STUDENT ARTICLE

Vernal Keratoconjunctivitis in an Adolescent Female

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CASE PRESENTATION

A 15-year-old African American female was referred to an academic ophthalmology clinic by her ophthalmologist for management of bilateral (OU) keratitis being treated with daily dexamethasone drops. She initially presented to her primary provider with bloodshot eyes with complaints of a foreign body sensation and episodes of "white spots" on her eyes. She was referred to a pediatric ophthalmologist who noted that she had severe photophobia, and her vision was 20/200 OU. She was subsequently started on dexamethasone drops which relieved many of these symptoms and partially improved her vision. Recently however she had worsening blurriness when opening her eyes that was partially resolved with blinking. She denied any visual disturbance, pain, or conjunctival injection. A review of systems was positive only for atopic dermatitis.

Visual testing showed uncorrected distance visual acuity of 20/20 OD (right eye) and 20/70 OS (left eye). Her intraocular pressures were within normal limits at 16 mmHg OD and 13 mmHg OS (normal range: 12-22mmHg). Neurologic examination showed that her pupils were equal, round, and reactive to light bilaterally with full visual fields and intact extraocular movements. Detailed examination via slit lamp revealed significant, large papillae and follicles over the upper tarsal conjunctiva OU (Figures 1 & 2). Corneal examination showed mild inferior scarring and thinning OD and central thinning and scarring OS with vessels encroaching into the cornea (Figure 3). The remainder of the examination including the anterior chamber, iris, and lens revealed no changes or pathology.

The patient was subsequently diagnosed with vernal keratoconjunctivitis (VKC) involving the corneas. It was presumed that she had previous shield ulcers that had healed with residual scarring of the corneas. The patient was switched to 0.05%

