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Pityriasis Lichenoides Chronica in Black Patients

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Pityriasis lichenoides chronica (PLC) is a cutaneous disease of unknown etiology that most commonly affects children and young adults. The highly variable presentation of this condition often poses a diagnostic challenge. The clinical presentation of PLC in black patients is not well described. We report a series of 5 black patients (4 children and 1 young adult) with PLC who presented with extensive hypopigmentation and prominent facial involvement. One patient had concomitant mycosis fungoides (MF). The diagnosis of PLC should be included in the differential diagnosis in dark-skinned patients who present with widespread hypopigmented macules and patches. The presence of MF in one of our patients underscores the potential relationship between MF and PLC.

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Pityriasis lichenoides is a T cell–mediated papular dermatosis of unknown etiology that lies along a continuum of acute and chronic eruption, both with similar but distinguishable clinicopathologic features. The chronic form of pityriasis lichenoides, pityriasis lichenoides chronica (PLC), most commonly occurs during the early decades of life and classically presents with red-brown dusky papules with adherent micaceous scale often giving a frosted glass appearance.[1-5] In contrast, the acute form of the disease, pityriasis lichenoides et varioliformis acuta (PLEVA), has a tendency for a more papulonecrotic appearance with hemorrhagic crusts and occasionally vesicopustules.[4] However, it is not uncommon for individuals to present with lesions characteristic of both the acute and chronic forms.[6-8] Individual lesions occur in crops and last weeks to months, but the disease typically has an overall chronic clinical course of several months to years.[6-8] In this report we describe a series of 5 black patients (4 children and 1 young adult) with PLC presenting as widespread dramatic and persistent hypopigmentation with prominent facial involvement. Additionally, one patient had concomitant mycosis fungoides (MF).

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The authors report no conflict of interest.

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Case Reports

Patient 1—An 8-year-old girl presented with a 3-month history of a minimally pruritic, generalized, hypopigmented rash. Review of the patient's medical record revealed that she had been treated with intramuscular penicillin G benzathine for a scarlatiniform rash 8 weeks prior to the onset of her skin hypopigmentation. She had been subsequently diagnosed with tinea versicolor, atopic dermatitis, pityriasis alba, and vitiligo. Prior treatments consisted of triamcinolone acetonide ointment 0.1%, emollients, selenium sulfide shampoo 2.5%, and oral antihistamines, all without benefit. On physical examination the patient had extensive ill-defined hypopigmented macules and patches with no infiltration, erythema, or scale, and sparse red-brown minimally scaly papules (Figure 1). Facial involvement was prominent (Figure 2). Histology of a thin papule from the shoulder revealed parakeratosis with focal interface alteration and lymphocytic exocytosis, slight spongiosis, and sparse perivascular lymphocytic infiltrate consistent with PLC (Figure 3). This patient was seen sporadically over the following 2 years, and despite treatment with topical corticosteroids and oral erythromycin (30 mg/kg daily), persistent pigmentary alteration was notable. She was subsequently lost to follow-up.

Patient 2—A 2-year-old girl presented with a several month history of hypopigmented macular lesions primarily involving the face and extremities with

