

# LUPUS ERYTHEMATOSUS PROFUNDUS (KAPOSI-IRGANG)

## REPORT OF A CASE INCLUDING A COMPARATIVE STUDY OF THE HISTOPATHOLOGY WITH THAT OF CHRONIC DISCOID LUPUS ERYTHEMATOSUS\*

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The purpose of this report is twofold: to describe a case in support of the contention that there is such an entity as lupus erythematosus profundus, and to present a comparative histologic study of this condition with the classical forms of the disease. A special survey of the microscopic findings in the cutis and subcutis in chronic discoid lupus erythematosus was made to shed light, if possible, on the relationship of lupus erythematosus profundus to the latter form.

Pautrier (1) has taken the view that there is no reason to assume that a variant such as lupus erythematosus profundus occurs and that cases so described by Irgang (2) and others are in reality examples of the coexistence of lupus erythematosus of the face and scalp with subcutaneous tuberculides or sarcoids in other parts of the integument. The concept of lupus erythematosus profundus as a definable dermatosis, on the other hand has many adherents (3-7), a view which we share on the basis of the present study and a review of the literature.

The striking involvement of the deep cutis and the subcutis in the case we are about to describe led to an inquiry as to the status of the deeper layers of the corium and the hypoderm in classical instances of lupus erythematosus. A review of the literature on the histopathology of chronic discoid lupus erythematosus (8, 9) and a perusal of the description of the microscopic findings in a number of standard dermatologic texts, disclosed that either the deep cutis and subcutis had not been examined or that the changes observed were not considered worthy of comment. Pautrier (1) noted that at most, as he put it, the pathologic process extended to the deepest portions of the cutis just "licking the fat" but that otherwise the fat was left undisturbed.

The microscopic changes in the deeper layers of the skin in acute lupus erythematosus, on the other hand, have received closer attention (10, 11). Lever (10) noted that the fat is often involved in the systemic form and that "it may show focal mucoid degeneration with reactive infiltration. The collagen bundles separating the fat lobules may be increased in thickness and show edema and fibrinoid degeneration similar to the corium".

In view of the sparsity of reference to the deep cutis and subcutis in chronic discoid lupus erythematosus, we reviewed the histologic specimens of unequivocal cases in our files with special attention to the deeper structures. Our findings and Lever's description will be used as a basis for the comparison of the histologic

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features of the classical forms with the atypical case presented in this study. Comparisons will also be drawn with the cases of lupus erythematosus profundus previously reported.

#### CASE REPORT

*History:* A. S., a white woman, aged 31, was first seen at the New York Skin and Cancer Unit of the University Hospital on April 17, 1952. She came because of an "inflamed" lesion of three years duration on the outer aspect of the left arm. The pertinent aspects of her earlier history were "sun-poisoning with blisters" in the summer of 1942 and recurrent episodes of fever since January 1949. She was hospitalized on three different occasions between January and April 1949 because of these febrile episodes which were associated with weakness and joint pains. An eruption, thought to be a "drug rash" accompanied the first elevation in temperature. While at the hospital, subcutaneous injections were administered in both arms, a fact which may or may not be significant. Investigation failed to disclose the cause of the fever, and on each occasion the diagnosis on discharge from the hospital was "fever of unknown origin."\*

The lesion on the left arm appeared in April, 1949 at the termination of the third febrile episode. The patient had no other complaints with the exception of "joint pains" in inclement weather. She was well enough to take care of her household and a family of four. In September 1951, approximately two and one-half years after the onset of the eruption, she gave birth to her third child without any apparent untoward effect on mother or offspring.

*Examination:* The patient appeared to be in good health. There were no objective changes in the fingers and wrists to account for the "joint pains." A deep indurated inflammatory mass measuring approximately six cm. in diameter, was found on the proximal third of the lateral aspect of the left arm. Closer inspection of the affected area disclosed an irregular plaque encircled by an ill-defined zone of dusky erythema. The surface of the plaque was somewhat depressed, except for the periphery, which was more infiltrated than the rest of the lesion. The hue was a violaceous red, except for a less vivid central portion where scales adhered to the surface. There was no visible evidence of follicular plugging and of scarring. Rather symmetrically distributed over the anterior and lateral aspect of both arms, were approximately 15 firm, subcutaneous nodules, hazel-nut in size. These were not visible and were demonstrable only on palpation. The overlying skin, which was freely movable, appeared normal. The scalp, face, torso and lower extremities were free of lesions.

*Laboratory Findings:* Repeated urinalyses and blood counts disclosed no abnormal findings. Two serologic tests for syphilis as well as the Coomb's test gave negative results. The total serum proteins measured 7.3 gms. % (normal 6-8 gms. %), albumin 4.6 gms. % (normal 4-5 gms. %) and globulin 2.7 gms. % (normal 2-3 gms. %). The sedimentation rate on three occasions varied between 16-22 mm./hr. (normal 15 mm./hr.—Westergren method). Repeated examinations of the peripheral blood for L.E. cells and L.E. factor gave negative results. Intradermal tests with serial dilutions of old tuberculin Koch (OTK) inclusive of a 1:10,000 dilution did not elicit any reaction. Two electrocardiographic studies within a period of six months showed, in leads AVL, V<sub>2</sub> and V<sub>3</sub>, ST segment and T wave changes which did not fit any diagnostic pattern.

