PURPURA ANNULARIS TELANGIECTODES (MAJOCCHI'S DISEASE)*

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In 1887, Majocchi for the first time observed, and in 1896 for the first time published, a report of a hitherto undescribed dermatosis, "purpura annularis teleangiectodes," and enlarged on it, in 1898, in the *Archiv für Dermatologie und Syphilis*, as a contribution to the Festschrift for Pick. In a memoir to the Royal Academy of Sciences of Bologna, in 1904, he collected reports of his cases, seven in all, and published them in book form in honor of Pick's anniversary. In 1912, at the International Congress of Dermatology and Syphilis at Rome, he reported a further series of six cases,¹ so that the first observer of this unrecorded skin malady had a collection of thirteen cases to report within a period of twenty-five years.

This rather small number evidences the comparative rarity of the disease. It was fully eight years before another observer, Brandweiner, published an exhaustive paper based on a case presented two years previously at a society meeting, including two new cases. The attention of dermatologists was now awakened and reports soon followed by Arndt, Herxheimer and Kohler, Balzer and Galoup, Vignolo-Lutatti and other observers, up to 1914. In 1912, Brandweiner and Lindenheim, in critical papers, contributed largely to the study of this interesting skin affection; but by far the most valuable contribution to this subject was rendered by George M. MacKee² of New York, who in 1914 and 1915 made a profound study of this disease entity, of which thirty-eight recorded cases, including his own, were known, and published it in a monograph which, as to completeness of illuminating features, critical review of all hitherto observed cases, and complete bibliography, is the most classical exposition of this topic and a fountain-head for future investigators.

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1. Majocchi: Tr. VII, Internat. Cong. Dermat. and Syph., Rome, 1912, p. 818.

2. MacKee, George M.: Purpura Annularis Teleangiectodes, Monograph in Three Parts, J. Cutan. Dis. **33**:129, 186 and 280 (March, April, May) 1915. For a complete bibliography the reader is directed to this article.

In the present paper I shall present two cases, one of about two years' the other of about one and one-half years' standing. They possess, both clinically and histologically, the main classical features set down by Majocchi. They deviate, as will be noted later on, in minor details, or show them in a less conspicuous manner. I shall endeavor to plead for a broader view of the dermatosis, by no means in a destructive but rather in a constructive manner. In accepting the diagnosis, one should consider that though there is a lack of well developed annular arrangement of the lesions, or well developed atrophy, there is the achromia and alopecia of the affected parts as well as other clinical and histologic features to demonstrate amply their identity with the dermatosis in question.

We cannot in reason expect a perfect conformity of symptoms to exist in any disease-type, which would in every instance tally with the description given by the first observer. A syndrome is not immutable; it may change in the direction of a plus or minus, part of it may be wanting entirely or may be slightly manifested, or one or the other of its component parts may outshine the others and stand out prominently. There are, however, in all such cases of this dermatosis, enough of the classical features to assure them the right to be included under the category of *purpura annularis teleangiectodes*.

REPORT OF CASES

CASE 1.—*History*.—A man, aged 53, a Russian, residing in the United States four and one half years, and married thirty years, father of five children, all of them well, presented a negative family history, and had always been well up to the beginning of the present affection, which had lasted about two years. On the anterior portion of the lower half of the left leg, the patient had noticed a patch the size of a silver quarter; it was bluish, dry, not raised, and was unaccompanied by any symptoms. There were no rheumatoid pains and no enlarged veins present at that time. (The patient was very definite about the last statement.) Within a month, a similar patch, at about the same location, appeared on the right leg. He went to a dispensary and was treated with ointments, but in spite of treatment the area spread in size and new lesions appeared during this time. At the end of about six month, enlarged veins began to appear on both legs; first on the right, later on the left. The general condition of the patient at the onset of the present condition was absolutely negative. At no time had the lesions ever entirely disappeared. The patient stated that there was a quarter-sized, bluish patch on the left leg for about three months. After treatment for about five or six months, the patch became sensitive and itchy. Minute reddish spots varying in size from a pin-point to a pin-head began to appear about the original lesion, extending to the middle of the leg. These spots slowly turned brown, and for the past five months had been gradually turning yellowish. Those on the middle of the leg were entirely gone, only a dark brown discoloration marking the site of the previous lesions. For the past three months no new lesions had appeared; at no time

was there any inflammation, infiltration or hyperemia present, nor were there any ulcers present at any time.

