Peripheral Ulcerative Keratitis in Relapsing Polychondritis: An Ophthalmic Challenge

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Abstract

Peripheral ulcerative keratitis (PUK) is an inflammatory condition leading to ocular morbidity. It can be associated with underlying autoimmune diseases which often goes undetected if the ocular manifestation precedes the systemic manifestation. The objective of this case report is to highlight that, the management of such entity is challenging. We present a case of a 36-year-old female with diminution of vision, PUK with necrotizing scleritis in both the eyes for which she was on regular treatment. Along with medications, she underwent multilayered amniotic membrane transplantation and scleral graft. Later on she developed tracheal stenosis and polychondritis for which she underwent tracheostomy. The ocular presentation of relapsing polychondritis can precede the systemic manifestations which may lead to delay in diagnosis and management of the disease. Unless the underlying disease is addressed, ocular morbidity progresses regardless the ocular treatment.

Keywords: Necrotizing scleritis; Peripheral ulcerative keratitis; Relapsing polychondritis.

Declarations

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Peripheral ulcerative keratitis (PUK) is a vision threatening condition arising in the juxta-limbal cornea characterized by its crescent shape epithelial defect and stromal melting. It is not associated with pain which delays the patient’s hospital visit. The most distinguished factor is its association with inflammation of the adjacent conjunctiva, episclera, and the sclera which makes the disease more aggressive. Most of the cases are associated with underlying systemic infectious or non-infectious diseases out of which rheumatoid arthritis tops the list followed by collagen vascular disease [1].

The most accepted explanation of PUK is both the cell-mediated and humoral mediated autoimmune processes in which the reactions maybe against the corneal antigens, exogenous antigens or circulating immune complex [2, 3]. When associated with underlying systemic disease, it becomes more challenging to an ophthalmologist to control PUK with necrotizing scleritis and aggressive treatment is mandatory. Unless the underlying disease is controlled, it’s quite impossible to control the ongoing ocular inflammation. Here, we present a case of PUK in a 36-year-old female associated with relapsing polychondritis (RP) challenging the management of the patient.

**CASE**

A 36-year-old female presented with the complaint of redness and pain in both eyes for last seven months associated with diminution of vision for last two months. This was associated with foreign body sensation, photophobia and watering (Fig. 1). There was no history of trauma, contact lens use and use of topical eye drops or systemic drugs. There was no history suggestive of systemic illness or autoimmune diseases. She complained of severe headache on and off associated with mild shortness of breath. She gave a history of right eye (RE) pterygium surgery 3 years back. For these complaints, she visited different eye hospitals but all in vain. She was on topical eye drops of steroids four times daily, 1% atropine thrice daily and antibiotics four times daily in both the eyes.

At presentation, the general examination showed saddle nose deformity with mild dyspnea. Her vitals were stable. The ocular examination showed the best corrected visual acuity of 6/9 in the RE and 6/12 in the left. There was diffuse swelling of both the upper and lower eyelids of both eyes Oculus Uterque (OU) associated with meibomianitis and blepharospasm.

Conjunctiva showed diffused ciliary congestion with underlying sclera necrosis in the superotemporal quadrant in the RE and superonasally in the Left eye (LE) (Fig. 2a). Examination of the RE showed a peripheral ulcer involving juxtalimbal area from 7 to 1 o’clock position with 2 mm width. It had a sloping outer margin and steep inner margin with thinning.

![Figure 1](image1.jpg)  
**Figure 1:** Photograph showing bilateral upper and lower eyelid swelling with mechanical ptosis.

![Figure 2a](image2a.jpg)  
**2a.** Necrotizing scleritis in the left eye.

![Figure 2b](image2b.jpg)  
**2b.** Ulcer of left eye involving juxtalimbal area extending from 7 to 1 o’clock position with sloping outer margin and steep inner margin with 70% thinning.
of 70% (Fig. 2b). Left eye (LE) examination showed an ulcer extending from 6 to 2 o’clock position of 2 mm width with 90% thinning (Fig. 3). There was presence of stromal oedema surrounding the ulcer with diffuse superficial punctate keratopathy OU. Anterior chambers were quiet and normal depth OU. The pupil of the RE was about 4 mm with normal reaction and in the LE slightly eccentric superonasally and was pharmacologically mid dilated and fixed non-reacting to light. The rest of the posterior segment examinations were within the normal limit (OU).

