

DUHRING (L.A.)

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Dermatitis Herpetiformis

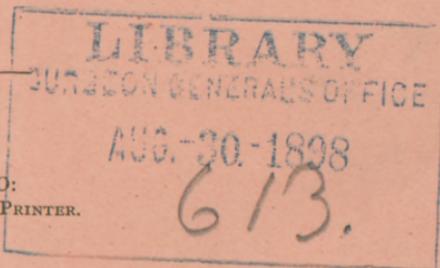
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DERMATITIS HERPETIFORMIS.

Under the name Dermatitis Herpetiformis I propose to place a number of cases of skin disease that I have encountered from time to time. These cases at present are for the most part nameless, having been regarded and diagnosed either as peculiar manifestations of one or another of the commoner and well-known diseases, as eczema, herpes or pemphigus, or, in some cases, as undescribed diseases. From these remarks it will be inferred that the disease is rare, and such in a measure is the fact. At the same time I have met with a sufficient number of cases during the last fifteen years, to warrant the view that the disease is worthy of a special description and a name. I first recognized the affection as being peculiar as far back as 1871, but with the few cases observed at that time was at a loss to classify or to treat them satisfactorily. Since this date I have encountered a number of other cases illustrating the same and other varieties of the disease. In the first edition (1877) of my Treatise on Skin Diseases I made no allusion to the subject, for the reason that my mind was not clear as to the relation that the several cases I had encountered bore one to another, nor that they were really all merely different manifestations of the same pathological process. In the light, however, of a number of marked cases that have now been under observation for a period of years, and of others that have been more recently noted, the statement may be made that, dissimilar as they may in some cases at first sight appear, they all represent varieties of one and the same disease, for which I propose the name dermatitis herpetiformis.

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In the present communication attention will be directed to the principal features of the disease, describing more particularly its symptoms and natural course. On another occasion cases illustrating the several varieties will be brought forward.

It may be premised here that dermatitis herpetiformis includes what Hebra¹ designated first "herpes impetiginiformis" and afterwards "impetigo herpetiformis;" that is to say, that the cases of Hebra constitute one of the varieties of the disease it is proposed to call dermatitis herpetiformis. And here it may be remarked that this name must not be confounded with the "dermatitis-circumscripta herpetiformis" of Neumann, a term introduced by this author a few years ago to designate lichen planus, which at that time he supposed to be an undescribed disease. It may be added that this term is now no longer used by Neumann.

In the second edition of my Treatise on Skin Diseases (1881) p. 276, under the title impetigo herpetiformis, will be found an abstract of Hebra's description of the disease he so named.

His account may be summarized as follows: It is a rare and grave form of skin disease, of which, at the date of his report, he had seen but five examples, four of which terminated fatally. The disease is characterized by the formation of yellowish pustules, arranged in groups or in an annular form, which tend to run together and to dry into yellowish, greenish, or brownish crusts, beneath which a red, excoriated, moist surface exists. On the periphery of the lesions and patches new groups and rings of pustules form. The course of the disease was similar in every case. Each outbreak of pustules was preceded by malaise, chills, fever, and systemic disturbance. The disease occupied all regions, with preference for the anterior

¹ Wiener Med. Wochensch. No. 48, 1872; see also Lancet, March 23, 1872.

surface of the trunk and the flexor surfaces of the thighs.

Single cases, under different names,¹ were before this date reported by Bærensprung,² Neumann,³ Auspitz,⁴ and Geber.⁵ Heitzmann⁶ has more recently also reported a case with the name impetigo herpetiformis. As supplementary to Hebra's description, I gave my own experience with hitherto undescribed varieties of the disease in the following language: "*Within the last ten years I have from time to time met with cases, occurring in both sexes, representing other phases of the disease than heretofore described. In some cases the lesions were vesicular and bullous; in others pustular; in still others, and in the majority of cases, bullous and pustular combined, or these lesions appearing alternately,—the disease being at one time vesicular and bullous, at another time pustular. In all instances the disposition to group or to extend about the periphery was more or less marked. A varied amount of constitutional disturbance, with violent itching, was always present. The disease manifested a disposition to constant recurrence, in the form of repeated attacks, extending in the majority of cases over years, and was but little influenced by treatment. None of the cases occurred in pregnant women; nor in any case has the disease proved fatal. The disease is liable to be confounded with eczema, ecthyma, and pemphigus, according as the lesions happen to be vesicles, pustules, or blebs. The etiology and pathology of the disease are both obscure. In some cases it possesses*

¹ Wiener Med. Wochensch., No. 48, 1872; The Lancet, March 23, 1872; Atlas der Hautkrankheiten, Heft ix, Tafeln 9 und 10. Wien, 1876.

