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CASE REPORT

Papular mycosis fungoides: Six new cases and association with chronic lymphocytic leukemia

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Abstract

Papular mycosis fungoides (MF) is a rare presentation of MF. Six illustrative cases of papular MF were retrospectively reviewed. Five of the cases studied by immunohistochemistry had variable numbers (range: 1%-20%) of CD30+ cells in the dermal infiltrate, a finding that is characteristic of lymphomatoid papulosis but may occasionally occur in typical early MF. Although none of our papular MF patients had progressive disease, lesions with relatively high numbers of CD30+ cells in 3 patients did not respond well to skin-directed treatments used for MF. Interestingly, these patients had evidence of co-existing clonal B cell populations in the blood (one with clonal B cell lymphocytosis and two with B-cell chronic lymphocytic leukemia). We conclude that: (1) papular MF may contain CD30+ cells, thereby causing confusion with lymphomatoid papulosis; and (2) papular MF, like more typical MF, may be associated with clonal B-cell proliferations including chronic lymphocytic leukemia.

Key words: Mycosis fungoides; Lymphocytosis; Chronic lymphocytic leukemia; Papule; Cutaneous lymphoma

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Core tip: Mycosis fungoides presenting with papules

as the only clinical manifestation is a rare variant of the disease. To date only 16 cases of papular mycosis fungoides have been described in the literature and none had CD30+ cells. We report 6 additional cases, 5 with 1%-20% CD30+ cells. Three cases had co-existing clonal B cell lymphoproliferation (2 with chronic lymphocytic leukemia). The possible pathogenic relationship between mycosis fungoides and chronic lymphocytic leukemia is discussed.

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INTRODUCTION

Mycosis fungoides (MF), a great masquerader of other skin diseases, can present with varied types of lesions that are confused with infectious and drug related eruptions among others[1,2]. Recently, Kodama reported 6 cases of "papular MF" that presented with persistent papules that had the histopathologic features of MF but without typical patch/plaque MF lesions nor evidence of a lymphomatoid drug reaction[3]. Lymphomatoid papulosis (LyP) was excluded by the absence of spontaneous regression of lesions and lack of CD30+ cells in the dermal infiltrate. With follow up, 2 cases subsequently developed typical skin manifestations of MF (one developed MF patches only 2 mo after the diagnosis of papular MF).

At the time of this report, 10 additional cases of papular MF have been published (Table 1)[4-11]. Collectively, these papular MF cases (8 men, 8 women, ages, 27 to 83 years) are characterized by the following: (1) persistent papules, sometimes only a few millimeters in diameter, that did not enlarge into nodules, plaques or tumors; (2) Pautrier microabscesses in 8 of 14 cases; (3) a CD4+ immunophenotype in 8 cases and a CD8+ phenotype in 2 cases; (4) negative staining for CD30 in all 16 cases; (5) clonal T cells demonstrated in 7 of 8 cases; (6) subsequent appearance of typical patch or plaque lesions of MF in 3 cases including Kodama's 2 cases; and (7) an overall non-progressive clinical course.

Herein we report our experience with 6 additional cases of papular MF. Unlike reported cases, variable numbers of CD30+ cells were observed in the dermal infiltrate in 5 cases and 3 cases had evidence of co-existing clonal B-cell proliferations in the blood. The significance of these findings is discussed.

CASE REPORT

The registry of patients with cutaneous T cell lymphoma (1481 patients diagnosed with MF excluding its erythrodermic variant) that is maintained by one of us with approval of the Institutional Review Board at Johns Hopkins University was reviewed for cases that fulfilled the clinical-pathological criteria for papular MF as defined by Kodama[3]. Information obtained at the time of initial presentation, subsequent staging and follow up provide the basis of this report. This includes the results of histopathology, immunohistochemistry on corresponding frozen sections, and PCR amplification of T cell receptor gamma (TCR-γ) chain gene for T cell clonality on representative lesions.

