STELWAGON (H.W.)

Reprinted from the "Journal of Cutaneous and Genito-Urinary Diseases," for February, 1890.

DERMATITIS HERPETIFORMIS.

A REPORT OF THREE CASES.

By HENRY W. STELWAGON, M. D., Philadelphia.

HE disease or condition implied by the name dermatitis herpetiformis is sufficiently uncommon and its nature as yet so obscure as . to merit a more or less extended report of each case met with. While I have had opportunity of seeing several cases of this rare disease coming under the observation of Dr. Duhring, in my own practice, in the past ten years, out of a total of about eight thousand skin patients but three cases are recorded. Two of these may be said to represent typically the vesicular variety; the other was of a multiform nature, at one time simulating pemphigus, at another herpes iris, and at another erythema multiforme. It is not my present purpose to enter into a discussion of the nature of the disease, the propriety of the name, or the advisability of grouping together the various cases described under this term. I can not but feel, however, that my distinguished townsman, in presenting and grouping these various conditions, which in the past would have been reported as several diseases, has made a striking advance, an important step toward a more intelligent study and a clearer understanding of these obscure and puzzling cases.

The following notes will, I think, portray the main character and course of the three cases coming under my observation.

CASE I .- William H., aged thirty-two, of English birth and in fair general health, applied for treatment at the Philadelphia Dispensary for Skin Diseases in November, 1883. The eruption for which he sought relief had, according to the statement of the patient, first made its appearance in June, 1881, and consisted of small solid and vesicular elevations, for the most part occurring in groups, and involving the face, neck, trunk, and limbs. Itching had been a troublesome symptom. Considerable arsenic had been taken, and this remedy, he stated, appeared to give him some relief, but the benefit was only temporary. At the time of his application at the Dispensary the following condition was noted: The face, neck, trunk, and upper part of the limbs were the seat of a vesico-papular and a vesicular eruption, the vesicular character being pronounced. Many of the lesions were discrete, elevated a few lines, and varying in size from a pin-head to a large pea; they were irregularly rounded, angular, or elongate. For the most part, however, the eruption appeared in groups of three or more lesions, the skin intervening between the groups remaining apparently normal. Here and there individual lesions, as they dried, showed slight umbilication. Especially on the breast and upper part of the back was the peculiar grouping of the vesicles to be noticed. In places the vesicles had coalesced, resulting in large, flat, irregularly rounded, quadrate, stellate, or puckered-looking blebs. The vesi-



cles and bullæ, although full, were not distended or tense. In some lesions could be seen a slight tendency of the contents toward milkiness, a few becoming sero-purulent. The same characters, but to a less conspicuous degree, obtained with the eruption on the limbs and face. On the latter, and particularly on the ears, the eruption was more scattered, and, although the same disposition to grouping was noticeable, the groups were small and rarely consisted of more than two or three lesions. In the main the individual lesions, as well as the groups, appeared to arise from apparently healthy skin, but not in all; in some they had their origin from a slightly inflamed base or hyperæmic spot, and this hyperæmic base persisted. These latter groups possessed a close resemblance to the groups of zoster. The covering of the vesicles and blebs appeared thin, but exhibited no tendency to spontaneous rupture. Here and there the lesions had been rubbed or scratched, and in addition to such exceriations a few linear scratch-marks could be seen. The burning and itching, the patient stated, were at times almost intolerable. The man claimed to know several days in advance of the approach of an outbreak by a peculiar burning and heated feeling in the skin. In spite of the cutaneous disturbance and its persistence, the general health remained good, although the man was depressed. There was, however, more or less irregularity of the bowels. Suitable local treatment and a prescription containing Fowler's solution and cascara sagrada were ordered. The patient continued this treatment of his own volition for several weeks, with, so he stated, marked relief, and did not reappear at the Dispensary until a relapse took place two months later. At this time the eruption was much more extensive and of a more severe grade than when the patient was first seen, but the same characters were preserved. As he had been constipated for several days past, cascara sagrada, without the arsenic, was again prescribed. He reported a month later, and, with the exception of one or two vesicular groups, the skin was entirely free. Two weeks afterward a dozen or more discrete vesicles and groups could be seen, and so the disease varied for seven or eight months. at the end of which time a fresh and more or less general outbreak occurred. Under the use of a saline laxative and arsenic improvement again set in.

