Mycosis fungoides presenting as hypopigmented and pigmented purpuric dermatosis-like lesions in a girl: a case report and literature review

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Mycosis fungoides (MF) is the most common skin subtype of T-cell lymphoma. Its underlying cause is not yet clearly understood, and diagnosis might be difficult since MF presents itself with symptoms similar to some other dermatologic disorders. In the current case presentation, we report a 12-year-old female with concurrent hypopigmented and pigmented purpuric dermatosis-like lesions that underwent skin biopsies and immunohistochemistry study. Microscopic findings of hyperkeratosis and mild acanthosis in addition to epidermotropism of lymphocytes with perivascular and interstitial infiltration led us to the probable diagnosis of MF. Intraepidermal lymphocyte immunohistochemistry assessments were positive for CD3 and CD4 while negative for CD8 and CD7. The peripheral blood smear revealed a normal CD4 to CD8 ratio, and the number of Sézary cells detected was insufficient to diagnose Sézary syndrome. Therefore, the final diagnosis of MF was made for this young patient.

Keywords: hypopigmented mycosis fungoides; mycosis fungoides; immunohistochemistry

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INTRODUCTION

Mycosis fungoides (MF) is the most common subtype of cutaneous T-cell lymphoma and was first described in 1806 by Alibert. In 1870, further investigations by Bazin led to the classification of different stages of the disease. Therefore, MF is also known as Alibert-Bazin syndrome. Although the underlying cause of MF is elusive, genetic, environmental or occupational exposure to chemical substances and agents that trigger T-cell lymphocyte activation (such as HTLV-1, HIV, or Herpes virus family infection) play a remarkable role in the incidence of the disease [1,2]. MF affects mostly old adults (between 55 to 60 years) and has an incidence rate of approximately 0.5 per 100,000 individuals per year [3,4]. Reports indicate that males are more affected than females, with a male to female ratio ranging from 1.7–2.2: 1. Patients usually have progressive skin lesions of varying size and shapes. Especially in the early stages, the diagnosis may be difficult due to the similarity of the skin biopsy results to other dermatologic abnormalities such as psoriasis, parapsoriasis, lichen planus, and vitiligo, so MF may often be misdiagnosed [1].

This report introduces a child with an atypical form of MF with concurrent hypopigmented and pigmented purpuric dermatosis (PPD)-like lesions. She had not been diagnosed for a relatively long period and therefore had not been treated or referred to the higher health care levels.

CASE PRESENTATION

The patient was a 12-year-old girl of Afghan

nationality. About a year ago, the patient developed numerous purpuric lesions (PPD-like lesions) on the lower limbs. These lesions gradually enlarged and did not recover over several months, and hypopigmented patches appeared. The lesions were scattered all over the body with different sizes. Before referring to us at Al-Zahra Hospital in Isfahan, Iran, the patient had not been evaluated, diagnosed or treated. The patient's skin type was Fitzpatrick skin 4 (Figure 1).

A skin biopsy was obtained from the hypopigmented and PPD-like lesions, and differential diagnoses such as vitiligo, MF, lichen sclerosus, morphea, and leprosy were considered. Microscopic findings of the skin samples, derived from hypopigmented lesions, showed hyperkeratosis and mild acanthosis with an elongation of rete ridges. There was an epidermotropism of lymphocytes with hyperchromatic nuclei in the lower portion of the epidermis in some areas with a perinuclear halo. Furthermore, in the pathological study of the PPD-like lesions, moderate perivascular and interstitial infiltration with some extravasated erythrocytes were present. According to these findings, MF was considered as the most probable diagnosis (Figure 2).

In the immunohistochemistry (IHC) study, the intraepidermal lymphocytes were positive for the CD3 and CD4 markers while negative for CD8 and CD7. The results of the complete blood count study did not show any abnormal findings. Furthermore, the CD4 to CD8 ratio assessed in the peripheral blood smear (PBS) was normal. As Sézary cells were not present in the PBS, we were reassured that Sézary syndrome can not be considered for this patient. Her chest X-ray was normal, and no