The Surveillance, Epidemiology, and End Results (SEER)-9 registry, which captures data from 9.4% of the total United States population, was analyzed using SEER*Stat 8.2.1 software to determine the relative risk of developing chronic B-cell leukemia (ICD-O-3 Site C42.0, C42.1, C42.4 and ICD-O-3 code 9823/3) in patients initially diagnosed with MF (ICD-O-3 code 9700/3) and *vice versa* between 1973 and 2012. The statistical significance of the standardized incidence ratio (observed/expected) was determined using a Poisson distribution to calculate 95% confidence intervals.

Our retrospective review identified 6 patients who presented with persistent papules and/or small nodules with histopathologic features interpreted as diagnostic or consistent with MF (Table 2 and Figures 1-7). With follow-

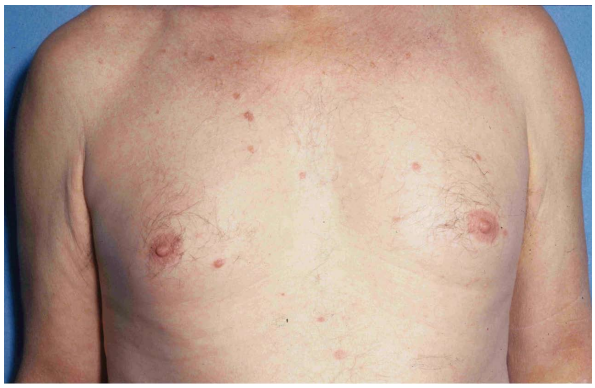

Figure 1 Patient 4 presented with a 6 mo history of persistent 2 to 8 mm papules of mycosis fungoides on the trunk and legs.

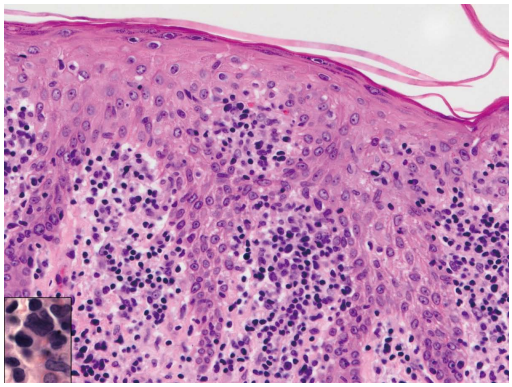

Figure 2 Skin specimen from patient 4 shows an acanthotic epidermis that contains atypical lymphocytes with hyperchromatic irregular nuclei (insert). The dermis has a superficial infiltrate composed of normal and atypical lymphocytes (H and E, × 400).

Table 1 Mycosis fungoides presenting as persistent papules in the literature

Case[1]	ARG; Dur	Distribution	Dermal infiltrate	Epidermal lymphocytes	Immunophenotype	PCR	Progression (time)[2]
Kodama1	57WM; NS	T	PV, F	Sm; PMAs	CD30-	ND	No
Kodama2	58WF; 2 yr	T, UE	Li, PV	Pleo Sm-med; No PMAs	CD4+30-	Pos	No
Kodama3	57F; Few mo	T, UE, LE	Li, PV	NS; No PMAs	CD30-	ND	Yes (3 yr)
Kodama4	41M; NS	LE	Li, PV	NS; No PMAs	CD30-	ND	Yes (2 mo)
Kodama5	59WM; 30 yr	T, UE, LE[3]	Li	NS; PMAs	CD30-	ND	No
Kodama6	61M; NS	T	PV	NS; PMAs	CD4+8-30-	ND	No
Uddin	31WF; 2 yr	T, UE, LE	Li, PV, SC, VA	NS; No PMAs	Mostly CD30-	ND	No
Martorell-Calatayud1	50WF; 2 yr	T[3]	A, P, Li, PV, F	Pleo Med-lg; PMAs	CD4+30-	Pos	No
Martorell-Calatayud2	55WF; 1.5 yr	T, LE[3]	NS	Sm-med; No PMAs	CD4+30-	Neg	No
Liu	27AM; NS	T[3]	Li	NS; PMAs	CD4+8-30-	ND	No
Neri	47WF; 1 yr	UE, LE	PV	Sm-med CL; PMAs	CD4-8+7+30-	Pos	No
Noe1	83WF; 3 yr	T, UE, LE	PV	NS; PMAs	CD4+30-	Pos	No
Noe2	65WF; 1mo	T	Li, PV	NS	CD30-	ND	Yes (NS)
Brajon	63WM; 10 mo	T, UE, LE	Li, PV	CL; NS	CD4+8-30-	Pos	No
Santamarina-Albertos	55WM; 1 yr	LE	Li	Sm-med CL; NS	CD4+30-	Pos	No
Balta	35WM; 2 yr	T, UE, LE	A, PV	NS; PMAs	CD4-8+30-	Pos	No (10 mo)

