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CASE REPORT

Beyond Eczema and Lupus: Sézary Syndrome with Chronic Cutaneous Presentation Mimicking Common Inflammatory Dermatoses

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Abstract

Sézary syndrome is an uncommon and aggressive leukemic variant of cutaneous T-cell lymphoma (CTCL), often misdiagnosed in its early stages due to its clinical resemblance to chronic inflammatory dermatoses such as eczema or cutaneous lupus erythematosus. We report the case of a 67-year-old woman who presented with chronic pruritic erythematous plaques initially diagnosed and treated as chronic eczema and later as systemic lupus erythematosus (SLE), delaying the correct diagnosis. The eventual identification of Sézary cells on peripheral smear and confirmatory immunophenotyping established the diagnosis of Sézary syndrome, underscoring the need for a high index of suspicion and a multidisciplinary diagnostic approach when evaluating chronic, treatment-refractory dermatoses.

Introduction

In recent years, there has been a growing interest in the relationship between hematologic diseases and autoimmune disorders. The association between hematologic malignancies and autoimmune conditions has been increasingly recognized, with documented links between antinuclear antibodies (ANA) and cancer, challenging the conventional belief that these antibodies are exclusive to autoimmune contexts.

It has been determined that the association between cancer and autoimmune disorders is bidirectional; in the case of systemic lupus erythematosus (SLE), age and disease duration have been found to be inversely associated with the overall risk of malignant neoplasms.

The presence of serum ANA reflects an immune response directed against self-nuclear antigens and, although commonly interpreted as a marker of autoimmunity, its detection through indirect immunofluorescence on HEp-2 cells has also been associated with neoplastic phenomena. Certain ANA patterns, such as the homogeneous or speckled types, are linked to a lower prevalence of cancer, whereas the nucleolar pattern has been associated with a significantly higher risk of malignancy.[1]

Among the group of non-Hodgkin lymphomas, cutaneous T-cell lymphomas (CTCL) represent a rare lymphoproliferative neoplasm that predominantly affects males, with a mean age between 50 and 60 years. It primarily involves the skin, blood, and lymph nodes. Two major subtypes are recognized: mycosis fungoides and Sézary syndrome, the latter being more aggressive and characterized by marked cutaneous involvement and a significant leukemic component.[2]

Several mechanisms have been proposed to explain this relationship. Among them is the possibility that certain neoplasms act as triggers for paraneoplastic autoimmune manifestations, as well as the fact that immunosuppressive therapies used in autoimmune diseases may contribute to oncogenesis. These observations reinforce the need for a comprehensive clinical evaluation that considers both immunologic and oncologic factors in patients with positive ANA, particularly when they present atypical clinical manifestations.[3]

Case Description

A 67-year-old female patient, with no significant past medical history, presented with a progressive 10-month history of pruritic skin lesions localized to the trunk and extremities, initially interpreted in primary care as chronic eczema. She was treated with topical corticosteroids and emollients without significant improvement.

Subsequently, the patient experienced progression of the lesions, which evolved into erythematous-scaly plaques with skin thickening, accompanied by symmetric peripheral arthralgias and chronic fatigue. ANA testing revealed a titer of 1:160, with positive antibodies against extractable nuclear antigens (ENA). Given these findings, she was referred to rheumatology, where a presumptive diagnosis of systemic lupus erythematosus (SLE) with cutaneous involvement was made, and oral hydroxychloroquine 200 mg every 12 hours was initiated.

During the following six months, the patient reported partial relief of arthralgias but progressive worsening of the skin lesions, which became "cardboard-like," erythematous, thickened, and "woody" to the touch, with intense pruritus and desquamation. A skin biopsy was performed, revealing findings consistent with discoid lupus erythematosus, and methylprednisolone 4 mg daily plus methotrexate 15 mg weekly were added to the treatment regimen.

During this time, the patient exhibited a progressive and sustained increase in leukocyte count with marked lymphocytosis (>25,000/mm³), without hematologic evaluation or complementary phenotypic studies. The diagnostic alert was finally raised after detecting hyperleukocytosis exceeding 50,000/mm³, hepatosplenomegaly, and a firm, fixed, non-tender axillary lymph node on the left side.