² Atlas der Hautkrankheiten, Tafel 8. Berlin, 1867.

³ Lehrbuch der Hautkrankheiten, III. Auflage, Wien, 1873, p. 173.

⁴ Archiv für Derm. und Syph., II. Heft, 1869, p. 246.

⁵ Jahresb. des K. K. Allg. Krankenhauses zu Wien, Jahrg. 1871.

⁶ Archives of Dermatology, January, 1878.

⁷ In Hebra's fifth case the disease was characterized by vesicles and blebs, from which circumstance he was inclined to regard the disease as a variety of herpes, and designated it "herpes impetiginiformis," Lancet, March 23, 1872.

many features in common with pemphigus; other cases, however, manifest but little disposition to the formation of blebs. It is therefore evident that the process is capable of appearing in the form of various lesions, and that the true impetigo herpetiformis represents but one variety of the disease. More information is needed before the disease can be assigned its proper place in classification."

These words, written four years ago, give a brief account of the disease which is the subject of this communication, and portray its important features. The description is succinct, but it embraces the leading points, and, it may be added, agrees with the views I hold at the present time.

It will be seen from this discription that the name impetigo herpetiformis is altogether inappropriate to express the condition in other and equally important varieties of the disease. The cases that fell under Hebra's observation were for the most part instances of the pustular variety, and he therefore regarded the name he selected as proper; but to call a vesicular or bullous disease "impetigo," with our present ideas of nomenclature, is of course most confusing. Because of the multiformity of the lesions manifested in the several varieties, which will be shown may exist, therefore I think the term dermatitis more suitable, allowing as it does all varieties of this most protean affection to come under its title. The adjective herpetiformis expresses the chief characteristic of the disease, and as in Hebra's cases of the pustular variety, has been present in all of my cases. The disease is unquestionably herpetic, especially in its typical and commonest manifestation, that is, the lesions tend to be vesicular and to occur in small groups or clusters. And for this reason it has on several occasions occurred to me that the affection might be regarded as a herpes, an observation that was also made by Hebra, and by the other reporters of cases mentioned,

All of these observers recognized its herpetic character. The name herpes pruriginosus, it might be suggested, would seem suitable and would answer well for most of the cases observed, itching being almost always a marked and constant symptom. At the same time, if this term were adopted, our present definition of herpes would require to be changed, and we should be obliged to regard pustular and bullous lesions as manifestations of herpes, an admission which would be confusing or even disastrous in its results.

For these reasons I think the name now introduced preferable. That such a protean disease as I have intimated exists, and that these varied cutaneous manifestations are all but forms of one pathological process, there can be no doubt, and I shall elucidate this point by describing the several important varieties, which, as in the case of eczema, are based upon the predominance of certain lesions. Before doing this, however, I may refer to certain symptoms common to all forms of the disease, to which no particular allusion has as yet been made. In severe cases prodromata are usually present for several days preceding the cutaneous outbreak, consisting of malaise, constipation, febrile disturbance, chilliness, heat or alternate hot and cold sensations. Itching is also generally present for several days before any sign of efflorescence. Even in mild cases slight systemic disorder may precede or exist with the outbreak. This latter may be gradual or sudden in its advent and development. Not infrequently it is sudden, one or another manifestation breaking out over the greater part of the general surface diffusely or in patches in the course of a few days, accompanied by severe itching or burning.