[1]References: [3-11]; [2]Time to progression of disease; [3]Some grouping or clustering of lesions. ARG: Age, race, gender; Dur: Duration to disease; PCR: Polymerase chain reaction for rearrangement of T cell receptor gamma chain; NS: Not stated; T: Trunk; UE: Upper extremities; LE: Lower extremities; Li: Lichenoid; PV: Perivascular; A: Acanthosis; P: Parakeratosis; F: Fibrosis; VA: Vacuolar alteration; SC: Subcutaneous; Sm: Small sized; Med: Medium sized; Lg: Large sized; Pleo: Pleomorphic; PMA: Pautrier microabscess; CL: Lymphocyte with cerebriform or infolded nuclei.

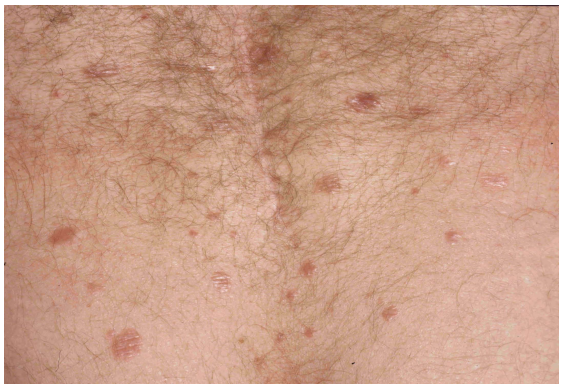

Figure 3 Persistent 2 to 14 mm papules of mycosis fungoides scattered on trunk and legs of patient 5.

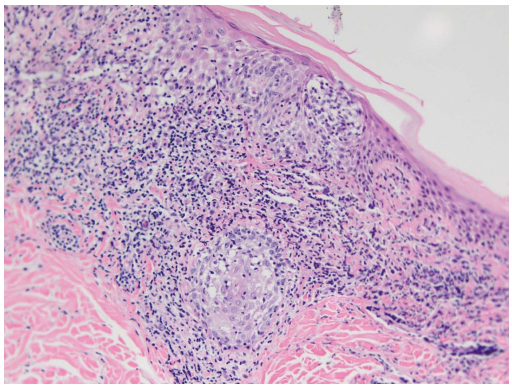

Figure 4 Skin specimen from patient 5 shows a moderately dense lichenoid infiltrate, wiry bundles of collagen in a thickened papillary dermis, and follicular mucinosis. Numerous atypical lymphocytes, some with large irregular nuclei, are located within the epidermis, both as solitary units and in aggregates, and dermal infiltrate (H and E, × 400).

up, none of these papular MF patients developed typical lesions of MF nor had disease progression. Pautrier microabscesses were described in skin specimens from 3 patients, and the immunophenotype of the neoplastic cells of 5 studied cases was CD4+CD8-. A dominant T cell clone was demonstrated by PCR in 3 cases.

