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CASE REPORT

Progressive and extensive hypomelanosis and extensive pityriasis alba: same disease, different names?

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ABSTRACT

We report the cases of five female patients with high skin phototype affected by relapsing, hypochromic, non-scaling macules occurring after the summer on the back and spreading over large areas of skin. Histological features disclosed decreased epidermal melanin. Psoralen plus ultraviolet A (PUVA) treatment proved to be beneficial, but new relapses were noted after stopping treatment. Clinical and histological features were consistent with the diagnosis of 'progressive and extensive hypomelanosis' described by Guillet in persons of mixed racial background. We discuss the differential diagnosis of the latter entity with respect to the other idiopathic acquired primitive hypomelanosis and hypothesize an overlapping with the so-called extensive pityriasis alba (EPA).

Key words: dyschromia, hypomelanosis, hypopigmentation, melanodermic metis, pityriasis alba, PUVA

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'Progressive and extensive hypomelanosis', also called 'creole dyschromia' or 'progressive and confluent hypomelanosis of the melanodermic metis', is a primary, acquired, widespread hypopigmentation described by Guillet in 1988 in females from 18 to 25 years of age of mixed ethnic origin.[1] Typical lesions consist of hypochromic, non-scaling macules developing on the back and abdomen.

We report five new cases consistent with this entity and discuss its relationship with other conditions showing similar clinical features.

Case reports

All our five patients were young adult females, 19–25 years of age, skin type IV–V, three of Caucasian, one of North-African (Morocco) and one of Afro-Caribbean origin. The duration of the eruption ranged from 3 to 6 years. Three of them referred a high number of sun exposures before the occurrence of the dermatosis. There was no seasonal variation in the condition, although four out of the five patients reported an improvement during summer with rapid relapse of the skin manifestations in September after stopping sun exposures. Four patients reported having previously performed several treatments with general and topical antimycotics due to a diagnosis of tinea versicolor, or with steroids and emollients without benefit.

Anamnesis and physical examination excluded the presence of atopic disorders; a mild xerosis was present in two patients. The clinical features were similar in all cases and consisted of numerous, hypopigmented, well-defined circular or oval non-scaling patches, 0.5–3 cm in diameter, which were asymptomatic and scattered over the dorsum and shoulders. Sometimes the chest, abdomen and the proximal portions of the arms were involved (figs 1 and 2). In one case the patches were scattered

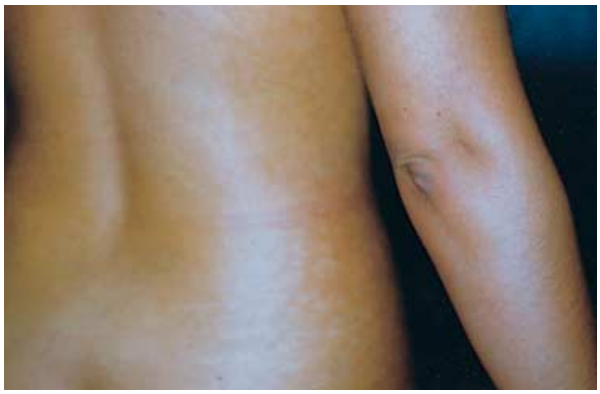

fig. 1 Multiple hypopigmented, non-scaling, non-coalescing patches involving the dorsum.

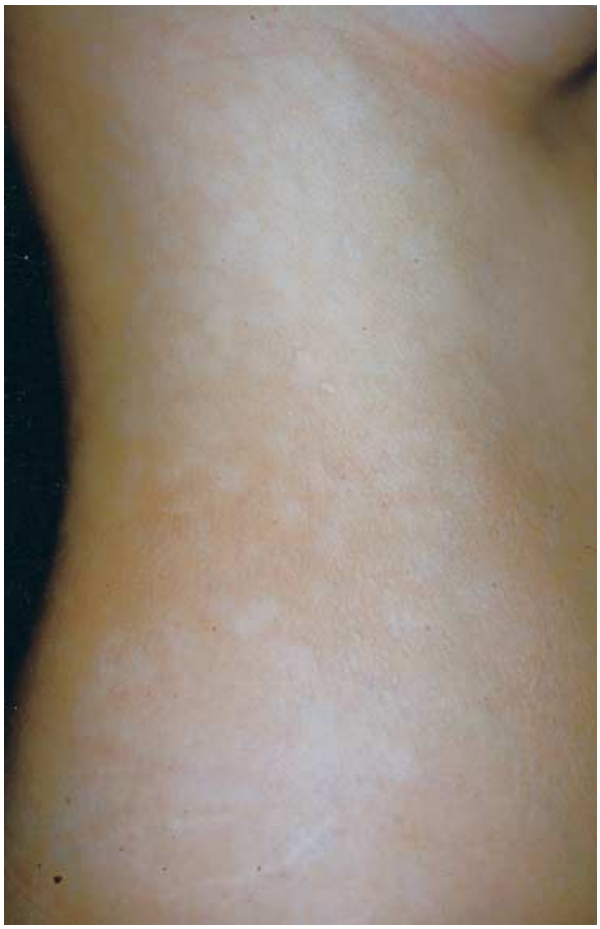

fig. 2 Numerous hypopigmented patches on the trunk, resembling tinea versicolor.

over the nape and thighs as well. No signs of inflammation were present on the borders of the lesions, which had gradually increased in number with a tendency to coalesce; however, the borders of the lesions remained well defined. No atrophy or scarring was noted. The remainder of the physical examination was negative. Family members did not show any similar skin manifestations.

