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Recognizing ophiasis pattern in discoid lupus: a rare diagnostic challenge

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Abstract

Lupus erythematosus is an autoimmune condition that can present in both cutaneous and systemic forms, with discoid lupus erythematosus (DLE) being the most prevalent type of chronic cutaneous lupus. Although the ophioid pattern is well documented in alopecia areata, we present a rare case of DLE exhibiting this pattern in a 51-year-old Saudi female patient with a complex medical history, including systemic lupus erythematosus (SLE). This case underscores the importance of recognizing atypical presentations of DLE, which can lead to misdiagnosis and delays in treatment. We aim to enhance awareness among dermatologists regarding the diverse manifestations of DLE and encourage further investigation into such rare patterns.

Introduction

Lupus erythematosus is an autoimmune condition that can manifest as either limited to the skin or have systemic involvement. Among the various types of cutaneous lupus, discoid lupus erythematosus (DLE) is the most prevalent form of chronic cutaneous lupus erythematosus. While the risk of DLE progressing to systemic forms is relatively low (5%-10% over a lifetime), it can still result in significant morbidity.¹ The discoid lesions associated with DLE have the potential to cause scarring, and over time, many patients may develop disfiguring scars. These lesions commonly involve the hair follicles, leading to follicular plugging and subsequent scarring alopecia. Furthermore, in longstanding lesions, dyspigmentation is often observed, typically presenting as hypopigmentation in the central area and hyperpigmentation at the periphery.² When considering the sites most affected by discoid lupus erythematosus, the scalp is notably impacted, with involvement reported in approximately 30% to 50% of cases.³

Although DLE lesions classically present as multiple scarring alopecic patches and are most commonly localized to the vertex of the scalp,⁴ it sometimes poses a diagnostic challenge, especially among patients with atypical presentations. In this report, we present a rare instance of the ophioid pattern presentation in a Saudi female patient diagnosed with DLE. This case is significant as it underscores the importance of recognizing atypical manifestations of discoid lupus, which can lead to misdiagnosis and delays in appropriate treatment. We aim to raise awareness among dermatologists and clinicians about the diverse clinical presentations of DLE, particularly those that deviate from the more commonly recognized forms. By documenting this unique case, we seek to contribute to the existing literature and encourage further investigation into the implications of such rare patterns.

Case Report

A 51-year-old female with a complex medical history, including systemic lupus erythematosus (SLE), hypertension, diabetes mellitus, and hypothyroidism, presented to the dermatology clinic with a primary complaint of hair loss. This issue had developed over the past two years, characterized by hairless patches, redness, and itching of the scalp. Her current medication regimen included rituximab, hydroxychloroquine, prednisolone, nifedipine, irbesartan/hydrochlorothiazide, amitriptyline, and levothyroxine.

Upon physical examination, two broad patches of scarring hair loss were noted over the bilateral occipital scalp, including the posterior hairline, accompanied by faint erythema (Figure 1).

A skin biopsy taken from the erythematous area demonstrated perivascular and periadenxal lymphocytic infiltrate along with follicular plugging (Figure 2).

Based on these findings, a diagnosis of scalp DLE with an ophiasis pattern was established. The patient received a treatment regimen consisting of Betamethasone scalp lotion combined with triamcinolone acetonide injections (1 mL; 5 mg/mL).

Discussion

The ophiasis pattern is characterized by a distinctive band-like distribution of hair loss. While this pattern is well-documented in alopecia areata, our case presents a novel perspective by illustrating the ophiasis pattern in discoid lupus erythematosus. To our knowledge, this is the first case describing such a presentation.

Hair loss is a common manifestation in SLE and is observed in over 50% of patients at various stages of the disease.^{5,6} While various patterns of hair loss can be observed in individuals with SLE, the underlying cause is not always directly attributed to lupus itself.⁷ It is of utmost importance to distinguish whether alopecia is an inherent manifestation of SLE or merely a coincidental finding, as this determination carries significant clinical implications.⁷

Lupus erythematosus (LE)-specific alopecia is commonly characterized by scalp DLE, which typically presents with scarring.^{6,8} Various forms of non-scarring hair loss, including conditions like lupus hair, alopecia areata, and telogen/anagen effluvium, do not exhibit biopsy characteristics specific to LE. Instead, these types of hair loss are generally regarded as either non-specific to LE or coincidental occurrences in individuals with LE.⁸

Uncommon patterns of alopecia associated with DLE have been reported in the literature, highlighting the diverse presentations of hair loss in this condition. Linear cutaneous lupus erythematosus following

lines of Blaschko, particularly DLE involving the scalp, is infrequently reported in medical literature. Only a few cases were reported that specifically describe isolated linear DLE affecting the scalp.^{9,10} This report presents the first documented case of DLE exhibiting the ophiasis pattern, a phenomenon that has not been previously reported in the literature. It is essential to acknowledge and identify the various forms of alopecia that may manifest in patients with SLE, recognizing their clinical patterns regardless of how atypical they may appear. As illustrated in our case, the patient presented with an ophiasis pattern typically associated with alopecia areata; however, biopsy results confirmed the diagnosis of DLE. The presented case highlights the importance of comprehensive evaluation and diagnostic investigations in accurately determining the underlying cause of alopecia in SLE patients, even when the clinical presentation is unusual. By enhancing awareness of such atypical presentations, we can improve diagnostic accuracy and optimize patient care.

Conclusions

Our case highlights an unusual manifestation of DLE that resembles other conditions, particularly alopecia areata. The ophiasis pattern, known for its distinctive distribution, may complicate the diagnostic process. Our findings underscore the importance of identifying these atypical presentations to prevent misdiagnosis and facilitate effective management.

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Figure 1. Band-like pattern of hair loss in the left parieto-occipital areas extending toward the temples.

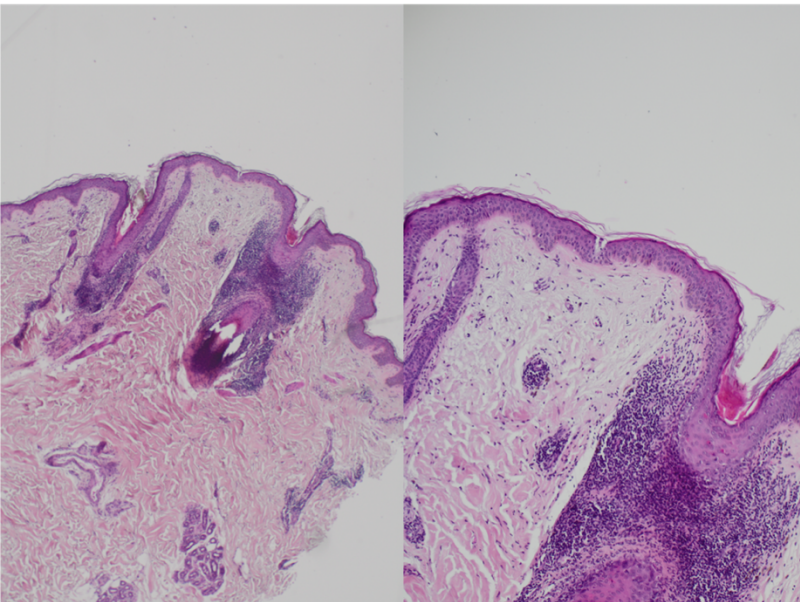


Figure 2. Pathological features associated with discoid lupus erythematosus, highlighting typical epidermal changes such as acanthosis and atrophy. Additionally, a perivascular and periadnexal lymphocytic infiltrate is observed, along with evidence of follicular plugging.