She was referred to hematology, where peripheral blood flow cytometry revealed a mature T-cell population (CD3+, CD4+, CD8−) with partial loss of CD7, consistent with mature T-cell neoplasia. Peripheral blood smear examination demonstrated cerebriform lymphocytes (Sézary cells), a key finding for establishing the diagnosis of Sézary syndrome.

The disease was classified as an aggressive variant of cutaneous T-cell lymphoma (CTCL), leukemic type, stage IVB, with diffuse hematologic and cutaneous involvement. Induction chemotherapy with CHOEP (cyclophosphamide, doxorubicin, vincristine, etoposide, and prednisone) was initiated. After the first cycle, the patient exhibited partial remission of cutaneous lesions, symptomatic improvement, and a decrease in leukocyte count.

Discussion

Diagnostic Challenges

The evaluation of chronic cutaneous lesions represents a significant clinical challenge, particularly when these mimic common inflammatory dermatoses such as eczema or cutaneous lupus erythematosus. The present case illustrates the diagnostic complexity of Sézary syndrome, a leukemic variant of cutaneous T-cell lymphoma (CTCL), clinically

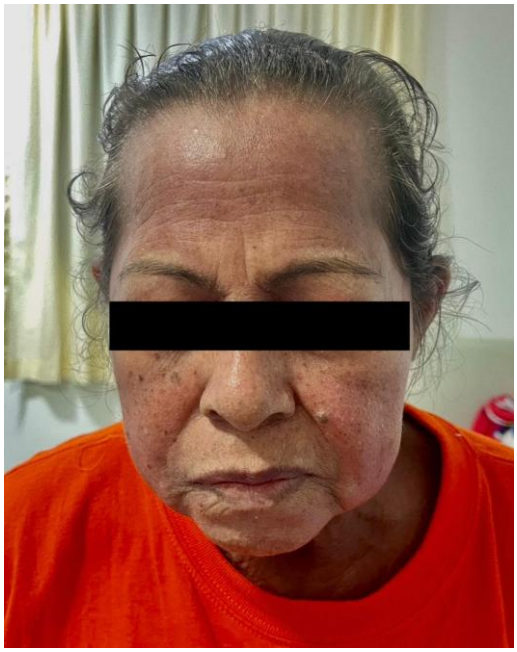

Figure 1. Erythematous, thickened skin lesions with a stiff, parchment-like appearance predominantly affecting the forehead, cheeks, chin, and upper neck. The skin displays a faint reddish tone, fine scaling, and loss of normal texture without shine.

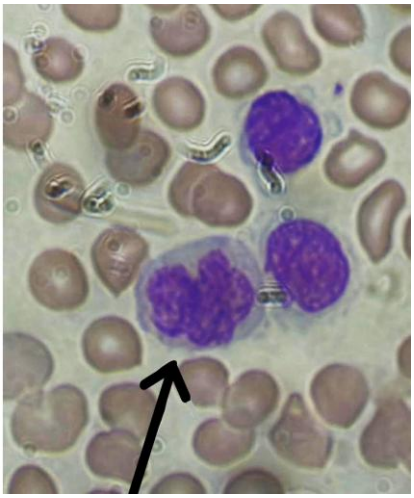

Figure 2. Wright–Giemsa–stained peripheral blood smear showing an atypical lymphocyte with a hyperchromatic, irregularly contoured, deeply convoluted nucleus with multiple folds and grooves resembling the surface of a brain, morphologically compatible with a Sézary cell.

characterized by the triad of erythroderma, generalized lymphadenopathy, and lymphocytosis with Sézary cells in peripheral blood. This rare and aggressive entity often goes unrecognized in its early stages, leading to diagnostic delays and inappropriate immunosuppressive treatments.

Misleading Autoimmune Markers

Given the presence of positive ANA, articular manifestations, and a skin biopsy showing focal moderate hyperkeratosis, hyperparakeratosis, mild epidermal atrophy with basal hyperpigmentation and focal vacuolization, papillary and reticular dermal edema without mucinosis, and a periadnexal lymphocytic inflammatory infiltrate with scarce melanophages, the patient was initially approached by rheumatology with a presumptive diagnosis of systemic lupus erythematosus (SLE).

