

Late-onset systemic lupus erythematosus associated with inverse discoid lupus erythematosus on the buttock



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INTRODUCTION

Discoid lupus erythematosus (DLE) is important to diagnose early, as treatment is critical for preventing permanent scarring and for identifying possible systemic involvement. Diagnosis is frequently aided by the presence of photosensitive lesions of the head and neck; however, in rare cases, photosensitive lesions may not be present, thus clouding the diagnosis. Importantly, more research is needed to highlight these atypical cases, since they can be associated with greater morbidity.

CASE REPORT

A 56-year-old Hispanic woman with a history of Hashimoto disease, gastritis, and hepatitis C was admitted for a multiday history of double and blurry vision with sharp, stabbing, bilateral eye pain, vertigo, and headaches. The patient also reported months of intermittent fevers, night sweats, and loss of appetite. Neurologic examination was significant for left abducens-nerve palsy, but she otherwise had normal strength, sensation, and motor skills. Skin examination revealed a large hyperpigmented scaly plaque with areas of atrophy on her superior aspect of left buttock and lower portion of the back, for which dermatology was consulted (Fig 1). Skin examination was otherwise unremarkable. No lesions were appreciated on the scalp, face, ears (including conchal bowls), or chest. Laboratory tests revealed normocytic anemia and elevated C-reactive protein and erythrocyte sedimentation rate. Magnetic resonance imaging without contrast was significant for 3 new subcentimeter hyperintense

Abbreviations used:

DLE: discoid lupus erythematosus
 SLE: systemic lupus erythematosus

lesions in the bilateral centrum semiovale. Lumbar puncture revealed sterile cerebrospinal fluid.

During initial dermatologic consultation, the patient noted that the rash on her back and buttock had been present for approximately 5 months and initially began as a tender pink plaque that enlarged and progressed to develop dyspigmentation and areas of scarring. The lesion was otherwise asymptomatic. She denied any history of similar lesions elsewhere on her body. After examination, 2 lesional skin punch biopsies were taken; one for hematoxylin-eosin staining, and the other for direct immunofluorescence test. The differential diagnoses at this time included cutaneous lupus (subacute cutaneous lupus erythematosus and DLE) versus dermatomyositis versus mixed connective tissue disease versus atypical allergic dermatitis. Histopathologic analysis of the hematoxylin-eosin staining revealed a patchy lichenoid interface dermatitis with superficial and deep periadnexal and perieccrine lymphoplasmacytic inflammatory infiltrate with background-increased dermal mucin deposition (Figs 2 and 3). Direct immunofluorescence test showed dense granular staining for immunoglobulin M, IgG, IgA, C3, C5b-9, and cytooid bodies along the epidermal and adnexal basement membrane zone. Given the extensive

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Fig 1. Discoid lupus erythematosus lesion isolated on the lower portion of the back and buttock.

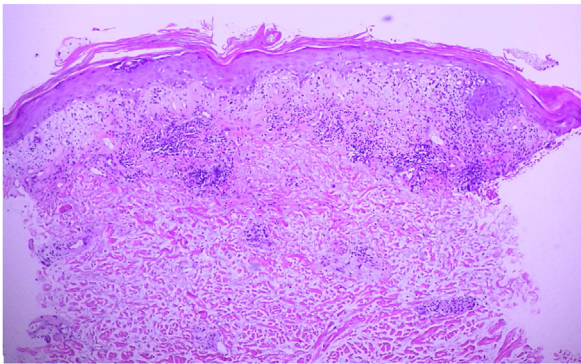


Fig 2. Discoid lupus erythematosus (hematoxylin-eosin stain; original magnification: $\times 4$).

immunoreactant deposition with adnexal involvement on direct immunofluorescence test and the clinical examination of an isolated, hyperpigmented, and atrophic plaque, DLE was favored over other diagnoses. Furthermore, the patient tested negative for anti-Ro antibodies, supporting the diagnosis of DLE over subacute cutaneous lupus erythematosus. Additional rheumatologic workup was significant for positivity for antinuclear, anti-double-stranded DNA, anti-Smith, and anti-ribonucleoprotein antibodies, and she was subsequently diagnosed with systemic lupus erythematosus (SLE) and lupus cerebritis in the setting of her neurologic manifestations. The patient's cutaneous disease improved after starting a topical calcineurin inhibitor in addition to systemic treatment with hydroxychloroquine and belimumab for her SLE.