Examination.—The patient, a man of small stature, was moderately well developed, of good musculature and a moderate amount of fat, and with a healthy color of the skin and mucous membranes. There was no adenopathy; the superficial veins were moderately well developed throughout. Extensive follicular keratoses were present all over the body; on the legs these were scant except on the upper part where the keratoses intermingled, though distinctly separated from the lesion to be described. The chest, abdominal organs and urine showed nothing pathologic. The Wassermann and tuberculin tests were refused. The patient denied any infection and did not remember having had any of the diseases of childhood. He had not been sick and did not complain of any illness.

On the lower third of the anterior surface of the left leg there was an irregularly circular patch, about 3 inches in diameter, mottled in appearance and predominatingly dark brown, containing an aggregation of discretely scattered or grouped dark red to dark brown spots, varying in size from a pin-point to a pin-head, which did not disappear on glass pressure. Some of them, on closer observation, showed a minute network of dilated capillaries. In putting the skin on the stretch there was a noticeable tendency for some of these spots to be arranged in circles and half-circles, and some coalesced to form irregular groups. In the centers of these circles and half-circles the skin was smooth and easily wrinkled, and when a fold was raised it was felt to be thinner than its surroundings. Some of these minute lesions were palpably raised above the surface of the surrounding skin. In the center of this area was a scar marking the site of a biopsy. In this scar area were a number of slightly raised, pin-head to pin-point sized, reddish elevations, the color of which did not disappear on glass pressure, almost resembling the apple-jelly nodules of lupus, though smaller, less succulent and somewhat redder in tone.

The margin of the patch described above was not sharply circumscribed but shaded off into the surrounding skin, extending down to and over the inner malleolus, less so over the external malleolus. Here we found minute reddish spots on normal, not inflamed nor discolored skin; these spots did not disappear on pressure.

Between the Achilles tendon and the internal malleolus, the skin was discolored in patches, yellowish, with a light brown tone, containing numerous pin-point to pin-head sized spots which by coalescence showed a tendency in places to slight configuration. Directly under the malleolus was a distinct annular lesion, composed of similar elements; on stretching the skin in this region, one could notice with a lens, an extremely fine network of blood vessels.

Above this primary area, occupying the middle and lateral surfaces of the leg, there were innumerable reddish dots, extending to the patella and over it, and downward over both malleoli. Posteriorly, from the insertion of the Achilles tendon, extending over the calf and well up to the popliteal space and invading the whole circumference of the whole lower half of the thigh, there were also a number of patches composed of innumerable brown red spots; some of them involuting with a change of color to yellowish brown, and with a tendency to circle and half-circle formation. These spots, in the area of yellowish discoloration, were not palpable, while the multitude of discrete puncta on the anterior surface were *palpable* as well as *visible*, and distinctly separated from the follicular keratosis mentioned before (Fig. 1).

Fig. 1.—Lesions in Case 1.

On the inner surface of the leg and lower half of the upper part of the thigh, these discrete puncta were similarly found in groups, showing a tendency to incomplete circular arrangement. There were no lesions on the trunk or upper extremities.

The lower third of the right leg, starting above the malleoli and upward toward the midcalf, posteriorly, showed a brownish-yellow discoloration, especially on the inner surface. Within the area just outlined were innumerable puncta, reddish yellow on the external surface, while on the inner surface they were reddish blue. The color disappeared on pressure. These puncta were only visible, not palpable, while on the upper portion of the leg, where there were innumerable discrete and slightly annular lesions, situated on more or less normal skin, they were palpable as well as visible. On both sides of the Achilles tendon, near its insertion, were a few of these scattered puncta. Scattered throughout the dark-brown, discolored areas of the middle of the legs, there were a few penny-sized, whitish, circular patches, of which I shall speak later on (Fig. 2).

