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Risk of Blindness in Rheumatoid Arthritis Patients: A Case Report

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Abstract

Background: Peripheral ulcerative keratitis (PUK) is a serious inflammatory disorder of the juxtalimbal corneal stroma, characterized by epithelial defects, infiltration of inflammatory cells, and progressive thinning of the stroma, potentially resulting in vision loss. Rheumatoid arthritis (RA) is the most common systemic autoimmune condition linked to non-infectious PUK, responsible for about 34% of cases, with nearly half exhibiting bilateral involvement. **Objective:** This case report presents a patient with bilateral PUK associated with RA and highlights the importance of early diagnosis and timely management to prevent vision loss. **Case presentation:** A 43-year-old female presented with a 12-month history of blurred vision and progressive ocular pain. Physical inspection revealed a skin rash, swan neck deformity, boutonniere deformity, and hallux valgus on both legs. Ocular examination showed a crescent-shaped peripheral corneal ulcer with an epithelized iris prolapse in the right eye, and nasal corneal thinning with peripheral ulceration in the left eye. Irregular pupils and distinct lens morphology were also noted. Serological tests were positive for antinuclear antibodies (ANA) and rheumatoid factor (RF). **Result:** The patient was diagnosed with RA-associated PUK and started on systemic and topical corticosteroids. Treatment led to initial improvement in ocular inflammation and stabilization of corneal structure. **Conclusion:** RA is the most frequently associated autoimmune condition in PUK. Early recognition of systemic autoimmune disease in patients with peripheral corneal ulcers is critical. Timely multidisciplinary management involving internists and ophthalmologists can preserve vision and improve patient outcomes.

Keywords: Autoimmune, Peripheral ulcerative keratitis, Rheumatoid arthritis

Case Report

INTRODUCTION

Peripheral ulcerative keratitis (PUK) is a destructive inflammatory disease of the peripheral cornea, characterized by epithelial defects and stromal thinning (Hassanpour et al., 2022). Peripheral ulcerative keratitis often contiguous with adjacent conjunctival, episcleral, and scleral inflammation. The presence of such adjacent tissue inflammation aggravates the course of PUK and causes potentially serious complications, such as perforation of the cornea (Das et al., 2025). It is most often associated with systemic autoimmune diseases, particularly rheumatoid arthritis (RA), where immune complex deposition and limbal vasculitis play central roles in driving corneal damage (Höllhumer, 2022)

Rheumatoid arthritis is a chronic systemic autoimmune disease characterized by persistent joint inflammation, pain, and swelling, but it may also involve extra-articular organs, including the eyes (Chauhan et al., 2023). Ocular manifestations are clinically significant because they can be severe and vision-threatening, particularly in the form of PUK, which occurs in association with autoimmune-mediated vasculitis (Hassanpour et al., 2022). In PUK, inflammatory cells and cytokines stimulate the release of matrix metalloproteinases that degrade stromal collagen, leading to corneal thinning and, in severe cases, perforation (Höllhumer, 2022). Furthermore, RA-associated vasculitis can compromise ocular perfusion, predisposing patients to ischemic complications such as scleritis or optic neuropathy, which may result in irreversible vision loss (Wang et al., 2022)

Ocular complications of RA are clinically important because they may progress rapidly and severely affect vision (Duchvenic et al., 2024). Epidemiological studies have shown that a significant proportion of non-infectious peripheral ulcerative keratitis (PUK) cases are associated with RA, and bilateral involvement is relatively common (Gupta et al., 2021). Early recognition, timely ophthalmological evaluation, and the initiation of appropriate systemic therapy are crucial to prevent blindness and improve long-term outcomes (Zemba et al., 2020). This case report describes a patient with bilateral PUK associated with RA and highlights the importance of early diagnosis, multidisciplinary collaboration, and aggressive management to prevent irreversible vision loss.

