A CASE OF PITYRIASIS RUBRA.

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IN bringing before the Academy a case of the rare skin disease pityriasis rubra, a few remarks on the use of the term may not be out of place. Etymologically the name pityriasis rubra is derived from *πίτυρον* (bran) and *ruber* (red), and from the time of Bateman it has been applied to various chronic affections of the skin characterised by scales and redness. In 1854 the term was used in a more restricted sense by Devergie, who applied the name *Pityriasis rubra aigue* to the disease, which Hebra, writing in 1860, called pityriasis rubra. Hebra's description is as follows:—"In pityriasis rubra there is nothing more than an intense redness diffused over a large part of the skin, or even universal, disappearing beneath the pressure of the finger (when it gives place to a yellowish coloration), and accompanied by the presence of fine, white, loosely-adherent scales, which result from the constant shedding of the most superficial layer of the cuticle." He insisted on the absence of infiltration, moisture, or of severe itching or excoriations, which were present in psoriasis, eczema, lichen, &c.

Kaposi writes:—"By pityriasis rubra we mean a peculiar disease, first described by Hebra, which is of extremely rare occurrence, and is characterised besides its chronic course by the fact that it presents no other form of eruption (neither nodules, nor vesicles, nor pustules) but that from the start, and during its entire course, there is nothing but redness and desquamation of the skin."

Crocker defines pityriasis rubra as "An inflammatory disease involving the whole surface of the body, characterised by deep redness with abundant flaky desquamation," and he states that it may be primary or follow some other form of dermatitis. He gives "dermatitis exfoliativa" as a synonym. Confusion would be avoided if the name pityriasis rubra were confined to the disease described by Hebra, and if the term "general exfoliative dermatitis" were reserved for cases secondary to other skin diseases—such as psoriasis, eczema, &c. The patient whom I exhibit will be found to be an example of Hebra's type of pityriasis rubra. His *clinical history* is as follows:—

Edward R., aged twenty, a clerk by occupation, was admitted to the Whitworth Hospital, on November 4th, 1897, suffering from general redness and scaling of his skin, with some itchiness. He has had the affection for six months. It appeared suddenly in May, 1897, in the shape of redness on the backs of both his hands and wrists, and, spreading from those regions, the eruption covered the whole body within a week. From the start the skin was bright red, and there was free desquamation of large, thin white flakes. The eruption was preceded by, or attended with, no general symptoms, but there was moderate itching. He had previously enjoyed excellent health, and never had any skin affection. He never had syphilis. He was a total abstainer till November, 1896, since which time he drank to excess, more especially during the month preceding the appearance of the disease. He was constantly exposed to cold, and the night before the onset of the eruption he was out all night and got a severe wetting. Alcohol and chill, therefore, may have had some ætiological influence in exciting the disease.

His family is free from any gouty, rheumatic, or phthisical taint, with the exception of an uncle who died of phthisis. He states that an aunt had eczema.

On admission to hospital he was fairly well nourished, though of rather slight build, and he enjoyed good health except for the condition of his skin. He sometimes complains of feeling chill when he goes out of doors, and he is troubled at night by moderate itching. He eats and sleeps well; his bowels are usually confined; urine normal. His lungs and heart are healthy; temperature normal

The inguinal and axillary glands are all enlarged, but only to a slight degree.

Examination of the blood shows a normal condition as to sp. gr., hæmoglobin and red corpuscles. He has marked leucocytosis, the white cells being increased to 37,000 per cubic mm. Further, the eosinophile leucocytes are relatively increased, constituting 10 per cent. of all the white cells. A differential count of 1,000 leucocytes gives the following result:—

Lymphocytes	...	...	...	20 per cent.
Large mononuclear and transitional forms...	4	„		
Polymorphonuclear neutrophiles	...	66	„	
Eosinophiles	...	...	...	10 „

The patient, therefore, has *eosinophilia*, a condition found by Neusser in many skin affections.