CASE 2.—History.—A woman, aged 44, Russian, married, with no history of any special previous illnesses, had had the eruption about one and one-half years. The affection began on both legs; on the lower third of the anterior surfaces she noticed a group of reddish spots, varying in size from a pin-point to a pin-head, situated on normal skin. In the course of a few months the lesions began to fade and the surrounding skin turned yellow, with new similar lesions appearing above and extending upwards to the middle of the leg. The patient did not complain at the time of onset, but now complained of itching. The general condition of the patient was good and the internal organs were negative. In this case, also, a Wassermann and tuberculin tests were refused after the biopsy was made.

Examination.—On the anterior surfaces of both legs, involving the middle and lower thirds, the skin showed a yellowish discoloration with numerous minute, in places grouped, mostly discrete, reddish spots. The site of the original lesions showed only a yellowish discoloration. There was no brownish discoloration or any evidence of hemorrhage whatsoever. The reddish spots did not disappear on pressure. The patient had noticed, for the last few months, a tendency to develop superficial veins on the upper half of the legs, but these caused no discomfort. It is worthy of recording the fact that the varicosities began some months after the onset of the lesions under consideration.

MICROSCOPIC REPORT

In Case 1, three biopsies were made: (1) Group of minute, reddish, pin-point spots in apparently normal skin, representing the most recent lesions. (2) Group of slightly raised, reddish spots surrounded by moderately discolored skin (yellowish brown), representing fully developed lesions. (3) Site of previous grouped lesions, only yellowish discoloration present; representing involuted lesions, part of area first involved when the disease began.

In Case 2, one biopsy was made: Group of reddish spots on yellowish skin, representing developing lesions, though present for some months.

Low Power.—In case 1 the section of group 1 shows dilatation of the lymphatics and edema of the entire corium, more marked in the upper part. There are more or less sharply circumscribed groups of vessels (small and somewhat dilated) in the subpapillary zone, surrounded by a moderate amount of cell infiltration. The surrounding connective tissue is reticulated as well as edematous, especially about these groups of vessels. The epidermis appears thinned, the rete pegs and papillæ are practically wanting.

Fig. 2—Right leg, Case 1.

In the section of Group 2 the lower corium the deep vessels just above the fatty tissue are dilated, the walls are somewhat thickened, and there is a moderate amount of perivascular edema. The perivascular lymphatic spaces are somewhat dilated. In the midcutis there is a group of closely aggregated, variously sized, somewhat dilated vessels, showing slight edema and with no cell infiltration about them. (Here there is already evidence of telangiectasis in the communicating branches, between the lower and upper systems of cutaneous vessels.) In the upper cutis or subpapillary zone, there are groups of large numbers of vessels, some dilated; in some the lumen is almost occluded. The walls of a great many are swollen, in a few, thinned; all are surrounded by a well marked

Fig. 3.—Location of pathologic process in upper corium, just beneath the epidermis. There is edema of the connective tissue and dilatation of some of the deeper vessels.

cell infiltration, containing, in places, a large amount of pigment granules, the entire pathologic process lying within moderately edematous, reticulated connective tissue. In one place, surrounding a group of vessels, is an irregular space, with ragged walls; within this space are free blood cells and some pigment.

In the deep corium the vessels are markedly thickened. In the midcutis very few vessels are present, and there is no tendency to group formation. In some areas the vessels can hardly be differentiated, on account of the number of infiltration cells. Those vessels which can be seen, have apparently thickened walls. The upper lymphatics are all dilated; pigment

granules are few in number; the epidermis as a whole is thinned, with absence of rete pegs and papillae. The connective tissue, as a whole, in places appears somewhat condensed, so that the fat appears up half way to the epidermis. The connective fibers in places are somewhat reticulated.

In the section of Group 3 the deep vessels are moderately dilated, their walls vary in thickness, but all are thickened. The connecting branches in the midcutis are dilated, the walls somewhat thickened and surrounded by dilated lymphatic spaces; no cell infiltration is present. In the upper cutis, across the entire section are groups of vessels, from two to five in each group; some are dilated, the walls apparently thickened; the connective tissue about them is reticulated;

Fig. 4.—Groups of vessels in upper corium surrounded by moderate amount of cell infiltration; dilatation of lymphatics and deeper vessels; edema of connective tissue and of epidermis.

there is very little cell infiltration; the lymphatic spaces are all dilated. The connective tissue as a whole is edematous and appears fragmented. The epidermis as a whole is thinned; the rete pegs and papillae are wanting.

