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CASE REPORT

Granulomatous slack skin syndrome coexisting with hypopigmented mycosis fungoides: a rare case report

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ABSTRACT

Background: Granulomatous slack skin syndrome (GSSS) is a rare variant of mycosis fungoides (MF), a form of cutaneous T-cell lymphoma, characterized by lax, pendulous skin folds predominantly affecting flexural areas. Hypopigmented mycosis fungoides (HMF) is another uncommon variant that presents with hypopigmented macules and patches, often in younger individuals with darker skin tones. The simultaneous occurrence of GSSS and HMF in a single patient is exceptionally rare, with only one prior case reported in the literature.

Case Presentation: We report the case of a 31-year-old woman presenting with clinical features consistent with both GSSS and HMF. The diagnosis was established based on clinical examination, histopathological findings, immunohistochemical analysis, and molecular studies.

Results and Conclusion: Clinical findings demonstrated overlapping features of GSSS and HMF, supported by histopathology and immunophenotyping consistent with MF despite negative T-cell receptor gene rearrangement. This case highlights the rare coexistence of these variants and underscores the importance of clinicopathological correlation and multiple biopsies in atypical presentations.

Keywords: Granulomatous slack skin syndrome, hypopigmented mycosis fungoides, cutaneous T-cell lymphoma, mycosis fungoides variants.

Introduction

Mycosis fungoides (MF), the most common subtype of cutaneous T-cell lymphoma, encompasses several rare variants. Granulomatous slack skin syndrome (GSSS) is a rare subtype characterized by progressive skin laxity, particularly in flexural areas such as the axillae and groin [1,2]. Histopathologically, it often demonstrates granulomatous lymphoid infiltrates with multinucleated giant cells and elastophagocytosis [3].

Hypopigmented mycosis fungoides (HMF) is another uncommon variant that typically affects younger individuals with darker skin phototypes and presents as hypopigmented macules or patches on sun-protected areas [4]. It often exhibits a CD8+ T-cell phenotype and may mimic benign dermatoses such as pityriasis alba or post-inflammatory hypopigmentation. Despite its atypical presentation, it generally carries a favorable prognosis [5,6].

Although both variants are well recognized, their coexistence in a single patient is exceedingly rare. To date, only one such case has been reported [7]. We describe an additional case demonstrating features of both variants, highlighting the diagnostic challenges and the importance of an integrated clinicopathological approach.

Case Report

A 31-year-old woman presented with a 6-year history of gradually progressive, asymptomatic hyperpigmented plaques involving intertriginous areas and proximal extremities. Over the past year, several lesions developed associated skin laxity, particularly over the axillae and

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upper arms. Simultaneously, she noted the appearance of new hypopigmented macules and patches on her limbs and back. She denied pruritus, pain, or systemic symptoms.

Physical examination revealed multiple hyperpigmented plaques with mild atrophy and skin laxity in the axillae and upper arms. Additionally, several ill-defined hypopigmented macules and patches with smooth surfaces and no scaling were observed on the upper and lower extremities and back. No palpable lymphadenopathy or organomegaly was detected. These findings are illustrated in Figure 1.

Routine laboratory investigations, including complete blood count, liver and renal function tests, and lactate dehydrogenase levels, were within normal limits.

Histopathological analysis of representative lesions revealed epidermal hyperplasia with focal parakeratosis and mild spongiosis, accompanied by lymphocytic exocytosis. The dermis showed edema, dilated superficial vessels, and a moderate perivascular lymphocytic infiltrate. No well-formed granulomas or overt cytologic atypia were identified. However, lymphocytes along the dermoepidermal junction exhibited mild nuclear atypia. Representative findings are shown in Figure 2.

