ERYTHEMA DYSCHROMICUM PERSTANS

A Hitherto Undescribed Skin Disease*

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During the year 1959–60 in the Department of Dermatology of the Vargas Hospital in Caracas, we studied 5 adult patients with a peculiar new pigmentary disorder. This preliminary article will describe our investigations and attempt to classify this disease.

DESCRIPTION

The skin eruption is characterized by numerous macules with a grayish color of varying intensity. This color is in marked contrast to the normal skin. When the lesions grow the macules have a slightly raised erythematous border which is firm and feels like a thin piece of string. The macules vary in size, the smallest being a few centimeters in diameter, while the larger ones, formed by the merging of the smaller macules, cover extensive areas of the trunk, limbs and face. The lesions tend to be present chiefly on the trunk and upper limbs but have been seen on all parts of the body except the scalp, palms and soles, and mucous membranes. When the macules occur on the face it is impossible to trace the borders of the individual lesions, giving the appearance of a diffuse poorly defined eruption.

Depending upon the age of the lesion and the pigmentation of the patient the macules vary in their color presenting hypo- and/or hyperpigmentation upon a grayish background.

Our 5 patients consisted of 3 males between the ages of 11 and 36 and 2 females aged 17 and 27. All 5 were from the interior part of the country of Venezuela.

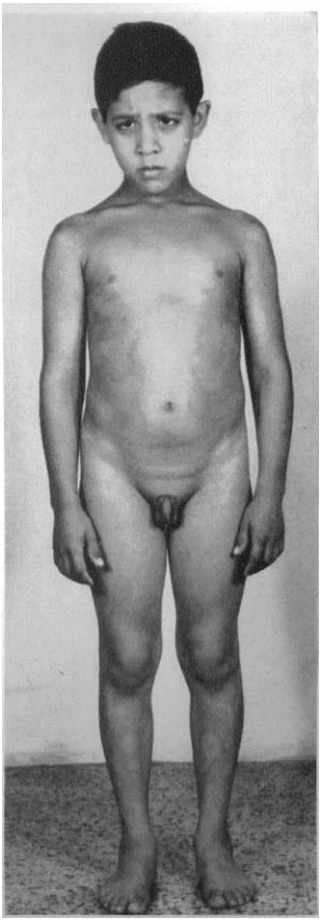

Fig. 1. The widespread distribution of the dyschromic lesions are seen on the trunk and extremities. These lesions have a thin infiltrated erythematous border.

COURSE

The condition is mainly asymptomatic except for occasional moderate pruritus during the evanescence of new macules. The condition is chronic and has a tendency to extend and to involve previously unaffected areas. New lesions have been observed to occur in previously old pigmented areas. These new macules undergo the same evolution and have the same descriptive appearance as described above.

HISTOPATHOLOGY

Skin biopsy specimens were taken from each patient and the following stains were used: Hematoxylin-eosin; Pearl; Hotchkiss MacManus (PAS) Gomori's trichrome stain; Gallego;

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Received for publication November 8, 1960.