Histologic examination of a specimen removed with an 8 mm. punch on 5/22/52 from the large plaque on the left arm revealed the following changes: The epidermis is, for the most part, thinned exhibiting short or absent rete pegs, Spotty liquefaction degeneration of the basal cell layer is evident. Distinct follicular plugging is present in one of the sections.

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\* We are indebted to the Jewish Hospital of Brooklyn for making these data available to us.

The corium presents a moderate patchy infiltrate which extends from the mid-portion to the subcutaneous fat septa. The infiltration is most marked in the deep corium and the subcutaneous fat. In these zones the reaction is present about the vessels, the lower two-thirds of the follicles and throughout the fat septa. In the latter areas the infiltration is more diffuse and is equal to that present in the deep corium. The infiltration consists of small round, wandering connective tissue cells and an occasional polymorphonuclear leucocyte. The vessels throughout, particularly those in the deep corium and fat septa, are thickened, their walls edematous, their intimal nuclei prominent. In some instances the lumen of the vessel is decidedly narrowed. Sweat glands are present in the deep corium, and while they present no noteworthy abnormalities, they are surrounded by a mild cellular infiltration similar in character to that previously described. In the upper corium the infiltration is sparse and diffuse and appears to bear little relationship to the vessels or adnexa. Basophilic strands are present throughout the corium and subcutaneous fat as a fine, lacy syncytium. The collagen bundles in the upper corium appear fragmented. While there is some edema of the connective tissue framework in the sub-epidermal zone, this is not a noteworthy feature in the remainder of the section. Some elastorrhexis is demonstrable with the van Weigert stain. Fibrinoid degeneration is not demonstrable either with the hematoxylin-eosin or with the Hotchkiss-McManus stain. There is no evidence of fat necrosis.

*Course:* The patient has been under observation up to the time of the present writing. Her only complaints have been "some aches and pains" in the wrist and finger joints in damp weather, and a transitory elevation in temperature, up to 100°F. in the late afternoon from time to time. In March 1954 she experienced an episode of fever, similar to that in 1949, associated with weakness, lasting about 10 days. The patient was admitted at this time to the University Hospital for further observation. The only change in her dermatologic status was a temporary augmentation of the inflammatory reaction in and around the large plaque on the left arm. The sedimentation rate became quite rapid, 87 mm./hr (Westergren) as compared with a previous high of 22 mm./hr. Tests for L.E. cells in the peripheral blood during the febrile phase yielded negative results.

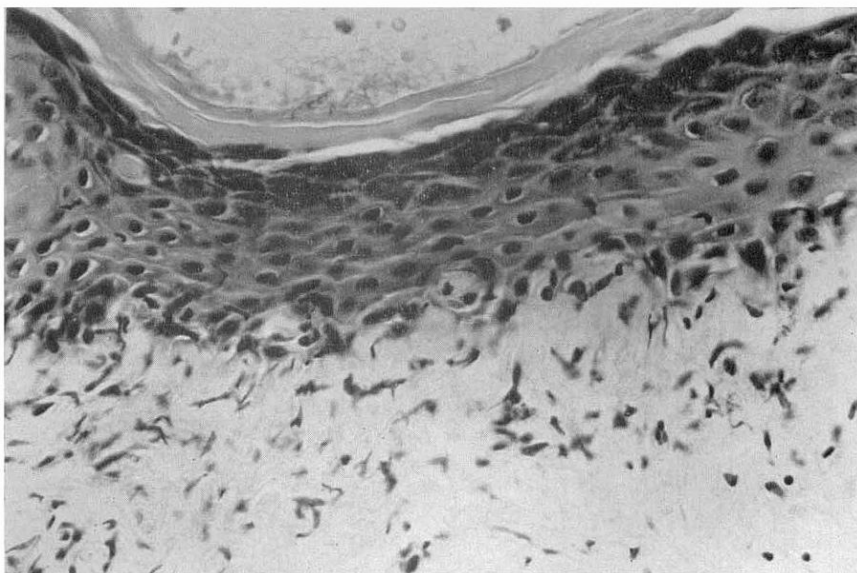


FIG. I. Hyperkeratinization, liquefaction of the basal cell layer and subepidermal edema are evident in the plaque over the left deltoid. 498X.

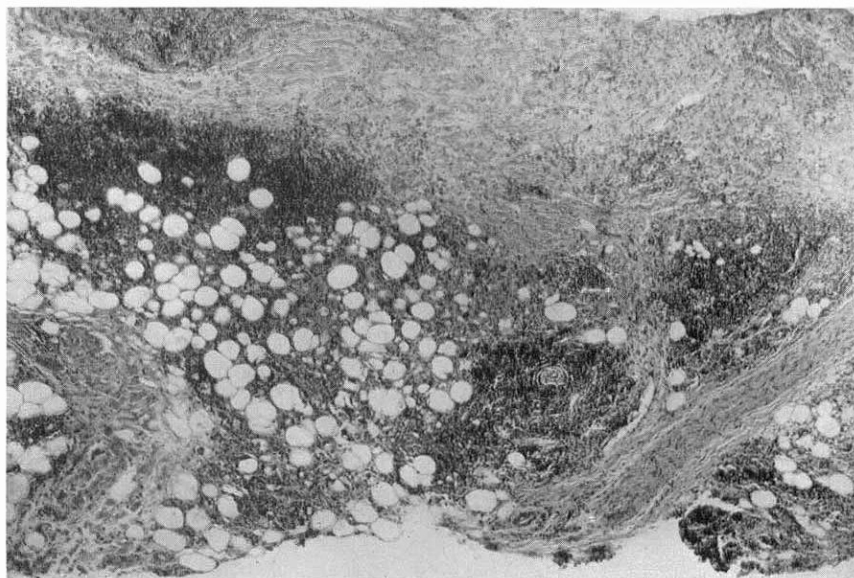


FIG. II. The inflammatory reaction in the deep corium and hypoderm is pronounced. Note the diffuse as well as the perivascular distribution of the cellular infiltrate. 65.4 $\times$ .

