THE BRITISH JOURNAL OF DERMATOLOGY.

FEBRUARY, 1902.

TWO CASES OF PRURIGO SIMPLEX CHRONICUS (BROcq).

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IN March of this year two women, a mother and her daughter, came under Dr. Unna's care on account of an extremely chronic and pruriginous affection, from which they had both suffered for many years.

I have to thank Dr. Unna for allowing me to publish the clinical details of the two cases, and also the results of a microscopical examination that I made of a piece of skin excised from the daughter.

The cases are by no means common, and they have not been much investigated. Clinically they are easy to recognise and fall into a natural group. The best description, that I know, of these cases is given by Brocq, and it is on this account that I have chosen the name given by him. I am well aware of the objections that can be raised to the name, and to a certain extent share them. The Prurigo simplex chronicus of Brocq is, according to him, simply the chronic manifestation of the P. simplex acutus, *i.e.*, Lichen simplex acutus of Vidal. The great majority of Dermatologists deny the separate existence of the latter, and without reserve class all the cases under eczema. I believe that Brocq is right in separating some of these cases from eczema, and I also believe the disease is manifested in both acute and chronic fashion. It is a question that admits of exact determination by histological examination, and, in fact, urgently requires it.

But when this point has been settled another difficulty remains. The wide difference of opinion among the representatives of the various schools as to the meaning of the word Prurigo, and the extension that can be given to it, makes it extremely hard to fix the position of cases such as I am about to describe. It would be quite out of place in a paper like this to discuss so wide a subject. It may be possible to make out if these particular cases bear any relationship to cases of Prurigo of Hebra, and that is all. It will then be understood that in adopting Brocq's name I do not necessarily adopt all his teaching on this question, and I use the term Prurigo more as a convenience, and in default of a better name. The whole subject requires careful and exact study. It may be that such cases will be finally and definitely rejected from the Prurigo group. It may well be, on the other hand, that further study may support much of the teaching of the French school, and that the present narrow and unsatisfactory definition of Prurigo may be widened in scope with great advantage.

CASE I.—Frau L., aged 47, a strong, healthy-looking, well-nourished, fair-skinned, pleasant-looking woman. She has always enjoyed good health. From infancy she has had a dry skin, much given to itching. She never sweats.

The present eruption commenced after the birth of her daughter, when she was 25, and since then has never entirely disappeared. Her state is always at its worst in winter.

When first seen the eruption was chiefly present on the trunk. There were a few papules on the arms and legs, but none on the face or on the scalp. The old papules were small, round, slightly elevated, and each was capped by a tiny reddish-brown crust. Two fresh papules were present. These were about the size of hemp-seeds, obtusely conical, of a pale red colour, with a paler, almost yellow, centre. The central pale spot certainly did not give the impression of containing fluid. The shape alone suggested that the papules surrounded a follicle. When they appear the papules itch violently, and are at once scratched. This gives instant relief, and is followed by the formation of the crust already mentioned. The crusts eventually fall off, leaving scarcely perceptible punctiform scars.

No obvious thickening of skin or excess of pigmentation was detected.

CASE II.—H. L., aged 22, daughter of the above patient. Like her

mother, she appears strong and healthy, makes no complaint of ill-health, and the only points of difference are that she is rather thinner than her mother, much more extensively affected, and that her skin is drier, rougher and more pigmented.

As a child also her skin was noticed to be dry. She never sweats, and the skin has always itched on the slightest provocation. Her skin is a little aggravated by cold weather, but to no great extent.

When five years of age she had red, dry patches all over the body which had no scales and were followed by the appearance on the back of water-blisters (? miliaria).

The present eruption appeared with the menses at the age of fourteen, and has never left her. It commenced on the neck and forehead, and now resembles that of her mother, only it is far more extensive.

The papules are most numerous on the trunk, and particularly over the whole of the back. They are present on the outer aspects of the legs and arms—scattered singly in the hairy scalp, and very abundantly round the hair margin—*i.e.*, on the upper part of the forehead, back of neck and parts behind the ears. In this way a band is formed. The papules run together, and produce an appearance resembling seborrhoic eczema as regards localisation and fattiness. With this exception, there is not the slightest suggestion of eczema either in this case or in her mother's.

The papules on other parts are invariably discrete, even where several occur close together. They exactly resemble those found on the mother, and need not be separately described.

Her skin feels and looks very dry. There is a general hyperkeratosis, and all the follicles project. In this she differs from her mother, whose skin appears of normal texture, and shows no elevation of the follicles. So, too, in this case the connection of the new papules with the follicles is evident, whereas in the case of the mother it had to be inferred.