The next note is made in March, 1885, about a year and a half after the case first came under my observation, and it was to the effect that another extensive and severe outbreak had just taken place. The character of the eruption remained the same, likewise the distribution. A tonic saline laxative, along with an antipruritic lotion, was prescribed, and some relief afforded. In June the eruption had again become much worse—sufficiently so as to impel the man to visit the Dispensary. His next visit to the institution was made the following May—eleven months later. He had continued the treatment prescribed at the former visit for several weeks, until the activity of the disease had diminished, the affection afterward remaining comparatively quiescent. A similar mixture, containing a small quantity of arsenic, was prescribed, and seemingly with benefit. Four months later—in September, 1886—he reappeared, and an examination showed another decided relapse. The patient was not seen from this date until the

past month, two years later, coming at this time at my request. He stated that during the two years his skin had been fairly free; at least it had not been sufficiently troublesome to lead him to sacrifice the time necessary to visit the institution. In the past week, however, a fresh general outbreak had made its appearance. In fact, the eruption in extent and severity was at this time about the same as that which presented when he first came under my observation. So far as could be ascertained, no cause for the disease existed. In all it had now lasted, with remissions, a period of eight years, the last five of which the case had been in my charge. The general health was seemingly good throughout, there was no organic disease, and the patient during the whole period of the disease had been engaged uninterruptedly at his occupation. The disease was so variable in its severity that the effect of remedies was necessarily doubtful, but at the same time the impression was formed that the process was more or less influenced either by arsenic or laxatives, more especially the latter.

Case II.—John B., aged thirty-four, laborer, of Irish birth, was admitted to the Hospital of the University of Pennsylvania, November 1, 1886. His condition when admitted was as follows: His whole integument was closely covered with discrete and confluent blebs, varying in size from that of a large pea to that of a hen's egg, and, as a rule, without areolæ. The lesions for the most part were distended; others were more or less flattened. The eruption was profuse, and spared no portion of the body. The skin between the bullæ was in some places normal in appearance, in other places it was of a pinkish or purplish tinge. The conjunctivæ, as well as the mucous membrane of the mouth, were likewise involved in the process. Symptoms were also present, such as coughing and the occasional spitting of small flakes of membrane, indicating that the laryngeal and tracheal mucous membranes were similarly involved. The eruption exhibited no attempt at symmetrical arrangement. The temperature was above normal, and the pulse accelerated. There were mental depression and profound general weakness. In short, every symptom of a serious pemphigus was present, and at this time, without the previous history, such it was considered. The history of the development of the disease and its further course, however, indicated a severe case of the multiform type of dermatitis herpetiformis. From the patient himself and from the notes sent me by Dr. Trimmer, of White Haven, Pa., who had been previously in charge of the case, the following history was obtained: About the last of July, three to four months before admission to the hospital, he contracted scarlet fever from one of his children. The attack was severe and of the anginose type. Ten days after thorough convalescence had begun, and when desquamation had about ceased, he left the house one evening to join some friends, and drank heavily of beer, which had been his custom before his illness. On the following day it was noticed that he was feverish. At the same time there appeared a band of papules, papulo-vesicles, and vesicles, like a collar, around the neck, a similar band around the ankles extending upward six inches, and a four-inch band around the wrists. The papules rapidly developed into vesicles, and these for the most part became confluent. The primary vesicles were small. New parts were soon involved, and finally the whole surface shared in the eruption, the lesions being at first similar to those which had appeared on the neck, wrists, and ankles. The eruption now consisted of discrete and confluent vesicles and bullæ, some of the latter being as large as a small hen's egg. This attack lasted nearly four weeks, at the end of which time there was, according to Dr. Trimmer's statements, scarcely a vesicle or bleb to be seen. He was then given a bath. The following evening he had a feeling of intense burning under the skin, followed the next day with a general crop of vesicles and bullæ. These were not quite so large or so distended as the bullæ of the former outbreak. From this time until his admission into the hospital, crop after crop developed, before one had subsided a new outbreak making its appearance. The constitutional symptoms thus far had been a variable elevation of the temperature, an increase in the pulse-rate, irregularity of the bowels, diarrhæa followed by constipation, and a coated tongue.

