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Case report

Self-induced lens subluxation with avulsion of ciliary processes in Tourette Syndrome

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ABSTRACT

Purpose: To report a case of self-induced eye injury resulting in lens subluxation combined with avulsion of ciliary processes in a patient with Tourette Syndrome.

Observation: A 14-year-old male had repeated involuntary trauma to the left side of his face. On exam, left eye lens subluxation combined with ciliary process avulsion were noted. Pars plana vitrectomy and lensectomy were performed.

Conclusions and Importance: This report of self-inflicted ciliary processes detachment in Tourette Syndrome is the first of its kind. Ocular injuries reported in the literature include isolated cases of lens luxation, retinal detachment and orbital hemorrhage. Additionally, iatrogenic conjunctival laceration and corneal abrasion have been reported due to involuntary movements during clinical examination. This case provides further evidence that patients with Tourette Syndrome may be at risk of eye injury due to the involuntary jerk movements associated with this condition. Patients with involuntary tics that put their eyes at risk should be advised to wear safety eye wear to avoid eye trauma and its deleterious visual consequences.

1. Introduction

Gilles de la Tourette syndrome is a neurological disorder characterized by abnormalities of movement and behavior. Motor tics are characterized by involuntary movements such as facial grimacing, blepharospasm, frequent eye blinking and arm jerking. Extreme forms of this disease manifest with self-mutilating behavior. Ocular injuries due to self-harm reported in the literature include isolated cases of lens luxation, retinal detachment and orbital hemorrhage. Additionally, iatrogenic conjunctival laceration and corneal abrasion have been reported during clinical examination due to involuntary movements.

2. Case report

A 14-year-old boy presented with vision loss after repeated self-inflicted punches to the left eye. He was diagnosed with Tourette Syndrome at age 8 and was taking Clonidine, Haloperidol and Clonazepam. Visual acuity (VA) was 20/20 in the right eye and hand motions (HM) in the left eye. He had normal intraocular pressure (IOP) and no afferent pupillary defect (APD). Examination and B-scan ultrasonography showed left subluxated lens with attached ciliary processes, presence of posterior hyaloid detachment and no retinal detachment (Fig. 1). The lens was attached at a hinge superonasally and remaining lens-ciliary processes complex was detached.

The lens and attached ciliary processes were removed with 23 gauge pars plana vitrectomy (PPV). Only 2 clock hours of ciliary processes were still on ciliary body. No retinal breaks or dialyses were seen. Given his propensity for self-harm, we elected to defer placement of a scleral fixated intraocular lens (IOL) implant and a contact lens was recommended for his aphakia. At 10 months of follow-up, he was well adapted to his contact lens (+13.00 sphere) in the left eye and his corrected VA was 20/40, IOP was 20 mmHg and retina was attached. The importance of good hygiene, proper contact lenses care and regular follow-up were discussed with the patient. On gonioscopy, 3 clock-hours of peripheral anterior synechiae (PAS) were noted. UBM demonstrated the loss of ciliary processes (Fig. 2).

3. Discussion

Most severe ophthalmic manifestations of Tourette Syndrome are accidental injuries. This includes risk of ocular trauma during eye examination and physicians should be aware of this risk before
approaching the eye with an instrument or hand. Unfortunately, self-inflicted injuries are common in Tourette syndrome with isolated cases of lens luxation, retinal detachment and orbital hemorrhage reported in the literature. To the best of our knowledge, ciliary processes detachment has not been described previously. Sponsel et al. quantified the impact energy thresholds to produce various eye injuries in a porcine model. They demonstrated that 2 J of energy can induce posterior lens dislocation and zonulolysis, and 7 J can induce severe angle recession and cyclodialysis. What is especially intriguing about our case is the combined ciliary processes avulsion and lens subluxation, which suggests that the zonular attachment to the lens and ciliary processes was stronger than the ciliary processes' attachment to the ciliary muscle. Hence the avulsion of the processes from the longitudinal ciliary muscle fibers by severe blunt trauma. Bornfeld et al. described that the number of zonule elements is markedly reduced with the aging process. Additionally, fibers inserting on the equator of the lens are found exclusively in young eyes. This might help explain the ciliary process avulsion in our case, considering the young age of our patient.

Ciliary body detachment can result in cyclodialysis cleft and hypotony. In our case, ciliary processes-lens complex disconnected at the radial ridges and ciliary body remained attached. Additionally, ocular trauma can cause PAS formation, which might lead to increased IOP due to angle-closure glaucoma by anterior pulling mechanism. The occurrence of trauma-related PAS formation is due to dialysis of the iris root combined with inflammation, cellular proliferation and, sometimes, hyphema that results in a membrane between the iris and the trabecular meshwork, creating the peripheral anterior synechiae. Our patient has a small area of PAS (3 clock-hours) and normal IOP, however, it is unknown if significant loss of processes may lead to decreased aqueous humor secretion or hypotony in the future.

Management of patients with Tourette syndrome is challenging and pharmacotherapy and behavior therapy are crucial to decrease the severity of motor tics. This unique case illustrates that patients with Tourette Syndrome are at risk of ocular injury due to the involuntary jerk movements. We highlight the challenges in visual rehabilitation in self-injury patients. In particular, continued self-injury is a risk factor for future dislocation of any scleral or iris fixated IOL. Physicians should consider recommending contact lenses for the management of aphakia in this scenario, since it may provide good visual results as described in this case report. Finally, patients should be advised to wear safety eye wear to avoid continued eye trauma and its deleterious visual consequences.

Patient consent

Consent to publish the case report was not obtained. This report does not contain any personal information that could lead to the identification of the patient.
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Conflict of interest

The following authors have no financial disclosures: CLMF, KM, PJK.

Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

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