A single variety, as for example, the erythematous or the vesicular, may appear, or several forms of lesion may exist simultaneously, constituting what

may very properly be designated the multiform variety. The tendency is, in almost every instance that I have observed, to multiformity. There is, moreover, in almost every case a distinct disposition for one variety, sooner or later, to pass into some other variety; thus, for the vesicular or pustular to become bullous or *vice versa*. This change of type may take place during the course of one attack or on the occasion of a relapse; or, as is often the case, it may not show itself until months or years afterwards. I have notes of several cases where, during a period of from two to five years, the erythematous, vesicular, and bullous varieties were all in turn manifested. Permit me, however, to state again that not only multiformity of lesion, but irregularity in the order of development, is the rule, whether during an attack or later in the course of the disease.

Itching, burning or pricking sensations almost always exist. When the eruption is profuse they are intense, and cause the greatest suffering. As in the case of eczema, before and with each outbreak they become most violent, abating in a measure only with the laceration or rupture of the lesions.

The disease is rare, but is of more frequent occurrence than I formerly supposed. I have encountered fifteen cases, during a period of as many years, drawn from hospital, dispensary and private practice. All, with one exception, were adults, including both sexes in about equal proportion. The natural history is interesting. The process is in almost all instances chronic, and is characterized by more or less distinctly marked exacerbations or relapses, occurring at intervals of weeks or months. The disposition to appear in successive crops, sometimes slight, at other times severe, is peculiar. Relapses are the rule, the disease in most cases extending over years, pursuing an obstinate, emphatically chronic course. All regions are liable to invasion, including both flexor and

extensor surfaces, the face and scalp, elbows and knees, and palms and soles. Excoriations and pigmentation, diffuse and in localized areas, are in old cases always at hand in a marked degree. The pigmentation is usually of a mottled, dirty-yellowish or brownish hue, and is persistent. I have seen it as pronounced as in chronic pediculosis corporis.

The more important forms of the disease may now be considered.

DERMATITIS HERPETIFORMIS (ERYTHEMATOSA).

The erythematous variety manifests itself in patches or as a diffuse efflorescence, as an erythema or superficial inflammation, usually of an urticarial or erythema-multiforme-like type. The urticarial element may be marked, the skin showing a disposition to acute œdematous infiltration in a diffuse form. Urticarial complication, rather than urticaria, is suggested by the condition of the skin; in like manner, a resemblance to diffuse erythema multiforme may be noted. At times the patches, whether discrete or confluent, are circumscribed, and later, by their coalition, show irregularly-shaped, marginate outlines, as in erythema multiforme. The color varies with the shape, being at first bright-red, but soon becoming deep-red or violaceous, mottled, and tinged with yellowish hues. The variegation is usually pronounced in the later stages of the process, at which period more or less diffuse pigmentation is also present. Together with the erythematous inflammation there may form maculopapules, or circumscribed or diffuse flat infiltrations, variable as to size and shape; also vesico-papules, the process now bearing a resemblance to the first stage of herpes iris. It will thus be noted that the eruption, in its general aspect and course, is much like that of erythema multiforme. In severe cases the outbreak is preceded by and accompanied with malaise, chilliness or slight febrile disturbance. The

itching is generally violent, the disease differing in this respect from erythema multiforme.

Its course is variable. It may continue for days or weeks, or, as is usually the case, it may pass into the multiform variety, to be described later. It may be the first manifestation, or it may follow other varieties as a relapse.

As a variety, it is not as clearly defined as the vesicular, bullous or pustular, in some cases it appearing to be but the first manifestation of one of the first-mentioned forms. But it is important that its features be described, for the reason that it is liable to be met with as a clinical picture, and may readily be confounded with other diseases, notably, urticaria, erythema multiforme and eczema. I recall two cases where the diagnosis was at first difficult, and it was not until other manifestations appeared on the skin that the true nature of the process became evident.

DERMATITIS HERPETIFORMIS (VESICULOSA).

