Clinics in Dermatology (2019) **37**, 255–267

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Clinics in Dermatology

Mycosis fungoides: A great imitator

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Abstract Mycosis fungoides (MF), the most common cutaneous T-cell lymphoma, typically presents in its early stage as inflammatory erythematous patches or plaques, with epidermotropism as the histopathologic hallmark of the disease. Over the past 30 years, numerous atypical types of MF, which deviate from the classic Alibert-Bazin presentation of the disease, have been described. These variants can simulate a wide variety of benign inflammatory skin disorders either clinically, both clinically and histopathologically, or mainly histopathologically. We have summarized the many faces of the disease, which set MF as a "great imitator," with special focus on the differential diagnosis and its benign mimickers.

Introduction

Mycosis fungoides (MF), the most common cutaneous T-cell lymphoma (CTCL) and often referred to by the latter term, is characterized by clonal proliferation of skin homing mature T cells, mostly CD4 positive, with special predilection for involving the epidermis.[1,2] Although the vast majority of the patients diagnosed are 50 years or older, children can also be affected by this lymphoma.[3–5] Patients with classic MF, as originally described by Jean Alibert (1768-1837) and Ernest Bazin (1807-1878), initially present with the early stage of the disease, characterized by persistent, progressive erythematous patches or thin plaques of variable size and shape, which have a scaly atrophic surface, located on sun-protected areas.[1,2] Patients may progress later into the advanced stage of the disease,[6] usually in the form of tumors.

The diagnosis of MF is based on the combination of clinical and histologic findings.[1,2,7] Epidermotropism, the histopathologic hallmark of MF, is characterized by the presence of T cells in the epidermis, not necessarily atypical, with only minimal spongiosis and is accompanied by superficial dermal lymphoid infiltrate[7,8]; however, histopathologic diagnosis of early MF is one of the most vexing and debated issues in dermatopathology, and not uncommonly the findings are subtle and focal. To facilitate the diagnosis of early MF, an algorithm has been proposed by the International Society for Cutaneous Lymphomas (ISCL), whereby clinical and histopathologic characteristics, as well as immunohistochemistry and T-cell receptor gene rearrangement studies, are combined in equivocal cases.[8]

It has long been recognized that the diagnostic pitfalls of early MF include some common benign inflammatory dermatoses. Over the past 30 years, the list of the differential diagnoses of MF has been considerably widened due to the description of numerous atypical types of MF, which deviate from the classic presentation. These variants can simulate a wide variety of inflammatory skin disorders either clinically alone, both clinically and histopathologically, or mainly histopathologically.[9–13]

The World Health Organization–European Organization for Research and Treatment of Cancer (WHO-EORTC) recognizes only three variants, or subtypes, of MF with different

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https://doi.org/10.1016/j.clindermatol.2019.01.004
0738-081X/

clinicopathologic features, clinical behavior, and prognosis compared with classic MF (folliculotropic MF, pagetoid reticulosis, and granulomatous slack skin).[1]

In addition, there are numerous variants that have clinical behavior similar to classic MF. These many unusual variants or types of MF are not unexpected, as in the early stage the number of malignant T cells is small, and the dermal infiltrate consists mainly of reactive T lymphocytes, which produce inflammatory cytokines that are partly responsible for the large variety of the histologic patterns. In fact, every one of the nine major patterns described by A. Bernard Ackerman (1936-2008) for the histologic diagnosis of inflammatory skin diseases was found in MF.[14]

In the following contribution, we shall summarize the wide clinicopathologic spectrum that set MF as a "great imitator," with special focus on the differential diagnosis (DD) and its benign mimickers detailed in Table 1.

Special diagnostic pearls

- Atypical MF is a diagnostic challenge, and there is a need for high index of suspicion and integration of all the clinicopathologic features with the ancillary studies for reaching the correct diagnosis. If feasible, in equivocal cases, the newly introduced molecular technique—high-throughput sequencing of the T cell receptor (TCR)—is warranted, given its higher sensitivity and specificity in discriminating MF from benign inflammatory diseases than the routine TCR-γ PCR analysis used in current clinical practice.[15]
- Meticulous whole-body examination is mandatory, not only to capture the accurate extent of involvement but also to detect the coexistence of any characteristic lesion of conventional MF as an important clinical clue for reaching the correct diagnosis.
- To enhance the likelihood of establishing a histologic diagnosis of early MF, topical corticosteroids should be discontinued at least 2 weeks before performing a biopsy, and multiple biopsies from a variety of lesions (preferably 6 mm in diameter), or alternatively one biopsy from the most developed lesion, should be obtained.
- Close observation and repeated periodic biopsies are needed in some cases.

Table 1 Differential diagnosis/mimickers of mycosis fungoides

Types of mycosis fungoides	Diagnosis differed
Folliculotropic	Alopecic lesions on the scalp: alopecia areata, trichotillomania, cicatricial alopecias Erythematous lesions on the scalp: seborrheic dermatitis, atopic dermatitis, psoriasis Follicular spiky papules: keratosis pilaris, lichen spinulosus, pityriasis rubra pilaris, lichen planopilaris Hairless patches/flat plaques: alopecia mucinosa Acneiform lesions: Favre-Racouchot syndrome, chloracne, follicular-comedogenic graft-versus-host disease, rosacea, adult onset acne
Hypopigmented	Vitiligo, tinea corporis, tinea versicolor, pityriasis alba, postinflammatory hypopigmentation, progressive macular hypomelanosis, idiopathic guttate hypomelanosis, sarcoidosis, leprosy
Hyperpigmented	Postinflammatory hyperpigmentation, fixed drug eruption, pigmented contact dermatitis, erythema dyschromicum perstans (ashy dermatosis), cutaneous amyloidosis, atrophoderma of Pasini and Pierini, idiopathic eruptive macular hyperpigmentation
Ichthyosiform	Sarcoidosis as a specific cutaneous finding of the disease Conditions that are associated with acquired ichthyosis as a secondary finding
Papular and pityriasis lichenoides chronica–like	Lymphomatoid papulosis type B, pityriasis lichenoides chronica, persistent arthropod bite reactions, lymphomatoid drug eruption
Palmaris et plantaris	Dermatophyte infection, psoriasis, eczematous dermatitides, secondary syphilis, hyperkeratotic lichen planus, verrucae, granuloma annulare
Psoriasiform	Psoriasis
Pagetoid reticulosis	Papulosquamous, eczematoid, infectious or neoplastic diseases
Unilesional	Papulosquamous, eczematoid, or dermatophytic diseases, Bowen disease
Poikilodermatous	Acquired poikiloderma such as a manifestation of collagen vascular disease, or radiation dermatitis
Chronic pigmented purpura–like	Chronic pigmented purpura
Figurate erythema–like	Erythema annulare centrifugum, erythema gyratum repens, Lyme disease, fungal infection
Verrucous	Inflammatory linear verrucous epidermal nevus, palmoplantar hyperkeratosis, verrucae vulgaris, keratosis lichenoides chronica, keratoacanthoma, seborrheic keratosis, porokeratosis of Mibelli
Bullous	Bacterial or viral infections, autoimmune bullous disorders, drug eruption
Pustular	Infections, pustular psoriasis, drug eruption
Interstitial	Interstitial granuloma annulare, inflammatory stage of localized scleroderma, interstitial granulomatous dermatitis
Granulomatous	Benign granulomatous skin diseases
Other	Atopic dermatitis, generalized nonspecific dermatitis

Variants of MF

Folliculotropic MF

Folliculotropic MF is the most common "atypical" clinicopathologic subtype of MF, comprising about 12% of all MF patients, according to some cohorts,[6] categorized as a distinct clinicopathologic variant by the current classification scheme of WHO-EORTC, and defined as the presence of follicle-based lesions and folliculotropism as the dominant histopathologic findings with or without follicular mucinosis.[1] It mostly affects adults but has also been reported in children and adolescents.[3,4]

Until recently, folliculotropic MF was generally considered a subtype with an aggressive biologic course and bad prognosis, comparable to tumor stage of classic MF[16–18]; however, folliculotropic MF has recently been described by the authors to present with two distinct patterns of clinicopathologic features with different prognostic implications, representing an early stage and tumor or advanced stage of this variant.[19] The clinical spectrum of early folliculotropic MF is broad and consists of follicle-based patch or flat plaques, keratosis pilaris–like lesions, or acneiform lesions, usually with alopecia, located mainly on the trunk and extremities, and histologically is characterized by intrafollicular and sparse or lichenoid perifollicular infiltrates of atypical lymphocytes that are confined to the adventitial perifollicular dermis.

