Lupus Erythematosus Profundus: Case Reports

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GOAL

To describe lupus erythematosus profundus

OBJECTIVES

Upon completion of this activity, dermatologists and general practitioners should be able to:

1. Delineate the epidemiologic features of lupus erythematosus profundus.
2. Describe the location, manifestations, and systemic findings of lupus erythematosus profundus.
3. Discuss the diagnosis of lupus erythematosus profundus.

CME Test on page 468.

This article has been peer reviewed and approved by Michael Fisher, MD, Professor of Medicine, Albert Einstein College of Medicine. Review date: May 2001.

This activity has been planned and implemented in accordance with the Essentials and Standards of the Accreditation Council for Continuing Medical Education through the joint sponsorship of Albert Einstein College of Medicine and Quadrant HealthCom, Inc. The Albert Einstein College of Medicine is accredited by the ACCME to provide continuing medical education for physicians.

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Lupus erythematosus profundus is a rare manifestation of lupus. We conducted a study to observe the epidemiologic and clinical aspects of lupus erythematosus profundus, its development into other types of lupus or systemic disease, and its response to treatment. Six patients were followed for variable periods. We conclude that lupus erythematosus profundus is a benign form of lupus, distinguishable by systemic manifestations, clinical evolution and resolution, and heterogeneity in clinical manifestations. A skin biopsy is necessary to make an accurate diagnosis.

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Lupus erythematosus profundus (also known as lupus profundus, lupus panniculitis, or Irgang-Kaposi panniculitis) is a rare manifestation of lupus present in 2% of cases.[1] As in other variants of lupus, the antigenic stimulus that starts the autoimmune response is unknown, as well as the reason why the infiltrate of lymphocytes is in the deep dermis and subcutaneous fat. It seems logical that if the tissular

Case Reports*

Patient No.	Localization	Clinical Manifestations	Renal Function	Hematologic	Autoantibodies	Other Findings	Classification
1	Face	Asymptomatic	Normal	Normal	—	—	CCL
2	Face	Pain	Normal	Normal	—	High C3 and CH50	CCL
3	Arms and neck	Asymptomatic	Normal	Leukopenia	ANA, anti-Ro, antimitochondrial	High C4	CCL
4	Perineum	Prurigo	Normal	Normal	ANA, anti-DNA, antimitochondrials, antithyroglobulin, anti–smooth muscle	Cold agglutinines	CCL
5	Knee and thigh	Pain	Normal	Normal	—	—	CCL†
6	Sacrum	Prurigo	Normal	Normal	—	—	CCL†

*ANA indicates antinuclear antibodies; Anti-Ro, Ro/Sjögren syndrome antigen A antibodies; Anti-DNA, anti–double-stranded DNA antibodies; CCL, chronic cutaneous lupus erythematosus.

†Only cutaneous manifestations of chronic lupus erythematosus.

injury is observed in an almost selective way in deep dermis and subcutaneous tissue, the pathogenic aspects and eliciting factors, although similar to other varieties of lupus, must have a hint of difference. Because the eliciting mechanism is different, systemic findings, conversion to other types of lupus, and the evolution of the lesions should be different. To investigate this, we conducted a study to observe the epidemiologic and clinical aspects of lupus erythematosus profundus, its development into other types of lupus or systemic disease, and its response to treatment.

Methods

We examined all skin biopsies that were obtained in our hospital from 1992 to 1999 that showed histopathologic markers of lupus. Of the 73 samples collected, only 6 were retrospectively diagnosed as lupus erythematosus profundus. The diagnosis was confirmed if one or more skin biopsies matched the typical changes of this condition[2]: (1) lobular infiltrates of lymphocytes, plasma cells, and histiocytes and (2) fat necrosis.

The following parameters were observed:

- Age at diagnosis
- Sex
- Follow-up period
- Site and clinical manifestations of panniculitis
- Systemic findings—arthritis, myalgia, and pleurisy
- Laboratory findings—renal function, blood counts, and autoantibodies, including: antinuclear antibodies (ANA), Ro/Sjögren syndrome antigen A antibodies (anti-Ro), anti–double-stranded DNA antibodies (anti-DNA), La/Sjögren syndrome antigen B antibodies (anti-La), antiphospholipid antibodies, and other non–lupus-specific antibodies
- Classification of the patient into acute, subacute, or chronic cutaneous lupus erythematosus. If panniculitis was the only physical finding of lupus, then the patient was included in the chronic group

Results

Seventy-three biopsies with histopathologic markers of lupus were obtained, 6 of which had lupus erythematosus profundus diagnoses (8.2%)(Table). Four of the 6 patients were females (female:male ratio, 2:1). The

mean age was 53.67 years (range, 43 years), and the mean follow-up period was 39.83 months (range, 13.4 years).

The sites of panniculitis were as follows: face (2), arms and neck (1), perineum (1), knee and thigh (1), and sacrum (1). Clinical findings were diverse; 2 patients had no symptoms, 2 had painful lesions, and 2 had pruriginous lesions.

All patients in our study were categorized as chronic lupus erythematosus. Four patients had typical cutaneous lesions and confirmatory biopsy, and the other 2 patients had panniculitis as the only cutaneous manifestation of chronic lupus. Other than these lesions, photosensitivity was the only cutaneous manifestation found. None of the patients developed any other type of lupus, and panniculitis appeared just once and was permanently resolved. No systemic findings were observed, except in one patient who had scleroderma, Sjögren syndrome, and primary biliary cirrhosis.

Laboratory findings included high titers of complement in 2 of 6 patients and leukopenia in another one. The rest of the parameters were in the normal range. Autoantibody results were positive in 2 patients.

All patients were treated with antimalarial drugs, topical corticosteroids, and photoprotection. The therapy resulted in scars that developed atrophic and pigmented areas.

Comment

The primary difference between lupus erythematosus profundus and other types of lupus is epidemiologic. In our study, lupus erythematosus profundus was more frequent in females. Some authors report that the mean age of appearance is between 20 and 40 years,[3] but we found a higher mean age in our group of 53.67 years.

As in previous studies, our study found the main sites to be the face and proximal areas of the arms and thighs (4 of 6 patients). There is no typical symptomatology, although panniculitis in skin folds develop pruritus, which is often the symptom that leads the patients to the hospital, making diagnosis possible. This heterogeneity of clinical manifestations shows the importance of using a skin biopsy to diagnose lupus erythematosus profundus.

None of the patients fulfilled the 1982 American Rheumatism Association criteria for systemic lupus erythematosus.[4] The only systemic finding associated was photosensitivity, which never coincided with positive anti-Ro antibodies. Photosensitivity is a frequent symptom in patients with lupus, with up to 63% being reported in some studies.[5]

Despite some reports that patients with lupus erythematosus profundus have positive ANA,[6] we found 2 such cases in this study and only one was permanent. This supports the theory of a different pathogenesis.

Díaz-Pérez et al[7] report that almost 50% of patients with lupus erythematosus profundus develop systemic abnormalities. Nevertheless, renal function is absolutely normal during the follow-up period. This, along with the lack of progression to other types of lupus and the absence of systemic findings, leads us to conclude that lupus erythematosus profundus is a benign form of lupus, with differences in systemic manifestations, clinical evolution, and resolution.

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