Lymphomatoid papulosis in a young adult of African descent

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INTRODUCTION
Lymphomatoid papulosis (LyP) is an indolent skin disease characterized by chronic, recurrent, self-healing papules and nodules with histologic findings suggestive of malignant lymphoma. LyP is classified as a CD30+ cutaneous lymphoproliferative disorder according to the 2005 World Health Organization/European Organisation for Research and Treatment of Cancer classification and has 5 histologic subtypes (A-E).1 Differential diagnoses of LyP includes other CD30+ cutaneous lymphomas, systemic T-cell or B-cell lymphomas, and reactive processes that exhibit CD30 expression.1 LyP rarely affects black skin, with only 2 previously reported cases.2,3 We present a case of LyP in a male of African descent.

CASE REPORT
A 24-year-old Afro-Caribbean man, with no significant medical history, was referred to the Howard University Hospital Dermatology Clinic with a 1-year history of multiple relapsing pruritic macules. He denied experiencing fever, chills, night sweats, or unexpected weight loss. Previous treatment with topical low and midpotency steroids was of no benefit.

On clinical examination, there were numerous hyperpigmented and erythematous, 3- to 5-mm macules on upper arms, trunk, and bilateral hips (Figs 1 and 2). There was no evidence of lymphadenopathy or other signs of systemic disease.

Histologic evaluation of a 4-mm punch biopsy specimen showed irregular epidermal hyperplasia, spongiosis, and a moderately intense inflammatory infiltrate of lymphocytes, eosinophils, and scattered atypical enlarged lymphocytes (Fig 3). Immunohistochemical stains found a prominence of CD3+ T cells, which showed some loss of expression of CD7 within the atypical cells. The CD4 to CD8 ratio was 4:1. CD30 shows increased positivity with some clustering within the atypical cells (Fig 4).

A diagnosis of LyP type B was made. Routine laboratory tests were normal, except for an initial antinuclear antibody, which was incidentally found to be positive, but ultimately revealed to be a false positive on repeat testing. Treatment with 3-times-a-week narrow-band ultraviolet B phototherapy resulted in improvement of pruritus and resolution of all lesions. There has been no evidence of new lesions since initiating therapy. Evaluation by the medical oncology team ruled out signs of malignancy. The patient continues to remain in close follow-up.

DISCUSSION
LyP is characterized by chronic, recurrent, self-healing papules and nodules. Because of this particular presentation, it is often misdiagnosed as folliculitis or arthropod bites. It may also be confused with pityriasis lichenoides. Other differential diagnoses to consider include cutaneous T-cell lymphoma, cutaneous anaplastic large cell lymphoma, leukemia cutis, drug eruption, or potential infectious entities, such as scabies, syphilis, or other bacterial or even fungal infections. Clinical pathologic correlation and a complete history are important in guiding a clinician to the proper diagnosis. In our patient, the lack of B symptoms
and signs of systemic disease favored a benign process over a malignant one. Clinically, suspicion for mycosis fungoides was very low, given the lack of prior or concurrent patches or plaques.

LyP predominantly affects whites from western countries, although many skin of color cases have been reported in East Asian countries\(^4\) as well as one in India.\(^5\) Our case represents the rare occurrence of LyP in a patient of African descent. After a thorough literature review, we found only 2 such cases reported.\(^2,3\) The first case describes a 47-year-old black man with LyP and concurrent plaque-stage mycosis fungoides who responded to topical carmustine therapy.\(^2\) The second case involves a 50-year-old black woman with LyP who presented with tuberculum sellae meningioma.\(^5\) Although both cases involved middle-age black patients, our patient was affected much earlier in adulthood. In general, adults are the most commonly affected group (median age, 45),\(^6\) although cases have been reported among children. Therefore, clinical suspicion is necessary in successfully diagnosing LyP in adolescents and young adults.

LyP is typically distributed over the trunk and extremities although may rarely affect the genital and oral mucosa.\(^8\) Our case was interesting in that it involved intertriginous and follicle-dense areas such as the axilla and inguinal regions. Because intertriginous areas are subject to a high degree of friction, such lesions may possibly be attributed to local trauma. Cases of localized LyP have been reported in the past, including 1 case describing a 20-year-old man with lesions in his left antecubital fossa, an intertriginous area.\(^9\) Some investigators have also described a follicular variant of LyP that is characterized by perifollicular infiltrates and folliculotropism on histology.\(^10\) However, our patient's histologic evaluation lacked such evidence.

Of the 5 histologic subtypes (A-E), LyP type A is the most common subtype and comprises 75% of all
LyP cases.\textsuperscript{1} The histology of our case is consistent with LyP type B, which is defined by a bandlike infiltrate consisting of small- to medium-sized cerebriform lymphocytes.\textsuperscript{1} Although CD30 is usually negative, a small number of positive cells may occasionally be seen in cases such as ours. Type A is characterized by a wedge-shaped infiltrate comprising large pleomorphic and anaplastic CD30+ lymphocytes.\textsuperscript{1} The admixed inflammatory infiltrate consists of histiocytes, neutrophils, and eosinophils. LyP type C is similar to type A but is characterized by sheets of large anaplastic CD30+ lymphocytes with fewer admixed inflammatory cells.\textsuperscript{1} LyP type D shows prominent epidermotropism of atypical small- and medium-sized CD8+ and CD30+ pleomorphic lymphocytes.\textsuperscript{1} The most recently described subtype, angioinvasive LyP (type E), consists of angiocentric infiltrates of pleomorphic CD30+ and CD8+ cells of variable size that form thrombi, which lead to vascular occlusion and ultimately cause necrosis and ulceration.\textsuperscript{1}

Despite sharing histologic features of malignant lymphoma, LyP fortunately has an excellent prognosis and some, therefore, recommend a “wait-and-see” strategy.\textsuperscript{1} Other strategies include phototherapy and low-dose methotrexate. LyP is associated with an increased risk of secondary lymphoma and thus requires lifelong surveillance in affected patients. We must have a high index of suspicion for diagnosing LyP in all skin types, including patients of African descent.

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