Tumor folliculotropic MF preferentially involves the head and neck area and is characterized by indurated plaques or tumors, usually with alopecia, with heavier perifollicular infiltrates often extending to the reticular dermis.[19]

Using a very similar clinicopathologic approach and very similar histologic criteria, the Dutch group validated this new distinction between early and advanced stage folliculotropic MF.[20] Early folliculotropic MF is typified with a favorable prognosis, whereas in tumor folliculotropic MF, survival rates were found to be similar to those of classic tumor-stage MF.[19,20] In virtually all cases, the neoplastic cells in folliculotropic MF have a $CD3^+/CD4^+/CD8^-$ T-cell phenotype, as in classic MF.[16–19]

The clinical DD of folliculotropic MF depends on the clinical presentation[3,9,10,21,22]:

- Alopecic lesions on the scalp: alopecia areata, trichotillomania, and cicatricial alopecia
- Erythematous lesions on the scalp: seborrheic dermatitis, atopic dermatitis, or psoriasis capitis
- Follicular spiky papules: keratosis pilaris, lichen spinulosus, pityriasis rubra pilaris, and lichen planopilaris (Figure 1)
- Hairless patches/flat plaques: alopecia mucinosa, also known as idiopathic follicular mucinosis
- Acneiform lesions: Favre-Racouchot syndrome, chloracne, follicular-comedogenic graft-versus-host disease, rosacea, and adult onset acne

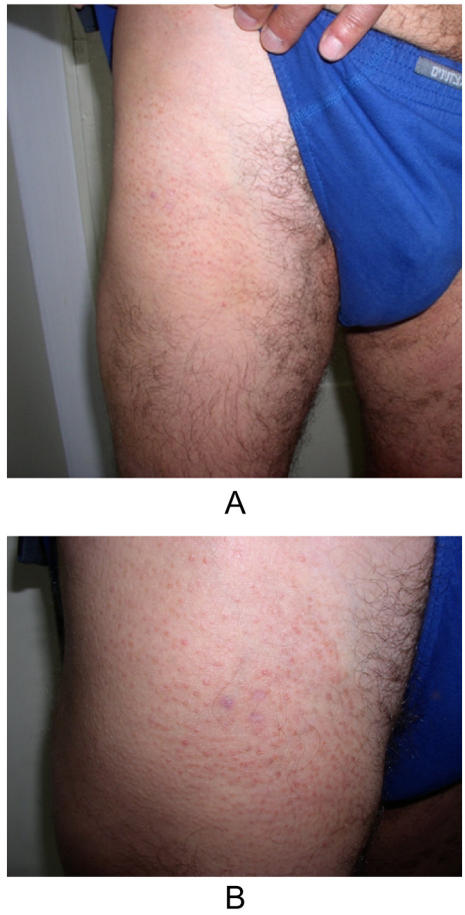

Fig. 1 **(A)** Early stage folliculotropic mycosis fungoides mimicking lichen spinulosus, pityriasis rubra pilaris, or lichen planopilaris, located on the thigh. Note the alopecia. **(B)** Close-up view showing spiky hyperkeratotic follicular papules.

In children, in whom folliculotropic MF most often presents with hypopigmented patches with follicular accentuation and often with alopecia,[3] the main DD is alopecia mucinosa, which may mimic folliculotropic MF clinically and histopathologically. Whether they truly represent two different entities or a disease spectrum is still a matter of debate.[23]

Clinicopathologic correlation is also required to differentiate folliculotropic MF from other types of CTCLs. In case the hair follicles have been destroyed or are completely obscured by diffuse dermal infiltrates, it may be difficult or even impossible to differentiate between folliculotropic MF and primary cutaneous peripheral T-cell lymphoma, unspecified.

Syringotropic MF

Syringotropic MF (also known as syringolymphoid hyperplasia with alopecia, syringotropic CTCL, or adnexotropic T-cell lymphoma) is a rare variant of the disease with many features that overlap with folliculotropic MF; thus, it is described as a subset of folliculotropic MF in the WHO-

EORTC classification.[1] Syringotropic MF presents with a solitary lesion or multiple erythematous papules, patches, and plaques with punctate follicular accentuation, often with alopecia overlying the lesions (63%-70%). Histopathologically, syringotropic MF is characterized by a prominent involvement of the eccrine glands, often associated with folliculotropism.

Whether it constitutes a separate entity or falls within the spectrum of folliculotropic MF is still a matter of debate.[24,25]

Hypopigmented MF

Hypopigmented MF predominantly affects people with darker complexion, including Asians but has been reported in Caucasians. According to one large cohort study, this variant accounts for 3.5% of all MF in adults,[6] whereas in the pediatric age group it is overrepresented and, in some series, even accounts for more than 50% of the cases.[3,26] Hypopigmented MF presents as round or irregular hypopigmented patches or flat plaques, some covered by fine scale, usually without atrophy. Subtle erythematous background can sometimes be observed. Lesions are typically asymptomatic or slightly pruritic and are mainly located on the trunk, buttocks, and extremities. On the arms, in contrast with classic MF, lesions show a predilection for the outer surfaces, rather than the inner sun-protected surfaces.[3,26–31] It may be the sole manifestation of MF or may occur in association with lesions of classic MF or other variants. In the pediatric age group, hypopigmented MF may be seen in combination with early folliculotropic lesions.[3] Histopathologically, lesions are indistinguishable from classic MF,[3,26–31] although striking epidermotropism of atypical lymphocytes,[26] as well as decreased melanin in the basal layer of the epidermis and melanin incontinence with melanophages in the dermis, have been observed in some cases.

In contrast with classic MF, it is not uncommon for the intraepidermal lymphocytes in HMF to exhibit a $CD8^+$ phenotype.[3,26,32]

This variant of MF has a close clinical resemblance to a wide range of dermatologic conditions presenting with hypopigmented macules or patches, including vitiligo, tinea versicolor, pityriasis alba, postinflammatory hypopigmentation, progressive macular hypomelanosis, idiopathic guttate hypomelanosis, sarcoidosis, and leprosy[3,11,26,31] (Figure 2).

The mechanism of loss of pigment in MF is still unclear, but it has been suggested that vitiligo and hypopigmented MF may share the same cytotoxic pathway of melanocyte destruction by $CD8^+$ T cells.[33] Immunohistochemic studies have demonstrated a reduction in the number of melanocytes and possible melanocyte dysfunction, as evidenced by the fewer tyrosinase, melan-A, CD117, and MiTF-positive melanocytes in the hypopigmented lesions versus normal skin and nonhypopigmented MF.[33,34]

Hyperpigmented MF

Hyperpigmented MF has been reported in only few case reports and series, and similar to hypopigmented MF, it is overrepresented in patients with pigmented skin.[35] It is characterized by hyperpigmented patches or plaques, some with ill-defined borders, with various degrees of skin atrophy and scaling. It may be the sole manifestation of MF, although some patients may have concomitant lesions of classic MF or other variants of MF.[35–37] Hyperpigmented MF may mimic a wide range of benign conditions:

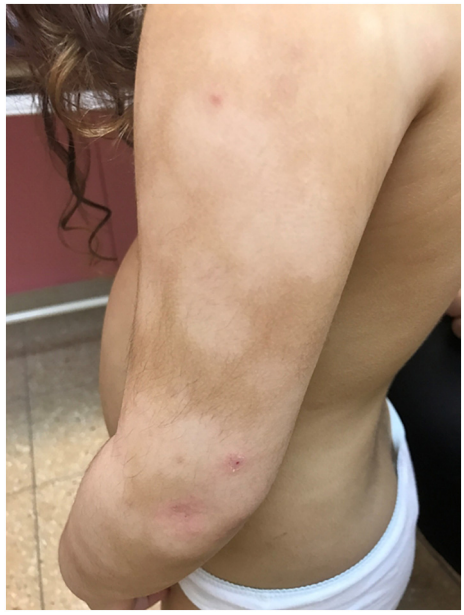

Fig. 2 Hypopigmented mycosis fungoides on the outer aspect of the arm in a light-skinned child masquerading as pityriasis alba.

- Postinflammatory hyperpigmentation
- Fixed drug eruption
- Pigmented contact dermatitis
- Erythema dyschromicum perstans (ashy dermatosis)
- Cutaneous amyloidosis
- Atrophoderma of Pasini and Pierini
- Idiopathic eruptive macular hyperpigmentation[35–39] (Figure 3).

Histopathologically, in addition to features of typical MF, diffuse vacuolar degeneration of basal keratinocytes mimicking "interface dermatitis," together with melanophages, has been found in the majority of cases. Immunohistochemically, most cases of hyperpigmented MF show a $CD8^+$ phenotype.[35] These cytotoxic T cells may affect neighboring melanocytes or basal keratinocytes, causing interface changes and marked melanin incontinence that results in hyperpigmentation.[35]

Ichthyosiform MF

Ichthyosiform eruption, as a specific manifestation of MF (ie, ichthyosiform MF), has been reported in only a few cases, although in one study this variant comprised 3.5% of all MF cases.[40] Ichthyosiform MF is typified by ichthyosis vulgaris–like eruption, or by less specific ichthyosiform

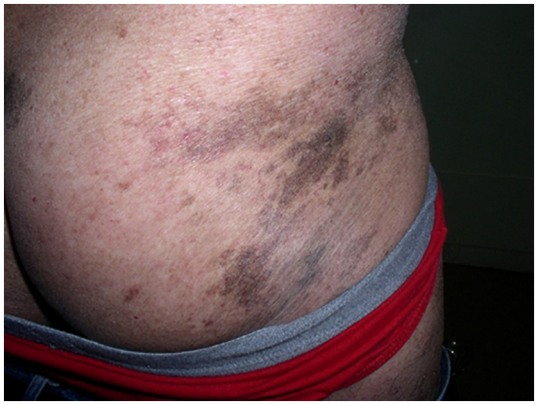

Fig. 3 Hyperpigmented mycosis fungoides: hyperpigmented patches of a dark-skinned patient, generating differential diagnosis with ashy dermatosis, pigmented contact dermatitis, and lichen planus pigmentosus.