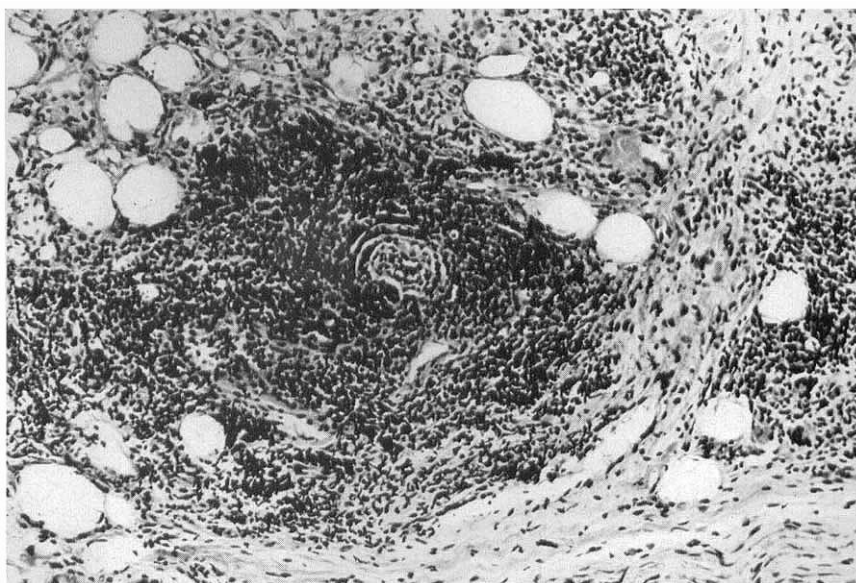


FIG. III. The vasculitis and perivasculitis are striking. The walls of the blood vessels show edema and proliferative changes with narrowing of the lumen. 185.5 $\times$ .

Resorption of the induration in and around the massive lesion on the left arm proceeded slowly and gradually except for the transitory exacerbation previously mentioned over a period of 18 months. By November 1953, there was little evidence of the deep-seated inflammatory reaction that was so striking early in 1952. As the induration waned, the clinical features became more and more typical of chronic discoid lupus erythematosus. At times there was more scaling and crusting than at others. In May 1955 a deep concave depression marked the site of the inflamed area. At the lower pole of this retracted scar, one could still make out the surface changes of lupus erythematosus.

The deep-seated nodules on the arms resorbed within a period of four months. Some left no trace, others healed leaving conspicuous concave depressions. The remainder, six in all, surfaced with the development of changes typical of discoid lupus erythematosus. Resolution of the deep-seated reaction in the large plaque as well as in the subcutaneous nodules was attributed to spontaneous evolution rather than to treatment. The patient took medication too irregularly for us to assume that this was of significant influence.

During November 1952, six months after the patient was first seen at the Skin & Cancer Unit, typical areas of chronic discoid lupus erythematosus appeared on the scalp. One of these extended to the back of the left ear and the other was near the vertex. Neither of these lesions has shown any progression or regression, in the last two and one-half years.

*Diagnosis:* The diagnosis of lupus erythematosus profundus was entertained because of the unusual degree of inflammation surrounding and underlying the plaque over the left deltoid and the presence of subcutaneous nodules on both arms. There was some question as to the nature of these deep-seated lesions before changes typical of chronic discoid lupus erythematosus appeared in the overlying skin. The possibility of a non-specific panniculitis or a foreign body reaction to the "injections" administered during the febrile episodes prior to admission, was also entertained.

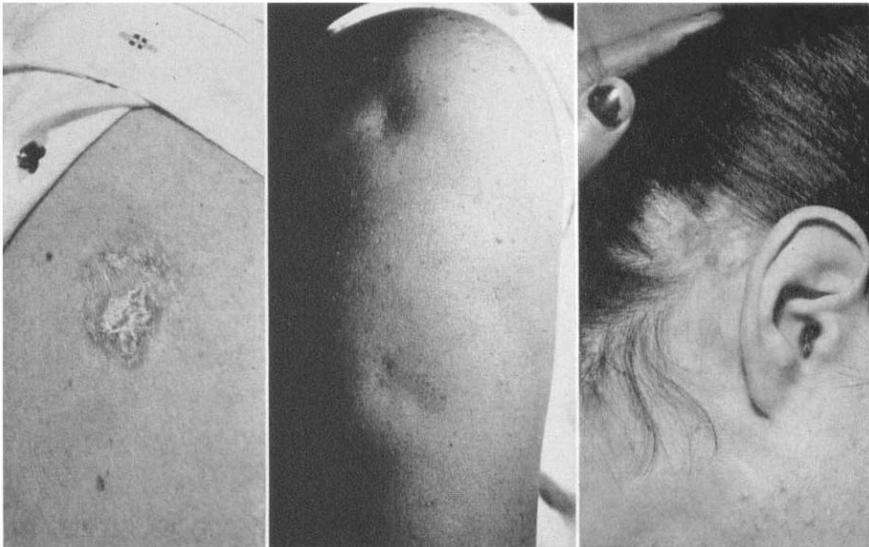


FIG. IV

FIG. V

FIG. VI

FIG. IV. The clinical features of the plaque on the left arm became more typical as the induration waned. Note retraction, crusting and scarring. Taken on 3/4/54.