High Power.—In Case 1, Group 1, the corneous layer is present throughout, is slightly thickened and has a tendency to lamination, especially in the depressions marking the probable sites of follicles. The keratohyaline layer is present throughout, and is composed of two layers of cells; the granules are very fine and difficult to distinguish. In the prickle cell layer in a few places there is evidence of moderate interstitial and parenchymatous edema; a few wandering

cells are present. The basal cell layer as a whole is disorganized, especially over the groups of vessels. The connective tissue in its subepidermic portion is finely reticulated, with a moderate degree of granular degeneration (very fine granules); the elastic fibers are fragmented, and for the greater part wanting. This granulated and reticulated appearance of the connective tissue is present about the groups of vessels, but in these areas there is more edema and dilatation of the lymphatics and spaces. The deeper connective tissue is edematous, the finer structure no longer distinguishable; the fibers appear more or less homogeneous and glassy. The lymphatics are dilated. The connective tissue cells are moderate in number in the upper part, and comparatively few in the lower part.

Vascular Apparatus: One of the deeper vessels, of moderate size, shows its lumen narrowed down to a small crescent, in some of the sections; in others, the lumen is no longer present. It is occupied by a globular mass composed of connective tissue cells, with little interstitial substance, the cells apparently derived from the subendothelial connective tissue. The adventitia is moderately thickened, due to edema—endarteritis.

Fig. 5.—Higher power of preceding illustration showing groups of vessels with their indistinct homogeneous wall, edema of the surrounding tissue

Upper Plexus, in Subpapillary Zone: Lying within an edematous, reticulated connective tissue are groups of vessels of various sizes and varying numbers in each group, some close together, with small bands of reticulated, connective tissue between them. The lumen of some is markedly narrowed, owing to the edematous condition of the walls. The individual structural elements cannot be differentiated; they appear somewhat homogeneous and do not take the stain properly (hyaline degeneration?). In some of the vessels the endothelial cells are swollen. In none of the vessels is there any proliferation of the intima, no thickening of the adventitia, and only a swollen condition of the entire wall, as described. Within the edematous, reticulated connective tissue in which these vessels lie is a moderate number of cellular elements, mainly fixed connective tissue cells with a few round cells. The granular appearance of the connective tissue noted before is more marked in this vicinity than elsewhere. Nowhere is there any evidence of pigment or blood cells (hemorrhage), or any rupture of vessel walls.

Summary: The tissue is characterized by groups of vessels of varying size and number, lying within a reticulated, edematous, degenerating, connective tissue, in the meshes of which are a moderate number of cells. The walls of the vessels are swollen, and appear homogeneous; there is no evidence of endarteritis or of hemorrhage. The deeper vessels show distinct endarteritis. The epidermis is thinned, probably because of the edematous condition of the entire connective tissue.

In Group 2 the epidermis is similar to the preceding. The subepidermic portion of the connective tissue is reticulated, fragmented and granulated. From the midcutis and deep cutis it is moderately edematous, with dilatation of the lymph-

Fig. 6.—High power of groups of vessels associated with cavity with its ragged walls, containing pigment and cell elements. Some of the vessels are filled with blood cells. The structure of the walls is indistinct. The endothelium in some vessels project into the lumen; in some the lumen is dilated, in others it is narrowed. There is reticulation of the surrounding connective tissue.

atics. The entire connective tissue appears to be broken, fragmented in varying sizes, and in the upper part and about the upper vessels, very finely granular. The elastic tissue in the upper cutis and about the vessels is fragmented, and in the subepidermic region, practically wanting. In the lower cutis it is broken in large strands. Nowhere in the sections is the elastic tissue normal. The vessels of the deep cutis in the fatty tissue show a fairly well marked panarteritis,

whereas in the tissue described above only endarteritis is present. The vessels just above the fatty tissue show a thickening of the walls, though no endarteritis or panarteritis (edema). The communicating branches of the midcutis are in a finely reticulated connective tissue. The walls of the vessels are swollen and appear somewhat homogeneous, the endothelium swollen. There are only a few infiltration cells present. No hemorrhage or pigment is present in this region. The groups of vessels of the upper corium lie in a reticulated, edematous, degenerating (granular) connective tissue, containing varying numbers of fixed connective tissue and round cells. The walls of the vessels are swollen and homogeneous and take the stain not as well as in the previous tissue. Scattered throughout the entire upper corium are very numerous, larger and smaller groups of pigment granules, most abundant in the region of the groups