lesions. Patients typically present with diffuse, dry, scaling skin; well-circumscribed scaly patches; or flat plaques affecting the trunk and extremities.[40–44] Histologically, in addition to the typical features of MF, findings suggestive of coexistent ichthyosis vulgaris, such as parakeratosis and focally compact orthokeratosis with thinning or absence of the granular layer[40,44] and diminished filaggrin expression in the thin granular layer, are noted.[45] Most cases of IMF have a $CD3^+$,$CD4^+$ phenotype, with only a few showing a predominance of $CD8^+$ lymphocytes.[40–44] The diagnosis of ichthyosiform MF can clinically be suspected in patients showing concomitant lesions of classic MF or other variants, usually folliculotropic MFlesions[3,40,41,44] (Figure 4). In cases with no other concomitant signs of MF, the DD includes a long list of conditions that are associated with acquired ichthyosis:

- Underlying malignancy (as a paraneoplastic eruption)
- Endocrinologic and autoimmune disorders
- Infectious diseases
- Nutritional disorders
- Renal impairment

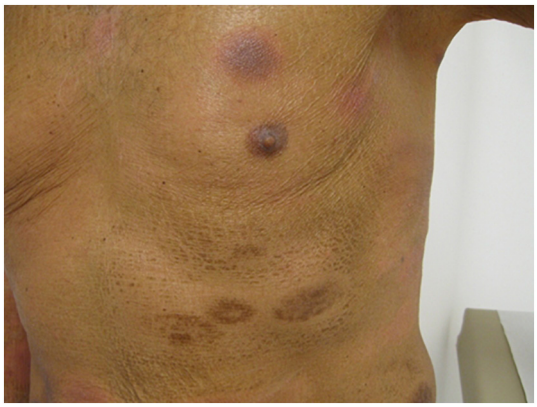

Fig. 4 Ichthyosiform mycosis fungoides adjacent to hyperpigmented and classic flat plaque type of mycosis fungoides.

- Drug eruption
- Sarcoidosis
- Other rare conditions[40]

Regardless of these underlying conditions, except for sarcoidosis in which a skin biopsy specimen may yield the noncaseating granulomas diagnostic for the disease,[46] the histopathologic changes are nonspecific, revealing no indication of the underlying condition.[40]

Papular and pityriasis lichenoide–like MF (PL-like MF)

The papular variant of MF was initially described in 2005 based on the following criteria:

- Papules with histopathologic findings of MF
- Absence of spontaneous regression of lesions
- No other evidence of MF or lymphomatoid drug reaction[47]

To date, less than 20 cases have been reported in the literature, with only a small fraction of them subsequently developing patches or plaques of classic MF. Lesions are either widespread or more localized, often symmetric, and without apparent predilection for sun-protected areas.[47–50]

The DD of papular MF includes mainly pityriasis lichenoides (PL), lymphomatois papulosis (LyP) type B, less commonly persistent arthropod bite reactions, and lymphomatoid drug eruption. In view of the overlapping clinical and histopathologic features of papular MF, LyP, and PL, some authors have suggested that they may represent a pathophysiologic continuum rather than three discrete entities.[47,51,52]

PL-like MF has been reported in a few adults and children. It is characterized by papules that resemble PL chronica and histopathologic findings consistent with MF, including a dermal infiltrate composed of lymphocytes that are $CD30^-$.[48,52–54]

MF palmaris et plantaris

Although specific involvement of the palms or soles can be seen in the course of classic MF in approximately 10% of patients,[55] MF that is limited predominantly to or initially presents on the palms or soles, a condition referred to as MF palmaris et plantaris, has been rarely reported.[55] Lesions are usually bilateral and present as erythematous hyperkeratotic patches and plaques with fissures and scales, with or without pruritus. Unusual clinical variations of MF palmaris et plantaris include:

- Annular
- Hyperpigmented
- Vesicular
- Dyshidrotic
- Pustular

- Verrucous
- Psoriasiform
- Ulcerative lesions
- Nail dystrophy[55–62]

The main DD of MF palmaris et plantaris includes dermatophyte infection or inflammatory skin diseases localized to the palms and soles, such as psoriasis and eczematous dermatitides[63] (Figure 5). The diagnosis of MF palmaris et plantaris should be suspected in long-standing cases of hand or foot dermatitis or "psoriasis" that are unresponsive to standard therapies or have atypical presentations.[29,58,59] Other possible differential diagnoses to be considered are secondary syphilis, hyperkeratotic lichen planus, verrucae, and granuloma annulare.[29,59]

On histology, the usual features of MF are found in MF palmaris et plantaris; however, in our experience, spongiosis may be more pronounced than in classic MF, making it difficult to differentiate MF palmaris et plantaris from a spongiotic dermatitis. Immunophenotyping and analysis of TCR clonality should be performed to confirm the diagnosis.[59]

In some cases, a definitive diagnosis cannot be reached, unless MF lesions in other areas of the skin are detected.

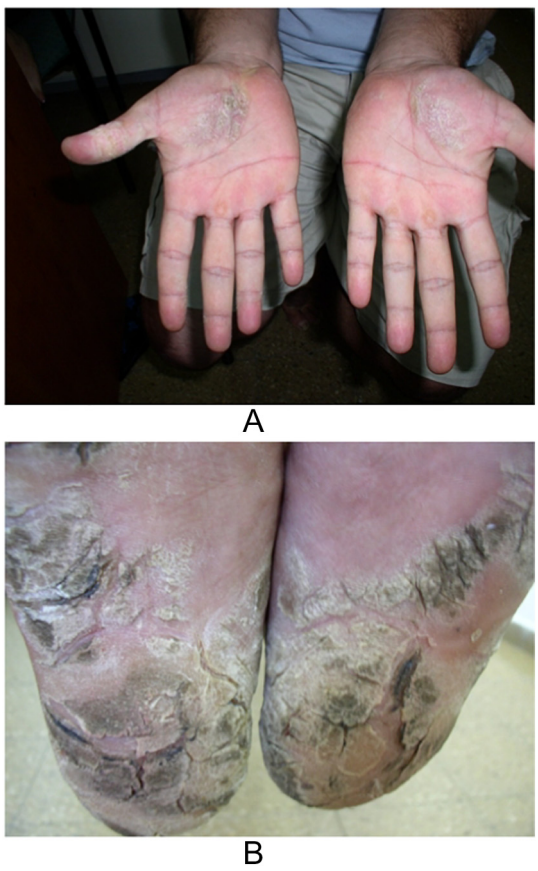

Fig. 5 Palmoplantar mycosis fungoides showing hyperkeratotic plaques with fissures, resembling severe palmoplantar psoriasis.

Psoriasiform MF

MF presenting with psoriasiform plaques (ie, psoriasiform MF) has been reported in only a few case reports and series in either adults[64–71] or children,[3] although psoriasis is not infrequently included in the initial DD of MF. Patients may be treated as having "psoriasis" for years before the correct diagnosis is rendered. Differentiating MF from psoriasis vulgaris is of importance, especially because systemic immunosuppressive treatments used for psoriasis, such as cyclosporine and tumor necrosis factor inhibitors, can worsen MF.[65,67] The possibility of coexistence of MF and psoriasis in the same patient should also be taken into account.

The clinical presentation of psoriasiform MF includes thick, scaly, well-demarcated, erythematous psoriasiform plaques, which are also typical of psoriasis; thus at times the DD between the two is challenging. Some patients may have additional alopecia and induration, which are clues for the diagnosis of MF. Other patients may have additional erosions and ulcerative lesions[66–71] (Figure 6). In atypical cases of psoriasis, a biopsy should be obtained, which may reveal psoriasiform hyperplastic epidermis and epidermotropic atypical lymphocytes, typical for MF. Other concomitant histopathologic findings reported in cases of psoriasiform MF are scant spongiosis and a lichenoid pattern.[66–71]

Pagetoid reticulosis

Pagetoid reticulosis, also called Woringer-Kolopp disease, is a variant of MF that presents with slowly growing, localized patches or plaques with a psoriasiform or hyperkeratotic appearance, usually located on the distal extremities.[1] Histopathologic study may reveal a psoriasiform and sometimes verrucous hyperplasia with marked pagetoid spread of highly epidermotropic, large atypical lymphocytes, singly or in nests, which occupy the entire thickness of the epidermis. The neoplastic T cells may show a $CD3^+/CD4^-/CD8^+$, or less commonly a $CD3^+/CD4^+/CD8^-$ or $CD3^+/CD4^-/CD8^-$, phenotype. Cases with a $CD8^+$ or $CD4^-/CD8^-$ phenotype express cytotoxic proteins. CD30 is often expressed.[72–74]

The clinical DD of pagetoid reticulosis may include:

- Chronic dermatitis
- Psoriasis
- Tuberculosis verrucose cutis
- Blastomycosis
- Verrucous squamous cell carcinoma

On histopathology, pagetoid reticulosis may mimic:

- Superficial spreading melanoma
- Pagetoid squamous cell carcinoma in situ
- Extramammary Paget disease

It may also histologically mimic other types of highly epidermotropic lymphoproliferative process, such as $CD30^+$

