Papular Mycosis Fungoides: A Case Report and Review in the Literature

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Sir,

A 35-year-old male patient presented with skin eruptions on his trunk and upper and lower extremities, which had appeared 2 years ago. The lesions were persistent and did not show any tendency for spontaneous resolution. There was no history of any other dermatological disease or drug intake. Dermatological examination revealed a great number of slightly scaly erythematous papules on his trunk and upper and lower extremities, clustered together or scattered, and varying in sizes between 2-3 mm and 8-9 mm [Figure 1]. Histopathological examination revealed mild acanthosis in the epidermis and exocytosis of lymphocytes with hyperchromatic convoluted nuclei exhibiting apparent epidermotropism. There were small collections of lymphocytes (Pautrier's microabscesses) within the epidermis in some areas [Figure 2]. Immunohistochemical examination revealed CD8 positive staining in the intraepithelial focal regions; CD3 positive staining in the perivascular lymphocytes and intraepithelial focal regions; CD45RO positive staining in the perivascular lymphocytes and CD4 and CD30 negative staining. Molecular analyses of the T-cell receptor gene rearrangement showed the presence of a monoclonal band. With these findings, the patient was diagnosed with mycosis fungoides (MF) and psoralen + ultraviolet A (PUVA) treatment was started. Complete regression was achieved after 2 months. The patient had no recurrence during a period of 10 months after first diagnosis. Common patches of MF were not observed during the follow-up visit.

Papular MF is a new variant of early MF, characterized with presence of papular lesions instead of conventional lesions of the disease. It was first demonstrated by Kodama *et al.*,[1] in 2005. Thus far, to our knowledge, only 11 patients with papular MF have been reported in the literature.[1-5]

Papular MF affects most often between 27 and 61 years of age, with an average age of 47.3. The lesions consist of discrete erythematous tender papules, ranging from 1 to 10 mm. The papules are usually localized and distributed symmetrically. Papular MF mostly occurs on the trunk, upper arms, and thighs.[3]

Papular lesions of MF may present in two different situations. Papules may ocur during the course of MF, which is frequently a sign of progression, similar to the onset of plaques or tumors. The same lesions can also occur at the onset of the disease without any evidence or history of preceding patches. These papules are nonspecific and the diagnosis of MF is established histopathologically.[3]

The differential diagnosis for papular MF includes lymphomatoid papulosis (LyP) type B, pityriasis lichenoides chronica (PLC), and pityriasis lichenoides et varioliformis acuta (PLEVA). LyP type B is characterized by waxing and waning papules and ulcerated, crusted nodules. Although histopathologic features of LyP type B are considered to be similar or even identical to those observed in MF, LyP type B shows variable