Fig. 7.—High power of cavity showing its walls, contents and vessels. The walls of the cavity are composed mainly of broken and ragged fragments of connective tissue. On the right side, group of vessels seen in the preceding slide filled with blood; the one projecting into the cavity shows a disintegration of one part of its wall (rupture?). In the cavity are numerous clusters of pigment granules, red, white blood cells and fragments of connective tissue. The connective tissue is for the greater part fragmented and granulated. The lymphatics are dilated. Section of fully developed lesion.

of vessels. In one area, about a group of ten vessels, is an irregular cavity with ragged walls, composed of broken and fragmented connective tissue. On one side, the wall is made up of a number of somewhat dilated vessels filled with blood cells. The outer wall of one of these vessels appears as though

ruptured. In the cavity and along its ragged walls are many groups of pigment granules, also a moderate number of scattered red and white blood cells and masses of degenerating connective tissue.

Summary: The process is further advanced than in the tissue previously described. There is a panarteritis of the deeper vessels; there are telangiectatic communicating branches and further degeneration of the upper telangiectatic vessels, with the presence of pigment and cavity formation, containing pigment and blood elements. The granulation (degeneration) of the connective tissue is somewhat more marked, as is the breaking up of the elastic fibers.

In Group 3 the section shows panarteritis, with degeneration of the vessels in the deep cutis, whereas in the previous tissue only panarteritis was seen. The

Fig. 8.—Endarteritis of the deep vessel with proliferation of the intima; from early lesion.

vessels in the midcutis show the lumina for the greater part almost closed. The other changes are as before noted. Most of the lumina of the upper cutis are narrowed, the connective tissue is more fragmented, and more vessels appear to be filled with blood cells, with vessel walls thinned. The connective tissue as a whole is more disorganized. The coil glands and muscles show evidence of degeneration. No evidence of hemorrhage or pigment is present.

Summary: The process is more advanced, with further degeneration of the parts enumerated in the last tissue

In Case 2 the findings are similar to those present in No. 1, excepting that the granulation of the connective tissue is somewhat more marked.

COMMENT

In analyzing the features of these cases, especially Case No. 1, to which I shall devote most of my attention, one can observe a coexistence of two of the customary three stages, namely, the telangiectatic and hemorrhagic-pigmentary; the atrophic stage is expressed both clinically and histologically in a slight degree only. Neither the arrangement nor the involution of the lesions conforms fully to the classical description as given by Majocchi, but the main clinical and

Fig. 9.—Same vessel in different section.

histologic features agree with his and the findings of all observers. I am able to add some minor features not mentioned by Majocchi, as slight *elevation* of the lesions above the surrounding skin, and confirm another feature (achromia) not observed by successive authors.

The center of the ring or figured lesions, and the places where the discrete lesions were involuting, showed a smooth, silky, often glistening surface with an alopecia here and there.

The distribution of the eruption in my case shows the total ring form only sparsely represented. Outlines of fading, ill-defined,

annular lesions, in the form of semicircles, serpentine lines, and also in grouped and linear configurations, are discernible. These spots exhibit all the retrogressive color changes of seeming hemorrhage. The most recent ones are bright red with a just discernible capillary ectasia, while the older ones are dark red and do not pale on diascopic pressure. The older ones show a gradual fading into brownish red or yellowish red. Telangiectatic, punctate, hemorrhagic and pigmentary spots are found throughout, representing two of the three classical stages, as mentioned before.

Fig. 10.—Panarteritis of deep vessel with degeneration of the wall, late lesion.

As to the annular arrangement of the lesions, which, in the original presentation of the disease occupied the third place of Majocchi's seven requirements, while they had been found and more or less noted by all observers, yet their presence is not an indispensable attribute of the eruption, as the title of the dermatosis would imply.

