Systemic lupus erythematosus overlap with morphea in a 26-year-old female: a case report

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ABSTRACT

Background: The coexistence of morphea and systemic lupus erythematosus (SLE) is rare and represents an important overlap of autoimmune disorders. This dual diagnosis highlights the complexity of immune dysregulation and poses challenges in clinical evaluation and management.

Case Presentation: A 26-year-old female presented with an 11-month history of progressive, recurrent erythematous plaques on her arms, back, and knees, accompanied by fatigue and polyarthralgia. She developed generalized swelling involving the periorbital regions and lower extremities, worsening in the morning. Physical examination revealed non-tender, well-defined macules and plaques, some with central hyperpigmentation and others with peripheral hyperpigmentation and central hypopigmentation, along with generalized swelling. Laboratory tests showed pancytopenia, significant proteinuria, hematuria, elevated creatinine and blood urea nitrogen, raised inflammatory markers, liver dysfunction, positive rheumatoid factor, and positive ANA and anti-double-stranded DNA antibodies. Skin biopsy demonstrated hyperkeratosis, epidermal atrophy, dermal collagenization, and periadnexal lymphocytic infiltrates.

Conclusion: This case underscores the rare coexistence of morphea and SLE in a young female, illustrating the complexity of autoimmune overlap syndromes. Early recognition through clinical, laboratory, and histopathological evaluation is crucial for accurate diagnosis and effective multidisciplinary management to improve patient outcomes.

Keywords: Systemic lupus erythematosus, Morphea, coexistence, case report, overlap.

Type of Article: CASE REPORT **Specialty:** Rheumatology

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Background

Systemic lupus erythematosus (SLE) is a chronic multisystem autoimmune disorder characterized by immune complex deposition resulting in widespread tissue damage. Morphea, also known as localized scleroderma, is a fibrosing disorder of the skin and subcutaneous tissues. Unlike systemic sclerosis, morphea does not present with sclerodactyly, Raynaud's phenomenon, and other cutaneous manifestations of systemic sclerosis [1]. While the exact etiology is still unknown, potential triggers include trauma, toxins, and some medications.

The coexistence of morphea and systemic lupus erythematosus represents a rare overlap of autoimmune disorders, characterized by distinct cutaneous and systemic manifestations. This dual diagnosis presented significant clinical challenges due to the complex immune dysregulation underlying both conditions.

Case Presentation

A 26-year-old female presented with an 11-month history of progressive, recurrent erythematous plaques initially appearing as a solitary raised lesion on her left upper arm, which later spread to her right arm, back, and knees. The rashes resolved with discoloration but recurred without pain, itching, fever, blistering, or ulceration. She also reported a 2-day history of generalized swelling, mainly affecting the periorbital regions and lower extremities, with swelling peaking in the morning and partially subsiding by evening. She denied a history of trauma, food, or drug allergies and has no history of any chronic illness. Physical examination revealed hard, non-tender reddish-brown patches on the bilateral malar regions sparing the nasal bridge, along with multiple well-demarcated, round to oval plaques and papules with central hypopigmentation and peripheral hyperpigmentation

symmetrically distributed on the back and upper extremities. (Figures 1 and 2).

Laboratory investigations revealed pancytopenia, significant proteinuria of 2089.64 mg/24 hours, hematuria, and elevated serum creatinine (155.4 µmol/l) and blood urea nitrogen (9.35 mmol/l). Inflammatory markers showed an elevated ESR (20.17 mm/hour) and CRP (16.4 mg/l), while CRP II was normal. Liver function tests indicated raised AST, ALT, and bilirubin levels with hypoalbuminemia (2.0 g/dl). Electrolyte imbalances included severe hypervolemic hyponatremia (123 mmol/l), and hypocalcemia (1.75 mmol/l). Thyroid function, lipid profile, coagulation, and Coombs test were normal or negative. Rheumatoid factor was positive. Immunological assays were positive for ANA (205.84 AU/ml) and anti-dsDNA antibodies (800 IU/ml).

Imaging studies, including chest X-rays, ECG, echocardiography, and renal Doppler ultrasound, showed no abnormalities. Skin biopsy histopathology revealed hyperkeratosis, epidermal atrophy, extensive dermal collagenization, and periadnexal lymphocytic infiltrates (Figs. 3 and 4).

