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Beyond The Itch: Unmasking A Rare Composite Lymphoproliferative Disorder With Coexistent Chronic Lymphocytic Leukemia And Folliculotropic Mycosis Fungoides

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Abstract
Background: Composite lymphoproliferative disorders, defined by the coexistence of two distinct lymphoid neoplasms of different cell lineages in the same patient, are exceptionally rare. The simultaneous occurrence of ***Chronic Lymphocytic Leukemia (CLL)****, a mature B-cell neoplasm, and* ***Folliculotropic Mycosis Fungoides (FMF)****, a variant of cutaneous T-cell lymphoma, has been infrequently reported. Their overlapping clinical manifestations, distinct biological behavior, and lack of standardized treatment strategies pose significant diagnostic and therapeutic challenges, making early recognition essential.*
Case: A male farmer presented with refractory generalized pruritus, erythematous plaques progressing to hyperpigmented scaly lesions. TLC was 45,900/µL with atypical lymphocytosis. Skin biopsy confirmed folliculotropic mycosis fungoides while peripheral blood flow cytometry demonstrated chronic lymphocytic leukemia. PET-CT showed metabolically quiescent nodal disease without visceral involvement. Conclusion: Multimodal evaluation established a rare composite lymphoproliferative disorder.
Keywords: *Chronic lymphocytic leukemia; Mycosis fungoides; Composite lymphoma; Cutaneous T-cell lymphoma; Flow cytometry.*

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I. Introduction

Composite lymphomas are uncommon neoplasms characterized by two distinct lymphoid malignancies. Concurrent chronic lymphocytic leukemia and folliculotropic mycosis fungoides is exceptionally rare and poses diagnostic challenges. We report this rare coexistence.

II. Case Report

A male farmer presented with a **6-month history of generalized pruritus** associated with erythematous, elevated skin lesions that progressively evolved into diffuse hyperpigmented scaly plaques. The pruritus was predominantly nocturnal, resulting in significant sleep disturbance. He had been treated empirically with oral antihistamines (montelukast and levocetirizine), oral fluconazole, topical clotrimazole cream, and liquid paraffin without symptomatic improvement. He had a past history of bronchial asthma for 10 years.

On dermatological examination, diffuse xerotic erythematous to hyperpigmented scaly plaques with multiple excoriations were noted over the trunk, extremities, and groin, suggestive of a chronic infiltrative dermatosis (Figure 1).

Initial laboratory investigations demonstrated **marked leukocytosis (total leukocyte count: 45,900/µL)** with **lymphocytic predominance and atypical lymphocytes** on peripheral smear, raising suspicion of an underlying lymphoproliferative disorder. Ultrasonography of the abdomen revealed no hepatosplenomegaly, while ultrasonography of the neck demonstrated only subcentimeter cervical lymph nodes.

To evaluate the persistent skin lesions, a punch biopsy was obtained from the back. Histopathological examination demonstrated features of **cutaneous T-cell lymphoma (mycosis fungoides spectrum) with focal folliculotropism** (Figure 2). Immunohistochemistry showed positivity for **CD3** with **predominant CD4 expression**, focal loss of **CD5**, preserved **CD7** expression, **CD30 negativity**, and a low proliferative index (**Ki-67 approximately 10%**). These findings were characteristic of **folliculotropic mycosis fungoides**, a recognized variant of cutaneous T-cell lymphoma.

In view of the marked lymphocytosis, peripheral blood immunophenotyping by flow cytometry was performed. A monoclonal B-cell population expressing **CD45 (bright), CD19, CD20 (dim heterogeneous), CD5 (dim heterogeneous), CD23 (moderate), CD200, CD43, FMC7 (dim)** with **lambda light-chain**

restriction was identified, while T-cell markers (**CD3, CD4, CD7, CD8**) and other B-cell markers (**CD10, CD38, CD79b, CD103, CD25, CD11c, CD123**) were negative. This immunophenotypic profile was diagnostic of **Chronic Lymphocytic Leukemia/Small Lymphocytic Lymphoma (CLL/SLL)**.

Whole-body **18F-FDG PET-CT** was performed for staging. The scan demonstrated **metabolically quiescent cervical and mediastinal lymphadenopathy** with **low-grade FDG-avid bilateral axillary and subpectoral lymph nodes** (SUVmax 1.4), while no hepatosplenomegaly or visceral organ involvement was identified. The low metabolic activity of the lymph nodes was consistent with the indolent biological behavior of CLL. Mild FDG uptake in the salivary glands and palatine tonsils was considered inflammatory, whereas pulmonary findings were attributable to chronic obstructive airway disease.