FIG. V. Deep hollows mark the site of subcutaneous nodules. Taken 7/21/55

FIG. VI. A typical plaque of chronic discoid lupus erythematosus appeared in the mastoid area, six months after admission. Taken on 3/4/54.

HISTOLOGIC STUDY OF THE CUTIS AND SUBCUTIS IN  
CHRONIC DISCOID LUPUS ERYTHEMATOSUS

Two hundred and thirteen histologic specimens of chronic discoid lupus erythematosus were reviewed before 100 examples were found in which the biopsy was deep enough to include the subcutaneous fat. Specimens that did not satisfy the accepted criteria for the histologic diagnosis of chronic discoid lupus erythematosus were excluded from the study. We found that the dermo-hypodermal zone and the fat were spared in 72/100 specimens. In the remainder, 28/100 these layers were involved to a greater or lesser extent. The prominence of the histologic change was graded as sparse, mild or moderate, depending on the depth and degree of the reaction. A sparse reaction in the deep corium and fat was found in 20/28 cases; a "mild" reaction in 5/28 cases and a "moderate" reaction in 3/28 cases. In no instance was the reaction as pronounced as in the case cited in detail, although in the group evaluated as moderate, the reaction in one of the cases (see figs. XI and XII) approached but did not display quite the same degree of response.

A description and microphotographs illustrating the various degrees of histologic change found in the deep corium and fat in discoid lupus erythematosus follow:

*"Sparse" reaction in the deep corium and fat*

The epidermis is thin and mildly irregular with obliterated rete pegs and corresponding papillary bodies. Areas of basal cell layer liquefaction alternating with well-defined basal cell layers are noted as are dilated follicular ostia partly filled with horny plugs. A mild perivascular, perifollicular and periglandular cellular infiltration, which extends at one or two points into a few of the upper fat septa is noted. The latter are involved to a minimal degree, the greater part of the fat is not infiltrated. The infiltrate consists for the most part of small, round, and wandering connective tissue cells. The vessels throughout are dilated, their walls edematous, their intimal nuclei prominent. Proliferation of intimal nuclei is not noted. Sub-epidermal edema of the connective tissue framework is present, the collagen fibers form a lacy network in which scattered strands of basophilic collagen are seen. Collagenization to a mild degree is seen in the fat.

*"Mild" reaction in the deep corium and fat*

The epidermis is thin, the rete pegs and corresponding papillary bodies for the most part are obliterated. Some basal cell layer disturbance is present at scattered points. Several dilated follicular ostia partly filled with horny plugs are noted. A perifollicular, periglandular and perivascular focal and diffuse cellular reaction is present consisting in the main of small round and wandering connective tissue cells. The infiltration extends down and into the upper portion of the fat septa, adjacent to the deep corium and involves several fat septa. The vessel walls are mildly edematous, their intimal nuclei prominent but not proliferative. Mild collagenization of the fat lobules is observed.

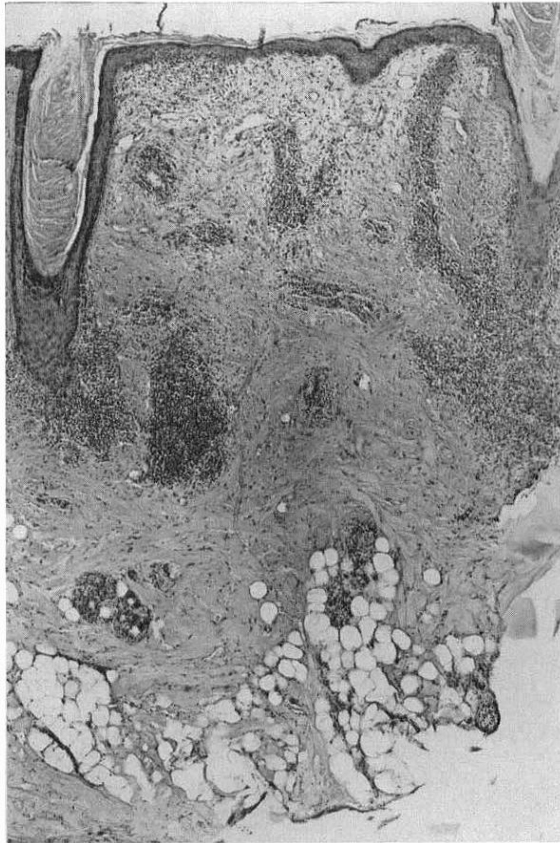


FIG. VII. Sparse reaction—collections of cellular infiltrate are seen in the upper fat septa in a typical example of chronic discoid lupus erythematosus. 63 $\times$ .