Routine laboratory tests were performed in all patients and did not disclose any abnormality. Scrapings for potassium hydroxide (KOH) examination and gram stain were taken from all patients and were repeatedly negative for fungi and bacteria. There was no fluorescence under Wood's light. Biopsy for routine examination from affected skin of four patients showed mild hyperkeratosis in the epidermis and a reduction in the amount of melanin in the basal layer; in the upper dermis there was mild papillomatosis and sometimes a slight perivascular inflammatory lymphocytic infiltrate with a few melanophages.

Four patients were treated with psoralen plus ultraviolet A (PUVA) therapy. After 8 weeks of treatment two patients showed a marked improvement, rapidly followed by relapsing hypopigmented patches after the treatment was stopped. Two patients showed no significant or persistent improvement but reported a spontaneous improvement after almost completely avoiding sun exposure during the follow-up. Because of pregnancy, one patient was not treated with PUVA and is still presenting the disease.

Discussion

Hypochromic macules on the back are a frequent complaint after summer, especially in tropical regions. 'Progressive and extensive hypomelanosis' was reported in young females of mixed ethnic origin in their twenties in 1988.[1] The condition is characterized by macular hypopigmented patches, 10–30 mm in diameter, appearing in the dorso-lumbar area and spreading towards the lumbo-sacral region, increasing in number and progressively coalescing over the whole trunk into larger patches surrounded by smaller well-defined macules. The resulting hypomelanosis is most apparent in dark-skinned individuals. Histological features show decreased epidermal melanin. Ultrastructural examination in two black patients revealed a variation in melanosome size and distribution with a decrease in production of stage IV melanosomes within the hypopigmented macules. A possible spontaneous improvement could be observed within 5 years. All patients were young females observed by French researchers between 1988 and 1994.[1–4] Although this condition appeared to be frequent, no further cases have been reported in the literature in the past 10 years.

In 1983 Zaynoun *et al.* reported, under the name of extensive pityriasis alba (EPA), nine cases of an acquired hypopigmentation occurring in dark-skinned young adult females from the Lebanon.[5] Although the single skin lesions resembled those of pityriasis alba, the widespread and symmetrical involvement of the skin of the trunk with numerous, round, non-scaling hypomelanotic patches without a preceding inflammatory phase and with a long-lasting duration were characteristic of EPA. Histological examination showed a decrease in epidermal melanin; spongiosis was absent. Ultrastructural studies suggested that this hypopigmentation resulted primarily from a reduced number of active melanocytes and a decrease in the number and size of melanosomes in the affected skin. No atopy or associated pathologies or familial cases were reported. In a later study, Zaynoun *et al.* reported good effects with PUVA treatment, although follow-up was not provided.[6] No further cases of EPA have been reported in the literature. The correct relationship between classical pityriasis alba and EPA remains controversial: not only the distribution of the skin lesions but also the age of occurrence, the sex ratio (female preponderance), the lack of a preceding inflammatory phase and the absence of spongiosis sharply differentiate EPA from the classical form.

In our study five patients in good health affected by a diffuse hypochromic dermatosis are reported. The clinical and histological

features were consistent with the diagnosis of both entities described, respectively, by Guillet and Zaynoun and c-workers.[1,5] All the patients were young adult females with high skin phototype. Multiple skin patches recurred for several years with alternate periods of exacerbation and remission without coexisting familial cases. No correlation with atopy was noted. All cases appeared refractory to therapy. Some of our patients reported a high number of sun exposures before the occurrence of the dermatosis. A significant but transient improvement was reported during sun exposure, as well as after PUVA treatment. The latter gave us the impression of inducing a significant remission, although this was followed promptly by new relapses of the skin manifestations.

In conclusion, we think that a consistent differential diagnosis cannot be made between the cases of EPA and the progressive and extensive hypomelanosis described by Guillet. In particular, the sex ratio, age of occurrence (15–25 years), seasonality (end of summer) and skin phototype of the affected patients (III–V) are the same. The clinical manifestations of the two conditions are indistinguishable and the persistent evolution with poor response to the therapy is common to both diseases. Similarly, a decrease in epidermal melanin is the same histological change, while a reduction in the size of the melanosomes can be found on ultrastructural examination of affected areas in both conditions.[1,5]

In the differential diagnosis it is necessary to consider tinea versicolor, pityriasis rotunda, postinflammatory hypomelanosis and classical pityriasis alba.

Skin patches in tinea versicolor are scaly and may have erythematous raised margins. They can become confluent, showing a red–brown change of colour. In addition, KOH preparations are positive for Pityrosporum orbiculare (Malassetia furfur). Pityriasis rotunda affects both men and women, can be familial, with endemic areas in some geographical areas, such as the isle of Sardinia in Italy, Japan, south of Africa, and the Antilles. It is considered a rare variant of ichtyosis and is characterized by round or polycyclic, uniformly scaling, sometimes hypopigmented patches. Patients may have summer remissions and winter exacerbations. Post-inflammatory hypopigmentation can be ruled out due to the absence of a preceding dermatitis or inflammatory rash. Pityriasis alba has an equal incidence in boys and girls. Skin patches show a slight overlying scaliness and elevated borders. Seasonal variation with exacerbation in the summer and the winter has been reported. Skin lesions often present a preceding inflammatory phase and are usually located on the face, although involvement of the arms and shoulders is common. Spongiosis is the most consistent finding on histological examination; focal parakeratosis and mild acanthosis can also be noted.[7]

In conclusion, progressive and extensive hypomelanosis seems to be a pathology that is common in clinical practice but is under-reported, not well clarified and is difficult to manage. Further studies are necessary to confirm the hypothesis of a possible overlap with EPA and the existence of the latter entity.

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