

# Mycosis fungoides beyond the classical Alibert-Bazin disease: report of seventeen atypical variants

## “Aarón Kaminsky Award 2007”

Alejandra Abeldaño<sup>1</sup>, Mariana Arias<sup>2</sup>, Karina Ochoa<sup>3</sup>, Adriana Benedetti<sup>3</sup>, María Cristina Kien<sup>4</sup>, Graciela Pellerano<sup>5</sup>, Edgardo Chouela<sup>5</sup>

## Abstract

**Introduction.** Mycosis fungoides (MF) is the most common subtype of cutaneous T-cell lymphomas (CTCL). Clinical manifestations in the classic form of the disease are multiple macules and plaques that may develop into tumors. Many cases of this CTCL differ substantially from this presentation and therefore are referred to as atypical or unusual variants, most of them resembling other dermatological diseases.

**Objective.** To evaluate the relative frequency of rare variants of MF and to estimate of clinical course of these variants in relation to stage at the time of diagnosis, prognosis, and response to treatment.

**Design.** Observational, retrospective, and descriptive.

**Material and methods.** Review of database of patients with primary cutaneous lymphomas (PCL) between November 1, 1995, and August 1, 2007. Included were all patients with clinical, histopathological, and immunophenotypic diagnosis comprised in the categories of the WHO/EORTC classification. Records included age, sex, date of definite diagnosis, treatment and clinical course.

**Results:** We analyzed 93 patients with PCL, 89 of whom were CTCL. Of these, 68 (73.12 percent) were MF (40 males and 28 females). Mean age at diagnosis was 53.9 years (range: 13-78 years). 17/68 (25 percent) were rare variants (11 males and 6 females) with a mean age of 51 years (range: 13-78 years). The average follow-up period after diagnosis of the rare variants was 21.94 months (range: 4-102 months). These cases included 5 folliculotropic MF, 4 erythrodermic MF, 3 poikilodermic MF, 2 hypopigmented MF, 1 granulomatous slack skin, 1 ichthyosiform MF and 1 unilesional MF. Most of the patients received a combined treatment regimen with IFN and PUVA, with 53 percent complete remissions. We observed one case of folliculotropic MF with progressive disease and visceral involvement.

**Conclusions.** Recognition of these atypical variants of MF is important, due to their similarity to other inflammatory dermatoses; therefore, correct diagnosis poses a true challenge. Misdiagnosis in the early stages will delay correct diagnosis and prompt treatment, which are directly related to disease prognosis (Dermatol Argent 2008;14(2):124-133).

**Key words:** *mycosis fungoides, atypical variants.*

**Reception date:** 8/1/2008 | **Approval date:** 8/2/2008

1. Head of Photo-Medicine Section. Ascribed Teacher of Dermatology, School of Medicine, UBA.
2. Head of Residents.
3. Dermatologist.
4. Dermopathologist. Head of Dermopathology Section.
5. Head of Dermopathology Unit.
6. Named Professor of Dermatology, UBA.

Hospital General de Agudos “Dr. Cosme Argerich”. Center of Dermatological Research. Autonomous City of Buenos Aires. Argentine Republic.

## Correspondence

Alejandra Abeldaño: Almirante Brown 280 – Autonomous City of Buenos Aires – Argentine Republic  
Phone: 4121-0856. E-mail: [dermatargerich@gmail.com](mailto:dermatargerich@gmail.com)

## Introduction

Cutaneous T-cell lymphoma (CTCL) is a monoclonal expansion of helper T-lymphocytes expressing strong epidermal affinity with early involvement restricted to skin. When the disease progresses, helper T-lymphocyte subclones with less affinity with epidermis, with greater tendency to disseminate into non-adjacent skin, lymph nodes, peripheral blood, and viscera are found.