There is the same history of itching and scratching.

A piece of skin was excised from this last patient from a spot near the angle of the scapula, where by good fortune an old and new papule were close enough together to be excised in one piece. The piece was hardened in alcohol and embedded in celloidin, and the sections illustrated were stained with polychrome methylene blue,

differentiated in glycerine-æther, and decolourised in absol. alcohol, cleared in bergamot oil and mounted in balsam.

They present for examination three points: (1) The presumably normal skin lying between the two papules; (2) the fresh papule with contents and surroundings; (3) the old, crust-covered papule.

1. *The Skin between the Papules.*—There are several points that attract attention and suggest a previously existing abnormal state.

The epithelium shows the folding that one finds in the neighbourhood of recent acute swellings. The horny layer is perhaps a little thickened and the prickle layer is irregular, being thin in some places and thick in others. The cells of the prickle-layer are often small, irregularly disposed and very closely crowded, so that they stain deeply. There is in fact a commencing acanthosis, and this extends to the follicles, which are in consequence most massive in appearance. Some allowance must be made for the situation of the follicles, namely, the back. But even then they are very large and striking.

In almost every instance the follicles are more or less blocked by horny plugs. This is well shown in Fig. A 1. There is other evidence of stagnation in that practically all the epithelium of the sebaceous glands is converted into fat cells. It is hard to resist the conviction that this hyperkeratosis of follicles with consequent stagnation of sebum is the elementary lesion, round which all the other appearances are grouped.

The acanthosis and hyperkeratosis extend to the upper part of the ducts, which below this appear exceedingly narrow and ill-developed, considering the age of the patient.

Unfortunately the piece excised, though cut fairly deep, did not include the coil glands. So a most interesting point is thus excluded from consideration.

These patients, according to their statement, never sweated and the skin was always dry. It is quite possible to imagine that the dryness was caused by the blocking of the ducts and follicles. The acanthosis in the upper part of the ducts must cause some obstruction, and if such a state existed in infancy it is also possible to imagine that in time the sweat glands would become atrophic.

On the other hand there is no evidence as to the state of the glands, and it is equally possible to assume some functional disturbance of them.

The blocking of the follicles, with the retention of sebum, certainly had much to do with the feeling of dryness.

2. *The fresh Papule*.—The beautiful illustration made by Herr Gummelt, of Hamburg, from a central section, shows, in the clearest possible manner, the anatomical features of the papule and renders a long description almost superfluous. One finds:—

(a) The centre occupied by a large oval plug of concentrically arranged horny layers, surrounding a mass of retained sebum. The plug fills and dilates the follicle and presses down on the gland below. The sebum extends down to the deepest recesses of the gland.

(b) The prickly-layer of the surface and that lining the follicle is greatly thickened. There has been an increase of cells (acanthosis), but the great part of the swelling is due to œdema of the cells (Parenchymatous swelling). The intercellular lymph spaces are not widened.

(c) An acute œdema of the sebaceous glands and infiltration of the same with leucocytes. The regular arrangement of the leucocytes shows that this is no abscess. The structure of the gland still remains and in places can be clearly seen. Deep in the glands one finds large plugs of sebum.

The swelling of the gland has forced the follicular walls apart from one another, and the gland lies in the space so formed.

At one point, B. 4, the leucocytes are just beginning to penetrate the wall of the follicle and to produce a dissociation of the cells.

(d) Marked œdema of the cutis and dilatation of lymph spaces. There is an increase of the perithelia of vessels and considerable infiltration of the tissues with leucocytes.

There seems to be a complete absence of mast cells.

(e) At the mouth of the follicle a few cocci are seen, separated from one another in such a fashion as to exclude the idea of recent activity. In the depths the only organism found is the Bottle Bacillus, and that in very large numbers. The organism can be seen in the sebum deep down in the gland. There is in the specimen illustrated one colony indicating active growth. In other preparations examined the same is found. The bottle bacilli swarm everywhere, and in some follicles are present in enormous numbers.

It is assumed that this fat-loving organism has no pathological

action. A single case like this proves nothing. And yet it might well be that the bacillus was not quite so harmless as is generally supposed.

So in the fresh papule the chief points of interest are:—

(1) The blocking of the follicle with retention of sebum.

(2) The acanthosis of the epithelium that lines the follicle, and of that part of the surface epithelium that surrounds the follicle mouth. The great œdema of the prickle cells is a conspicuous feature.

(3) The acute œdema and leucocytic infiltration, centred in the sebaceous gland, and so strongly suggesting a local cause for the same.

