CASE REPORT
Iran J Allergy Asthma Immunol
August 2021; 20(4):505-508.
Doi: 10.18502/ijaai.v20i4.6961

A Case of Shield Ulcer Due to Vernal Keratoconjunctivitis

Dilek Özkaya, Gülşah Usta, and Umut Karaca

Department of Ophthalmology, Faculty of Medicine, Süleyman Demirel University, Isparta, Turkey

Received: 30 June 2020; Received in revised form: 14 December 2020; Accepted: 26 December 2020

ABSTRACT

Corneal shield ulcer is an uncommon but serious complication of vernal keratoconjunctivitis (VKC) that can threaten visual acuity. We present a 12-year-old case with a corneal shield ulcer on the superior part of the cornea in the right eye. We learned from his history that he was treated with topical cyclosporine A (CsA) and corneal debridement was performed for the same complaints six months ago. His complaints recurred six months after ceasing topical CsA voluntarily. Topical anti-allergic and CsA treatments were commenced, we also performed corneal debridement. During his follow-ups, the corneal ulcer healed leaving a scar as opacity and neovascularization. This case highlights the role of the anti-inflammatory effect of CsA in preventing the recurrence of shield ulcers.

Keywords: Cyclosporine; Vernal keratoconjunctivitis

INTRODUCTION

Vernal keratoconjunctivitis (VKC) is a recurrent, bilateral, allergic disease of the ocular surface with intermittent seasonal aggravations. The disease is mostly seen in males younger than 10 years of age, and symptoms especially increase between April and August. VKC is a member of a disease group, called allergic conjunctivitis. The other members are atopic keratoconjunctivitis, perennial and seasonal rhino-conjunctivitis, and giant papillary conjunctivitis. The symptoms of VKC are severe itching, burning, irritation, conjunctival redness, increased tear production, swelling of conjunctiva and eyelids, photophobia, and mucous secretion. The clinical findings are commonly related to conjunctival involvement. Conjunctival signs are hypertrophy, infiltrates and nodules in the limbus, and giant papillary hypertrophy in the superior palpebral conjunctiva.[1-2]

Corneal shield ulcer is a rare and painless manifestation that occurs in 3–11% of patients with VKC. Both mechanical injury of corneal epithelium by the giant papillary structures and toxic effects of inflammatory agents released by mast cells and eosinophils are thought to form the pathogenesis of the disease. Shield ulcers should be managed exactly and appropriately for avoiding vision-threatening complications. According to the grade of shield ulcer, treatment alters from topical eye drops to surgical applications.[3] We present a case of corneal shield ulcer treated with topical cyclosporine A (CsA) and corneal debridement.

Corresponding Author: Dilek Özkaya, MD;
Department of Ophthalmology, Faculty of Medicine, Süleyman Demirel University, Isparta, Turkey. Tel: (+90 50) 5478 1377, Fax: (+90 24) 6211 2830, E-mail: drdilekozkaya@yahoo.com

CASE REPORT

A 12-year-old boy presented to our clinic with complaints of stinging, itching, and redness in his right eye for two weeks. He had a history of seasonal attacks due to VKC. His best-corrected visual acuity was 7/10 in the right eye and 10/10 in the left eye. On slit-lamp examination, the conjunctiva was hyperemic, and giant papillae were found on the superior tarsal conjunctiva (Figure 1).

Corneal examination showed a shield ulcer with a plaque (grade-3 shield ulcer), measuring 2x2 mm, staining with fluorescein, between 11-12 o'clock adjacent to the limbus in the right eye (Figure 2).

