WHITE (C. J.) with the writer's compliments.

ÉRYTHRODERMIE PITYRIASIQUE EN PLAQUES DISSÉMINÉES.

By Charles J. White, M.D.,

Instructor in Dermatology at Harvard University.

Reprinted from The Journal of Cutaneous Diseases
for April, 1903.

LIBRARY
SURGEON GENERAL'S OFFICE
JUL 21 1903
771.

THE GRAFTON PRESS
PUBLISHERS
70 FIFTH AVENUE
NEW YORK

Copyright 1902 by The Grafton Press

ERYTHRODERMIE PITYRIASIQUE EN PLAQUES DISSEMINEES.

By Charles J. White, M.D.,

Instructor in Dermatology at Harvard University.

THE rarity of this disease and the fact that two similar cases have been reported from the Department of Skin Diseases at the Massachusetts General Hospital leads me to place on record our third case within three years.

The patient, I. S., 44 years of age, was born in Russia, near Poland, and until his departure for the United States was a peddler of milk and had experienced but one illness, the grippe. He is married, and his wife and six children are living and free from any skin diseases. In 1888 he came to this country, and has kept a variety store in Boston ever since. This occupation requires him to stand for many hours each day.

In 1898 the patient came to the skin clinic and the following entry was made in our books: "Extraordinary macular, deep red lesions of eight years' duration. Symmetrical glandular infiltration. Beginning mycosis fungoides?"

In April, 1901, the patient felt that the boot on his right foot was too tight. His foot became œdematous and the second toe turned blue, cold and very painful, and dry gangrene developed, on account of which the toe was amputated on July 17 in the surgical out-patient department by Dr. C. A. Porter. A thorough physical examination was made at this time, in which no sugar or albumen were found in the urine, and his viscera were declared normal. On account of this negative examination and the persistence of the dorsalis pedis pulse the diagnosis of "*hebräische Krankheit*" was made.

The unusual diagnosis and possible ætiological importance of this intercurrent malady should detain us for a few moments. In the British *Journal of Dermatology* (February, 1901, p. 41) F. Parkes Weber reports a strikingly similar case in a man who noticed swelling, cyanosis and pain in his left foot. These conditions were all increased in severity whenever the patient stood for a long time and especially after exposure to heat or to cold. Gangrene developed later

LIBRARY
SURGEON GENERAL'S OFFICE
JUL - 1
771

in several points and amputation of the foot was performed. The pathological examination showed that the arteries were patent and showed no end-arteritis or thrombosis, but the microscope revealed great thickening of the arterioles and a similar condition of the middle coat of the arteries in the vicinity of the gangrenous spots. The nerves were found to be quite normal, but the muscles and bones of the foot were atrophied. On the strength of these findings Weber made the diagnosis of obliterating arteritis of the arterioles—the so-called "*hebräische Krankheit.*"

In July, 1901, our patient appeared at the nerve department of the hospital and was treated successfully for pain in the right foot and toes, and four weeks later he again presented himself, complaining of pain behind the knee of the left leg. Examination proved that the pain increased whenever the man stood or was exposed to the cold, but remained unaltered in warm atmospheres.

On November 12, 1902, the patient came to the skin department again on account of a severe lymphangitis which did not yield to ambulatory treatment, and at the end of a few days he was sent to the wards of the hospital. The house records state that at entrance the man's temperature was $99\,^3/_5$ degrees F., pulse 80, and respiration 20. General physical examination again revealed nothing abnormal of importance outside of the local disturbance in the leg. The right leg was not tender to pressure, but the toes were cold and blue. The left leg, on its anterior, internal face, showed a cord-like mass, red-blue in color, easily palpable and painful to pressure. Tenderness was most noticeable, however, in the calf, popliteal space and just below Scarpa's triangle. No œdema of the foot was present. An X-ray examination showed nothing abnormal and after two weeks the man was discharged improved.

On November 29 the man returned to us for a thorough cutaneous investigation. He stated that the spots appeared on his body twelve years ago, during the winter months, and he became aware of their presence merely from sight, as they did not itch or burn or cause any pain. He is well nourished and feels well. On his scalp there is a mild degree of seborrhœa, but the skin is not congested. Over the very numerous pinkish-red, irregularly circular macules about one-lower and upper arms, especially on the extensor surface, appear half inch in diameter, which disappear on pressure. The hands show nothing.

