CHEST Original Research

SARCOIDOSIS

# The Treatment of Lupus Pernio*

## Results of 116 Treatment Courses in 54 Patients

*Eleni Stagaki, MD; William K. Mountford, PhD; Daniel T. Lackland, DrPH; and Marc A. Judson, MD, FCCP*

***Background:*** **Lupus pernio is a disfiguring sarcoidosis skin lesion that is difficult to treat and often causes a major psychosocial impact that may adversely affect the patient's quality of life. We reviewed the treatment outcome of 54 patients with lupus pernio who received 116 individual courses of treatment in our sarcoidosis clinic.**

***Methods:*** **Lupus pernio patients were identified from an institution-approved database. All patients were assessed at each clinic visit with facial photographs. By examining the photographs, the percentage of face involved (< 10%, 10 to 25%, > 25 to 50%, > 50%) was determined as was the effect of therapy (resolution, near resolution, improvement, no change, worsening). Medications included infliximab-containing regimens; systemic corticosteroids; noninfliximab, noncorticosteroid agents; and corticosteroids plus noncorticosteroid agents.**

***Results:*** **In terms of achieving resolution or near resolution, infliximab regimens were superior to all others (infliximab, 77%; corticosteroids plus noncorticosteroids, 29%; corticosteroids, 20%; noncorticosteroids, 11%; infliximab vs other therapies: corticosteroids plus noncorticosteroids, $p = 0.0015$; corticosteroids, $p = 0.0005$; noncorticosteroids, $p = 0.0002$). The percentage of facial involvement also improved most with infliximab. Evaluating a secondary analysis of achieving resolution, near resolution, or improvement, infliximab (92%) was superior to noncorticosteroids (20%; $p < 0.0001$) and corticosteroids plus noncorticosteroids (56%; $p = 0.0098$), but not corticosteroids (72%; $p = 0.2456$); and noncorticosteroid agents were inferior to all other regimens.**

***Conclusions:*** **Infliximab appears superior to systemic corticosteroids with or without additional agents for the treatment of lupus pernio. Noninfliximab, noncorticosteroid-containing regimens are of limited use for this condition.** ***(CHEST 2009; 135:468–476)***

**Key words:** infliximab; lupus pernio; skin; systemic corticosteroids; therapy

**Abbreviations:** MUSC = Medical University of South Carolina; SASI = Sarcoidosis Activity and Severity Index

Lupus pernio is an indolent, red-to-purple, or violaceous nodular or plaque-like sarcoidosis skin lesion that may affect the cheeks, nose, chin, forehead, ears, perioral, or periocular regions.[1] The lesions are often disfiguring, and their psychosocial impact may adversely affect the patient's quality of life. This form of sarcoidosis often portends a poor prognosis.[2–4]

Despite the gravity of this clinical entity, there is a paucity of information concerning its treatment. Treatment studies of lupus pernio include case reports,[5–10] case series,[11–16] and one small (n = 14) open label trial.[17] This article summarizes our experience with the treatment of lupus pernio over the past 8 years in our sarcoidosis clinic, describing the outcome of 54 sarcoidosis patients with lupus pernio who received a total of 116 separate treatments.

## Materials and Methods

This study was approved by the Medical University of South Carolina (MUSC) Institutional Review Board.

### *Study Population*

The patients were identified retrospectively from a clinical sarcoidosis database approved by MUSC. Subjects were evaluated in the MUSC Sarcoidosis Clinic from May 12, 2000, until January 07, 2008. All patients had biopsy-proven sarcoidosis and definite lupus pernio based on the organ involvement instrument from A Case Control Etiologic Study of Sarcoidosis.[18] That is,

