Case Report

ANNALS OF MEDICINE & SURGERY

OPEN

# Coexistence of severe peripheral ulcerative keratitis and lichen planus: case report

Hamza Lazaar, MD*, Taha Boutaj, MD, Meryem Sefrioui, MD, Najoua ElMoubarik, MD, Boutayna Azarkan, MD, Saad Benchekroun, PhD, Abdellah Amazouzi, PhD, Noureddine Boutimzine, PhD, Lalla Ouafa Cherkaoui, PhD

**Introduction and importance:** Ocular involvement in lichen planus is highly uncommon, primarily affecting the eyelids, conjunctiva, and less frequently, the cornea. Peripheral ulcerative keratitis (PUK), a rare subtype form of corneal lichen planus, has been reported only once in the literature.

**Case presentation:** The authors report details of a 34-year-old man with confirmed cutaneous lichen planus who developed severe PUK, a rare ocular manifestation of lichen planus. Despite initial worsening with corticosteroids, successful resolution of PUK was achieved with topical Cyclosporin and azathioprine over 2 months but with a final visual acuity limited to light perception.

**Clinical discussion:** To the best of our knowledge, very rare cases have been reported of the coexistence of severe PUK and lichen planus. Lichen planus should be considered in any case of PUKs associated with cutaneous-mucosal manifestations cyclosporin and azathioprine are crucial for effective management and favorable outcomes in such cases.

**Conclusion:** This case aims to show the importance of dermatological examination in the presence of any peripheral ulcerative keratitis. It also sheds light on the therapeutic difficulties and the prognosis of this rare form of ocular lichen planus.

**Keywords:** corticosteroids, cyclosporin, ocular lichen planus, peripheral ulcerative keratitis

## Introduction

Lichen planus (LP) is an autoimmune, idiopathic, chronic, and inflammatory disorder typically involving the skin, mucous membranes, and nails[1]. The exact prevalence of Lichen Planus is unknown. Nevertheless, the estimated prevalence of LP is in the range of 0.22–0.8%[2]. Its pathogenesis is still unknown, but it appears to represent a T-cell-mediated autoimmune disease.

Ocular lichen planus is an extremely rare presentation, affecting the eyelids, conjunctiva, and, exceptionally the cornea[3]. This manifestation can compromise the functional prognosis of the eye.

In this paper, we present an unusual case with confirmed cutaneous lichen planus histology, without any other systemic diseases, presenting with peripheral ulcerative keratitis (PUK).

Our case report adheres to the SCARE Guidelines 2023[4].

### HIGHLIGHTS

- Ocular lichen planus is a very rare but vision-threatening condition.
- The peripheral ulcerative keratitis with lichen planus is exceptional, and only one case has been described in the literature.
- Azathioprine and topical cyclosporine should be in the first line for the treatment of severe ocular lichen planus.
- Suspect lichen planus in the context of any peripheral ulcerative keratitis (PUK) with cutaneomucous manifestations."

## Case presentation

We report the case of a 34-year-old patient, with no particular medical or family history or recent ocular surgery, presenting to the ophthalmic emergency department of Ibn Sina University Hospital in Rabat with a painful red eye and decreased visual acuity in the right eye over the past 2 weeks.

The best-corrected visual acuity was 20/20 in the left eye and 20/400 in the right eye. Intraocular pressure was within the normal range in both eyes. No abnormalities of the eyelids or ocular adnexa were observed. Slit-lamp examination revealed ocular surface dryness with an 8-sec break-up time in both eyes. Conjunctival hyperemia, superficial punctate keratitis, and a deep peripheral ulcer involving the external 1/3 of the nasal peripheral region, reaching the posterior stroma, extending from 1 to 5 o'clock. No conjunctival scarring or stromal infiltrate was present. The anterior chamber was of good depth without Tyndall, and a nuclear grade II cataract (Fig. 1A). The rest of the examination was unremarkable.

*Ophthalmology A, hôpital des Spécialités, Faculty of Medecine and Pharmacy of Rabat University Mohammed V, CHU Ibn Sina RABAY, Rabat, Morocco*

*Sponsorships or competing interests that may be relevant to content are disclosed at the end of this article.*

**Corresponding author. Address: Ophthalmology A, hôpital des Spécialités,Faculty of Medecine and Pharmacy of Rabat University Mohammed V, CHU Ibn Sina, Souissi, BP 6220, Rabat, Morocco. Résidence safaa II immeuble 24 appartement 12 Opera Mohamed V Salé, Morocco. Tel.: +212 623 204 877. E-mail: lzhmz22@gmail.com*
*(H. Lazaar).*



