Keratoconus With Acute Hydrops

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This 14-year-old boy with autism presented with sudden visual loss in the right eye. For a week, the eye had been red, irritated, and painful. Three days earlier, "a white bubble" had developed on the cornea and had begun to obscure his vision. The patient had a habit of rubbing his right eye. The conjunctiva was diffusely injected. The cornea had a central area of thinning and bulging with surrounding edema (A). A qualitative visual acuity assessment could not be obtained. A Seidel test to detect an aqueous leak or vitreous exposure (performed by applying fluorescein drops to the eye) was negative. There were no signs of corneal perforation. Keratoconus with acute hydrops was diagnosed.

Keratoconus is a corneal abnormality characterized by a conical shape and central stromal thinning. It has been associated with Down syndrome¹ and eye rubbing.² More recently, it has been reported in 2.3% of persons with intellectual disabilities.³ Hydrops develops when the Descemet membrane ruptures and aqueous fluid flows into the cornea; this causes edema and opacification. The
differential diagnosis for an opacified cornea includes keratoconus with hydrops, corneal ulcer, and increased intraocular pressure, among many other conditions. Ophthalmological examination is advisable to facilitate proper diagnosis and appropriate management. Improper treatment can lead to complications, such as infection and corneal perforation.

Hydrops is not an indication for immediate surgery. With proper treatment, it is reversible and leaves minimal scarring. In patients with severe scarring, corneal transplant may be necessary. Hyperosmotic agents, such as isotonic sodium chloride ointment or drops, can help reduce corneal edema.

Sodium chloride 5% drops, given 3 times a day, and a protective eye shield were prescribed. However, the drops were discontinued 1 week later because the caregiver had difficulty in administering them. Oral acetazolamide, 250 mg twice daily, was started. Three weeks later, the corneal edema was markedly reduced. Six months later, the acute hydrops had resolved, leaving a small central stromal corneal scar, which remained unchanged 1 year later (B).


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