The course of the disease after admission may be given in the notes made from time to time. It may be here stated that from week to week the urine was examined, but, further than an abundance of urates, nothing abnormal was found. Mild external applications were employed with a view chiefly to relieve the intolerable burning and itching. The blebs were punctured as soon as formed. Arsenious acid was prescribed in one-thirtieth-grain doses, three times daily, and also twelve grains of quinine in divided doses each day, together with plain nutritious diet and some malt extract. The diet was full but unstimulating. On November 6th, five days after admission, the patient was much more comfortable, although new blebs continued to make their appearance both on the surface and on the buccal mucous membrane. The dose of arsenic was increased to one twentieth of a grain. November 9th: New blebs continue to appear on the trunk, and several erythematous spots on the hands. The vesicles and blebs at this time arose from apparently healthy skin; later, as it will be seen, they had for the most part their origin in erythemato-papular or papulo-vesicular patches. November 18th: New crop of blebs on the face and trunk, with a tendency to form festoons and circles. A small vesicle on the conjunctiva of the left eve near inner edge of the cornea; the neighboring conjunctival vessels injected. Dose of arsenic increased to one twelfth of a grain. Old lesions rapidly disappearing. December 6th: Two large blebs-one on the foot, the other on the thigh-and an extensive general erythematous and papular eruption, assuming the shapes of rings, segments of circles, and in places forming festoons; in the segments of circles there is a strong disposition to vesiculation. Old eruption almost gone. The palms and soles were painful and tender, and of a light pinkish or purplish hue. A laxative pill at night was ordered. December 20th: Marginal keratitis in the left eye. General skin symptoms much improved, and less disposition to the appearance of new lesions. Four days later the improvement had been so marked that the patient desired to pass Christmas at home, and at his request he was discharged. At this time, with the exception of a dozen erythematous spots and several small vesicles, the skin was free. The skin of the hands and feet, however, remained tender, and of the same pinkish or purplish tint, resembling the tint of passive congestion. The dose of arsenic had been increased to one eighth of a grain.

On the 2d of the following April the patient was again admitted to the hospital. Since his return home, three months previously, I had heard from him irregularly, and his condition, while upon the whole much improved, was far from satisfactory. The eruption had been to a great degree in abeyance, but from time to time new lesions had made their appearance. The constitutional treatment had been during this time of a general tonic nature. On admission at this date there was more or less general weakness, and the skin was irregularly covered with an erythematopapular eruption, with here and there a few small and large discrete and grouped vesicles and blebs. A feeling of burning was complained of, but itching was at this time slight. One or two ruptured blebs could be seen in the mouth. Arsenic was again administered and in increasing doses. The patient's condition varied from day to day and week to week, a more or less profuse erythemato-papular eruption, scattered blebs, and groups of vesicles making their appearance from time to time. The stomach rebelled against the arsenic, the dose of which had been increased to one eighth of a grain three times daily, and it was discontinued. Although apparently robust in appearance, the man was weak and debilitated, and, with a view of toning up, cod-liver oil was, after a few days' intermission, ordered. The middle of May there was another general outbreak, but partaking more of the nature of a well-marked erythema multiforme. The trunk, limbs, and face were covered with erythematous and papular rings and segments. Later, especially upon the segmental lesions, bullæ made their appearance, maintaining in a great measure the segmental shapes. There were also a few scattered, flattened erythematous elevations upon which could be seen irregularly distributed vesico-papules and vesicles, somewhat similar to the patches of herpes zoster. There was considerable itching, although at this date burning was the more prominent subjective symptom. All this time the formation, at irregular intervals, of abortive bullæ on the tongue and buccal mucous membrane continued. The palms and soles, which also showed the different phases of the eruption, remained painful and tender-so much so that standing or walking was almost impossible. The temperature at the beginning of this, as with previous outbreaks, showed an elevation of from one to three degrees; and until the outbreak had well subsided, a tendency to slight elevation, especially in the evening, could be noticed. The urine, so far as pertained to the presence of sugar or albumin, remained normal. There was but little disposition toward pustule formation, the eruption at this time partaking of the features of erythema multiforme, herpes iris, and pemphigus, more especially the first named. On July 5th twenty to thirty new dime-sized blebs made their appearance, seemingly arising from healthy skin.

