THE BRITISH JOURNAL OF DERMATOLOGY.

SEPTEMBER, 1895.

A CASE OF PITYRIASIS RUBRA PILARIS (DEVERGIE), WITH A SHORT ACCOUNT OF THE DISEASE.

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(*Illustrated.*)

JOHN JONES, aged 58, came to the Hospital complaining of an affection of the skin, of which he gave the following account:—Two years ago some small pimples appeared on the forehead, which itched a good deal. The rash rapidly spread to the whole face. From the face it gradually spread over the head and body, so that in five or six weeks the whole body was covered. The rash was scaly from the first, never moist or running, and always dry, though not so dry at first as it is now.

The general health had not been in any way affected. He had been under treatment for the last seven months, the diagnosis, he stated, being made of eczema. No improvement or change had taken place, and so he came to St. Bartholomew's Hospital.

He had had syphilis at the age of 20, but had never had any return of it. He was very intemperate when young, but had been steady for many years. There was no similar complaint in his family, nor anything of interest in the family history.

The following description of his condition was taken on 16th April:—The patient is a well-nourished man, looking in fairly

good health, and except for the rash has nothing the matter with him.

The eruption covers the whole body, from the crown of the head to the sole of the foot. Its general character is red, the redness being most marked below the waist, between the thighs and upon the legs, while on the upper part of the abdomen it is of a brownish colour. Upon the shoulder, arm, neck and face, the skin is covered with bright glistening scales. These scales occur over the whole body, but are most marked on the shoulder, the back and the elbows, and about the sacrum between the natal folds. In the latter place they form large patches of a mother-of-pearl appearance, but nowhere have they the characters of the scales in psoriasis, and there is hardly any scaling over the knees. Over the head, neck, back, thighs and arms the normal lines of the skin are greatly exaggerated, so that the case resembles one of ichthyosis. Over the whole body, even in the parts which are most affected, there are small patches of healthy skin, some of them as large as a shilling, but many of them so small as to be easily overlooked. They stand out in strong contrast with the redness round them, so that at a distance they look as if they were small scars. In some of these patches the skin appears to be perfectly normal, in others there are small papules to be seen. The hairy scalp is covered with fine scales, which give it much the appearance of seborrhœa, but on the crown, which is bald, the skin is somewhat cracked and fissured. The patient has been bald for many years, but does not think that he has lost more hair recently. The conjunctivæ are somewhat congested, and the lower lids swollen and everted. There is but little thickening of the skin anywhere, even in the parts most affected.

The eruption begins as a papule. The initial lesions are best seen over the lower part of the front of the abdomen and in the epigastric region. Elsewhere the disease has passed beyond the papular stage, but in most of the small patches of healthy skin, as already stated, papules are to be seen. The individual papules are quite small, round, about $\frac{1}{16}$ th inch in diameter, distinctly raised above the surface, with a red base and a cap of scales. They are for the most part arranged in lines which follow the normal lines of the skin. When they run together they form at first long ridges along these lines. The scales are very adherent, and when picked off leave a little

depression in the top of the papule, which is red, but does not bleed. The penis seems to have escaped entirely. The hands and soles are covered with horny scales, which have cracked in places, and look like the skin of a crocodile. The cracks, however, do not bleed, nor are they painful. The nutrition of the nails on the hands is affected, for they have grown irregularly and are ridged, but they are firmly fixed. On the feet the nails are for the most part black and dropping off, their place being taken by masses of scales of a dirty grey colour.

All the functions are performed naturally, and the patient looks and feels well. He is out of work, for the reason that his employers will not have him at his work, and not that he is unable to work.

I took him into the Hospital, intending before treatment was commenced to excise a piece of the skin and make microscopical specimens of the initial papules. Unfortunately, in the few days that were allowed to elapse, the disease spread so rapidly that all the initial papules were lost, and an examination of other parts of the skin seemed unnecessary.

I ordered the patient to be soaked daily in a hot soda bath (8 ozs. of washing soda to thirty gallons of water) for about twenty to thirty minutes, to soften the skin and remove the scales, after which he was to be well rubbed down with a bath-towel and thoroughly oiled all over with carbolated oil, 1 per cent. At the same time I placed him upon a mixture containing 5 minims of *Liq. arsenicalis*, which was to be taken three times daily.

The bathing and oiling were continued throughout his stay. The arsenic was rapidly increased up to 15 minims three times a day, and continued for several weeks, but without any real improvement; for, although the scales were removed by the bathing and oiling, and so far the appearance of the patient was improved, the disease seemed to be really as active as ever. In all other respects he was well, and the arsenic in no way upset him.

After nine weeks of the arsenic treatment I decided to place him upon thyroid extract, and gave him at first two and later three tabloids daily. He took them without any general disturbance, and almost immediately improvement commenced in the skin.

His present condition (August 12), after taking to thyroid extract for five weeks, is as follows:—

There is little to be seen in the skin now but a general redness—the colour is not vivid as it was, but of a dull brownish-red, and is largely due to pigment staining, such as is seen after erythema—for on pressing the blood out of the skin the impression is not white but brown.

There is still slight general desquamation, but the skin has lost its hardness, is moist and supple, and, except that a little thickening still remains, it feels normal. The palms and soles are soft, moist and supple, and so are the buttocks, back and shoulders. The nails of the fingers are growing normally, and the scaly masses on the toes are almost gone; and here too the nails are becoming normal. The ectropion, which was so marked at first, has now gone entirely in the left eyelid, and is hardly perceptible in the right.

