

Lupus pernio as an important feature of different cutaneous diseases in a case series of 57 cases

Khalifa E. Sharquie, Raed I. Jabbar

¹Department of Dermatology, College of Medicine, University of Baghdad. Medical City Teaching Hospital, ²Department of Dermatology, Fallujah Teaching Hospital, Al-Anbar Health Directorate, Anbar, Iraq

Corresponding author: Prof. Khalifa E. Sharquie, MD PhD, E-mail: ksharquieprof@yahoo.com

ABSTRACT

Background: Lupus pernio has traditionally been associated with sarcoidosis, presenting as purple nodules or plaques on the cheeks, ears, and nose. Now, it has been identified alongside different diseases, such as cutaneous tuberculosis, cutaneous leishmaniasis, rosacea, and discoid lupus erythematosus. **Objective:** The objective was to record all dermatological conditions possessing the same clinical features as lupus pernio. **Patients and Methods:** This is a case series, descriptive study conducted from 2014 to 2024 where all patients with different skin diseases associated with lupus pernio were included. This study was mainly concerned with nasal involvement having features of lupus pernio. The diagnosis of each disease was based on clinical features supported by histopathological examination and confirmed by specific investigations. **Results:** Fifty-seven patients with typical clinical features of lupus pernio were included in this study, with their ages ranging from 2 to 60 years, with a mean of 33 years, with 30 (52.63%) females and 27 (47.36%) males. The patients had different skin diseases with typical features of lupus pernio in the form of bulky swelling of the nose with associated dusky erythematous rash on the cheeks together with cutaneous leishmaniasis in 23 (40.35%) patients, rosacea in 16 (28.07%) cases, cutaneous tuberculosis in 7 (12.28%), sarcoidosis in 6 (10.52%), and discoid lupus erythematosus in 5 (8.77%). **Conclusion:** This was the first study recording lupus pernio in association with different skin diseases. All patients presented with a dusky red, bulky nose with nasal mucosa invasion in many cases but, in particular, patients with cutaneous leishmaniasis and tuberculosis. This was in contrast with the dermatological literature in which only sarcoidosis was well defined to be associated with lupus pernio.

Key words: Lupus pernio, Sarcoidosis, Cutaneous leishmaniasis, Histopathology, Cutaneous tuberculosis

INTRODUCTION

Sarcoidosis is a multi-system disorder of unknown etiology that may affect nearly any part of the body [1]. The skin is the most common part of extra-thoracic involvement. Cutaneous sarcoidosis (CS) is present in up to 30% of sarcoidosis cases, while it might be pure CS in 25% [2]. Sarcoid-specific lesions typically exist histologically with a granulomatous alignment in the tissue [3-5].

Lupus pernio is one of the skin manifestations of sarcoidosis [6]. It was first described by Besnier in 1889 who suggested the term *lupus pernio* to describe a patient with multiple purple nodules or plaques of a

chronic nature usually affecting the cheeks, nose, ears, and extremities [7].

Lupus pernio tends to be associated particularly with other forms of chronic fibrotic sarcoidosis, including upper respiratory tract sarcoidosis, lacrimal gland, bone cysts, and renal sarcoidosis, and with hypercalcemia and hyperglobulinemia. Lupus pernio tends to persist as lesions for more than two years, seldom to resolve [8]. The facial disfigurement may cause emotional scarring, which may justify aggressive lines of treatment, including plastic surgery [9].

Despite intensive investigation, the cause of lupus pernio and other forms of CS is unknown. Sarcoidosis is

How to cite this article: Sharquie KE, Jabbar RI. Lupus pernio as an important feature of different cutaneous diseases in a case series of 57 cases. Our Dermatology Online. 2026;17(2):167-171.

Submission: 13.05.2025; **Acceptance:** 15.08.2025

DOI: 10.7241/ourd.20262.4

identified as a chronic cell-mediated immune response to an unknown antigen, in which CD4 T-lymphocytes and activated macrophages release cytokines that trigger the formation of granulomas. At least some cases of CS may be due to an unusual host reaction to one or more infective agents, such as *Mycobacterium tuberculosis*, histoplasmosis, and other fungi. Yet, lupus pernio itself is not infectious [10-12].