Above the nipples there are no lesions, but below them appear small, sparsely scattered macules similar to those on the arms. Lower

down the spots grow larger, reaching the diameter of one inch, but losing their distinctly round appearance. They are velvety to the touch and soft, and do not rise above the surface, are slightly scaly and disappear when compressed.

On the back, the lesions begin at the lower edge of the scapulæ, and are large, irregular in shape and sparsely scattered, do not appear over the spine, and are most abundant on the flanks. The lesions on the thighs, anteriorly and posteriorly, are so numerous that they practically coalesce, especially on the buttocks, where they appear blue-red in color and slightly furfuraceous. The penis and scrotum are unaffected. The lower legs are almost free. In the cervical region no glands are to be felt, but the right epitrochlear gland forms an oval body about one-half inch in length, while the left feels like a good-sized pea. There are small glands in the axillæ, but none appreciable in the inguinal region.

When the patient rests in bed for any length of time the macules disappear entirely from the arms, but no connection between the seasons of the year and the disappearance of the lesions can be discovered. The scaling everywhere is fine, dry and very slight. Pruritus and any evidence of papulation are positively absent.

The blood examination (made by our assistant, Dr. F. S. Burns) reveals the following percentages:

Polynuclear neutrophiles	62.8	per cent.
Small lymphocytes	20.0	"
Large lymphocytes	7.6	"
Eosinophiles	5.6	"
Transition forms	4.0	"
Hæmoglobin	85.0	"

For microscopical examination a small macule was excised from the thigh, hardened in alcohol and stained by various methods.

Epidermis. The stratum corneum consists of seven to eight layers of elongated, non-nucleated cells, forming an open network.

The stratum lucidum is absent.

The stratum granulosum is reduced to one layer of attenuated spindle-shaped cells, with long, oval nuclei. In places even this thin layer is absent and small, very fine granules seeming to replace the normal layer.

The stratum spinosum appears much compressed. The cells are flattened and elongated and reduced to three or four layers in thickness. The whole stratum seems very œdematous, the protoplasm and

nuclei appearing cloudy and receiving the stain but poorly. The palisade layer is almost universally absent and in certain areas the nuclei seem to have fallen out, leaving but a protoplasmic network. In other places at the same level the nuclei remain, but the tissue receives only the acid stains. In the upper layers of the rete, near the granular layer, the tissue is cloudy, bearing non-nucleated or vacuolated cells.

As we leave, on either side, this focus of highly degenerated epidermis, the conditions gradually become more normal, the layers increase in thickness, the tissue stains more clearly, the spines of the rete cells reappear and mitoses are observed.

Corium. Just below the highly abnormal epidermic area the papillæ are practically obliterated and, with the exception of an occasional connective tissue nucleus, no structures appear. Here the collagen itself seems abnormal. The bundles consist of œdematous, poorly staining, slightly waving fibers with elongated nuclei. The bundles abandon their more usual direction and run vertically and obliquely as well as horizontally and between them large empty lymph spaces appear.

In the more normal parts of the sections, papillæ reappear but are short and comparatively far apart and bear somewhat dilated capillaries, with no undue amount of cellular extravasation. Subpapillary vessels appear with no apparent endo or perivascular changes. The farther we leave the distinctly affected region, however, the vessels become more and more numerous and exhibit more and more cellular extravasation.

In the deeper layers of the corium the collagenous bundles become larger and somewhat tortuous, but they also are swollen, œdematous, mostly a-nuclear, and are surrounded by dilated lymph spaces. In addition, an occasional elongated, greatly dilated lymph vessel, with unusually vesicular nuclei, presents itself.

Deep down in the corium sweat glands and hair follicles appear, but their cells also show the effects of this universal œdema and appear vacuolated or have crescentic nuclei.

Elastin is everywhere reduced in amount, especially in the papillary and subpapillary layers, but it retains its normal staining reactions to a marked extent, considering the highly œdematous condition of the skin.