The diagnosis of **composite lymphoproliferative disorder** was established based on the demonstration of **two independent lymphoid neoplasms**. The skin biopsy with immunohistochemistry confirmed **folliculotropic mycosis fungoides**, representing a mature **T-cell lymphoma**, while peripheral blood flow cytometry demonstrated a distinct monoclonal **B-cell population** diagnostic of **Chronic Lymphocytic Leukemia**. The presence of lymphocytosis with cervical and axillary lymphadenopathy in the absence of anemia, thrombocytopenia, hepatomegaly, or splenomegaly fulfilled the criteria for **Rai Stage I CLL**. Collectively, the clinical, histopathological, immunophenotypic, and imaging findings established the final diagnosis of **composite lymphoproliferative disorder comprising Chronic Lymphocytic Leukemia (Rai Stage I) and folliculotropic Mycosis Fungoides**.

Considering the indolent nature of both malignancies, the patient received **PUVA phototherapy combined with weekly oral methotrexate** (7.5mg) for mycosis fungoides, while CLL was managed with active surveillance. He has shown a favorable clinical response with significant improvement in cutaneous manifestations and remains clinically stable on follow-up.

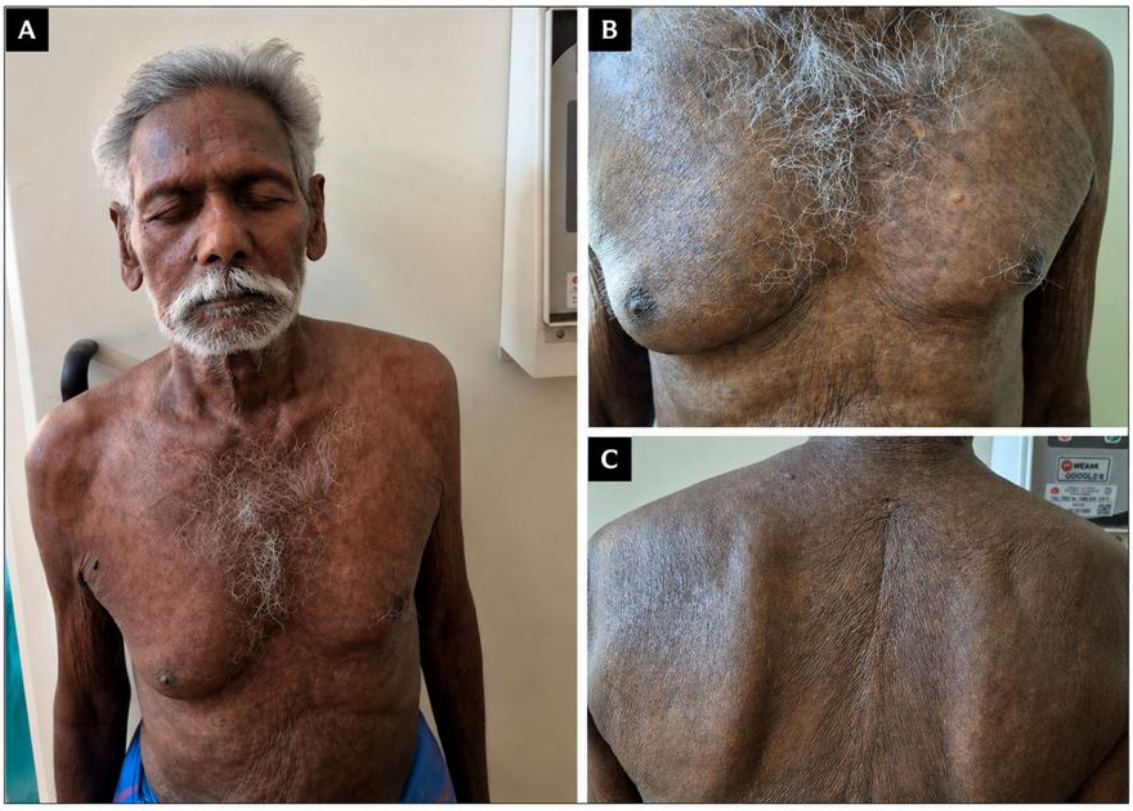

Figure 1: Clinical photographs showing diffuse hyperpigmented macules with associated follicular prominence and alopecia.
(A) Frontal view showing hyperpigmented macules over the face, neck, and upper chest with sparse gray-white hair.
(B) Anterior chest showing diffuse hyperpigmented macules with prominent hair follicles and sparse hair.
(C) Back showing similar hyperpigmented macules with follicular prominence and alopecia.