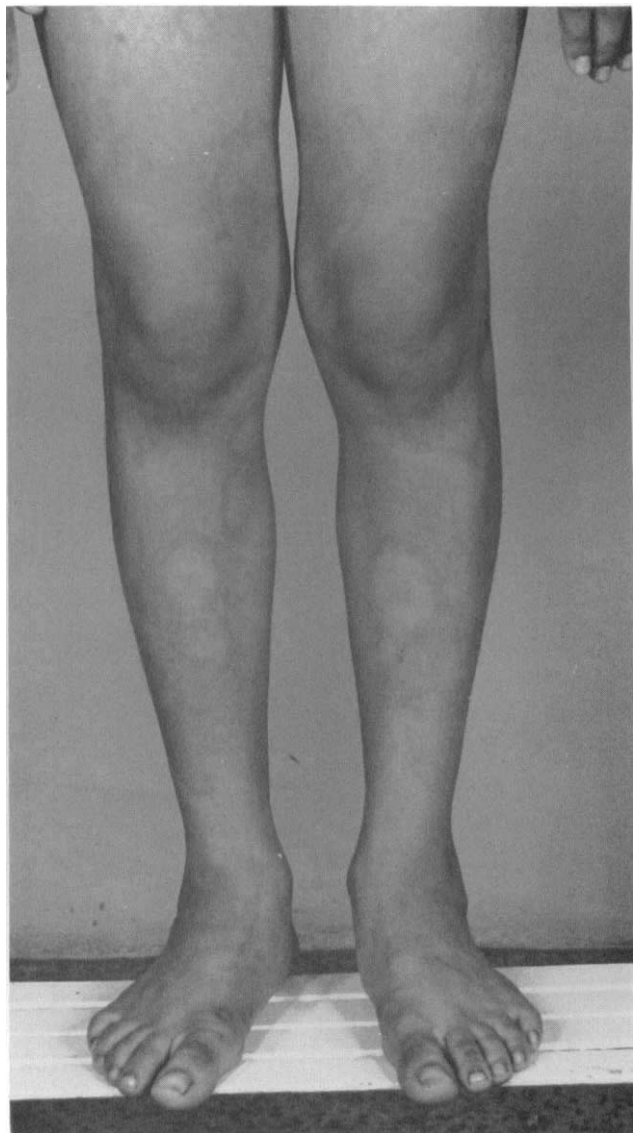

FIG. 2. A closer view of the dyschromic lesions on the legs and thighs showing the hyperchomic borders and the hyperchromic centers

Masson-Fontana (silver impregnation) and Dieterle treponeme stain.

The findings were uniform and consistent with the following histopathologic picture. There is a slight follicular hyperkeratosis and hydropic degeneration of the basal layer of the epidermis.

Incontinence of the pigment is present. There is a sleevelike perivascular infiltrate of small cells, histiocytes, and numerous macrophages filled with malanin-carrying granules. These granules are also seen outside the cells.

The histopathologic changes in the cutis are seen mainly in the upper third, especially in the papillary and subpapillary zones.

INVESTIGATIONS

Each patient was carefully examined medically and dermatologically and the following routine laboratory procedures were performed; complete bloodcounts, urinalysis, X-ray of the thorax, and serological tests for syphilis (Kahn, Kolmer,

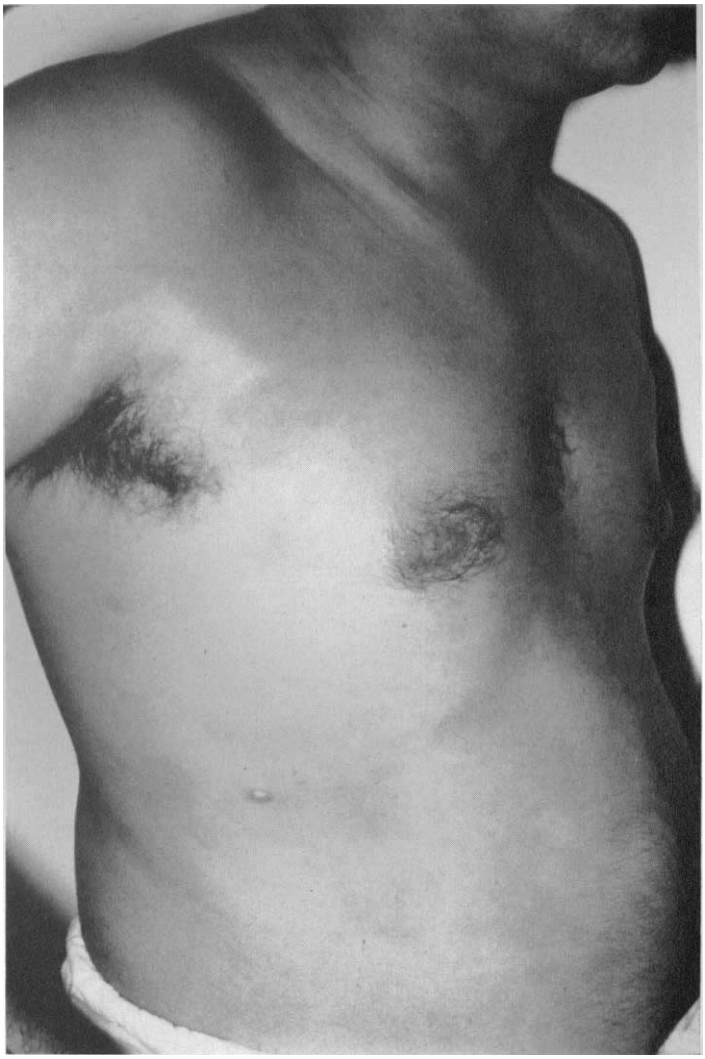

FIG. 3. Extensive involvement of the anterior part of the body showing sharp demarcation between hyperpigmented and normal skin.

VDRL and Reiter PCF). These were all within normal limits.

Frequent attempts to find treponemes using darkfield examination, with material obtained from the lesions were repeatedly negative.

Plasma protein electrophoresis was normal.

Cultures were taken from skin, urine and feces and revealed no pathogenic fungi.

Antistreptolysin titres were normal.

Bacteriological examination of the pharyngeal mucosa revealed the constant presence of alpha and beta hemolytic streptococci. Both these organisms were sensitive to penicillin.

