VERRUCOUS LUPUS ERYTHEMATOSUS

B. Žgavec, H.P. Soyer, W. Salmhofer, S. Hödl

ABSTRACT

Chronic cutaneous lupus erythematosus is a cutaneous expression of lupus erythematosus including a broad spectrum of clinically and histopathologically distinct types of lesions. Among them the verrucous or hypertrophic type represents an uncommon cutaneous manifestation resembling clinically as well as histopathologically keratoacanthoma or verrucous lichen planus. The course of the disease is – in most but not in all instances — marked by chronicity, absence of regression and resistance to treatment. The diagnosis is based on clinicopathologic findings. We herein present a patient with a longstanding history of DLE with keratoacanthoma-like verrucous lesions on both forearms and hands.

KEY WORDS

discoid lupus erythematosus, verrucous lupus erythematosus, keratoacanthoma-like lupus erythematosus

INTRODUCTION

Verrucous lesions in patients with discoid lupus erythematosus were first mentioned in the literature by Bechet in 1940 (1). However, the clinical, histopathologic, immunofluorescent and ultrastructural features were studied in detail by Uitto et al. nearly 40 years later (2,3)

CASE REPORT

A 42-year-old man presented with several verrucous lesions on the dorsal aspects of both forearms and hands resembling keratoacanthomas.

Histopathology (Fig. 1 and Fig. 2) revealed irregular epidermal hyperplasia with hypergranulosis and compact orthohyperkeratosis. There was also a slight vacuolar degeneration of the basal layer and, in addition, few necrotic keratinocytes were present in the epidermis. In the upper dermis a rather dense lichenoid infiltrate of lymphocytes was observed. PAS staining showed a thickened basement membrane. The histopathologic differential diagnoses included keratoacanthoma, lichen ruber verrucosus, prurigo nodularis, verruca vulgaris, and verrucous lupus erythematosus.

Clinical correlation revealed that the patient had, besides the verrucous lesions on the hands (Fig. 3), also skin lesions on the face and neck typical for

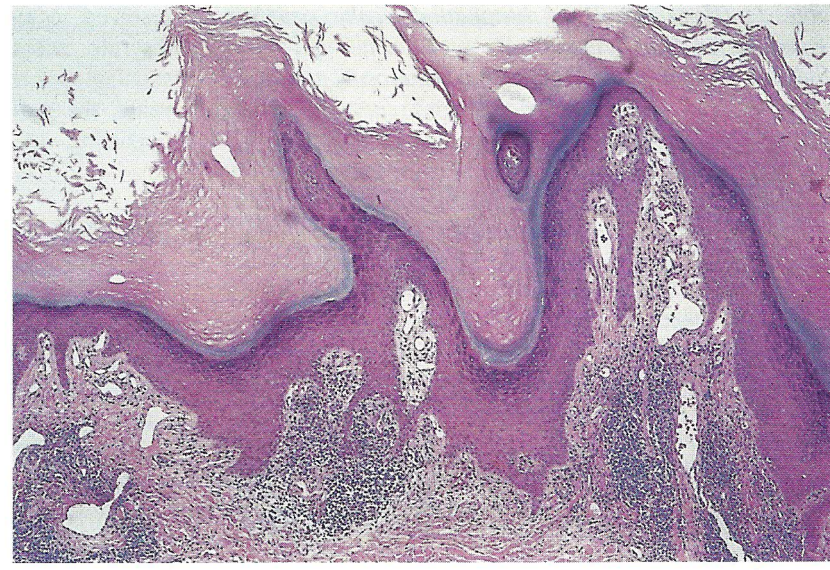

Fig. 1. This photomicrograph shows irregular epidermal hyperplasia, hypergranulosis with compact orthohyperkeratosis and a rather dense, lichenoid infiltrate of lymphocytes in the upper dermis. (Magnification 100x)

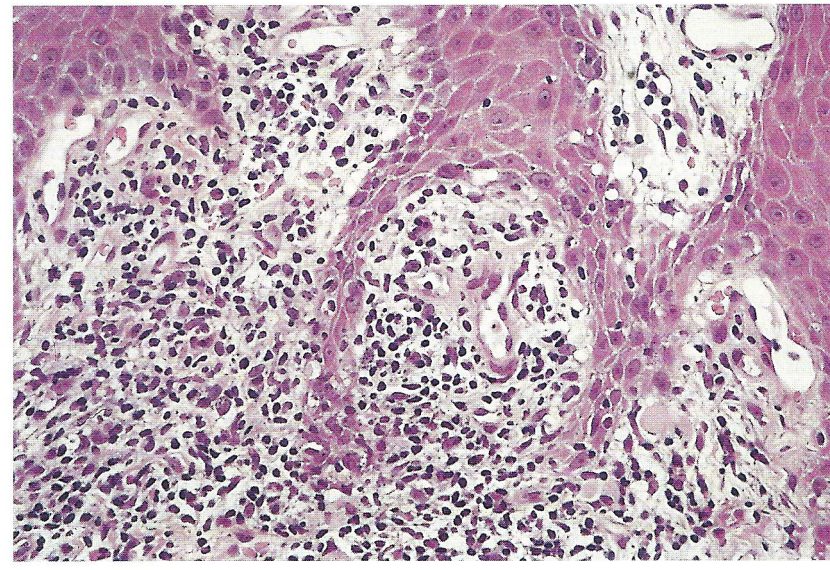

Fig. 2. Higher magnification reveals a predominantly lymphocytic infiltrate, a slight vacuolar degeneration of the basal layer and few necrotic keratinocytes. (Magnification 250x)