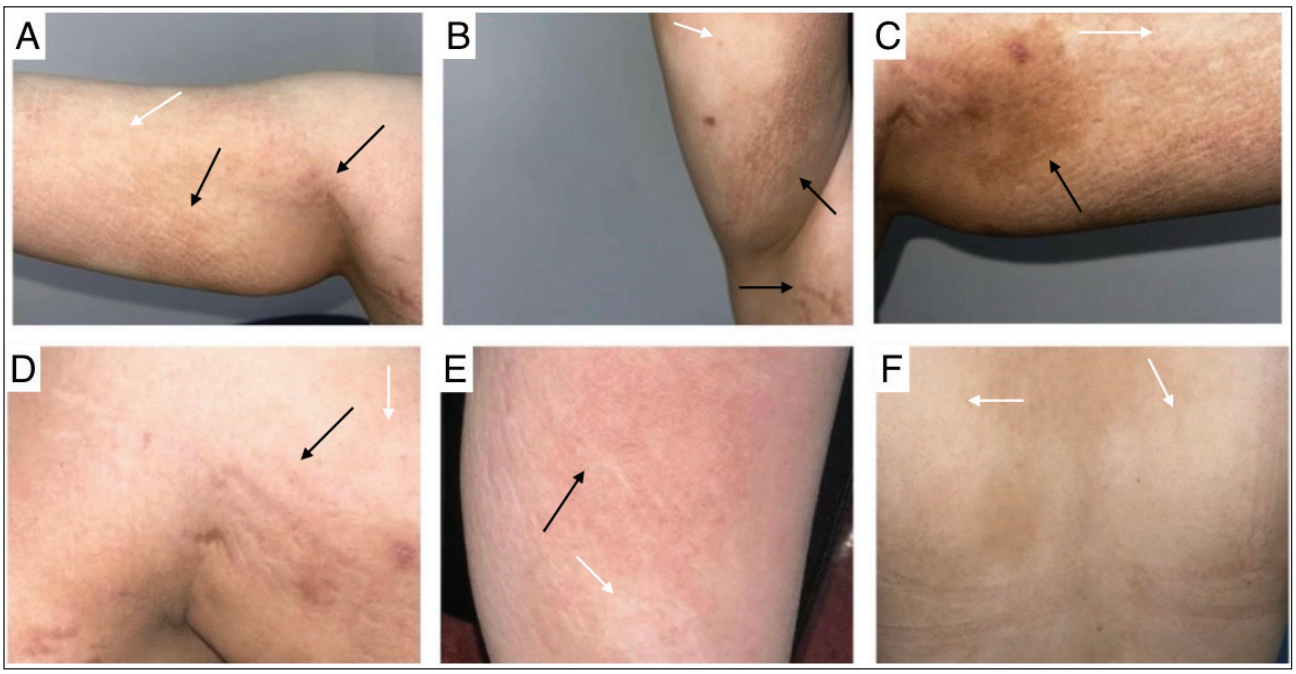

Figure 1. *Clinical presentation of overlapping variants of mycosis fungoides. (A–E) Black arrows indicate hyperpigmented plaques with mild atrophy and skin laxity involving the axillae, proximal upper arms, and legs, consistent with granulomatous slack skin syndrome. (A–F) White arrows indicate multiple ill-defined hypopigmented macules and patches on the extremities and back, representing hypopigmented mycosis fungoides.*

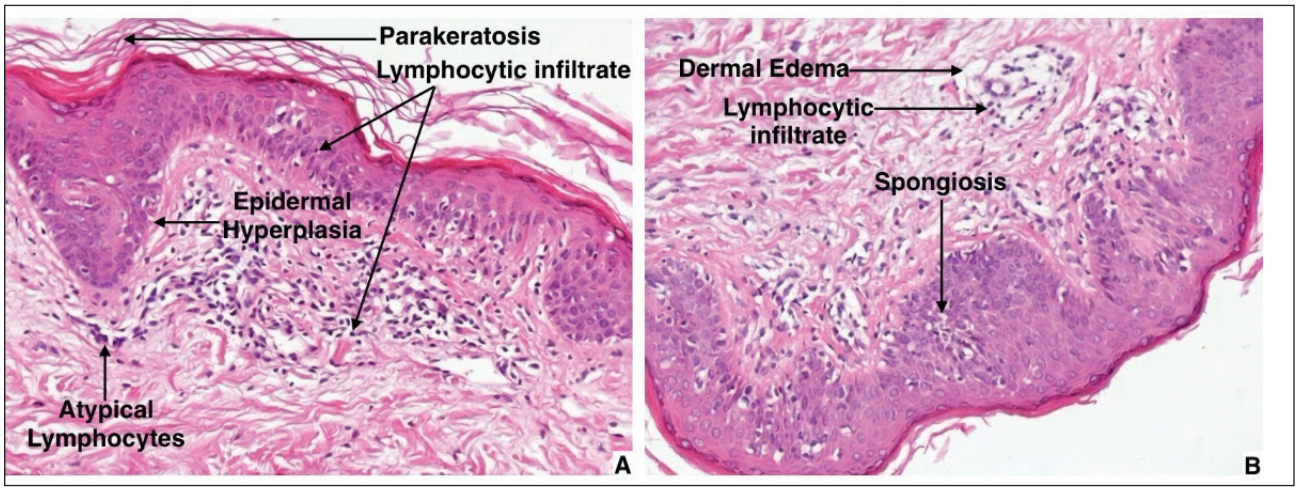

Figure 2. *Histopathological features of two lesional biopsies (hematoxylin and eosin staining, original magnification ×200). (A) Biopsy from a hypopigmented lesion showing epidermal hyperplasia with parakeratosis and lymphocytic infiltrate with epidermotropism. (B) Biopsy from a hyperpigmented lesion showing spongiosis, superficial dermal edema, and perivascular lymphocytic infiltrate.*

Immunohistochemical analysis demonstrated a predominance of CD3+ and CD4+ T-cells, with partial loss of CD7 expression and an elevated CD4:CD8 ratio. Special staining for fungal organisms was negative. T-cell receptor gene rearrangement testing did not demonstrate clonality; however, this does not exclude MF, particularly in early-stage disease, where clonal populations may fall below detection thresholds.