Mycosis fungoides (MF) is the most frequent CTCL. In a series of 91 patients studied, they accounted for 72.52 percent,<sup>1</sup> a somewhat higher prevalence than the 44 percent found in the WHO/EORTC study on 1905 patients.<sup>2</sup>

The first case of MF was described by Alibert in 1806. In 1832, it received the name by which it is currently known, given the morphological feature of fungus-like tumors. In 1876, Bazin, a disciple of Alibert, divided the clinical progression into the stages of patch, plaque, and tumor.<sup>3</sup> This classical Alibert-Bazin MF affects more frequently males of average age between 50

and 60 years, and appears clinically with non-infiltrated erythematous-squamous lesions (macular stage), and/or infiltrated lesions with sharply demarcated borders leaving areas of healthy skin (plaque stage), which may develop into tumors tending to ulcerate. Classically, these lesions are seen more frequently on the trunk, on non-sun-exposed areas. However, the face is one preferred location in tumor growth.

This disease may have different aspects in its clinical and histopathological presentation; a series of variants are summarized in Table 1. It should be noted that in the new consensual classification by WHO and EORTC<sup>2</sup> the only recognized MF variants are folliculotropic, pagetoid reticulosis, and granulomatous slack skin (Table 2).

### Objective

- To determine prevalence of rare MF variants in the population of patients with CTCL.
- To assess the behavior of these variants in relation to stage at the time of diagnosis, progression, and treatment response

### Design

Observational, retrospective, and descriptive study.

### Materials and methods

Review of database on primary cutaneous lymphomas (PCL) from November 1, 1995 to August 1, 2007. It included all patients with clinical, histopathological, and immunohistochemical diagnosis complying with the classification categories of WHO/EORTC. Reported are age, gender, date of definite diagnosis, type of treatment, and clinical course.

### Results

The study included 93 patients with PCL, whereof 89 had CTCL. Of these, 68 (73.12 %) were affected by MF (40 males and 28 females), with mean age 53.9 years (range: 13 a 78 years). Seventeen/68 (25 %) were rare forms (11 males and 6 females), with a mean age of 51 years (range: 13 to 78 years). The average follow-up period after diagnosis of the rare variant was 21.94 months (range: 4-102 months). Distribution of these variants was 5 folliculotropic MF, 4 erythrodermic MF, 3 poikilodermic MF, 2 hypopigmented MF, 1 granulomatous slack skin, 1 ichthyosiform MF, and 1 unilesional MF.

The stage at the time of diagnosis was early (Ia-IIa) in 8 (47 %) and late (IIb-IV) in 9 (53 %) patients. Most fulfilled a combined treatment regimen with IFN + PUVA. Complete resolution (absence of signs and symptoms for at least 12 weeks) occurred in 53 % of the cases. Progressive disease with visceral involvement appeared in one case of folliculotropic MF, as well as node involvement by lymphoma in one patient with erythrodermic MF.

**TABLE 1.** CLINICAL AND HISTOPATHOLOGICAL VARIANTS OF MYCOSIS FUNGOIDES (MF).

• Bullous MF
• Dyshidrotic MF
• Erythrodermic MF
• Follicular mucinous MF
• Follicular or folliculotropic MF
• Syringotropic MF
• Granulomatous slack skin
• MF palmaris et plantaris
• Pagetoid reticulosis
• Poikilodermic MF
• Pustular MF
• Verrucous/hyperkeratotic MF
• Acanthosis nigricans-like MF
• Hypopigmented MF
• Hyperpigmented MF
• Ichthyosiform MF
• MF with eruptive epidermoid cysts
• Perioral dermatitis-like MF
• Pigmented purpura-like MF
• Zosteriform MF
• Angiocentric MF
• Granulomatous MF
• Invisible MF

**TABLE 2.** CTCL WHO/EORTC CLASSIFICATION

<b>Mycosis fungoides (MF)</b>
<b>Variants or subtypes of MF</b>
• Folliculotropic MF
• Pagetoid reticulosis
• Granulomatous slack skin
<b>Sézary syndrome (SS)</b>
<b>Primary cutaneous lymphoproliferative disorders CD30+</b>
• Primary cutaneous anaplastic large T-cell lymphoma
• Lymphomatous papulosis
<b>Subcutaneous panniculitis-like T-cell lymphoma</b>
<b>Extranodal NK/T cell lymphoma, nasal type</b>
<b>Not otherwise specified primary cutaneous peripheral T-cell lymphoma</b>
• Aggressive epidermotropic CD8+ T-cell lymphoma (provisional)
• Gamma/delta T-cell lymphoma (provisional)
• Pleomorphic small/medium sized T-cell lymphoma (provisional)
<b>Adult T-cell lymphoma/leukemia</b>

### Case history

Our case history is listed in Table 3.