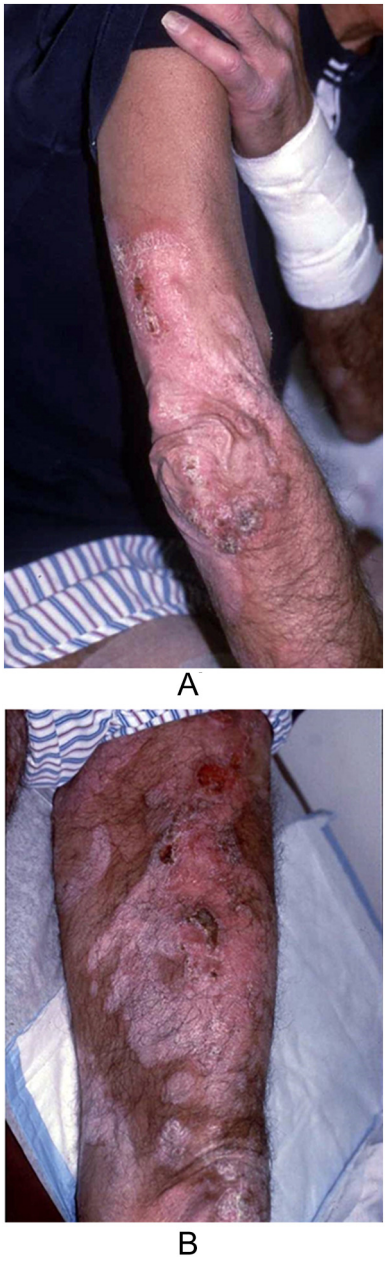

Fig. 6 Mycosis fungoides masquerading as psoriasis vulgaris: thick, scaly, well-demarcated erythematous plaques located on the external aspects of arm **(A)** and the leg **(B)**. An important clinical clue for MF is the presence of ulcers and crusts.

lymphoproliferative disorders, in particular LyP type D, and CD8-positive aggressive epidermotropic cytotoxic CTCL.[9] Useful criteria for pagetoid reticulosis include the characteristic clinical presentation and the often strictly epidermal localization of the neoplastic T cells.

Unilesional MF

Unilesional MF, although rare, is well documented in the literature. It is characterized by a solitary lesion that is clinicopathologically indistinguishable from patch and plaque classic MF,[75–77] although hypopigmented, eczematoid, psoriasiform, poikilodermatous, and folliculotropic solitary lesions have also been reported.[75–83]

In many patients, the lesion has been present for several years before the diagnosis has been confirmed. The main DD of unilesional MF involves:

- Papulosquamous or eczematous lesions
- Dermatophyte infection
- Bowen disease

Only pagetoid reticulosis, but not unilesional MF, is listed as an MF subtype in the 2005 WHO-EORTC classifications.[1] It remains to be seen whether the historic term pagetoid reticulosis will be included within unilesional MF in future classifications.

Poikilodermatous MF

Poikiloderma is the combination of cutaneous atrophy, telangiectasia, and macular pigmentary changes, which result in a mottled skin appearance.[84] Poikilodermatous MF, accounting for approximately 11% of all MF cases according to one study,[6] is usually a clinicopathologic variant of patch-stage MF, predominantly located on the breast, hips, and buttocks, and may present concomitantly with patches or plaques of classic MF or unusual variants.[85] It may mimic acquired poikiloderma, which can be related mainly to collagen vascular disease and radiation.

On histopathologic examination, poikilodermatous MF shows findings of classic MF, combined with changes of poikiloderma, such as epidermal atrophy, basal hydropic degeneration, pigment incontinence, and telangiectatic vessel formation.[86,87] Immunophenotypic analysis may reveal a $CD8^+$ phenotype.[88,89]

Pigmented purpuric dermatosis (PPD)–like MF

This rare clinicopathologic variant of MF, reported in both adults and children, manifests with persistent purpuric lesions and golden-brown discoloration simulating true PPD (Figure 7). Clinical clues to the diagnosis of PPD-like MF are the extensive distribution of lesions (in PPD, lesions are usually limited to the legs), the presence of reticular arrangement, and the coexistence of other lesions suspicious of large plaque parapsoriasis, classic MF, or of other unusual variants.[90–97]

Histopathologic examination, reveals features of MF, as well as extravasation of erythrocytes in the papillary dermis and the presence of siderophages typically seen in PPD. Immunohistochemic analysis performed in a minority of the published cases showed $CD4^+$, $CD8^+$, or $CD4^-$,$CD8^-$ double-negative phenotypes.[35,97,98]

Although MF-like histopathologic patterns can be observed in classic cases of PPD, the presence of intraepidermal

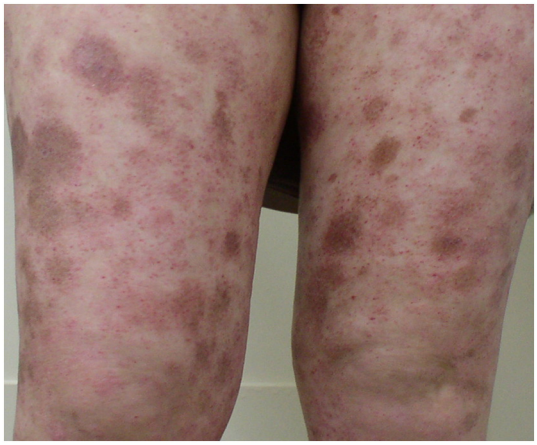

Fig. 7 Pigmented purpuric dermatosis–like mycosis fungoides: purpuric patches, some with golden-brown discoloration, located on the legs.

lymphocytes larger than those in the dermis along with atypical lymphocytes in the dermis and papillary dermal fibrosis favor the diagnosis of MF. The finding of a monoclonal TCR gene rearrangement can be helpful to confirm the diagnosis, although T-cell clonality may also be detected in otherwise classic cases of PPD.[96,99,100]

Figurate erythema–like MF

Erythema annulare centrifugum (EAC), and erythema gyratum repens (EGR)–like MF

Several case reports of MF have been described in the literature, exhibiting annular or polycyclic erythematous patches and/or plaques, which expand outward with or without central clearing, and trailing scale typical to true EAC. The DD of this atypical presentation of MF may include Lyme disease and superficial fungal infection (Figure 8). Unlike EAC, EAC-like MF does not show "coat-sleeve-like" perivascular lymphocytic infiltration, but rather a common typical histology of MF.[101–103]

EGR-like MF manifested by symmetrically distributed red patches with multiple concentric, almost targetoid, rings with trailing scale that radiated from the center was reported in one case report. Unlike EAC, there are no obvious central clearing, and the entire patch was composed of bandlike rings.[104]

Verrucous MF

Verrucous MF is an extremely rare variant; manifests as hyperkeratotic pruritic, warty plaques located on the legs, face, and trunk; and may or may not be accompanied by classic MF lesions.[105–107] Histopathologically, this type of MF is characterized by marked epidermal hyperplasia that may even resemble carcinoma.

The DD includes:

- Palmoplantar hyperkeratosis
- Verrucae vulgaris

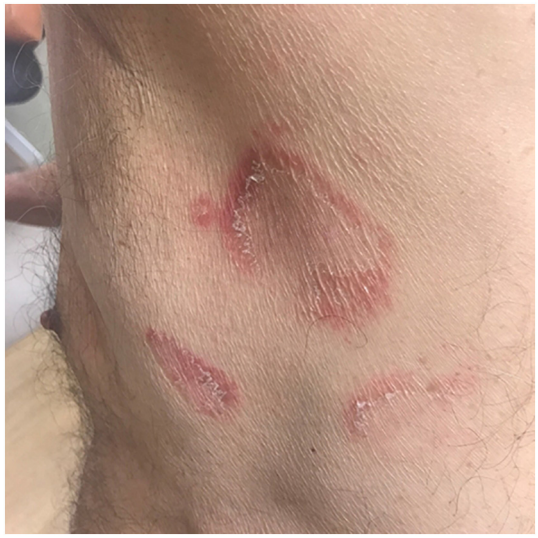

Fig. 8 Mycosis fungoides masquerading as erythema annulare centrifugum, showing annular erythematous flat plaques with a trailing scale at the inner border of the lesions.

- Keratosis lichenoides chronica
- Keratoacanthoma
- Seborrheic keratosis
- Porokeratosis of Mibelli

An unusual case of MF with clinicopathologic features mimicking inflammatory linear verrucous epidermal nevus has also been described.[108]

Acanthosis nigricans–like MF (vegetating or papillomatous MF)

A rare variant has been described in conjunction with classic MF lesions. This is a papillomatous variant with brownish pigmentation and a velvety appearance mimicking acanthosis nigricans, involving the antecubital folds, axillae, groins, popliteal fossae, intergluteal, and sometimes the nipple, areola, and peri-umbilical areas.[109–111]

Histologically, in addition to typical findings of MF, there are hyperkeratosis, papillomatosis, and acanthosis with interconnected rete ridges and horny pseudocysts with a seborrheic keratosis–like appearance.

Bullous MF

Vesiculobullous lesions are a rare manifestation of MF, usually appearing months to years after the onset of classic MF or Sézary syndrome.[112–116] Bullous MF presents with flaccid or tense, often multiple blisters appearing on lesional and normal-looking skin, sometimes with a tendency to form ulcers. The DD includes bacterial or viral infections, autoimmune bullous disorders, and drug eruptions. The coexistence of bullous pemphigoides or pemphigus foliaceous with MF

has been reported in a few cases.[112,117] Bulla formation can be an adverse effect of treatments for MF, such as topical mechlorethamine, systemic interferon, and psoralen and ultraviolet A.

