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Initial Diagnosis of Psoriasis and Final Diagnosis of Cutaneous T-Cell Lymphoma/Sézary Syndrome

Estefania G. Piedrahita[1], Maria A. G. Abad[1], Ángela M. L. García[1] and José D. S. Ramírez[2]

[1]Department of Dermatology, CES University, Medellín, Colombia
[2]Department of Oncology, Clínica Vida, Medellín, Colombia

Abstract

Cutaneous T-cell lymphomas are part of non-Hodgkin lymphomas that mainly affects the skin but can compromise blood, lymph nodes and other internal organs in patients with advanced disease. There are many types of cutaneous T-cell lymphoma. The two main subtypes are mycosis fungoides (MF) and Sézary syndrome (SS). The diagnosis of the latter is a challenge due to the clinical and some histological findings, which makes them similar to benign dermatoses. The clinical case of a patient with an initial diagnosis of erythroderma associated with psoriasis with joint involvement is presented, then the etiology is re-evaluated and an extra cutaneous nodal T lymphoma with bone marrow involvement (Sézary syndrome) is confirmed.

Keywords: Psoriasis • Psoriatic arthritis • Erythroderma • Lymphoma • Mycosis fungoides

Introduction

Mycosis fungoides and Sézary syndrome are the most common forms of cutaneous T-cell lymphoma. In early stages, the diagnostic approach is challenging, given the clinical similarity between benign dermatoses, such as psoriasis [1,2]. A patient who had an initial diagnosis of psoriasis is presented with poor therapeutic response, and a final diagnosis of cutaneous extranodal T lymphoma and Sézary syndrome is made.

Clinical Case Presentation

A 34-year-old male with 3-year erythroderma, biopsy confirmed the diagnosis of psoriasis. He was treated with phototherapy without improvement. Later, he started with arthritis in the knees, heel pain and dactylitis of the right hallux, which was determined as psoriatic arthritis. He received treatment with sulfasalazine and methotrexate, with improvement of arthritis, but persistence of skin lesions (Figure 1).

A new skin biopsy reports a severe lymphocytic infiltrate, suggesting cutaneous T-cell lymphoma. Immunohistochemistry reports: CD2 conserved, CD3 positive, CD4 positive in a 1: 3 ratio with respect to lymphocytesT CD8, CD5 decreased, CD7 conserved, CD20 negative, CD30 positive in activated lymphocytes (Figures 2 and 3). Bone marrow study, biopsy, myelogram and flow cytometry, confirms infiltration by T lymphocytes with immune-phenotype compatible with mycosis fungoides with expression of CD4 and CD2, finally making the diagnosis of cutaneous extranodal T lymphoma and Sézary syndrome. He received treatment with systemic CHOP protocol and second-line pralatrexate, and is currently in remission.

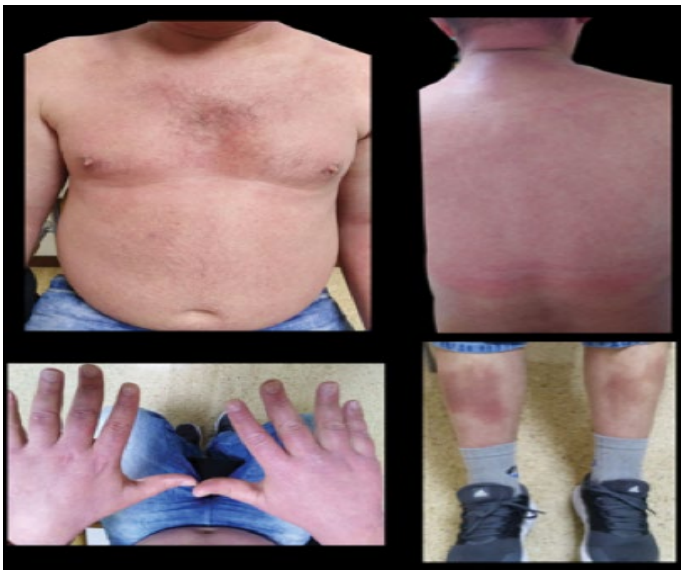

Figure 1. Physical examination of the patient.

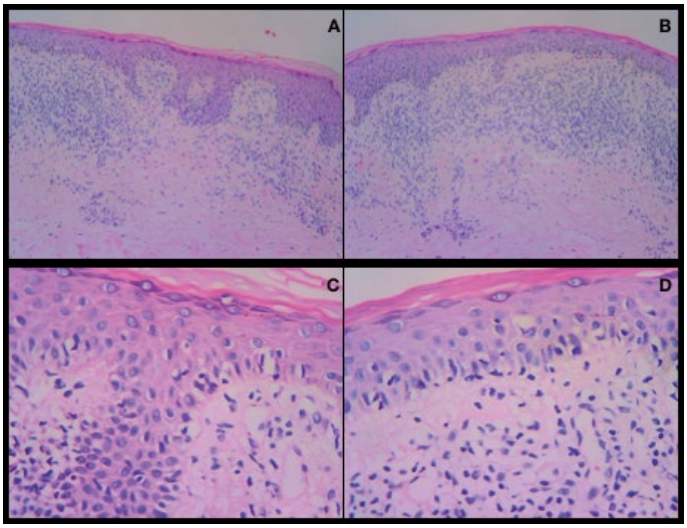

Figure 2. Histological study with hematoxylin and eosin (A and B 10x magnification, C and D 40x magnification).