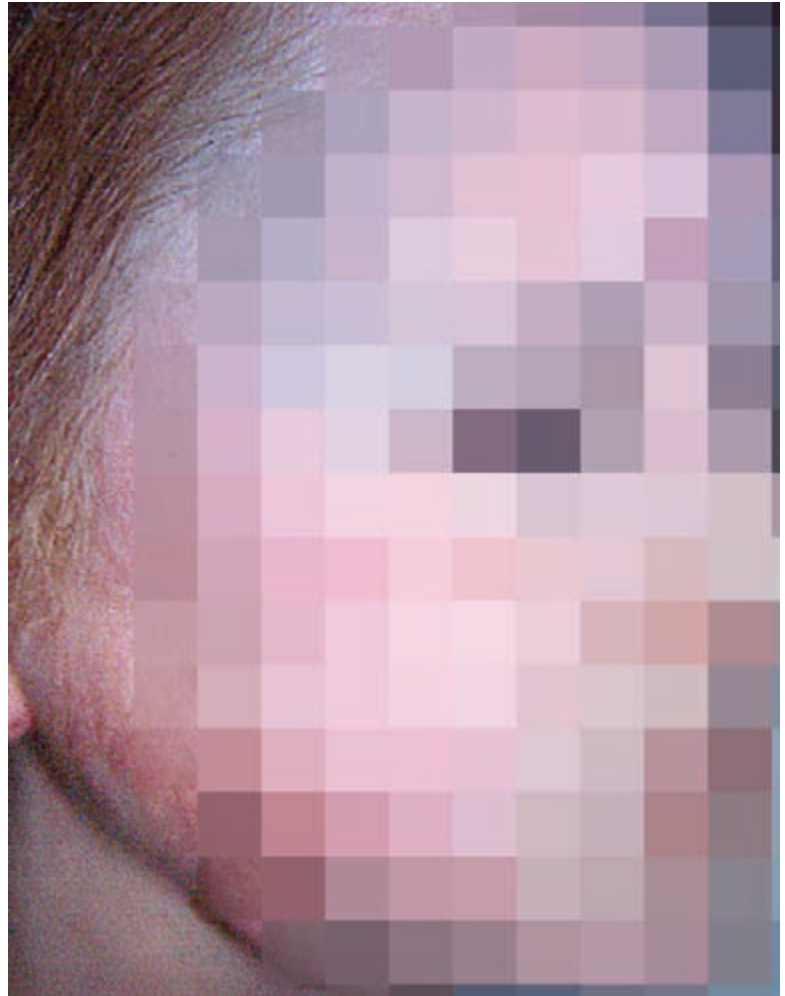
### Discussion

The described MF variants illustrate different presentation forms of this primary cutaneous lymphoma. In the studied population, mean age and gender distribution were similar to the observations in the whole group of MF patients. Generally, patients show classical MF clinical features (MFC) in other parts of the body, and this favors diagnosis, as in Cases 4, 6, 7, 8, 13, and 15. On other occasions, the disease begins with an atypical presentation that makes initial diagnosis difficult; thus, diagnosis is reached at a later stage, thus complicating prognosis and

survival, as in Cases 2, 3, 5, and 9. A recent study published by Barengo et al.,<sup>4</sup> on 7 patients with atypical variants of the disease, concluded on the utmost importance of reaching diagnosis at early stages, and this requires clinical awareness.

