Unilateral acute hydrops in a child with bilateral microcornea and iridofundal coloboma

Rinky Agarwal, Grisilda Nongrem, Prafulla K Maharana, Namrata Sharma

A 15-year-old female child with history of bilateral poor vision since childhood presented with sudden onset pain, photophobia, and diminution of vision OD for 10 days. Visual acuity was hand motion OD and 1/60 OS. Slit lamp examination revealed microcornea OU with multiple intrastromal fluid clefts OD and an irregular cornea and iridofundal coloboma OS. A clinical diagnosis of acute corneal hydrops OD was made, and the child was subjected to intraoperative optical coherence tomography guided intrastromal fluid drainage with air tamponade. The corneal edema resolved completely within 2 weeks resuming visual acuity to 3/60 allowing laser delimitation of fundal coloboma OD.

Key words: Acute hydrops, iridofundal coloboma, microcornea

Acute corneal hydrops (CH) resulting from sudden disruption of Descemet's membrane (DM) is a visually disabling complication of corneal ectatic disorders associated with a prolonged natural course of healing. Microcornea with iridofundal coloboma (IFC) is a rare ophthalmological condition associated with poor vision since childhood. Here, we report a case of CH in a child with IFC coexistent with microcornea managed by intraoperative optical coherence tomography (i-OCT) guided intrastromal puncture with air tamponade.

Case Report

A 15-year-old systemically healthy girl with pre-existing poor vision OU (OS worse than OD) along with inward deviation of left eye since birth presented to our emergency department with sudden onset pain, photophobia, and whitish opacification OD for the past 10 days without any associated redness, history of trauma, previous ocular surgeries, or similar episodes in the past. Presenting visual acuity (VA) was hand motion OD and 1/60 OS. Slit lamp biomicroscopic examination revealed corneal diameter of 8 × 8.5 mm, deep angle recess and quiet anterior chamber (AC), pear-shaped pupil with inferiorly deficient iris, normal intraocular pressure (IOP) and clear crystalline lens OU, central corneal edema with multiple intrastromal clear fluid-filled spaces OD [Fig. 1a and b] and a steep cornea, fundal coloboma (FC) involving disc and macula [Fig. 2a and b] and convergent squint OS. Clinical suspicion of acute CH OD was confirmed on anterior segment optical coherence tomography (ASOCT) which showed a central corneal thickness (CCT) of 0.98 mm along with large multiple anterior and mid stromal clear fluid clefts and on pentacam (Oculus, Inc. GmbH, Germany) which revealed steep cornea with 89.4D/91.2D at 93°/87° dioptric values [Fig. 1c and d]. CCT, keratometric values and axial length OS were 0.52 mm, 35.2D/52.7D at 79°/101° and 20.81 mm, respectively [Fig. 2c]. Patient was managed medically with hypertonic saline 5% QID, homatropine 2% QID, timolol maleate 0.5% BD, loteprednol etabonate 0.5% QID, carboxymethylcellulose 0.5% QID and moxifloxacin hydrochloride 0.5% QID for 2 weeks with no response following which i-OCT (OPMI Lumera 700 and RESCAN 700, Carl Zeiss, Meditec, Germany) guided anterior stromal punctures with intracameral injection of air was performed under general anaesthesia along with laser delimitation of FC OS in the same sitting. The procedure resulted in dramatic resolution of corneal edema followed by resumption of VA to 3/60 and minimal scarring allowing laser delimitation of FC 2 weeks later [Fig. 1e-h].

Discussion

Acute CH is associated with sudden onset pain, photophobia, and diminution of vision resulting from sudden seepage of aqueous fluid into corneal stroma secondary to abrupt rupture of DM.[1] It is most commonly caused by trivial trauma from ocular rubbing and has been reported to occur in numerous ocular conditions such as keratoconus (KC), pellucid marginal degeneration, keratoglobus, and Terrien's marginal degeneration. Microcornea is a rare corneal anomaly presenting with smaller and flatter cornea and IFC is a type of rare congenital malformation associated with defective development of iris and choroid tissue.[2,3] Both microcornea and IFC can lead to poor vision since childhood and have been reported to occur concurrently.[4] Although unilateral acute hydrops has been described in a case of bilateral KC and microcornea in Crouzon's syndrome and unilateral IFC has been reported in an eye with bilateral posterior KC and microcornea, CH in a child with microcornea coexistent with IFC, to the best of our knowledge, has not been yet reported in the literature.[5,6]

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Dr Rajendra Prasad Centre for Ophthalmic Sciences, New Delhi, India

Correspondence to: Dr. Namrata Sharma, Dr Rajendra Prasad Centre for Ophthalmic Sciences, All India Institute of Medical Sciences, Ansari Nagar, New Delhi, Delhi - 110 029, India. E-mail: namrata.sharma@gmail.com