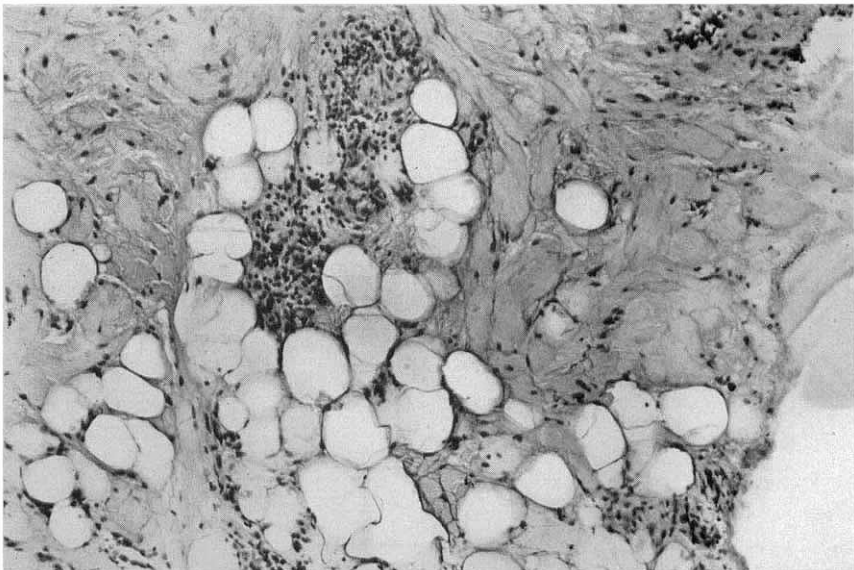


FIG. VIII. Sparse reaction—the cellular infiltrate consists principally of lymphocytes. 185.5 $\times$ .



FIG. IX. Mild reaction—the cellular infiltrate is more pronounced and involves a larger number of fat septa. 65.4 $\times$ .

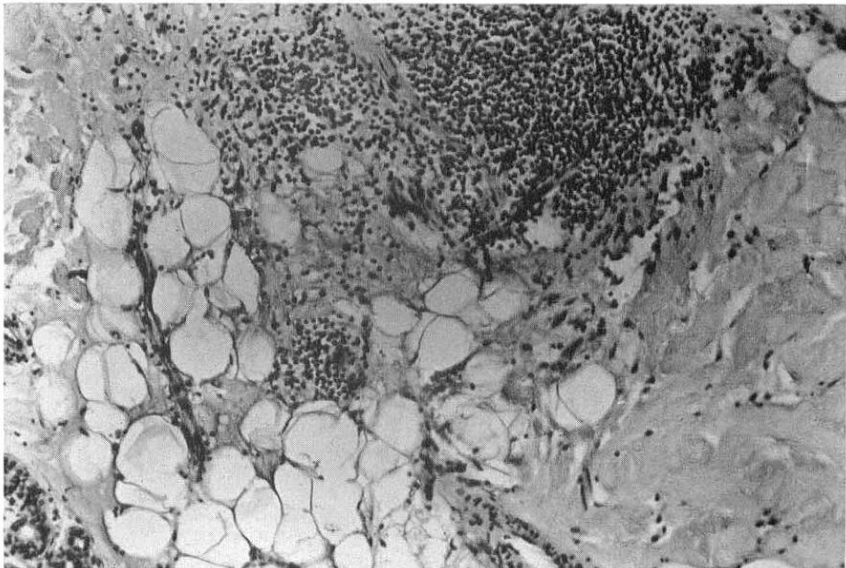


FIG. X. Mild reaction—collections of lymphocytes are distributed diffusely as well as in circumscribed areas in the fat. 185.5 $\times$ .





FIG. XI. Moderate reaction—the epidermis is verrucous, hyperkeratotic and acanthotic. The cellular reaction involves the fat septa in many areas. 65.4X.

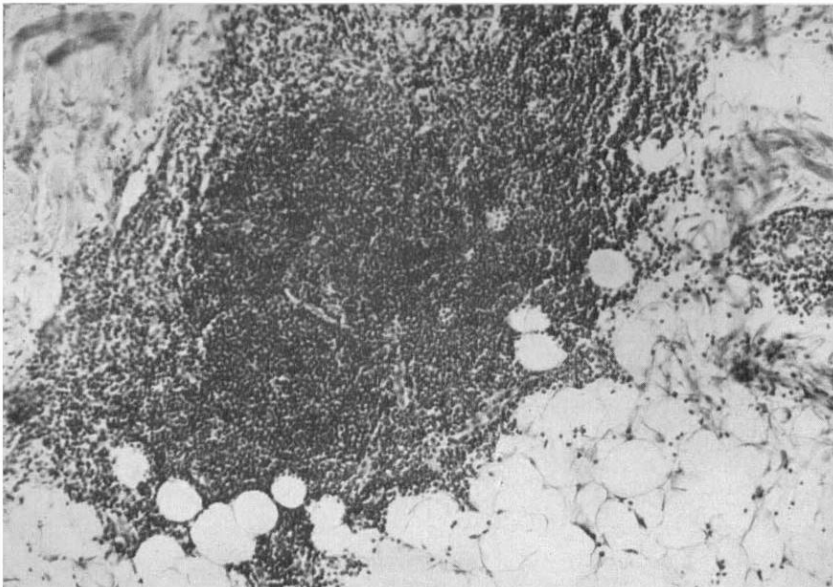


FIG. XII. Moderate reaction—the infiltrate consists of lymphocytes and wandering connective tissue cells. 185.5X.

