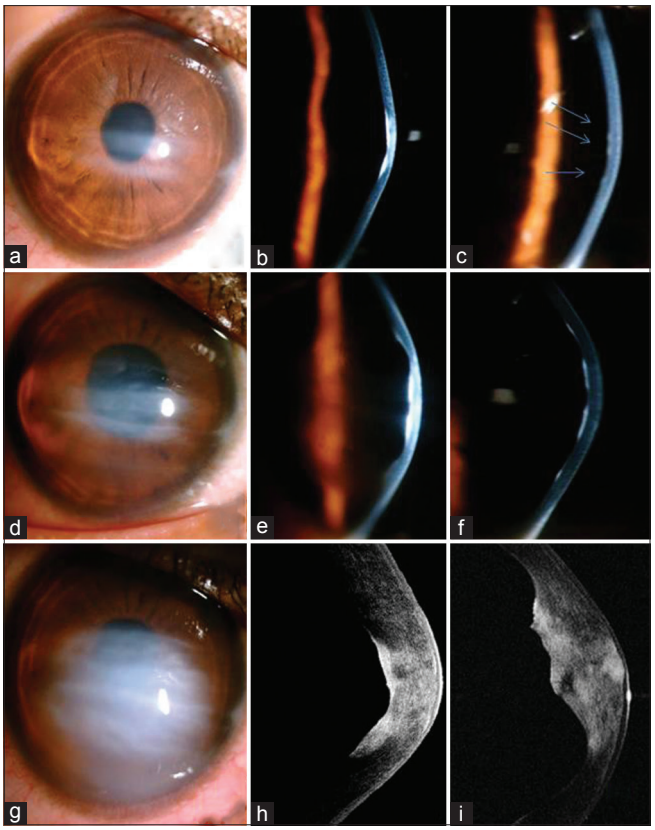

Figure 3: Top row: first appearance of edema, front view (a), slit view of edema bands (b), elevation at Descemet's level (c), middle row: progression after 5 weeks, front view (d), slit at central area (e), slit at peiphery (f), bottom row: full blown hydrops after 4 months (g), optical coherence tomography at first appearance of edema band (h), optical coherence tomography after 2 months of edema progression (i)

Even OCT findings were unusual, unlike reported by Fuentes *et al*.[7] According to this study, predictive OCT findings of hydrops were marked epithelial thickening, stromal thinning, and hyperreflective anomalies occurring at the Bowman's layer level. In our case, epithelium appeared normal in thickness, stroma was not very thin and Bowman's did not show a hyperreflective shadow. OCT in our case had hyperreflective areas corresponding to posterior stromal edema [Fig. 3h]. Even at later stages, OCT findings did not show hyperreflective Bowman's[Fig. 3i].

Conclusion

To the best of our knowledge, creeping presentation of hydrops has not been reported in the literature. Reason for this creeping course of hydrops is not clear. One hypothesis may be that Descemet's microtears lead to endothelial dysfunction, which resulted in very limited edema. As these microtears progressed to adjacent sites, edema may have appeared in the adjacent areas.

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.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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