The results of the following tests were all within normal limits: histamine and the intradermal injections of PPD, Ito, Mitsuda, Leishmanin, and Frei antigen. Neurological examinations for paraesthesias were normal and dermagrophism was not present. We were unable to perform any successful auto- or hetero-inoculations.

THERAPY

The condition is resistant to large doses of penicillin (15–20 million units) as well as to

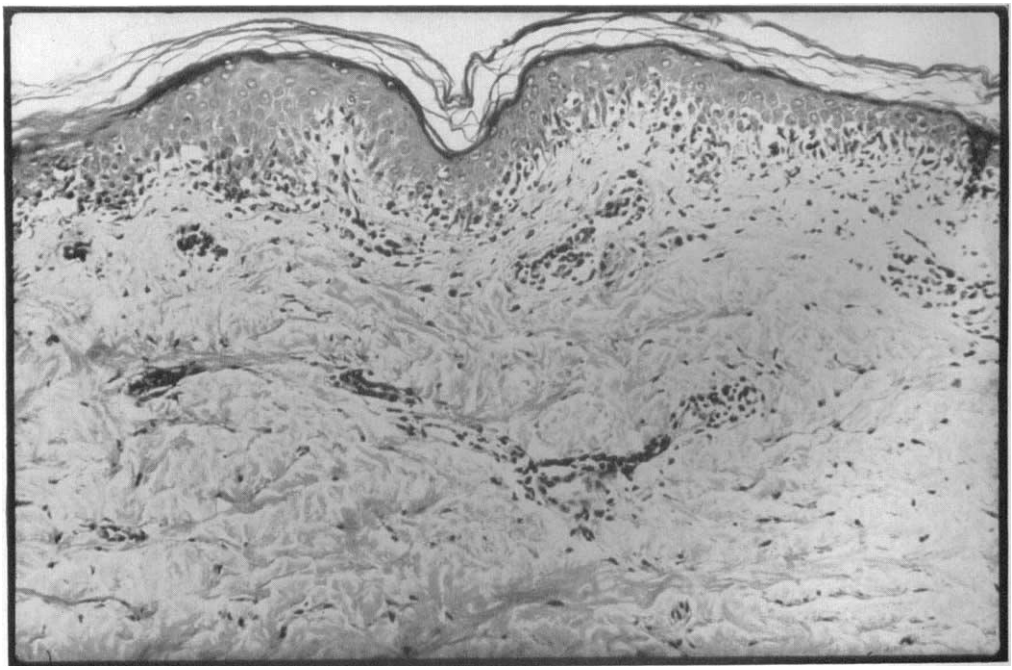

FIG. 4. Photomicrograph—× 60. The intense hydropic degeneration of the basal layer of the epidermis is noted. There is a perivascular cellular infiltrate in the upper and mid-cutis composed of small round cells, histiocytes and chromatophores.

broad spectrum antibiotics. Sulfones and corticosteroids have been of no value.

CLINICAL AND HISTOPATHOLOGICAL DIFFERENTIAL DIAGNOSIS

We have tentatively correlated this new condition with the erythema perstans group (2, 16), which consists of various erythematous polycyclic skin eruptions of a chronic nature and of unknown etiology. It differs, however, from this group by the predominant gray or slate-colored appearance, which is caused by the hydropic degeneration of the basal layer.

This phenomenon is not seen in the disease of the erythema perstans group.

When we first saw these lesions with their peculiar color that varies from a bluish to a grayish steel, we felt that we were dealing with a variety of pinta (called carate in Venezuela).

Repeated darkfield examination of the lymph fluid from the lesions revealed no treponemes. All the serological tests were negative and the condition did not respond to treatment with penicillin. We, therefore, discarded the diagnosis of pinta.

In pinto there are mottled dyschromias, and in later stages achromia as well. We have observed our patients who have had their condition for years and have not seen any of the above manifestations.

The lesions are not affected by sun light and are identical in both the exposed and unexposed parts of the body. In this respect they form a marked contrast to other pigmentary conditions which are affected to some extent by sun exposure, *e.g.* chloasma, Riehl's melanosis, melanodermatitis toxica and agyria.

Cañizares has written (1) about the work of Ramírez on "los cenicientos" or "the ash-colored ones" suffering from a curious disease, that has been observed in El Salvador and is characterized by an ash-colored eruption on the neck, in the armpits and on the mucous membranes. Ramírez presented a case of this disease to the second Central American Congress of dermatologists held in Guatemala City, Nov. 5–8, 1959. (This could possibly be similar to the cases we are describing.)

