CORRESPONDENCE

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ON ATOPIC AND IDIOPATHIC EXTENSIVE PITYRIASIS ALBA

To the Editor:

We read with interest the description of extensive pityriasis alba (EPA) in a child with atopic dermatitis (AD) reported recently by Sandhu et al (1). In that article the main differences between classical pityriasis alba (PA) and EPA, a condition described by Zaynoun et al in 1983 in nine dark skinned young women from Lebanon (2) are discussed. Although the single skin lesions of EPA do not differ substantially from those of pityriasis alba, consistent differences are a widespread, symmetric involvement of the skin of the trunk by numerous, round, nonscaly, hypomelanotic patches without a preceding inflammatory phase and with long-lasting duration. Histologic examination shows a decrease of epidermal melanin; spongiosis is 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. No atopy, associated pathologies, or familial occurrences were reported.

In our opinion the term EPA is a misnomer. There is no clear evidence of a relationship between EPA and classical PA: 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 the two conditions. In addition, PA usually presents with skin patches showing small amounts of overlying scale and elevated borders. Seasonal variation with exacerbation in the summer and the winter is common. Skin lesions are often located on the face, although involvement of the arms and shoulders sometimes occurs. Widespread lesions of classical PA can be observed in atopic dermatitis, but they should not be confused with the disorder described by Zaynoun et al (2). Current knowledge would lead us to suggest that EPA overlaps with another condition described by Guillet et al in 1988 as progressive and extensive hypomelanosis in persons of mixed racial background (3) and reported also as progressive and confluent hypomelanosis of the melanodermic metis or Creole dyschromia (4–6). This is a primary, acquired hypopigmentation observed in females from 18 to 25 years of age of mixed ethnic origin and characterized by hypochromic, nonscaly macules that develop on the back and abdomen, increase in number and progressively coalesce over the whole trunk into larger patches surrounded by smaller, well-defined macules. The resulting hypomelanosis is most apparent in dark-skinned individuals. Comparison between this condition and EPA does not show any substantial differences regarding age and sex distribution, seasonality (end of summer), and skin phototype of the affected patients (3–5). The clinical manifestations of these two hypomelanotic disorders are indistinguishable and the persistent evolution with a poor response is common to both. The same histologic change, a decrease of epidermal melanin, and a reduced size of the melanosomes, can be found on ultrastructural examination of affected areas in both conditions (2,3).

Recently we observed five female patients with high skin phototype affected by relapsing, hypochromic, nonscaly macules that occurred after the summer on the back and spread over wide areas of skin. The clinical and histologic features were consistent with the diagnosis of both these entities. Details of these patients are published elsewhere (7).

In conclusion, we think that the condition described by Zaynoun et al under the name of EPA (2) is a distinct disorder that cannot be considered an uncommon variant of PA. The latter can occur with multiple and diffuse skin lesions in atopic dermatitis, as in the patient of Sandhu's et al (1). From this point of view their observation that the morphology of EPA lesions may not be different from that of classical PA is correct. The denomination of EPA suggested by Zaynoun et al (2) is a misnomer that can create confusion. It is a condition not related to atopy, occurring in young adult females with chronic duration. To avoid misunderstanding, it should be regarded and identified possibly under the term progressive and extensive hypomelanosis, which is a different descriptive term defining the same disease.

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EARLY SKELETAL SIGNS IN NETHERTON SYNDROME

To the Editor:

Only a few authors (1–3) have reported radiologic data in patients with Netherton syndrome (NS): those patients were more than 3 years old and all showed increased bone density. We suggest that in this condition a generalized disturbance of the osseous development is present, noticeable soon after birth as small foci of osteosclerosis in knee epiphyses and vertebral bodies.

We report a 4-month-old boy born at term after an uneventful pregnancy. He was the first child of healthy, unrelated parents. Family history revealed atopy in the mother's family (mild atopic dermatitis and asthma) and psoriasis in the father's family (two uncles). At birth the boy was normal, with growth parameters between 50th and 75th percentile. At age 2 weeks he developed perioral erythema with subsequent spreading to the entire body. In spite of breast feeding and oral and topical steroids his condition progressively deteriorated. At 4 months he was admitted to the hospital in a very serious condition. He was lethargic, with hepatomegaly, severe anemia (Hb 5.2 g%), thrombocytopenia (56,000/mm^3), and hypoalbuminemia (1.3 g/dl).