*Annals of Medicine & Surgery (2024) 86:4803–4806*

*Received 6 March 2024; Accepted 8 May 2024*

*Published online 17 May 2024*

*http://dx.doi.org/10.1097/MS9.0000000000002179*

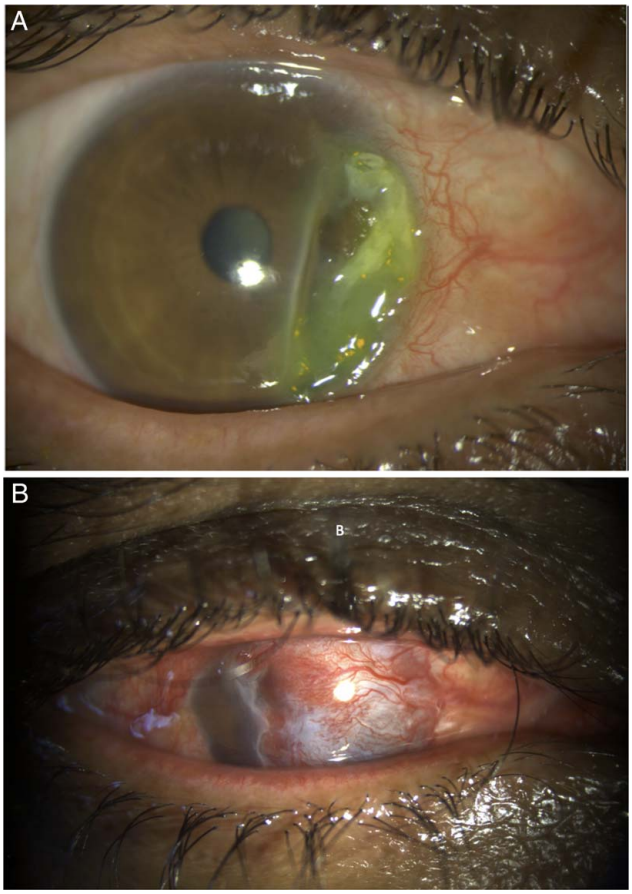


**Figure 1.** A, photograph of the Anterior Segment showing a peripheral Ulcerative keratitis of the 1/3 nasal of the cornea. B, photograph of Anterior segment Showing a worsening of the ulcer extending to 3/4 of the cornea with a severe corneal neovascularization.

A general examination revealed pruritic, violaceous plaques, flat and polygonal, on the forearms, legs, and the patient's back, evolving for 20 days before the ocular symptoms (Fig. 2).

Histological examination of skin biopsy reveals a basal layer damage marked by an inflammatory infiltrate of the chorion, arranged in a sub-epithelial band. In the basal layer, the lowest cells are vacuolated, and the epithelial ridges are tapered consistent with a Lichen Planus (Fig. 3).

A paraclinical assessment in collaboration with the internal medicine department was conducted to exclude other diagnoses associated with lichen planus: serologies for B and C, HIV, and syphilis were negative; negative P-ANCA and C-ANCA; normal anti-CCP and antinuclear antibodies; normal IDR; and normal chest CT scan.

We diagnosed severe peripheral ulcerative keratitis associated with lichen planus.

The patient received a corticosteroid bolus of 1 g/day for 3 days, followed by oral prednisone at a dose of 1 mg/kg, topical 0.1% preservative-free dexamethasone four times daily, oral doxycycline 200 mg per day, and ocular lubricants. Two weeks later, there was a dramatic evolution with the destruction of 3/4 of the cornea (Fig. 1, image B). We stopped topical corticosteroids and added topical Cyclosporin at a dose of 4 drops per day and

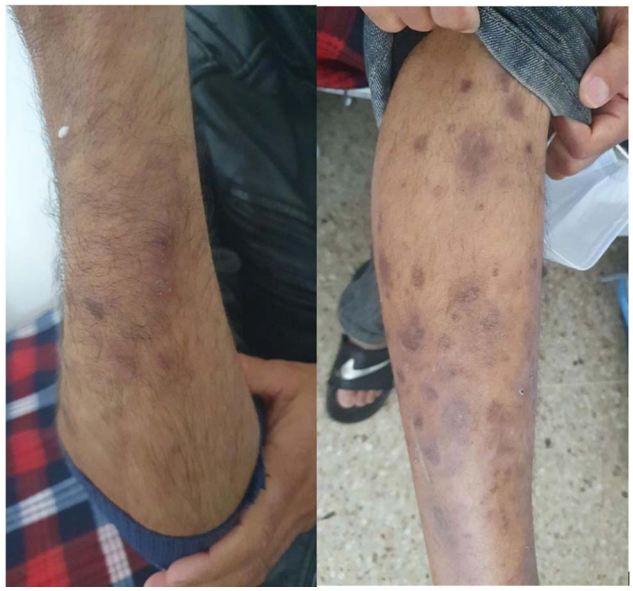

**Figure 2.** Multiple violaceous papules and Plaques on forearms and legs of the Patient.

oral azathioprine 80 mg twice daily (2 mg/kg per day). The evolution showed ulcer healing and re-epithelialization with the development of corneal neovascularization after 2 months (Fig. 4).