CASE PRESENTATION

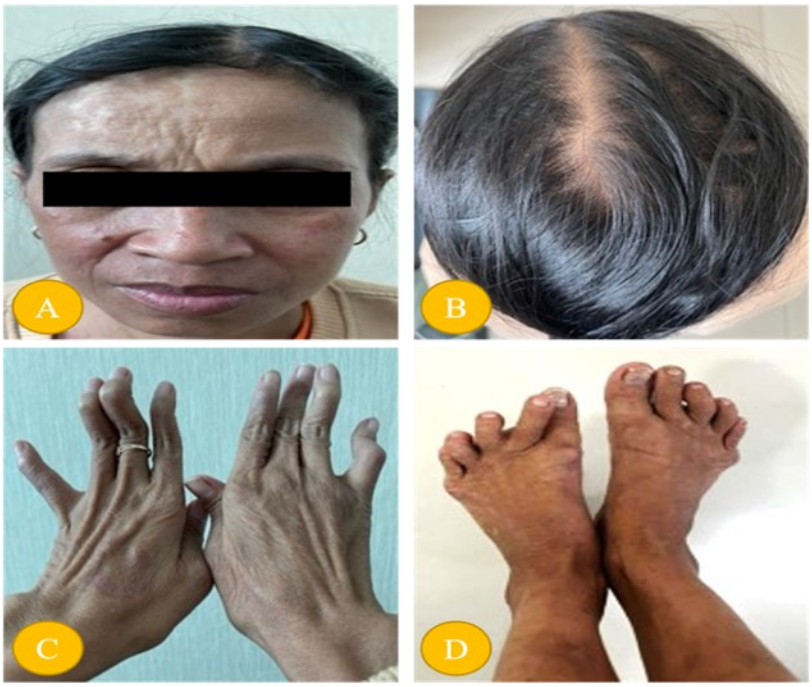

Figure 1. On the face, there was a skin rash and skin depigmentation (A). On the head area, central alopecia was seen; (B) both hands showed swan neck deformity and boutonniere deformity; (C) On both legs, hallux valgus was found on both lateral sides (D).

A 43-year-old female presented to the Infection and Immunology Department of the National Eye Center, Cicendo National Eye Hospital, with complaints of blurred vision and discomfort in both eyes. The patient reported a history of progressive blurry vision for the past year, which had worsened in the weeks prior to presentation. She also described bilateral eye pain, photophobia, tearing, redness, and a persistent foreign body sensation in the right eye. A white corneal spot had been developed in the right eye (RE) one month earlier and had not resolved. Systemic complaints consisted of morning stiffness, joint pain in both hands and feet, hair loss, fatigue, and weight loss over the past year. The patient had a previous history of hospitalization for rheumatoid arthritis. She denied any history of diabetes mellitus, hypertension, or similar ocular disease in her family.

On general examination, the patient's vital signs were within normal limits. Extra-ocular findings revealed a facial rash, central alopecia, and depigmentation of the frontal scalp. Musculoskeletal examination demonstrated deformities consistent with chronic RA, including swan neck and boutonniere deformities of both hands and hallux valgus in both feet. Other systemic examinations were unremarkable.

Ophthalmological examination revealed orthotropic alignment with full range of ocular movements in all directions. Visual acuity was 1/60 in the RE and 0.63 in the left eye (LE), with normal intraocular pressure in both eyes. Slit lamp examination of the RE showed a quiet lid, grade 1 ciliary Efron injection, a peripheral ulcer on the cornea with a cresentic-shaped, stromal thinning extending from one to four o'clock, prolapse of the iris, which was epithelialized at five o'clock, and peripheral corneal ulcer. The Van Herick grade III was allocated to the temporal side of the anterior chamber and grade II nasally. The iris has anterior synechiae, the pupils was irregular, and the lens was clear. The LE examination showed that the conjunctiva and lids were normal, but the cornea had a peripheral ulcer in the nasal region from four to nine o'clock with stromal thinning. An anterior chamber was Van Herick Grade III. The iris appeared clear, the lens was transparent, and the pupils were irregular in shape. Posterior segment evaluation was normal in both eyes.