His growth parameters were all below 3 SD. He had alopecia and generalized exfoliative erythroderma with skin induration. Serum IgE was greatly increased (410 IU/ml, which rose to 8000 IU/ml during the hospitalization) and eosinophil counts were elevated (20%). A graft versus host reaction was excluded (no circulating maternal cells). Serum levels of IgG, IgA, and IgM, serum zinc level, T lymphocyte subpopulations, mitogenic responses, complement components, and karyotype analysis had normal results. Histologic examination of the skin showed findings compatible with those of

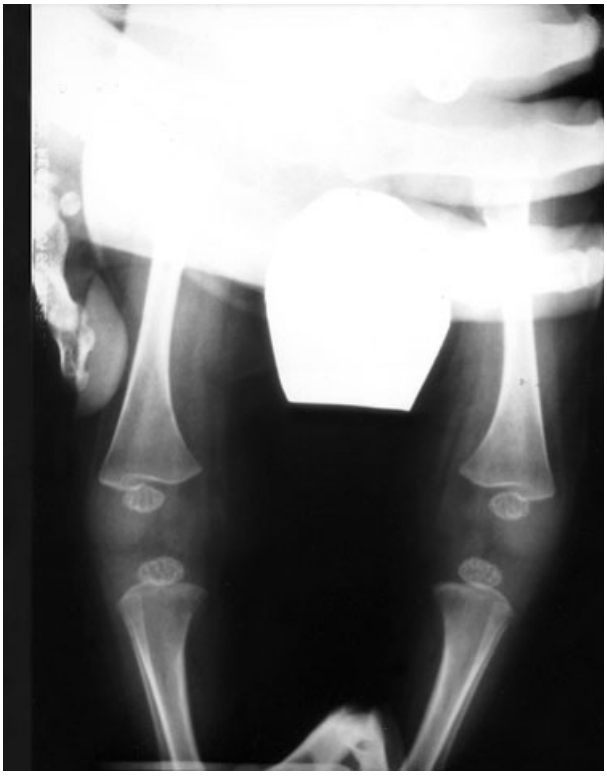

Figure 1. The knees show grains in both epiphyses.

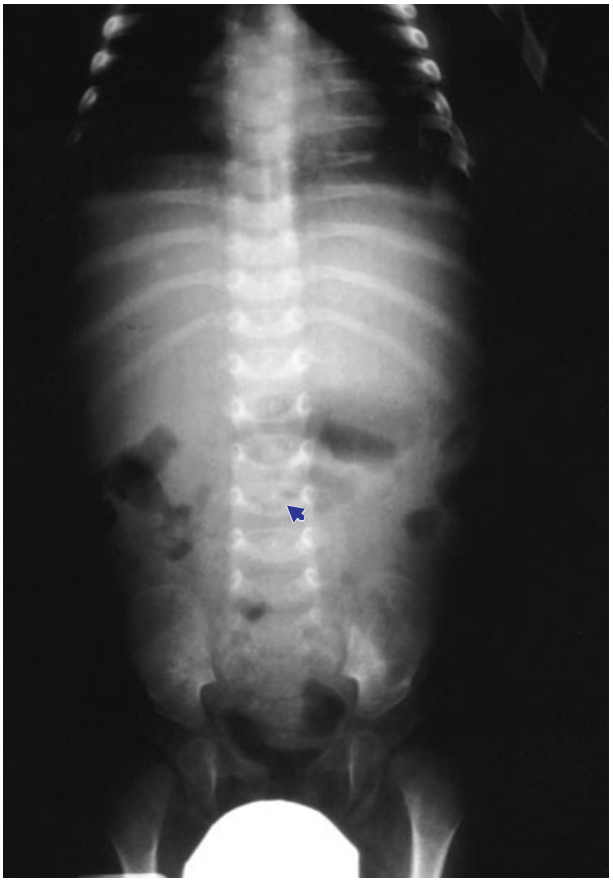

Figure 2. Anterioposterior view of the spine: hyperdense marks are visible in the middle of the vertebral bodies (arrow).