The **folliculotropic variant** may appear with a very proteiform clinical picture including patchy alopecia, epidermal cysts, comedones, follicular keratosis-type follicular plugs, and mucinorrhea when associated with follicular mucinosis.<sup>5</sup> More typical locations include face, neck, and upper trunk (**Figures 1 and 2**). Patients, males in a 4-5:1 ratio, may or may not have MFc lesions. Our series showed similar behavior, with dominance of male cases in a 3:2 ratio. Strong folliculotropism is noticeable in the histopathology (**Figure 3**), with scarce or none epidermotropism, and with or without follicular mucinosis.<sup>5-7</sup> The immunophenotype analysis shows atypical CD4+ lymphocytes with a phenotype similar to MFc. Folliculotropism may be explained by an increase of ICAM-1 expression in follicular epithelium, associated with LFA-1 lymphocytes, concurrently with reduction of ICAM-1 expression in epidermal keratinocytes.<sup>8</sup> Although the small number of reported cases makes a prognosis assessment difficult, folliculotropic mycosis fungoides seems to have a prognosis similar to MFc. Some authors suggest that, because of the depth reached by the infiltrate in its intra- and perifollicular arrangement, folliculotropic mycosis fungoides should be staged as T3 (tumor), independently of its clinical appearance; this suggests a more aggressive prognosis towards tumor and erythroderma.<sup>8</sup> In the analyzed cases, we see how Case 1 evolved as MFc for its stage, and Case 2 had a slower growth, with frequent relapses, and refractory to treatments. In addition, it should be noted that the disease was underdiagnosed for several years, and at the time of certainty diagnosis, the patient had reached tumor stage. The extension, initial stage, and treatment resistance resulted in an unfavorable prognosis for this patient, who progressed toward visceral involvement of the disease.

The consensus of the International Society for Cutaneous Lymphomas (ISCL) clearly defines the **erythrodermic variants** (E-CTCL). Here, erythrodermic MF differs from Sézary syndrome and non-specified E-CTCL because usually there is previous history of macular or plaque MF, and minimal or none peripheral blood involvement appears in staging (5 Sézary cells/100 lymphocytes plus PCR, or 20 Sézary cells/100 lymphocytes).<sup>9</sup> As every erythroder-



**Figure 1.** Case 2 (folliculotropic mycosis fungoides). Infiltrated erythematous tumor lesions in glabella and malar area.



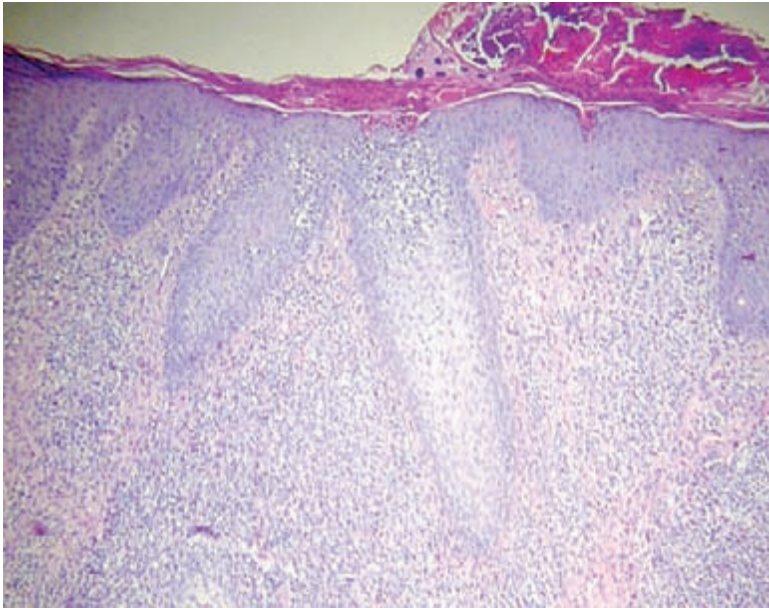
**Figure 2.** Case 2 (folliculotropic mycosis fungoides). Multiple follicular hyperkeratotic papules in trunk.

mic clinical picture, prognosis is serious. Frequently there are adenopathies, given the extension of skin involvement and itching, although not always with categorical histopathological confirmation, a situation we only found in Case 9, even though patients 6 and 7 also had lymphadenopathies. While