The vesicular variety is that most frequently met with. It is characterized by variously sized, varying from a pin-head to a pea, flat or raised, irregularly shaped or stellate, glistening, pale-yellowish or pearly, usually firm or tensely distended vesicles, as a rule, unaccompanied by areolæ. In their early stages they can be seen only with difficulty, and are liable to be overlooked in the examination. Sometimes they can only be detected or seen to advantage in an oblique light. This observation I have repeatedly noted, and arises from the fact of the lesions being flat, translucent and without areolæ. In size they vary extremely, large and small being formed side by side, and in this respect they differ from the vesicles of eczema. Here and there papules, papulo-vesicles, vesico-pustules and small blebs will sometime be encountered. Concerning their distribution, the eruption as a whole is disseminate, the lesions existing

scattered more or less profusely over a given region, as, for example, the neck or the back, but they are for the most part aggregated in the form of small clusters or groups of two, three or more ; or there may be patches here and there as large as a silver dollar, upon which a number will be seated. When in close proximity they incline to coalesce, as in herpes zoster, forming multilocular vesicles or small blebs. Where this occurs they are generally slightly raised and are surrounded with a pale or distinctly reddish areola, which shows forth the irregular, angular or stellate outline of the lesion. At this stage, moreover, the little cluster will generally present a "puckered" or "drawn up" appearance, indicative of its herpetic nature.

The eruption is usually profuse, sometimes to the extent of the upper extremities, trunk and thighs being well covered. It may attack any region, the neck, chest, back, abdomen, upper extremities, and thighs all being especially liable to invasion.

The most striking symptom is the itching. Not infrequently burning is also complained of. Itching, however, predominates, and is in all cases violent or even intense. Patients state that it is altogether disproportionately in excess of the amount of eruption. It is, moreover, a persistent itching, causing the sufferer to scratch constantly. It generally precedes the outbreak, and does not abate until the lesions have been ruptured. Old sufferers, familiar with the natural course of the process, have informed me that they can obtain no relief until the lesions have been ruptured. From my observation I should say that the itching was both more severe and more lasting than in vesicular eczema. The vesicles make their appearance slowly, so that several days or a week may be required for their complete development. Notwithstanding that scratching is indulged in in the early stages of the disease, excoriations are not prom-

inent, owing to the fact that the walls of the lesions are tough, and do not rupture, and that they incline to re-fill immediately on being evacuated.

The diagnosis in some cases is attended with difficulty, on account of the resemblance to vesicular eczema. I recall the embarrassment experienced in the classification of the earlier cases encountered, and the provisional diagnoses of "vesicular eczema?" made at the time. But the irregularity in the size and form of the vesicles; their angular or stellate outline; their firm, tense walls, with no disposition to rupture, and their herpetic character, will all serve to aid in the diagnosis. In some cases the constitutional disturbance and the magnitude of the eruption, as regards profusion, distribution, and multiplicity, showing a more formidable disease than eczema, will also be striking. The itching and burning will usually be found to be more continuous and intenser than in eczema. The obstinacy of the disease to the ordinary treatment of eczema, moreover, must also soon become apparent, the usual milder remedies so frequently of service in acute vesicular eczema, being of little or no benefit in this disease. Finally, the tendency to repeated relapses, and the chronicity of the affection, must strike the observer as peculiar. This variety cannot be confounded with herpes zoster, herpes iris, or pemphigus. Its relations to the "herpes gestationis" of some authors will not be considered in this paper, further than to state that in my opinion they are probably one and the same disease. On a future occasion I shall deal more at length with this point.

DERMATITIS HERPETIFORMIS (BULLOSA).

In the bullous variety the lesions are more or less typical blebs, tense or flaccid, rounded or flat, usually the former, filled with a serous or cloudy fluid, seated upon a non-inflammatory or slightly inflamed

base. In size they vary from a pea to a cherry or walnut, and are for the most part irregular or angular in outline. They incline to group in clusters of two or three, the skin between them in this event being reddish and puckered. Sometimes in immediate proximity—almost contiguous—will exist one, two or three, or a part of a circle of small, pin-head sized, flat, whitish pustules. Vesicles of all sizes, flat or raised, are also generally found near by or disseminated over the affected surface. As in the other varieties, all regions may be attacked, especially the trunk, upper extremities and thighs. In several cases I have seen the greater part of the general surface invaded most profusely, in which event the lesions are usually smaller than where comparatively few exist. They incline to appear in crops at irregular intervals, as in the other varieties. The lesions are generally ruptured in the course of a few days and then crust over with a yellowish, greenish or brownish crust. They are accompanied by burning and itching, which may be very severe. They bear resemblance to those of pemphigus vulgaris, with which of course they may be readily confounded, but they are more herpetic in character. They differ in that they incline to group, and have a more inflammatory herpetic aspect, the type of which picture is seen in herpes zoster. Moreover, around and near the bleb will usually be found vesicles and pustules, the latter often in close proximity, the whole manifestation being quite different from that of pemphigus.