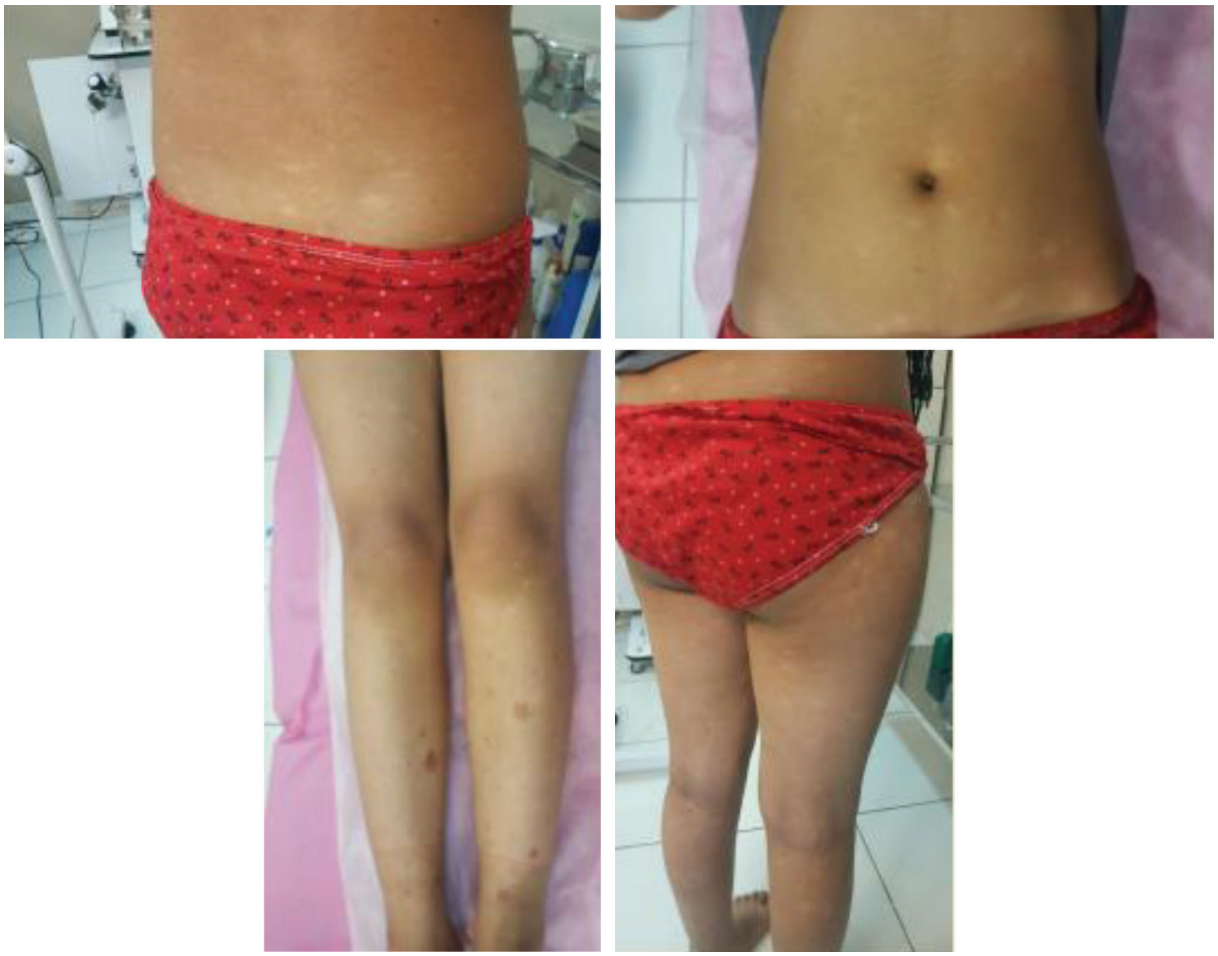

Figure 1. The skin lesions of the patient.

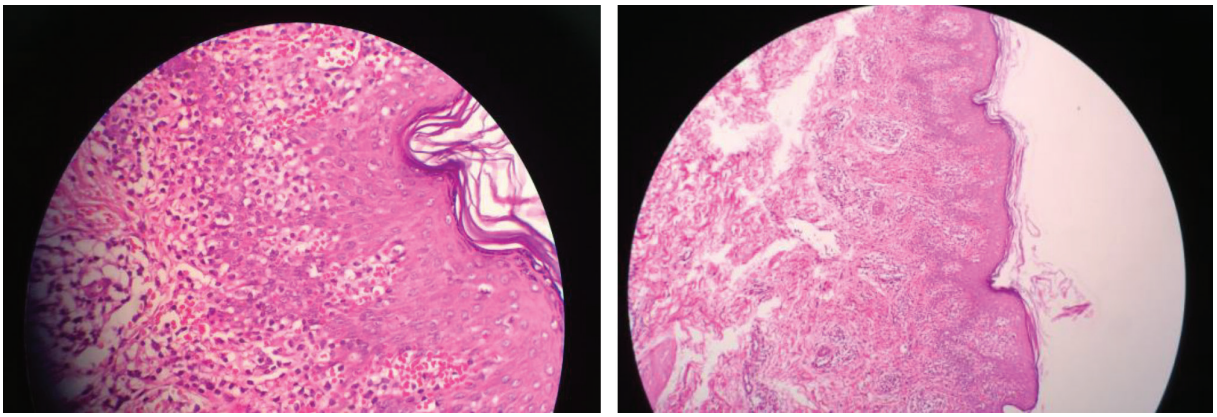

Figure 2. Pigmented purpuric dermatosis-like lesions (H & E; ×100 & ×400).

lymphadenopathy was detected. The serum lactate dehydrogenase level was within the normal range.