TABLE 3. CLINICAL DATA OF PATIENTS INCLUDED IN THIS STUDY

Patient Nº / Initials	Mycosis fungoides variant	Age Gender	Previous or co-existing MF	Evolution time to definite diagnosis of atypical MF/Previous diagnosis	Physical examination	Initial staging	Treatments/evolution	Follow-up after diagnosis of atypical MF
1 AC	Folliculotropic	51 M	No	12 months Alopecia areata	2 erythematous-squamous alopecic plaques in occipital region and vertex	Ia	4000 cGy electron beam. IFN- $\alpha$ 2b 3 mil. IU 5/wk for 19 mos. Complete response (CR). No relapses	102 mos.
2 RM	Folliculotropic	57 F	No	60 mos. Rosacea	Hyperkeratotic follicular papules on trunk and limbs. On face, infiltrated erythematous tumor lesions. Moderate itching	IIb	IFN- $\alpha$ 2b 3 mil. IU 3/wk + PUVA. Continued w/ IFN completing 18 mos. CR. Relapse: total body electron beam (EB). Relapse w/ node involvement and nodular images at lung level (IVb): poly-chemotherapy (CHOP). Diagnosis of cervical invasive carcinoma requiring radiotherapy (RT)	51 mos.
3 VB	Folliculotropic	30 F	No	72 mos. Follicular mucinosis. Jessner's lymphocytic infiltration	Infiltrated scaling erythematous plaques with internal tumor lesions on face (pre-serving center-facial area). On back, follicular keratotic erythematous papules forming plaques	IIb	Focused RT and IFN- $\alpha$ 2b 3 mil. IU 3/wk	4 mos.
4 FC	Folliculotropic	50 M	Yes (IIb stage) Previous	24 mos. after diagnosis of classic MF adds follicular lesions on trunk	Follicular erythematous papules on trunk. Multiple tan erythematous lesions involving over 10% of body surface. Right axillary lymphadenopathies	IIb	PUVA + IFN w/ partial response (PR). PUVA-therapy and bexarotene 200 mg/m <sup>2</sup> /day with PR. PUVA suspended, increases bexarotene dose to 300 mg/m <sup>2</sup> /day, and add intralesional IFN 3/wk, with good response	19 mos.
5 IC	Folliculotropic	53 M	No	Psoriasiform eczema since adolescence	Infiltrated facies. Erythematous-scaling isolated plaques on trunk and limbs	IIb	RePUVA with PR, then continues w/ IFN- $\alpha$ 2b 3 mil. IU 3/wk CR	14 mos.
6 FQ	Erythrodermic	70 M	Yes (IIa stage) Previous	10-year-evolution, non-specific chronic dermatosis, and then diagnosis of classic MF. Evolves towards erythroderma w/ lymphadenopathies	Generalized erythema and scaling. Neck and armpit lymphadenopathies	III	PUVA and IFN 3 mil. IU 5/wk for 18 mos. CR.	4 mos.
7 TA	Erythrodermic	50 M	Yes (Ib stage) Previous	Onset w/ small plaque parapsoriasis evolving to classic MF. 14 mos after MF diagnosis evolves towards erythroderma	Erythema and diffuse scaling in trunk and 4 limbs (upper limb dominance). Negative lymphadenopathies	IIIO	PUVA-therapy	4 mos.