The left eye examination was normal except for giant papillae on the superior tarsal conjunctiva. Six months ago he was treated for the same complaints and corneal debridement was performed for corneal shield ulcer in another clinic. After healing of the ulcer, treatment was continued with topical CsA 0.1% (Depores X, Deva, Kocaeli, Turkey) eye drop. When his symptoms improved completely, he stopped topical CsA voluntarily. However, his complaints recurred after six months. Treatment was commenced with topical moxifloxacin, olopatadine, loteprednol, artificial tear drops without preservatives, and CsA 0.1% eye drops. Under topical anesthesia, ulcer plaque was extracted with the tip of a 26-gauge needle and the basis of the ulcer was also scrapped at the slit lamp. The right eye was patched for twenty-four hours. His symptoms improved and the epithelial defect healed in two weeks completely. His best-corrected visual acuity was 10/10 in both eyes. Three weeks later, the shield ulcer healed completely leaving neovascularization and a corneal opacity that stained negatively with fluorescein (Figure 3).

The topical steroid was tapered gradually. Treatment was continued with topical CsA 0.1% and he was advised not to stop topical CsA. The follow-up duration was twelve months, and no recurrence was observed.

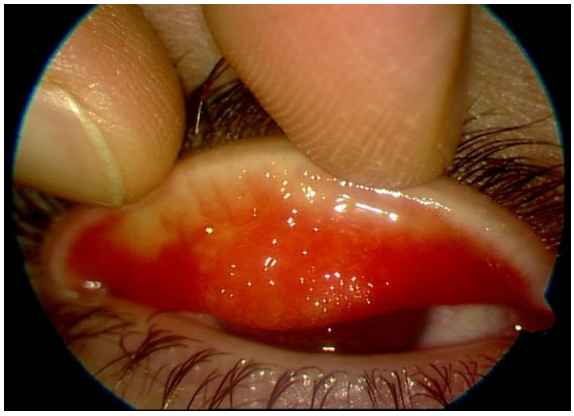

Figure 1. Giant papillae on superior tarsal conjunctiva giving cobblestone appearance

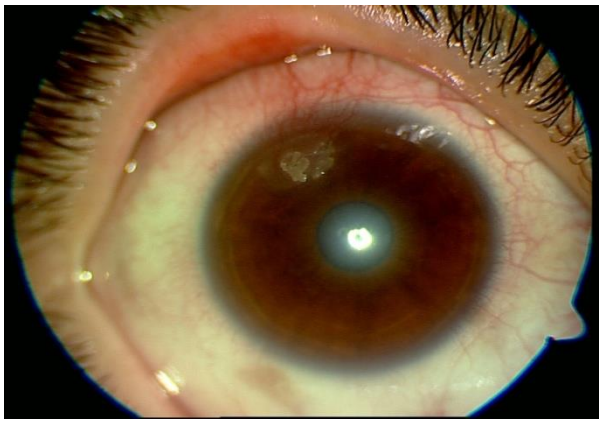

Figure 2. Shield ulcer with a plaque (grade-3 shield ulcer), between 11-12 o'clock adjacent to the limbus in the right eye

