

ATYPICAL SCABIES IN SYSTEMIC LUPUS ERYTHEMATOSUS

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Recent reports have increasingly linked scabies with congenital or acquired immunosuppression. It is important to be aware of this form of infestation, because its unusual clinical signs may be easily overlooked. Scabies can mimic several diseases, including cutaneous or systemic lupus erythematosus (SLE).¹ However, the infestation has rarely been reported in patients with SLE. The objective of this report is to describe a rare case of atypical (exaggerated) scabies in SLE that illustrates a pitfall in the recognition of the infestation, since such recognition took place belatedly. We hope that this case will draw attention to the possibility of scabies whenever a widespread erythematosquamous dermatosis develops in a case of SLE.

Case Report

A 39-year-old woman had SLE for about 17 years, which was manifested as malar eruption, oral ulcers, leukopenia and arthritis. She also had a history of several episodes of pleural effusion. The patient was being treated with prednisone (30 mg/day), enalapril maleate (10 mg/day), and chloroquine (250 mg/day).

In September 1995, she presented with nonpruritic erythematosquamous plaques, without alopecia or scars in the scalp. She was treated with topical application of betamethasone valerate. Two months later, she developed a generalized papulosquamous eruption (Figure 1). Physical examination revealed erythematous and erythematosquamous papules without itching, particularly affecting abdomen, arms, legs and buttocks. Routine laboratory findings, including creatinine level, were normal. Serum protein electrophoresis demonstrated a polyclonal increase in gammaglobulin, and IgA was 663 mg/dL (normal, 90-450). Cryoglobulins were trace-positive (IgG, IgA). ANA was positive at 1:1280, with a homogenous pattern. The serum levels of C3 and C4 were within the normal

range. The rest of the immunologic studies were normal or negative. Although there was no clinical or serological activity of the disease, the lesions were presumed to be secondary cutaneous lupus erythematosus. Two punch biopsies were performed and scabies was diagnosed. Treatment with 1% lindane lotion in two courses resolved the cutaneous manifestations of the patient. She remained asymptomatic 24 months without relapse of her skin lesions.

Materials and Methods

The patient fulfilled the revised 1982 criteria for SLE of the American Rheumatism Association. The surgical specimens were fixed in 10% buffered formaldehyde solution, embedded in paraffin, and routinely processed. Consecutive 4 µm sections were stained with hematoxylin and eosin. Immunohistochemistry stains were performed by using the streptavidin-biotin-peroxidase method and a Techmate 500-220 automated immunostainer (Biotek, Santa Barbara, CA, USA). With the use of a pressure cooker, sections were boiled for a short time in sodium citrate buffer prior to the staining. Diaminobenzidine was used as chromogen. The following antibodies were utilized: CD3 (monoclonal, Dako, Glostrup, Denmark,

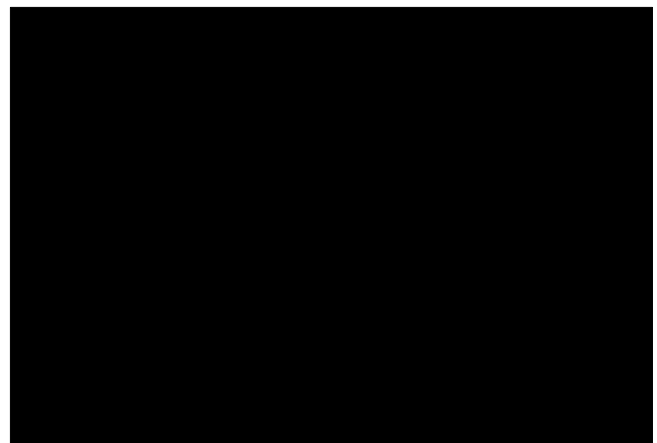


FIGURE 1. Clinical appearance of scabies manifesting as a disseminated erythematosquamous papular dermatosis. The lesions are evident in the trunk and in the right upper arm.

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1:100) for T-lymphocytes; CD20 (monoclonal, Dako, 1:50) for B-lymphocytes; CD68 (KP1, monoclonal, Dako, 1:25) for macrophages; and carcinoembryonic antigen (CEA, polyclonal, Dako, 1:500) for neutrophils.

Pathologic Findings

Two punch biopsy specimens of the erythematous-squamous papules were obtained from the abdomen and left leg, respectively. Histologic examination revealed a slightly acanthotic epidermis with focal parakeratosis, intracorneal pustules, and a mite of scabies in a burrow within the epidermal cornified layer (Figure 2). In the superficial dermis, there was a perivascular inflammatory infiltrate of mixed cell type with mononuclear cells, mainly macrophages, a few lymphocytes, neutrophils, and sparse



FIGURE 2. Papular lesion of scabies showing a scale-crust composed of parakeratotic cells and eosinophilic material, a scabie egg residing in the stratum corneum (on the left), and an adult mite within a subcorneal burrow (on the right). A mixed-cell perivascular infiltrate is present in the dermis (H&E, 25x).