Histopathology reveals subcorneal, intraepithelial, or subepidermal blisters, which may contain atypical T cells. The presence of typical features of MF (eg, epidermotropism with atypical lymphocytes) is the main clue to the diagnosis.[113–118]

Pustular MF

An extremely rare variant, initially described by Ackerman et al,[119] is marked by a pustular eruption that may be limited to the palmoplantar areas. Pustular MF can be regarded as an unusual manifestation of MF palmaris et plantaris. The DD may include infection, pustular psoriasis, and a drug reaction. Histologically, there are intraepidermal spaces filled with a mixture of atypical lymphocytes, neutrophil, and eosinophils. IL-8 has been suggested to play a role in the development of pustular lesions in MF.[57,119,120]

Interstitial MF

Interstitial MF usually presents as flat or slightly elevated plaques that, in contrast to classic MF, lack scaling and atrophy.[121–123] Histopathologically, it is characterized by dermal infiltrates of lymphocytes dissecting the collagen bundles, mimicking the pattern of inflammatory dermatoses such as interstitial granuloma annulare, inflammatory stage of localized scleroderma, and interstitial granulomatous dermatitis (Figure 9).

Epidermotropic lymphocytes are present at least focally in many, but not in all, cases, and bandlike dermal infiltrate may be missing, thus representing a diagnostic pitfall.

Some collagen fibers are surrounded by neoplastic lymphocytes, resembling the "rosetting" observed in interstitial granulomatous dermatitis. Immunohistology shows that most interstitial cells are T lymphocytes, in about half of the cases with a cytotoxic phenotype, with variable amounts of $CD68^+$ histiocytes that do not outnumber the lymphocytes, with occasional mucin deposition.[121–124]

Interstitial granuloma annulare and interstitial granulomatous dermatitis can be differentiated from interstitial MF by the predominance of histiocytes. Inflammatory morphea is differentiated from interstitial MF by the relative predominance of B cells and plasma cells in the interstitium and presence of sclerosis. The detection of T-cell clonality by polymerase chain reaction favors the diagnosis of MF; however, clonality has been detected, although rarely, also in granuloma annulare.[125] A rare coexistence of MF and interstitial granuloma annulare has been reported.[126]

Granulomatous slack skin

Granulomatous slack skin (GSS) is an extremely rare clinicopathologic subtype of MF.[1] It is characterized by the slow development of bulky, infiltrated, pendulous folds of atrophic skin in the axillae and groins reminiscent of cutis laxa.[127,128]

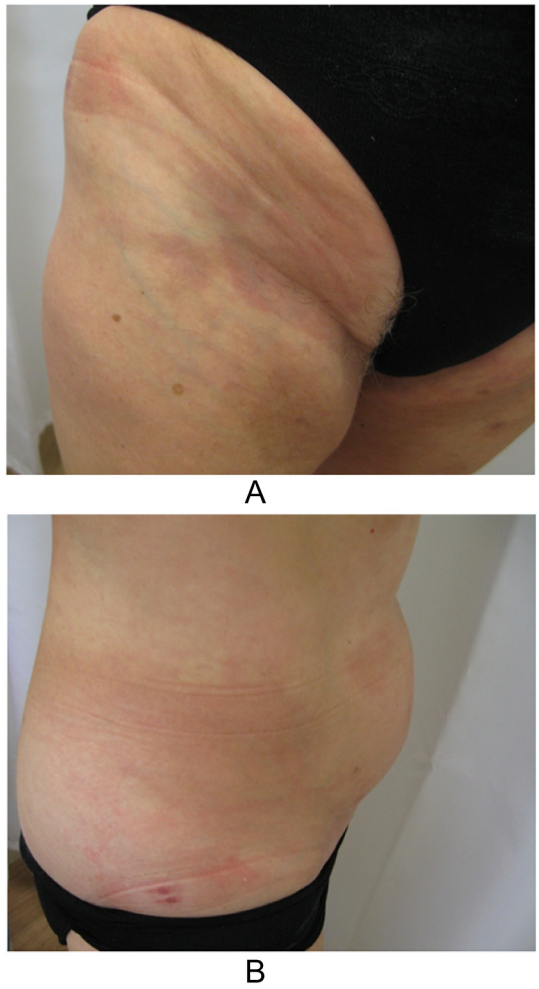

Fig. 9 Interstitial mycosis fungoides devoid of the scales or atrophy, which are characteristics of early stage conventional mycosis fungoides. The main differential diagnoses include macular or interstitial granuloma annulare, inflammatory stage of morphea, and interstitial granulomatous dermatitis.

Histopathologically, GSS is characterized by dense infiltrates throughout the entire dermis of small to medium-sized T cells admixed with numerous macrophages and many scattered multinucleated giant cells. Loss of elastic tissue, elastophagocytosis, and emperipolesis (engulfment of lymphocytes) by multinucleated cells are commonly observed. The epidermis may be infiltrated by small atypical T cells with cerebriform nuclei, as in classic MF. Most cases have a $CD3^+/CD4^+/CD8^-$ T-cell phenotype and show clonal TCR gene rearrangement.[1,11,127]

Granulomatous MF

Granulomatous MF is mainly a histopathologic variant that may be found either at the time of the initial diagnosis or years later in histologic sections from patients with otherwise classic

MF (including erythrodermic MF), as well as in unusual variants.[14,127,129,130] Treatment of MF with interferon or bexarotene has been associated with the development of a granulomatous reaction.[131,132]

In some patients, the clinical features may be suggestive of a granulomatous disease, as it may present as nonscaly, thick plaques or nodules, without the cutis laxa–like features typical of GSS. Granulomatous MF may mimic benign granulomatous skin diseases, such as granuloma annulare, sarcoidosis, or granulomatous rosacea.[10,133]

The EORTC pathologic criteria for granulomatous MF include prominent granuloma formation or numerous histiocytic giant cells or a histiocyte-rich infiltrate defined by histiocytes accounting for more than 25% of the entire infiltrate.[127] The histopathologic patterns of granulomatous MF may vary and include epithelioidm, sarcoidal, tuberculoid, periadnexal, granuloma annulare–like, palisaded, necrobiotic granuloma–like, and diffuse granulomatous infiltrate. Loss of elastic fibers is a common finding, but elastophagocytosis is rare. Immunohistochemically, most cases have a $CD4^+$ phenotype.[127,129]

The diagnosis of granulomatous MF may be difficult, particularly in cases in which the atypical lymphoid infiltrate is obscured by a predominant granulomatous component.[134] In some cases, epidermotropism, which is the major diagnostic clue for MF, may be absent, leading to a significant delay in the diagnosis and treatment.[127,129]

The detection of monoclonality can support the diagnosis, although a monoclonal T-cell clone can occasionally be found in nonneoplastic granulomatous disorders.[135]

Other unusual manifestations of MF

Although not regarded as a variant, cases with early stage or advanced MF, including erythrodermic MF and Sézary syndrome, misdiagnosed as atopic dermatitis or generalized nonspecific dermatitis are reported in series of MF and Sézary syndrome.[3,136–138]

Any atypical findings in a patient with seemingly atopic dermatitis, such as absence of pruritus (a mandatory criterion for the diagnosis of atopic dermatitis) and detection of infiltrated plaques or the presence of hypopigmented patches and plaques with follicular accentuation (neither of which are an expected feature of atopic dermatitis), are among the findings that should raise the suspicion of MF (Figure 10).

Pathologic criteria alone may be insufficient, as morphologic findings in early stage MF, in erythrodermic MF, as well as in Sézary syndrome, may show "nonspecific" findings.[7,64] In addition, spongiosis, which is usually observed in patients with dermatitis, can be seen in approximately 30% of MF cases.[14]

The combination of all data, including the personal and family history, the clinical manifestations and the clinical findings, the histopathology (with preferably several biopsies from different sites), and ancillary studies, are crucial in any patient with atypical atopic dermatitis. In patients with extensive nonspecific dermatitis, assessment of blood involvement (flow cytometry analysis) should also be considered.

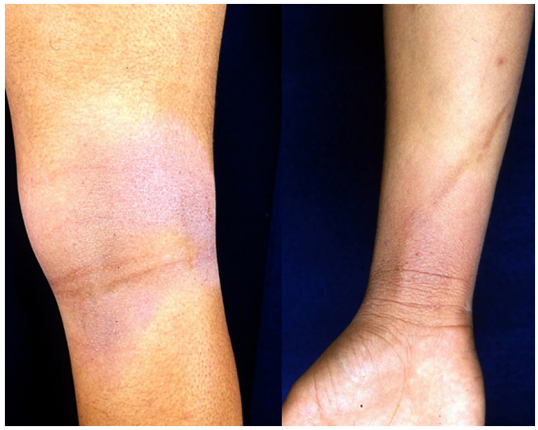

Fig. 10 A 16-year-old patient with mycosis fungoides resembling typical atopic dermatitis. The red flag was raised, because pruritus, a mandatory criterion for atopic dermatitis, was lacking.

Conclusions

Recognition of the many faces of MF is a major challenge for clinicians and pathologists. This most common cutaneous lymphoma, especially in its early stage, can mimic clinically and histopathologically a wide spectrum of common and rarer inflammatory skin conditions. High index of suspicion and awareness of all the clinical and histopathologic pitfalls, together with the correct integration of the ancillary studies, are mandatory to avoid undiagnosis of MF. Regular followup and repeated biopsies often help in arriving at a diagnosis in suspicious cases.