There are many skin diseases sharing the same clinical features with lupus pernio of sarcoidosis, such as cutaneous tuberculosis (CTB), rosacea, cutaneous leishmaniasis, discoid lupus erythematosus (DLE), granulomatous secondary syphilis, and others, which is poorly documented in the medical literature [3,10,13-15]. Hence, reaching a definite diagnosis can be difficult, and the presence of specific histological features and other laboratory tests of each disease may be the ideal method to reach a definite diagnosis.

As lupus pernio remains not well determined and only well documented in patients with sarcoidosis, the aim of this study was to record all dermatological conditions that possess the same clinical features as lupus pernio.

PATIENTS AND METHODS

This was a case series, descriptive study conducted from 2014 to 2024 in which all patients with different skin diseases associated with lupus pernio were included. Full demographic and clinical features were thoroughly described and analyzed. The study was mainly concerned with nasal involvement having features of lupus pernio. The study was conducted in accordance with principles of the Declaration of Helsinki, and informed consent was taken from each participant or his/her guardian. A thorough history was taken from each patient regarding their name, age, sex, address, disease onset, associated symptoms, and medical and drug history.

A close physical examination was done, including the site of involvement, color, associated signs, and size of the lesion.

The diagnosis of each disease was based on clinical features supported by histopathological examination and confirmed by specific investigations as well as other techniques to detect any forms of systemic involvement, especially in sarcoidosis patients.

RESULTS

Fifty-seven patients with typical clinical features of lupus pernio were included in the study, with their ages ranging from 2 to 60 years, with a mean of 33 years, with 30 (52.63%) females and 27 (47.36%) males. The patients had different skin diseases with typical features of lupus pernio in the form of bulky swelling of the nose with associated dusky, erythematous rash on the cheeks together with the following:

Cutaneous leishmaniasis included 23 (40.35%) patients, 14 (60,87%) males and 9 (39,13%) females; their ages ranged from 2 to 60 years, with a mean of 30 years. 12 (52.17%) cases presented with bilateral, indurated, erythematous cheeks in addition to nasal involvement, while 11 (47.82%) cases presented with an erythematous, bulky nose only. In 9 (39.13%) cases, there was the invasion of the nasal mucosa (Figs. 1a and 1b).

Rosacea was recorded in 16 (28.07%) cases, with 13 (81.25%) females and 3 (18.75%) males; their ages ranged from 30 to 63 years, with a mean of 45 years. All patients presented with multiple papules and pustules distributed on persistent erythema of the central face (Figs. 2a and 2b). Bulky nasal involvement was seen in all cases, but there was no nasal mucosal invasion.

Cutaneous tuberculosis was observed in 7 (12.28%) patients, with their ages ranging from 12 to 60 years, with a mean of 37 years, with 5 (71.42%) males and 2 (28.57%) females. In all cases, the nose was affected, including the nasal mucosa of the nostril presenting with erythematous, papulonodular lesions (Figs. 3a and 3b), apart from one case presenting with



Figure 1: a) Cutaneous leishmaniasis with lupus pernio in a male patient. b) Bulky nose with some invasion of the nasal mucosa in a female patient.



Figure 2: Rosacea with lupus pernio in a) a male patient and b) a female patient.

ulcerative lesions. The histopathological features of CTB ranged from diffuse lymphocytic infiltrate throughout the entire dermis without granuloma formation to well-developed granuloma with central necrosis surrounded by lymphocytes (Fig. 3c).

Sarcoidosis was detected in 6 (10.52%) patients, 4 (66.66%) females and 2 (33.33%) males; their ages ranged from 25 to 60 years, with a mean of 41 years. The lesions presented as erythematous to violaceous papules, nodules, or plaques involving the cheeks, nose, and eyelids. In 2 (33.33%) cases, the lesions extended to involve the extremities and trunk. (Figs. 4a and 4b). All patients demonstrated nasal skin involvement, with 3 (50%) cases exhibiting the invasion of the nasal mucosa. The histopathological assessment of sarcoid lesions showed a marked granulomatous reaction consisting of multiple non-caseating granulomas with sparse lymphocytic infiltrate at the border of the granulomas (so-called naked granuloma). These granulomas were loaded with foamy cells, and there were different types of giant cells (Fig. 4c).

Discoid lupus erythematosus was detected in 5 (8.77%) cases, with 3 (60%) males and 2 (40%) females; their ages ranged from 20 to 45 years, with a mean of 38 years. The skin lesions were characterized by multiple, erythematous, scaly plaques with bulky nasal involvement but without nasal mucosal invasion (Figs. 5a and 5b).