However, the final visual acuity was limited to light perception, and the follow-up was without particularities.

## Clinical discussion

Lichen planus is a chronic inflammatory pathology affecting primarily the skin, oral and genital mucosa, and rarely the eyes[1]. The pathophysiology remains unknown, but an autoimmune mechanism mediated by CD8 + cytotoxic T lymphocytes is presumed, recruiting into the skin and mucous membranes of genetically predisposed individuals[5].

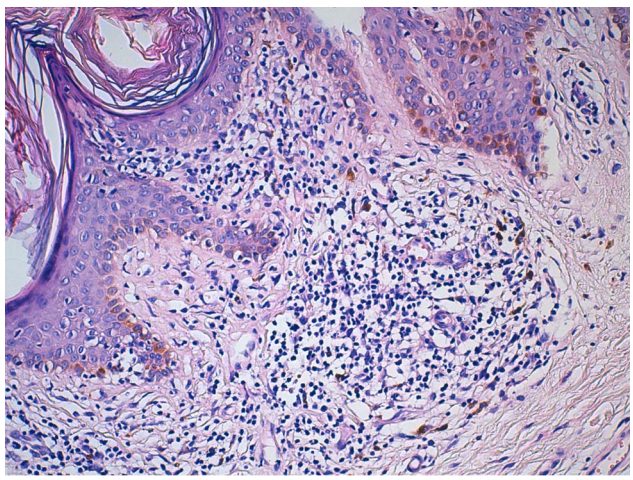

**Figure 3.** (HxE) x20: Histological section of skin biopsy with lichen planus. Vacuolated basal cells. Sub-epithelial band of lymphoplasmacytic cells.

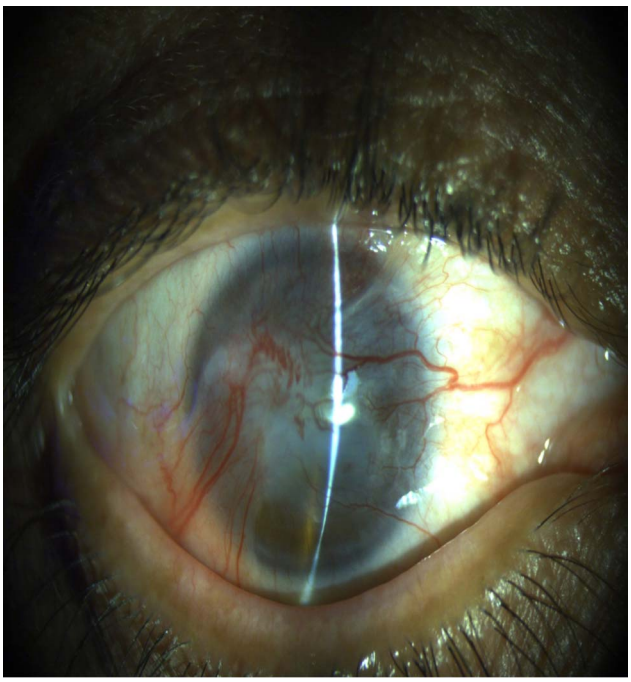

**Figure 4.** Photograph of the anterior segment showing complete healing of the ulcer with conjunctivalization and corneal neovascularization.

Ocular involvement in lichen planus is extremely rare, affecting the eyelids, lacrimal ducts, conjunctiva, and cornea.

Keratitis associated with lichen planus is even rarer, with only a few cases described in the literature.

The origin of corneal involvement in the context of lichen planus is controversial. It is unclear whether this involvement is directly caused by lichen planus as a systemic pathology, by the scarring process in the conjunctiva, by abnormalities associated with the tear film, or by a complex combination of all these factors[6].

The clinical presentation is highly polymorphic and can manifest as keratoconjunctivitis, with multiple pigmented lesions limited to the corneal epithelium[7]. It can also present as variable-severity superficial punctate keratitis associated with corneal pannus, with or without the involvement of the eyelids or conjunctiva[8]. Rhee *et al*.[9] reported a case of genital and oral cutaneous lichen planus associated with keratouveitis and cicatricial conjunctivitis. Like our case, lichen planus can also have a severe presentation, threatening the eye's functional prognosis with corneal perforation, as reported by Gomez-Elizondo *et al*.[10]. However, these manifestations lack specificity for lichen planus, posing a challenge in clinically differentiating it from other types of cicatrizing ocular conditions, particularly in cases where there is no concurrent skin or mucosal involvement.