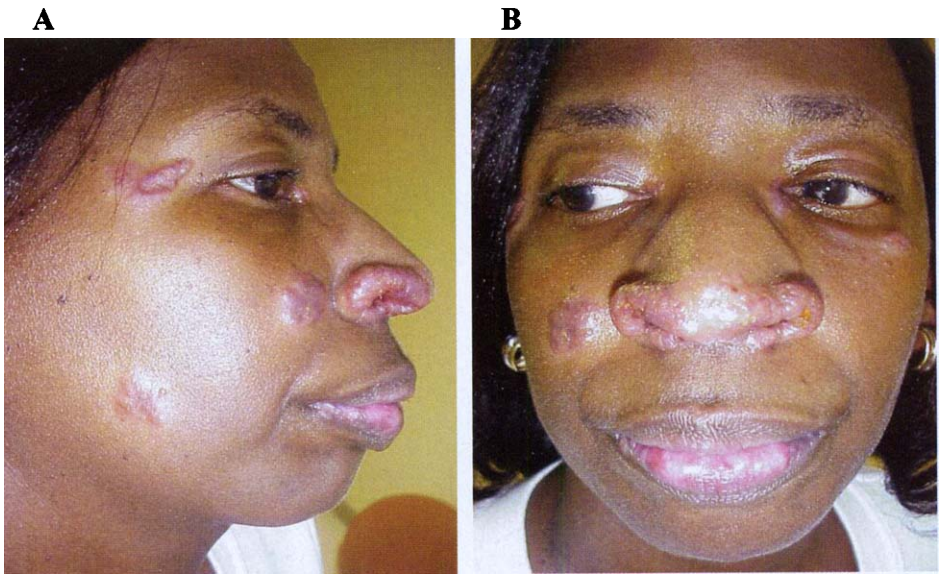


FIGURE 1. A patient with lupus pernio skin lesions on the nose, forehead, and around the eyes: 10 to 25% of the face surface. The patient has given permission to use these pictures.

patients were defined as having lupus pernio if they had biopsy-proven sarcoidosis and indolent, red-purple or violaceous, indurated, nodular, or plaque-like skin lesions affecting the nose, cheeks, chin, forehead, ears, and perioral or periocular regions that could not be attributed to another cause.[1]

*Data Collection*

Demographic data and clinical data related to extracutaneous sarcoidosis were obtained from a review of each medical record. The evaluation of lupus pernio lesions was based on information extracted from the medical record and photographs of the lupus pernio lesions taken at clinical visits. By using the medical record and photographs, lupus pernio lesions were assessed by the following: (1) the onset of lupus pernio; (2) the specific locations of the face that were involved (forehead, cheeks, periocular, perioral, chin, ears, and nose); and (3) the extent of the lesions characterized by the amount of face surface involved (< 10%, 10 to 25%, 25 to 50%, and > 50%). Photographic assessment of the skin lesions was done with knowledge of which photograph was performed before and after therapy. However, photographic assessment was blinded in terms of the treatment regimen that was used. Figure 1 shows an example of a lupus pernio patient assessed by this method.

*Follow-up and Treatment*

Subjects were examined and treated by the same physician (M.A.J). Patients who were primarily receiving sarcoidosis treatment because of organ involvement other than lupus pernio were excluded from the analysis. The schedule of follow-up visits and the treatment plan were not standardized but were tailored to the individual patient. The dosing of the major drugs administered is outlined as follows. Corticosteroid dosing was usually 20 to 40 mg/d of prednisone equivalent. The dose of methotrexate ranged between 10 to 20 mg/wk. Hydroxychloroquine doses ranged between 200 mg/d and 400 mg/d. Infliximab dosing basically followed a previously published dosing regimen[19,20]: 5 mg/kg IV at weeks 0, 2, and 6, and then every 6 weeks. For the purposes of this analysis, we characterized the treatment regimens used for the lupus pernio patients as follows: (regimen 1) systemic corticosteroids alone; (regimen 2) noninfliximab, noncorticosteroid drugs including methotrexate, hydroxychloroquine or chloroquine, minocycline, azathioprine, thalidomide, pentoxyfylline, intralesional and topical corticosteroids, topical tacrolimus, and topical retinoic acid; (regimen 3) systemic corticosteroids plus one or more of the agents above in regimen 2; and (regimen 4) treatment regimens in which infliximab was used: infliximab alone, infliximab plus systemic corticosteroids, and/or plus agents above in regimen 2.

Treatment remained unchanged and uninterrupted in each treatment course. If there was an addition or discontinuation of one or more drugs, the treatment course was considered terminated and a new treatment regimen started. As a result, each subject could have one or more treatment courses. If treatment was interrupted more than a week, this was considered the end of a treatment course. Patients were only analyzed if they had evidence of lupus pernio after May 12, 2000 confirmed at our institution. If such a patient had complete resolution of skin lesions, a new treatment course would be analyzed if there was a de-escalation of therapy in terms of the number of drugs and/or drug dosages. All patients who received a course of infliximab had previously failed at least one previous course of therapy for lupus pernio.

*From the Third Pulmonary Department (Dr. Stagaki), Sismanoglio General Hospital, Athens, Greece; and Department of Biostatistics, Bioinformatics, and Epidemiology (Drs. Lackland and Mountford), and Division of Pulmonary and Critical Care Medicine (Dr. Judson), Department of Medicine, Medical University of South Carolina, Charleston, SC.

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*Correspondence to: Marc A. Judson, MD, Division of Pulmonary and Critical Care Medicine CSB-812, Medical University of South Carolina, 96 Jonathan Lucas St, Charleston, SC 29425; e-mail: judsonma@musc.edu*

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