The skin all over his body is of a dull red colour, and covered with thin, papery scales. It feels hot, rough, and dry; he never sweats. The redness fades on pressure, leaving a yellowish hue. The scales are thin, whitish, easily separated from the subjacent skin, which is red, but neither moist nor infiltrated. The scales vary in size from a fine branny desquamation on the face and hands, to large flakes as big as a sixpenny piece on the back and on the extensor surfaces of the limbs and dorsal surface of the feet. During the earlier months of the disease the scales were much larger; for the last few weeks they have diminished in size. Every morning about a pint of scales and powdery detritus is found in his bed. The nails of both big toes and of the little finger of his right hand are thickened and nearly separated from their bed by an accumulation of epithelium beneath. The other nails are thin, and longer than the fingers and toes—the thinning being specially conspicuous in the toe-nails. The scalp is covered with whitish-yellow masses of scales, and the hair is falling out. The surface of the palms is a little reddened, and covered with fine, powdery desquamation, and the epidermis appears thin (as if newly formed) and traversed by superficial cracks. The skin of his soles is thicker and desquamating in white flakes. He states that the epidermis peeled freely from his palms and soles at an early stage. When the skin is relaxed, especially at the bends of his elbows, the sides of his neck and trunk, &c., the red and scale-covered surface presents the appearance of crepe, being traversed longitudinally and transversely by fine ridges and furrows; but if the skin is made tense in the same positions, the ridges and furrows are obliterated,

and fine, linear red streaks become apparent. The mucous membranes are not affected, and the tongue is normal.

The above description fairly represents his condition on admission to hospital a fortnight ago; since then he has improved considerably; the amount of the desquamation and the size of the scales have diminished, and the itching is less. The scales are now most evident on the extensor surfaces of his arms, on his back, and on his legs.

The *diagnosis* of the case depends on the presence of universal redness of the skin, associated with loosely-adherent, thin, papery scales, and on *the absence of* thickening, moisture, or of any primary lesion. The case differs from lichen ruber in the absence of papules at any stage, and in the absence of thickening; from pemphigus foliaceus, in the absence of laminated crusts, and of flaccid bullæ, beneath which the corium would be moist and raw; from general eczema, in the absence of exudation and of severe itching, of infiltration, vesicles, pustules, and crusts; from diffuse psoriasis, by the absence of silvery scales adherent into crusts, and the absence of red points when the scales are removed; and from *all* those affections it differs in its absolute universality.

The *prognosis* of the case, both in reference to its probable duration and likelihood of cure, and as regards its danger to life, must be considered serious, but by no means as hopeless as Hebra's and Kaposi's experience would indicate. Those observers record a fatal termination in almost all the cases which came under their notice. On the other hand, Crocker considers 50 per cent. far too high a figure for the mortality, and Pye-Smith refers to forty cases from different sources, with fifteen recoveries and only eight deaths. It is probable that such discrepancy in the proportion of fatal cases in the practice of different dermatologists is due to the fact that general exfoliative dermatitis secondary to psoriasis, eczema

or other skin diseases is not nearly as intractable or as fatal as Hebra's type of pityriasis rubra, whereas some authors classify all cases under the latter category. Most fatal cases appear to occur in children or in elderly patients.

The *rarity* of the disease is indicated by the fact that Hebra met with fifteen cases, and Kaposi only six, whilst Crocker saw only fourteen instances of the affection amongst a total of 10,000 skin cases in his hospital out-patient department.

The *course* is generally very chronic, and the usual cause of death is marasmus or some intercurrent complaint. Cases which apparently recover are very liable to recur.

The *cause* of the disease is unknown.

Regarding the *pathological anatomy* in recent cases there is a moderate amount of cellular infiltration in the cutis and papillary layer. No special changes occur in the epidermis apart from those associated with desquamation. In the later stages there is much atrophy of the skin, the rete Malpighii is thinned, the papillæ disappear, and the skin appendages are obliterated. Where the atrophied skin is tense fissures may occur.

The *treatment* of pityriasis rubra, as may be inferred from the chronic course of the disease and high mortality, is not satisfactory. Such general measures as prolonged rest in bed, avoidance of cold, repeated employment of tepid baths, milk and easily assimilable diet, are indicated, together with the local use of oleaginous applications and emollient ointments. The internal administration of arsenic may be tried in the later stages, though it frequently fails; bark and mineral acids, iron and cod-liver oil have benefited some cases. Perhaps the employment of thyroid extract might be of use. I should add that my patient has been treated with arsenic, ointments and baths for the last few months without any apparent improvement. Since he came under my care a

fortnight ago, I have adopted no more active treatment than keeping him in bed and giving him an aperient mixture containing sulphate of magnesia. Whatever the result may be attributed to, his condition has certainly improved since his admission to hospital, but it remains to be seen if the improvement will be permanent.