It will be seen that the changes found here are merely exaggerations of the conditions found in the unaffected follicles, with the addition of œdema and of leucocytes.

3. *The Old Papule with Crust.*—This presents at first sight a very different appearance, and yet a careful examination shows that the lesion is closely allied to that just described, and dependent on the same causes.

The central horny plug and dilated follicle are here missing—but there is the same retention of sebum—the same great increase of follicle- and surface-epithelium, and the same œdema and infiltration of the sebaceous glands.

In this instance three or four follicles lying close together are affected. They show a great degree of acanthosis, and have more or less joined to form a single mass of epithelium. Successive sections show clearly the implication of several follicles, and this is further shown in the sections stained for elastin, where remains of elastin are found embedded in the block of cells. This extraordinary increase of epithelium in the depths and fusion of contiguous masses is a most remarkable feature, and one that seems to place the disease in a class by itself.

The illustration was taken from a section that showed best the connection between the old and new papules. Other sections show far better the œdema and leucocytic infiltration of the glands, exactly resembling that encountered in the new papule, and only differing in that it is in a more advanced stage.

In this case blocks of necrosed epithelium are seen. The disassociation of cells that in the new papule is only just commencing has advanced much further, and now reaches the surface.

The second great interest of the specimen lies on the surface. The papule is crowned by a large vesicle, due, apparently, to a necrosis and solution of the cells of the prickle-layer. The vesicle is formed entirely at the expense of the prickle-layer. In the centre the whole layer is destroyed, while at the circumference the vesicle occupies the centre of the layer.

The contents have coagulated and form a crust, in which are seen granules of blood pigment. These are very abundant, and probably arise from disintegration of red blood corpuscles. No red blood corpuscles, however, are seen in the section.

In the centre the horny layer has in part disappeared, but all round the circumference it is well preserved.

It will be noticed that the crust does not project above the surface, even where the horny layer has disappeared. In this respect there is a marked distinction from eczema.

Where the horny layer has gone there are several clumped colonies of cocci, but as these are not found in places where the horny layer is intact it is possible that they have nothing to do with the causation.

In the centre can also be well seen the leucocytes coming from the depths. They appear in columns that apparently correspond to the mouths of the affected follicle. They are by no means evenly distributed through the crust.

Consideration of the appearances found :—

Apparently every stage of the lesion is represented in these specimens. There is the initial hyperkeratosis of the follicles and stagnation of sebum. Then also a marked tendency to acanthosis of the follicle wall and neighbouring epithelium. This helps to still further block the exit from the gland. As the result of some irritation there is a sudden œdema of the sebaceous gland, with a rush of leucocytes to the part. The œdema extends to the neighbouring epithelium. As a consequence the cells, already crowded, became much swollen (parenchymatous œdema) without any corresponding dilatation of the epithelial lymph spaces such as one meets with in eczema. Such a condition appears to lessen the power of resistance of the cell. Hence the addition of some trauma, *e.g.* scratching, is sufficient to determine a necrosis, a colliquation of the cells leading to the formation of the half vesicle, half crust so characteristic of the disease.

It is probable that the liquefaction of cells occurs without the intervention of scratching. Brocq has obtained a drop of fluid from these papules on puncture. Also the same evolution occurs in places that it would be almost impossible to reach by scratching. However produced, the contents immediately coagulate, and no further serum exudes. The crust lies on a level with the surface, and does not stand out boldly above it, as would happen in Eczema or Impetigo. The necrosed part is immediately sequestered by a growth of new horn-cells, and is eventually cast off, leaving a tiny scar.

It is important to notice that the first manifestations occur in the sebaceous glands. The gland is swarming with leucocytes before there is any attempt to pass to the surface, or invade the follicular wall. This is well shown in Fig. B. There the structure of the gland is nearly obscured by the great crowd of leucocytes. Yet none have penetrated the surface epithelium, and only at one spot deep in the follicle has the invasion of the prickly-layer commenced. This suggests in the strongest possible manner that the exciting cause lies in the gland itself. The hyperkeratosis, the tendency to acanthosis and stagnation of sebum, are predisposing causes, but the actual cause of the outbreak appears to lie in the gland. Probably it is some organism and the intimate association of the bottle bacillus in such enormous numbers with the lesions prompts the thought that they may not be so entirely innocent, as is generally supposed, when they find deep in the follicles collections of fat in which to thrive.

Two clusters of cocci, and only two, occur in the crust. But as these are away from the centre of the lesion, and only occur in spots from which the horns sheath has been removed, their influence is extremely doubtful.