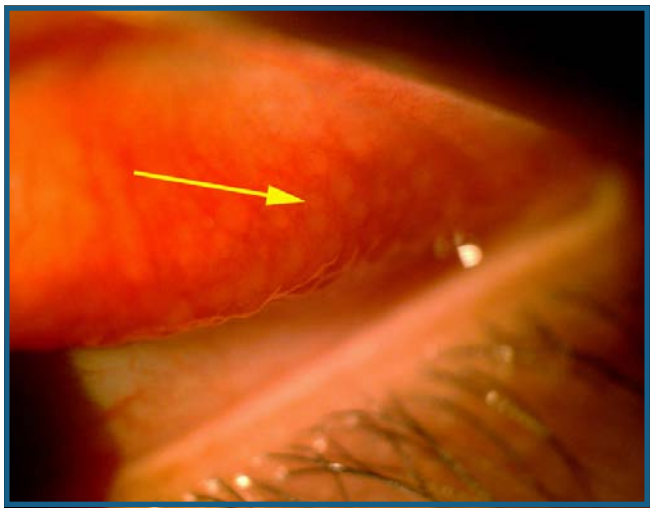

Figure 1: Follicular reaction on the upper tarsal conjunctiva

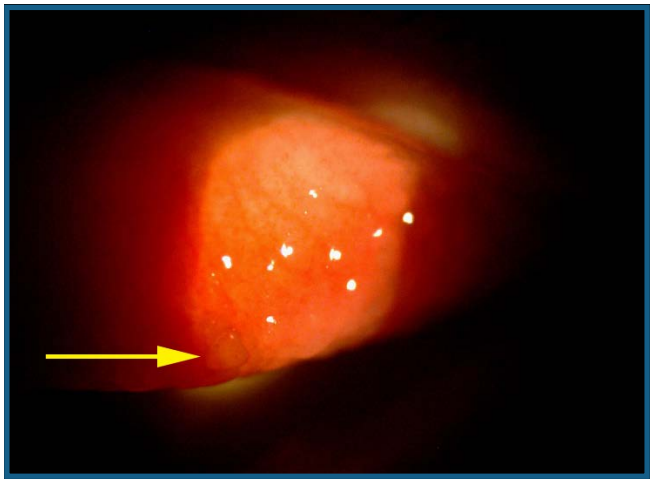

Figure 2: Giant papillae (arrow) on the upper tarsal conjunctiva

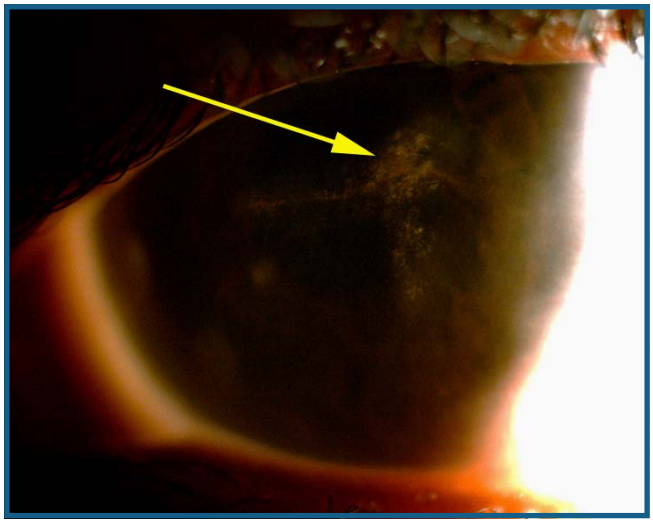

Figure 3: Central corneal scarring from healed shield ulcer

cyclosporine (Restasis®, Allergan) and olopatadine (Pataday®, Alcon) drops to reduce her long-term steroid dependence with the understanding that she would occasionally require steroids (e.g. dexamethasone drops) for future disease flares. Given the vision changes in her left eye, she was counseled that she could possibly benefit from contact lens fitting in that eye in the future if desired.

DISCUSSION

Vernal keratoconjunctivitis (VKC) is a condition of chronic allergic inflammation of the external eye. The condition is more common in hot, arid regions of the world, but is also seen in the United States.[1,2] It tends to occur seasonally (hence "vernal" referring to the Spring) though many patients have recurrent symptoms throughout the year. The condition tends to affect young patients and is more common in males than females.[3] Many patients outgrow the condition as they progress through puberty, while some have symptoms that persist into adulthood. As seen in this patient, a personal or family history of atopy is common. Published studies suggest nearly half of patients with VKC have a family history of autoimmune or immunologic disorders.[4,5]

The pathophysiology of VKC has not been fully elucidated, but immunoglobulin E (IgE)-mediated Type 1 hypersensitivity, T helper cell type 2 (Th2)-mediated Type 4 hypersensitivity, and eosinophil-mediated processes are all believed to be involved.[6,7,8] Due to the difference in prevalence between boys and girls, there may also be an endocrine component, but no formal pathways have yet been identified.[1,9] Ocular allergy testing and the development of ocular biomarkers may help better elucidate the underlying processes.[10]

Patient-reported symptoms often include severe itching, photophobia, tearing, foreign body sensation, discharge (often described as thick or "ropy"), pain, and blurry vision.[11] Three subtypes of VKC exist: conjunctival, limbal, and mixed. The classic examination finding in VKC is giant (greater than 1mm in diameter) "cobblestone-like" papillae on the upper tarsal conjunctiva (Figures 1 & 2) that are visualized by flipping over the upper lid. Other signs include hyperemia, thick discharge, ptosis (droopy eyelid), blepharospasm, opacification of the limbal conjunctiva and peri-limbal Horner-Trantas dots. These punctate white dots represent accumulations of epithelial cells and eosinophils and are consistent with the history of "white dots" reported in this patient report.[1,11] Often most concerning is that progression of the disease can lead to corneal involvement and subsequent vision loss. Corneal involvement ranges from mild punctate erosions to large erosions and ulcers. Some erosions heal completely, others can progress to large shield ulcers (Figure 3) with subsequent neovascularization (as seen in this patient) and vision loss.[12]

As VKC is a chronic condition, treatment similarly requires long-term medication use and routine follow-up. Basic non-pharmacologic approaches include the avoidance of nonspecific triggers such as allergens (e.g., environmental, dander, etc.), the use of cool compresses, and the use of preservative-free artificial tears.[13,14] A proposed grading system exists and may aid in treatment decisions, however the first-line treatment is typically a dual-acting mast cell stabilizer and antihistamine such as olopatadine as prescribed to this patient.[14,15] If the patient does not respond to initial therapy within a few weeks, referral to an ophthalmologist is warranted for possible corticosteroid or other therapeutic considerations. Topical calcineurin inhibitors have been shown in randomized trials to significantly improve signs and symptoms of VKC.[16] Topical cyclosporine is especially preferred in patient with shield ulcers, as was seen in our patient, since they do not inhibit ulcer healing. Even with improved control with cyclosporine, patients may still require occasional topical corticosteroid treatment for flares, as was discussed with this patient.[16] A wide range of other therapies have been studied for VKC including vasoconstrictors, NSAIDs, antimetabolites, and newer monoclonal antibodies such as omalizumab (anti-IgE) with varying success.[13, 17, 18]

Many patients with VKC experience resolution of the disease with puberty. Chronic treatment until resolution may be required and treatment regimens should be tailored based on individual response to treatment and symptom severity.

CONCLUSION

Vernal keratoconjunctivitis is an uncommon, chronic inflammatory condition of the external eye that presents with severe itching, photophobia, tearing, foreign body sensation, discharge, pain, and blurry vision. Examination findings classically include large "cobblestone-like" papillae of the upper tarsal conjunctiva and possible disease involvement of the limbus and/or cornea. Initial treatment includes avoidance of allergens and treatment with a topical dual-acting mast cell destabilizer and antihistamine. Patients who do not respond to initial treatment should be referred to an ophthalmologist to explore other treatment options including calcineurin inhibitors, corticosteroids, and more.

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