However, these clinical manifestations lack specificity for lichen planus, presenting a considerable challenge in distinguishing it from other forms of keratoconjunctivitis conditions, especially in instances where there is an absence of concurrent skin or mucosal involvement, In this conjunctival biopsy can prove beneficial. However, in our case, conjunctival biopsy was not performed due to the absence of conjunctival scarring lesions and the confirmed histological evidence of cutaneous lichen planus.

To our knowledge, this is the second case to note an association between lichen planus and Peripheral ulcerative keratitis. Neo *et al*.[11] reported a case of bilateral peripheral ulcerative keratitis with confirmed oral lichen planus by biopsy of the oral mucosa.

Corticosteroid therapy and cyclosporine are typically regarded as first-line treatments for ocular lichen planus, showing favorable outcomes in the majority of studies. However, our case represents a corticosteroid-resistant ocular lichen planus, and it is noteworthy that only one case of keratitis associated with a lichen planus corticosteroid-resistant and refractory to cyclosporin case has been reported in the literature[12]. A tectonic graft was recommended in this case but could not be carried out due to a lack of grafts in the center.

The favorable response to azathioprine and cyclosporin reinforces our diagnosis of lichen planus for peripheral ulcerative keratitis, as found in several publications[8,11,13].

A general examination, including examination of the skin and mucous membranes, should be systematic in the presence of any peripheral ulcerative keratitis.

## Conclusion

Corneal involvement in lichen planus is rare and can jeopardize the functional prognosis of the eye.

Lichen planus should be considered in any case of peripheral ulcerative keratitis associated with cutaneous-mucosal manifestations.

Prompt initiation of corticosteroids and immunosuppressants such as cyclosporin and azathioprine is crucial for effective management and favorable outcomes in such cases.

## Ethical approval

Not applicable.

## Consent

Consent was taken from the next of kin due to the death of the patient.

## Source of funding

No specific grant was received from any public, commercial, or non-profit funding agency for this research.

## Author contribution

All authors contributed to the interpretation and assembly of data and media, drafted or revised the article, gave final approval of the manuscript, and agreed to be accountable for all aspects of the work.

## Conflicts of interest disclosure

The authors declares no conflicts of interest.

## Research registration unique identifying number (UIN)

Not applicable.

## Guarantor

Hamza Lazaar.

## Data availability statement

No dataset has been generated and/or analyzed in this article.

## Provenance and peer review

Not commissioned, externally peer-reviewed.

## References

[1] Gorouhi F, Davari P, Fazel N. Cutaneous and mucosal lichen planus: a comprehensive review of clinical subtypes, risk factors, diagnosis, and prognosis. Sci World J 2014;2014:1–22.
[2] Boyd AS, Neldner KH. Lichen planus. J Am Acad Dermatol 1991;25: 593–619.
[3] Mohebbi M, Mirghorbani M, Banafshe Afshan A, *et al*. Lichen planus in an ocular surface: major presentations and treatments. Ocul Immunol Inflamm 2018;27:987–94.
[4] Sohrabi C, Mathew G, Maria N, *et al*. The SCARE 2023 guideline: updating consensus Surgical Case Report (SCARE) guidelines. Int J Surg Lond Engl 2023;109:1136.
[5] Farhi D, Dupin N. Pathophysiology, etiologic factors, and clinical management of oral lichen planus, part I: Facts and controversies. Clin Dermatol 2010;28:100–8.
[6] Diniz CM, Vieira LA, Rigueiro MP, *et al*. Bilateral Cicatricial Keratoconjunctivitis Associated with Lichen Planus: Case Report. Arq Bras Oftalmol 2008;71:881–5.
[7] Soleimani M, Tabatabaei SA, Mirshahi R, *et al*. Corneal involvement by lichen planus pigmentosus. Ocul Immunol Inflamm 2017;26:897–9.
[8] Huang S, Channa P. Cicatricial keratoconjunctivitis associated with lichen planus. GMS Ophthalmol Cases 2015;5:Doc08.
[9] Rhee MK, Mootha VV. Bilateral keratoconjunctivitis associated with lichen planus. Cornea 2004;23:100–5.
[10] Gomez-Elizondo DE, Lopez-Martinez M, Ruiz-Lozano RE, *et al*. Corneal perforation associated with isolated ocular lichen planus: a case report. Eur J Ophthalmol 2020;31:6.
[11] Neo YN, Farwana R, Ahmoye S, *et al*. Peripheral ulcerative keratitis associated with lichen planus. Cornea 2023;42:1458–60.
[12] Hutnik CM, Probst LE, Burt WL, *et al*. Progressive, refractory keratoconjunctivitis associated with lichen planus. Can J Ophthalmol 1995;30: 211–4.
[13] Verma K, Mittal R, Ma K Y. Azathioprine for the treatment of severe erosive oral and generalized lichen planus. Acta Derm Venereol 2001;81: 378–9.