Bilateral Disciform Keratitis with Immune Corneal Ring in a Patient with HLA-B27 Positive Reactive Arthritis

Niveditha Narayanan, Pavani Murthy
Medical Research Foundation, Sankara Nethralaya, 18 College Road, Chennai, Tamil Nadu, India

Abstract

Purpose: *To report a presentation of bilateral disciform keratitis with immune corneal ring in a young male with HLA B 27 positive reactive arthritis.*

Case Report: *A 21 year old man suffered an attack of bacterial UTI a month back with concurrent self-limited bilateral scleritis for 3 days. A week later, he developed multiple joint aches and a rheumatologist diagnosed HLAB27 associated reactive arthritis for the first time, though he had occasional mild joint aches for 8 years and was on irregular aurvedic medication. Specific treatment led to remission of arthritis. About 2 weeks later, he had second episode of bilateral red eye with blurry vision along with self-limited genital ulcer. With no improvement using topical and systemic antivirals for 3 days, he reported to us with bilateral disciform keratitis with incomplete immune rings. Preliminary smear reports of corneal scraping and conjunctival swab were negative and the patient was started on topical antibiotics and later tapering dose of topical steroids were added that led to complete resolution in 45 days.*

Conclusion: *This is the first report of immune corneal ring in bilateral disciform keratitis in a patient with HLAB27 positive reactive arthritis to the best of our knowledge.*

Delhi J Ophthalmol 2019;29;87-89; Doi http://dx.doi.org/10.7869/djo.453

Keywords: *Disciform keratitis, Immune corneal rings, Reactive arthritis, Scleritis*

Introduction

The classical triad of reactive arthritis include urethritis, arthritis and conjunctivitis. After initial mucosal infection, arthritis usually occurs around a month later and ocular involvement coincides with arthritic flare ups. Common ocular associations are self-limited papillary conjunctivitis, non-granulomatous uveitis and rarely punctate keratitis. We present a case of HLAB27 positive reactive arthritis with bilateral disciform keratitis surrounded by incomplete immune corneal rings.

Case Report

A 21 year old man had sudden onset of painful UTI a month back, the urine culture confirmed E.coli infection and he recovered with systemic antibiotics. Concurrently he had bilateral red eye suspected as scleritis that resolved spontaneously in 3 days. A week later, the patient had pain in his left hip, both knees and few metacarpal joints. Rheumatological investigations including blood tests, MRI pelvis and left femur biopsy confirmed HLA B27 related spondyloarthritis for the first time though the patient gave history of occasional mild joint aches for the past 8 years and was on irregular aurvedic medication. Remission was achieved with appropriate treatment and maintenance dose was continued as advised by rheumatologist. Two weeks later he developed bilateral red eye with glare and was treated by local ophthalmologist with topical and systemic antivirals for 3 days but no improvement was noted and he was referred to higher centre. Around that time, he developed genital ulcer that resolved spontaneously.

On reporting to us, BCVA was 20/30 and 20/20 in the right and the left eye respectively. Slitlamp examination showed mild lid edema with diffuse conjunctival congestion both eyes, the right eye had two corneal lesions, a large temporal lesion with 5/3.5 mm^2 and a second lesion inferonasally with 2.5/1.5 mm^2 of epithelial defects, both with underlying infiltrates surrounded by stromal edema and thin incomplete immune rings. The left eye showed a paracentral lesion 2.5/1.5 mm^2 of epithelial defect over a mild infiltrate and a larger surrounding stromal edema ending in multiple tiny infiltrates and faint incomplete immune ring (Figure 1). Both eyes had mild AC flare, no cells, clear lens, normal finger tension and dilated fundus.

Routine corneal scraping and conjunctival swab including for chlamydia showed negative smear reports. Topical hourly instillation of both moxifloxacin 1% and fortified cefatoxime 50mg/ml were started and after 2 days, the right eye (Figure 2) showed AC exudate while the left eye was same. Topical fluoromethalone 0.5% twice daily was added until epithelial healing, then replaced with weekly tapering of topical betnesol 0.1% starting 6 times daily in both eyes, leading to complete resolution with faint scar by 45 days (Figure 3 & 4). Final culture reports were negative for both eyes.

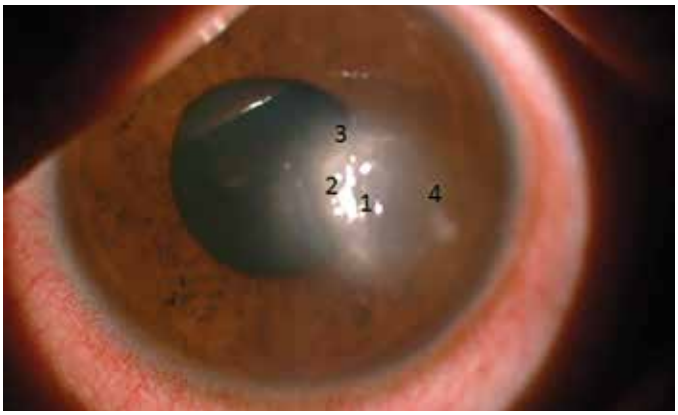

Figure 1: *1-Epithelial defect, 2-infiltrate, 3-stromal edema, 4-tiny infiltrates*

