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Case Report Open Access

Acute Corneal Hydrops in Down Syndrome

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Abstract

We report a case of severe acute corneal hydrops in a 32-year-old woman with keratoconus associated with Down syndrome. The anterior segment of her right eye showed an extremely diffuse edematous ectasia of acute corneal hydrops. She was observed rubbing her right eye because of itchiness and discomfort in the eye. Complete resolution of the corneal edema and a central corneal scar was seen at the 6th week of follow-up.

Keywords: Acute corneal hydrops; Keratoconus; Down syndrome; Eye rubbing

Introduction

Keratoconus is a condition in which the cornea assumes a conical shape as a result of noninflammatory thinning of the corneal stroma. The corneal thinning induces irregular astigmatism, myopia, and protrusion, leading to mild to marked impairment of the quality of vision. Keratoconus has been reported in various clinical settings. It may be an isolated sporadic disorder, associated with Down syndrome or Leber congenital amaurosis or due to connective tissue disorders, hard contact lens wear and eye rubbing [1]. An incidence of up to 15% in patients with Down syndrome has been reported in the literature [1]. Acute keratoconus or corneal hydrops is a frequent feature in these patients leading to a further decrease in visual acuity and a mostly central corneal opacification [2]. Herein, we report a case of severe acute corneal hydrops in Down syndrome.

Case Report

A 32-year-old woman with Down syndrome and keratoconus was referred to our hospital with a one week history of ocular pain due to corneal hydrops in her right eye. On initial slit lamp examination, a marked edema of the central corneal stroma was apparent in the right eye (Figure 1).

Best-corrected visual acuity was light perception in the right eye and 0.5 in the left eye. She had a cataract operation 8 years earlier. The postoperative period was uneventful. On further questioning, her family reported that she had been rubbing her right eye because of itchiness and discomfort in the eye. Initial treatment consisted of topical hypertonic agent (4% NaCl). The medical therapy continued for 6 weeks. Six weeks after the initial visit, slit lamp evaluation revealed central corneal scarring with complete resolution of the corneal edema (Figure 2). Her visual acuity improved to 0.1.

Discussion

Keratoconus is a progressive disorder ultimately affecting both eyes. Early in the disease there may be no symptoms, and keratoconus may be noted by the ophthalmologist simply because the patient cannot be refracted to a clear corrected vision. In advanced disease, there is significant distortion of vision accompanied by profound visual loss. Patients with advanced disease may occasionally present with a sudden onset of visual loss accompanied by pain. On slit lamp examination, the conjunctiva may be injected and a diffuse stromal opacity may be noted in the cornea. This condition, referred to as acute corneal hydrops, is caused by the rupture of Descemet's membrane, followed by an influx of aqueous humor into the weakened corneal stroma that leads to a marked edema and the formation of cystic spaces [1,3,4]. The edema may persist for weeks or months, usually diminishing gradually, with relief of pain and resolution of the redness and corneal edema ultimately being replaced by scarring [1]. Down syndrome has been reported to have a high association with keratoconus [5,6], with a reported incidence ranging from 0.5% to 15% [1]. Similarly, there is a high incidence of keratoconus in patients with Leber congenital amaurosis. The frequent occurrence of keratoconus has been attributed to a high incidence of eye rubbing in these two disorders, due to increased blepharitis in Down syndrome and an oculo-digital sign in Leber congenital amaurosis [1]. Eye rubbing has been implicated in the pathogenesis of keratoconus [7-10]. A study of eye rubbing prevalence found that it was reported in 80% of keratoconic subjects and in 58% of controls [6]. Habitual eye rubbing, which is frequently observed in patients with Down syndrome and other forms of mental deficiency, has been postulated as an important factor, not only for the

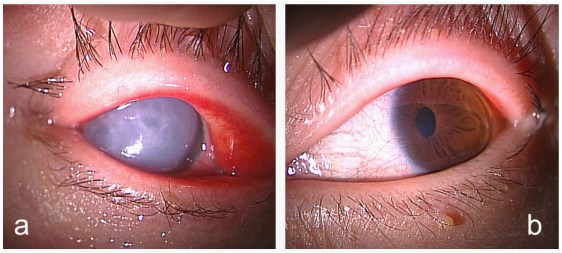

Figure 1: Diffuse lamp photograph of the right cornea (a) showing severe acute corneal hydrops and stromal edema. The left eye (b) showed no acute corneal hydrops.

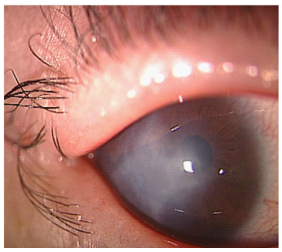

Figure 2: The central corneal scar with complete resolution of stromal edema after medical treatment in the right eye.

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development of keratoconus itself, but also for progression to the acute condition of the disease [1,11]. Occasionally, patients with keratoconus require penetrating keratoplasty for corneal scars [12]. While the development of corneal hydrops in association with keratoconus is well documented, acute hyrdops with perforation is exceedingly rare [3,11]. In this patient, conventional therapy was successful in its management. Regrettably, however, she might require interventional surgery for the progression of corneal opacity in the future.

Finally, in adults with Down syndrome, acquired sensory deficits are significant problems, including loss of vision due to early onset of adult cataracts, recurrent keratitis or keratoconus, and significant hearing loss. In addition, behavioral problems, loss of cognitive abilities, and onset of symptoms of Alzheimer disease pose ongoing challenges for care [13]. The quality of life for adults with Down syndrome can be improved by routine, systematic health care screening to identify treatable diseases.

Conclusions

We suggest keratoconus and/or acute corneal hydrops should be considered as a diagnosis in Down syndrome patients presenting with ocular pain and corneal opacity. Additionally, these patients should be cautioned about rubbing their eyes.

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