PATCH STAGE MYCOSIS FUNGOIDES IN A 21-YEAR-OLD FEMALE PATIENT

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Abstract

Mycosis fungoides (MF) is a relatively rare type of lymphoma that represents over half of all cutaneous T-cell lymphoma (CTCL). An increase in the annual global incidence of MF was observed during the last decades. Clinically, skin lesions of variable forms and dimensions, persistent, or slowly progressive, with the appearance of erythematous macules, plaques, and tumors tend to appear over sun-protected areas. Herein, we report the case of a 21-year-old Romanian female patient presenting with patch stage MF, and discuss clinical, histological and treatment issues in this case.

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Keywords:

Mycosis fungoides, female, Romania, cutaneous T-cell lymphoma.

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Introduction

Mycosis fungoides (MF) is the most frequent variant of cutaneous T-cell lymphoma (CTCL). Together with Sezary syndrome (SS), which was considered to be a leukemic variant of MF for a long time, they represent about two thirds of the total primary cutaneous lymphomas (1, 2).

Even though MF is a relatively rare type of lymphoma, it represents over one third of all primary cutaneous non-Hodgkin lymphomas and over half of all cutaneous T-cell lymphomas (2).

During the last decades, an increase in the annual global incidence of MF was observed (2, 3). This increase could be partly explained by the improvement of diagnostic and treatment standards (4) and also by the modification of reporting methods and classification of lymphoproliferative diseases over time.

Mycosis fungoides has been observed in patients infected with the human lymphotropic virus T (HTLV-1). These patients frequently develop T cell leukemia, having skin involvement which is indistinguishable from MF, thus leading some to consider that CTCL is a consequence of HTLV-1 infection (5).

Although MF can start at an early age, it usually occurs between the fifth and seventh decades of life, having a median age of diagnosis of 57 years. World-wide, an incidence of four cases *per* million inhabitants is reported; the disease is 2.2 times more common in men and has a slightly higher incidence among African Americans in comparison to Caucasians, as well as in people exposed to petrochemicals (6).

Mycosis fungoides is clinically manifested by skin lesions of variable forms and dimensions, persistent, or slowly progressive, with the appearance of erythematous macules, plaques and, more rarely, tumors or generalized erythroderma. The lesions are often pruriginous, with profound impairment of the patient's quality of life (7). In most cases, the distribution of lesions overlaps sun-protected areas, but any anatomical area may be affected, including palms and soles (8, 9). Patches and plaques affect less than 10% of the body surface area in about 30% of patients, while about 35% of patients experience a more generalized impairment (10). Twenty percent and 15% of patients, respectively, develop tumors or reach the erythrodermic status.

A definitive diagnosis of MF is often preceded by a "premycotic" phase, which can last from several months to several decades, during which the patient can manifest erythematous, slightly squamous, non-specific lesions with inconclusive biopsy (11, 12). These lesions may fluctuate over the years, so it is often misdiagnosed as non-specific dermatitis or plaque parapsoriasis (13). Sometimes repeated biopsies can be necessary for a final diagnosis, especially when the initial biopsy is inconclusive (14).

The diagnosis of MF in its initial phases is made according to an algorithm established by the International Society of Cutaneous Lymphomas (ISCL): a certain number of points is given for each category of data (clinical, histopathological, biomolecular and immunopathological data). The diagnosis of MF can be formulated when a score over four points is obtained (8, 15).

Histopathological examination in more advanced stages represents the diagnostic gold standard in case of clinical suspicion of a MF. An immunohistochemical study is performed in order to complete the diagnostic examination and to indicate the best course of treatment and establish prognosis. Herein, we present a case of mycosis fungoides in a young Romanian female patient and discuss clinical, histologic and treatment issues in this case.

Case presentation

A 21 year-old Caucasian female presented for the first time in a dermatology clinic six years ago. She was then described as having a scaly erythematous cutaneous lesion localised on the right posterior thigh with progressive appearance of similar lesions on the contralateral posterior thigh. There was no history of skin disease in her known relatives.

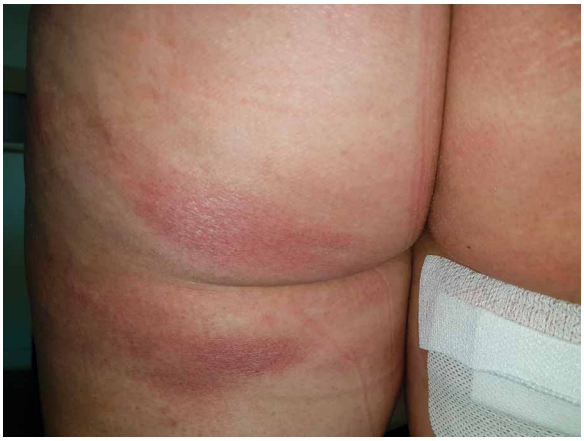

Figure 1. Clinical image showing erythematous patches on buttocks

She was seen again in a dermatology clinic two years ago, when a skin biopsy from the lesion on the right thigh was performed, with an inconclusive result. At the time being, she was diagnosed with cutaneous lupus and received treatment with Hydroxychloroquine 400 mg *per* day (distributed in two doses each day) and topical corticosteroids for three months. Due to no clinical benefit, the patient decided to stop the treatment without consulting with her dermatologist.

