Contact Lens & Anterior Eye 36 (2013) 313–317

Contents lists available at ScienceDirect

Contact Lens & Anterior Eye

journal homepage: www.elsevier.com/locate/clae

Case report

Peripheral ulcerative keratitis in association with sarcoidosis

Jennifer S. Harthan*, Renée E. Reeder

Illinois College of Optometry, United States

ARTICLE INFO

Article history:
Received 30 April 2012
Received in revised form 22 July 2013
Accepted 25 July 2013

Keywords:
Peripheral ulcerative keratitis (PUK)
Collagen vascular diseases
Sarcoidosis

ABSTRACT

Peripheral ulcerative keratitis (PUK) is a sight-threatening condition characterized by an epithelial defect, crescent-shaped stromal inflammation, and progressive stromal thinning. Peripheral ulcerative keratitis as a purely inflammatory entity is most commonly associated with collagen vascular diseases, including rheumatoid arthritis, polyarteritis nodosa, Wegener granulomatosis, systemic lupus erythematosus, and relapsing polychondritis. PUK can also be associated with infectious and inflammatory conditions such as hepatitis, syphilis, herpes simplex keratitis, fungal keratitis, Mooren ulcer, and marginal keratitis. We describe a case report of PUK associated with the inflammatory condition of sarcoidosis.

1. Background

Peripheral ulcerative keratitis (PUK) is a sight-threatening condition characterized by a corneal epithelial defect, crescent-shaped stromal inflammation, progressive stromal thinning, and often with adjacent conjunctival, episcleral, or scleral inflammation [1,2] PUK presents bilaterally in greater than one third of patients.

Peripheral ulcerative keratitis has long been associated with autoimmune and collagen vascular diseases, most commonly with the systemic disease of rheumatoid arthritis (RA) [1–5]. PUK commonly manifests in RA patients later in the disease process rather than at the onset of the disease, suggesting that the disease is worsening. It is hypothesized that the presentation of PUK may signify the transformation of RA into the systemic vasculitic phase [2–4]. The presentation of PUK in a patient with RA suggests a life threatening stage of the disease and should be treated as an emergent situation with immunosuppressants and cytotoxic therapy [2]. Rheumatoid factor and autoantibodies to IgG are often found in the serum. Very high titers of IgM-rheumatoid factor are also typically present in patients with RA-associated vasculitis [1]. Although rheumatoid factor is used in the diagnosis of RA, it is not specific for the disease as it may also be present in patients with scleroderma, polyarteritis nodosa, Wegener's granulomatosis, systemic lupus erythematosus, sarcoidosis and certain infections [5,6].

In addition to RA, Wegener's granulomatosis (WG) is a leading autoimmune condition associated with PUK. Wegener's granulomatosis is a rare disease, of unknown etiology, that is characterized by vasculitis of the upper and lower respiratory tracts, often in combination with glomerulonephritis. Wegener's granulomatosis may affect multiple organs including the skin, eye, heart, nervous system and gastrointestinal tract and may cause a variety of ocular complications such as scleritis, proptosis, PUK, and conjunctivitis [7]. Peripheral ulcerative keratitis experienced in a patient with WG is a non-specific disease causing conjunctival and scleral inflammation which eventually leads to corneal thinning if systemic therapy is not initiated. In contrast to RA, PUK often manifests at the onset of WG, leading to the diagnosis of the systemic condition. Wegener's granulomatosis is highly sensitive and specific for cytoplasmic antineutrophil antibodies (c-ANCA) when systemic disease is present. When the disease is limited, the sensitivity drops and fluctuation in the c-ANCA titer may correlate with the disease state [4].

Conditions like RA and WG may also be associated with vasculitis, a condition characterized by inflammation and damage of vascular endothelium, which causes necrosis of the blood vessel wall. As a result, there may be vessel destruction, occlusion, or ischemia to the affected organs. Vasculitis is associated with many systemic conditions and can affect various organs including the eye [8]. Rheumatoid arthritis, systemic lupus erythematosus, relapsing polychondritis, Wegener's granulomatosis, polyarteritis nodosa, and Churg-Strauss syndrome are the most common systemic diseases associated with small-vessel vasculitis that causes corneoscleral disease [2,4,7–9]. Peripheral ulcerative keratitis may be an early manifestation of the systemic development of vasculitis [2,4,7–9].

The presentation of corneal ulceration along with a known or unknown systemic disease requires immediate work-up as it may represent a potentially sight and life threatening condition. When a patient is suspected of having a peripheral ulcerative keratitis, immediate treatment and referral to rheumatology is warranted due the serious nature of the condition, as it may lead to stromal

* Corresponding author at: Illinois College of Optometry/Illinois Eye Institute, 3241 S Michigan Avenue, Chicago, IL 60616, United States. Tel.: +1 312 949 7137; fax: +1 312 949 7655.
E-mail address: jharthan@ico.edu (J.S. Harthan).

1367-0484/$ – see front matter
http://dx.doi.org/10.1016/j.clae.2013.07.013

necrosis, corneal perforation, and blindness [2,3,9–13]. In these severe cases, a penetrating keratoplasty may be needed. For RA patients, current treatment involves systemic corticosteroids plus a cytotoxic agent (any agent or process that kills cells, such as chemotherapy and radiation) in the acute phase of the disease. In cases where no underlying systemic disease is identified but is suspected, only topical treatment may be called for and could include corticosteroids, antibiotics and aggressive lubrication.