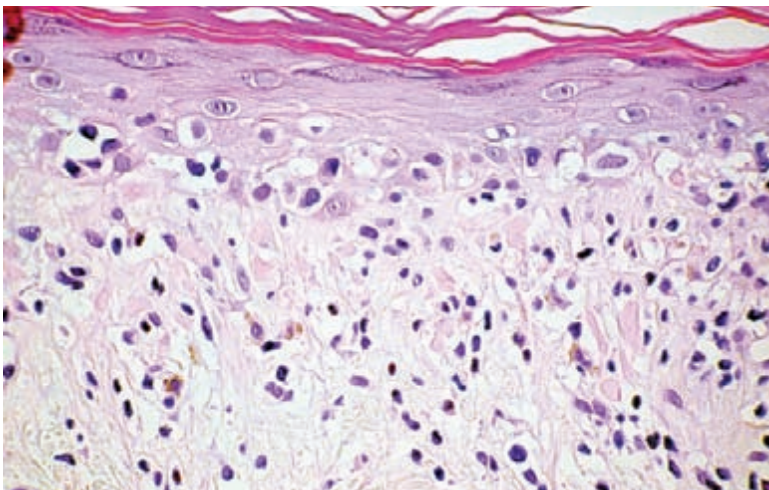
Patient N° / Initials	Mycosis fungoides variant	Age Gender	Previous or co-existing MF	Evolution time to definite diagnosis of atypical MF/Previous diagnosis	Physical examination	Initial staging	Treatments/evolution	Follow-up after diagnosis of atypical MF
8 VC	Erythrodermic	78 M	Yes Previous	81 mos. after diagnosis of classic MF, begins w/generalized erythema and scaling	Erythema and diffuse scaling on trunk and 4 limbs. No lymphadenopathies	III	Loss of follow-up prior to onset of therapy	6 mos.
9 JM	Erythrodermic	54 M	Yes Previous	Begins with pruriginous abdominal lesions w/ non-specific HP progressing to involvement of all tegument associated w/ groin lymphadenopathies	Involvement of all tegument w/ scaling and erythematous to violaceous skin color. Lichenification areas. Alopecia. Onychodystrophy, Beau lines. Groin lymphadenopathies	IVa	PUVA-therapy + systemic GCs, w/o response. Poly-chemotherapy (CHOP) w/o response. RePUVA w/o response. Electron beam w/ PR. Loss of follow-up while doing re-staging studies	52 mos.
10 JS	Vascular atrophic poikilodermic	49 M	No	48 mos.	Scaling salmon-colored patches on buttocks. Poikilodermiform patches on left groin, left arm, and low back.	Ib	Treatment with IFN- $\alpha$ 2b and PUVA therapy is suggested. Follow-up is lost within the year of reaching diagnosis	4 mos.
11 MV	Vascular atrophic poikilodermic	60 F	No	15 days	Eroded erythematous-squamous plaque on right breast	Ia	Heliotherapy and topical corticosteroids. Due to the impossibility of heliotherapy, thalidomide 100 mg/day is indicated. CR.	19 mos.
12 DC	Vascular atrophic poikilodermic	31 M	No	36 mos.	Erythematous-squamous macules on both buttocks	Ib	Heliotherapy and topical corticosteroids. CR	8 mos.
13 GC	Hypopigmented	65 M	Yes (Ib stage)	Develops hypopigmented lesions 24 mos. after diagnosis of MFC	Hyper- and hypopigmented lesions over skin not previously involved in trunk and face	IIb	RT on tumor lesions. IFN- $\alpha$ 2b 3 mil. IU SC 3/wk + PUVA. PR. Stationary	24 mos.
14 CB	Hypopigmented	13 F	No	12 mos.	2 hypopigmented macules, left submammary, and suprapubic areas	Ia	Heliotherapy and topical corticoids, CR. No relapses.	22 mos.
15 JA	Granulomatous slack skin	45 F	Yes (Ib stage) Previous	Begins with lesions in armpit and right groin, 48 mos. after diagnosis of classic MF	Erythematous-squamous plaques coincident with hanging folds of slack skin located in right axilla and groin	Ib	PUVA, IFN, acitretine, EB with PR. Surgical removal of slack skin. Relapse and new lesions in abdomen: recently begins with bexarotene p.o. 200 200 mg/m <sup>2</sup> /day. PR. Multiple relapses.	70 mos.
16 AS	Ichthyosiform	67 M	No	48 mos. Ichthyosiform dermatitis. Poikiloderma with strong lichenoid reaction	Fine generalized scaling with adherent tan scales in lower limbs. Multiple erythematous-squamous plaques in upper and lower limbs, abdomen, and lower back area.	Ib	IFN- $\alpha$ 2b 3 mil. IU 3/wk + PUVA. CR.	13 mos.
17 CG	Unilesional	44 F	No	12 mos.	4 erythematous-scaling plaques with diffuse borders in rib area and right breast	Ia	Heliotherapy and topical corticoids. PR. Rotation to IFN- $\alpha$ 2b + PUVA therapy, with CR.	13 mos.



