An unusual case of acute hydrops in a case of regressed retinoblastoma

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Key words: Hydrops, keratoconus, retinoblastoma

An 11-year-old boy, who was a known case of bilateral chemo-reduced intraocular retinoblastoma, presented to our center with sudden onset diminution of vision, whitish discoloration, photophobia, and watering in the right eye for past two days. On examination, he had bilateral periocular fat atrophy, deep superior sulcus, and horizontal jerk nystagmus along with diffuse corneal edema, grossly visible corneal ectasia, and Munson's sign in the right eye

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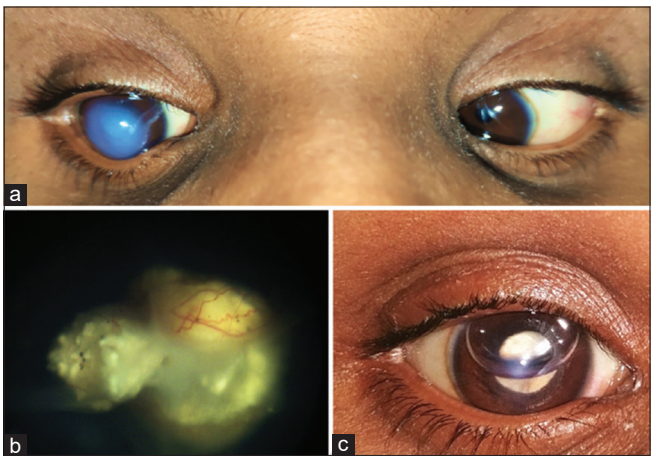

Figure 1: (a) Clinical picture showing periocular fat atrophy and deep superior sulcus in both eyes and acute hydrops with Munson's sign in the right eye along with a left convergent squint. (b) Fundus image (RetCam3, Natus Medical Inc., USA) of the left eye showing a calcified mass at the posterior pole suggestive of regressed retinoblastoma (similar findings present in the right eye). (c) Postoperative clinical picture revealing the drastic resolution in corneal edema and improvement in corneal clarity with a gas bubble in the anterior chamber

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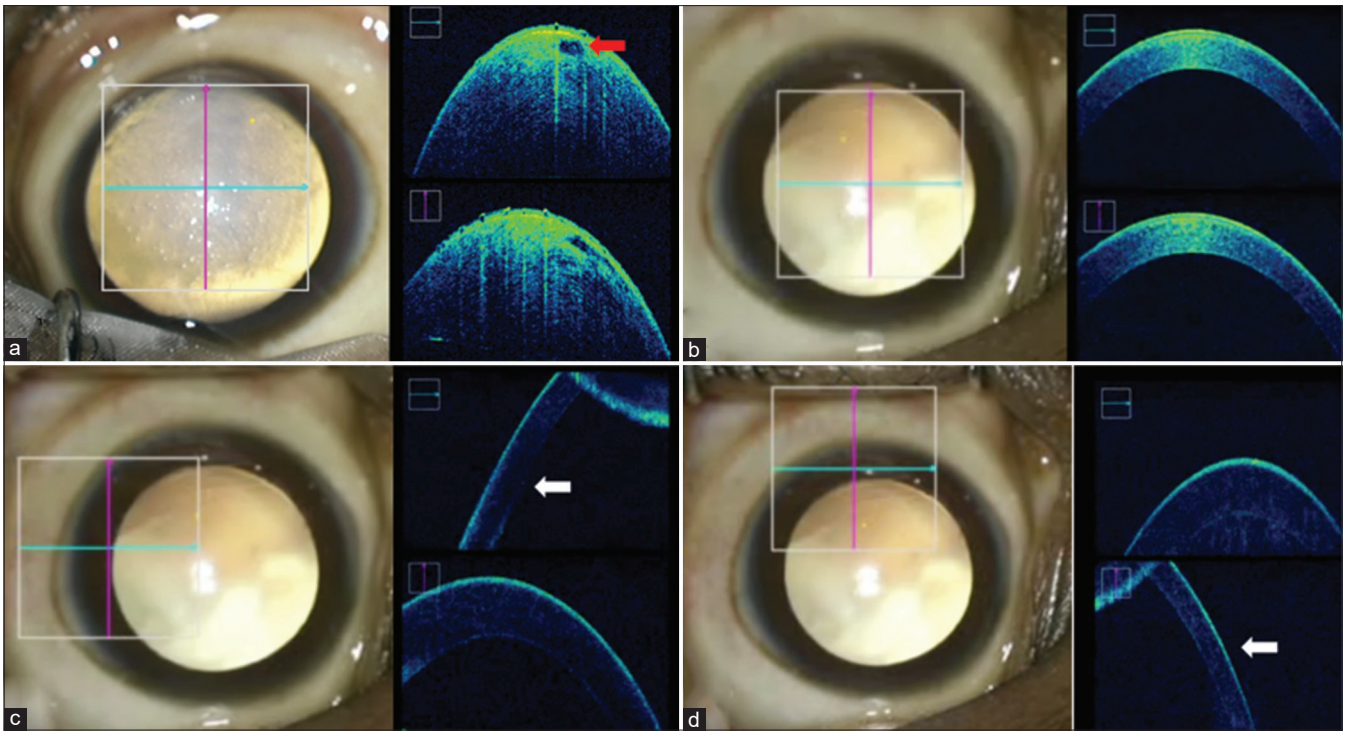

Figure 2: (a) Intraoperative OCT picture of the right eye showing corneal epithelial and stromal edema along with fluid clefts (red arrow). (b) Intraoperative OCT picture of the left eye focussed at the center depicting a decrease in corneal thickness toward the periphery (c and d) Intraoperative OCT pictures of the left eye focussed at the temporal and inferior cornea, respectively, showing significant corneal thinning (white arrows)

and esotropia in the left eye [Fig. 1]. He was provisionally diagnosed as acute hydrops and planned for examination under general anesthesia, during which a diagnosis of bilateral keratoconus with right acute hydrops was confirmed using microscope-integrated Optical Coherence Tomography [Fig. 2]. Intracameral gas injection was performed in view of severe edema and to provide early visual rehabilitation in the only seeing eye, following which hydrops resolved, and since then, the ocular condition has remained stable.

Discussion

Keratoconus, a common ectatic corneal disorder, has been linked with numerous ocular comorbidities, including atopy, allergic conjunctivitis, and congenital retinal dystrophies such as retinitis pigmentosa and Leber congenital amaurosis.[1] Chronic and persistent eye rubbing is well-known to be associated with keratoconus and increased risk of hydrops and various mechanisms of rubbing-related corneal trauma have also been previously described.[2,3] Further, keratoconus is known to be associated with hereditary retinal dystrophies, where-in oculodigital sign and eye rubbing due to poor vision are postulated to be the contributing factors for development of keratoconus.[4,5]

In our case, no relevant ocular or systemic association or family history of keratoconus was present. However, there was significant history of eye rubbing and oculodigital phenomenon due to poor vision, supported by the presence of severe periocular fat atrophy, and in the absence of any other risk factors, it may have contributed to the development of keratoconus and subsequent hydrops. To the best of our knowledge, there is no reported case of hydrops, or even keratoconus, in association with retinoblastoma in literature.

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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