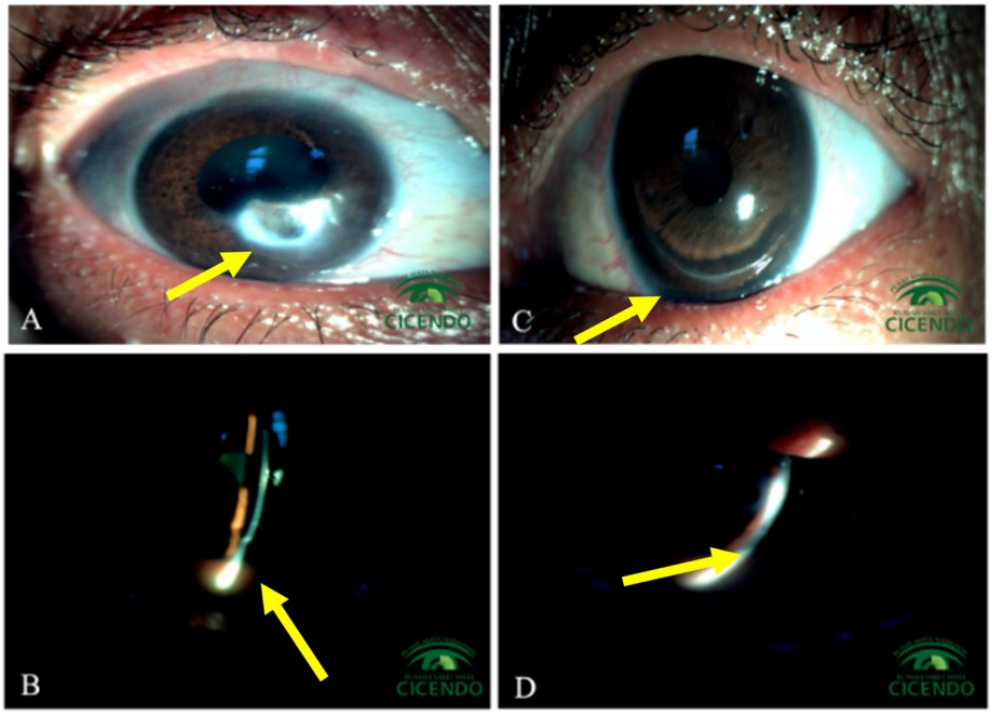

Figure 2. Examination of the anterior segment of the right eye showed prolapse of the epithelialized iris (A) with thinning in the form of a crescent at one o'clock to four o'clock in the peripheral cornea (C). The anterior segment of the left eye shows peripheral corneal thinning at four o'clock to nine o'clock (B-D).

Based on the clinical findings, the patient was diagnosed with bilateral peripheral ulcerative keratitis associated with rheumatoid arthritis. Laboratory investigations were performed, revealing reactive results for both rheumatoid factor (RF) and antinuclear antibody (ANA), supporting an autoimmune etiology. Treatment was initiated with oral methylprednisolone 40 mg daily gradually tapered, oral ranitidine hydrochloride 150 mg daily, prednisolone acetate eye drops six times daily in both eyes, cyclopentolate hydrochloride eye drops three times daily, and artificial tears eye drops six times daily. The patient was referred to the rheumatology division for systemic disease management.

At the one-week follow-up, systemic and topical corticosteroids were tapered further, with continued supportive treatment. Corneal thinning and ocular surface inflammation were stabilized, although the visual prognosis remained poor.

DISCUSSION

The morphology and physiology of the peripheral cornea differ from those of the central cornea. The cornea, conjunctiva, episclera, and sclera come together to form the peripheral cornea (Sridhar, 2018). Histologically, the stroma peripherally has looser collagen bundles, and limbal and lymphatic vascular arcades extend about 0.5 mm into the central cornea (Hassanpour et al., 2022). The peripheral cornea harbors a higher density of Langerhans cells and immune mediators, including immunoglobulins and complement components, which contribute to heightened local and systemic immune responses (Cao et al., 2017).

Peripheral ulcerative keratitis can arise from a variety of ocular and systemic disorders, with autoimmune diseases being the predominant non-infectious cause (Zemba et al., 2020). Collagen vascular diseases, particularly rheumatoid arthritis, are the most frequently reported systemic associations and contribute substantially to the burden of PUK (Wang et al., 2022). Although the condition more often manifests unilaterally, bilateral cases have been described and are strongly linked to underlying systemic autoimmune activity (Kochhar et al., 2022). In this case, however, bilateral PUK was observed, which strongly suggests an association with an underlying systemic autoimmune condition such as rheumatoid arthritis.

Although the exact mechanism of PUK in RA remains unclear, it is thought to be triggered by autoantigen alterations influenced by both genetic and environmental factors (Cao et al., 2017). Autoimmune processes are closely linked to epigenetic modifications and dysregulation of both cellular and humoral immunity (Gupta et al., 2021). In this context, T and B lymphocyte activation promotes the release of complement factors such as C3a and C5a, which increase vascular permeability and recruit neutrophils to the corneal periphery (Hassanpour et al., 2022). Activated neutrophils then release proteases and matrix metalloproteinases, leading to stromal collagen breakdown, progressive thinning, and peripheral corneal destruction (Guney et al., 2021)

Diagnostic evaluation for PUK includes anamnesis, general physical examination, slit-lamp examination, and laboratory tests to identify the underlying etiology and rule out conditions that may mimic PUK (Hassanpour et al., 2022). A complete history is essential for diagnosing PUK as its progression is closely linked to RA (Czarnacka et al., 2023). A swan neck deformity, a boutonniere deformity, and hallux valgus, which are characteristic deformities in RA (Guo et al., 2018), were among the patient's symptoms and signs in this case. Furthermore, the rheumatoid factor test showed reactive results, supporting the diagnosis. A detailed systemic history is also crucial to distinguish RA from other autoimmune conditions with ocular involvement, such as systemic lupus erythematosus, which may present with skin rash, alopecia, and facial depigmentation (Fava & Petri, 2019).

