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THE NATURE OF DISCOID LUPUS ERYTHEMATOSUS

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Abstract. The nature of discoid lupus erythematosus (DLE) and its relationship with the systemic form of the disease (SLE) were studied in 37 cases with SLE. Ten out of the 37 patients had cutaneous lesions which were morphologically and histologically diagnostic of DLE. In 8 cases the skin lesion preceded the systemic manifestations by $2\,^1/_2$ to 28 years. In one case both occurred simultaneously and in another the DLE developed during a fatal course of SLE. This observation is in conformity with the concept that DLE and SLE are one and the same disease.

The exact nature of discoid lupus erythematosus (DLE) and its relationship with the systemic form of the disease (SLE) is still not clear. Burch & Rowel (4, 5) think that there are genetic differences between them. Others believe that the two forms are of different nosology (2). There is much evidence that DLE and SLE represent different ends of one spectrum (9, 16). Support for the latter hypothesis is the occasional change from one form to the other (8, 10, 15). Kaposi in the year 1872 suggested that DLE and SLE are one and the same disease (11). This view is now becoming more and more accepted.

In this paper we will present the problem through the study of cases with SLE which we have seen.

REPORT OF CASES

37 cases with SLE were studied. Their diagnosis was based upon clinical symptoms, laboratory investigations and histological examination of the skin. The diagnosis in some cases was further confirmed by post mortem examination.

There were 33 females and 4 males. Their average age was 37.9 years. The youngest was a 12-year-old girl, who died within a year of the onset of her disease and the oldest was a male aged 37. Fourteen patients were born in this country, nineteen in Europe and four in Eastern countries. The various manifestations of the disease in the skin were as follows:

Skin manifestation	*No. of cases*
Discoid lesion	10
Erythema on the butterfly area	11
Allergic conditions	7
Pyogenic infection	4
Purpuric lesion	9
Monilial infection	4
Alopecia	3
Depigmentation	2
Generalized pruritus	2

Generalized exfoliative dermatitis, morbiliform rash, scarlatiniform eruption, ulceration and lichen planus like lesions were met with in one case each. Only in two out of the 37 cases was the disease not presented in the skin.

The following 3 cases are examples of the relationship between the discoid and the systemic form of the disease:

Case 1

A married woman aged 23. Her disease started at the age of 12 years and presented with a DLE patch on the face. She was under continuous medical care. No evidence for a systemic manifestation was revealed and the repeated laboratory investigations were always reported as normal. Treatment with chloroquine and topical applications were said to have improved her skin disease. Two months before admission to our department following an uncomplicated pregnancy and normal delivery, the patient developed for the first time arathralgia and bouts of fever. The physical and laboratory examinations revealed the following pathologies. Temp. 38°C; An old standing DLE patch on the face which was confirmed by a histological examination; Few erythematous macules on the dorsum of the fingers; Moderate enlargement of the spleen. The erythrocyte sedimentation rate (BSR) 124/126 (Westergreen), Haemoglobin 9.0 gm%, W.B.C. 9000 per mm^3 37% of which were lymphocytic cells. An obvious increase in the gamma segment of globulin. Presence of L.E. cells in the peripheral blood.

The diagnosis of acute SLE was made. The patient received prednisone and was discharged improved in order to continue the treatment under the supervision of her physician. Following an abortion during the second month of her second pregnancy, her symptoms re-occurred, she developed ileus and died in hospital in a

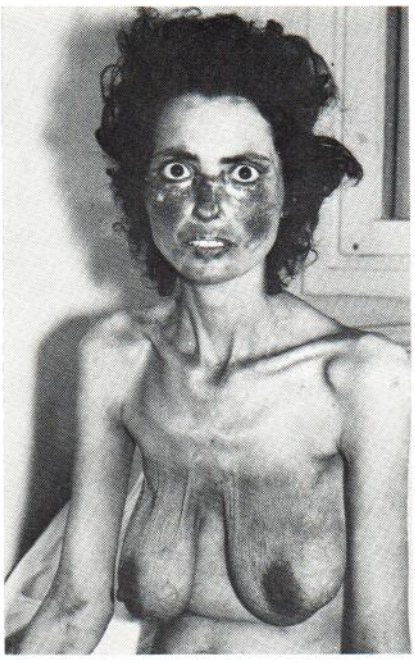

Fig. 1. A cachectic patient with marked pigmentation of the skin. Note the scars of an old standing DLE on the butterfly area of the face.

cachectic state. Prior to her death, the skin manifestations were presented with vast pigmented areas (Fig. 1). Post mortem examination confirmed the diagnosis of SLE.

Case 2

The patient was a 19-year-old girl when first admitted in June 1958. She presented with an allergic rash, probably due to penicillin. Apart from the rash there had been nothing seen on physical or laboratory examinations. Antihistamine treatment was given and the patient was discharged.

