Hutchins (M. B.)

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A CASE OF "POROKERATOSIS" (MIBELLI).

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THIS case first came under my observation on the 5th day of October, 1892. A description was then written, the diagnosis being left open. I recognized it as dissimilar to any hyperkeratosis with which I was familiar or of which I had read. Fearing that my knowledge of the literature might be incomplete, I hesitated to publish the case.

I did not see Mibelli's report in The International Atlas of Rare Skin Diseases of October 28, 1893, but as soon as I saw his plates and description in the Monatshefte für praktische Dermatologie of November 1, 1893, I recognized the identity of his cases with mine.

My report was further postponed until I should see how his cases were received. Nothing concerning them came to my notice. On January 1, 1894, Respighi's cases were reported in the Monatshefte, after which nothing appeared until March 1, 1895, when Mibelli's second article was published in the same journal.

On May 16, 1895, I succeeded in getting new notes of my case, with two photographs. At intervals up to the present time I have used every effort to get my patient to permit the excision of a portion of the diseased skin for microscopic study; and, after a year of futile effort, I am compelled to report simply the clinical features of the case because of failure to get even the smallest part of the tissue. I hope that the opportunity for a histological examination will yet come.

The first record of my case is as follows:

October 5, 1892.—Mr. W., aged thirty-two years; commercial traveler; wholesale liquors.

The disease began at about the age of two years, just inside of the border of the left palm, at the radial side of the base of the index finger, as a "seed-wart"-like growth. This gradually spread, and other patches, or points, developed until about the age of fifteen, when the one on the palm at the base of the index and middle fingers appeared. He does not know the date or form of development of the dorsal lesions. Now, the original point is the center of a smooth patch of palmlike skin, extending halfway to the base of the metacarpal bone of the first finger on its dorso-radial side, also forward, narrowing, nearly to the first phalangeal joint of the index finger. The patch is irregularly bordered on the radial side of the dorsum of the hand and finger and extends internally nearly on the palm. The boundary of the described area is irregular, wavy, about one line in width and height (two millimetres), and is formed of horny epidermis. It is like the outside of a seam, with a longitudinal, thread-like line dividing its lateral halves. (See diagrams.) Here and there in this line are round, millet-seed, or smaller, sized, blackish epidermic concretions which can be picked out. The "seam" can be cut to the *niveau* without pain or injury to the papillæ. On the dorsum of the first phalanx, external to this "seam," the hairs are broken in their follicles, and the latter show dirty, horny plugs.

On the left palm, corresponding to the lesion which appeared at fifteen, at the base of the first and second fingers, there is a patch the size of a nickel five-cent piece, having the peculiar seamlike border, here scarcely elevated—being more of the nature of a fine fissure. This patch is a smooth hypertrophy of horny epidermis and shows a number of the millet-seed-sized bodies in its surface. Between the radial side of the wrist and the base of the index finger, dorsal surface, is an oblong area surrounded by a "seam" similar to that first described, the included space being an average of one inch wide by two inches long, but the "seam" is tortuous. The skin within looks normal save for one pea-diametered verrucous growth toward its distal end. The hairs in and about the patch are normal. Another patch, similar to the other dorsal ones, lies to the ulnar side of the unaffected surface between the first and third described, about one half by three quarters of an inch in area, included skin normal. The border of all the dorsal patches is markedly seamlike, or like a mole burrow, or a bird's-eye view of a mountain range.

The patient does not remember any recent extension of the disease. There is occasional itching, as upon exposure to the sun.

Just below the left zygoma is a roundish, smooth patch the size of a silver quarter of a dollar, bordered by a fine fringe of internally loosened epidermis. It is of a faintly brownish color, with slight telangiectases and a faintly atrophic appearance. He thinks this lesion is contemporaneous with the lesions on the dorsum of the hand. There are no other lesions.

Fig. 1.

The patient's health is excellent.

As he came to me for treatment, I attempted the removal or destruction of the abdominal growths, first with a twenty-per-cent acid salicylic-flexible-collodion mixture. This took away the excess of horny tissue, but left the thin central line *in statu quo*, save in a point between the two dorsal patches. Fuming nitric acid was afterward penciled on, as a result of which parts of the seam appeared to entirely disappear.

