Diffuse Systemic Large-B-Cell Lymphoma With Secondary Skin Involvement

Linfoma B difuso de células grandes sistémico con afectación cutánea secundaria

To the Editor:

Diffuse large B-cell lymphoma (DLBCL) is the most common type of systemic lymphoma and accounts for 40% of all new cases of this disease. Prognosis has improved considerably since the introduction of rituximab, which combined with chemotherapy is associated with a cure rate of up to 50%\(^1\); R-CHOP is the regimen of choice. Secondary cutaneous involvement is uncommon in DLBCL, although it has been observed in up to 20% of cases in some series\(^2\).

We present the case of an 81-year-old man in good health who presented with an indurated, erythematous-violaceous plaque-like lesion with a diameter of \(7 \times 4\) cm on the right leg (Fig. 1). The plaque had appeared 3 weeks earlier.

A biopsy was performed to investigate the suspected diagnosis of primary cutaneous lymphoma, with results showing a diffuse, dense infiltration of large, highly pleomorphic atypical basophilic cells in the dermis (Fig. 2). Immunohistochemical stains were positive for B-cell markers (CD20, CD79a), negative for T-cell markers (CD3, CD5), and strongly positive for Bcl-2. Less intense staining was observed for Bcl-6 and MUM-1, and the results were negative

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A computed tomography staging study revealed multiple enlarged infradiaphragmatic and supradiaphragmatic lymph nodes and a tumor measuring 6 cm on the left kidney. Bone marrow biopsy showed an infiltration of small cells with positive B-cell markers. Unlike the skin biopsy, the bone marrow biopsy showed CD5 expression and absence of Bcl-6 and MUM-1 expression, leading to a diagnosis of chronic lymphocytic leukemia. The definitive diagnosis was thus systemic DLBCL with secondary cutaneous involvement due to a previous unknown transformation of chronic lymphocytic leukemia (Richter syndrome or transformation). The patient was treated with R-CHOP, which initially resulted in a reduction in the size of the skin tumor and the lymph nodes. However, he died 4 months later due to neutropenic sepsis.

DLBCL is a heterogeneous entity with multiple clinical and pathologic variants. It is most common in individuals aged over 60 years. It is also the most common type of lymphoma in patients with human immunodeficiency virus infection. Because of its high proliferative activity, it is sometimes diagnosed earlier than other more indolent lymphomas. Immunophenotypically, all DLBCLs express B-cell markers, and other markers are expressed in a variable proportion of tumors (Bcl-6 in 60%-90%, MUM-1 in 35%-65%, and Bcl-2 in 50%). Although Bcl-2 is expressed in over 60% of tumors, Bcl-2 rearrangement is observed in under 20% of cases, suggesting the existence of other genetic pathways for the expression of this protein. There has also been a description of a double-hit variant involving both Bcl-2 and Myc rearrangement associated with worse prognosis and a greater tendency for secondary cutaneous involvement.

The primary cutaneous variant of DLBCL, primary cutaneous diffuse large B-cell lymphoma (PCDLBCL), leg type, is one of the 3 most common types of primary cutaneous B-cell lymphomas, together with primary cutaneous follicle center lymphoma and marginal zone B-cell lymphoma. PCDLBCL, leg-type presents as solitary or multiple violaceous plaques that typically occur on the legs of elderly patients. It is by far the primary cutaneous B-cell lymphoma with the worst prognosis, with a 5-year survival of 50%. It is therefore essential to initiate treatment with multi-agent chemotherapy and rituximab as early as possible. Because the immunohistochemical markers are the same for PCDLBCL, leg type and its systemic variant, DLBCL, according to the current WHO-EORTC criteria.