Diagnosis.—The cases of this disease that I have seen hitherto have been called Prurigo Mitis. This implies that they are instances of Prurigo Mitis of Willan or P. Simplex (Hebra).

But the clinical description of the two does not really correspond. In Prurigo Mitis the original papules are described as being very small, seated in the depth of the epidermis, and more perceptible to touch than to sight. Later, when scratched, they become inflamed and red, and then rise up from the surface. They are mostly associated with urticaria, and occur early in life. This description corresponds very closely to some cases of urticaria papulosa in which the papule

formation is more in evidence than the urticaria, but it does not at all correspond to the cases in question.

In these the papule is rosy-red from the start, obtusely conical, obvious to the eye, even more than to the touch, and apparently seated on the surface. The pale centre is very characteristic. The disease occurs later in life, is not associated with urticaria, and not so apt to be associated with secondary changes in the skin. Further, these cases are distinctly relieved by scratching, whereas the others are aggravated and become inflamed. Anyone who will trouble to read case M.X. given by Brocq (*Le Prurigo Simplex et sa série, Annales de Derm. et de Syph.*, 1894, p. 29) will at once see that it is identical with these under consideration. This case showed the same peculiarity—that it was worse in winter.

At present it is difficult to fix the true position of these cases.

They appear to fall into the group of follicular inflammations as do also, I believe, Brocq's cases of P. Simplex acutus and many of the other affections to which, at various times, the name of P. Simplex has been given.

Touton ("Über Neurodermitis Chronica Circumscripta," *Archiv. f. Derm. u. Syph.*, 1895, p. 109), goes so far as to place all cases of papular urticaria in this group.

Brocq appears to keep them apart, and this, in the present state of our knowledge, appears the best thing to do.

It is my impression that several entirely distinct disorders are included under the name papular urticaria, and that whereas some are purely follicular and belong of right to this group, the others are not so and are of an essentially urticarial nature.

So far I feel inclined to agree with Touton.

If one could define Prurigo Simplex as a condition of the follicles and sebaceous glands tending to produce attacks of sudden congestion with formation of itching red papules that do not coalesce, usually subside rapidly and recur readily—then a considerable number of similar cases could be conveniently brought together. But I imagine that Brocq would not accept the purely follicular origin of P. Simplex.

The cases I am bringing forward show an acute œdema, centred round a follicle and tending to formation of vesicles. The œdema comes suddenly and passes away with comparative rapidity under the influence of scratching, pressure or cooling applications. The œdema

does not show the slightest resemblance to that of urticaria. The papules are red, tense, and shining, and itch severely. Such are encountered commonly in the seborrhoic, and are by most considered to be merely a manifestation of eczema. This view is open to question, and I think Brocq presents a remarkably strong case for keeping them apart.

The papules in question, however numerous they may happen to be, almost always remain discrete. In the few instances when they run together it is much more reasonable to suppose that a secondary eczematization has occurred, and this may be supported by the result of treatment; for often the eczematous patch remains long after the surrounding papules have disappeared.

Anyone who cares to study the cases I have described will realise how easily they might develop eczema. The daughter actually did show some eczematization in the neighbourhood of the scalp. A case I saw in England behaved in the same way. She had had the disease for years without the slightest evidence of eczema and then she suddenly developed a typical seborrhoic eczema of the scalp and face.

Some cases of acne have close affinities with this group. The blocked follicles, in certain instances, become surrounded by an acute œdema that often subsides as quickly as it comes. Such cases are particularly obstinate. In one a dose of alcohol, in another some indigestible food, in another change of temperature, exposure to heat or cold, in another the application of some unsuitable remedy—these and a hundred other causes are sufficient to convert a plugged but otherwise unaltered follicle into a large, conical, intensely red, itching papule.

These cases do not resemble the ordinary cases of inflamed or pustular acne. They require a different treatment and are of far graver prognosis.

It is not surprising that the French school looks to internal causes in trying to explain these cases. They are diathetic, autotoxic, reflex nervous disorders, etc. And yet when one examines the exciting causes it will be found that they are quite as often external as internal. When a case shows an eruption of itching, red papules after taking alcohol or sugar, or any other article of diet, or after some nervous disturbance, one is apt to give the particular thing

named the credit, or rather discredit. But the same eruption in the same case may follow exposure to sun or to cold wind, or any other external trauma, and then it is classed in the still more obscure group of angio-neuroses.

But it is quite as probable, seeing that the assumed cause may be either internal or external, that the real cause lies in the skin itself, and may be capable of demonstration.

The two cases forming the subject of this paper are good instances in point.