As no impression upon the course of the disease had been made with cod-liver oil, it was now discontinued, and phosphorus in small doses administered. The case progressed in the same variable manner, however, the skin never being free, and with, at irregular periods, fresh, more or less general erythematous and erythemato-papular outbreaks. On account of the sensitiveness of the stomach, and for the reason that no positive influence had been exerted, the phosphorus toward August 1st was discontined, and a general tonic treatment instituted. In the middle of this month the patient was upon the whole better, and, in fact, for the past several weeks the earlier tendency of the disease to take on a serious pemphigoid aspect, as the notes have indicated, had lessened. From this date till September 13, 1887, the disease remained comparatively quiescent, and, at my suggestion, the patient then went to the sea-shore, where he remained until the first week in October. While here he at first improved, but later the disease manifested renewed activity, and the patient, being thoroughly discouraged, and at the suggestion of some of his friends, sought, I have understood, treatment in New York.* In all, the disease had up to this time existed fourteen months, the last eleven of which the patient had been in my charge. While treatment has been incidentally mentioned, it may be stated, as probably already inferred, that it was largely experimental and apparently without result from first to last. The local measures employed were those usually prescribed in acute and subacute eczema.

Case III.—Susan W., a domestic, aged forty-five, of American birth, tolerably robust and apparently in fair general health, applied at the Skin Dispensary of the Hospital of the University of Pennsylvania, December 14, 1886. On examination, the face, scalp, trunk, and arms were found to be the seat of an irregularly scattered, discrete, and grouped vesicular eruption. The lesions were distinctly vesicular, averaging the size of a small pea, some without areola, others showing a slightly inflammatory base. Many of the vesicles had been scratched, and these points appeared as superficial excoriations. If undisturbed, there seemed very little tendency toward spontaneous rupture; in this respect, and also in the disposition toward umbilication of the older and drying lesions, resembling ordinary herpetic vesicles. The groups were probably more numerous than the discrete lesions, and consisted of from three to six vesicles, seated either upon an inflammatory base or

^{*} I have since learned that this case was subsequently admitted to the Skin and Cancer Hospital, New York, coming under the care of Dr. Fox. At this time the patient was shown before the New York Dermatological Society as a case of chronic erythema multiforme. (See Journal of Cutaneous and Genito-urinary Diseases, February, 1888, p. 68.)

upon apparently sound skin. In some of these groups the vesicles had coalesced and given rise to irregularly shaped bullæ. The individual vesicles were peculiar in that they were of a strikingly irregular outline, some angular, some stellate, and, when drying had set in, assuming a puckered appearance. In places could be seen, also, vesico-papules, although these were not numerous. Itching was intense, at times being almost unbearable. The disease, so the patient stated, had made its appearance nine months previously, being somewhat variable in its course. So far as the patient was aware, there had been no predisposing or exciting cause. Her general condition when first seen was fair, but she was nervous and more or less depressed, probably from loss of sleep. A lotion of carbolic acid and an arsenic mixture were prescribed. Fifteen days later the patient was again seen, but the severity of the disease had in no wise abated; some of the old lesions had disappeared, but as many new ones had arisen. On December 21st there had been practically no improvement. The dose of arsenic was increased, and liquor carbonis detergens prescribed for local application. A week later a decided change for the better was noticed, the itching was less violent, and there were fewer new lesions. The case improved up to January 18, 1887, three weeks later, when a fresh general outbreak occurred, much greater in extent than when the patient was first seen.

There were, in addition to the previously described lesions, several blebs, and in places discrete and grouped pustules. The treatment was now changed, but the progress of the disease was in all probability in no way influenced, as the patient failed to return for additional advice, and the case unfortunately escaped further observation. Up to this date the affection had existed eleven months, the last seven weeks of which the patient had been under my care.

In briefly recapitulating, it will be seen that these three examples of dermatitis herpetiformis had much in common. They were all characterized by a remarkable chronicity as well as rebelliousness to treatment. Moreover, the variable nature of the eruption, its herpetic character, the intense pruritus, and the absence of a tendency to atrophy, ulceration, or scarring were striking features. Further, it will be noticed that there was, excepting possibly in the second case, no apparent antecedent or existing cause for the disease, the patients otherwise enjoying tolerably good health. In short, in their character and course and general symptoms these cases were essentially similar to the cases reported by Duhring and others.