DERMATITIS HERPETIFORMIS (PUSTULOSA).

The pustular variety is generally less clearly defined than the vesicular, because the lesions are often intermingled with vesicles, vesico-pustules and blebs. In typical cases the pustules are acuminate, rounded or flat, tense or flaccid, usually the former, and vary in size from a pin-point to a pea or silver quarter-

dollar. Vesicles and blebs in some cases precede the pustules. The smallest lesions are generally flat, or on a level with the surrounding skin, and, as stated, are frequently not larger than a pin-point or pin-head. Larger pustules, the size of a pea, are generally rounded or acuminate, and are surrounded by a reddish inflammatory areola. Later they incline to flatten, and to increase in size by spreading peripherally, and drying in the centre.

Sometimes they are seated on a slightly raised base. When fully matured they generally present an "angry" appearance, the skin immediately around them having a "puckered" look, from the fact that the pustule itself is irregular in outline, as sometimes is the case in herpes zoster.

They incline to form in groups of two, three or more, and moreover, often appear in patches of two or more groups. Such an arrangement is generally met with on the trunk. The grouping is further peculiar, in that a central pustule will often be immediately surrounded by a variable number of smaller pustules, sometimes in a circinate form, as in herpes iris.

In other localities, however, no such peculiarity occurs, the lesions being discrete, and even disseminate. The pustules are usually opaque, and of a whitish color; sometimes they are yellowish, though they are seldom so yellowish as in pustular eczema. Not infrequently slight hæmorrhagic exudation occurs, as in the later stages of herpes zoster, giving them a reddish, bluish or brownish hue. They are generally accompanied by sensations of heat, pricking or itching; in some cases these symptoms precede for several days the eruption. They pursue a slow course, from one to two weeks being usually necessary for their full development; in other cases their maturation occurs more rapidly. In some cases, together with the pustules are found vesicles and blebs of va-

rious shapes and sizes, and these often form immediately by the side of, or in close proximity to, the pustules. Papules and papulo-vesicles may also be present. In a given area, say of a few square inches, as for example upon the abdomen, there may exist all of these lesions in various stages of evolution. This multiformity is striking, and presents a curious and peculiar mixture of lesions. The attacks last from two to four weeks, after which there generally follows a comparative respite of from one to six weeks. The disease may thus be kept up indefinitely, the outbreaks being at one time slight, at another time severe.

Sometimes it has preceded other varieties; in other cases it has followed the bullous variety; while in some instances that have been under observation for a long period, it has at intervals of months alternated between the vesicular and bullous varieties. After what has been previously said, it need scarcely be stated that this variety is identical with the "impetigo herpetiformis" of Hebra, although in but few of the cases observed by me have the symptoms been so pronounced as in Hebra's experience, if I may judge from the portraits in his Atlas of Skin Diseases. The account given by me relates to the disease as I have encountered it. Hebra's account has been already given, and need not be repeated here. The difference of experience concerns chiefly the severity of the process. Thus, in none of my cases has it proved fatal, while it will be remembered four out of five of Hebra's cases died, and according to Kaposi¹ (Hebra's successor), ten out of eleven cases observed by him have perished. Finally, I have observed it to occur about as often in men as in women, and also in the latter apart from pregnancy. Hebra, on the other hand, met with it only in women, and, moreover, only during the parturient state.

¹Pathologie und Therapie der Hautkrankheiten, 2te Auflage, Wien, 1883.

Concerning the constitutional symptoms, they may be stated to vary with the gravity of the attack. Usually, however, they are more marked than in the other varieties, rigors and alternate paroxysms of heat and cold being especially noticeable. It is the most serious phase of the disease.

DERMATITIS HERPETIFORMIS (PAPULOSA).