For CD10 and CD30 (Fig. 3A and B), Ki-67 staining showed high proliferative activity (90%). Fluorescent in situ hybridization was negative for Bcl-2 rearrangement with the t(14;18) translocation.
definitive diagnosis should be based on the presence or absence of extracutaneous disease at diagnosis. Testicular B-cell lymphoma can also affect the skin and its pathologic and immunophenotypic characteristics are indistinguishable from those of PCDLBCL, leg type; routine testicular examination is therefore recommended in appropriate cases.\textsuperscript{6}

We have reported a case of systemic DLBCL diagnosed following the detection of a cutaneous lesion without other signs or symptoms. One particularly interesting aspect of this case was the presence of a solitary lesion that clinically resembled a primary cutaneous lymphoma.

References


C. Lloret-Ruiz,\textsuperscript{*} P. Molés-Poveda, N. Barrado-Solís, E. Gimeno-Carpio
Servicio de Dermatología, Hospital Arnau de Vilanova, Valencia, Spain

*Corresponding author. E-mail address: cllloretruiz@gmail.com (C. Lloret-Ruiz).

Psoriatic Cheilitis: A Report of 2 Cases Treated Successfully With Topical Tacrolimus and a Review of the Literature

Quelitis psoriásica: Un reporte de 2 casos tratados con éxito con tacrolimus tópico y revisión de la literatura

Dear Editor,

Psoriasis is a chronic inflammatory skin disease that typically affects the extremities, trunk, scalp, and nails. Psoriatic cheilitis as an exclusive presentation is very rare,\textsuperscript{1} and to our knowledge, only 5 cases have been reported to date.\textsuperscript{2-4} The absence of cutaneous lesions causes diagnostic difficulties that can result in misdiagnosis and inadequate treatment.

We report the cases of 2 young adults who presented with psoriasis of the vermilion of the lips as the only disease manifestation. Response to topical tacrolimus 0.1% treatment was good in both cases.

The first case involved a 28-year-old white woman referred to our clinic with a 3-month history of scaly plaques on the vermilion of her lips and a clinical diagnosis of contact cheilitis. Clinical examination revealed an erythematous fissured plaque over the entire upper and lower lip surface, covered by thick white-yellowish scales (Fig. 1A). Full body examination did not reveal any evidence of intraoral or cutaneous involvement. Apart from a 10-year history of Hashimoto thyroiditis, the patient’s medical history was unremarkable. However, she did mention that her brother had psoriasis. Contact cheilitis was excluded following negative patch tests. Subsequent biopsy and histological examination confirmed the clinical suspicion of psoriasis.

The patient received initial treatment with salicylic acid 5% ointment twice daily for 5 days, followed by a 1-month course of topical tacrolimus 0.1% twice daily, resulting in adequate control of disease activity (Fig. 1B). The patient is currently on maintenance treatment with twice-weekly application of tacrolimus 0.1% ointment.

The second case involved a 20-year-old white man referred to our clinic for further assessment of a 2-year history of recurrent erosive cheilitis, characterized by fissures and white-yellowish scales on the vermilion borders of both lips (Fig. 2A). Intraoral and cutaneous lesions were absent. The patient reported exacerbations, particularly during winter. His medical history was unremarkable; there was, however, a positive family history of psoriasis (his mother). After excluding contact cheilitis by patch testing, we performed a lip biopsy. Histopathology showed mounds of parakeratosis and hypergranulosis, acanthosis, and dilated and vertically elongated papillary vessels (Fig. 2B), leading to a diagnosis of psoriatic cheilitis. The patient was started on salicylic acid 5% twice daily for 3 days, followed by topical tacrolimus 0.1% twice daily for the next month. The treatment resulted in the gradual remission of lesions, but discontinuation led to a flare-up, which was managed with the same regimen. To avoid recurrences the patient was advised to use tacrolimus 0.1% twice weekly, with excellent results.

Perioral psoriasis is an unusual presentation of psoriasis. It is clinically characterized by cracking and scaling of the lips and can have a profound emotional, social, and physical impact on patients’ lives.\textsuperscript{1,4} In the vast majority of the cases, coexistence of typical psoriatic lesions elsewhere on the body facilitates diagnosis. To the best of our knowledge, exclusive lip involvement is very uncommon, with only 5 cases reported in the literature.\textsuperscript{2-4} In 2 of these cases, involvement of the vermilion of the lips preceded cuta-