***Address for Correspondence:** Estefania Galeano Piedrahita, Department of Dermatology, CES University, Carrera, 39# 5A-20, Medellín, Colombia, Tel: +57-3157787875; E-mail: estefigalea90@hotmail.com*

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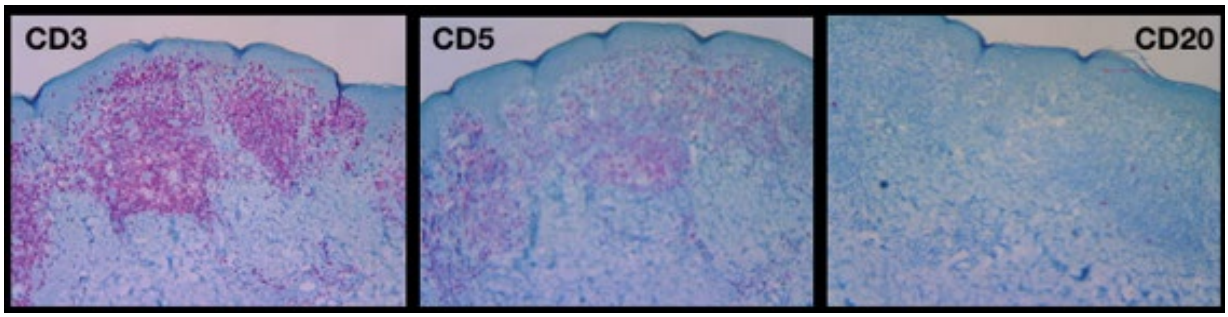

Figure 3. Immunohistochemistry study (10x magnification).

Discussion

Psoriasis is a chronic inflammatory disease, affecting 1% to 3% of the world population and treatment in a moderate to severe manner is carried out with systemic medications, such as methotrexate, cyclosporine, and biologics. In addition, several associations and comorbidities related to this pathology have been described, such as: Lymphomas, esophageal cancer, liver cancer, pancreas cancer, and keratinocyte carcinoma [3,4].

Mycosis fungoides (MF), represents 50% of cutaneous T-cell lymphomas (LCCT), with an incidence of approximately 10 cases per 1 million people, there are multiple clinical variants and is is very similar to benign dermatoses. Sézary syndrome is one of the most frequent forms of LCCT and is caused by the presence of erythroderma with at least 1000 circulating Sézary cells/mm^3 evaluated by flow cytometry. It´s difficult to make a diagnosis only with a skin biopsy and this can delay the diagnosis [1,5,6].

Several clinical studies have associated the appearance of MF in patients who have been systemically treated for psoriasis [7-9]. However, this has been controversial, because there is still no clear association between these two pathologies. It has been considered that there is a causal relationship due to the use of medications or if there is any immunological relationship between the diseases [5].

There are several reports in the literature of patients with psoriasis who present with MF lesions, but few studies of patients with an initial diagnosis of MF and with psoriasis lesions; furthermore the controversy becomes even more difficult given that there is a psoriasiform variant of MF [7].

On the other hand, researchers like Doniganand collaborators describe the association between mycosis fungoides and psoriasis, supported by the immune dysregulation of Th1 cells that occurs in both diseases. In addition, Th17 lymphocytes and the IL23/Th17 axis are involved in psoriasis and recently, Th17 cells and their cytokines (IL17A/F, IL-21, IL-22) have also been implicated in MF and they even found that the prevalence of psoriasis was higher in these patients (19.8%) than in the general population (1% to 3%) [10,11].

However, there is also a clear relationship between chronic inflammation and malignancies, which may explain this association, in addition to the chronic lymphocyte proliferation that occurs in psoriasis [12]. The risk of presenting LCCT is increased when psoriasis is moderate to severe, when they have received systemic treatment or phototherapy, and in elderly patients [11,13].

Among other causes of association between these two entities, is the use of immunosuppressants and immunomodulators, which have been used for psoriasis and may facilitate the development of cutaneous T-cell lymphoma or even favor the transformation of psoriasis to MF, as there are some cases reported in the literature with the use of methotrexate, PUVA, cyclosporine, and biologics [14]. It is noted that, in some studies, there were errors in the classification of psoriasis and MF, possibly because these two entities may share histopathological characteristics such as epidermal hyperplasia and exocytosis of CD4+T lymphocytes in the dermal infiltrate [15].

More studies are needed to associate and relate these two entities and that evaluates if the use of immunosuppressive/immunomodulatory drugs can influence psoriasis, generating a transformation to mycosis fungoides.

Conclusion

The clinical and histological similarity of MF and Sézary syndrome with other benign dermatoses, in addition to the possibility of overlapping between them, represents a challenge for the dermatologist when it comes to making an appropriate approach and diagnosis, in addition to long and unnecessary treatments for the patient. The relationship that exists so far between MF and psoriasis opens up an interesting field of study and for the development of future diagnostic aids and therapeutic targets.

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