Case report

BULLOUS SYSTEMIC LUPUS ERYTHEMATOSUS WITH MALIGNANT PAPULOSIS ATROPHICANS AND SCHÖNLEIN-HENOCH VASCULITIS SYMPTOMS

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ABSTRACT

A 36-year old female patient suffering from systemic lupus erythematosus (SLE) with SLE specific vesiculo-bullous lesions together with symptoms of malignant papulosis atrophicans Degos and of Schönlein-Henoch purpura is reported. Such a combination of symptoms seems to be extremely rare and probably has not been described in the literature. The fact that the papulosis atrophicans Degos lesions were benign and that the treatment of the vesiculo-bullous lesions with sulphons was effective deserves to be mentioned.

KEY WORDS

systemic lupus erythematosus, vesiculo-bullous lesions, papulosis atrophicans maligna symptoms, purpura Schönlein, case report

INTRODUCTION

Dermatological symptoms can be observed in about two-thirds of all cases of systemic lupus erythematosus (SLE). Vesiculo-bullous lesions are considered however to be rare. Several non-specific vascular lesions connected with the basic illness may also occur: blood vessels of the dermis as well as vessels of larger diameter may be affected (1). A rare case of a patient with SLE who later developed vesiculo-bullous symptoms, dermal infarcts of the papulosis atrophicans Degos type as well as Schönlein-Henoch purpura is described. The authors are not aware that such a case was published in the literature.

CASE REPORT

A 36-year old patient had a history of toxaemia during pregnancy in 1974, later on clinical and laboratory findings revealed a SLE. She became symptom-free following a treatment with Prednisolon and Immuran and remained so for two years after this medication. In the summer 1980 she developed vesiculo-bullous symptoms and an atypical bullous pemphigoid was diagnosed. Remissions followed repeated treatment for some years, however in summer 1987 in spite of ongoing treatment the patient developed a new blisterous eruption.

On admission she was in a good general condition. On the

hairy part of scalp, on the face, on the upper parts of the limbs and around the knees solitary and grouped tense vesicles and bullae containing serum were expressed on an erythematous urticaria-like skin (Fig. 1). Some scattered papules with a diameter of 2-4 mm as well as slightly depressed scarred lesions of a china-white colour surrounded by a teleangiectatic erythema were also present (Fig. 2). These symptoms were present for some years.

Results of routine laboratory studies were negative. There was a slightly expressed cryoproteinemia. a positive Latex test, positive test for LE cells, positive anti-SSA/Ro and anti-SSB/La, essentially decreased C_3 fraction of complement (0.57%), alpha 2 and beta globulins were slightly raised.

Histologic examination of a blisterous lesion revealed a slight spongiosis and intracellular edema of the epidermis, a moderate edema of the upper part of the dermis with a chronic inflammatory infiltrate around the blood vessels; in the papillae neutrofils were present as well as formation of microvesicles. In the vesicles some neutrofils, a few mononuclear cells and eosinophyls could be seen (a histology corresponding to dermatitis herpetiformis Duhring).

Histologic examination of the atrophic scarred papulae: Below the hyper keratotic, atrophic epidermis an eosinophylic homogeneus fibrotic tissue with only a few cells was found in place of normal collagen fibres. The blood vessels displayed thick hyalinized walls. By toluidin blue a small quantity of acid mucopolysaccharide could be detected at the edge of lesions. Around the blood vessels a mononuclear infiltrate could be observed. Such histology was consistent with the late stage of a Degos papule (Fig. 3).

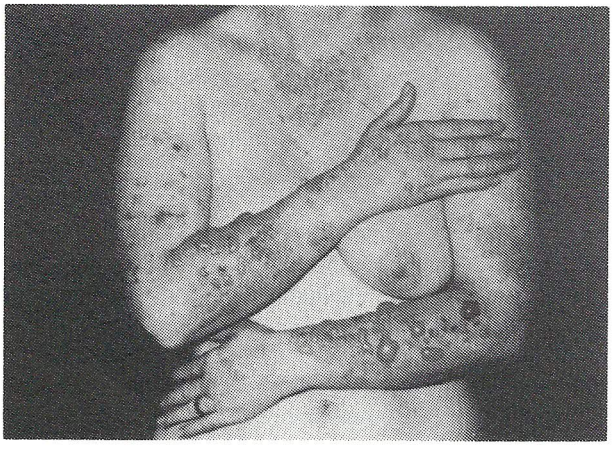

Figure 1.
Vesiculo-bullous eruption on an erythematous basis

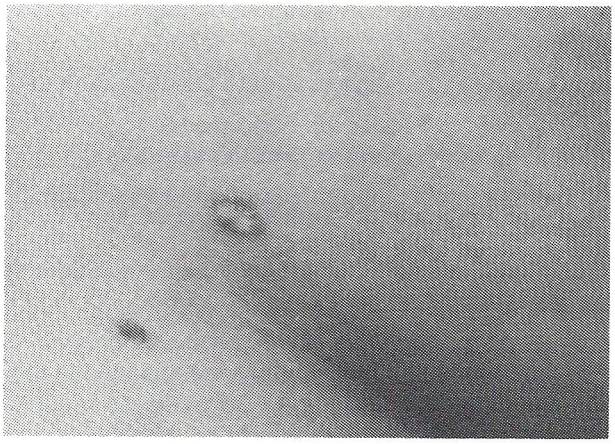

Figure 2.
A china-white slightly depressed lesion with an erythematous halo and brownish-red papule (Degos papule)

