Case Report

Bilateral keratoconus, acute hydrops and unilateral corneal perforation due to Tourette syndrome

Melis Palamar a,⇑; Gulsah Dincer b; Mehmet Esat Teker a; Bulent Kayahan b; Ali Saffet Gonul b

Abstract

A 27-year-old male with a repetitive behavior of eyeball pressing for six months presented with decreasing visual acuity for three months. Upon arrival his best corrected visual acuity was 0.2 on the right and 0.6 on the left eyes. Scheimpflug camera system demonstrated grade 4 and grade 2 keratoconus, respectively. Psychiatric consultation revealed Tourette syndrome and treatment was started. Despite psychiatric treatment, acute hydrops occurred in both eyes decreasing visual acuity to hand motions bilaterally. Left corneal perforation due to ongoing habit of eyeball pressure was experienced which led the loss of light perception and phthisis bulbi. Although Tourette syndrome is rare, it might cause bilateral acute onset keratoconus when the repetitive movements affect pericocular region. Psychiatric treatment should be considered as early as possible in these patients in order to prevent unfavorable complications.

Keywords: Cornea, Eye, Hydrops, Keratoconus, Tourette syndrome

Introduction

Tourette syndrome (TS) is a neurologic disorder that is characterized by repetitive muscle contractions that produce stereotyped movements (motor tics) or sounds (vocal tics) which is first described in 1885 by Gilles de la Tourette. Approximately 60% of individuals with TS also exhibit some kind of self injuries behavior and 50% of TS patients have obsessive-compulsive behaviors.1 Eye tics (blinking, alterations in gaze, rubbing of the eyes, blepharospasm, etc.) are well known early manifestations of TS.2 There are reports of self-induced keratoconus, self-induced bilateral retinal detachment and visual field defects as well.3–6

Herein, a case of bilateral self-induced keratoconus in a patient with TS, associated with compulsive eyeball pressing (compulsive pressing over his eyeballs). To the best of our knowledge, this is the most rapid progression (4 months) of keratoconus due to TS in the English literature.

Case report

A 27-year-old male presented with decreasing visual acuity for three months. It was learned the first jerky movements were started when he was 12 years old. In the following 10 years multiple vocal tics (coughing, throat clearing, sniffing) were added. When the patient was 22 years old, he started to yell, swear and hit other people (especially to his
mother) involuntarily. Finally, for the last 6 months the patient started to press severely over his eyeballs until he fainted in order to prevent aggressive impulses like hitting others.

Upon arrival his best corrected visual acuity was 0.2 (−6.50 × 100) on the right and 0.6 (−2.00–6.75 × 75 D) on the left eyes. Corneas were bilaterally clear and posterior segment examinations were unremarkable. Pentacam Scheimpflug camera system (Oculus Optikgeräte GmbH, Wetzlar, Germany) demonstrated grade 4 keratoconus on the right and grade 2 keratoconus on the left eye (Fig. 1). Central corneal thickness (CCT) was 448 and 490 micrometer, respectively. Spectacles were prescribed for visual rehabilitation and he was told not to make pressure to his eyes. Psychiatric consultation revealed TS and the patient was put on aripiprazol 2.5 mg/day, klonazepam 2 mg/day treatment.

Two weeks later on follow-up right BCVA was hand motion (HM), and left BCVA was 0.6. Acute hydrops was evident on the right eye, the cornea on the left eye was clear (Fig. 2A, B). Hypertonic saline solution and hyaluronic acid (Eyestil monodose, Teka, Italy) 8 times/day, cyclopentolate...
hydrochloride (Sikloplejin, Abdi Ibrahim, Turkey) 3 times/day, loteprednol acetate (Lotemax, B&L, US) 4 times/day were started.

On the first month BCVA was HM on the right and 0.5 (−6.00–6.00 x 85 D) on the left eyes. The right cornea was totally opaque. Pentacam evaluation showed that the keratoconus on the left eye progressed to grade 3–4 (Fig. 2C). CCT was 449 micrometer on the left eye. As the patient had no benefit from outpatient follow-up, he was hospitalized in the psychiatry clinic. A combination antidepressant (sertraline 200 mg/day), antiepileptic (carbamazepine 800 mg/day) and antipsychotics (risperidone 2 mg/day and olanzapine 10 mg/day with biperiden 4 mg/day) were initiated. Zuclopentixol 50 mg IM was applied in p.r.n. in case of aggressive behavior. In case of further necessity, he was strained with his and his mother’s approval to prevent further damage to his eyes. After 4 weeks of follow-up, the patient’s symptoms gradually improved and he could resist his urges to press over his eyes. He was discharged with the same treatment and was still under follow-up by the same psychiatric team.

On the fourth month acute hydrops on the left eye as well was evident with a BCVA of HM. Acute hydrops treatment was started. However, as the patient kept pressing his eye balls the left cornea was perforated one week later. A bandage contact lens was inserted and moxifloxacin (Vigamox, Alcon, US) 8 times/day was started. The perforation healed with corneal scar formation with the loss of light perception and phthisis bulbi (Fig. 2D). This case presentation adheres to the tenets of Declaration of Helsinki and the informed consent was obtained.

Discussion

Keratoconus is a bilateral corneal ectasia and the exact pathophysiological mechanism of the disease is still not known. However, eye rubbing is one of the accused factors that lead to keratoconus by triggering apoptosis.7 Keratoconus due to habitual eye rubbing in TS was previously reported.3,4 Kandarakis et al.3 reported a case of bilateral self-induced keratoconus that occurred in 4 years of period in a patient with TS. To the best of our knowledge this is the most rapid bilateral keratoconus and acute hydrops formation due to habitual eye pressing in less than a year. Despite ongoing psychiatric treatment, the repetitive ocular pressing movements could not be prevented and the left eye ended up with corneal perforation which led loss of light perception. As these habitual ocular pressing movements were still continuing a keratoplasty surgery could not be considered.

Herein, we report a patient who developed rapidly progressing bilateral corneal ectasia, acute hydrops and perforation and consequent loss of light perception in one eye during a time period of 4 months. TS is a rare entity, however might cause bilateral acute onset keratoconus when the repetitive movements effect periocular region. As the treatment for TS is difficult and challenging, psychiatric interventions should be considered as soon as possible for similar patients in order to prevent irreversible complications.

Conflict of interest

The authors declared that there is no conflict of interest.

References

1. American Psychiatric Association. Diagnostic and statistical manual of mental disorders American Psychiatric Association; 1994.
2. Golden G. Tourette syndrome: recent advances. Pediatr Neurol 1986;2:189–92.
3. Kandarakis A, Karampelas M, Soumplis V, et al. A case of bilateral self-induced keratoconus in a patient with tourette syndrome associated with compulsive eye rubbing: case report. BMC Ophthalmol 2011;11:28.
4. Mashor RS, Kumar NL, Ritenour RJ, Rootman DS. Keratoconus caused by eye rubbing in patients with Tourette Syndrome. Can J Ophthalmol 2011;46:83–6.
5. Lim S, Rezai KA, Abrams GW, Elliott D. Self-induced, bilateral retinal detachment in Tourette Syndrome. Arch Ophthalmol 2004;122(6):930–1.
6. Enoch JM, Itzhaki A, Lakshminarayanan V, Comerford JP, Lieberman M. Visual field defects detected in patients with Gilles de la Tourette syndrome: preliminary report. Int Ophthalmol 1989;13(5):331–44.
7. Kenney MC, Brown DJ. The cascade hypothesis of keratoconus. Contact Lens Anterior Eye 2003;26:139–46.