In all patients, physical examination, in addition to appropriate investigations, showed no obvious systemic involvement at diagnosis.

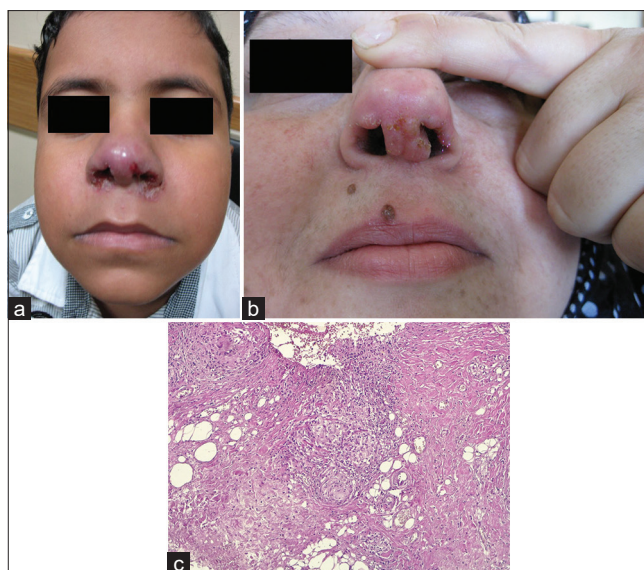


Figure 3: (a and b) Cutaneous tuberculosis with lupus pernio involving the nose with nasal mucosal invasion in both patients. c) H&E-stained section from the patient in Fig. b showing diffuse lymphocytic infiltration with poorly developed granuloma with multiple giant cells (10x).

DISCUSSION

Lupus pernio has traditionally been associated with sarcoidosis, presenting as purple nodules or plaques on the cheeks, ears, and nose [16]. However, in the current study, it was identified alongside diseases such as CTB, cutaneous leishmaniasis, rosacea, and DLE. This raises important questions about the interaction of lupus pernio with other skin diseases.

Lupus pernio in sarcoidosis was detected in 10.52% of the patients and found to produce isolated skin lesions with some invasion of the nostril mucosa in 50% of the cases, but without systemic involvement. These results were in line with another Iraqi study, in which systemic involvement was not an important feature [10], while they differed from previous studies that recorded 40–62% of patients with CS developing systemic involvement [17,18]. The exact etiology beyond this discrepancy could not be well explained. Still, it could be racial, or the early presentation with rapid diagnosis before systemic involvement could be the most important cause of this difference.

Still, cutaneous leishmaniasis is the most common cause (40.35%) of lupus pernio, as reported in this study. This is because cutaneous leishmaniasis is endemic in Iraq and involves mostly the exposed parts of the body, including the face [19,20].

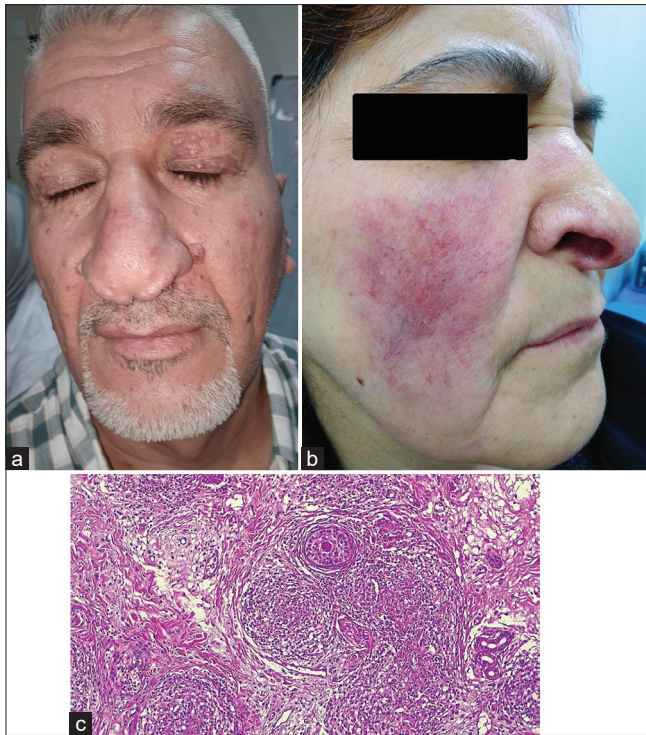


Figure 4: Sarcoidosis with lupus pernio involving the eyelids and nose a) and cheeks and nose b) with nostril mucosal invasion in both cases. c) H&E-stained section from the patient in Fig. a showing multiple non-caseating granulomas (naked granuloma) loaded with foamy cells (10x).