References

1. Willemze R, Jaffe ES, Burg G, et al. WHO-EORTC classification for cutaneous lymphomas. *Blood* 2005;105:3768-3785.
2. Olsen E, Vonderheid E, Pimpinelli N, et al. Revisions to the staging and classification of mycosis fungoides and Sézary syndrome: a proposal of the International Society for Cutaneous Lymphomas (ISCL) and the cutaneous lymphoma task force of the European Organization of Research and Treatment of Cancer (EORTC). *Blood* 2007;110:1713-1722.
3. Hodak E, Amitay-Laish I, Feinmesser M, et al. Juvenile mycosis fungoides: cutaneous T-cell lymphoma with frequent follicular involvement. *J Am Acad Dermatol* 2014;70:993-1001.
4. Reiter O, Ben Amitai D, Amitay-Laish I, et al. Pediatric mycosis fungoides: a study of the human leukocyte antigen system among Israeli Jewish patients. *Arch Dermatol Res* 2017;309:851-856.
5. Virmani P, Levin L, Myskowski PL, et al. Clinical outcome and prognosis of young patients with mycosis fungoides. *Pediatr Dermatol* 2017;34:547-553.
6. Agar NS, Wedgeworth E, Crichton S, et al. Survival outcomes and prognostic factors in mycosis fungoides/Sézary syndrome: validation of the revised International Society for Cutaneous Lymphomas/European Organisation for Research and Treatment of Cancer staging proposal. *J Clin Oncol* 2010;28:4730-4739.

7. Cerroni L. Mycosis fungoides-clinical and histopathologic features, differential diagnosis, and treatment. *Semin Cutan Med Surg* 2018;37:2-10.
8. Pimpinelli N, Olsen EA, Santucci M, et al. Defining early mycosis fungoides. *J Am Acad Dermatol* 2005;53:1053-1063.
9. Willemze R. Mycosis fungoides variants-clinicopathologic features, differential diagnosis, and treatment. *Semin Cutan Med Surg* 2018;37:11-17.
10. Nashan D, Faulhaber D, Ständer S, et al. Mycosis fungoides: a dermatological masquerader. *Br J Dermatol* 2007;156:1-10.
11. Martínez-Escala ME, González BR, Guitart J. Mycosis fungoides variants. *Surg Pathol Clin* 2014;7:169-189.
12. Zackheim HS, McCalmont TH. Mycosis fungoides: the great imitator. *J Am Acad Dermatol* 2002;47:914-918.
13. Ahn CS, ALSayyah A, Sangüeza OP. Mycosis fungoides: an updated review of clinicopathologic variants. *Am J Dermatopathol* 2014;36:933-948.
14. Shapiro PE, Pinto FJ. The histologic spectrum of mycosis fungoides/Sézary syndrome (cutaneous T-cell lymphoma). A review of 222 biopsies, including newly described patterns and the earliest pathologic changes. *Am J Surg Pathol* 1994;18:645-667.
15. Kirsch IR, Watanabe R, O'Malley JT, et al. TCR sequencing facilitates diagnosis and identifies mature T cells as the cell of origin in CTCL. *Sci Transl Med* 2015;7, 308ra158.
16. van Doorn R, Scheffer E, Willemze R. Follicular mycosis fungoides, a distinct disease entity with or without associated follicular mucinosis: a clinicopathologic and follow-up study of 51 patients. *Arch Dermatol* 2002;138:191-198.
17. Gerami P, Rosen S, Kuzel T, et al. Folliculotropic mycosis fungoides: an aggressive variant of cutaneous T-cell lymphoma. *Arch Dermatol* 2008;144:738-746.
18. Lehman JS, Cook-Norris RH, Weed BR, et al. Folliculotropic mycosis fungoides: single-center study and systematic review. *Arch Dermatol* 2010;146:607-613.
19. Hodak E, Amitay-Laish I, Atzmony L, et al. New insights into folliculotropic mycosis fungoides (FMF): a single-center experience. *J Am Acad Dermatol* 2016;75:347-355.
20. van Santen S, Roach RE, van Doorn R, et al. Clinical staging and prognostic factors in folliculotropic mycosis fungoides. *JAMA Dermatol* 2016;152:992-1000.
21. Muniesa C, Estrach T, Pujol RM, et al. Folliculotropic mycosis fungoides: clinicopathological features and outcome in a series of 20 cases. *J Am Acad Dermatol* 2010;62:418-426.
22. Tomasini C, Kempf W, Novelli M, et al. Spiky follicular mycosis fungoides: a clinicopathologic study of 8 cases. *J Cutan Pathol* 2015;42:164-172.
23. Mitteldorf C, Stadler R, Sander CA, et al. Folliculotropic mycosis fungoides. *J DDG* 2018;15:543-557.
24. Pileri A, Facchetti F, Rütten A, et al. Syringotropic mycosis fungoides: a rare variant of the disease with peculiar clinicopathologic features. *Am J Surg Pathol* 2011;35:100-109.
25. de Masson A, Battistella M, Vignon-Pennamen MD, et al. Syringotropic mycosis fungoides: clinical and histologic features, response to treatment, and outcome in 19 patients. *J Am Acad Dermatol* 2014;71:926-934.
26. Castano E, Glick S, Wolgast L, et al. Hypopigmented mycosis fungoides in childhood and adolescence: a long-term retrospective study. *J Cutan Pathol* 2013;40:924-934.
27. Rodney IJ, Kindred C, Angra K, et al. Hypopigmented mycosis fungoides: a retrospective clinicohistopathologic study. *J Eur Acad Dermatol Venereol* 2017;31:808-814.
28. Amorim GM, Niemeyer-Corbellini JP, Quintella DC, et al. Hypopigmented mycosis fungoides: a 20-case retrospective series. *Int J Dermatol* 2018;57:306-312.
29. Kazakov DV, Burg G, Kempf W. Clinicopathological spectrum of mycosis fungoides. *J Eur Acad Dermatol Venereol* 2004;18:397-415.
30. Lambroza E, Cohen SR, Phelps R, et al. Hypopigmented variant of mycosis fungoides: demography, histopathology, and treatment of seven cases. *J Am Acad Dermatol* 1995;32:987-993.
31. Furlan FC, Pereira BA, Sotto MN, et al. Hypopigmented mycosis fungoides versus mycosis fungoides with concomitant hypopigmented lesions: same disease or different variants of mycosis fungoides? *Dermatology* 2014;229:271-274.
32. El-Shabrawi-Caelen L, Cerroni L, Medeiros LJ, et al. Hypopigmented mycosis fungoides: frequent expression of a CD8+ T-cell phenotype. *Am J Surg Pathol* 2002;26:450-457.
33. Furlan FC, de Paula Pereira BA, da Silva LF, et al. Loss of melanocytes in hypopigmented mycosis fungoides: a study of 18 patients. *J Cutan Pathol* 2014;41:101-107.
34. Singh ZN, Tretiakova MS, Shea CR, et al. Decreased CD117 expression in hypopigmented mycosis fungoides correlates with hypomelanosis: lessons learned from vitiligo. *Mod Pathol* 2006;19:1255-1260.
35. Pavlovsky L, Mimouni D, Amitay-Laish I, et al. Hyperpigmented mycosis fungoides: an unusual variant of cutaneous T-cell lymphoma with a frequent CD8+ phenotype. *J Am Acad Dermatol* 2012;67:69-75.
36. David M, Shanon A, Hazaz B, et al. Diffuse, progressive hyperpigmentation: an unusual skin manifestation of mycosis fungoides. *J Am Acad Dermatol* 1987;16:257-260.
37. Kikuchi A, Shimizu H, Nishikawa T. Mycosis fungoides with marked hyperpigmentation. *Dermatology* 1996;192:360-363.
38. Trattner A, Hodak E, David M. Screening patch tests for pigmented contact dermatitis in Israel. *Contact Dermatitis* 1999;40:155-157.
39. Beard JS, Smith KJ, Skelton HG. Combination chemotherapy with 5-fluorouracil, folinic acid, and alpha-interferon producing histologic features of graft-versus-host disease. *J Am Acad Dermatol* 1993;29:325-330.
40. Hodak E, Amitay I, Feinmesser M, et al. Ichthyosiform mycosis fungoides: an atypical variant of cutaneous T-cell lymphoma. *J Am Acad Dermatol* 2004;50:368-374.
41. Ryan C, Whittaker S, D'Arcy C, et al. Juvenile folliculotropic and ichthyosiform mycosis fungoides. *Clin Exp Dermatol* 2009;34:e160-e162.
42. Kütting B, Metze D, Luger TA, et al. Mycosis fungoides presenting as an acquired ichthyosis. *J Am Acad Dermatol* 1996;34:887-889.
43. Marzano AV, Borghi A, Facchetti M, et al. Ichthyosiform mycosis fungoides. *Dermatology* 2002;204:124-129.
44. Jang MS, Kang DY, Park JB, et al. Clinicopathological manifestations of ichthyosiform mycosis fungoides. *Acta Derm Venereol* 2016;96:100-101.
45. Morizane S, Setsu N, Yamamoto T, et al. Ichthyosiform eruptions in association with primary cutaneous T-cell lymphomas. *Br J Dermatol* 2009;161:115-120.
46. Miura T, Kato Y, Yamamoto T. Ichthyosiform sarcoidosis: report of three cases from Japan and literature review. *Sarcoidosis Vasc Diffuse Lung Dis* 2016;33:392-397.
47. Kodama K, Fink-Puches R, Massone C, et al. Papular mycosis fungoides: a new clinical variant of early mycosis fungoides. *J Am Acad Dermatol* 2005;52:694-698.
48. Vonderheid EC, Kadin ME, Telang GH. Commentary about papular mycosis fungoides, lymphomatoid papulosis and lymphomatoid pityriasis lichenoides: more similarities than differences. *J Cutan Pathol* 2016;43:303-312.
49. Uddin A, Bennett M, Nayeem K, et al. A case of papular mycosis fungoides: new clinical variant of early mycosis fungoides. *J Eur Acad Dermatol Venereol* 2007;21:685-687.
50. Martorell-Calatayud A, Botella-Estrada R, Sanmartín-Jimenez O, et al. Papular mycosis fungoides: two new cases of a recently described clinicopathological variant of early mycosis fungoides. *J Cutan Pathol* 2010;37:330-335.
51. 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;140:441-447.
52. de Unamuno Bustos B, Ferriols AP, Sánchez RB, et al. Adult pityriasis lichenoides-like mycosis fungoides: a clinical variant of mycosis fungoides. *Int J Dermatol* 2014;53:1331-1338.
53. Ko JW, Seong JY, Suh KS, et al. Pityriasis lichenoides-like mycosis fungoides in children. *Br J Dermatol* 2000;142:347-352.