Following consultation with the Internal Medicine Departments of Kairuki Hospital and one of the tertiary hospitals in Tanzania, initially, the patient was treated with clobetasol propionate ointment [9] to reduce inflammation and slow morphea progression. After being diagnosed with SLE, the patient's treatment was tailored to include methylprednisolone, cyclophosphamide [7,8] and hydroxychloroquine [7,8,10] for systemic lupus erythematosus. The patient was supplemented with calcium and vitamin D to correct the hypocalcemic state. The existing hypervolemic hyponatremia was corrected appropriately through the use of diuretics, salt restriction, and fluid restriction.

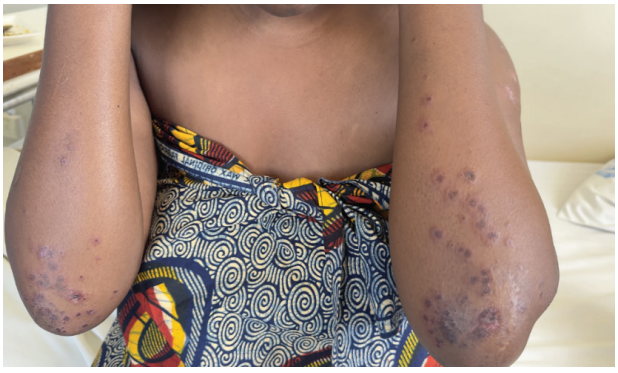

Figure 1. *Multiple, hyperpigmented, well-demarcated, roundish macules and papules on the extensor surface of the upper extremities.*

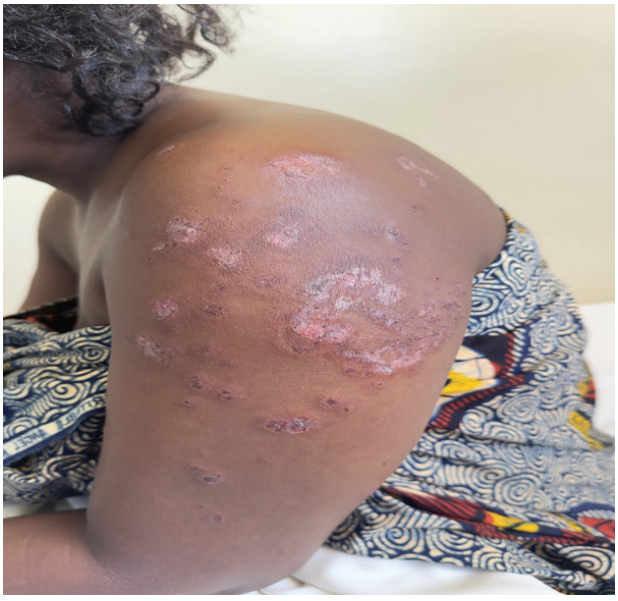

Figure 2. *Multiple, well-demarcated, roundish macules and papules on the upper limb with central hypopigmentation and peripheral hyperpigmentation.*

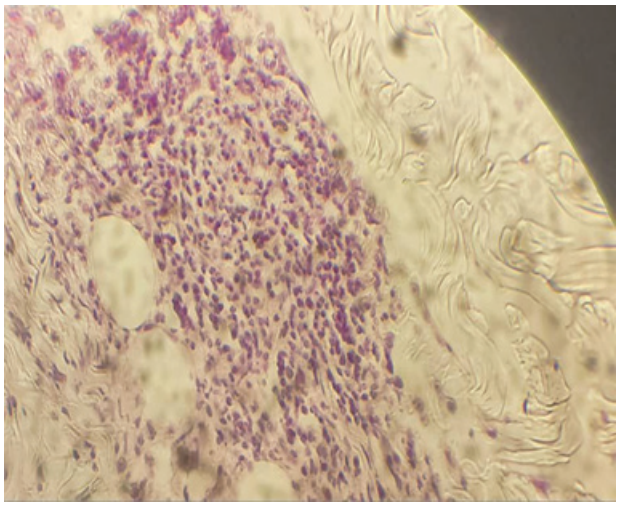

Figure 3. *Prominent lymphoplasmacytic infiltrate in the dermis typical of the inflammatory stage of morphea.*