**Figure 3.** Case 3 (folliculotropic mycosis fungoides). Atypical lymphocyte infiltrate with marked folliculotropism.



**Figure 4.** Case 10 (poikilodermic mycosis fungoides). Scaling annular salmon-colored patch with superficial atrophy.



**Figure 5.** Case 12 (poikilodermic mycosis fungoides). Epidermal atrophy, Vacuolization of basal layer with individualized large lymphocyte infiltration.

the disease is limited to a B1, non-aggressive treatment (PUVA, IFN, methotrexate, retinoids, in sequence or concomitant) is advisable. Case 6 benefited from this therapeutic option with minimum side effects, reaching complete resolution verified up to 4 months of follow-up. Case 9 appeared refractory to multiple treatments, with further specific node involvement. In the remaining cases, we could not reach conclusions, because follow-up was short, and treatments insufficient.

**Poikilodermic MF** is characterized by hypo- and hyperpigmentation, xerosis, atrophy, and telangiectasias (**Figure 4**). Generally, it appears on preexisting macular lesion sites, in chronic rubbing areas associated with classical lesions. Occasionally, it may dominate or even be the only manifestation of the disease.<sup>8,10</sup> The histopathological study is similar to MFc, but includes epidermal atrophy with rete ridge flattening, mild to moderate basal layer vacuolar degeneration with loss of pigment, melanophages in dermis, and superficial vasodilation with red blood cells in vessels (**Figure 5**).

The **hypopigmented MF** may appear with hypopigmented patches or plaques (**Figure 6**) that may be mistaken for pityriasis versicolor, vitiligo, or achromic eczematides, but an appropriate histopathology may ascertain the presence of atypical lymphocytes with epidermotropism, together with pigment disorders. Most frequent in black individuals, it is very often seen in youngsters,<sup>11-13</sup> as in Case 14, only 13 years old. Generally, treatments succeed in repigmentation. Clinical course and prognosis are similar to MFc, and depend on the initial stage.<sup>14</sup> Case 13 developed pigment disorders in the MF course already in tumor stage. This conditioned the partial response to established treatments.

**Granulomatous slack skin (GSS)** is now a MF variant classified as such by WHO/EORTC.<sup>2</sup> It presents plaques and infiltrated tumors in folding sites such as groins and axillae, which progressively form hanging folds of slack skin (**Figure 7**). In this presentation, the substrate comprises a more or less epidermotropic lymphoid infiltrate of small to medium size cells, with generation of giant multinucleated cell granulomas with elastophagia, and lack of elastic fibers (**Figure 8**). It is important to differentiate GSS and granulomatous MF histopathologically, where the latter is characterized by histopathological presence of a granulomatous reaction clinically related to a papule or nodule, but that may not

have a clinical equivalent, and account for a finding from MFc, erythrodermic, follicular, hyperpigmented MF.<sup>8</sup> The immunohistochemical analysis usually shows CD3+, CD4+, and CD8-, and TCR clonal rearrangement may occur. As in our case, it is more frequent in females. Prognosis and clinical significance of the granulomatous reaction are uncertain. For some authors, granulomas may imply an aggressive course with rapid extracutaneous dissemination and death.<sup>8</sup> About one third of GSS cases report a subsequent diagnosis of Hodgkin's lymphoma.<sup>15</sup> Patient N°15 had a long progression of MF in early IB stage that in spite of the established treatments (IFN + PUVA, total body electron beam, retinoids) has not achieved complete resolution. Although the disease does not progress (no evidence of extracutaneous involvement), treatment resistance and appearance of GSS imply a reserved prognosis.