54. Wang SH, Hsiao CH, Hsiao PF, et al. Adult pityriasis lichenoides-like mycosis fungoides with high density of CD8-positive T-lymphocytic infiltration. *J Eur Acad Dermatol Venereol* 2007;21:401-402.
55. Topf S, Lüftl M, Neisius U, et al. Mycosis fungoides palmaris et plantaris—an unusual variant of cutaneous T-cell lymphoma. *Eur J Dermatol* 2006;16:84-86.
56. Resnik KS, Kantor GR, Lessin SR, et al. Mycosis fungoides palmaris et plantaris. *Arch Dermatol* 1995;131:1052-1056.
57. Spieth K, Grundmann-Kollmann M, Runne U, et al. Mycosis-fungoides-type cutaneous T cell lymphoma of the hands and soles: a variant causing delay in diagnosis and adequate treatment of patients with palmoplantar eczema. *Dermatology* 2002;205:239-244.
58. Wang CM, Duvic M, Dabaja BS. Acral erosive mycosis fungoides: successful treatment with localised radiotherapy. *BMJ Case Rep* 2013;2013.
59. Kim ST, Jeon YS, Sim HJ, et al. Clinicopathologic features and T-cell receptor gene rearrangement findings of mycosis fungoides palmaris et plantaris. *J Am Acad Dermatol* 2006;54:466-471.
60. Toritsugi M, Satoh T, Higuchi T, et al. A vesiculopustular variant of mycosis fungoides palmaris et plantaris masquerading as palmoplantar pustulosis with nail involvement. *J Am Acad Dermatol* 2004;51:139-141.
61. Nakai N, Hagura A, Yamazato S, et al. Mycosis fungoides palmaris et plantaris successfully treated with radiotherapy: case report and mini-review of the published work. *J Dermatol* 2014;41:63-67.
62. Lambert TJ, Prieto VG, Duvic M. Multiple plaques on the hands and feet. Mycosis fungoides (MF) palmaris et plantaris (MFPP). *Arch Dermatol* 2007;143:109-114.
63. Magro CM, Nguyen GH. Keratoderma-like T cell dyscrasia: a report of 13 cases and its distinction from mycosis fungoides palmaris et plantaris. *Indian J Dermatol Venereol Leprol* 2016;82:395-403.
64. Kelati A, Gallouj S, Tahiri L, et al. Defining the mimics and clinicohistological diagnosis criteria for mycosis fungoides to minimize misdiagnosis. *Int J Womens Dermatol* 2017;3:100-106.
65. Martinez-Escala ME, Posligua AL, Wickless H, et al. Progression of undiagnosed cutaneous lymphoma after anti-tumor necrosis factor-alpha therapy. *J Am Acad Dermatol* 2018;78:1068-1076.
66. Azizpour A, Ghanadan A, Nasimi M, et al. Psoriasiform mycosis fungoides: a rare form of the disease with review of the literature. *Dermatol Online J* 2017;23(9).
67. Lafaille P, Bouffard D, Provost N. Exacerbation of undiagnosed mycosis fungoides during treatment with etanercept. *Arch Dermatol* 2009;145:94-95.
68. Zackheim HS, Koo J, LeBoit PE, et al. Psoriasiform mycosis fungoides with fatal outcome after treatment with cyclosporine. *J Am Acad Dermatol* 2002;47:155-157.
69. Weenig RH, Comfere NI, Gibson LE. Fatal cytotoxic cutaneous lymphoma presenting as ulcerative psoriasis. *Arch Dermatol* 2009;145:801-808.
70. Doukaki S, Arico M, Bongiorno MR. A rare presentation of mycosis fungoides mimicking psoriasis vulgaris. *Case Rep Dermatol* 2009;1:60-65.
71. Jinno N, Yamana Y, Kawashima M, et al. Mycosis fungoides with psoriasiform lesions. *Jpn Dermatol Assoc* 2015;42:227-229.
72. Haghighi B, Smoller BR, LeBoit PE, et al. Pagetoid reticulosis (Woringer-Kolopp disease): an immunophenotypic, molecular, and clinicopathologic study. *Mod Pathol* 2000;13:502-510.
73. Larson K, Wick MR. Pagetoid reticulosis: report of two cases and review of the literature. *Dermatopathology (Basel)* 2016;3:8-13.
74. Steffen C. Ketron-Goodman disease, Woringer-Kolopp disease, and pagetoid reticulosis. *Am J Dermatopathol* 2005;27:68-85.
75. Hodak E, Phenig E, Amichai B, et al. Unilesional mycosis fungoides: a study of seven cases. *Dermatology* 2000;201:300-306.
76. Cerroni L, Fink-Puches R, El-Shabrawi-Caelen L, et al. Solitary skin lesions with histopathologic features of early mycosis fungoides. *Am J Dermatopathol* 1999;21:518-524.
77. Ally MS, Robson A. A review of the solitary cutaneous T-cell lymphomas. *J Cutan Pathol* 2014;41:703-714.
78. Amitay-Laish I, Feinmesser M, Ben-Amitai D, et al. Unilesional folliculotropic mycosis fungoides: a unique variant of cutaneous lymphoma. *J Eur Acad Dermatol Venereol* 2016;30:25-29.
79. Ally MS, Pawade J, Tanaka M, et al. Solitary mycosis fungoides: a distinct clinicopathologic entity with a good prognosis: a series of 15 cases and literature review. *J Am Acad Dermatol* 2012;67:736-744.
80. Heald PW, Glusac EJ. Unilesional cutaneous T-cell lymphoma: clinical features, therapy, and follow-up of 10 patients with a treatment-responsive mycosis fungoides variant. *J Am Acad Dermatol* 2000;42:283-285.
81. van Santen S, Jansen PM, Vermeer MH, et al. Folliculotropic mycosis fungoides presenting with a solitary lesion: clinicopathological features and long-term follow-up data in a series of 9 cases. *J Cutan Pathol* 2018;45:122-128.
82. Magro CM, Telang GH, Momtahen S. Unilesional follicular mycosis fungoides: report of 6 cases and review of the literature. *Am J Dermatopathol* 2018;40:329-336.
83. Kaufmann F, Kettelhack N, Hilty N, et al. Unilesional plantar mycosis fungoides treated with topical photodynamic therapy—case report and review of the literature. *J Eur Acad Dermatol Venereol* 2017;31:1633-1637.
84. Nofal A, Salah E. Acquired poikiloderma: proposed classification and diagnostic approach. *J Am Acad Dermatol* 2013;69:e129-e140.
85. Mataix J, Bañuls J, Lucas A, et al. Poikilodermatous mycosis fungoides. *Int J Dermatol* 2007;46:950-951.
86. McKee PH, Calonje E, Granter S. *Pathology of the skin with clinical correlations. 3rd ed.* Philadelphia, PA: Mosby. 2005.
87. Raznatovskiĭ IM. Brocq's parapsoriases, their relationship to pokiloderma, mycosis fungoides and skin reticulosis. *Vestn Dermatol Venerol* 1976:42-46.
88. Abbott RA, Sahni D, Robson A, et al. Poikilodermatous mycosis fungoides: a study of its clinicopathological, immunophenotypic, and prognostic features. *J Am Acad Dermatol* 2011;65:313-319.
89. Nikolaou VA, Papadavid E, Katsambas A, et al. Clinical characteristics and course of CD8+ cytotoxic variant of mycosis fungoides: a case series of seven patients. *Br J Dermatol* 2009;161:826-830.
90. Barnhill RL, Braverman IM. Progression of pigmented purpura-like eruptions to mycosis fungoides: report of three cases. *J Am Acad Dermatol* 1988;19:25-31.
91. Georgala S, Katoulis AC, Symeonidou S, et al. Persistent pigmented purpuric eruption associated with mycosis fungoides: a case report and review of the literature. *J Eur Acad Dermatol Venereol* 2001;15:62-64.
92. Martínez W, del Pozo J, Vázquez J, et al. Cutaneous T-cell lymphoma presenting as disseminated, pigmented, purpura-like eruption. *Int J Dermatol* 2001;40:140-144.
93. Ladrigan MK, Poligone B. The spectrum of pigmented purpuric dermatosis and mycosis fungoides: atypical T-cell dyscrasia. *Cutis* 2014;94:297-300.
94. Hanna S, Walsh N, D'Intino Y, et al. Mycosis fungoides presenting as pigmented purpuric dermatitis. *Pediatr Dermatol* 2006;23:350-354.
95. Coulombe J, Jean SE, Hatami A, et al. Pigmented purpuric dermatosis: clinicopathologic characterization in a pediatric series. *Pediatr Dermatol* 2015;32:358-362.
96. Kim DH, Seo SH, Ahn HH, et al. Characteristics and clinical manifestations of pigmented purpuric dermatosis. *Ann Dermatol* 2015;27:404-410.
97. Ugajin T, Satoh T, Yokozeki H, et al. Mycosis fungoides presenting as pigmented purpuric eruption. *Eur J Dermatol* 2005;15:489-491.
98. Hodak E, David M, Maron L, et al. CD4/CD8 double-negative epidermotropic cutaneous T-cell lymphoma: an immunohistochemical variant of mycosis fungoides. *J Am Acad Dermatol* 2006;55:276-284.
99. Lor P, Krueger U, Kempf W, et al. Monoclonal rearrangement of the T cell receptor gamma-chain in lichenoid pigmented purpuric dermatitis of gougerot-blum responding to topical corticosteroid therapy. *Dermatology* 2002;205:191-193.
100. Toro JR, Sander CA, LeBoit PE. Persistent pigmented purpuric dermatitis and mycosis fungoides: simulant, precursor, or both? A study by

