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Peripheral ulcerative keratitis secondary to severe hidradenitis suppurativa

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ABSTRACT

Purpose: To describe a unique case of peripheral ulcerative keratitis secondary to isolated, severe hidradenitis suppurativa (HS).

Observation: A 31-year-old male with HS presented with a red painful right eye with best corrected visual acuity of count fingers at 3 feet with peripheral corneal thinning, inferior descemetocele, and adjacent infiltrate. Work-up revealed negative corneal cultures and positive ANA, ANCA, and rheumatoid factor without other autoimmune or rheumatologic history or symptomatology. He was treated with topical corticosteroids with improvement until he was lost to follow-up before tumor necrosis factor-a inhibitor therapy could be started. Upon re-presentation, he was found to have corneal perforation.

Conclusions and importance: Coexistence of inflammatory eye disease and HS is known but rare, and most commonly manifests as anterior uveitis. Here we present a unique case of peripheral ulcerative keratitis secondary to HS and demonstrate the importance of ophthalmologists' familiarly with this systemic disease and its variety of ocular manifestations.

1. Introduction

Hidradenitis suppurativa is a rare cause of inflammatory eye disease and when present most commonly manifests as anterior uveitis. Herein we report a unique case of peripheral ulcerative keratitis secondary to isolated, severe hidradenitis suppurativa.

2. Case report

A 31-year-old African American man with history of diabetes mellitus, alcohol abuse, and hidradenitis suppurativa (HS) presented with decreased vision and red, painful right eye. His history of HS included open lesions of the bilateral scalp, postauricular areas, axillae, groin, scrotum, buttocks, and perineum with no prior anti-tumor necrosis factor (*anti*-TNF) therapy. His initial examination revealed visual acuity of count fingers at 3 feet OD and 20/20 OS, moderate right conjunctival injection, peripheral infratemporal epithelial defect with 75% stromal thinning and involvement of adjacent limbus and sclera, inferior descemetocele with neovascularization, and temporal stromal infiltrate. There was no anterior chamber reaction and ocular ultrasound revealed no vitritis and no choroidal or scleral thickening to suggest posterior involvement. External examination was notable for multiple nodules and fistulas with purulent drainage of the inguinal folds, medial things, scrotum, and perineum with extensive scarring and lymphedema.

Given severe HS with this unilateral painful corneal ulceration involving the adjacent limbus and sclera, peripheral ulcerative keratitis was considered as the leading diagnosis. Corneal cultures were obtained to rule out infectious etiology and returned negative. There was no inciting history to suspect a traumatic or neurotrophic etiology. The patient underwent temporal conjunctival recession to distance inflammatory infiltrate from the affected cornea. Further, given the risk of progressive thinning, the cornea was glued, and an overlying bandage contact lens was placed (Fig. 1).

Alternative etiologies for peripheral ulcerative keratitis were investigated. Serologic work-up revealed positive ANA 1:40, ANCA 1:80, and Rheumatoid Factor, albeit at low titers. Inflammatory markers ESR and CRP were elevated. The patient was negative for HIV, HBV, HCV; he had no other autoimmune or rheumatologic history or symptomatology. Serum chemistry panel including creatinine was normal, and urinalysis demonstrated proteinuria without hematuria. This was attributed to his concurrently poorly controlled type 2 diabetes mellitus. Complete blood count was demonstrative of microcytic anemia attributed to blood loss from his chronic lesions. Chest X-ray was within normal, without hilar lymphadenopathy.

He was started on topical prednisolone acetate as well as oral doxycycline and ascorbic acid for prevention of corneal melt. His vision

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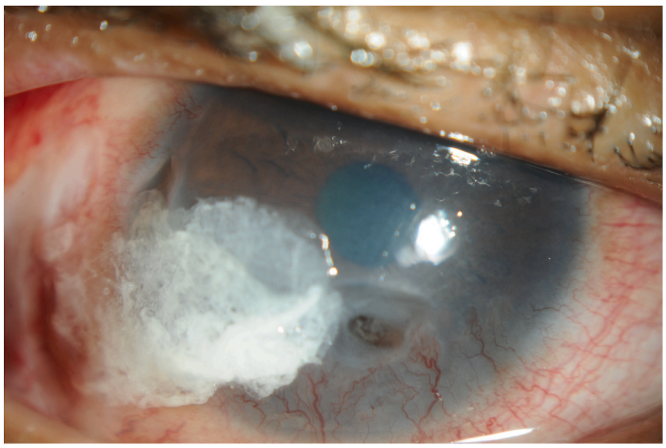

Fig. 1. Appearance of the ocular surface on presentation following temporal conjunctival recession and corneal gluing. Descemetocele is pictured with inferior neovascularization.