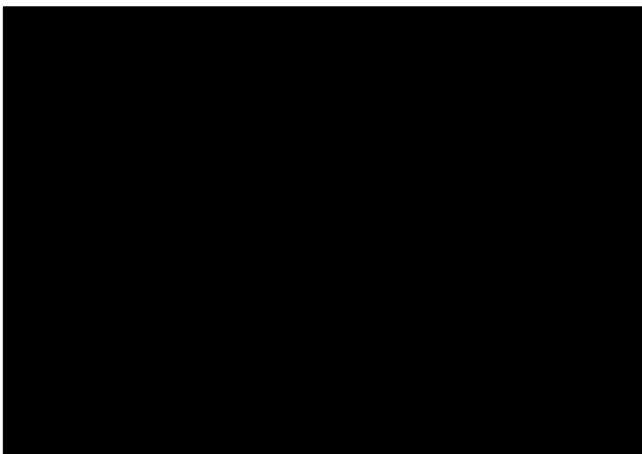


FIGURE 3A. The inflammatory-cell infiltrate consists of histiocytes, lymphocytes, and neutrophils arranged around blood vessels of both superficial and deep plexuses (H&E, 80x).

eosinophils (Figure 3). Serial sections were required to demonstrate eggs and mites. Immunohistochemical stains showed that macrophages were the predominant cells in the infiltrate. Fewer T-lymphocytes and very few B-lymphocytes and neutrophils were detected.

Discussion

In classic scabies, the lesions are few and roughly symmetrical. These are frequently eczematous and occur in the hands, flexor surface of the wrists, elbows, and the anterior axillary folds. Other typical locations are female breasts, around the umbilicus, penis, and lower portion of the buttocks. However, there are special forms of scabies resulting in unusual clinical presentations, atypical locations and unusual extension of the disease closely simulating other entities. Some unusual forms of scabies are the crusted (Norwegian) scabies and the atypical (exaggerated) scabies.² In many cases of these unusual forms, the pruritus of classic scabies lessens or disappears, the sites of involvement are different, i.e., the scalp, face, back, and nails, and burrows are less apparent. It has been said that scabies is a great imitator. Thus, it can masquerade as psoriasiform dermatosis, contact dermatitis, Darier disease, dermatitis herpetiformis, bullous pemphigoid, malignant lymphoma, and systemic or cutaneous lupus erythematosus,¹ among other entities.

Crusted scabies has been associated with a wide group of disorders, including mental retardation, diabetes mellitus, and immunosuppressed status. New features associated with severe *Sarcoptes scabiei* infestations exist. Severe forms of infestation develop preferentially in cases of imbalance in host-parasite relationships. Immune deficiency is one etiologic factor that is being increasingly recognized. Forms of scabies associated with congenital, acquired or iatrogenic suppression of the immune response

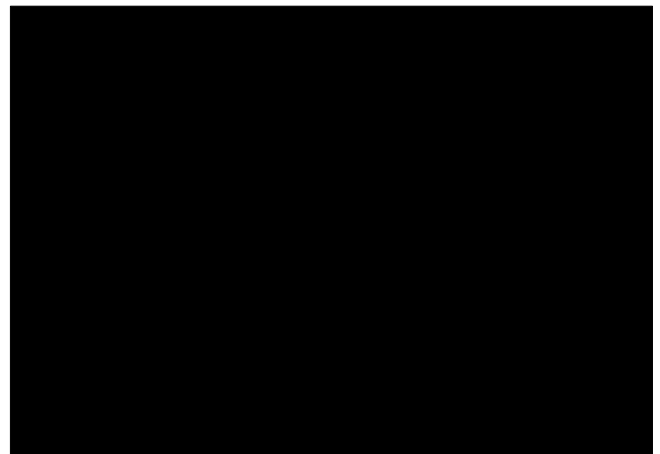


FIGURE 3B. Monoclonal antibody CD68 (KP1) decorating the cytoplasm of most of the cells of dermal infiltrate (160x).

include the following: 1) spontaneous essential immunodeficiency with *Pneumocystis carinii* infection in patients who do not have acquired immune deficiency syndrome (AIDS);³ 2) HTLV-I-associated T-cell leukemia;⁴ 3) HTLV-III infection (AIDS);² 4) lepromatous leprosy;⁵ 5) dialysis with or without immunosuppression;⁶ 6) corticosteroid therapy, including topical treatment;⁷ 7) renal transplantation;⁸ 8) bone marrow transplantation;⁹ 9) Bloom's syndrome;¹⁰; and 10) SLE.¹¹⁻¹⁶

Scabies has rarely been described in patients with SLE. A review of the literature revealed eleven reported cases.¹¹⁻¹⁶ Scabies in these eleven cases appeared to be more severe than usual, and seven of the patients developed crusted scabies. Atypical or exaggerated type of scabies was observed in only four patients.^{11,13}

An exaggerated form of scabies with generalized papulosquamous lesions occurred in our patient upon treatment for SLE. This special form can be designated scabies incognito, as the lesions were ascribed to SLE. In our case, systemic and topical corticosteroid therapy aggravated a primary cutaneous involvement, resulting in a severe form of infestation. The role of immunologic mechanisms in limiting the number of scabies mites is not well defined. However, the cellular immunity of the host appears to play a significant role.¹⁷ Histology suggests that a cell-mediated reaction causes the lesions of classic scabies. Lymphocytes constitute the majority of cells in the inflammatory infiltrate and T-cells dominate.¹⁷ The ratio of T-cells to B-cells in the lesions is greater than in the peripheral blood.¹⁸ This suggests a selection of T-lymphocytes in the inflammatory infiltrate. SLE is characterized by an alteration of humoral and cellular immunity,¹⁹ with loss of suppressor T-cell function.²⁰ Corticosteroid therapy decreases T-cell response. It is not surprising, therefore, that in our case macrophages were predominant in the infiltrate. The exaggerated form of scabies in the present case may be the result of underlying SLE and oral and topical usage of corticosteroids. Secondary bacterial infection may complicate scabies. The mite scybala and the fissures probably contribute to bacterial colonization.

It is necessary to emphasize the importance of a rapid diagnosis of scabies and that of the secondary bacterial

infection, because four patients reported with this complication in SLE died of septicemia.^{12,13,15,16} These patients had developed crusted scabies.

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