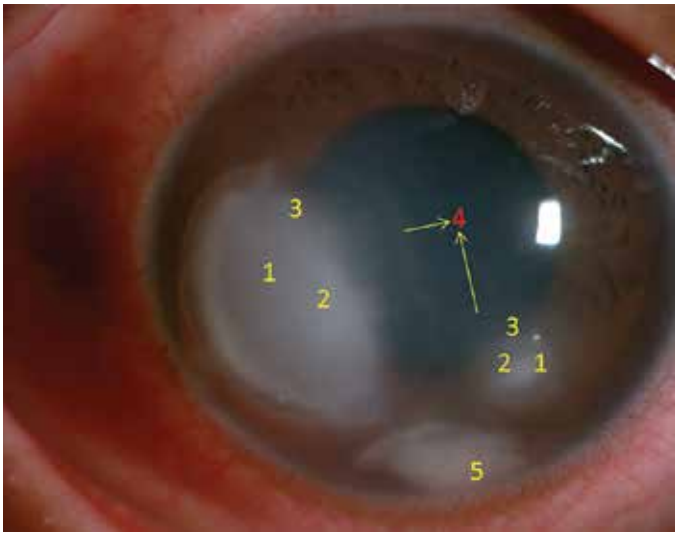

Figure 2: *1-Epithelial defect, 2-infiltrate, 3-stromal edema, 4-immune ring, 5-Anterior chamber exudates*

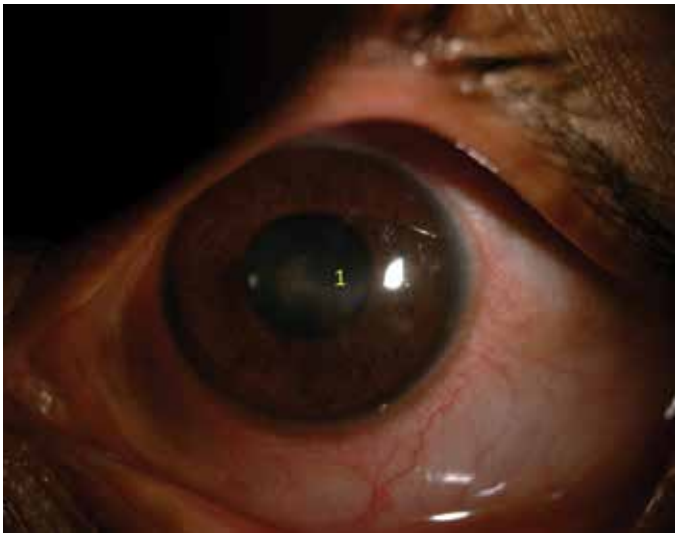

Figure 3: *1-Nebulomacular scaring*

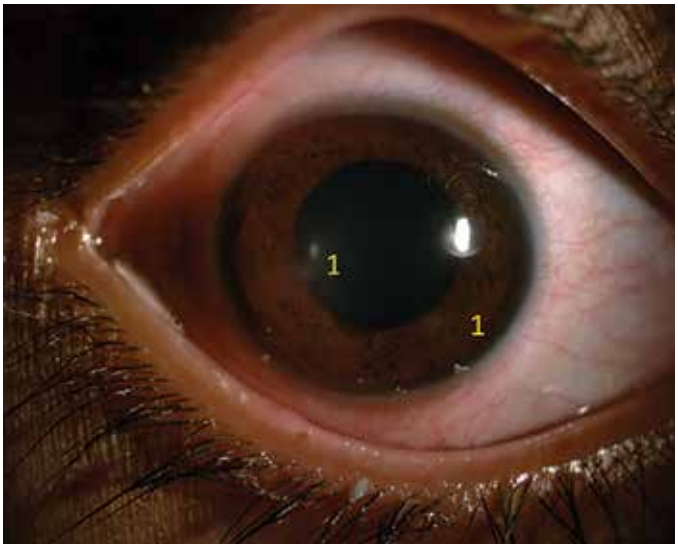

Figure 4: *1-Scaring*

Discussion

Reactive arthritis, so renamed from Reiter's disease by Spondyloarthritic society of America due to Nazi past of Hans Conrad Julius Reiter, is an immune-mediated synovitis triggered by bacterial infection of the mucosal surface and later develops as a triad of autoimmune oligoarthritis, urethritis and conjunctivitis. The prevalence of reactive arthritis is 0.1% in the general population and 5 times more in HLAB27 positives. The postvenereal form is common in developed nations with a male preponderance by 9 times and the postenteric form is common in developing nations with no sex predilection.