*"Moderate" reaction in the deep corium and fat*

The epidermis is irregular, verrucous, hyperkeratotic and moderately acanthotic. Areas of basal cell layer liquefaction are seen at several points along the basal margin. A moderate perifollicular and periglandular infiltration is present in the upper, and mid parts of the deep corium, and extends in the fat septa in many areas forming several irregular cellular masses. The cellular reaction consists of small round and wandering connective tissue cells. The collagen bundles of the upper corium are edematous and lacy with scattered strands of basophilic degeneration. A very mild collagenization is seen in the fat. The vessel walls throughout, including those in the fat septa, are edematous, their intimal nuclei prominent but not proliferative.

## DISCUSSION

The case presented in this report disclosed an unusual combination of symptoms; namely, fever, joint pains, a deep-seated and indurated plaque of lupus erythematosus on the left arm, subcutaneous nodules of both arms and typical plaques of discoid lupus erythematosus of the scalp.

The first episode of fever antedated the appearance of the eruption by a period of three years and continued to recur intermittently after the eruption appeared. An exacerbation in the degree of the inflammatory reaction in the plaque on the left arm was noted during one of these febrile episodes.

During the first few weeks of observation the superficial changes overlying the plaque were suggestive rather than typical of discoid lupus erythematosus. As time went on however, the appearance of the surface became characteristic. Involution of the plaque was followed by the development of an unusually deep, concave atrophic and scarred area.

The subcutaneous nodules on both arms were demonstrable only on palpation, detached from the overlying skin. These did not run a uniform course. Some were resorbed leaving no visible trace, a number surfaced with the development of small discoid lesions and others healed leaving retracted scars.

It is interesting to speculate as to what role, if any, the "injections" played in the localization of the subcutaneous nodules as well as the deep-seated plaque on the left arm. One may hypothesize that the trauma of injection was in some measure responsible, in the nature of a modified Koebner phenomenon. On the other hand, as we shall see later, the arm is one of the areas of predilection in so-called lupus erythematosus profundus (Kaposi-Irgang).

Histologic examination of the principal plaque on the left arm disclosed that the deep cutis and subcutis as well as the epidermis and superficial layers of the cutis were involved. In addition to the features of discoid lupus erythematosus in the upper part of the section, there were striking non-specific changes in the deep corium and fat. Invasion of these layers with a pronounced cellular infiltrate made up chiefly of lymphocytes and inflammatory changes in the walls of the blood vessels were the essential features of the reaction. There was no evidence of fat necrosis, fibrinoid degeneration or collagenization. The histologic changes in

the deep corium and hypoderm conformed with those noted by other authors (2-4), in this aberrant form of lupus erythematosus.

The changes in the deep corium and subcutaneous fat were regarded as non-specific in character, simulating the histologic picture in certain forms of panniculitis, nodular vasculitis and erythema nodosum. Without the pathognomonic features of lupus erythematosus in the epidermis and upper half of the corium, one could not possibly have said on histologic grounds that the panniculitis was diagnostic of any particular entity.

Comparison of the histopathologic changes in this atypical case with those of discoid lupus erythematosus disclosed a distinct resemblance between the two. To make this comparative study possible, 100 specimens of discoid lupus erythematosus were reviewed with special attention to the corium and fat. Contrary to expectation, changes in the deep corium and fat were found in 28/100 or in slightly more than 1/4. The degree of involvement was graded as sparse in 20/28, mild in 5/28 and moderate in 3/28 in accordance with the intensity and depth of the inflammatory reaction. A cellular infiltration made up principally of lymphocytes in the fat septa and among the fat cells along with inflammatory changes in the walls of the blood vessels were the major features. These findings have been described in detail and illustrated (see Figs. VII-XII).

Descriptions of the corresponding clinical lesions and photographs, when available, were then analyzed for possible clues to these microscopic changes. There was no hint of any histologic involvement of the deep cutis and subcutis in those classed as sparse or mild. In those evaluated as moderate on the other hand, the infiltration was notable. Sarcoid and lupus vulgaris respectively were considered in the differential diagnosis in two and in the third, lupus erythematosus hypertrophicus was the provisional diagnosis. In the last case the intensity of the inflammatory reaction in the deep cutis and in the fat approached but did not equal that in case A.S. (see Figs. II, III, XI, XII).

Three pertinent facts were gleaned from the investigation of the status of the deep corium and subcutaneous fat in typical discoid lupus erythematosus. Firstly, these layers may be involved contrary to the experience of Pautrier (1) and contrary to what the dearth of reference in the literature would lead one to expect. Secondly, the inflammatory reaction is of a non-specific character and generally insufficient magnitude to be registered clinically. Thirdly, the differences between the histologic changes in the deep corium and fat in chronic discoid lupus erythematosus and in lupus erythematosus profundus may be only quantitative.

The diagnosis of lupus erythematosus profundus in our case was based on the deep-seated nature of the pathologic process in the prominent plaque on the left arm, the presence of subcutaneous nodules and the resemblance to other cases so designated in the literature. We found reason to hesitate, however, before adopting this terminology because of the divergent views in the literature on the nature of lupus erythematosus profundus and the doubt expressed by Pautrier that such an entity occurs.

Before attempting to deal with the pros and cons of the suitability of the term

lupus erythematosus profundus in this instance, perhaps we had better try to cope with Pautrier's skepticism.