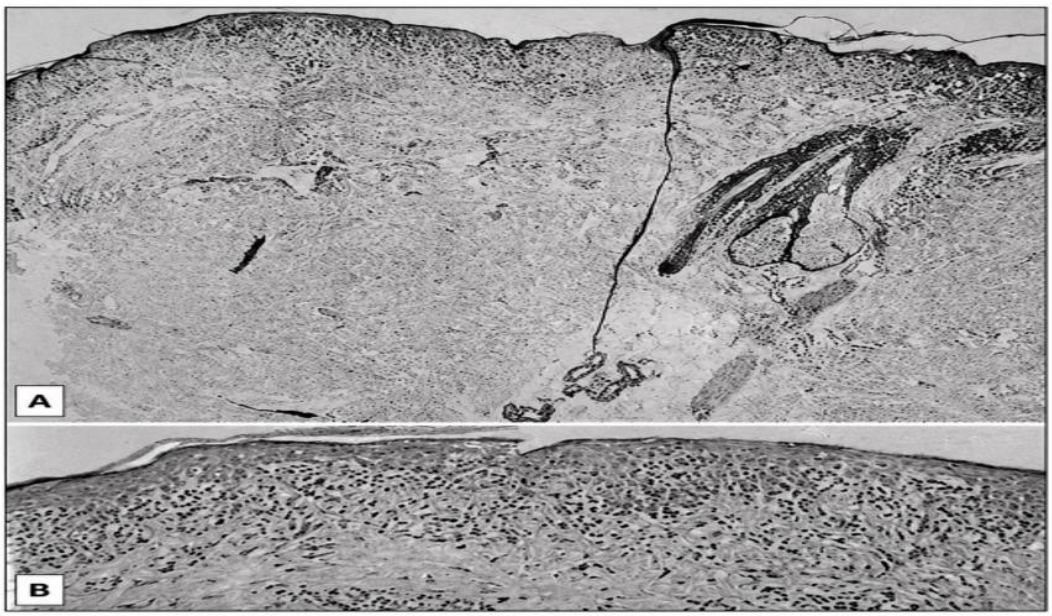

Figure 2: Histopathology (H&E).
(A) Low power view showing epidermal infiltration by atypical lymphocytes with folliculotropism (follicular epithelium surrounded by lymphocytes).
(B) High power view showing epidermotropism of atypical lymphocytes with minimal spongiosis.

III. Discussion

The coexistence of **chronic lymphocytic leukemia (CLL)** and **mycosis fungoides (MF)** is exceptionally rare and represents the occurrence of two distinct mature lymphoid neoplasms arising from **B-cell and T-cell lineages**, respectively. Although patients with CLL have an increased risk of secondary malignancies due to immune dysregulation, the development of cutaneous T-cell lymphoma remains distinctly uncommon. The pathogenesis is not fully understood but has been attributed to impaired immune surveillance, chronic antigenic stimulation, genetic susceptibility, or treatment-related immune modulation.

Our patient initially presented with **persistent generalized pruritus and diffuse hyperpigmented scaly plaques**, a presentation easily mistaken for common dermatological disorders such as eczema, xerosis, or superficial fungal infections. Failure to respond to conventional therapy, together with progressive lymphocytosis, prompted further evaluation. Skin biopsy demonstrated epidermotropic atypical T-cell infiltrates consistent with **mycosis fungoides**, while peripheral blood flow cytometry independently confirmed **CLL**, establishing the diagnosis of two synchronous lymphoid neoplasms rather than cutaneous involvement by CLL.

An important diagnostic challenge was differentiating MF from **leukemia cutis**, which may also present with cutaneous lesions in patients with CLL. In our case, the characteristic histopathological findings and immunophenotype supported a diagnosis of primary cutaneous T-cell lymphoma rather than secondary skin infiltration by CLL. Furthermore, PET-CT showed only low-grade FDG uptake in lymph nodes, consistent with indolent CLL without evidence of Richter transformation or aggressive systemic disease.

This case highlights the importance of a **multidisciplinary clinicopathological approach**, integrating dermatologic evaluation, histopathology, immunohistochemistry, flow cytometry, and functional imaging to accurately identify coexisting lymphoid malignancies. Recognition of this rare association has significant therapeutic implications because management should be directed at the clinically dominant disease rather than assuming a single pathological process.

IV. Conclusion

This case illustrates the rare coexistence of **mycosis fungoides and chronic lymphocytic leukemia**, presenting initially as persistent refractory pruritus. Early skin biopsy, comprehensive immunophenotyping, and flow cytometric analysis were crucial in establishing two independent lymphoid neoplasms. Clinicians should maintain a high index of suspicion for secondary hematologic malignancies in patients with atypical or treatment-resistant dermatoses accompanied by unexplained lymphocytosis. Prompt diagnosis enables appropriate risk stratification and individualized management.

Learning Points

• Persistent pruritic dermatoses warrant biopsy.
• Unexplained lymphocytosis requires flow cytometry.
• Consider composite B-cell and T-cell lymphomas.
• Multidisciplinary evaluation improves diagnosis.

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