On October 17, 1892, I lost sight of the patient, not examining him again until May 16, 1895.

The description made upon the latter date can be much abbreviated and simplified by a study of the plates (Figs. 1 and 2) which accompany this article, the photographs from which the plates are made having been obtained at that time.

On May 16, 1895, the patch seen between the bases of the index and the middle finger, in the palm, appears slightly horny and shows two or three of the minute plugs or globules (Fig. 1). Its boundary is slightly raised—wavy. Along the free edge of the interdigital fold this border is a very faint, fine seam; the rest of it is more marked and appears composed of two lateral halves divided by a longitudinal, central line of horny tissue of a yellowish or blackish color. The

horny hypertrophy anywhere on the palm is of the usual callous-yellow color. The adjacent part of the patch on the side of the finger and hand is separated from the palmar lesion by a small, horny, epidermic elevation about one eighth of an inch wide.

The boundary of this second patch continues around on the finger and the dorsum of the hand, back to the starting point, as will be seen in an examination of both plates, following the edge of the finger, looping back on its dorsum to the first interosseous muscle and over the edge of the fold between the thumb and index finger (Fig. 2). The

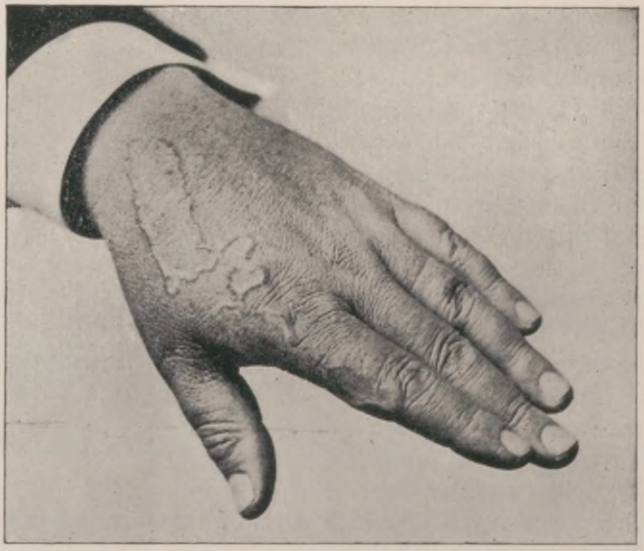

FIG. 2.

"seam" along the dorsum of the finger is the most decided of all, though it is the least distinct in the photograph (Fig. 2). There is a slight break in the "seam" over the middle of the metacarpal bone —on the dorsum. The entire included patch has nothing save a little smoother appearance than the same area on the healthy hand, excepting an epidermic thickening on the dorsal side of and adjacent to the border in the digito-*pollical* fold. The palmar part of this boundary is much like that of the first patch, with here and there within its circumscribed area a few of the minute plugs. The boundary on the dorsal surface is about one twelfth of an inch in thickness, appearing a simple horny hypertrophy—seamlike—with the characteristic

central line, showing many yellowish-black, millet-seed, or smaller sized concretions. The "seam" along the finger may have been increased in thickness by the patient biting there for occasional itching.

Where the "plugs" are absent the "seam" has a simply flattened line in the center, as if a fine thread were laid in.

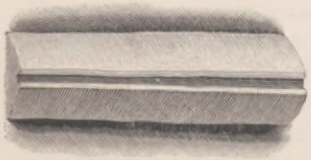

FIG. 3.—Magnified diagrammatic view of a dorsal "seam."

FIG. 4.—Magnified transverse sectional view of dorsal "seam."

FIG. 5.—Magnified transverse sectional view of palmar "seam."

One fourth of an inch to the ulnar side on the dorsum is a dumb-bell-shaped area bounded by the usual "seam," broken for one fourth of an inch in one point, the whole lesion like two half-dime pieces, connected by a small isthmus. This is the patch which was one half by three fourths of an inch in area in October, 1892. It appears to have undergone some mesial change. Between this and the previously mentioned patch is a pinhead-sized horny elevation, doubtless a new lesion. The skin within the last-described patch has a slightly atrophic appearance, but shows a number of normal hairs, one of which is directly in the "seam."