Notably, all 5 patients evaluated for CD30 expression had variable numbers of scattered atypical CD30+ cells in the dermal infiltrate (estimated range: 1%-20%), a finding that suggested the possibility of type A LyP with epidermotropic T cells or possibly type B LyP. This was particularly true for the specimen from patient 2 (Figure

Table 2 Six additional patients with papular mycosis fungoides

Pt	ARG; Dur[1]	Lesions and Size (mm)	Distribution	Dermal Infiltrate	Epidermal Lymphocytes	Immunophenotype[2]	TCR-γ (method)	Course and Status (Duration FU)
1	68WM; 15 mo	Pa (1-2)	T, UE, UE	PA, VA, F, PC, Eos, LVC	Focal basilar Ep, Med CLs; No PMAs	ED: NS D: CD4 > CD8, CD30 10%	Pos Sk + Bd (SSCP)[3]	Controlled on prednisone; developed pancytopenia; DwD (99 mo)
2	47 WM; 19 yr	Pa (3-5)	H/N, T	A, PV, F, Eos, PC, MtF, LVC	Focal basilar Ep, Med-lg CLs; PMAs	ED: CD4+8-7-62L-30- D: CD4 90%, CD8 < 10%, CD7< 10%, CD62L < 1%, CD30 5%-10%	Pos Sk + Bd (DGGE)[3]	Poor or partial response to PUVA, MTX, isotretinoin, XRT, IFNα; DwD (171 mo)
3	57 WF; 5 yr	Pa (2-5)	LE, UE	P, A, Li, PV, CLs	Basilar Ep, CLs; No PMAs	ED: CD4+8-7+62L+30+/-[4] D: CD4 80%, CD8 20%-30%, CD7 40%, CD62L 50%, CD30 1%-2%	Neg (DGGE)	PUVA/NBUVB: CR; breast CA; RA, HT; A, NED (156 mo)
4	68 WM; 6 mo	Pa (2-8)	T, LE	A, Li, F, EE, Neu (v), CLs	Diffuse Ep, Med CLs; No PMAs	ED: CD4+/-8-7+62L-30- D: CD4 60%-70%, CD8 20%, CD7 70%, CD62L 70%, CD30 1%-2%	Pos (DGGE)	PUVA: CR ; no progression; AwD (210 mo)
5	58 WM; 2 mo	PaNd (2-14)	T, LE	Sp, Li, F, FM, CLs	Basilar Ep, Med-lg CLs; PMAs	Not available	Neg (SSCP)[5]	TopHN2: CR; No progression; A, NED (171 mo)
6	81 WM; 15 mo	PaNd (5-15)	T, LE	Li, F, CLs, MtF	Diffuse Ep, Med-lg CLs; PMAs	ED: CD4+8-7-62L-30- D: CD4 99%, CD8 1%, CD7 10%, CD62L 99%, CD30 20%	Neg (DGGE)[3,5]	PUVA: PR; DwD/MI (12 mo)