However, it is crucial to remember that ANA are not exclusive to autoimmune diseases; their presence can also be associated with paraneoplastic phenomena or even underlying lymphoproliferative processes, as documented in recent literatura.[4,5] This immunologic overlap underscores the need for critical interpretation of serologic findings within the patient's clinical context.

Histopathologic Clues

Although skin biopsy is fundamental in the diagnostic process, its interpretation must be approached with caution. The histopathology of early CTCL lesions may mimic inflammatory patterns such as discoid lupus, lichen planus, or even psoriasis, due to dense lymphocytic infiltrates that may not exhibit overt atypia. Lymphoid cellularity can overlap across different entities, limiting diagnostic specificity in the absence of immunophenotypic or molecular studies.

In this sense, the present case highlights the relevance of integrating biopsy results with complementary hematologic studies, including flow cytometry and peripheral smear examination, particularly in the presence of persistent lymphocytosis.[6]

Importance of Multidisciplinary Evaluation

The complete blood count, though basic, proved critical in this case. Progressive leukocytosis with lymphoid predominance, initially deemed non-relevant, became a key warning marker that should prompt hematologic evaluation in patients with atypical, treatment-refractory dermatoses. Lymphocytosis greater than 50,000/mm³, in association with hepatosplenomegaly and firm lymphadenopathy, constitutes a highly suggestive pattern of systemic hematologic involvement, as confirmed here by the identification of cerebriform lymphocytes characteristic of Sézary síndrome.[6,7,8]

The initially low index of suspicion can be partly explained by the low prevalence of this entity, which represents less than 5% of cutaneous lymphomas.[9] Nevertheless, Sézary syndrome should remain in the differential diagnosis when evaluating elderly patients with progressive, treatment-refractory skin lesions and concurrent hematologic abnormalities.

This case reinforces that systemic diseases with cutaneous manifestations require an integrative approach combining clinical expertise, hematologic surveillance, immunologic evaluation, and critical interpretation of histopathologic studies. Only through a multidisciplinary perspective can timely diagnosis be achieved and therapeutic interventions that mask or worsen underlying neoplastic diseases be avoided.

This case is notable for its exceptionally deceptive presentation—combining positive ANA, lupus-like histopathology, and chronic eczema-like lesions—which led to an unusual and prolonged misdiagnosis before revealing Sézary syndrome, a scenario scarcely documented in the literature.

Conclusion

The evaluation of chronic cutaneous lesions presents significant diagnostic challenges, as they may mimic common inflammatory diseases such as systemic lupus erythematosus (SLE). This can lead to initial misdiagnoses and considerable delays in definitive diagnosis. For example, Majocchi's granuloma lesions were initially misdiagnosed as lupus-associated, delaying appropriate treatment and resulting in severe disseminated lesions.[11]

Sézary syndrome, a rare leukemic variant of cutaneous T-cell lymphoma (CTCL), accounts for approximately 3% of all primary cutaneous lymphomas.[11] Its diagnosis is often delayed due to nonspecific clinical presentation and resemblance to other inflammatory dermatoses. A retrospective study found that the average diagnostic delay was 4.2 years, being longer in cases without initial erythroderma.[7,12]

The integration of diagnostic tools such as skin biopsy, complete blood count, and immunophenotypic studies (e.g., flow cytometry) is essential for the timely identification of underlying hematologic diseases such as cutaneous lymphomas.[6,13]

Flow cytometry, in particular, has proven useful for assessing blood tumor burden, which is crucial for diagnosis, staging, and management of CTCL.[6,8]

Since CTCL diagnosis is often delayed due to its indolent course and nonspecific symptoms, maintaining a high index of clinical suspicion in atypical or refractory chronic dermatoses is essential. Multidisciplinary collaboration and comprehensive evaluation can help avoid diagnostic and therapeutic delays in rare entities such as Sézary syndrome.[14,15,16,17]

Disclaimer (Artificial Intelligence)

None.

Consent

It is not applicable.

Ethical Approval

None.

Competing Interests

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