The patient is in excellent health, and appears to be on the high road to complete recovery.

There can be no doubt, I think, that the thyroid extract has been the active agent in the cure of the case, for the improvement was coincident with the administration, and continued steadily day by day.

Attention was first drawn to this affection by Devergie in 1854, after whom the disease is usually called Pityriasis rubra pilaris of Devergie. The most complete monograph is that by Besnier,* published in 1889, which is based upon twenty-eight cases, most of them of French origin, three described in this country, one in New York, and two in Christiania. Hilton Fagge, under the name of Rhinoderma, also records a case in the Guy's Hospital Reports, a model of which Dr. Pringle informs me is in the Guy's Museum, No. 327.† Recently, besides the case which is the subject of this article, another has been recorded by Dr. Abraham during the present year.

The disease is, therefore, a very rare one, for in forty years hardly more than thirty cases in all have been put on record. The initial lesions consist of small papules which give the skin, at a distance, the appearance of "cutis anserina." These papules are capped with a small mass of scales which project from the mouth of the dilated

* *Annales de Dermatologie et de Syphiligraphie*, 2nd Ser., t. X, 1889. Nos. 4, 5, 6.

† This case appears to have been missed in most of the references to this disease, and the credit of rescuing it from oblivion is due to Dr. Pringle.

hair-follicles. The papule is, indeed, nothing more than the hair-follicle dilated with these epidermic accumulations and surrounded by a little inflammatory infiltration, giving the papule its red colour. These epidermic scales form blunt conical masses with their base upwards, and they extend down the hair-follicle as far as the entrance of the ducts of the sebaceous glands. They can be detached by the finger or the forceps without causing bleeding, and then leave a cup-like depression in which the vessels of the deeper layers of the skin can be seen, but which do not bleed. As these papules increase in number the exudation in the skin round them increases, and the normal lines of the skin become greatly exaggerated, so that in the later stages, when the infiltration is considerable, the skin lines look very much like those met with in Ichthyosis.

As the disease progresses the papules become confluent, and the whole skin assumes a diffuse red colour, becoming thickened and covered with scales. In this stage the appearance is somewhat like that of diffuse Psoriasis, or even Dermatitis Exfoliativa.

The four stages of the disease are the following:—(1) The period “*anserina*”; (2) the perifollicular hyperæmia; (3) the superficial infiltration of the papillary layer of the dermis; (4) the diffuse smooth redness with exaggeration of the folds of the skin, and desquamation (Pityriasis).

The initial lesion may appear in almost any part of the body, but its favourite seat is upon the exposed parts, viz., the hands and the face, and occasionally the neck and extensor surface of the forearms. Upon the hands the common seat is the backs of the second and third phalanges and the dorsum of the hand. Upon the face it often appears first on the forehead, eyelids, or chin. The initial papules do not occur upon the scalp or upon the parts of the body covered with large hairs, but they may be found on any part of the body where the lanugo-like hairs are found.

Even when the disease is fully developed, and in most parts of the body has passed beyond the papular stage, still patches of skin may be found here and there which, except for the initial lesions, are otherwise healthy. This is important in respect of diagnosis. Attention was drawn to these patches of healthy skin in the present case.

Once started, the disease usually extends locally for some time, and often quite slowly. Subsequently it develops with greater

rapidity, and may then, in a very short time indeed, become general.

The disease may be met with at almost any age, but generally, it is stated, commences in infancy or during youth. It is, as a rule, most common in males. There is nothing in the family history with which the disease can be connected, and its cause is quite unknown. The general outbreak may be preceded by an initial stage of varying duration in which no characteristic lesion may be present to give the diagnosis; nothing, perhaps, but a diffuse redness, the cause of which is obscure. As soon as the typical lesion appears, a few weeks may be sufficient for it to spread very widely.

There is generally no constitutional disturbance of any importance throughout the disease, and but few symptoms in the skin itself.

The affection runs a very chronic course, but its duration is very variable. It is liable to relapse, but any given attack may develop very suddenly and terminate as rapidly, though it usually lasts for some months, and occasionally even some years.

When once the disease begins to resolve it may rapidly disappear.

Finally, it is never fatal.

The essential lesion is an accidental anomaly of the cornification of the epidermis, especially at the mouths of the follicles, affecting first the walls of the follicular infundibulum, the mouths of the sebaceous glands, and very frequently the bed of the nail. When the initial lesions are present the diagnosis is easy.

In the more advanced conditions of the disease, or when the initial lesions are not evident, the diagnosis may be very difficult.

The diseases with which it is most likely to be confused are, in the first place, Ichthyosis; (2) Lichen planus; (3) other forms of Pityriasis rubra; and (4), lastly, Psoriasis.

Nosologically its closest affinities are with Psoriasis.

The treatment which appears to be most useful is that commonly employed in Psoriasis, namely, arsenic in full doses, frequent bathing, with inunction and ointment of chrysarobin, salicylic acid, tar, and its derivatives, &c. In my own case the success which followed the administration of Thyroid Extract was very striking.

In conclusion, I wish to express my thanks to Dr. Colcott Fox for the interest he has taken in the case, and for his great kindness in placing the literature of the subject at my disposal.