[1]Duration: Time from onset of disease to evaluation; [2]Estimated percentage of positively labeled cells in dermal infiltrate (frozen sections). CD2, CD3, and CD5 expressed by all cases (data not shown); [3]Clonal B-cell population detected by flow cytometry for patients 1, 2 and 6 and confirmed by PCR for patients 1 and 2 (see text for details); [4]30% of epidermotropic T cells expressed CD30; [5]Blood sample also negative for T cell clone. Pt: Patient; ARG: Age, race, gender; TCR-γ: T-cell receptor gamma chain rearrangement; FU: Follow up; Pa: Papule; Nd: Nodule; T: Trunk; UE: Upper extremity; LE: Lower extremity; A: Acanthosis; Sp: Spongiosis; P: Parakeratosis; VA: Vacuolar alteration; Ep: Epidermotropism; Li: Lichenoid (band-like); PV: Perivascular; PA: Periadnexal; PDE: Papillary dermal edema; F: Fibrosis; FM: Follicular mucinosis; Neu (v): Neutrophils in vessels; PC: Plasma cells; Eos: Eosinophils; EE: Extravasated erythrocytes; CL: Lymphocytes with cerebriform or infolded nuclei (mycosis cells); MtF: Mitotic figures; LVC: Lymphocytes with vesiculated nuclei and prominent nucleoli; PMA: Pautrier microabscess; Sm-med: CLs with small-medium sized nuclei (5 to 7 μm in diameter); Med-lg: CLs with medium-large sized nuclei (7 to 9 μm in diameter); ED: Epidermis; D: Dermis; >: Greater than; <: Less than; NS: Not stated; Sk: Skin; Bd: Blood; DGGE: Denaturing gradient gel electrophoresis; SSCP: Single-stranded conformation polymorphism; MTX: Methotrexate; XRT: Local field radiation therapy, IFNα: Interferon alfa; PUVA: Methoxsalen-ultraviolet A photochemotherapy; NBUVB: Narrow band ultraviolet B phototherapy; TopHN2: Topical mechlorethamine; CA: Carcinoma; RA: Rheumatoid arthritis; HT: Hashimoto's thyroiditis; MI: Mycocardial infarction; AwD: Alive with active disease; DwD: Dead with active disease; A: NED, alive, no evidence disease; LTF: Lost to follow up.

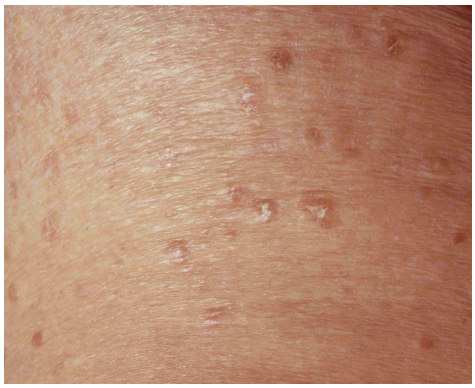

Figure 5 Patient 6 presented with a 15 mo history of persistent papules and small nodules, some with scaling, disseminated on the trunk and legs.

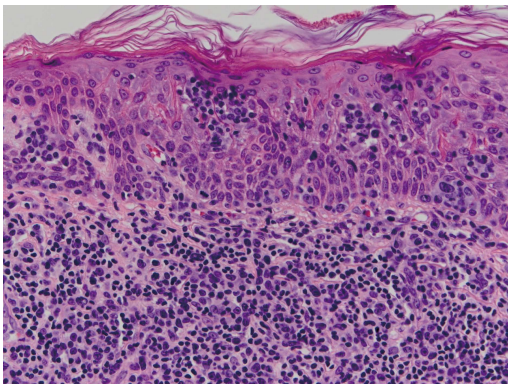

Figure 6 Skin specimen from patient 6 shows typical histopathologic features of mycosis fungoides. Nests of medium to large sized neoplastic lymphocytes with pleomorphic and cerebriform nuclei are observed within the epidermis (Pautrier microabscesses) and adjacent superficial dermis (H and E, × 400).

7). However, his skin lesions did not spontaneously regress as expected in LyP. In addition, CD30 also was expressed by 30% of the epidermotropic CD4+T cells of patient 3 (discussed below). Therefore, other than persistence of lesions, the histo-immunopathologic findings of papular MF overlap with those of LyP[12,13].