Here there is evidence that from a very early period the skin was not normal. The eruption developed in both cases at important developmental periods—namely, childbearing and menstruation.

The disease developed in the daughter at an earlier age, and was more severe. In both it had never disappeared.

The French school would most certainly have ascribed these cases to internal conditions, either diathetic or nervous. They do so ascribe them.

It is extremely hard to believe that any and every chance substance that irritates the stomach or passes into the blood can, of itself, cause papule formation unless there are already existing foci in the skin that must first be called into activity.

So, too, with the diathetic prurigos described by Besnier, one feels that whereas great attention is paid to the diathesis and to the action of trauma, very little is given to the skin itself, in which probably lies the immediate cause and explanation of the eruption.

As Besnier has shown, the removal of the trauma is far more efficacious in the cure, or at least alleviation of the skin eruption, than the modification, by diet or otherwise, of the diathesis supposed to cause the same.

It is practically certain that in these cases the true cause is a local change, brought about, it may well be, in course of time by general conditions. When these skin-changes are better known and distinguished we shall be in a much better position to permanently cure these affections than we are at present.

It is still more difficult to accept the neurodermitic theory of Brocq for explaining these pruriginous eruptions.

As Neisser so aptly observes, it makes a great difference as to

whether a disease, with the same external manifestations, and one may add with the same histological features, occurs in a nervous, highly-strung Frenchman (and one can also add American as well), or in the more phlegmatic German or Englishman.

Naturally, in the first instance the nervous phenomena are most striking, and receive undue attention—as also the digestive disturbances.

The cases described afford a good illustration. Had this disease occurred in a Frenchwoman there would almost certainly have been far more nervous disturbance, and, with equal probability, digestive disturbance to give colour to the nervous and autotoxic theories of this disease.

And yet these German ladies bore it with equanimity. Their general health was not in the least disturbed. They ate their food with relish and digested it well. They slept well, looked strong, and were well-nourished, and happy, with an amount of irritation that would have driven many other people frantic.

Hence, in these cases, the idea of a local cause, lying in the skin itself was at first sight far more probable than any other.

I think my preparations show clearly that such a local cause did exist.

I have a strong impression that the eruption depended largely on a retention of fat and on decomposition or changes in that substance. I believe that the more this group of cases is investigated the more clearly will local changes be demonstrated, and that it will be in the fat that the chief changes will be found.

Chemistry alone can solve these problems, and in Dermatology chemistry has a great opening.

Treatment.—I need not enter into the treatment usually adopted for such cases. It is almost invariably successful for the time, and then later on there are fresh outbreaks. But the histological examination impresses on us the need in these cases of systematic bathing and massage of the skin. If this be done constantly and well I am inclined to fancy that these cases would give very little further trouble.

EXPLANATION OF PLATES.

PLATE A.—*Sebaceous gland from a part not inflamed.*

1. The commencing horny plug.
2. A spiral follicle, hyperkeratotic.
3. Sebaceous gland, completely converted into fat cells.

PLATE B.—*The New Papule.*

1. The horny plug containing colonies of bottle bacilli.
2. The sebaceous gland, infiltrated with leucocytes.
3. A plug of sebum deep in gland, showing bottle bacilli.
4. Commencing infiltration of epithelium with leucocytes.
5. Cell-infiltration and increase of connective tissue cells round the vessels.

PLATE C.—*The Old Papule.*

1. Enormous epithelial mass by fusion of several follicles.
 2. Part of a sebaceous gland, showing plug of sebum and leucocytic infiltration.
 3. Columns of leucocytes coming to surface.
 4. Edge of vesicle.
 5. The crust.
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SOCIETY INTELLIGENCE.

DERMATOLOGICAL SOCIETY OF LONDON.

A MEETING of this Society was held on Wednesday, January 8th, 1902, Dr. STOWERS in the chair.

The following cases and specimens were shown:—

Dr. DORE showed (for Mr. MALCOLM MORRIS) (1) A case of *Myoma multiplex cutis* affecting the right leg and thigh of a woman, aged 29 years. The patient's father was shown in December, 1900, with a similar condition of the left side of the chest. A further account will be given of these cases.

(2) A case of *Erythema iris* in a man, aged 27 years, by occupation a cellarman. His first attack had occurred four years previously, and he had had fifteen or sixteen since at fairly regular intervals of two months. The attacks had all been similar in character, affecting the hands, wrists and forearms, and mucous membranes of the sides and roof of the mouth. No other part of the body had been affected. He had been laid up with inflammation of the lungs and dropsy, and had worked for four years in lead previous to the onset of his disease.