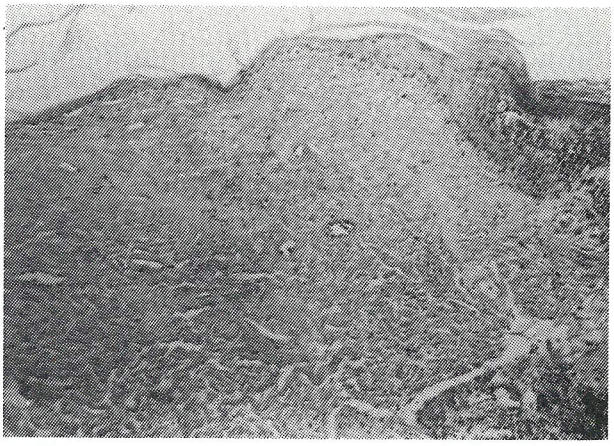

Figure 3.
Atrophic epithelium, fibrosis of the papillary dermis with only a few cells, some thick walled blood vessels.

Direct immunofluorescence of skin adjacent to a blister revealed continues fine fibrillar to granular deposits, consisting mainly of IgA, IgG and C3. By direct immunofluorescence of a Degos papule a pronounced deposit of immune-complexes of mainly granular type were detected along the basal membrane and in the small papillary blood vessels. The deposits were composed predominantly of IgM, but IgA, IgG and C_3 were also present.

By indirect immunofluorescence antinuclear antibodies at a low titer were detected.

Follow up. Dramatic improvement occurred 24-48 hours after the previous treatment (2 tablets of Prednisolon and 1 tablet of Immuran every second day) had been supplemented

by 2x50 mg of sulphon daily. The patient became symptom-free in a few days and remained so for 10 months observing the same treatment. She had to be readmitted because of methemoglobinemia and when sulphon was discontinued a serious recurrence of bullous lesions was noted. As steroids in a daily dosis of 60 mg and 100 mg Immuran daily were reintroduced her condition improved. During the process of lowering the dosis of steroids palpable purpurae appeared on the lower limbs (Fig. 4). Histology confirmed a

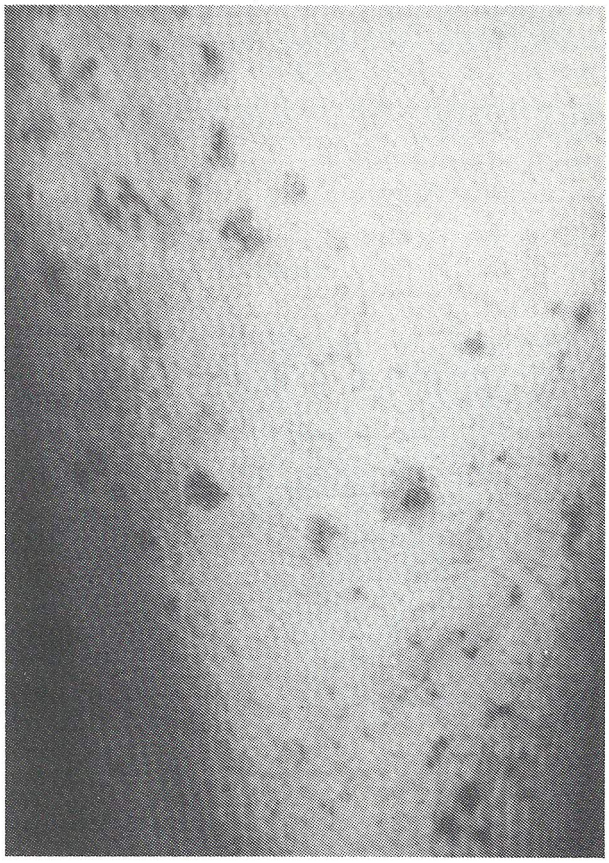

Figure 4.
Irregulary shaped purpuric lesions on the calf

leukocytoclastic vasculitis with extravasation of erythrocytes. After the introduction of plasmapheresis (altogether 4700 ml) the purpurae regressed.

On maintaining dose of 50 mg Immuran and 25 mg sulphon the patient remained symptom free for two years.

DISCUSSION

The vesiculo-bullous lesions in SLE have been described earlier (2, 3). Such symptoms partly correspond to SLE and partly to other bullous dermatoses (4-6). The differential diagnosis between the blisterous SLE and the bullous pemphigoid or dermatitis herpetiformis presents a serious difficulty. The criteria for a differentiation were defined by Hall et al and Olanski et al by applying immunohistology and immunoelectronmicroscopy (7-9). So far 33 cases of bullous SLE were published.