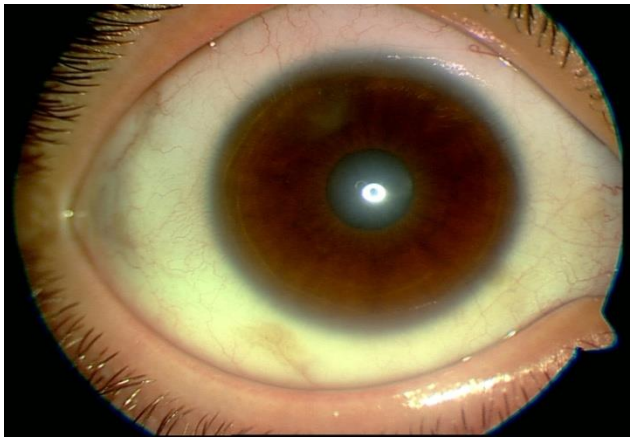

Figure 3. Shield ulcer healed leaving neovascularization and a corneal opacity

DISCUSSION

VKC is an allergic inflammation of the cornea and conjunctiva, primarily affecting children and teens. The etiology of VKC is not completely known. However, VKC is considered as an acute, type 1 Ig E-mediated hypersensitivity reaction. Nearly half of (49%) VKC cases have a family history of atopic disorders. There may be also atopic diseases such as asthma, allergic rhinitis, and atopic dermatitis in the medical history of patients. The presence of giant papillae (cobblestoning appearance) on the superior palpebral conjunctiva is the characteristic sign of the disease. Additionally, papillae at the limbus and deposits of epithelial cells and eosinophils are called 'Horner Trantas dots'.[1,2] The giant papillary structure of the superior palpebral conjunctiva leads to constant mechanical damage, consequently, corneal involvement occurs. Also, inflammatory mediators may increase this injury even more. Shield ulcers, corneal plaques, and opacities, keratoconus, and keratitis may develop due to this persistent damage.[4]

Vernal shield ulcer is characteristically oval or pentagonal shaped, superficial, and locates mostly superior to the cornea. A grayish opacification may be seen at the basis of the ulcer, and the borders may be elevated slightly. If inflammatory material and debris accumulate based on an ulcer, an opaque plaque occurs. Eosinophil major basic protein (MBP) has been determined in these plaques by Trocme et al. They claimed that MBP (a cytotoxic protein) is an important factor in the formation and constancy of shield ulcers. Cameron suggested a classification system for shield ulcers depending on the clinical features. The basis and borders of Grade-1 shield ulcers are clear and there is no apparent inflammatory material. Only medical treatment is generally enough for the regeneration of the epithelium. As a result of inflammatory material at the basis of ulcer, re-epithelization of Grade-2 ulcer is prolonged. They have a poor response to medical therapy, so surgical intervention may be necessary for management. Grade-3 shield ulcers have an elevated plaque above the level of the surrounding epithelium. These ulcers are generally refractive to medical treatment and surgical intervention is necessary.[5-6]

CsA is an immunosuppressive agent, which reduces ocular inflammation by inhibiting T helper 2 lymphocyte proliferation and interleukin-2 production. It inhibits histamine release from mast cells and basophils through a reduction in interleukin-5 production, reduces eosinophil recruitment, and affects the conjunctiva and cornea. Topical CsA is effective and safe in patients with moderate to severe VKC and shield ulcers with a good steroid-sparing effect. Studies have shown that the symptoms and signs of patients treated with 0.1% CsA improved and steroids were stopped in some patients.[7-8] The efficacy of topical CsA in different dosages and installation frequencies has been shown in several studies. Westland and et al. reported three patients with shield ulcers due to VKC who responded quickly with complete re-epithelialization after adding topical CsA to conventional treatment including steroids.[9-10] The surgical debridement aims to extract the inflammatory material and debris from the basis of the ulcer. The inflammatory material and debris are composed of several cationic proteins released from eosinophils and have cytotoxic properties delaying wound healing and

re-epithelialization. In this way, surgical debridement helps the corneal re-epithelialization. The benefits and effects of corneal debridement have been reported in different studies.[11-12] We also observed the efficacy of debridement with topical CsA in our case, as the corneal epithelial wound healed completely in two weeks. After surgical debridement, using topical CsA prevents recurrences by inhibiting T-cell transduction and reducing inflammation. Topical CsA in 0.1% or 0.2% concentrations are more effective in shield ulcer management. Recurrences have been reported by reducing the CsA concentrations to 0.05%. Similarly, after cessation of topical cyclosporine therapy shield ulcer also recurred in our patient. Therefore, using topical CsA in 0.1% concentration is proposed in VKC treatment for preventing the recurrence of clinical signs and shield ulcers.[9]

In conclusion; because of vision-threatening complications, shield ulcers should be treated aggressively. The combination of topical CsA and surgical debridement is the most appropriate treatment for Grade 2-3 shield ulcers. As in the present study, if inflammation due to VKC cannot be effectively suppressed, shield ulcers may recur.

CONFLICT OF INTEREST

There are no conflicts of interest.

ACKNOWLEDGEMENTS

There are no sources of funding.

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