DISCUSSION

DLE is the most common clinical variant (50%) of chronic cutaneous lupus erythematosus and disproportionately affects those with skin of color.^{1,2} Since DLE can lead to scarring and dyspigmentation, it is

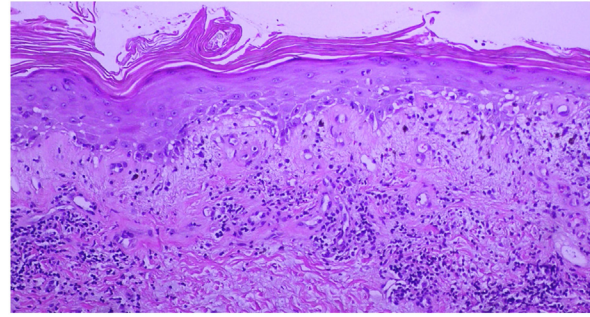


Fig 3. Discoid lupus erythematosus (hematoxylin-eosin stain; original magnification: $\times 10$).

critical to diagnose the condition early to prevent morbidity. DLE has a strong tendency to appear in photosensitive areas, such as the face, ears, and scalp; a feature that frequently aids in the diagnosis. DLE that occurs on photo-protected sites is rare and, when present, is most often associated with lesions concurrently present on photosensitive sites.³ This variant of discoid lesions appearing both above and below the neck is otherwise known as generalized/disseminated DLE and represents roughly 20% of all DLE cases. In contrast, localized DLE is commonly accepted as the presence of lesions confined to the head and neck. While there is no established classification for DLE lesions solely below the neck, our patient's presentation of an isolated lesion on the trunk might be best categorized as an atypical generalized DLE without head or neck involvement.

Another rare feature of this case is that our patient, with an isolated discoid lesion, went on to develop systemic lupus. Systemic lupus in association with DLE is uncommon; only approximately 50% of patients with discoid lupus are positive for antinuclear antibody, and only 5% to 20% develop systemic features of SLE.^{2,4} Those with DLE who do go on to develop systemic lupus mostly have mild, predominantly musculoskeletal, symptoms.⁵ Additionally, most patients with DLE who develop systemic symptoms have multiple discoid lesions located throughout the body.⁶ This patient, with an isolated lesion, developed systemic lupus in the form of lupus cerebritis, a more severe manifestation of systemic lupus. Lupus cerebritis describes the neuropsychiatric manifestations of SLE and most commonly manifests as headaches, cognitive dysfunction, and psychiatric disorders.⁷ Vision problems are less common and are considered a more severe symptom.⁷ Associated abducens-nerve palsy has been described in the literature, though rarely so.⁸

Overall, our case represents an unusual case of SLE in association with DLE. The patient presented

with an isolated discoid lesion on the photo-protected area of her lower portion of the back and buttock and subsequently went on to develop lupus cerebritis, a severe manifestation of systemic lupus. SLE can be difficult to diagnose given its frequent nonspecific and overlapping symptoms with many other different conditions. Photosensitive lesions can be a helpful clue to assess the diagnosis; however, our case demonstrates that overreliance on this identifying feature can lead to a situation where more insidious cases are missed. Hence, our case emphasizes the importance of high clinical suspicion and critical thinking in the absence of photosensitive lesions. Notably, given our patient's age, she technically qualifies as having late-onset SLE, which is when the disease develops after the age of 50.⁹ Late-onset SLE has been known to present with atypical SLE symptoms, and this may explain the unusual features of this case.¹⁰ Lastly, previous studies have highlighted that, although there is extensive data on patients with isolated DLE, there are few studies that have analyzed the clinical characteristics of SLE associated with DLE.² Of the few studies that do exist, the largest study found that these patients are at particularly increased risk of developing serious manifestations such as vasculitis and chronic seizures.² For this reason, identifying and surveilling these patients is critical, and more research is needed to further characterize this susceptible population.

Conflicts of interest

None disclosed.

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