2. Case report

A 65-year-old African American female, referred for conjunctivitis, presented in August 2008 complaining of epiphora, foreign body sensation, pain (8 on a scale up to 10), and photophobia of her left eye for two weeks. She had a history of previous eye infections OU, two failed penetrating keratoplasties secondary to infection and graft failure OD, and band keratopathy OU. Review of systems revealed a history of congenital syphilis, sarcoidosis, hypertension, diabetes mellitus (Type II), sleep apnea, and an allergy to iodine. Her only reported systemic medication was metoprolol for hypertension.

Presenting unaided visual acuities were light perception OD and 20/100 OS, pinhole no improvement. Slit lamp examination revealed corneal scarring, endothelial pigment and neovascularization of the right eye. The left eye had 2+ diffuse conjunctival hyperemia, 1+ anterior chamber reaction and an inferior epithelial defect with thinning (Fig. 1). All other ocular findings were unremarkable. Intraocular pressures measured 15 mmHg OU by Goldmann applanation tonometry. The Visante anterior segment OCT showed corneal thinning OS on high resolution cornea scan and global pachymetry readings (Fig. 2). Her condition was diagnosed as sclerosing keratitis of unknown etiology and was managed with 1% prednisolone acetate q.i.d., ciprofloxacin ointment t.i.d., and non-preserved gel tears every hour while awake. She was referred for immediate blood work to test for inflammatory and infectious etiologies.

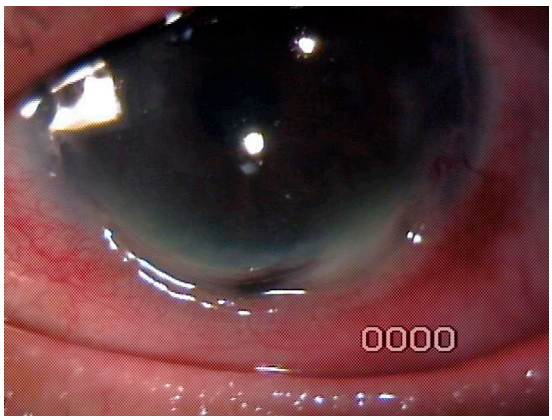

Fig. 1. Diffuse conjunctival hyperemia and inferior corneal thinning OS, August 2008.

While blood work was requested immediately, it took several weeks for the results to be received. During these first few weeks, her ocular condition and visual status were monitored closely. Ultimately, testing revealed no etiologic conditions, either infectious or inflammatory such as RA, WG, or SLE. Thus sarcoidosis was considered as the possible etiology of her ocular presentation. She was maintained on the topical treatment regimen and her symptoms and vision improved significantly over the course of the next four months.

Upon follow-up in December 2008, the patient only complained of mild photophobia as all other symptoms had resolved. She was compliant with her medication regimen of 1% prednisolone

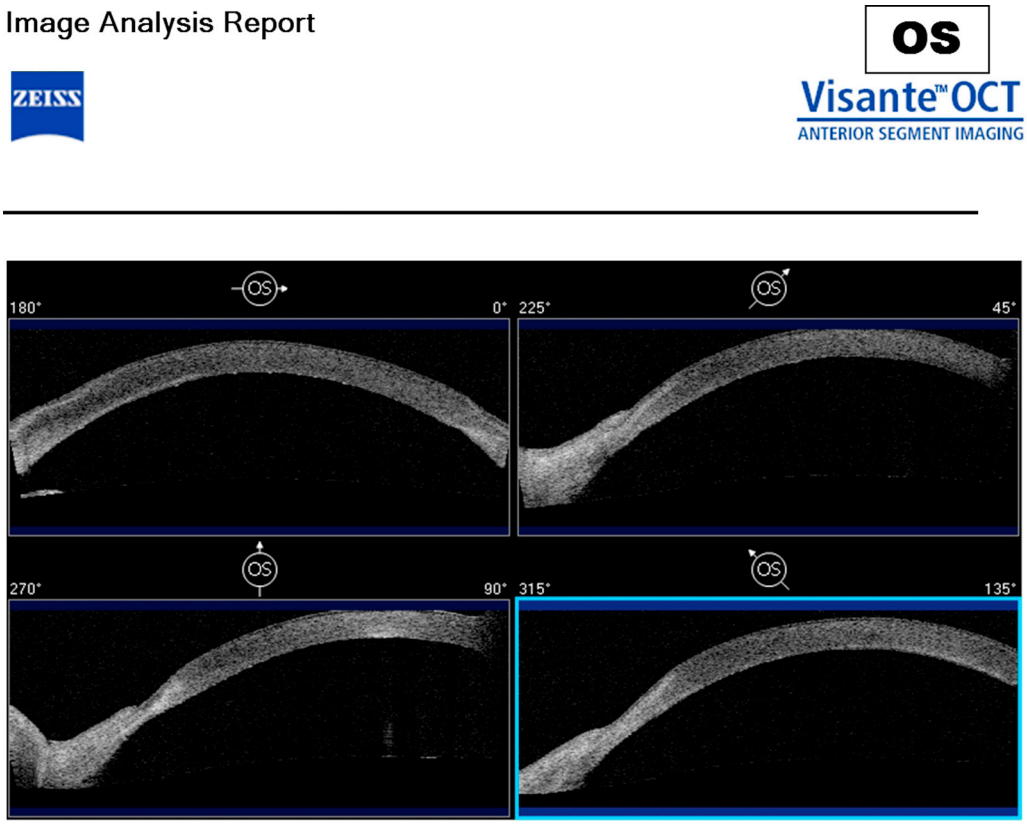

Fig. 2. Visante anterior segment OCT OS, demonstrating inferior corneal thinning, August 2008.