Brandweiner was the first to draw attention to this deviation, as did Ferrari, who speaks of "patches of all forms." Majocchi, in 1912, stated that the lesions were "mostly" annular. Vignolo-Lutati

was unable to observe a conversion of punctate lesions into an annular one. Ossola speaks of the lesions also occurring in linear formation; Lindenheim also mentions irregular arrangements; Noble speaks of solid disks or patches of various size; MacKee's case presented on the left leg mostly discrete and also grouped lesions. In my case there were indications of annular arrangements, some of them coalescing into gyrate configurations, but for the most part, the discrete and grouped lesions prevailed.

Fig. 11.—Dilated lymphatic with contents.

An involvement of the hair follicles or other appendages of the skin could be observed neither clinically nor histologically. That this involvement is not one of the necessary requirements is evidenced by Majocchi's statement in the first point of his characteristics of the disease, that the hair follicles are "usually" involved. (This was noted by only a few observers.)

In this case I wish to emphasize especially the extensive dark brown, yellowish brown, and yellowish green discoloration, and the silky sensation to touch, at the site of the fading old lesions, either figurated or grouped. As described before, a smaller part of the eruption, judging

from the appearance of formerly affected areas, must have developed in ring formation, in the earlier part of their life history. Within and outside of the dark brown discolored areas (occupying the front and lateral regions of the left, and in a lesser degree, the right leg) there are round clearings the size of a copper cent piece, visible within a dark frame of extensive pigmentation, or like rounded, whitish islands, in an area of dark brown discoloration. These round spaces or clearings—the probable sites of former annular lesions—present a striking

Fig. 12.—Granular degeneration of the connective tissue below epiderm.

whiteness, resembling a rounded vitiligo spot within the confines of its dark pigmentation. They show the characteristic wrinkled, cigaret paper-like appearance, are smooth and silky to the touch, and are devoid of hairs. This achromia, mentioned by Majocchi in Point 7 of his description, has been noted by only a few observers. Lindenheim mentions that "the pigmented areas are not solid, but contain minute clear spaces." In the case under discussion, the clear spaces had, as mentioned before, the size of a cent piece. In Lindenheim's case, these interspaces enlarged until all that was left of the lesions were a few

grouped "freckles" (disappearing pigment). In my case the white spaces had not as yet demobilized the surrounding pigment to such an extent.

The yellowish green hue of the skin occupying the site of the once grouped and discrete lesions is still in evidence. This peculiar discoloration is mentioned by almost all observers. It extends over different sized areas of an inch or more in length and width. In some of these areas, traces of involuting spots can yet be discerned. Although they look as if caused by disintegrating pigment, and have

Fig. 13.—High power. Parenchymatous edema of prickle cells and degeneration of connective tissue.

been defined as pigmentations, a careful study of the sectioned tissue shows no trace of blood extravasation or pigmentation. The discoloration is due to a distinct degeneration of the *connective* and *elastic* tissue, similar to that seen in pseudoxanthoma elasticum. In searching the available literature I have not found any special histologic reference to this degeneration of the connective and elastic tissue as the cause of the yellowish tone, noted clinically. To my knowledge, this is the first attempt to explain this clinical picture.

A fact worth mentioning is the reappearance of lesions at the site of one of the biopsies. This would amplify the statements of Brandweiner and Ossola. The former succeeded in causing the appearance of red puncta by scratching or scraping the skin. The latter, by applying an elastic band over the arm. Although not typical for the disease in question, this phenomenon shows only an increased vasomotor excitability of that particular skin.

Provocation of lesions, similar to already existing parent lesions, is frequently observed in other clinical conditions, as in lichen planus.