[*Addendum*.—The subsequent progress of the case was one of uninterrupted improvement. Patient was kept in bed for a further period of three weeks, during which the saline aperient mixture was continued, and the scales were removed by the daily administration of alkaline baths, combined with the use of Hebra's *Spiritus Saponatus Viridis* as a soap. The tendency to scale-formation lessened every week, the redness of the skin gradually faded, and the hair of the scalp took on new growth. Patient's skin was practically normal towards the end of December, and he left hospital apparently cured early in January, 1898.]

DR. WALLACE BEATTY said the diagnosis between the two varieties of pityriasis rubra—that was, the variety which began acutely with red spots and rapidly became general, and the variety preceded by an eczema or lichen—could be made out only by the clinical history. When one saw a universal case of pityriasis rubra which had succeeded an eczema or psoriasis, the appearances of such a case were precisely similar to the appearances in Dr. Coleman's case, so that it just came to be a question whether to call this case pityriasis rubra, when the case is acute and not preceded by any other skin disease, and whether to call the other cases general psoriasis or general lichen. He was inclined to look on pityriasis as a condition which might be primary or which might be secondary to some other skin affection. He described a few cases of skin disease last year, one of which presented all the characters of pityriasis rubra in the beginning; however, the course since shows that it is a case of pemphigus foliaceus. Looking at Dr. Coleman's case for the first time one might think that it was a case of generalised seborrhœa, but the history must show that it was a case of pityriasis rubra.

DR. FINNY said that the case did not conform typically to many of the described symptoms of the disease, and it was hard to say to what group it belonged, except to the class of pityriasis rubra. There were in this case symptoms which he had not seen in any case of pityriasis rubra—namely, great thickening of the scalp, with a great quantity of scales, and with alopecia. These symptoms were absent in the only two cases he had ever seen. Also, the scales in those cases were very much larger than those in Dr. Coleman's case. This fact made one look on the case as one in which scaling was taking place after universal erythema. It struck him that in taking up the skin of the patient's arms and legs it felt extremely tight and hard, and the skin did not seem to him to be the soft skin which is found in pityriasis rubra, but over the tibia it could not be moved at all, and one might think it was eczema with dried exudation. The patient himself said he was itchy and had to scratch himself, and he (Dr. Finny) thought that this was one of the peculiarities wanting in pityriasis rubra, as there was generally no itching at all.

THE SECRETARY (DR. R. TRAVERS SMITH), said that he should like to support Dr. Coleman when he said that the disease was much better marked some time ago when he (Dr. Smith) had seen the patient. The scales were then much more abundant, and the erythema was more intense, especially over the arms and anterior surface of the chest. He asked what Dr. Coleman thought was the significance of the leucocytosis in the case, as he believed it had not been demonstrated in a case of pityriasis before.

DR. KNOTT said he had seen a case of pityriasis rubra. The patient was an old man, eighty years of age, and the one symptom that he complained of was sleeplessness, due chiefly to the itching, although not violent itching.

DR. COLEMAN, replying, said he agreed with Dr. Beatty that secondary cases of pityriasis rubra or dermatitis exfoliativa were indistinguishable clinically, except for their history, from a case such as he had brought forward, but he thought that a distinction might with advantage be made on account of the relatively better prognosis of the cases which are secondary to eczema, for instance. Regarding Dr. Finny's remarks as to the alopecia of the patient's head, he (Dr. Coleman) thought that some alopecia always existed in pityriasis rubra, and Unna laid stress on this as a symptom of the disease; the accumulation of scales on the patient's scalp was due to the hairs fixing the scales, and to the

fact that the patient's head had not been washed for some weeks. Neither some thickening of the skin nor itching was sufficient to upset the diagnosis of pityriasis rubra; in fact, in some of the very chronic cases a considerable amount of induration of the skin had been found. Itching had also been noticed now and then as a prominent symptom, although it was not usually present. With regard to Dr. Travers Smith's question about the significance of the leucocytosis, he (Dr. Coleman) supposed that in this case it was due to the inflammatory condition of the skin. Neusser had found an increased proportion of eosinophiles in a number of skin diseases, and he (Dr. Coleman) had found such a condition in a case of pemphigus latterly. In reply to Dr. Knott, it was, he thought, true that no other form of skin disease had such an absolutely universal distribution as pityriasis rubra, although one occasionally sees cases of psoriasis diffusa and pemphigus foliaceus affecting almost the whole surface of the body.