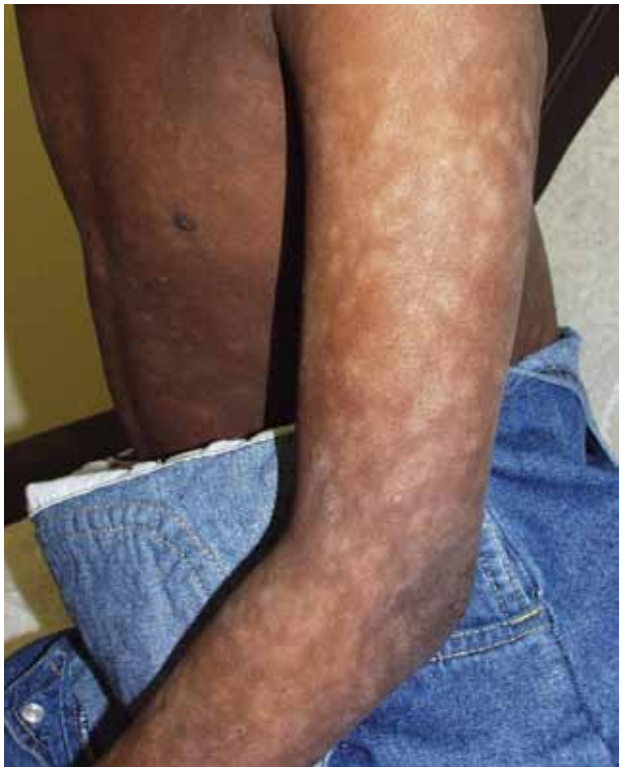

Figure 1. Generalized ill-defined hypopigmented macules and patches, and sparse red-brown minimally scaly papules.

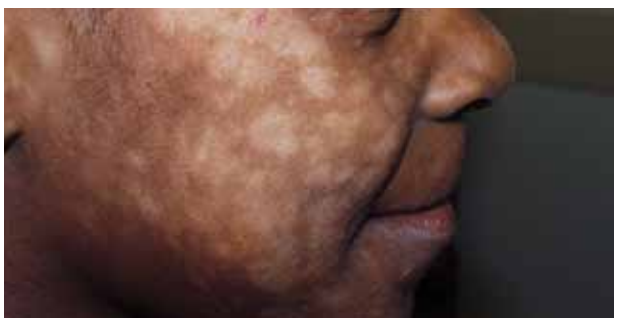

Figure 2. Scattered hypopigmented macules and thin scaling papules with prominent facial involvement.

relative sparing of the trunk. Upon further questioning, family members reported an antecedent pruritic papulovesicular eruption of the involved areas as well as concomitant cough, rhinorrhea, diminished appetite, and fever. On examination the patient had numerous hypopigmented ill-defined macules and sparse scaling hyperpigmented thin papules. Tentative diagnoses were Gianotti-Crosti syndrome versus PLC, and the patient was prescribed oral erythromycin (40 mg/kg daily) and triamcinolone acetonide ointment 0.1%. Histology of a hyperpigmented scaly papule from the anterior thigh revealed parakeratosis,

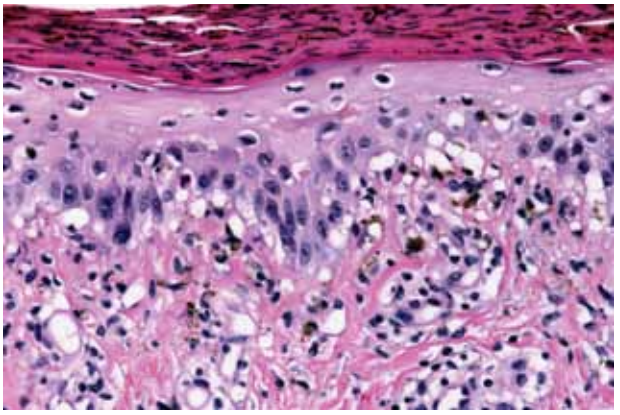

Figure 3. Parakeratosis with focal interface alteration, slight spongiosis with lymphocytic exocytosis, and sparse perivascular lymphocytic infiltrate (H&E, original magnification ×20).

spongiosis, focal vacuolar interface alteration, superficial perivascular lymphocytic infiltrate, and few scattered extravasated red blood cells in the papillary dermis consistent with pityriasis lichenoides. Her therapies included oral erythromycin (40 mg/kg daily), triamcinolone acetonide ointment 0.1%, and tacrolimus ointment 0.1% over 5 months. At follow-up 5 months later, the hypopigmented macules were still present. She subsequently failed to return for reevaluation.

Patient 3—A 27-year-old woman presented with a 4-year history of progressively worsening hypopigmentation on her face and extremities. She denied associated symptoms, but upon further inquiry she reported that many of the hypopigmented macules were preceded by small itchy scabs. She had no history of atopic dermatitis, and her medical history was unremarkable. Prior therapies, including griseofulvin, selenium sulfide shampoo 2.5%, lactic acid lotion 12%, and triamcinolone acetonide ointment 0.1%, were ineffective. Examination revealed numerous ill-marginated, 2- to 4-mm, hypopigmented macules distributed on the face (Figure 4), arms, and thighs. Amidst the hypopigmented macules were rare 2-mm red papules, several of them with an overlying adherent scale. Potassium hydroxide preparations were negative. Two skin biopsies, each from papular lesions from her extremities, revealed parakeratosis, mild spongiosis, focal vacuolar interface alteration, and a superficial and mid-dermal perivascular lymphocytic infiltrate consistent with PLC. No therapy was instituted due to the patient's newly noted pregnancy status; however, UV therapy was planned after delivery.

