SKIN

BRIEF ARTICLE

# Coexistent of Morphea and DLE in a Patient With Beta Thalassemia Leading to a Diagnosis of Systemic Lupus Erythematous

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## ABSTRACT

Patients with autoimmune disorders are predisposed to developing a second autoimmune condition. This can be applied to cutaneous conditions as well. Morphea or localized scleroderma is an autoimmune inflammatory and fibrosing skin disorder due to increased collagen deposition.[1,2] Patients with morphea are four times likelier of having a concomitant autoimmune disease.[1] There have been cases of systemic lupus erythematosus (SLE) with morphea, but the co-occurrence of discoid lupus erythematosus (DLE) with morphea has been rarely reported, and never reported in patient with Beta-thalamessmia.[1,2,7] Morphea and discoid lupus erythematosus can be found within one lesion or as separate diagnoses. In this case, we describe a patient with morphea and discoid lupus erythematous in the setting of beta thalassemia leading to diagnosis of SLE.

## INTRODUCTION

An 18-year-old female with beta thalassemia presented with a six-month history of rash. Physical exam showed two depigmented patches with surrounding hypopigmentation on the left lower back (Figure 1), and indurated, annular pink and brown plaques on the scalp (Figure 2A) and left temple (Figure 2B).

## CASE REPORT

Two biopsies were performed of the clinically distinct lesions on the scalp and left lower back. Histopathology of the scalp demonstrated irregular epithelial acanthosis, perifollicular and band-like lymphocytic infiltrate, thickened basement membrane

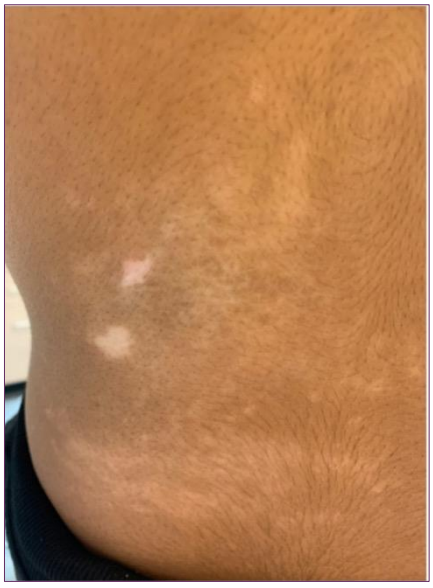

**Figure 1.** Depigmented patches with surrounding hypopigmentation on the left lower back with histology consistent with guttate morphea.

zone, slight mucin which was consistent with the diagnosis of discoid lupus erythematous


**Figure 2.** Indurated light brown plaques **A)** with localized alopecia of the scalp and **B)** left temple

lupus (Figure 3A). The left lower back consisted of sclerotic collagen bundles within the dermis with perivascular interstitial mononuclear cell infiltrate that was consistent with morphea (Figure 3B).

Labs were obtained to evaluate for SLE, which were significant for positive ANA, anti-Smith antibody, and anti-RNP antibody. She was subsequently diagnosed with SLE by Rheumatology following the American College of Rheumatology (ACR) with discoid lupus findings (+4 points) and SLE specific antibodies (+6), with a total score of 10, fulfilling the criteria for SLE. She was subsequently started on hydroxychloroquine 5 mg/kg and topical augmented betamethasone dipropionate ointment twice daily and educated on sun protective measures. At her three month follow up, she reported no new lesions or progression of her current lesions.