The final diagnosis of MF was made considering clinical, histopathological, and IHC findings; two clinical and two histopathological criteria were met. Based on the mentioned findings, the staging of MF was IB. Therefore, she was treated with emollients and Narrow-Band Ultraviolet-B (NBUVB) radiation. The treatment continues, and the lesions have improved.

DISCUSSION

The incidence of skin-lymphoma increases dramatically as the age rises. It occurs almost four times more in the age group of above 70 years compared with lower ages. Approximately two-thirds of cases of cutaneous T-cell lymphoma are individuals with MF or Sézary syndrome; the incidence of these diseases has increased in recent years. Studies in Iran have shown an estimated rate of 3.91 per million cases of skin lymphoma in this community [5]. Considering the remarkable mortality rate in these patients due to problems such as sepsis, it is essential to diagnose and treat these patients in the early stages [6,7]. Fatemi *et al.*'s study of MF in the Iranian population showed that the disease occurred at an earlier age and was more prevalent in females [8]. Another study by the same authors in Isfahan showed that most patients were at an early stage of the disease, and women were more likely to be affected [9]. The contamination of the living environment with industrial waste, the availability of healthcare centers, and variations in ethnic groups might have caused such results, though the sample size in these studies was relatively small. Therefore, under-aged people in this area of Iran should be investigated accurately.

Mycosis fungoides typically appears in the form of the progressive development of patches, plaques or tumors, or might involve visceral components. These lesions are characterized as the "classic" state of the disease. Although MF can be observed in different parts of the skin, areas that are not typically exposed to sunlight (such as buttocks, back, or breasts) are more likely to be affected [2]. Microscopic findings may show the proliferation of small to medium-sized pleomorphic lymphocytes, and prominent epidermotropic lymphocytes are often detected. Besides, hyperconvoluted cerebriform nuclei may also be present in these lymphocytes. Diagnosis in the early stages of the disease is usually unspecific as it may overlap with other inflammatory conditions [10]. Hypopigmented lesions are widely detected among those diagnosed as MF in earlier ages. On the other hand, these hypopigmented lesions can pose misdiagnosis due to the wide range of differential diagnoses and overlap syndromes [8].

The findings of IHC may indicate epidermotropic peripheral T-helper memory lymphocytes; CD2+, CD3+, CD4+, CD8-, CD7-, CD45RO+, CD20- and CD30- are common phenotypes. Cytotoxic T-lymphocyte CD4- with CD8+ or CD4 and CD8 double-negative or double-positive cases can be found rarely without significant clinical differences compared to other similar cases. Also, decreased CD7 expression may be noted [8].

Mycosis fungoides can present in atypical forms, among which are pagetoid reticulosis,

hypopigmented MF, hyperpigmented MF, ichthyosiform MF, pustular MF, and PPD overlap MF [10,11]. The diagnosis can be more complicated in case of the presence of the above categories. In this case, we observed the presence of hypopigmented MF along with PPD-like lesions in a female child, representing a rare coincidence. PPDs are a group of chronic treatment-resistant capillary lesions that may have different manifestations with unknown etiology [12]. As a type of T-cell dyscrasia, PPD might be characterized in the spectrum of cutaneous T-cell lymphoma, making our case quite challenging [13].

Some studies have reported cases of atypical PPD with findings such as cerebriform nuclear contours and CD4 predominance and significant loss of CD7 expression, evolving into MF. Therefore, it is critical to distinguish other conditions in which histopathologic findings are similar to PPD (such as trauma) due to the similarity of the clinical features in PPD lesions with MF manifestations (especially atypical forms) and the probability of progression of PPD cases to MF. Hence, critical considerations should be made to avoid misdiagnosis of atypical MF in situations similar to the patient of our study.

Our study's novel points include the concurrent presence of hypopigmented lesions with PPD-like ones in a female child that has not been previously reported. As the rarity and variety of clinical findings may mislead dermatologists in making the final diagnosis of MF, skin biopsies are strongly recommended is similar situations.

CONCLUSION

Mycosis fungoides (MF) can have different clinical features; even concomitant dermatologic findings may be observed. Our patient was diagnosed as MF with PPD-like and hypopigmented lesions, which was not probable, especially in a young female. A definite diagnosis can be achieved through multiple biopsies, and early treatment initiation is of immense importance.

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