A striking observation in this study was the presence of a lupus pernio lesion with nasal mucosal involvement in patients with cutaneous leishmaniasis and CTB. Nasal mucosal invasion was recorded in 39.13% of cases with cutaneous leishmaniasis and in 100% of cases with CTB, which was in contrast to DLE and rosacea, where such involvement was absent. This suggests that granulomatous infections such as TB and leishmaniasis may share some pathogenic mechanisms with sarcoidosis in eliciting lupus pernio lesions as some cases of sarcoidosis had a previous history of tuberculosis in the same patient [10].

Rosacea was another important condition that was associated with a lupus pernio lesion in the present study. Although rosacea typically manifests as centrofacial erythema and phymatous changes, its ability to present with a lupus pernio lesion is well-documented [3,10]. These findings propose that chronic inflammatory changes in rosacea may induce a tissue reaction resembling lupus pernio of sarcoidosis.

Similarly, DLE was found to exhibit a lupus pernio lesion involving the face and nose resembling lupus pernio of sarcoidosis. Both DLE and sarcoidosis can

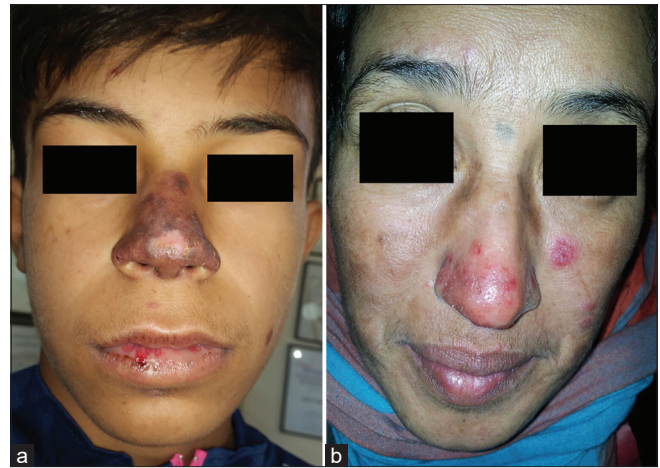


Figure 5: (a and b) Discoid lupus erythematosus with lupus pernio.

present with chronic erythematous plaques, and the overlap of skin findings in both conditions was reported in this study.

In both DLE and rosacea, no nasal mucosal involvement was recorded for unknown reasons, but the most acceptable explanation is that nasal mucosal invasion appears in granulomatous skin diseases such as TB, leishmaniasis, and sarcoidosis rather than in other inflammatory conditions such as rosacea and DLE.

One of the most important implications of our results is the need for pathological confirmation in cases of suspected sarcoidosis. Given that cutaneous diseases like CTB, DLE, rosacea, and leishmaniasis can present with the same cutaneous features, especially when involving the face, a misdiagnosis can lead to mismanagement. This highlights the necessity of taking a biopsy, and microbiological examinations, particularly in endemic areas for TB and leishmaniasis.

CONCLUSION

This was the first study that recorded lupus pernio in association with different skin diseases. All patients presented with a dusky red, bulky nose with nasal mucosa invasion in many cases, but in particular, patients with cutaneous leishmaniasis, tuberculosis, and sarcoidosis. This is in contrast with the dermatological literature, in which only sarcoidosis was reported to be associated with lupus pernio. This study focused on the clinical and histopathological features of lupus pernio, a hallmark feature of sarcoidosis, and its relation to different skin diseases other than sarcoidosis. The results are fascinating and provide insight into the diversity of lupus pernio presentations and clinical

features that are shared with other dermatological conditions. Accordingly, lupus pernio is not a specific feature of sarcoidosis but can be more observed in diverse cutaneous diseases.

Statement of Human and Animal Rights

All the procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the 2008 revision of the Declaration of Helsinki of 1975.

Statement of Informed Consent

Informed consent for participation in this study was obtained from all patients.

