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to Pemphigus.*

BY

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FROM

THE AMERICAN JOURNAL OF THE MEDICAL SCIENCES,  
FEBRUARY, 1897.





RELATION OF DERMATITIS HERPETIFORMIS TO ERYTHEMA  
MULTIFORME AND TO PEMPHIGUS.<sup>1</sup>

BY LOUIS A. DUHRING, M.D.,

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CONSIDERING that there has been, and that there still exists, some diversity of opinion among dermatologists regarding dermatitis herpetiformis, especially as to the cutaneous manifestations which should be included under this caption, and those that should be relegated elsewhere, a few remarks concerning the relation of this dermatosis to several well-known diseases may be made, with a view of making the subject plainer. It is not my intention to discuss the literature of the past decade, to criticise the cases that have been recorded as examples of dermatitis herpetiformis, nor to discuss the views of the reporters of such cases, beyond remarking that in some instances gross injustice has been done to this disease. It is not out of place to state here that the writer, in the light of additional clinical experience, sees no reason for changing his views as expressed in his earlier communications. Well-defined, typical cases are from time to time still presenting themselves to him as formerly. Cases with less clearly defined features are also sometimes met with, as well as occasionally cases in which it is difficult in the beginning of the attack to decide whether they should be classified with dermatitis herpetiformis or with some other disease. Such being the case, it is proper to note and label all the typical cases met with. There are enough of these to make the subject interesting and to enable us to say that they represent a special cutaneous disease—a disease *sui generis*. There exists among observers, however, but little difference of opinion concerning typical cases. These have been recognized and described in all dermatological centres, and the reports are so uniformly alike that no time need be spent here in a recapitulation of this subject.

The two diseases that bear the most likeness to dermatitis herpetiformis are erythema multiforme and pemphigus. Dermatitis herpetiformis possesses clinical features common to both, but it is more closely related to erythema multiforme than to pemphigus. It also possesses features in common with herpes, in a sense that the eruption is herpetic and also neuritic. By the term neuritic I mean that the cutaneous

<sup>1</sup> Read before the American Dermatological Association, September 11, 1896.



manifestation is obviously under the control of the nerves of the skin, as in the case of herpes simplex and herpes zoster. With the term neurotic, on the other hand, I would convey the idea that the disease of the skin was due to nerve-influence, but that this cause was not necessarily patent on the skin. Thus, alopecia areata and vitiligo are both neurotic diseases, but they do not display unmistakable signs of the cutaneous nerves being involved, and hence would not be called neuritic. Upon the herpetiform character of dermatitis herpetiformis I have always insisted. This feature is striking in all cases, and is a just reason for taking exception to the term dermatitis multiformis for this disease, as has been suggested by some observers. The term multiformis fails to convey any idea of the chief characteristic of the disease, namely, its herpetiformity. The term multiformis is general and vague, and is without special significance beyond the point that it indicates polymorphism, a feature common to certain other diseases, notably eczema. Herpetiformity, on the other hand, as stated, is eminently characteristic of this disease. Without it there can exist no dermatitis herpetiformis.

I have stated that dermatitis herpetiformis possesses features in common with erythema multiforme. Not only are both diseases strikingly polymorphic in their manifestations, but they are, moreover, allied in nature. In some cases of dermatitis herpetiformis this observation is conspicuously manifest. Many cases will be found at one period or another in their course to possess certain features common to erythema multiforme. There are instances, however, in which no resemblance to erythema multiforme occurs at any period in the course of the disease. It will be understood that erythema multiforme is employed in its broadest sense, and that it includes herpes iris. In dermatitis herpetiformis the cutaneous manifestations, it may be remarked, are in most cases more intense, more persistent, and more chronic than in erythema multiforme. The formation of pustules, especially miliary and acuminate, moreover, so common in dermatitis herpetiformis, is a feature that is wanting in erythema multiforme. In some cases the series of symptoms in dermatitis herpetiformis are such as to suggest the idea of a chronic erythema multiforme. But, even in these instances, the process will be found to be considerably more than a chronic erythema multiforme, so that even if the use of this term were sanctioned, a correct idea of the process of dermatitis herpetiformis would not be conveyed. While, therefore, some cases of dermatitis herpetiformis are allied to erythema multiforme, and simulate that disease, it would be unjust to both to regard them as being one and the same disease. It may be remarked here that, in my experience, dermatitis herpetiformis simulates erythema multiforme more frequently than *vice versa*. I have seen cases of undoubted dermatitis herpetiformis (as proved by the history and the subsequent course of the disease) resemble in the beginning erythema mul-



tiforme, but usually only for a short period. I have also met with rare cases in which a threatened dermatitis herpetiformis proved to be an erythema multiforme. Such experience long ago led me to the conclusion that these diseases were cognate.