light microscopy and molecular methods. *Am J Dermatopathol* 1997;19:108-118.
101. Imafuku K, Nomura Y, Nakayama C, et al. Erythema annulare centrifugum-like mycosis fungoides after unrelated bone marrow transplantation. *Br J Haematol* 2015;170:140.
102. Ceyhan AM, Akkaya VB, Chen W, et al. Erythema annulare centrifugum-like mycosis fungoides. *Australas J Dermatol* 2011;52:e11-e13.
103. Bernardini ML, Brandozzi G, Campanati A, et al. Bullous-vesicular variant of mycosis fungoides presenting as erythema annulare centrifugum: a case report. *J Eur Acad Dermatol Venerol* 2009;23:839-840.
104. Notay M, Petukhova TA, Kiuru M, et al. Mycosis fungoides presenting as symmetric concentric patches mimicking figurate erythema. *JAAD Case Rep* 2017;3:288-290.
105. Hallopeau H, Bureau G. Sur un cas de mycose fungoide avec localisation initiale d'eruptions polymorphes et vegetations axillaires et inguinales. *Bull Soc Fr Dermulol Syhiligr* 1896;7:480.
106. Nicolis GD, Stratigos JD, Tosca AD, et al. Mycosis fungoides with verrucous lesions. *Acta Derm Venereol* 1979;59:80-82.
107. Price NM, Fuks ZY, Hoffman TE. Hyperkeratotic and verrucous features of mycosis fungoides. *Arch Dermatol* 1977;113:57-60.
108. Jang JG, Sim HJ, Kim SH, et al. Mycosis fungoides mimicking inflammatory linear verrucous epidermal nevus. *J Eur Acad Dermatol Venereol* 2004;18:218-220.
109. Willemze R, Scheffer E, Van Vloten WA. Mycosis fungoides simulating acanthosis nigricans. *Am J Dermatopathol* 1985;7:367-371.
110. Neill SM, Monk BE, du Vivier A. Mycosis fungoides associated with acanthosis nigricans. *J R Soc Med* 1985;78:79-81.
111. Puig L, Musulen E, Fernandez-Figueras MT, et al. Mycosis fungoides associated with unusual epidermal hyperplasia. *Clin Exp Dermatol* 1996;21:61-64.
112. Korekawa A, Kaneko T, Nakajima K, et al. Mycosis fungoides bullosa associated with bullous pemphigoid. *Int J Dermatol* 2015;54:e366-e368.
113. Bowman PH, Hogan DJ, Sanusi ID. Mycosis fungoides bullosa: report of a case and review of the literature. *J Am Acad Dermatol* 2001;45:934-939.
114. Gantcheva M, Lalova A, Broshtilova V, et al. Vesicular mycosis fungoides. *J Dtsch Dermatol Ges* 2005;3:898-900.
115. Kamran B, Fatemeh M, Ahmadreza R. Bullous mycosis fungoides: a case report. *Dermatol Online J* 2008;14:11.
116. Ueda C, Makino T, Asano Y, et al. An ultrastructural examination of a blistering lesion of mycosis fungoides bullosa. *Br J Dermatol* 2011;165:213-214.
117. Sarnoff DS, DeFeo CP. Coexistence of pemphigus foliaceus and mycosis fungoides. *Arch Dermatol* 1985;121:669-672.
118. Sato S, Okamoto O, Kawamoto M, et al. Bullous mycosis fungoides associated with an extensive ulcer and a severe leukemoid reaction. *Dermatol Rep* 2011;3, e54.
119. Ackerman AB, Miller RC, Shapiro L. Pustular mycosis fungoides. *Arch Dermatol* 1966;93:221-225.
120. Camisa C, Aulisio A. Pustular mycosis fungoides. *Cutis* 1994;54:202-204.
121. Reggiani C, Massone C, Fink-Puches R, et al. Interstitial mycosis fungoides. A clinicopathologic study of 21 patients. *Am J Surg Pathol* 2016;40:1360-1367.
122. Su LD, Kim YH, LeBoit PE, et al. Interstitial mycosis fungoides, a variant of mycosis fungoides resembling granuloma annulare and inflammatory morphea. *J Cutan Pathol* 2002;29:135-141.
123. Jouary T, Beylot-Barry M, Vergier B, et al. Mycosis fungoides mimicking granuloma annulare. *Br J Dermatol* 2002;146:1102-1104.
124. Ferrara G, Crisman G, Zalaudek I, et al. Free-floating collagen fibers in interstitial mycosis fungoides. *Am J Dermatopathol* 2010;32:352-356.
125. Pfaltz K, Kerl K, Palmedo G, et al. Clonality in sarcoidosis, granuloma annulare, and granulomatous mycosis fungoides. *Am J Dermatopathol* 2011;33:659-662.
126. Koochek A, Fink-Puches R, Cerroni L. Coexistence of patch stage mycosis fungoides and interstitial granuloma annulare in the same patient: a pitfall in the clinicopathologic diagnosis of mycosis fungoides. *Am J Dermatopathol* 2012;34:198-202.
127. Kempf W, Ostheeren-Michaelis S, Paulli M, et al. Granulomatous mycosis fungoides and granulomatous slack skin: a multicenter study of the Cutaneous Lymphoma Histopathology Task Force Group of the European Organization For Research and Treatment of Cancer (EORTC). *Arch Dermatol* 2008;144:1609-1617.
128. Clarijs M, Poot F, Laka A, et al. Granulomatous slack skin: treatment with extensive surgery and review of the literature. *Dermatology* 2003;206:393-397.
129. Li JY, Pulitzer MP, Myskowski PL, et al. A case-control study of clinicopathologic features, prognosis, and therapeutic responses in patients with granulomatous mycosis fungoides. *J Am Acad Dermatol* 2013;69:366-374.
130. Gangar P, Venkatarajan S. Granulomatous lymphoproliferative disorders: granulomatous slack skin and lymphomatoid granulomatosis. *Dermatol Clin* 2015;33:489-496.
131. Ruiz-de-Casas A, Carrizosa-Esquivel A, Herrera-Saval A, et al. Sézary syndrome associated with granulomatous lesions during treatment with bexarotene. *Br J Dermatol* 2006;154:372-374.
132. Haruyama S, Sugita K, Kawakami C, et al. Development of a prominent granulomatous eruption after interferon-gamma therapy in a patient with mycosis fungoides. *Acta Derm Venereol* 2010;90:190-191.
133. Gutte R, Kharkar V, Mahajan S, et al. Granulomatous mycosis fungoides with hypohidrosis mimicking lepromatous leprosy. *Indian J Dermatol Venereol Leprol* 2010;76:686-690.
134. Scarabello A, Leinweber B, Ardigó M, et al. Cutaneous lymphomas with prominent granulomatous reaction: a potential pitfall in the histopathologic diagnosis of cutaneous T- and B-cell lymphomas. *Am J Surg Pathol* 2002;26:1259-1268.
135. Dabiri S, Morales A, Ma L, et al. The frequency of dual TCR-PCR clonality in granulomatous disorders. *J Cutan Pathol* 2011;38:704-709.
136. Mangold AR, Thompson AK, Davis MD, et al. Early clinical manifestations of Sézary syndrome: a multicenter retrospective cohort study. *J Am Acad Dermatol* 2017;77:719-727.
137. Miyagaki T, Sugaya M. Erythrodermic cutaneous T-cell lymphoma: how to differentiate this rare disease from atopic dermatitis. *J Dermatol Sci* 2011;64:1-6.
138. Olek-Hrab K, Silny W. Diagnostics in mycosis fungoides and Sézary syndrome. *Rep Pract Oncol Radiother* 2013;19:72-76.