Eight years from the first examination, she presented to our dermatology department. Clinical examination revealed erythematous patches, with minimal pruritus, localised on her both thighs and bottocks (Figure 1). Due to the prior histopathological result, we decided to perform a skin biopsy from a lesion on the left thigh, which was interpreted as mycosis fungoides. The histopathology examination showed a bandlike upper dermal infiltrate of lymphocytes and other inflammatory cells, epidermotropism of mononuclear cells, slight spongiosis of the epidermis and lymphocytes with hyperchromatic and cerebriform nuclei (Figure 2 A, B). The diagnosis was established as MF in the early stages, stage IA (T1N0M0B0) according to the modified tumor-node-metastasis-blood (TNMB) classification.

The patient has no history of chemical exposure or radiation, and no alcohol consumption or smoking history. We decided to perform further investigations: a thoraco-abdomino-pelvic computed-tomography (CT) and a haematology consult. The complete blood count and the CT were within normal limits. Test for antinuclear antibodies was negative. Viral screens for HIV, hepatitis B and Hepatitis C were also negative.

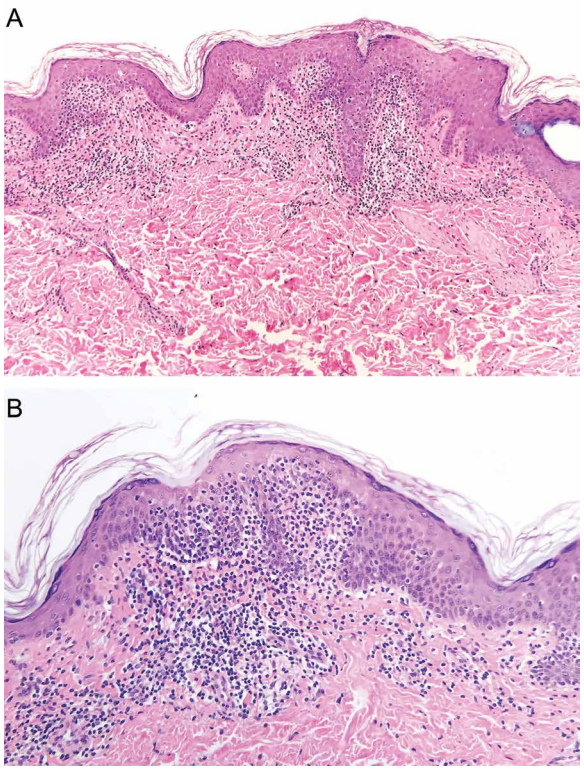

Figure 2. A, B. Histopathological aspect showing a bandlike upper dermal infiltrate of lymphocytes and other inflammatory cells, epidermotropism of mononuclear cells, spongiosis of the epidermis and lymphocytes with hyperchromatic and cerebriform nuclei (Hematoxylin and eosin stain, magnification 40x and 200x, respectively)

Treatment was started with Acitretin 10 mg/day for one month and then increased to 20 mg/day in association with hydrocortisone lotion and phototherapy with narrow-band UVB 3-4 times/week, for a regimen of 12 weeks. Follow-up visit was scheduled at one month.

Discussions

An important discussion is whether or not early onset of MF leads to a more aggressive course than onset at adult age. Some studies suggest that MF with onset at an early age can be more aggressive in comparison to MF with onset during adult life. Paller *et al* reported an aggressive nodular, primary CTCL in a 11-year-old girl, and correlating this case with the other cases reported at that time, reached the conclusion that the course of cutaneous lymphomas is less indolent in children in comparison to adults (16).

Conversely, Zackheim *et al* suggest that (13) sometimes better outcomes after specific treatment for MF is correlated with an earlier onset of the disease.

Taniguchi *et al*. present a case of tumoral stage MF in a 12-year-old girl treated with PUVA for one month after which the clearance of the nodular lesions was observed (17). Meister *et al* presented a case of a 11-year-old girl diagnosed with Sezary syndrome treated initially with systemic chemotherapy and PUVA three times a week for 24 months. Due to her skin lesion and alopecia improvement, the chemotherapy was stopped and PUVA treatment was reduced to once a week (18).

Burns *et al*. performed a study including 246 patients with a history of cutaneous lesions consistent with mycosis fungoides and with onset of the symptoms before 30 years of age (19). This study shows that even though MF is a disease of elderly people, we should take into consideration this diagnosis in young people with skin lesions resembling cutaneous lymphoma. Also there are some various reports that suggest that early-onset MF can be associated with non-T-cell lymphomas such as Hodgkin disease (20-22), even though this kind of association is usually found in patients with later onset of MF (23).

Conclusion

Mycosis fungoides is not considered a disease of the elderly anymore, a great part of these patients having had their first symptoms years earlier than the age of diagnosis. Crowley *et al* (24) developed a study using a cohort of 58 patients diagnosed with mycosis fungoides younger than 35 years of age. In comparison to elderly people with MF, the younger group had a smaller percentage of patients with stage T4 (erythroderma) and a higher percentage with stage T1 (limited patches), which is also the case of our patient. When looking at the patients in the tumoral stage, age was not a good predictor of survival rate. The response to therapy was similar between the patients with early diagnosis of MF and those with regular diagnosis of MF, therefore giving our patient a good prognosis under treatment.

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