**Mycosis Fungoides, Lymphomatoid Papulosis and Hodgkin’s Lymphoma in the Same Patient: Apropos of a Possible Monoclonal Origin**

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**Abstract**

A 59-year-old man with Hodgkin’s lymphoma was referred by a hematologist for consultation for cutaneous issues. Physical examination revealed generalized scaling and erythematous scaly patches located in the groin, abdomen, and arms. The biopsy was compatible with mycosis fungoides (MF). At his next medical visit, painful nodules with erythematous halo and scabby surface were noted, and a subsequent biopsy was compatible with lymphomatoid papulosis (LyP). Mycosis fungoides, the most common primary cutaneous T-cell lymphoma, is usually defined in its classic form as a CD4⁺ non-Hodgkin lymphoma; LyP corresponds to a CD30⁺ lymphoproliferative disorder; and Hodgkin’s lymphoma (HL) constitutes a lymphoid neoplasia characterized by the presence of Reed–Sternberg cells and its variants. Although these entities have been defined independently, evidence suggests the possibility of a common monoclonal origin. To our knowledge, this is the first case of MF, LyP, and HL in a single patient.

**Key Words:** Hodgkin’s lymphoma, lymphomatoid papulosis, monoclonality, mycosis fungoides

**Introduction**

Mycosis fungoides (MF) constitutes the most common primary cutaneous T cell lymphoma. Its classic form is defined as an indolent CD4⁺ non-Hodgkin lymphoma which manifests itself initially as scaly patches that may evolve into plaques and/or tumors. Multiple clinicopathological variants have been reported. Lymphomatoid papulosis (LyP) is a CD30⁺ lymphoproliferative disorder. It is considered a disease with a benign clinical course, but with a histological malignant appearance, classically characterized by self-limiting papules and nodules. In 20% of the cases it is associated with other lymphomas, mainly MF. Hodgkin’s lymphoma (HL) is a neoplasia characterized by the presence of Reed–Sternberg cells and its variants. In its classic form, it is considered to be a lymphoma of B lineage origin. Although these three entities have been defined independently, the evidence and the reports of cases in which two of these diseases coexist, suggest the possibility that in some patients there might be a common pathogenesis underlying the two conditions. But presence of all the three conditions in the same patient has not yet been reported.

To our knowledge, our presentation is the first case report of MF, LyP, and HL coexisting in the same patient.

**Case Report**

A 59-year-old man was referred by a hematologist for cutaneous lesions. His hematological history began 3 months prior to his dermatological visit and consisted of diarrhea, diffuse eczema, and a weight loss of 12 kg, without fever or night sweat. An etiological study was performed to assess the diarrheic episode to rule out an infectious disease. An axillary adenopathy was discovered and the biopsy was compatible with HL [Figure 1]. Enzyme-linked immunosorbent assay (ELISA) test for HIV was negative. Positron emission tomography–computed tomography (PET-CT) showed a supra and infra-diaphragmatic nodal compromise with an associated pleural effusion. A bone marrow biopsy was performed which reported normal hematopoiesis. A diagnosis of nodular sclerosis–type Hodgkin lymphoma stage IIIB was made, and chemotherapy was started with adriamycin, bleomycin, vinblastine, and dacarbazine (ABVD) regimen.

At the time of his dermatological consultation, the patient was still under this treatment, with no other relevant...
medical background. He had a history of 1.5 years of generalized xeroderma and skin desquamation, initially asymptomatic and subsequently pruritic. The physical examination revealed a generalized ichthyosiform desquamation and the presence of erythematous, scaly plaques on the left groin area, abdomen, and upper extremities. A provisional diagnosis of MF was made and 5 punch biopsies were taken. The serology for human T-lymphotropic virus (HTLV) type 1 and type 2 and Epstein Barr virus were negative; LDH level was normal. The patient came for his dermatology follow-up having finished his hematology treatment. The result of the skin biopsy showed orthokeratosis, mild spongiosis, and multiple foci of epidermotropism with some roughly shaped Pautrier’s microabscesses. The dermis had a superficial infiltrate of histiocytes and atypical lymphocytes with large, hyperchromatic nuclei with an irregular contour. Immunohistochemistry was positive for CD4 in 80 percent of the lymphocytes and positive for CD8 and CD7 in 15 and 5 percent of the lymphocytes, respectively. These findings were compatible with the diagnosis of MF [Figure 2]. During his medical visit, the patient complained of appearance of some new painful nodules. Physical examination revealed nodules with an erythematous halo and superficial crust on his abdomen and lower extremities [Figure 3]. An infectious component was suspected, a cutaneous culture was obtained, and our empirical treatment began with cefadroxil and cold cream, having the intention of starting psoralen and ultraviolet A (PUVA) therapy once the infection was resolved. Vitamins D and B12 levels were also requested. Due to a nonresolving clinical course of the painful nodules after 10 days of treatment, a biopsy was performed. Histological studies showed orthokeratosis, irregular acanthosis, moderate spongiosis, and exocytosis of small and medium lymphocytes, with an ulcerated area. The dermis had a nodular, perivascular and interstitial, superficial and deep infiltrate, mainly composed of neutrophils and atypical lymphocytes containing irregular, vesicular nuclei and visible nucleoli. Periodic acid–Schiff (PAS) staining was negative for fungi. Immunohistochemistry was positive for CD3 in 90 percent of the lymphocytes and positive for CD25 and CD30 in 50 percent of the lymphocytes. These findings were compatible with the diagnosis of LyP [Figure 4]. The skin culture showed normal skin flora. Vitamin D level was 7.5 ng/ml and that of B12 was 262 pg/ml.

Molecular clonality testing was performed using polymerase chain reaction (PCR) to assess for T cell receptor (TCR) gene rearrangement. The result was positive for monoclonal TCR-gamma rearrangement, a
possible evidence of a same clonal origin for both the MF and the LyP [Figure 5]. Due to technical limitations, we could not assess the clonality of the Reed–Sternberg cells in this case.

During next medical visit, the physical examination of the patient showed clinical improvement, but with a diffuse desquamation on his extremities, abdomen and torso, and an erythematous scaly plaque on his left leg. PUVA-therapy was started with vitamin D supplementation and cold cream application. A good clinical response to phototherapy could be observed.

After a year of remission, the patient presented with a relapse of his HL. Second line chemotherapy was started with anifosfamide, carboplatin, and etoposide (ICE) regimen. After completing two cycles, an autologous bone marrow transplant was performed. One month later, new scaly plaques and nodules appeared on his arms. Biopsy was compatible with mycosis fungoides with large cell transformation. The patient was subsequently planned to begin treatment with systemic bexarotene.

Discussion
There is a biological plausibility for a unique neoplastic clone of T lymphocyte to manifest a clinical concomitance of more than one type of lymphomas. Likewise, epidemiology supports these associations. Notwithstanding such findings, when it comes to clinical corroboration, there is scarcity of publications, for which reason the monoclonal origin hypothesis is still controversial, requiring yet greater amount of evidence to establish a possible pathological spectrum. Our case represents, to our knowledge, the first report of MF, LyP, and HL diagnosis in the same patient.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information.
to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**
There are no conflicts of interest.

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