Patient 4—An 11-year-old boy presented with a several year history of asymptomatic, erythematous, scaly patches with hyperpigmented margins on the

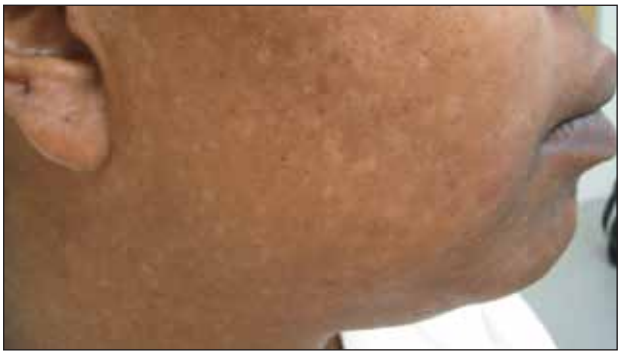

Figure 4. Hypopigmented macules on the face with focal thin scaling papules.

trunk. Lesional skin biopsies from an erythematous patch on the buttock revealed an epidermotropic lymphocytic infiltrate with marked lymphocyte atypia consistent with MF. T-cell receptor γ gene rearrangement analysis on paraffin-embedded tissue was negative. On a subsequent visit months later he was noted to have ill-defined macular areas of hypopigmentation on his cheeks and proximal extremities with sparse, widely scattered, thin, red-brown papules with micaceous scale that were not present at the initial visit. The patient's parents reported that the erythematous patches on the trunk were longstanding and persistent but that the hypopigmented macules developed subsequent to papular lesions that occurred in crops. Based on his history and clinical presentation, dual diagnoses of MF as well as PLC were made. On follow-up 2 months later he was noted to have dozens of red-brown papules amidst hypopigmented macules on the upper chest, face, and extremities, again consistent with PLC. The truncal lesions, consistent with MF, remained unchanged, as the patient had not yet begun treatment (Figure 5A). Nitrogen mustard ointment 0.01% was initiated, and on follow-up 9 months later, marked improvement in the MF lesions was noted (Figure 5B). However, the patient and his parents noted no effect on the PLC lesions. He was then started on doxycycline monohydrate for the PLC and nitrogen mustard ointment was continued. He currently is awaiting follow-up.

Patient 5—A 4-year-old boy presented for evaluation of a widespread hypopigmented, minimally pruritic, macular eruption involving the face, trunk, and extremities. On close inspection approximately 10 hyperpigmented thin papules with overlying scale were noted on his extremities and back in addition to the hypopigmented macules. The pigmentary alteration was first noted at approximately 6 months of age, at which time no clear diagnosis was made. Due to progression of the hypopigmentation, reevaluation was again sought at 2.5 years of age. Skin biopsies at that time and again at 4 years of age both revealed a superficial and mid-dermal perivascular lymphocytic infiltrate with mild spongiosis, focal vacuolar interface alteration, and overlying parakeratosis. Rare extravasated red blood cells were present. A 6-month course of treatment with oral erythromycin was ineffective. Fluticasone propionate ointment 0.005% reportedly had minimal effect. A 6-month course of UVB therapy resulted in improvement but was discontinued when the patient moved. Currently he reports continued benefit, particularly with regard to repigmentation of the facial lesions, with frequent regular intervals of natural sunlight exposure.