The literature upon this rare disease is very meager, and so far as I have been able to discover is limited to the references which may be found at the end of this article. There seem to be but two affections

at all akin to *érythrodermie pityriasique*. The first is parakeratosis variegata, first described by Unna;[2] but even this disease offers to my mind one insuperable barrier to the identity or even very close relationship of the two processes, namely, the presence of papules which were never present in the three cases of *érythrodermie pityriasique* of which I am cognizant (Brocq's, J. C. White's and the present one).

The microscope also reveals a condition which separates the two affections, that is the nucleated horny cells are present in practically all the sections of parakeratosis variegata and are never observed in the examples of *érythrodermie pityriasique* thus far examined.

The second disease which presents somewhat similar features is mycosis fungoides in its initial stages, the so-called pre-mycosic dermatitis. Here again one feature practically always present in one disease is universally absent in the other; I mean pruritus, which forms so distressing and persistent a factor in mycosis fungoides, and which is so totally wanting in *érythrodermie pityriasique*.

The discussion and differential diagnosis of these three affections have been so recently and so thoroughly described in this journal by J. C. White and by Colcott Fox and J. M. H. Macleod[3] that it seems hardly necessary to speak further here. It only remains for me to emphasize the great clinical similarity of the two previously reported cases of Brocq[4] and of J. C. White[1] to the case now presented by me and to summarize the histological findings of J. C. White's case (unfortunately no biopsie was possible in Brocq's case), which are so nearly identical with the microscopical conditions of the present case detailed above.

1. Open network formation of the stratum corneum composed of non-nucleated horny cells.

2. Absence of the stratum lucidum.

3. Great atrophy or even total absence of the stratum granulosum.

4. In places, compression of the rete cells and reduction of the layers composing the stratum spinosum; absence of the palisade layer; and finally, greatest divergence from the normal directly over the parts of the corium mostly affected.

5. Œdematous condition of the corium, and

6. Reduction in the amount of elastin.

In truth, the two specimens are almost identical, the one and only dissimilarity being the absence of vessels and their accompanying perivascular infiltration in my case, but here again the identity of the

two cases returns the moment we leave the most seriously affected areas.

In closing I wish to make a suggestion with regard to the possible origin of this unusual dermatosis. Is not the intercurrence of this strange arteritis of the arterioles of the toe a significant fact in relation to the general cutaneous disease? In other words, have not we here a possible or even probable clue to the genesis of the whole process? It seems to me fair to assume that there may be everywhere (where the macules appear) a localized ischæmia produced by the deeper arterial twigs which don't appear in our sections, but which produce a localized starving of the skin, manifesting itself as œdema and consequent atrophy of the integument. This does not lead to actual necrosis and gangrene, as it did in the patient's toe (which is farthest from the heart and suffers most from faulty nutrition), for artificial anastomosis may come to the rescue and keep alive the areas normally dependent upon the diseased vessels.

I wish to express here my indebtedness to Mr. L. S. Brown, of the Massachusetts General Hospital, for the photographs which emphasize so well the various histo-pathological facts which I have endeavored to describe in the text.

BIBLIOGRAPHY.

[1]James C. White. Jour. Cut. and Gen.-Urin. Dis., 1900, p. 536.

[2]Unna, Santi and Pollitzer. Monatshefte f. prakt. Dermat., vol. 10, pp. 405, 444.

[3]Colcott Fox and J. M. H. Macleod. Jour. Cut. and Gen.-Urin. Dis., 1901, Sept., p. 424.

[4]Brocq. Revue générale de Clinique et de Thérapeutique, 1897.

ILLUSTRATIONS.

FIG. 1. *Low power.* Represents the section as a whole with the most gravely affected regions of the corium and of the epidermis in the centre of the photograph. The great atrophy of the epidermis and the disorganized condition of the corium are well shown. On each side of the photograph the corium is beginning to appear more normal and on the left hand the various normal deeper structures are present. Hæmatoxylin-eosin.

FIG. 2. *High power.* Represents the most severely affected area of the epidermis. The rather increased stratum corneum and the marked changes of the other layers are clearly seen. Hæmatoxylin-eosin.

PLATE XVIII.—To Illustrate Dr. C. J. White's Article.

Fig. 1.

Fig. 2.