The dorsal metacarpal patch, as seen in the plate, comprises an irregular, tortuously bordered area about one by three inches. Its typical border is connected with patch No. 3 by a horny hypertrophy one fourth of an inch wide. The inclosed skin has the normal appearance, the lesion described in the first notes having disappeared. The hairs are slightly fewer than those without the patch, and some of them are *white*. About an inch from the posterior loop, on the ulnar side, is a pea-diametered widening of the "seam" callouslike. In the "wall" or "seam" are some apparently normal hairs, but there is a keratosis of their follicles, and one or two of them are broken. This, like all the dorsal "seams," has rather pinkish sides, sloping up and inclosing between them the horny thread and a few "plugs." These central formations can be picked out quite easily. The "thread" is distinct and black on the finger.

Save in the *palmar* patches, the round plugs occur only in the center of the "seams."

The disease causes no sensation save an occasional slight itching

in the "seams," especially that of the index finger, exposure to the sun somtimes inducing it. His mother tells him that the disease first appeared in the palm as a " seed wart," which could be picked out.

As to the nerve supply of the regions affected, we find the median in the palm and the radial and part of the ulnar in the dorsum. The disease follows the course of certain parasitic affections rather than the distribution of the nerves.

The face lesion—no photograph of this was obtained—below the anterior extremity of the left zygoma, is oblong, three fourths of an inch across, one inch from the upper to the lower margin in dimensions. It is now faintly brownish, with a tint of red from fine telangiectases, slightly depressed, but non-atrophic to the touch. The lens shows thin, adherent, shining scales. There is no abnormal sensation in the lesion, and I have no information as to absence of secretion.

The patient is of the brunette type, short, robust. His height is five feet seven inches; weight, two hundred and nine pounds. Has gained in weight in the past two years. The general health always good.

In the fall of 1895 he stated that he had carefully watched the hand areas for signs of perspiration and has seen none, though perspiring freely elsewhere. However, the skin, especially in the larger dorsal patch, remains soft and pliant and feels to me a little moist.

Clinically it is presuming somewhat to consider the face lesion as of the same nature as those of the hand; a comparison of the histological structure of all would be necessary to decide this point. I was inclined to consider it only coincident with the others and not as related, but must leave the question *sub judice*, in view of Mibelli's description of his cases.

Changes in the disease of my patient were but slight in over two years and a half. One destroyed dorsal lesion remained away; and possibly the " pinhead lesion " mentioned in the last notes is the only new one. Here and there in the dorsal " seams " is a slight hiatus, whether idiopathic or from my treatment in 1892 it is difficult to decide.

A Comparison of Cases.

V. Mibelli's case (International Atlas of Rare Skin Diseases, October 28, 1893) had lesions on the face and neck; mine only the one on the face to correspond. The face lesion in my case had no marked hyperkeratosic symptom. His " dike " is practically the same as my " seam." There was no peripheral " dike," " wall," or " seam," about

my smallest lesions, save on the palm patch, nor any exaggeration of the process over the joints. His "wide space" on the back of the right hand is similar to the large patches in my case, but mine have no smoother, thinner appearance, and the hairs remain. His "wall" or "dike" showed a tendency to disappear in places; in mine there were, latterly, complete breaks. A little warty lesion on the dorsum in my case is somewhat like his "conical, hard, etc.," elevations. His only palmar lesion was not different from an ordinary callosity. It would be of interest to know if this might develop a condition like the palmar lesion in my case.

His case began at the age of two years, the same as mine, but on the dorsum of the hand—apparently in the same way. The history of the development of mine can not be obtained fully enough for a further comparison. Mibelli's case, in the Atlas, had no subjective symptoms; mine a little itching. No other member of my patient's family had the disease.

In Monatshefte für praktische Dermatologie, Bd. xvii, No. 9, November 1, 1893, Mibelli's article, Beitrag zum Studium der Hypokeratosen der Knaueldrüsengänge—Porokeratosis, is very full and exhaustive, and he has been able to support his views with histological studies from his cases. He also reviews very thoroughly the literature of the hyperkeratoses.