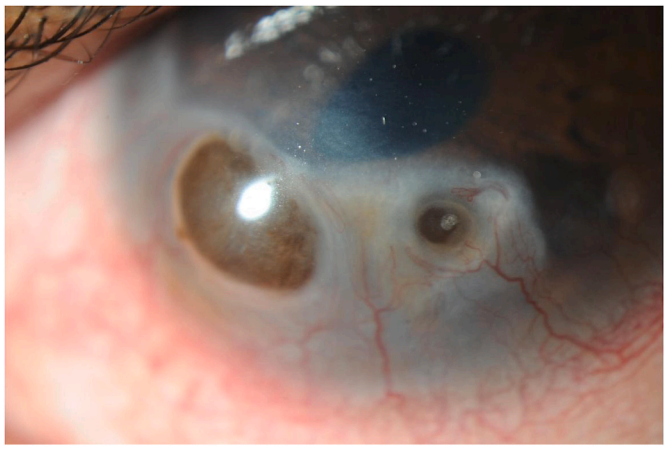

Fig. 2. Appearance of ocular surface at two-month follow-up. Inferior perforation at two sites with iris plugging are pictured, both seidel negative, with flattened anterior chamber and peaked pupil.

improved to 20/25 over 11 days without progressive thinning of the inferior cornea. During this admission, he also underwent management of underlying HS on the recommendation of the internal medicine, dermatology, general surgery, medicine teams including counseling on smoking cessation and weight loss, initiation of metformin, recommendation of TNF-a inhibitor therapy to begin as an outpatient, and surgical excision of HS tracts and lesions of scalp, postauricular areas, axillae, perineum, scrotum, and anal margins.

Following discharge, the patient was lost to follow-up before he could be transitioned to TNF-a inhibitor therapy. He re-presented two months later with inferior corneal perforation with iris plugging after discontinuing all topical and oral therapy (Fig. 2). He was offered emergent surgical repair but declined and elected to be managed conservatively. The site of perforation was glued, and an overlying bandage contact lens was again placed. The patient was started on topical moxifloxacin with oral doxycycline and ascorbic acid. He has been subsequently followed with stable iris plugging and now reformed anterior chamber, however he continues to decline surgical repair.

3. Discussion

Hidradenitis suppurativa is a chronic follicular inflammatory skin disorder characterized by recurrent nodules, sinus tracts, abscesses, and scarring within primarily intertriginous regions.[1–3] Its pathogenesis is believed to be initial occlusion of the folliculopilosebaceous units of apocrine gland-bearing skin that subsequently rupture, prompting both an innate and adaptive immune response to the release of antigens and pro-inflammatory stimuli.[1–3] Coexistence of inflammatory eye disease and HS is rare. In prior studies, the most common manifestation was anterior uveitis, followed by episcleritis/scleritis.[2,3] Forms of keratitis that have been reported include interstitial keratitis, Mooren-type ulceration, and peripheral ulcerative keratitis.[4–7] Previous studies have also suggested topical corticosteroids as first-line treatment, however there is particular interest in *anti*-TNF agents given higher rate of remission for ocular inflammatory disease. Further, there is the potential for a single *anti*-TNF agent to treat ocular inflammation and HS concurrently.

This case is unique in that our patient presented with peripheral ulcerative keratitis, a particularly rare ophthalmic manifestation of HS without concurrent autoimmune or rheumatologic disease making HS the likely cause. Similar to prior reports, our patient initially improved with topical steroid therapy. Unfortunately, he was lost to follow-up prior to transition to *anti*-TNF therapy and suffered corneal perforation, a feared complication of ulcerative keratitis. Ophthalmologists should be aware of HS and its variety of ocular manifestations. Further, coordination between ophthalmology, dermatology, and/or rheumatology is necessary to ensure prompt diagnosis and treatment of not only manifested inflammatory eye disease but also underlying HS.

Patient consent

This patient consented to publication of the case in writing. This report does not contain any personal information that could lead to identification of the patient.

CRedIT author statement

Liane O. Dallalzadeh – Conceptualization, Data Curation, Writing – Original Draft, Project Administration; Michael J. Ang – Data Curation; Alex P. Beazer – Data Curation; Doran B. Spencer – Writing – Review & Editing, Supervision; Natalie A. Afshari - Writing – Review & Editing, Supervision.

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Authorship

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

Declaration of competing interest

None.

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