Fig. 14. Adnexa, edema of connective tissue about them.

urticaria, etc., in individuals so predisposed. It is this predisposition, and not the trauma itself, that does the provoking. Therefore I do not think that Brandweiner's contention is tenable, that trauma and the predisposition created by a vasomotor excitability may cause the affection in question. I am rather inclined to assume that—analogueous to the etiology of erythemas like pityriasis rosea,³ and erythema mul-

3. Weiss, Ludwig: Pityriasis Rosea, and Erythematous Eruption of Internal Origin, *J. A. M. A.* **41**:20, 1903.

tiforme, etc.—pathogenic bacteria of different kinds, absorbed by the circulation, are causing disturbances in the skin. The sudden outbreak, the petechial character of the eruption, its cyclic course (the three stages), its disinclination to recurrences, all point toward a toxic origin. In the transmission of the circulating toxins, alterations of the vessel walls take place. Naturally, the process will invade the vessels of the deeper strata first and, extending its force, the main lesions will be found there and only later the vessels of the upper portion of the skin, with its surrounding tissue, will be affected. This is

Fig. 15. Coiled glands—vacuolization of some of the lining cells and cloudy swelling of the cells (parenchymatous edema).

exactly what happens in the development of *purpura annularis telangiectodes*. It seems as if a toxic and angioneurotic process can eventually cause mild inflammatory changes in the skin, as evidenced in the different erythemas, urticarias, and also slightly in the affection under discussion.

The scanty extravasation of blood cells cannot possibly cause the clinical observable hemorrhage. All observers noticed the discrepancy between the clinical and histologic findings in this respect. Some

explain this as an optical delusion due to the fact that the superficial, thrombosed capillaries are not emptied entirely by the diascopic pressure and are therefore taken for the tiny hemorrhage visible. In those rare instances in which a rupture of the vessel-wall had taken place, the clinical evidences of hemorrhage will find their natural explanation.

As to the etiology of the affection: Almost all diatheses have been suspected and ruled out — tuberculosis, syphilis, rheumatism, gout, diabetes, heart affections, hemophylia and lead poisoning — especially the latter were suspected in the cases of Brandweiner and Lindenheim.

Fig. 16.—The elastic tissue and degeneration of the connective tissue (special stain, Weigert elastic stain).

Their patients were printers. But this may have been a coincidence, as the other patients had widely different occupations and environments.

The male sex, according to MacKee's tabulation, contributed thirty-one cases of the recorded thirty-eight. That twenty-three of the thirty-eight patients were Italians is probably due to the fact that through Majocchi's publications Italian dermatologists were on the alert for such cases. Since MacKee's monograph on this affection was published a wave to recognize this disease has been permeating American dermatologists.

Autointoxication as the causative agent has as yet the largest number of adherents. With the finer laboratory technic and more extensive knowledge of metabolism and of focal diseases, it seems that this direction offers a favorable working basis for further investigations.

The treatment will and can be only symptomatic. Rest and an elevated position of the limb in the acute onset, with the use of embrocations of Burrow's solution will prove helpful. To increase the viscosity of the blood; calcium lactate should be exhibited, and later iron

Fig. 17.—Loss of elastic tissue. The elastic tissue in the upper cutis and about the vessels is fragmented, and in the subepidermic region practically wanting. Nowhere is the elastic tissue in this section normal.

and strychnin administered. The use of a porous weave bandage without rubber to exert an even pressure will prove rational.

There has been some controversy in reference to certain stasis-dermatoses presenting a few clinical features of the affection in question. Klotz,⁴ in 1904, described a case of chronic symmetrical diffuse hyperemia of the extremities, also calling it erythromelie. Here the

4. Klotz, Hermann: A Case of Chronic Symmetrical Hyperemia of the Extremities (Erythromelie), *J. Cutan. Dis.* **22**:170, 1904.

main features were redness over the fingers and toes, spreading over the hands and feet, arms and legs; a network of dilated capillaries, the color fading on pressure; the affected parts show all stages of hyperkeratosis, from the formation of an insignificant xeroderma to the formation of horny shields, resembling fish-scales, associated with thickening of the skin; no pigmentation or discoloration is present. There is very little resemblance in this description to the disease in question. Majocchi's disease is characterized by grouped or annular lesions, associated with pigmentation or discoloration and no thickening, but with a tendency to atrophy. Klotz thought that the condition that he described was probably a vasomotor neurosis, allied to the local asphyxia or Raynaud's disease. The two cases of Herxheimer and Kohler were regarded by Blaschko and others as a stasis or congestive dermatoses, and a third one was considered as a transition case of purpura annularis telangiectodes. These stasis-dermatoses, which are found on the legs, associated with varicosities, are due to extravasations from the papillary venous capillaries, resulting from a thinning of the epidermis. This affection is also called dermatitis-cruis et varicibus by the Vienna school and the other kindred ones, like Schamberg's progressive pigmentary⁵ dermatosis,⁶ are clinically and histologically different from purpura annularis telangiectodes.