Comment

Pityriasis lichenoides is a benign T cell–mediated dermatosis of unclear etiology that lies along a disease continuum of acute (PLEVA) and chronic (PLC) eruptions, both with similar but distinguishable clinicopathologic features. Histologically, PLEVA and PLC exhibit varying degrees of dermal inflammation, perivascular lymphocytic infiltrates, parakeratosis, focal vacuolar interface alteration, epidermal necrosis, and extravasated red blood cells.[9] Both eruptions contain lesional T cell infiltrates, with PLEVA being primarily $CD8^+$ dominant and PLC being principally a $CD4^+$ infiltrate.[3] Because T cell monoclonality has been demonstrated in the infiltrate in some cases of pityriasis lichenoides and rare cases of association with lymphoproliferative disorders have been reported, a relationship between pityriasis lichenoides, lymphomatoid papulosis, and even cutaneous T cell lymphoma has been proposed.[4,9]

The classic clinical presentation of PLC consists of asymptomatic, red-brown, lichenoid papules with adherent micaceous scale. Transient leukoderma or dyspigmentation also is characteristic but rarely has been reported as the presenting feature, as was the case in several of our patients.[1] The distribution of lesions varies, but until recently, the vast majority of reported cases of PLC involved the extremities and trunk, with relative sparing of the face. In a review of 22 pediatric cases of pityriasis lichenoides, Romani et al[9] found a striking minority (1/14 [7%]) demonstrated involvement of the head and neck. In fact, this topographic trend has been offered as a distinctive clinical feature to differentiate PLC from the pigmentary alteration induced by other primary processes such as pityriasis alba and vitiligo.[1,6] However, a retrospective review of 57 patients by Wahie et al[8] suggested that facial involvement in children may not be as uncommon as previously recognized.

In our case series, all patients were black, each presented with prominent facial involvement, and