In May 1959 we again admitted the patient. She stated that she had been healthy during the past year, no fever, no joints pain or skin rashes. However, on the butterfly area of her face an erythematous patch developed two months earlier. The patient looked apparently healthy, body temperature was normal, but the physical examination revealed the following pathologies: On the butterfly area of the face a scaly erythematous brownish plaque covered with scars and also keratotic and telangiectatic lesions. A biopsy examination confirmed the diagnosis of DLE. Also there were few palpable glands in the axillary regions and the spleen was enlarged by 2 cm. Erythrocyte sedimentation rate was 89/105 (Westergreen). W.B.C. 3250 per mm^3 43% of them were lymphocytic cells. Thrombocyte count was 100,000 per mm^3. There was a reverse of the Albumin/Globulin ratio and a large number of L.E. cells was revealed in the peripheral blood.

Thus, subjectively her disease was latent and then presented with a plaque indicative of DLE. Later only the patient developed bouts of fever, arathralgic pain and skin rashes. Corticosteroid therapy was started and in the last 8 years the patient has had complete remission of the disease. She is married and a mother of 2 healthy children.

Case 3

A woman aged 49 was referred because of arathralgic pain, swelling of the joints and fever of 4 months duration. A few years before that, the patient had similar symptoms which were said to have been cured by corticosteroid therapy. Three months before admission she noticed that she had few red patches on her chest and one on her skull.

On admission: she looked ill, cachectic and unable to move in bed because of pain in her joints. The dermatological manifestations were: few erythematous scaly patches on the V area of the chest. The boundaries were raised and sharp and in the centre was a scar which was depressed. Keratotic and telangiectatic lesions were noted on the patches (Fig. 2). A similar patch with marked cicatrization was seen on the temporal region. The histological examination of a biopsy specimen was diagnostic of DLE. Steroid and antibiotic therapy were of no avail. The patient's condition deteriorated rapidly, she developed renal failure and pulmonary edema and died 12 days after admission. The laboratory investigations before her death and the post mortem examination revealed findings diagnostic of SLE.

DISCUSSION

Our cases demonstrate clearly the relationship of the systemic form of lupus erythematosus with its chronic discoid type. Ten out of the 37 described cases with SLE presented a skin lesion which was morphologically and histologically diagnostic of DLE. In 8 cases, as in case no. 1, the cutaneous lesion preceded the systemic involvement by $2\,^1/_2$ to 28 years. In case no. 2, both types of the dis-

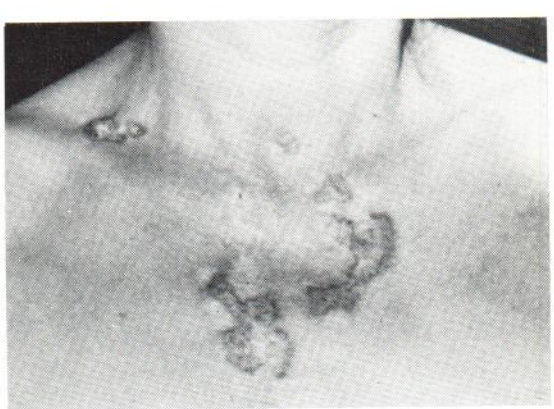

Fig. 2. Few sharply defined erythematous plaques on the exposed area of the chest showing telangiectasis and scars.

ease appeared simultaneously. In case no. 3, the discoid lesion occurred during a fatal course of the systemic disease.

According to Csermely (6), only the eccentric superficial type of the chronic lupus erythematosus may show a clinical continuity with the systemic form, whereas the fixed discoid type may show exacerbation but is almost never transformed into true acute form. This concept has not been borne out by our cases and the chronic fixed discoid type did show continuity with a true acute form of the disease. Moreover, we were unable to make clear histological distinction between the different types of the chronic skin lesions of lupus erythematosus. It becomes evident that the diagnosis of DLE is based mainly upon a morphological appearance of a skin lesion in patients who do not reveal clinical and laboratory evidence of a systemic involvement. The cutaneous pathology of DLE and SLE is quite similar and is of little help in differentiating between the two forms. Those accepted minor histological differences are quantitative rather than qualitative in nature. It is said that DLE reveals more marked hyperkeratosis, keratotic plugging and edema of the upper dermis. Prunieras & Montgomery (13) consider these relative differences as related more to the age of the lesion than to the clinical type of the disease. It can be assumed, therefore, that unless full hematological and immunological investigations are performed, patients with latent systemic involvement may appear to have uncomplicated lupus erythematosus (1). In fact, such was the case in our second patient.

The advance in technique is revealing more and more hematological and immunological disturbances in cases of DLE. Recent reviews of cases with DLE are revealing various incidences of intermediate and transitional forms of the disease. Shrunk & Doniach (14), in a study of 80 patients with DLE found that 14 had manifestations mid-way between the cutaneous and the systemic form and half of the patients had positive Fluorescent Antibody Test. The latter test was also reported positive in 28 out of another 34 eases with DLE (12). Dubois & Mastel (7) found in 62% of their patients with DLE, symptoms reminding one of a systemic involvement. Antibodies for different tissues of the body have been demonstrated in DLE lesions (3). We, therefore, think that it is justified to conclude that the entire classification of lupus erythematosus is arbitrary. A cutaneous lesion in this disease should always be suspected as being indicative of a disease more serious than DLE is considered to be.

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