The three central aspects in the pathogenesis of reactive arthritis are presence of bacteria or its products in joints, bacterium-host interaction and local immune response. Arthritigenesis has been explained by many theories like the molecular mimicry, misfold, unusual homodimer and heterodimer formation, non-elimination of pathogen infected macrophages by HLAB27, bacterial adhesion molecules attaching to synovial cells using HLAB27 as ligand and cytokines Th1-Th2 imbalance with impaired Th1 antibacterial response and exaggerated Th2 bacterial retaining effect.[1]

The cornea possesses unique anatomical, physiological and immunological adaptations to limit immune mediated inflammation preventing corneal damage maintaining clarity and this is the corneal immune privilege. Anatomically, it is paucity of blood and lymphatic vessels, physiologically it is the molecular mass based gradient of cell distribution between periphery to central cornea.[2]

Immunologically, it is the anterior chamber associated immune deviation (ACAID). In this, an allogen in anterior chamber triggers a systemic immune response that leads to antigen specific suppression of delayed-type hypersensitivity and deviation to non-complement fixing antibody isotope. Sometimes the best defence is a good offence. Like apoptosis of infiltrating neutrophils and lymphocytes by certain ligands expressed in corneal layers, the blocking of efferent arm of cell-mediated immune response by cell membrane-bound molecules, blunting of natural killer cells by factors in aqueous humour eventually preventing apoptosis of MCH class 1 negative cells and inhibition of natural killer cell mediated cytolysis by HLA-G expressed by corneal endothelium are few examples. At times, immunologically isolated sites can also become targets of autoimmune response, like the sympathetic ophthalmia and autoimmune diseases.

Transient conjunctivitis occurs in one third of patients, iritis occur in 5% of patients, while other forms of ocular involvement like keratitis, corneal ulcer, episcleritis, retrobulbar neuritis and hyphema occur only in chronic and persistent reactive arthritis. Keratitis is rare as an initial presentation, but seen in 64% of recurrent arthritis. Disciform keratitis was first reported by Mark and Mculley in 1982, 2 of his cases had typical infiltrates, the third case was suspected chlamydial disease.[3] Suresh in 2016 reported bilateral disciform keratitis in a 13 year old boy, with intact epithelium in one eye and entire corneal involvement with epithelial defect and hypopyon in the other.[4] Khandgave

in 2015 reported bilateral disciform keratitis in a 13 year old girl, nonresponsive to antivirals but improved only by systemic and topical steroids.[5]

Though our patient had occasional mild joint pain for 8 years and was on improper treatment, HLAB27 related spondyloarthritis was diagnosed after multiple joint involvement occurred a week following bacterial UTI. The patient had the first episode of eye involvement as self limiting bilateral scleritis concurrently with UTI and had recurrence of ocular involvement a month later as bilateral disciform keratitis with concurrent genital ulcer and had no recurrence of arthritis. He had two lesions in the right eye and one paracentral lesion with multiple tiny edge infiltrates in the left eye. All three lesions had incomplete immune rings that has not been reported so far to the best of our knowledge. Though the patient was initially started on empirical topical antibiotics, quick resolution was achieved by careful addition of tapering dose of topical steroids.

Conclusion

In cases of unusual bilateral disciform keratitis which can often be mistaken for viral keratitis, lack of clinical response to antivirals, association with arthritis, carefully eliciting history of prior bacterial mucosal infection commonly UTI or GIT can aid the clinician in the diagnosis of reactive arthritis. Prompt initiation of tapering dose of topical steroids can bring faster resolution along with a rheumatologist's intervention for arthritis management which is vital for recovery.

References

1. Colmegna I, Cuchacovich R, Espinoza LR. HLA-B27-associated reactive arthritis: pathogenetic and clinical considerations. *Clinical Microbiology Reviews* 2004; 17:348-369.
2. Niederkorn JY. Cornea: Window to Ocular Immunology. *Curr Immunol Rev* 2011; 7:328-335.
3. Mark DB, McCulley JB. Reiter's keratitis. *Arch Ophthalmol* 1982; 100:781-784.
4. Suresh PS. Bilateral disciform keratitis in Reiter's syndrome. *Indian J Ophthalmol* 2016; 64:685-687.
5. Khandgave TP, Puthran N, Kulkarni VN. Bilateral disciform keratitis: A rare feature of Reiter's syndrome. *J Clin Ophthalmol Res* 2015; 3:102-104.

Cite This Article as: *Narayanan N, Murthy P. Bilateral Disciform Keratitis with Immune Corneal Ring in a Patient with HLA-B27 Positive Reactive Arthritis.*

Acknowledgments: Nil

Conflict of interest: None declared

Source of Funding: None

Date of Submission: 22 March 2019
Date of Acceptance: 06 April 2019

Address for correspondence

Niveditha Narayanan FRCS
Medical Research Foundation
Sankara Nethralaya
18, College Road, Chennai 600 006,
Tamil Nadu, India
Email id: drnvn@snmail.org

Quick Response Code