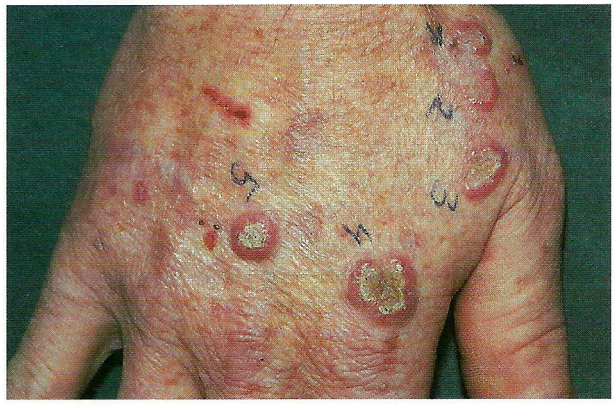

Fig. 3. Verrucous lesions on the dorsal aspect of the hand resembling multiple keratoacanthomas.

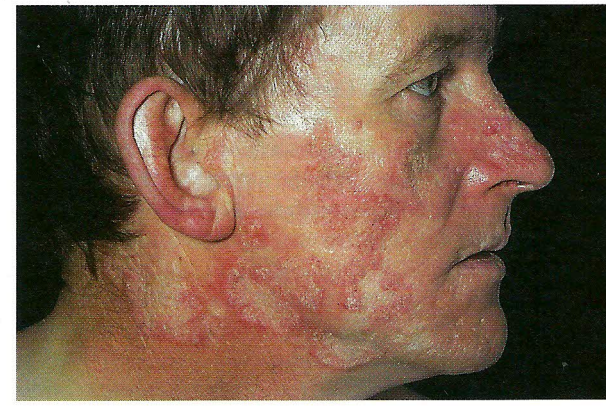

Fig. 4. Typical DLE lesions on the face and neck.

Table 1. Classification of histopathologically specific LE-associated skin lesions.
Modified from Dubois' Lupus erythematosus textbook (6)

Histopathologically specific LE-associated skin lesions
Acute cutaneous LE
Localized
Generalized
Subacute cutaneous LE
Annular
Papulosquamous
Chronic cutaneous LE
"Classical" DLE
Localized
Generalized
Hypertrophic (verrucous) DLE
Lupus panniculitis (profundus)
Mucosal LE
Lupus tumidus
Chilblains lupus
DLE-lichen planus overlap

DLE (Fig. 4). Histopatologic examination of the facial lesions as well as direct immunofluorescence studies confirmed the clinical diagnosis of DLE.

All clinical investigations and examinations of internal organs and organ systems were within normal limits. Laboratory data, ANA and ANA subsets, anti-ds DNA, as well as Ro/SS-A and La/SS-B, were negative. On the basis of clinical, histopathologic and immunofluorescence findings the diagnosis of verrucous or keratoacantoma-like lupus erythematosus was established.

The patient was treated with antimalarics and topical applications of steroid ointments on the facial lesions. The keratoacanthoma-like lesions on the hands were treated with intralesional injections of 0,1% triamcinolone acetonide. Following treatment, the verrucous lesions on the dorsa of the hands resolved completely within a few months leaving discrete scars. Also, the facial lesions regressed satisfactory. However, after a follow-up period of 12 years, the patient still has some erythematous lesions persisting on the face and neck.

DISCUSSION

Lupus erythematosus may affect the skin in several different, heterogeneous forms. According to the late J.N. Gilliam (4,5) the LE-associated skin lesions could be divided on the histopathologic basis into two main categories: histopathologically specific (LE-specific) and histopathologically non-specific (LE-nonspecific) skin lesions. LE-nonspecific skin diseases are those which are quite common in patients with LE, but are not characteristic for LE itself (e.g.: leucocytoclastic vasculitis, urticaria, and papulonodular mucinosis). The histopathology of LE-specific skin lesions is, however, more characteristic and pathognomonic for the disease.

Three categories of LE-specific skin diseases have been described (Table 1), namely, acute, subacute and chronic cutaneous LE. Chronic cutaneous LE comprises several distinct forms and verrucous or hypertrophic lupus erythematosus is one of them. (6).

Verrucous lupus erythematosus is a rare variant of chronic cutaneous LE that is characterized by verrucous lesions resembling keratoacanthomas, hypertrophic lichen planus, or prurigo nodularis (2,3,7). Lesions are most common on the dorsa of the hands, but may be present also on other sites (extensor aspects of the arms, upper back and face) (2,7). Usually, besides the verrucous lesions the patients have also typical discoid lupus erythematosus of the face. Epidermal hyperplasia (sometimes even cup-shaped with a central crater filled with keratin), hypergranulosis, orthohyperkeratosis and a dense lichenoid infiltrate are characteristic for the histopathology of verrrucous LE. All these features were also present in our patient.

In conclusion, verrucous lesions in a patient with otherwise typical DLE represent nothing but a distinct variant within the broad spectrum of cutaneous expressions of lupus erythematosus.

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