A rare variant, for the few reported cases, is **ichthyosiform MF**.<sup>16-19</sup> It is a specific expression of CTCL, not a secondary skin sign. It follows an acquired ichthyosis course, with keratotic or comedones-like follicular lesions. Even though it may be a general condition, the preferred location is on lower limbs, as in Case 16 (**Figure 9**). There is prominent itching, and consequently, excoriations.<sup>8</sup> Although paraneoplastic acquired ichthyosis has been described in Hodgkin and no Hodgkin lymphomas, as well as in MF and lymphomatoid papulosis patients, histopathology shows, in addition to epidermic changes associated with ichthyosis such as orthokeratosis and hypogranulosis, a bandlike epidermotropic infiltrate of small lymphocytes. A review of 7 cases by Hodak et al.,<sup>19</sup> shows male dominance with mean age of 50 years and an average of 5 years evolution time before diagnosis (as our case). In 5 of the 7 cases, 3 evidenced MF, and 2 had follicular MF. The immunohistochemical analysis showed CD3+, CD4+, and CD7- in 6; the last had CD3+, CD8+, CD7-. In addition, CPR was positive for TCR gamma cells in 3 of the 7 patients.

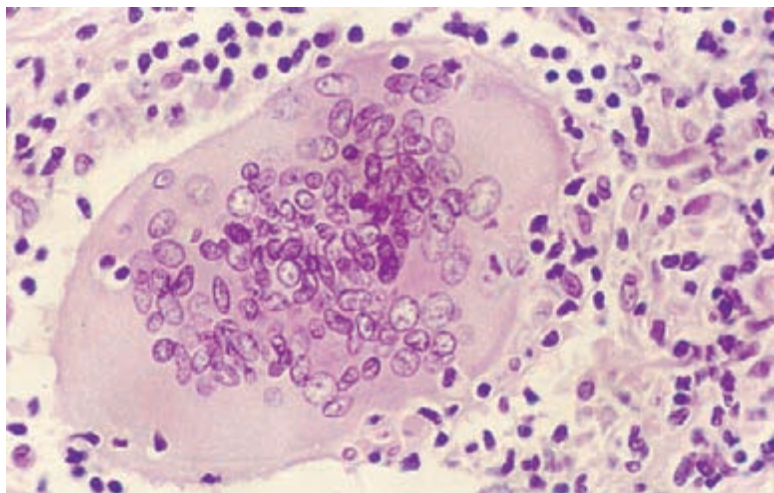
The **unilesional MF** is a rare variant that are expressed as single typical MF lesions, or lesions limited to an isolated individual area involving less than 5% of the body surface,<sup>8</sup> as Case 17 (**Figure 10**), whose few lesions were limited to an anatomic region. It may affect any age group, including children. The histopathological and immunophenotypic features cannot be differentiated from the classical forms, and it usually has a benign course.<sup>20</sup> It may be the reflection of an active antitumor response that minimizes malignant T-cell dissemination.<sup>4</sup>



**Figure 6.** Case 13 (hypopigmented mycosis fungoides). Hypopigmented lesions on skin previously not involved of the trunk.



**Figure 7.** Case 15 (granulomatous slack skin). Hanging fold of slack skin located in the axilla.



**Figure 8.** Case 15 (granulomatous slack skin). Detail of multinucleated giant cell in the infiltrate.



**Figure 10.** Case 17 (unilesional MF). Erythematous scaling plaques of diffuse borders on rib cage area and right breast.



**Figure 9.** Case 16 (ichthyosiform MF). Adherent tan scales and erythematous-squamous plaques on lower limbs.

## Conclusion

As we can see, the MF clinical spectrum is very extensive, and in about 25 percent of the cases, it may simulate or imitate different inflammatory dermatoses, according to the studied population. This diversity turns diagnosis of MF variants into a challenge for the dermatologist. The most frequent variants in our experience, which more often lead to wrong diagnosis, were folliculotropic mycosis fungoides; this group included the only case with disease progression and visceral involvement. Although histopathology is definite in most cases, final diagnosis should not be based on one single criterion, but be the result of an integral clinic, histopathology, and molecular biology concept; furthermore, in many cases, follow-up with periodically repeated studies and the natural evolution of the disease are determinant for certainty diagnosis. Understanding these variants and acquiring skills for early diagnosis have prognosis and therapeutic implications.

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