REFERENCES

- Drent M, Crouser ED, Grunewald J. Challenges of sarcoidosis and its management. *New Eng J Med.* 2021;385:1018-32.
- Boch K, Langan EA, Zillikens D, Ludwig RJ, Kridin K. Evaluation of clinical and laboratory characteristics of patients with cutaneous sarcoidosis: A single-center retrospective cohort study. *Front Med.* 2022;9:980507.
- Haimovic A, Sanchez M, Judson MA, Prystowsky S. Sarcoidosis: A comprehensive review and update for the dermatologist: part I. Cutaneous disease. *J Am Acad Dermatol.* 2012;66:e691-9.
- Grunewald J, Grutters JC, Arkema EV, Saketkoo LA, Moller DR, Müller-Quernheim J. Publisher correction: Sarcoidosis. *Nat Rev Dis Primers.* 2019;5:45.
- Ezeh N, Caplan A, Rosenbach M, Imadojemu S. Cutaneous sarcoidosis. *Dermatol Clin.* 2023;41:455-70.
- Kerdel FA, Moschella SL. Sarcoidosis: An updated review. *Jam Acad Dermatol.* 1984;11:1-19.
- Besnier E. Lupus pernio de la face: synovites fongueuses symétriques des extrémités supérieures. *Ann Dermatol Syphilol.* 1889;10:333-6.
- DJ. Sarcoidosis. In: Burns T, Breathnach S, Cox N, Griffiths C, editors. *Rooks Textbook of Dermatology.* Vol. 3. Massachusetts: Blackwell Science Ltd.; 2010. p. 61.1-21.
- Yanardag H, Pamuk ON, Pamuk GE. Lupus pernio in sarcoidosis: Clinical features and treatment outcomes of 14 patients. *J Clin Rheumatol.* 2003;9:72-6
- Sharquie K, Kubaisi TA, Sharquie IK. Cutaneous sarcoidosis as upsurging mimicking granulomatous diseases. *J Pak Assoc Dermatol.* 2024;34:698-70.
- Loke WSJ, Herbert C, Thomas PS. Sarcoidosis: Immunopathogenesis and immunological markers. *Int J Chron Dis.* 2013;2013:928601.
- Facco M, Cabrelle A, Teramo A, Olivieri V, Gnoato M, Teolato S, et al. Sarcoidosis is a Th1/Th17 multisystem disorder. *Thorax.* 2011;66:144-50.
- Ishak R, Kurban M, Kibbi AG, Abbas O. Cutaneous sarcoidosis: Clinicopathologic study of 76 patients from Lebanon. *Int J Dermatol.* 2015;54:33-41.
- Noe MH, Rosenbach M. Cutaneous sarcoidosis. *Curr Opin Pulm Med.* 2017;23:482-6.
- Bimbi C. Sarcoidosis mimicking seborrheic dermatitis: Another case of Sherlockian Dermatology. *Our Dermatol Online.* 2014;5:423-5.
- Howard A. Non-infectious granulomas. In: Bologna J L, Scaffer J V, Ceroni L. *Text book of dermatology*, 5th edition. China, Elsevir. 2024;93:1660-67.
- Paolino A, Galloway J, Birring S, Brex P, Larkin G, Patel A, et al. Clinical phenotypes and therapeutic responses in cutaneous-predominant sarcoidosis: 6-year experience in a tertiary referral service. *Clin Exp Dermatol.* 2021;46:1038-45.
- Byrne B, Goh A, Izham NF, Porter E, Field S. Systemic evaluation of cutaneous sarcoidosis: 15-year dermatology experience at University Hospital Limerick. *Clin Exp Dermatol.* 2022;47:850-7.
- Sharquie KE, Jabbar RI. Triple therapy for acute and chronic cutaneous leishmaniasis using oral zinc sulfate, oral ketoconazole and topical podophyllin. *J Pak Assoc Dermatol.* 2022;32:502-9.
- Sharquie KE, Jabbar RI. Leishmania recidivans could be induced by intralesional infiltration of cutaneous leishmaniasis by sodium stibogluconate, with new therapeutic trial. *J Clin Exp Invest.* 2022;13:em00788.

Copyright by Khalifa E. Sharquie, et al. This is an open-access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.

Source of Support: This article has no funding source.

Conflict of Interest: The authors have no conflict of interest to declare.