The other well-known disease to which dermatitis herpetiformis bullosa is more distantly related, and to which it bears a likeness, is pemphigus, but from which it differs in important particulars. In dermatitis herpetiformis blebs occur in certain varieties. They appear especially in the bullous and in the multiform varieties. In pemphigus, it need not be said, they are constant lesions. The existence of blebs in dermatitis herpetiformis does not in itself signify a relationship to pemphigus any more than the blebs in herpes iris indicate a kinship to pemphigus. No one, I believe, at the present date holds the view that herpes iris and pemphigus are closely related; nor does anyone contend that pemphigus and herpes simplex or herpes zoster are akin. In this connection I may refer to the subject of the coexistence and the combination of morbid processes, giving rise to deviations in type and to modifications, which I believe is a much commoner occurrence than is generally considered to be the case. By admitting that morbid processes in general may coexist or combine, the numerous variations from types of diseases so constantly met with may be accounted for. Some so-called "anomalous cases" may thus be explained. The point of practical importance in all such instances is to determine the prevailing morbid process, to distinguish between essentials and accidentals, in order that therapeutics may be directed against the predominant process. I believe that dermatitis herpetiformis may in rare instances occur in combination with certain other diseases, particularly with erythema multiforme and with pemphigus; and also that it may merge or lapse into these diseases. Clinical experience seems to warrant this statement, and I can see no objection to accepting this belief. Cases of dermatitis herpetiformis sometimes occur that partake more or less distinctly of erythema multiforme, others of pemphigus; that is to say, they possess clinical features that are strongly suggestive of one or the other of these diseases, yet holding fast to the peculiar features of the type. It may be said that dermatitis herpetiformis occupies a position between erythema multiforme and pemphigus.

It will be borne in mind that dermatitis herpetiformis possesses the peculiarity of manifesting itself with varied primary lesions, and of pursuing an irregular course. Irregularity or even capriciousness in the production of the lesions is in most cases a notable feature. They may be erythematous, papular, vesicular, pustular, bullous, or a mixture, and sometimes they all occur together. Polymorphism is usually a prominent feature, generally more so even than in eczema. On account of this peculiarity a general likeness to eczema is occasionally



met with. I recall one of my earlier cases, that was under observation for a long period, the lesions being small herpetiform vesicles and pustules, occurring in patches, accompanied by intense itching, and was for a long time regarded as a multiform eczema of an unusual type. Throughout a period of several years it certainly bore more resemblance to eczema than to any other established disease of the skin. At no time was there any likeness to erythema multiforme (erythematous, vesicular, or bullous), nor to pemphigus. In dermatitis herpetiformis polymorphism is even more notable than in erythema multiforme. It is not, however, a constant feature. Occasionally cases are met with in which it does not occur, the lesions being, it may be, vesicular, pustular, or bullous throughout the attack. It is especially in relapses and recurrences that this feature is most conspicuous. In this respect the disease is altogether different from pemphigus. The evolution of the lesions, moreover, is irregular, and is peculiar. In probably no other disease is this so singular. One notable feature is that they incline to make abrupt and radical changes, as, for example, a vesicle into a pustule or bleb; and, furthermore, that vesicle, pustule, and blebs at times may appear simultaneously, and often are seated side by side. The lesions also tend markedly to vary in kind in relapses and in recurrences. This is a striking point in the course of the disease. I have referred to these characteristics because they are totally different from those which mark the symptomatology of pemphigus. One cannot conceive of a pemphigus without blebs, perfect or imperfect in formation. There are but few of the exudative diseases that are more uniform in the production of their cutaneous lesions and that are more regular in their evolution than pemphigus. The evolution of the lesions of a disease, I contend, plays an important part in the history of that disease. It constitutes an integral part of the malady, and one that must not be lost sight of. It is this characteristic that I desire to lay stress upon in differentiating dermatitis herpetiformis from pemphigus. I believe that, if in a case of bullous dermatitis herpetiformis the course of the disease be watched for a long period, polymorphism of lesions will in most every instance sooner or later manifest itself to such a degree that pemphigus must be excluded in the diagnosis. But even in cases in which the evolution of the lesions is ignored there exists in dermatitis herpetiformis more or less conspicuous herpetiform features that are wanting in pemphigus vulgaris. If, therefore, the herpetiform features, the evolution of the lesions, and the course of the disease in dermatitis herpetiformis be kept in mind, the question of pemphigus will seldom arise in diagnosis. It is altogether unreasonable and irrational to hold, as some observers have done, that dermatitis herpetiformis bullosa is merely a pemphigus. If such observers will closely observe and follow the process of the evolution to its termination, and, further, if they would note

the relapses and recurrences which so commonly take place, they would not be so likely to confound these diseases.

CONCLUSIONS. The conclusions I would arrive at, briefly expressed, are :

1. That dermatitis herpetiformis is in most instances a disease with well-defined, tolerably constant clinical features which enable it to be distinguished from other cutaneous diseases.

2. That it is in most instances more closely allied to erythema multiforme than to any other generally recognized disease.

3. That one variety, the bullous, possesses features which resemble those of pemphigus vulgaris, from which disease, however, it differs in the peculiar inflammatory and herpetiform character of the cutaneous lesions, as well as in the tendency to polymorphism, the irregular evolution of the lesions, and in its course.











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