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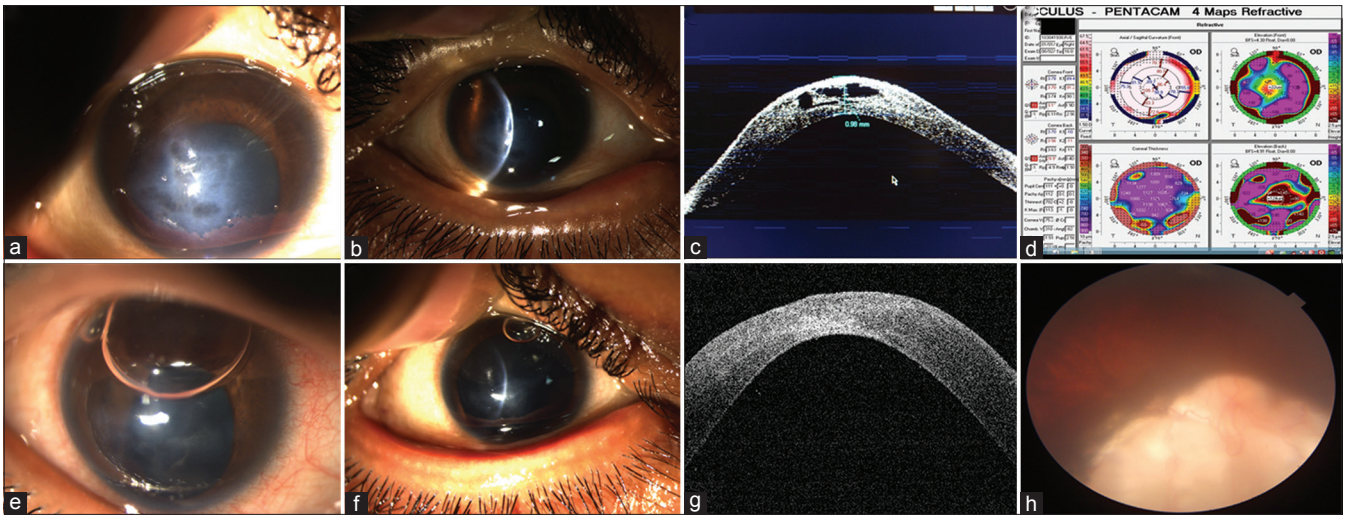

Figure 1: Clinical data OD; (a) Slit lamp photograph showing centrally localized corneal edema; (b and c) Anterior and mid stromal fluid spaces as seen on slit view and anterior segment optical coherence tomography (ASOCT); (d) Pentacam image at presentation; (e and f) Resolving edema on post-operative day (POD) 1 and 3; (g) Resolved edema on ASOCT on POD-3; (h) Laser delimited fundal coloboma on POD-14

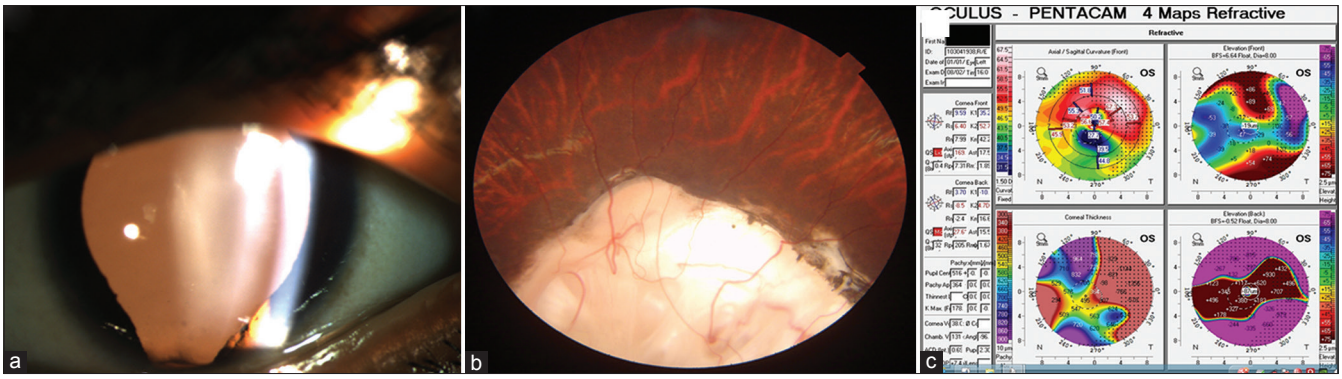

Figure 2: Clinical data OS; (a) inferior iris coloboma as seen on slit lamp; (b) Fundal coloboma involving disc and macula; (c) Pentacam showing irregular cornea

Various differential diagnoses to acute CH in our case included congenital hereditary endothelial dystrophy, corneal abscess, necrotizing viral keratitis, acute angle closure attack, and iridocorneal endothelial syndrome which were ruled out clinically by unilateral presentation, acute onset, young age, lack of circumciliary congestion, purulent discharge, epithelial defect, and keratitic precipitates, presence of deep angle recess and quiet AC, normal IOP and bilaterally symmetrical iris defect. ASOCT and pentacam confirmed our clinical diagnosis. Although most cases of CH resolve spontaneously without any intervention over a period of 2–4 months and surgical intervention such as intracameral injection of air/isoexpansile gases is indicated in eyes with extensive edema and multiple cystic clefts, urgent need for visual rehabilitation (in view of amblyopia in other eye evident from history of worse vision and presence of convergent squint OS), no improvement for two weeks despite medical management and urgent posterior segment examination, made us proceed with i-OCT guided anterior stromal puncture of intrastromal fluid pockets along with intracameral air tamponade.[7,8] Rapid resolution of corneal edema by means of i-OCT guided direct visualization and drainage of stromal fluid pockets away from visual axis followed by air injection not only accelerated visual recovery in our case but also allowed early prophylactic laser delimitation of FC by an experienced retina surgeon.

As prophylactic laser delimitation of FC is a simple procedure known to limit spread of retinal detachment from colobomatous to non-colobomatous area, bilateral laser delimitation of the FC was done as early as possible in our case.[9] Also, our patient has been advised regular follow up with corneal tomography and topography studies to monitor for recurrence of hydrops in same eye and progression of ectasia (in view irregular cornea on pentacam) in other eye to prevent further ocular morbidity.

To conclude from our case, acute CH in a case of microcornea with IFC involving macula needs to be managed attentively and vigilantly to prevent further visual deterioration from anterior segment disturbances in these eyes with already existing poor vision from posterior segment pathology.

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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Conflicts of interest

There are no conflicts of interest.

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