During the initial evaluation of patients with RA clinicians should specifically inquire about ocular symptoms suggestive of PUK, such as redness, ocular discomfort, excessive tearing, photophobia, and reduced visual acuity (Boonsopon et al., 2022). According to the anamnesis, the patient had been undergoing treatment for rheumatoid arthritis and reported progressive visual disturbances over the past year; however, ophthalmologic evaluation was only performed recently. Peripheral ulcerative keratitis is a severe extra-articular manifestation of RA, often unrelated to articular disease activity, and may present with scleritis and dry eye symptoms (Zlatanović et al., 2010). Early detection followed by appropriate management is essential to reserve vision in ocular involvement related to rheumatologic diseases (Al-ghamdi, Abdullah, Almoallim & Cheikh, 2021). Once the diagnosis of PUK has been made, the treatment must be aggressive and aimed at avoiding both the local and the systemic complications, since if untreated, PUK may lead to poor visual outcomes and a mortality of around 30%, usually because of the development of a subsequent rheumatoid vasculitis (Navas et al., 2021).

The purpose of treating PUK patients is to control the inflammatory response, halt disease progression, promote epithelial healing, repair stromal damage, prevent complications, and reduce the risk of recurrence (Fu & Jones, 2024; Hassanpour et al., 2022). Management includes both medical and surgical approaches (Sabhapandit et al., 2022). The initial medical regimen usually involves oral and/or topical corticosteroids to suppress acute inflammation (Fu & Jones, 2024). In severe cases or when corticosteroid monotherapy is insufficient, immunosuppressive agents such as methotrexate (5–

25 mg weekly), cyclophosphamide (2–2.5 mg/kg/day), and cyclosporine (2.5–5 mg/kg/day in divided doses) may be added to prevent corneal perforation and achieve systemic disease control (Hassanpour et al., 2022; Shyamsundar et al., 2011). Careful monitoring is essential to minimize adverse effects and preserve visual function, particularly in patients with underlying autoimmune disease (Maleki et al., 2024).

The recommended treatment protocol in this case was the initial treatment of systemic and topical corticosteroids in gradually reduced doses, which remain the primary approach for acute inflammatory control in PUK (Hassanpour et al., 2022). The patient showed clinical improvement at the one week follow-up, as the corneal ulcers in both eyes did not progress and no further signs of ocular inflammation, such as conjunctival ciliary injection, were observed. TNF-α promotes ocular tissue destruction by inducing the expression of matrix metalloproteinases (MMP-1 and MMP-13) through the activation of NF-kB and MAPK signaling pathways. These pathways facilitate the transcriptional regulation of MMP genes, leading to increased MMP production. This elevated MMP expression results in the degradation of collagen in the extracellular matrix, contributing to the breakdown of stromal collagen and ultimately causing tissue damage in inflammatory conditions (Vincenti & Brinckerhoff, 2002). Inhibiting TNF-α could potentially reduce MMP activity, thereby preventing corneal damage and preserving ocular integrity (Wu et al., 2015). Furthermore, biologics such as infliximab and adalimumab as well as conventional disease-modifying antirheumatic drugs (DMARDs), have demonstrated efficacy in treating PUK related to RA, supporting their role as adjunctive therapy to standard corticosteroid treatment (Maleki et al., 2024).

Following the stabilization achieved with systemic and topical corticosteroid therapy in our patient, further local and surgical options were considered to preserve corneal integrity. Conjunctival resection represents an adjunctive approach, particularly in RA-associated peripheral ulcerative keratitis, as it removes the adjacent limbal conjunctiva and eliminates collagenolytic enzymes and inflammatory mediators that perpetuate stromal melting (Zemba et al., 2020). Amniotic membrane transplantation and corneal glue, with or without keratoplasty, are also potential therapeutic modalities to restore tectonic stability, although they generally do not improve visual function and may only serve as temporizing measures (Hick et al., 2005). In more advanced cases with progressive thinning or impending perforation, tectonic surgical interventions such as C-shaped lamellar corneal grafts using donor tissue are recommended to reinforce the corneal structure, limit ectasia, reduce astigmatism, and minimize the risk of graft failure (Clarke et al., 2023; Hassanpour et al., 2022). In this case, medical therapy was prioritized as the first line of protection while awaiting a comprehensive systemic management plan from the rheumatology division.

In this patient with RA associated PUK, the presence of corneal perforation and progressive stromal thinning has resulted in severe visual impairment, leading to a prognosis of *quo ad functionam dubia ad malam* in both eyes. Given that RA is a chronic, progressive autoimmune disease with ongoing systemic activity, the prognosis *quo ad sanationam* is considered *dubia*, as complete recovery cannot be expected. Moreover, because uncontrolled systemic inflammation in RA carries the risk of life-threatening complications, the overall prognosis *quo ad vitam* is also *dubia*.

CONCLUSION

Rheumatoid arthritis is the most common autoimmune disease associated with peripheral ulcerative keratitis. Ocular complications can be vision-threatening and therefore require early recognition and timely intervention. Optimal management relies on close collaboration between ophthalmology and internal medicine to ensure comprehensive and effective patient care.

CONFLICT OF INTEREST

All authors have no conflicts of interest to disclose

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