The papular variety is in my experience rarer than any other form. I have met with only two cases, one in an adult, the other in a boy, and in both of these the eruption was scanty. It is characterized by the formation of small or large pea-sized, irregularly-shaped, usually firm, reddish or violaceous papular lesions, occurring for the most part in groups of two or three, scattered here and there over the affected surface. They are of variable size, large and small ones not infrequently existing side by side, and are as a rule ill-defined, being neither acuminate nor flat, but resemble the papular infiltrations sometimes met with in abortive herpes zoster. They are of an acute or subacute inflammatory type, and, as stated, bear resemblance to abortive herpetic lesions, and also to certain phases of chronic relapsing papular eczema. Their surfaces are generally excoriated, from scratching, and may be covered with blood-crusts, or with slight, adherent, thin epidermic scales.

The itching is severe, so much so that comparatively few lesions may so torment the patient as to interfere with the night's rest.

Ill-defined papulo-vesicles are also met with here and there, as in the case sometimes of papular eczema. The lesions pursue a slow, chronic course, lasting from one to two or four weeks, when they gradually disappear, leaving more or less pigmentation. Relapses are the rule at longer or shorter intervals.

As I have intimated, this variety bears a close resemblance to chronic papular eczema, and the earlier

cases encountered by me were so diagnosed, but the irregularity in the size and form of the lesions; the disposition to group; the usually slow evolution of the lesions; the tendency to appear in crops at irregular intervals; the chronicity of the process; and the extreme obstinacy to local and internal treatment, are all peculiar. It is the mildest phase of the disease.

DERMATITIS HERPETIFORMIS (MULTIFORMIS).

This may be regarded in the light of clinical variety, one that is eminently useful from a practical standpoint. It is a variety of the disease in the same sense that eczema rubrum is a variety of eczema. It is important to consider it for the reason that it is a picture which may at any time show itself in the course of the disease. It is a phase not infrequently met with, and on account of the great mixture of lesions and the difficulties presented in diagnosis, is worthy of special description. The multiform variety consists of erythematous, sometimes slightly raised, urticarial patches of variable size and outline, often confluent, of a reddish or violaceous, yellowish, dusky, sometimes variegated, color, not unlike that of erythema multiforme. In addition, there often exist more or less well-defined, irregularly shaped or rounded, maculo-papules and flat infiltrations, papules and vesico-papules, all in various stages of evolution. Small blebs and pustules, pin-point sized or larger, may also be present, though with the vesicular element predominating it is not usual to meet blebs or pustules of large size nor in numbers. As the disease is in an early or late stage of its existence, will pigmentation and excoriations be slight or marked. Briefly, then, to recapitulate, there exists a mixture of lesions, with no single type predominating, calling to mind in its behavior eczema, although it is far more capricious and protean. It must also be remembered that the process may at any period change its type, such an occurrence usually taking place with an ex-

acerbation or a relapse. Thus, the vesicular variety may exist for a variable period, to be followed in a few weeks or months by an attack of blebs, or it may be pustules, or by a mixture of the blebs and pustules. This mingling of several varieties is a marked and peculiar feature of the disease, giving a very striking dermatological picture, different from that seen in any other disease of the skin.

In conclusion, permit me to summarize by saying that I have endeavored to show:

1. The existence of a distinct, well-defined, rare, serious, inflammatory disease of the skin, manifestly of an herpetic nature, characterized by systemic disturbance, a great variety of primary lesions, by severe itching and burning, and by a disposition to appear in repeated successive outbreaks.

2. That the disease is capable of exhibiting itself in many forms, all having a tendency to run into or to succeed one another irregularly in the natural course of the process.

3. The principal varieties are the erythematous, papular, vesicular, bullous and pustular, which may occur singly or in various combinations.

4. That it is a remarkably protean disease.

5. That the pustular variety is the same manifestation as the disease described by Hebra under the name "impetigo herpetiformis," this being the only form hitherto described.

6. That the several other and equally important forms are worthy of special remark.

7. That the term "dermatitis herpetiformis" is sufficiently comprehensive and appropriate to include all varieties of the process.

8. That it may occur in both sexes, and in women independent of pregnancy.

9. That it usually pursues a chronic, variable course, often lasting years, and is exceedingly rebellious to treatment.