Table 3 Frequency of B-cell chronic lymphocytic leukemia occurring after a diagnosis of cutaneous T cell lymphoma

Ref.	Cohort	No. patients (Dx)	No. secondary B-CLL (%)
Olsen *et al*[27]	One institution	63 (CTCL)	0 (0)
Kantor *et al*[28]	One institution	519 (MF)	2 (0.39)
Väkevä *et al*[29]	Finnish cancer registry	319 (MF/SS)	1 (0.3)
Barzilai *et al*[24]	Two institutions	398 (MF)	2 (0.50)[1]
Huang *et al*[30]	One institution	429 (MF/SS)	1 (0.23)
Huang *et al*[30]	SEER-9 registry	1798 (MF/SS)	0 or 4 (0 or 0.22)[2]
Hallerman *et al*[31]	One institution	62 (CTCL)	0 (0)
Brownell *et al*[32]	One institution	672 (CTCL)	0 (0)
Hodak *et al*[33]	One institution	343 (MF)	2 (0.59)
Hodak *et al*[33]	Israeli population registry	683 (MF)	1 (0.15)
Lindahl *et al*[34]	Population-based	386 (MF)	0 (0)[3]
Current study	SEER-9 registry	3,977 (MF)	10 (0.25)

[1]One case with B-CLL preceding MF was excluded; [2]Surveillance, Epidemiology, and End Results (SEER)-9 Cohort between 1984 through 2001 included 4 cases of leukemia, not defined; [3]The 2 cases of hematologic cancer exclusive of non-Hodgkin lymphoma in this cohort were not B-CLL (Lindahl, personal communication, 2016). MF: Mycosis fungoides; SS: Sézary syndrome; CTCL: Cutaneous T cell lymphoma; B-CLL: B cell chronic lymphocytic leukemia; Dx: Diagnostic groups in cohort.

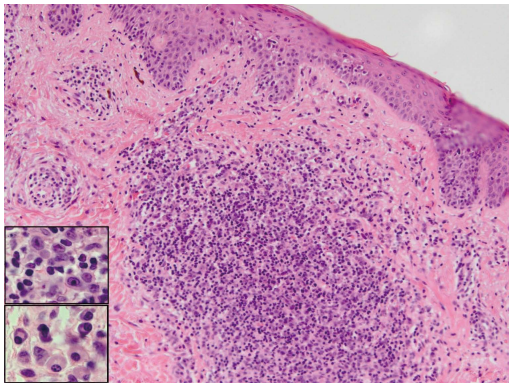

Figure 7 Skin specimen from patient 2 shows a perivascular and dense nodular infiltrate in the superficial and mid-dermis. A fibrotic papillary dermis and scattered epidermotropic lymphocytes aligned along the basal layer in the absence of spongiosis (H and E, × 400). The dermal infiltrate is composed of lymphocytes, some with large hyperchromatic cerebriform nuclei, large immunoblast-like cells (top insert), small clusters of plasma cells (bottom insert), and occasional eosinophils.

A second observation is that 3 of the papular MF patients had evidence of an associated clonal B-cell lymphoproliferation. Patient 1 had a T cell clone in skin and blood plus 6% of blood lymphocytes with a CD5+CD19+CD23+ phenotype and B cell clone demonstrated by PCR of the IgH gene in the blood, but not the skin. The small B cell population remained unchanged with follow-up and is therefore classified as clonal B cell lymphocytosis. Patient 2 had a T cell clone in skin and blood plus 65% of his blood lymphocytes were CD19+CD20+ B cells (absolute lymphocyte count: 770 cells/mm^3) and evidence of a B cell clone by PCR in the blood but not the skin. A subsequent bone marrow analysis revealed 20% B cells co-expressing CD5 and CD23 characteristic of chronic lymphocytic leukemia (B-CLL). Patient 6 also had a B cell clone in the blood by flow cytometry (21% of lymphocytes with a CD5+CD19+CD20+ phenotype; absolute lymphocyte count: 2490 cells/mm^3) but a negative PCR study when initially evaluated. However, a diagnosis of B-CLL was confirmed 6 mo later. These patients with clonal B cells tended to have higher percentages of CD30+ cells in their skin lesions and their response to treatment was partial or transitory compared to the other papular MF cases.