## DISCUSSION

Overlap syndromes are common in connective tissue diseases.[4] There have been few cases of morphea and DLE presenting within the same site. Mir et al. presented a case of 20-year-old woman with a spreading hypopigmented plaque on her right arm with limited range of motion with biopsy confirming DLE and morphea.[5] Pascucci et al. reported a case of a 60-year-old man with a progressive pruritic rash on the upper back for 30 years with biopsy revealing evidence of coexisting DLE and morphea.[4]

Our patient had separate lesions of DLE and morphea, with no overlap. To this date, only one other case of DLE and morphea in two different sites has been reported. In that case, a 46-year-old woman with history of depression and lithium-induced hypothyroidism was diagnosed with DLE for her facial lesions and linear morphea for right axilla.[3]

Furthermore, there have not been any reported cases of DLE and morphea in patients with beta thalassemia to our knowledge. There have been reports of patients with beta thalassemia with diagnosed SLE. Castellino et al. published a case series of 177 patients with SLE, and 17 patients had beta thalassemia. Their study demonstrated that the prevalence of SLE in patients with beta thalassemia was far less when compared to the general population. However, when SLE and beta thalassemia coexist, SLE seems to be more severe.[6,7] The association of SLE and beta thalassemia is not fully understood but theories include that the increased need for blood transfusions in beta-thalassemia patients can lead to formation of alloantibodies which can place patients at risk for infections and collagen vascular disorders.[8] Whether or not having beta-thalassemia increases the risk of autoimmune skin disorders remain unclear.

## CONCLUSION

In conclusion, we report the first case of DLE and morphea appearing simultaneously

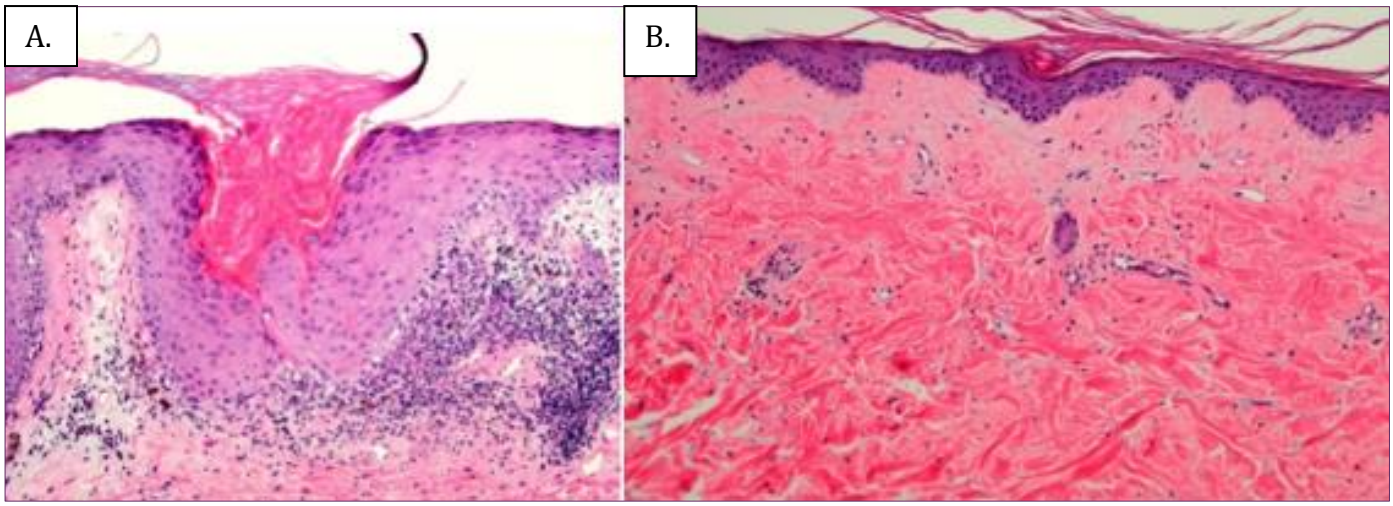


**Figure 3.** H&E of scalp with perifollicular, band-like lymphocytic interface with vacuolar changes with mucin and follicular plugging (A) and superficial perivascular lymphocytic (B) consistent with the diagnosis of cutaneous lupus.

in a patient with beta-thalassemia leading to a diagnosis of SLE. An association with SLE with beta thalassemia has been studied, however the extent with autoimmune skin conditions remains unknown.

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