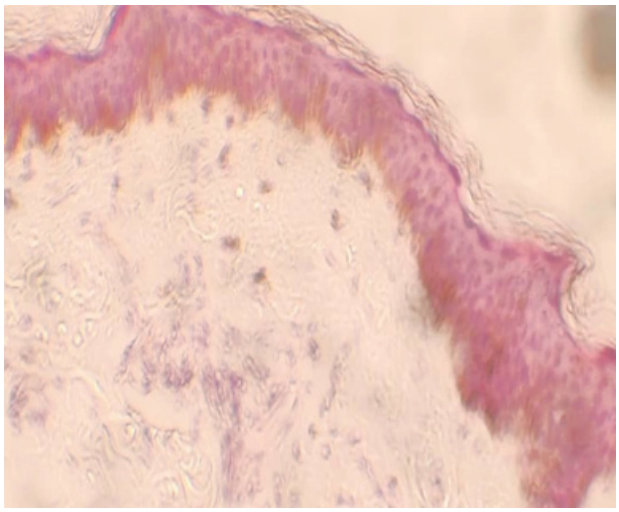

Figure 4. *Dense dermal collagen with sclerosis, loss of adnexal structures, and sparse lymphoplasmacytic infiltrate consistent with morphea.*

The patient's response to this immunosuppressive regimen has been positive, with reduced systemic inflammation, control of flares, and preservation of kidney function. Systemic steroids also helped manage widespread or active morphea by rapidly reducing inflammation, supplemented by other immunomodulatory agents to maintain remission.

Discussion

This case demonstrates a rare overlap syndrome involving SLE and morphea, a localized scleroderma variant. The patient's clinical presentation with progressive erythematous plaques and papules exhibiting central hypopigmentation and peripheral hyperpigmentation, along with histopathological findings of epidermal atrophy, hyperkeratosis, and dermal collagenization with lymphocytic infiltrates, is characteristic of morphea [2]. Concurrently, systemic features including pancytopenia, proteinuria, hematuria, elevated renal function markers, positive ANA and anti-dsDNA antibodies, and fulfillment of the American College of Rheumatology criteria confirm the diagnosis of SLE [3].

Although uncommon, the coexistence of these conditions highlights the complex autoimmune mechanisms affecting both systemic and localized tissues [4-6]. While SLE typically involves multisystem inflammation, including renal and hematologic manifestations, morphea primarily affects the skin with localized fibrosis [8]. The overlap in this patient underscores the importance of thorough clinical and immunological evaluation in patients presenting with atypical or mixed dermatologic and systemic symptoms and also highlights the importance of skin biopsy to make a correct diagnosis [4,5]. Management requires a multidisciplinary approach targeting both the systemic autoimmune activity of SLE and the localized skin fibrosis of morphea.

Conclusion

Coexisting systemic lupus erythematosus and morphea highlight the diagnostic challenges posed by overlapping autoimmune conditions. Early identification of such overlap syndromes facilitates timely intervention, improving patient outcomes and preventing irreversible organ damage. Continued awareness and reporting of similar cases will enhance understanding of the pathogenesis and necessitate multidisciplinary management to address both cutaneous and systemic manifestations. This case underscores the importance of considering overlapping autoimmune syndromes.

What's new?

This case highlights the rarity of the overlap of autoimmune disorders and how their co-occurrence poses diagnostic and therapeutic challenges. It emphasizes the need for careful multidisciplinary management to achieve effective treatment. The case underlines the in-depth understanding required to manage such complex autoimmune overlap conditions.

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Conflict of interest

The authors declare that there is no conflict of interest regarding the publication of this article.

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Consent for publication

Written informed consent was obtained from the patient.

Ethical approval

Ethical approval is not required at our institution to publish an anonymous case report.

Take home message

The coexistence of morphea and systemic lupus erythematosus (SLE) represents a rare but important overlap of autoimmune disorders that physicians should be aware of to ensure effective management and improve patient outcome.

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Summary of the case

1	Patient	26 years, female
2	Final diagnosis	Systemic lupus erythematosus and morphea
3	Symptoms	Skin lesions, fatigue, and polyarthlagia
4	Clinical investigations	Laboratory tests, Immunological assays, radiographic evaluations and histopathological examination
5	Medications	Cyclophosphamide, methylprednisolone, clobetasol ointment, hydroxychloroquine
6	Clinical procedure	None
7	Specialty	Rheumatology