Pautrier (1) expressed the opinion that the cases of so-called lupus erythematosus profundus previously reported were in reality examples of lupus erythematosus in co-existence with subcutaneous tuberculides or Darier-Roussy sarcoid. He stated moreover, that he had never encountered a case that could properly be called lupus erythematosus profundus, nor was he aware of any example in the experience of other dermatologists including Brocq. This eminent observer also felt that if the term lupus erythematosus profundus was used, it was as a histologic term to indicate infiltration reaching down to the cutaneous-subcutaneous border and not to designate a clinical form. Despite the plausibility of Pautrier's argument in Irgang's case (2) at least (active tuberculosis was a feature of the latter's case), one cannot dismiss the fact that the hypoderm may be conspicuously involved in lupus erythematosus and give rise to characteristic clinical features.

The sources of the term profundus quoted by various authors were read in the original. Brocq (12) used the term "lupus érythémateux profond" to contrast the deep-seated, indurated and usually asymmetrical nature of the fixed form of lupus erythematosus as compared with the more superficial form "l'érythème centrifuge typique". He also used "profond" as an adjective in describing a type of lesion which presumably was a tuberculoderm rather than an example of lupus erythematosus. No mention was made of subcutaneous nodules.

As far as we can determine, the first authors to use lupus erythematosus profundus as the designation for an unusual degree of induration and penetration of a fixed lesion of discoid lupus erythematosus were Oppenheim (13) and Scherber (14). The term is used similarly in at least one standard dermatologic text (15).

Irgang (2) it appears was the first to apply the term lupus erythematosus profundus for the combination of lupus erythematosus and subcutaneous nodules. Although there was no macroscopic evidence of lupus erythematosus in the skin over the subcutaneous lesion in his case, Irgang felt that the histologic changes in the epidermis were sufficiently characteristic to justify this diagnosis. Thus this author's use of profundus was again different from Brocq's or Oppenheim's. Although Kaposi (16) was the first to record the association of deep-seated doughy nodules and tender subcutaneous swellings with acute and subacute lupus erythematosus, he did not use the appellation lupus erythematosus profundus for these lesions. Incidentally, Kaposi also described the progressive transition of some of these subcutaneous lesions into typical plaques of lupus erythematosus.

Bechet (17, 18) used the term profundus in still another sense, namely to describe a verrucous, hypertrophic lesion that healed with cribriform scarring, located on the chin and around the mouth. Examination of the photomicrograph of his first case showed that the hypoderm was not involved. Unfortunately, the biopsy in the second case did not extend beyond the cutis. Peck (19) in turn applied Bechet's terminology, namely lupus erythematosus hypertrophicus et profundus for an example of discoid lupus erythematosus in which the subcu-

taneous infiltration was a striking feature. The consensus is that Bechet should not have linked "profundus" with his cases.

Thus we find that the term profundus has been associated with lupus erythematosus in at least four different ways: 1) for the fixed or discoid form of lupus erythematosus (Brocq) 2) for a discoid lesion exhibiting an *inordinate* degree of infiltration (Oppenheim, Scherber, Andrews); 3) for the combination of discoid lupus erythematosus and subcutaneous nodules (Irgang) and finally, 4) by Bechet for a rather unusual form of discoid lupus erythematosus which strictly speaking belongs to the category of lupus erythematosus hypertrophicus.

Irgang credited Brocq with having described the prototype for his case. Others (3, 4) were of the opinion that Kaposi's name, rather than Brocq's, should be associated with lupus erythematosus profundus because it was the former who first mentioned subcutaneous nodules. The trend in recent years has been to limit the term lupus erythematosus profundus (Kaposi-Irgang) (3-6) to the concurrence of subcutaneous nodules with lupus erythematosus either in situ or in some other part of the skin. In some instances the overlying superficial involvement was demonstrable only on histologic examination. For the sake of completeness, presumptive cases under various titles were reviewed and those that satisfied this criterion were included in the bibliography (20-24). Irgang's case, it should be pointed out, was accepted by the authors cited (3-6) despite the presence of active tuberculosis.

It appears equally appropriate to us to apply the term lupus erythematosus profundus to an indurated, deep-seated lesion (as exemplified by the large plaque over the deltoid in our case) in which there is unmistakable evidence of lupus erythematosus in the superficial layers and a pronounced inflammatory reaction in the deep cutis and hypoderm, whether or not subcutaneous nodules are also present. In the first place it is conceivable that such a lesion may precede the appearance of subcutaneous nodules, or may appear after the subcutaneous lesions have waned. Secondly, what if our patient had not come under observation until four months later, when the indurated plaque in the left arm was still evident and the subcutaneous nodules as such had disappeared. Would this case, which so closely resembles those described by Arnold (3), Costa and Janqueira (4), Silva and Portugal (5) have been less likely to be an instance of lupus erythematosus profundus? Perhaps one can resolve the problem of nomenclature by using the term lupus erythematosus profundus with eponyms for deep seated discoid lesions associated with subcutaneous nodules and lupus erythematosus profundus divorced from its eponyms in the absence of subcutaneous nodules.

It is not within the scope of this paper to discuss lupus erythematosus hypertrophicus to which category Bechet's cases probably belong. We wish to point out however that the deep corium and subcutaneous fat need not be exempt in such a case (see Fig. XI and XII). Of the three examples showing "moderate" involvement of the deeper structures, one was associated with the histologic changes in the epidermis and the clinical features consistent with the diagnosis of lupus erythematosus hypertrophicus. Consequently, lupus erythematosus hypertrophicus et profundus may not be such a misnomer after all. Nevertheless, the

latter must not be confused with lupus erythematosus profundus (Kaposi-Irgang).