Bullous SLE in the majority of cases imitates dermatitis herpetiformis and more rarely bullous pemphigoid (10-13). The histology corresponds to dermatitis herpetiformis, but in some cases a leucocytoclastic vasculitis in the upper dermis and an accumulation of neutrophils at the base of the blisters without a leukocytoclasia were described (9, 12, 13). By immunohistology deposits of globulins at the dermo-epidermal junction were demonstrated in all instances (7, 8, 11, 12, 13); they were granular in half of all cases, while linear in the other half (9). Attention has to be paid to the high ratio of IgA in the deposits (up to 67%). It is accepted that the IgA play a role in the formation of blisters and in the activity of the disease (7, 9, 12, 13). Indirect immunofluorescence is usually negative in bullous SLE (12, 13). Circulating anti-basal membrane antibodies were found in 5 patients out of 33 (10, 11). At present the immunoelectronmicroscopy is considered to be the most reliable method: in bullous pemphigoid the deposits can be localized in the lamina lucida, above the basal lamina and in SLE in the uppermost region of the dermis (8, 10).

The diagnostic criteria for bullous SLE can be summarized according to Camisa with slight modifications as follows:

1. The diagnosis of SLE set up on the basis of the ARA criteria.

2. Vesiculo-bullous eruptions are not limited to areas exposed to light.

3. Histology corresponds to dermatitis herpetiformis.

4. Direct immunofluorescence displays a basal membrane-zone reaction.

5. By immunoelectronmicroscopy immune reactants are localized below the basal lamina (12, 13).

Besides the SLE specific skin symptoms, the non-specific which indicate the activity of the disease should be considered: thrombophlebitis, Raynaud's syndrome, livedo-vasculitis, chronic ulcers, digital gangrenes, dermal vasculitis (urticaria

vasculitis, leukocytoclastic vasculitis), Degos-like dermal infarcts, vascular lesions leading to atrophie blanche, palmoplantar erythema, various teleangiectasias (2, 3, 14, 15, 16, 17).

The incidence of lesions corresponding to malignant papulosis atrophicans in SLE is rare. This vaso-occlusive disease is usually of a systemic character and can be recognized by papulae surrounded by a china-white teleangiectatic fringe (3, 18). Malignant papulosis atrophicans can be differentiated into: 1. systemic type (Degos disease); 2. symptomatic form which usually accompanies an autoimmune disease; 3. hereditary type which indicates a probable genetic background; 4. benign cutaneous form which, even if present for years, affects only the skin (19, 20). This is why it is still more appropriate to use the technical term "Degos syndrome" even today.

The symptomatic form was described in three cases of SLE while in two cases it was accompanying progressive sclerosis and polymyositis respectively (21-23). In these patients - except for one in which the central nervous system was affected - the characteristic polyorganotropic manifestations were not observed even after the disease being present for several years. The same occurrence of malignant-type papulosis atrophicans did not affect the course of the basic disease. Immunologically in the cases accompanying SLE both in the affected and in the normal skin at the dermo-epidermal junction linear deposits of IgM and complement as well as a granular fluorescence composed of IgM, IgG and C-3 were observed (21-24).

The Schönlein-Henoch vasculitis (3, 25) occurring in the form of palpable purpurae is rather rare in SLE. It usually indicates systemic (central involvement, central nervous system, kidney). Lately in the cases of patients suffering from Schönlein-Henoch purpurae accompanying SLE an increased number of cardiolipin antibodies and ANA-positivity have been described (25). It is likely that Schönlein-Henoch purpura is a heterogeneous disease, in which various immune complexes - in the formation of which perhaps the antiocardiolipin antibodies take part provoke the leukocytoclasic vasculitis.

In our own case the vesiculo-bullous symptoms are regarded as SLE-specific symptoms even without electronmicroscopic examinations, on the basis of photosensitivity, the Duhring-like clinical, histological and direct immune histological findings (granular deposition along the basal membrane), the SLE-specific laboratory findings and the dramatic improvement following sulphon treatment.

In the course of the disease further dermal vasculitis symptoms - rare in SLE - were observed: continuously occurring malignant papulosis atrophicans lesions and, on one occasion, palpable purpurae on the lower limbs. The view of various authors that in the case of histologically typical "papulosis atrophicans maligna" accompanying the autoimmune disease the diagnosis of Degos disease should not be made, can be confirmed, since the extra-cutaneous symptoms are lacking. In such a case the Degos papulae can be considered as a symptom of SLE (21, 22). This is confirmed by the perilesional skin and the similar immune deposit composition and pattern of the Degos papulae. Simultaneous incidence of vesiculo-bullous SLE and Degos papulae has not yet been published, as far as we know. The occurrence of Schönlein-Henoch type vasculitis, can be considered as a further rare symptom of SLE. In the case of vesiculo-bullous SLE with mainly IgA immune deposits routine treatment should be supplemented by sulphons, in the case of Schönlein-Henoch type symptoms, by plasmapheresis.

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