DISCUSSION

Papular MF is a very rare presentation of the disease, occurring in 0.4% of non-erythrodermic MF cases referred to our center. However, our patients differed from published cases with regard to the presence of atypical CD30+ cells in the dermal infiltrate in 5 studied specimens. Specimens from 3 patients had estimated numbers of dermal CD30+ cells that ranged from 5% to 20% such that LyP would be an alternative diagnosis. However, unlike LyP as currently defined, these lesions did not undergo spontaneous regression. Atypical CD30+ cells may also be encountered in clinically early lesions of MF so this finding does not exclude papular MF from the differential diagnosis[14]. The clinical significance of CD30+ cells in this context is unclear. It has been reported that CD30 expression in non-transformed patch or plaque phase MF has an adverse prognostic significance[14]. Although none of the patients in our small series developed more typical lesions of MF nor had disease progression, the 3 cases with 5% or more CD30+ cells in the dermal infiltrate did not respond adequately to various skin-directed therapies used to treat early MF.

An unexpected and previously unreported observation was that 3 patients with papular MF had an associated B-cell lymphoproliferative disorder (one with monoclonal B cell lymphocytosis and two with B-CLL). This raises the possibility that some of our papular MF cases might be examples of pseudo-MF reactions associated with B-CLL as described by Ingen-Housz-Oro[15]. In that paper, the

authors reported 4 patients that presented with localized papules in concert with B-CLL. Three patients were diagnosed to have a pseudo-MF reaction and one had papular MF. All cases had evidence of folliculotropism by lymphocytes and 3 had follicular mucinosis including the papular MF case. Of note, a T cell clone could not be demonstrated by PCR of the TCR-γ chain gene in all cases, whereas clusters of neoplastic B cells were observed in 3 cases including the papular MF case. CD30 staining was not performed. Therefore our papular MF cases differ from Ingen-Housz-Oro's cases in several ways: (1) in our patients, lesions were more widespread; (2) folliculotropic T cells and a B cell component in the infiltrate were not present; and (3) T cell clonality was demonstrated in two cases. Of interest, mature appearing plasma cells were observed in the dermal infiltrate of skin specimens obtained from patients 1 and 2 who had evidence of clonal B cells in the blood but not the skin (Figure 7). In addition, a prior skin specimen from patient 6 and studied elsewhere also showed numerous plasma cells. This phenomenon may be the result of a homing process as suggested by Ingen-Housz-Oro[15].

The association of MF and B-CLL may not be a fortuitous event. A review of the literature uncovered 23 cases of classic patch, plaque or tumor phase MF (erythrodermic MF excluded) co-existing with B-CLL[15-26]. Of interest CD30 staining was performed on skin specimens from only 2 cases and both were reported to be negative. Nevertheless, it has not been established that the risk of developing secondary B-CLL in MF patients is significantly higher than for the general population (Table 3)[27-34].

In the SEER-9 database, 1973 to 2012, there are 3977 cases coded as MF as the primary cancer for analysis. Of these, B-CLL was subsequently diagnosed in 10 cases compared to an expected frequency of 6.77 cases. Therefore, the relative risk (observed/expected) is 1.48 (95%CI: 0.71-2.71). Conversely, of 34160 cases with B-CLL as the primary cancer, 7 developed MF as a second cancer for a relative O/E of 7/4.02 or 1.74 (95%CI: 0.7-3.59). Although these relative risks are increased, they are not statistically significant. However, the possibility that the number of MF cases in the SEER registry might be under reported must be considered for several reasons: (1) some MF cases may be diagnosed as cutaneous T cell lymphoma and therefore coded by registrars as such (ICD-0-3 code 9709/3); (2) cases of MF and B-CLL that are diagnosed concurrently are coded separately as primary cancers; and (3) perhaps not all cases of MF are reported to the SEER registry by private dermatologists or dermatopathology laboratories[35].