Some have rejected lupus erythematosus profundus (Kaposi-Irgang) as the proper designation for like cases because of the disputations about "profundus". Grund (25) for example preferred "An Unusual Case of Chronic Lupus Erythematosus with Concomitant Subcutaneous Nodes and Swellings"; Facio and Pomposiello proposed lupus erythematosus sarcoidiforme (7) instead. We are more inclined to side with those (3-6) who have accepted the terminology lest "confusion be more confounded" by adding alternative appellations. Furthermore, it would not be difficult to find objections to these alternatives.

The dermatologic features of our case are of the same order as those described in recent years by other observers.\* A *sine qua non* for the diagnosis of lupus erythematosus profundus (Kaposi-Irgang) is the presence of typical lesions of discoid lupus erythematosus somewhere on the scalp or body alone or over the subcutaneous lesions. In the absence of such evidence, the characteristic histologic features of this disease in the skin overlying the nodule should be clearly demonstrable.

An analysis of the cases conforming to this criterion discloses that the subcutaneous nodules characteristically are distributed over the face, arms, buttocks or thighs. These lesions are firm, circumscribed, deep-seated, somewhat tender and of variable size, from hazel-nut to pigeon-egg. They may be resorbed without leaving any trace; they may extend to involve the overlying skin with the development of typical discoid lupus erythematosus or healing may be followed by a hollow or retracted scar of varying size and depth.

With few exceptions, this unusual form of lupus erythematosus has been limited to women between the ages of 22 and 45. Some of the subjects were free of general symptoms whereas others, like our own exhibited suggestive systemic manifestations. Examination for L.E. cells was mentioned only in one other report, namely by Silva and Portugal (5). These cells were demonstrable in one out of three examinations, the former during a febrile phase. Repeated tests in our patient, including a search during a period of fever, gave negative results.

The histologic features of lupus erythematosus profundus (Kaposi-Irgang) also require discussion. The changes noted in the deep cutis and subcutis by various authors fall in two categories: A) findings analogous to our case (2-4, 25) and B) alterations analogous to those seen in acute or subacute lupus erythematosus (5-7). We have already shown that the histologic findings in our case were similar to those found in chronic discoid lupus erythematosus and that the differences were quantitative rather than qualitative.

The essential features of group A were the presence of a predominantly lymphocytic infiltrate in the fat septa and among the fat cells, and pronounced vasculitis of the deep vessels of the skin. The changes were essentially proliferative and inflammatory. In group B the changes were primarily degenerative and

\* We are indebted to Dr. Orlando Canizares for calling the presentation of four additional cases by Arnold before the American Dermatologic Association in April '55 to our attention.

destructive. Fibrinoid degeneration and necrosis of the collagen in the deep cutis and hypoderm, obliterative and destructive changes in the vessel walls and collagenization of the fat were the striking features. The cellular infiltrate was on the whole scant, distributed principally about the foci of necrosis. The discrepancy in these findings may be a matter of the stage during which the histologic examinations were made or the natural variation in the severity of the same disease in different individuals or at different times or sites.

In neither group were the histologic changes in the deeper sections of the skin and hypoderm in our opinion sufficiently unique to make a histologic diagnosis of lupus erythematosus profundus possible, unless the pathognomonic features of lupus erythematosus were also present in the upper layers.

The absence of epithelioid cells in all reports merits emphasis in view of the association of the term sarcoid with these subcutaneous nodules in some reports.

#### SUMMARY AND CONCLUSIONS

An unusual case of lupus erythematosus has been presented to illustrate the fact that the hypoderm may be involved in this disease, giving rise to subcutaneous nodules and deep-seated indurated plaques with typical surface changes. This case supports those who contend that a variant such as lupus erythematosus profundus (Kaposi-Irgang) exists. The clinical and histologic features have been compared with similar cases previously reported and the terminology has been discussed in detail.

Although the subcutaneous nodules conform to a rather characteristic pattern, a definitive diagnosis of lupus erythematosus profundus (Kaposi-Irgang) on clinical grounds may be difficult unless a typical discoid plaque can be demonstrated in the scalp or in some part of the glabrous skin. Similarly a histologic diagnosis cannot be made with certainty unless the pathognomonic features of lupus erythematosus are demonstrable in the upper part of the section.

A study was made of the histopathologic findings in the deep cutis and hypoderm in one hundred biopsy specimens of typical discoid lesions of lupus erythematosus. Contrary to expectation, these layers were involved to some extent in 20/100 cases and to more than a negligible degree in 8/100 cases. The changes noted differed quantitatively rather than qualitatively from those described in our case as well as in other examples of lupus erythematosus profundus (Kaposi-Irgang).

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#### ADDENDUM

In a comprehensive study of the histology of lupus erythematosus, Ellis and Bundick noted that a deep infiltrate often occurs to the "bottom of the section" in the discoid type of lupus erythematosus. The hypoderm as such, however, was not mentioned. (ELLIS, F. A. AND BUNDICK, W. R.: *Histology of lupus erythematosus. Arch. Dermat. & Syph.*, **70**: 311-324, 1954.)