On the other hand, there are some unrecorded cases of what seems to have been Majocchi's disease, by Harris, Engman and Mook, and others.

CONCLUSION

The case in question presents the following deviations from the cases reported by other observers:

1. There is very intensive, dark-brown pigmentation over the middle of the legs, with whitish achromia in the center.
2. There are almost numberless discrete lesions, with the yellowish discoloration in the area of the fading spots, which discoloration is *not* due to pigment deposit, but to a degeneration of the connective and elastic tissues.
3. The evolution and the involution of the lesions and all their stages are observable simultaneously (coexistence of lesions).
4. A slight raising above the level of the skin is present, *independently* of the keratotic follicles.
5. The coil glands show evidence of a cloudy swelling and some degeneration, while the follicles are not involved.
6. Lesions appear at the site of a biopsy, which lesions in every respect are similar to the original ones.

5. Schamberg, J. F.: Brit. J. Dermat. **13**:1, 1901.

6. Kingery, L. B.: Schamberg's Progressive Pigmentary Dermatoses, Case Report with Histologic Study, J. Cutan. Dis. **36**:166 (March) 1918.

7. This fact shows almost to a certainty a vasomotor trophic influence on the peripheral vessels.

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ABSTRACT OF DISCUSSION

DR. MACKEE said that he had had several opportunities of observing the cases reported by Dr. Weiss, and that he could not identify them as examples of purpura annularis telangiectodes. As Dr. Weiss had mentioned, the differential diagnosis between examples of chronic purpura, such as Schamberg's progressive pigmentation, the so-called hemostatic dermatitis of Klotz, the common chronic telangiectasia of the legs due to deep-seated or superficial varicose veins, arteriosclerosis, cardiovascular and renal disturbances, and Majocchi's purpura annularis telangiectodes was often confusing both clinically and histologically. Dr. Weiss held a broader conception of the latter disease than did the speaker. In all diseases there were borderline cases, and until such time that cases like those so ably presented by Dr. Weiss were differently classified, it would seem preferable not to include them in such a definite entity as Majocchi's purpura. The main objections to such inclusion were the absence of clean-cut recurrent attacks, the paucity of annular lesions, the fact that lesions were always present, the presence of varicose veins and other causes of local purpura, and the absence of hyaline degeneration, histologically.

DR. ZEISLER thought these cases must be very rare, because in thirty-five years in dermatology he had not been called on to make such a diagnosis until recently, when in the Chicago Dermatological Society a case was shown which was thought to be that disease. The appearance was very much like a lichen planus moniliformis. His son and others who were more modern in their views of dermatology thought it was a case of Majocchi's disease, but he could not identify a case in thirty-five years' practice.

DR. WEISS thought that there were borderline cases existing. It must be remembered, however, that other purpuric conditions usually were accompanied by constitutional disturbances, fever and malaise, while in these so-called borderline cases, as well as in these of the true Majocchi type, the concomitant symptoms were negative. Some slight rheumatoid pains were present in some cases; but as Majocchi himself said, they were coincidental. There was a low grade of inflammation with a moderate pigmentation present in these borderline cases, including the stasis dermatosis.

In Majocchi's diseases there was no inflammation, no pain or other constitutional symptoms observable, and the pigmentation was extensive. He emphasized that the hemorrhage in the stasis dermatoses was due to capillary rhexis and diapedesis.

He tried to show—as he thought, for the first time—that the pigmentation in Majocchi's disease was not due to blood stasis and its sequelae, but to the degeneration of the elastica.

He felt that he was within the limits of the disease entity by counting his case as one of the Majocchi type, even if some of the symptoms were not sufficiently expressed. The syndrome of the first observer was not immutable: a plus here, or a minus there would invariably be observed. He thought that enough of the clinical and histologic features were present to declare the case in question to be purpura annularis telangiectodes.