Intra-ocular pressure (under eye drop timolol 0.5% bid) was 10.6 and 12.4 mm of Hg in the RE and the LE respectively. The laboratory investigations showed: hemoglobin: 9.6 gm/ dl, PCV/ hematocrit: 29.6%, total leucocyte count 16,400 cell/ mm³, differential count: N63 L37, platelets: 267,000 cells/ mm³, erythrocyte sedimentation rate: 62 mm in 1st hour, random blood sugar: 64 mg/dl, RA Factor: 128 IU/ ml, peripheral smear for cytology (PBS): normochromic to hypochromic red blood cells with mild anisocytosis was present. Microscopy examination of urine showed trace albumin, white blood cells: 3-5/ high power field (HPF) and epithelial cells: 1-2/ HPF. Stool test was normal.

Due to financial problem, the patient denied further investigations like antinuclear antibodies (ANA) and antineutrophil cytoplasmic antibodies (ANCA). The patient was treated for anemia with oral iron supplements. With the presumed diagnosis of bilateral peripheral ulcerative keratitis of rheumatic etiology, the patient was treated bilaterally with topical eye drops of 1% fluorometholone six hourly, tropicamide 1% thrice daily, timolol maleate 0.5% twice daily and artificial tear substitutes one hourly. Oral Vitamin C one gram twice daily, doxycycline 100 mg twice daily for 7 days, prednisolone 60 mg once daily (tapering dose) and ranitidine 150 mg twice daily were also prescribed. Topical 20% autologous serum eye drops were instilled in OU four times daily. Since the condition was static after a week, the patient underwent 360° conjunctival peritomy with multi-layered fresh amniotic membrane graft in the LE (Fig. 4a). The necrotizing scleritis of the LE was treated with scleral patch graft on second setting (Fig. 4b).

With this treatment, there was a dramatic improvement in ocular signs and symptoms. At one month follow up, there was decrease in corneal thinning with macular corneal opacity. However, the status of necrotizing scleritis was static in the RE with scleral graft in situ in the left.

Again, after a month, the patient presented to the emergency with severe shortness of breath with tracheostomy tube in situ performed in another hospital. Computed tomography scan of the chest, done outside, showed tracheal stenosis and polychondritis suggestive of relapsing polychondritis. All the ocular
findings were static (OU). She expired three days after her admission in the Intensive Care Unit due to cardiac failure.

**DISCUSSION**

PUK might be the initial manifestations of underlying autoimmune diseases. Necrotizing scleritis adjacent to PUK is commonly found in these patients [1]. RP is a severe progressive inflammatory condition of the cartilaginous structures (nose, ears, laryngotraceobronchial tree) and may affect eyes, cardiovascular system, joints, skins and central nervous system. Ocular manifestations of RP usually consist of decreased visual acuity, conjunctivitis, episcleritis, scleritis, ocular inflammation, diplopia, and eye lid swelling [4]. Scleritis occurs in about 47% of patients with RP [5]. In our case, the patient didn’t have such noticeable systemic features other than saddle-shaped nose, shortness of breath (on and off) and headache at presentation which caused the delay in systemic diagnosis.

After initiation of oral corticosteroids, there was improvement of the redness, pain and photophobia along with healing of the keratitis. Oral corticosteroid is the mainstay therapy of RP. As with other systemic diseases leading to PUK, cyclophosphamide and methotrexate gives a good outcome and fast recovery in RP if the patient is unresponsive to steroids. Topical corticosteroids are avoided in cases with associated systemic disease as it decreases the collagen synthesis [2, 3].

A case of orbital inflammatory disease in RP has been reported in a 73-year-old patient who showed improvement with systemic steroids. Similar to our patient, there was a delay in diagnosis of the disease [6]. A study showed that patients with rheumatoid arthritis (RA) associated PUK with/out necrotizing scleritis had decreased mortality and ocular morbidity when treated with immunosuppressive medication. The patients treated with cyclophosphamide or antimetabolite agents had lower mortality compared to those treated with glucocorticosteroids and non-steroidal anti-inflammatory drugs alone [7]. Since the patient was from low socioeconomic background, frequent visits to multiple hospitals may have led to poor compliance and delay in follow ups.

**CONCLUSION**

PUK and necrotizing scleritis can be an initial manifestation of RP. A thorough systemic evaluation in co-ordination with a Rheumatologist is essential for all patients presenting with PUK associated with scleritis to prevent potential morbidity.

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**References**