With regard to the first point, of 1304 patients coded initially as having cutaneous T cell lymphoma, 18 patients were subsequently coded as MF compared to an expected number of 0.15. The observed/expected ratio was 121.58 (95%CI: 72.06-192.15) was significantly high ($P < 0.05$). It is therefore conceivable but not proven that some patients with MF might be coded initially in the broader diagnostic category of cutaneous T cell lymphoma.

Incidentally our review also uncovered a case reported in 1983 that was characterized by disseminated therapeutically resistant papules with histopathologic features of MF in a patient with B-CLL[36]. We propose this case could represent the first example of papular MF associated with B-CLL.

The underlying basis for the uncommon but well documented association of MF and other forms of cutaneous T cell lymphoma with various B cell lymphoproliferations is unclear. Our hypothesis, which also has been suggested by others[18,19], is that an inherited genetic attribute that predisposes a patient to lymphoma (such as a nucleotide polymorphism)[37] or an acquired mutation is present at the level of the common lymphoid progenitor cell. If additional genetic alterations that promote lymphoma occur later in both the B and T cell developmental pathway, this would account for the observed associations of various T and B cell lymphoproliferations. It would also explain why B-CLL may precede, follow or present concurrently with cutaneous T cell lymphoma and the increased familial risk of lymphomas in family members of patients with cutaneous T cell lymphoma[38]. The increased risk of non-hematologic cancers in patients with cutaneous T cell lymphoma could be explained by the immunosuppression related to the disease and/or use of oncogenic treatments[27-30,33,34,39].

Alternatively, the interaction between stimulatory ligands such as CD30-CD30L and CD40-CD40L expressed by T and B cells may provide an explanation for the co-existence of T and B cell lymphoproliferative diseases in susceptible patients. For example, the interaction between CD40L, which is expressed by neoplastic T cells of MF[40], and CD40, which is constitutively expressed by B cells, could result in up-regulation of genes involved in B cell survival and proliferation[41,42]. The frequent expression of CD30 in some of our papular MF cases (and most LyP variants) with possible increased levels of soluble CD30 in the blood that we have observed in typical early MF patients could in theory contribute to the risk of developing B-CLL[43-45].

We conclude that MF may rarely present with persistent papules, but that there is considerable clinical and histo-immunopathologic overlap with LyP including a favorable prognosis[12,13]. Indeed the main difference is the persistence of lesions in papular MF and spontaneous regression of lesions in LyP. Considering that typical MF lesions may undergo partial or even complete regression[46], we wonder if the differences between papular MF and LyP may be related to differences in factors that mediate lesion regression such as the host immune response. In addition, in this small series, there appears to be an association of papular MF with B-cell CLL that requires confirmation and further investigation.

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relative risk of secondary B-CLL in patients with MF.

COMMENTS

Case characteristics

Mycosis fungoides (MF), a great masquerader of other skin diseases, can present with varied types of lesions that are confused with infectious and drug related eruptions among others.

Clinical diagnosis

Lymphomatoid papulosis (LyP) was excluded by the absence of spontaneous regression of lesions.

Differential diagnosis

The registry of patients with cutaneous T cell lymphoma (1481 patients diagnosed with MF excluding its erythrodermic variant) that is maintained by one of the authors with approval of the Institutional Review Board at Johns Hopkins University was reviewed for cases that fulfilled the clinical-pathological criteria for papular MF as defined by Kodama.

Imaging diagnosis

This retrospective review identified 6 patients who presented with persistent papules and/or small nodules with histopathologic features interpreted as diagnostic or consistent with MF

Experiences and lessons

Papular MF is a very rare presentation of the disease, occurring in 0.4% of non-erythrodermic MF cases referred to their center.

Peer-review

This is an interesting case series of papular mycosis fungoides. The authors described the clinical and histological features of this clinical entity, and its association with chronic lymphocytic leukemia. In general, the manuscript is well-written, and the content is clinically relevant and scientifically informative.

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