His Tafel vi, Figs. 1 and 3, shows how exactly his cases resemble mine, though it appears that the inclosed areas in his are thinner—atrophic-looking. There are no "disks" in mine which can be compared to his. There was no limiting "seam" or "wall" about the smallest hyperkeratosic lesions in mine. The "grat" in the center of his wall, as well as the little plugs in its middle and elsewhere, are practically the same as mine, but the central thread in mine was not higher than the sides of the seam—generally it was depressed a little.

A comparison as to development in my case is impossible, because the patient does not know, and I have not been able to obtain any information of his mother, who lives in another State. The only information he obtained from her is regarding the original "seed-wart" lesion. The lesions in my case are much fewer than those in Mibelli's, and they appear to have reached quite a fixed state of development.

As in the "Atlas" case, so here I have to say that the hairs persist in my large patches, none in his; his included skin appeared thinner, mine not different from that outside the "seam." The only small

lesion in my case with a peculiar border is the palmar one, so well shown in the plate, with its fine "seam," level, and its longitudinal, blackish-yellow thread and the blackish globules.

A brother of Mibelli's man also had the disease, beginning at eight; a sister, slightly, beginning at two. His Case II—Monatshefte—had lesions on the forehead, face, temple, mastoid region, and neck; also on the extensor surfaces of the forearms, dorsi of the feet and fronts of legs. The "wall" was well marked, also the thin "grat" or ridge. Many of the small lesions had the circumscribing "wall." There were only a few illy developed lesions over the flexors, and his only palmar lesions—on the fingers—were not different from ordinary callosities.

The disease began very late in this case, first on the hairy regions. Case III was much like Case I.

Having no histological specimens from my case, I feel that I am not at all competent to follow Mibelli and Respighi into that part of the subject which deals with the pathology of the disease, save in so far as the clinical features of my case would warrant an opinion. But an opinion under such circumstances can be of little value, and it is best to wait in hope of obtaining material at some future time, with which to study out the pathology of my case and thus get comparisons. My case would not be published even at this late day without the histology if I saw any near prospect of getting specimens, and having to publish it without this necessary part is the source of much regret to me.

Respighi (Monatshefte für praktische Dermatologie, Bd. xviii, No. 2; Ueber eine noch nicht beschriebene Hyperkeratose) mentions five kinds of lesions in his seven cases. His illustrations and descriptions would not alone enable me to identify his cases with mine, though I am able to see a slight resemblance between his and Mibelli's cases. He mentions a special border, but no "dike," "wall," or "seam."

Mibelli (Monatsh. für prak. Derm., Bd. xx, No. 6, Ueber die Porokeratose, neuer Beitrag) says that Respighi's cases are substantially the same as his. This article is mainly an answer to the criticisms of Tommasoli (Comment. clin. d. mal. cut. e gen.-ur., ii. Jahrg., No. 1, 1894). Without having read Tommasoli's article, but with the knowledge of my own case, and with the complete descriptions of Mibelli, I have no hesitation in believing that the former is mistaken in his views, and that he has been perfectly answered by the latter. I think a simple clinical view of the cases would convince Tommasoli

that the process is altogether different from the conditions with which he has endeavored to identify it. The disease is undoubtedly one hitherto unrecognized, having an identity of its own, the clear definement of which we owe to Mibelli.

Mibelli's description of a primary lesion is interesting, but I can not follow him with my case, because there are no lesions which can be positively compared with his as types of the original condition. I agree with him that the "seam," or "wall," composed of two lateral halves separated by a central, longitudinal line, thread, or "grat"—crest—with its horny globules here and there, constitutes the only distinctive and typifying element in the disease. The various lesions of hyperkeratosis, without the bordering "seam," are not characteristic. Absence of hyperæmia or inflammation is a symptom. Figs. 1, 2, and 3, in the last-quoted Monatshefte article, show clearly the appearance of Mibelli's "wall." The central part of mine has less elevation. See diagrams of mine (Figs. 3, 4, and 5). The plaque form, with visible hyperkeratosis within it, and the horny concretions, was present only on the palm in my case, and the limiting "seam" there is non-elevated, simply a fine fissure with its median "thread" and "globules." The normal, extra thickness of the epidermis in the palm would prevent a well-marked elevation of the "seam."