SUMMARY

A new skin disease is described and two names have been suggested. One is erythema chronicum

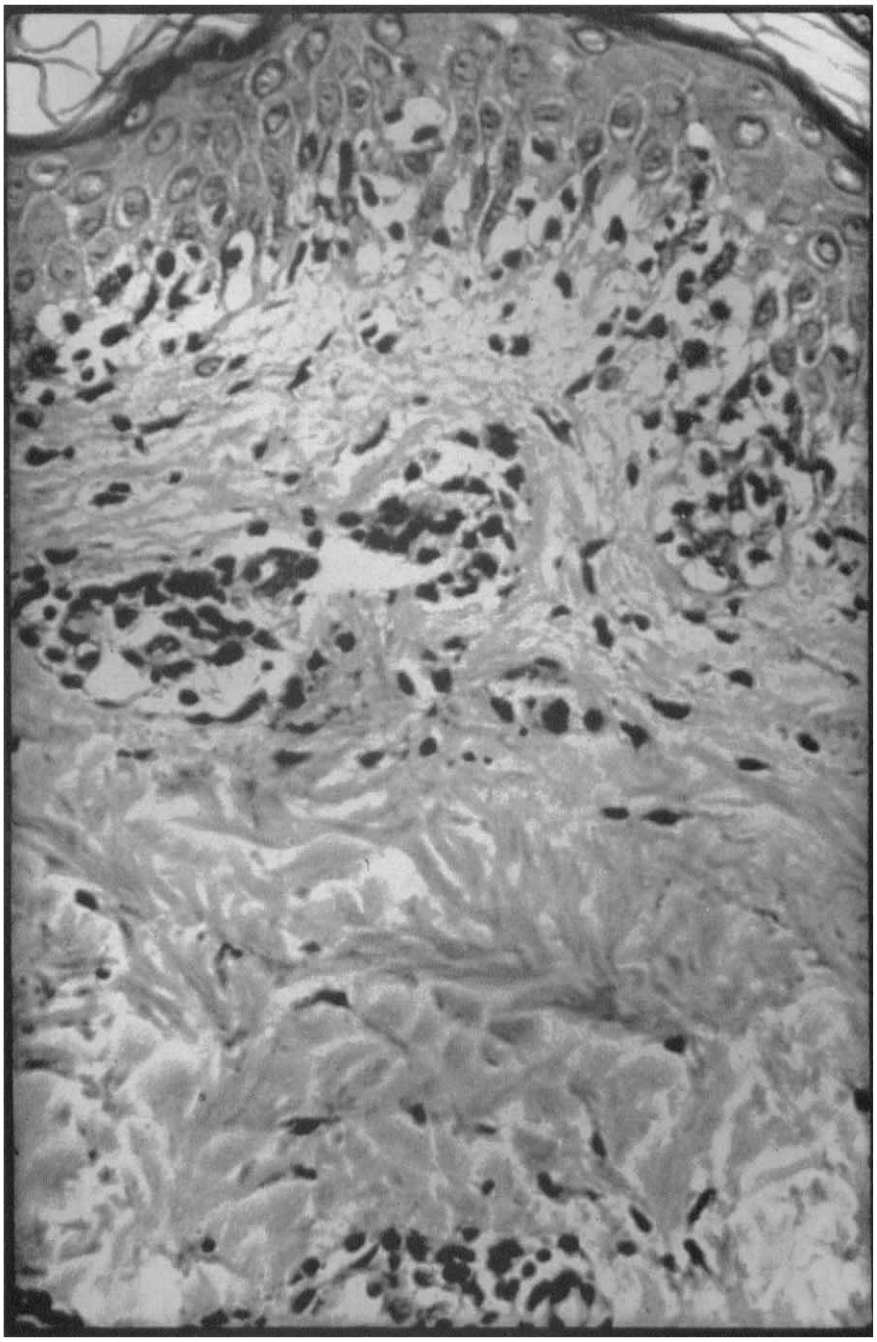

FIG. 5. Photomicrograph—× 200. Higher magnification demonstrating the hydropic degeneration of the basal layer of the epidermis.

figuratum melanodermicum and the other is erythema dyschromicum perstans (the latter suggested by Dr. M. B. Sulzberger).

Five cases have been seen and examined in Caracas, Venezuela. The condition is characterized by grayish slate-colored macules with slightly raised, erythematous borders. The histopathologic picture is reported. The differential diagnosis as regards to pinta and erythema perstans are discussed. The authors believe the condition belongs to the latter group. The etiology is unknown and the disease is chronic and capable of diffuse spread over the body. It is essentially asymptomatic. The condition is resistant to large doses of penicillin as well to broad spectrum antibiotics and corticosteroids.

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