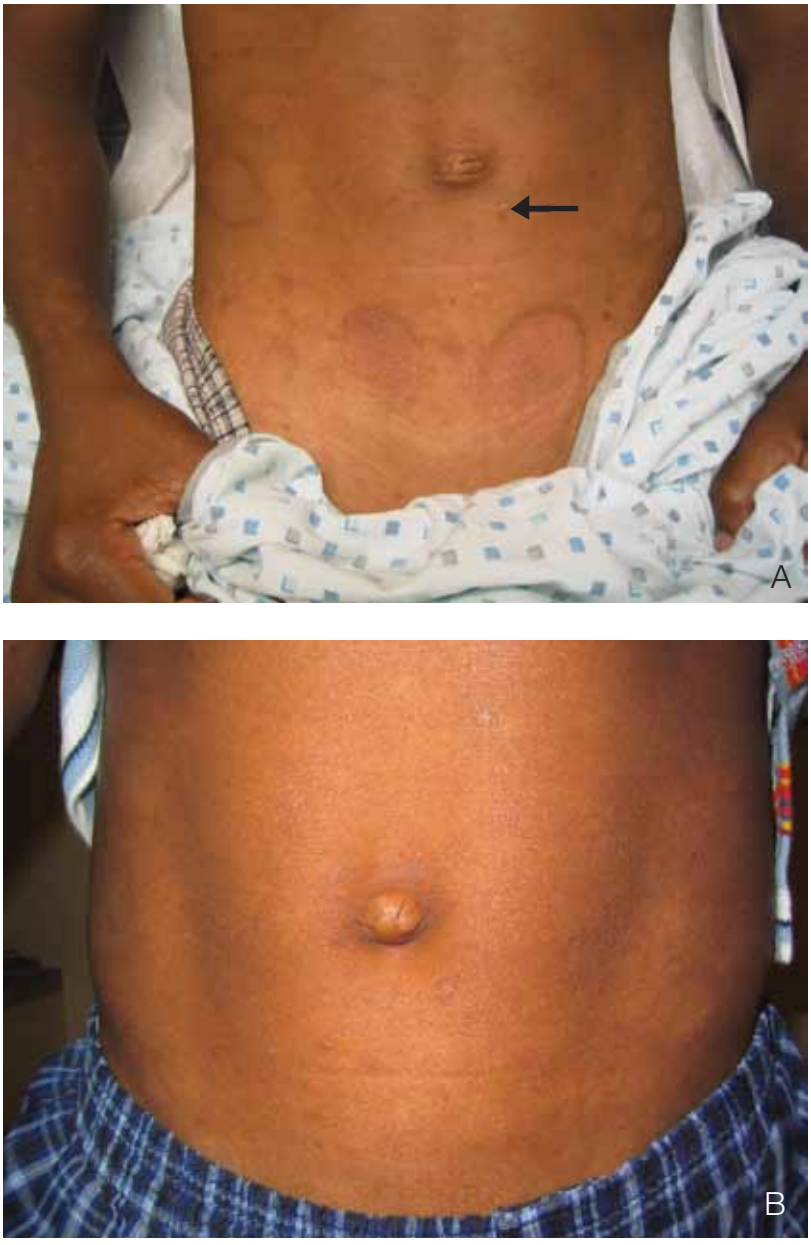

Figure 5. Scaly patches with hyperpigmented margins on the trunk and thin red-brown papules with micaceous scale (arrow)(A). The patient exhibited improvement in mycosis fungoides patches after treatment with nitrogen mustard ointment 0.01%, with persistence of pityriasis lichenoides chronica papules (B).

hypopigmentation was the predominant clinical feature. In at least 4 of our cases (patients 1, 2, 3, 5), the diagnosis of PLC was not entertained at the initial presentation. In 4 of our cases, histology confirmed the diagnosis. Although follow-up data from some of our cases is limited, PLC-related hypopigmentation was evident for as long as 2 years in at least one of our patients.

Supporting the possible relationship between MF and pityriasis lichenoides, patient 4 had coexisting clinically and morphologically distinct lesions. Long-standing, discrete, erythematous, nummular patches characteristic of MF were distributed on the mid and lower trunk. Cutaneous biopsies of these lesions were consistent with MF. Subsequent to his initial visit, this patient also developed ill-defined hypopigmented macules on the face, extremities, and upper chest admixed with crops of papules characteristic of PLC. While hypopigmented MF was in the differential diagnosis, the history of an antecedent papular eruption in the involved areas was supportive of our clinical impression of PLC. In addition, at follow-up 9 months after therapy with nitrogen mustard ointment was instituted, the MF lesions were markedly improved, yet no change in the course of the papular lesions was evident, which further supported the presence of 2 potentially related but distinct disorders in our patient. We acknowledge the possibility that this patient may have had a papular variant of MF in addition to the morphologically more characteristic type

of MF lesion.[10,11] The complete lack of response to therapy of one type of lesion (papules) but near complete resolution of the other type (patches) makes this scenario unlikely. Another potential diagnostic consideration in this patient was lymphomatoid papulosis, which is not uncommon in patients with MF.[11] Based on the clinical history, the morphology of the lesions, and the pigmentary alteration, this patient's diagnosis was most consistent with PLC. We recognize, however, that it is often difficult, both clinically and histologically, to differentiate pityriasis lichenoides from lymphomatoid papulosis in some cases. Because the clinical presentation and histology of these conditions overlap, they may be pathogenically related processes, along with MF.

Although lesional skin of MF often has a clonal T-cell population, a clone may not be detectable in up to 10% to 15% of cases,[12,13] as was the case in patient 4. Molecular analysis of a lesion characteristic of PLC was not performed in our patient. However, given the absence of a detectable clone in his lesional skin with characteristic MF histology, this information would not have altered the diagnosis or the recommended therapy.

Based on the clinical presentation in our patients, the differential diagnosis of PLC with prolonged pigmentary alteration includes tinea versicolor, pityriasis alba, resolving guttate psoriasis, leprosy, syphilitic leukoderma, vitiligo, idiopathic guttate hypomelanosis, and hypopigmented MF. Examination with potassium hydroxide preparation and Wood lamp are useful in excluding the diagnosis of tinea versicolor and vitiligo, respectively. The diagnosis of PLC can be made on clinical grounds; however, skin biopsy for histopathologic confirmation is useful and often necessary.

Conclusion

We report a series of 5 black patients (4 children and 1 young adult) with PLC who presented with extensive hypopigmentation and prominent facial involvement. This presentation may not be atypical, particularly in darkly pigmented individuals, yet it is relatively underreported in the literature. Pityriasis lichenoides chronica should be considered in the differential diagnosis of any generalized hypopigmented macular eruption, particularly in dark-skinned individuals. Careful clinical examination is required, as the primary papular lesions can be quite subtle, undergo spontaneous involution, and frequently may go unnoticed by the patient. Although considered benign, patients with PLC require careful monitoring because of the remote possible relationship to lymphoproliferative disorders such as MF, as seen in one of our patients.

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