The coexistence of hyperpigmented plaques with skin laxity and newly developed hypopigmented lesions raised suspicion of overlapping variants. Clinical findings supported a diagnosis of GSSS in the flexural plaques and HMF in the hypopigmented lesions. Histopathology and immunophenotyping confirmed MF with CD4+ predominance in both presentations.

The patient was treated with topical betamethasone dipropionate and pimecrolimus 1% cream, in addition to narrowband ultraviolet B phototherapy three times weekly. She was referred for oncologic evaluation and staging, as well as for lymph node assessment. Follow-up was arranged to monitor treatment response.

Results and Conclusion

This case illustrates the rare coexistence of GSSS and HMF in a single patient, confirmed through integrated clinical, histopathological, and immunophenotypic evaluation. Clinically, the patient exhibited characteristic lax, hyperpigmented plaques in flexural areas consistent with GSSS, alongside widespread hypopigmented macules and patches suggestive of HMF.

Histopathological findings supported early-stage MF, demonstrating epidermotropism and superficial perivascular lymphocytic infiltrates, although classic granulomatous features were not prominent. Immunohistochemistry revealed CD4+ T-cell predominance with partial loss of CD7 expression. Despite negative T-cell receptor gene rearrangement, the diagnosis was supported by clinicopathological correlation.

This case is significant because of the rare coexistence of these variants and the atypical immunophenotype observed in the hypopigmented lesions. Compared with the previously reported case, these findings support the concept that MF variants may represent a clinicopathological spectrum rather than distinct entities.

Discussion

GSSS is a rare subtype of cutaneous T-cell lymphoma characterized by the gradual development of localized, erythematous, and lax skin folds, most commonly involving intertriginous areas. Histologically, it may show granulomatous infiltrates and loss of elastic fibers; however, these features may be subtle or absent, particularly in early disease [3,8].

HMFs, in contrast, typically presents with multiple hypopigmented macules and patches distributed over the trunk and extremities. It is more frequently observed in individuals with darker skin phototypes and generally follows an indolent course [4]. While most cases demonstrate a CD8+ T-cell phenotype, the present case showed CD4+ predominance, highlighting the variability of this variant [6]. A recently reported case also described a CD4-predominant phenotype, further supporting this variability [7].

Molecular testing for T-cell receptor gene rearrangement is useful for assessing clonality; however, it should not be used in isolation. A clonal result does not confirm malignancy, and a negative result does not exclude it. Findings must be interpreted in conjunction with clinical, histopathological, and immunophenotypic features [9]. In early-stage disease, the neoplastic T-cell population may be too small for detection, leading to false-negative results [10].

Only one prior case describing the coexistence of these two variants has been reported [7]. In that case, distinct biopsies demonstrated classic features of both entities. In contrast, the present case showed less pronounced histopathological features, without well-formed granulomas, suggesting that GSSS may present with subtle or evolving changes. Additionally, the broader distribution of hypopigmented lesions and absence of tumorous masses further highlight the variability of presentation.

These findings support the concept that MF variants may represent a clinicopathological spectrum rather than entirely distinct entities.

Learning Points

- GSSS and HMFs are rare variants that can coexist in a single patient.
- The absence of classic histopathological features does not exclude the diagnosis, particularly in early or evolving disease.
- HMFs may demonstrate CD4+ predominance, indicating variability beyond the traditionally described CD8+ phenotype.
- Negative T-cell receptor gene rearrangement does not rule out MFs and must be interpreted in the appropriate clinical and histopathological context.
- Multiple biopsies from clinically distinct lesions are essential to improve diagnostic accuracy.
- These variants likely represent a clinicopathological spectrum rather than distinct disease entities.

List of Abbreviations

CD	Cluster of differentiation
CTCL	Cutaneous T-cell lymphoma
GMS	Grocott methenamine silver
GSSS	Granulomatous slack skin syndrome
H&E	Hematoxylin and eosin
HMF	Hypopigmented mycosis fungoides
IHC	Immunohistochemistry
LDH	Lactate dehydrogenase
MF	Mycosis fungoides
TCR	T-cell receptor

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

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Consent to participate
Written informed consent was obtained from the patient.

Consent for publication
Example: Written informed consent was obtained from the patient.

Ethical approval
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Supplementary content (If any) is available online.

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