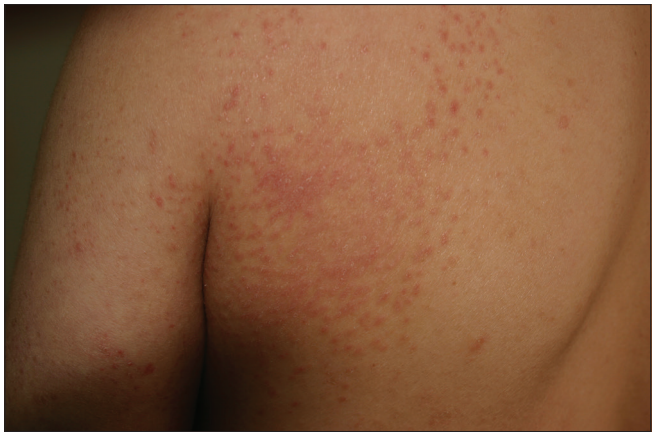

Figure 1: Erythematous papules on the left scapular region

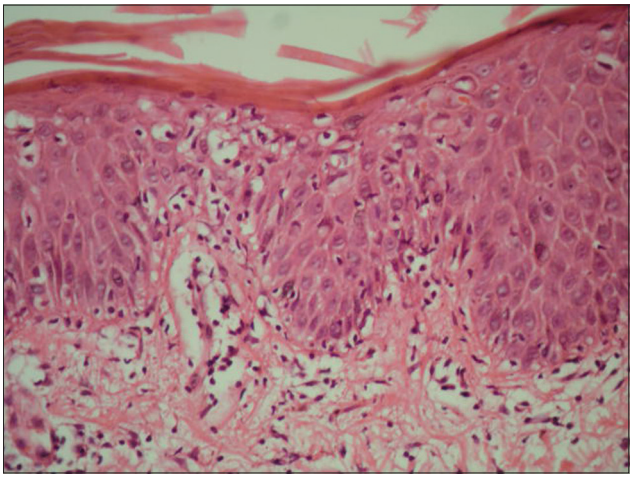

Figure 2: Lymphocytes with hyperchromatic convoluted nuclei exhibiting apparent epidermotropism and small collections of lymphocytes (Pautrier's microabscesses) in some areas within the epidermis (50 x 10, H and E)

positivity for CD30. Our case can be distinguished from LyP type B by lesions not being pleomorphic, the lack of tendency towards ulceration and spontaneous resolution and by CD30 negative staining.[6]

PLEVA and PLC are among the various dermatoses that are clinically confused with papular MF. PLEVA is frequently characterized by eruptions of pink, orange, or purpuric papules that undergo central vesiculation, may ulcerate, and resolve with hemorrhagic crusts. Histopathological features of PLEVA are different from papular MF: Thickened horny layer with parakeratosis, intracorneal neutrophils, and a cytotoxic T-cell infiltrate. However, It should be kept in mind that a pityriasis lichenoides et varioliformis acuta-like type of MF has been reported rarely in children.[7] PLC may clinically present as similar to papular MF, and rare cases of PLC eventuating into MF have been reported.[8] However, histopathologic features of PLC are different from those observed in papular MF.

The etiopathogenesis of MF is not exactly known. In early stage MF lesions, T helper 1 and $CD8^+$ cells are intensively found and they control malignant clonal proliferation. In advanced stage MF, T-helper 2 cytokine such as IL-4 and IL-10 increase. $CD8^+$ cells decreases and thus neoplastic cells increase.[8] CD8 positive staining was defined in our case, which seemed to support the early stage MF lesions. CD8 positive staining was also determined only in one of the cases previously reported in the literature.[5]

The diagnosis of papular MF was made in our case, by the presence of only erythematous papules, by absence of patch and plaque lesions before these lesions, and upon confirmation with histopathologic and immunohistochemical methods. This case is presented in order to keep in mind papular MF, which is a new and very rare variant of early stage MF, in evaluating patients presenting with pruritic, erythematous papules.

References

1. Kodama K, Fink-Puches R, Massone C, Kerl H, Cerroni L. Papular mycosis fungoides: A new clinical variant of early mycosis fungoides. J Am Acad Dermatol 2005;52:694-8.
2. Uddin A, Bennett M, Nayeem K, Marren P, Abushaira H. A case of papular mycosis fungoides: New clinical variant of early mycosis fungoides. J Eur Acad Dermatol Venereol 2007;21:685-7.
3. Martorell-Calatayud A, Botella-Estrada R, Sanmartín-Jimenez O, Requena C, Guillén-Barona C, Sangüeza OP. Papular mycosis fungoides: Two new cases of a recently described clinicopathological variant of early mycosis fungoides. J Cutan Pathol 2010;37:330-5.
4. Liu ZH, Wang YL, Chen SY, Zheng JH, Qiao G, Shen H, *et al*. Papular mycosis fungoides: A new clinic variant of early and benign mycosis fungoides? J Clin Oncol 2011;29:e381-3.
5. Neri I, D'Acunto C, Pileri A, Reggiani C, Patrizi A. Papular mycosis fungoides: A new case expanding the spectrum of phenotypic and clinical findings. G Ital Dermatol Venereol 2011;146:505-7.
6. El Shabrawi-Caelen L, Kerl H, Cerroni L. Lymphomatoid papulosis: Reappraisal of clinicopathologic presentation and classification into subtypes A, B, and C. Arch Dermatol 2004;140:441-7.
7. Ko JW, Seong JY, Suh KS, Kim ST. Pityriasis lichenoides-like mycosis fungoides in children. Br J Dermatol 2000;142:347-52.
8. Tomasini D, Zampatti C, Palmedo G, Bonfacini V, Sangalli G, Kutzner H. Cytotoxic mycosis fungoides evolving from pityriasis lichenoides chronica in a seventeen-year-old girl. Report of a case. Dermatology 2002;205:176-9.

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