The reader who is interested in following Mibelli's histological studies and his arguments is referred to his articles, which it would be superfluous to introduce here.

The British Journal of Dermatology, November, 1895, has a selection from an article by E. Respighi, Hypokeratosis Eccentrica (Il Jour. ital. del. mal. ven. del. pel., fasc. 1, 1895). The limiting sulcus described doubtless corresponds to the "seam" in the palmar lesions of my case. Respighi says the term "porokeratosis" is too limited. His name describes better the clinical course of the disease, while Mibelli, on the other hand, has constructed a strong chain of histological evidence to support *his* name. In my case some of the hairs were unaffected, some were broken in the follicles, the latter showing horny plugs, and there were also those remarkable white ones. Sweating did seem to be absent in my case, but the large patches remained soft, slightly moist to the touch.

My patient first showed the disease in the *palm*, where there are no hair nor sebaceous gland; so, at least, the hairs and sebaceous glands did not furnish *its* starting point. That the disease began in the sweat glands I have no proof, though it began there if in any appendage of the skin.

The editor who made the last-quoted selection, in the British Journal, is somewhat arbitrary in his objection to a special name for the disease. It is as clearly entitled to a special name as the verrucæ, ichthyosis, keratosis pilaris, pityriasis rubra pilaris, or any other well-defined disease.

As to Mibelli's name for it, it is well justified by his histological discoveries, but further study may necessitate an expansion of the term. All of the epithelial structures are involved in the progress of the lesions, and unless the process is shown definitely and absolutely to begin only in the sweat glands "panokeratosis" or "poro-panokeratosis" must take the place of "porokeratosis."

In the British Journal of Dermatology, February, 1896, another editor appears to have "abstracted" the same article of Respighi, using some different parts of it than are quoted in the November, 1895, issue. The father of the patient had similar lesions. In each quotation the patient is stated to have had the disease for forty years, for thirty years of this time limited to a single patch on the under surface of the left *heel;* other lesions developing in the last ten years. Lesions were on the soles, the palmar surfaces of the fingers, and on the scrotum. It may not be amiss to mention that during the past winter I saw a peculiar, narrow, raised line on the scrotum of a little negro in my clinic, but he disappeared before I could examine it. The description of Respighi's case may have been marred in the translation, or its clearness sacrificed to editorial brevity; but as I get it, it is of no value in identifying the disease as similar to that under consideration.

It only remains to be said, in conclusion, that mine is the only case upon record which has been observed and identified outside of Italy. The records of my case antedate the publication of Mibelli's, but he has the full credit of having worked out the disease and of having been the first to publish his conclusions.

My case is the only one which has typical lesions on the *palm* and which *began* on the palm. One of Respighi's had its beginning on the under surface of the *heel*, but I have not been able to find whether it was a typical lesion. The palmar lesions in several of the cases present, when taken alone, nothing characteristic of the disease.

Much which I might have said at the end of this article has been interspersed among the comments made, in the body of the paper, upon the other cases. It might be repeated that further study is necessary to the final naming of the disease, to decide whether we shall denominate it "porokeratosis," "hyperkeratosis eccentrica," "pano-

keratosis," or "poro-panokeratosis," with or without the adjective "eccentrica."

LITERATURE.

1. International Atlas of Rare Skin Diseases, October 28, 1893.
2. Monatshefte für praktische Dermatologie, Band xvii, No. 9.
3. Monatshefte für praktische Dermatologie, Band xviii, No. 2.
4. Monatshefte für praktische Dermatologie, Band xx, No. 6.
5. The British Journal of Dermatology, November, 1895.
6. The British Journal of Dermatology, February, 1896.
7, 8. Some Italian literature, perhaps, of which I have no knowledge, and a reference in Unna's Histopathologie der Haut.

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