Atypical clinical presentation of mycosis fungoides as a unilesional hypopigmented patch

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Introduction

Mycosis fungoides (MF), a low-grade lympho-proliferative disorder, is the most common type of cutaneous T-cell lymphoma. Diagnosis of MF can be difficult due to highly variable presentations and sometimes nonspecific nature of histological findings. Nevertheless, clinical experience is of substantial importance as MF can resemble a wide variety of skin diseases. We performed a literature review and found that MF can mimic >50 different clinical entities. We present a case report of a 24 year old female who presented with hypopigmented patch on the right shoulder for 1 year and treated as tinea corporis without much improvement. Skin biopsy was performed and immunohistology was strongly supportive of mycosis fungoides.

Case report

A 24 year old female presented with a mildly itchy ill-defined non scaly hypopigmented patch over the right shoulder for 1 year. She was treated as tinea corporis with miconazole cream for 1 month without response. The skin biopsy was performed to exclude other clinical diagnoses such as leprosy.

Skin biopsy showed heavy atypical lymphocytic infiltration in the superficial dermis and significant numbers of atypical lymphocytes going into the epidermis and forming multiple Pautrier's micro-abscesses in the epidermodermal junction. Immuno-markers revealed highly positive CD8 cells with less CD4 positive cells. Skin biopsy was repeated because of the aggressive nature of the histology which wasn't compatible with the clinical picture. However second biopsy also revealed same histological picture. Hence she was diagnosed as having mycosis fungoides presenting as a unilesional hypopigmented patch. Currently she is being managed with a moderate potent topical steroid. Clinic follow up has been arranged regularly to see the response and to pick up early relapse.

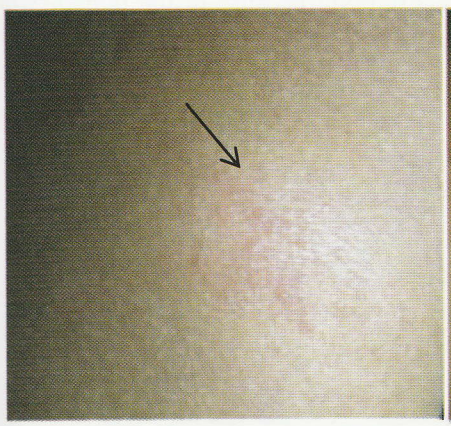

Figure 1.

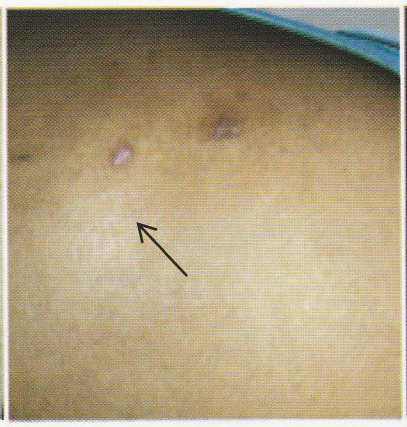

Figure 2.

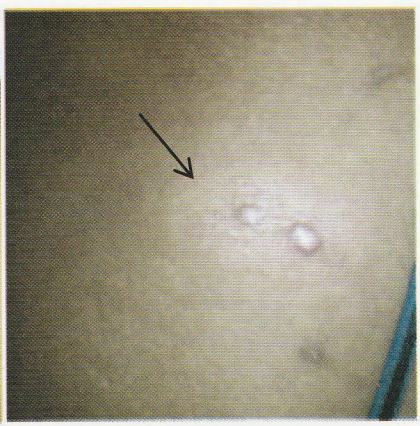

Figure 3.

Ill-defined hyopigmented patch on the shoulder (before taking biopsy, after taking 1st biopsy, before starting treatment respectively).

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Discussion

Diagnosis of MF is based on a combination of clinical presentation, histopathology and gene rearrangement. None of these factors exclusively determines the diagnosis. Histologically, MF is characterized by the presence of large atypical lymphocytes and a lymphocytic infiltrate in the papillary dermis. Clinically unusual variations of mycosis fungoides are common[1,2].

A real clinical challenge is provided by unusual and newly described variants of MF comprising hyperpigmented, hypopigmented, urticarial, bullous, solely papular, pustular and hyperkeratotic variants for which diagnosis is easier in conjunction with typical MF lesions or a positive history of MF. Hypopigmentation and a vitiligo-like outcome are predominantly described in individuals with dark skin[3]. In this subset asymptomatic ill-defined hypopigmented patches are commonly seen.

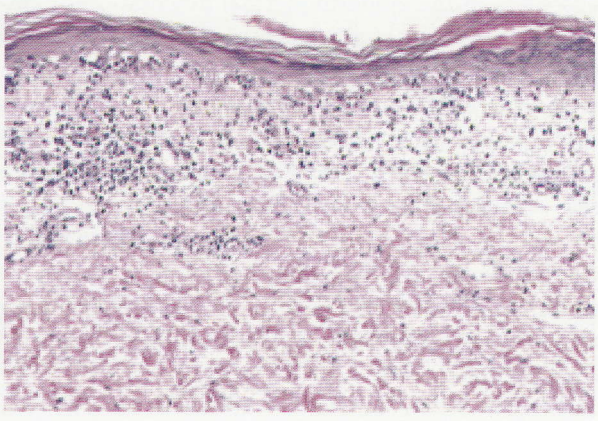

Figure 4. *Atypical lymphocytic infiltration in the epidermodermal junction and forming pautriersmicro-abscess and band like atypical lymphocytic infiltration in the superficial dermis.*

Some case reports point out single atypical lesions as a clue for a false diagnosis, e.g. a warty lesion misdiagnosed as a seborrhoeic keratosis or Bowen's disease. Other clinical impressions of atypical MF have simulated diagnoses such as purpura pigmentosa, vasculitis and pyoderma-gangrenosum[4]. However unilesional hypopigmented patch of mycosis fungoides was reported rarely in literature due to its benign clinical nature and spontaneous regression[5,9]. It also shows good response to phototherapy[10].

In every chronic skin condition resistant to treatment lymphoma has to be considered. Therefore, if lymphoma is suspected then the diagnosis must be confirmed by clinical, histological and molecular examinations perhaps repeated in the course of the disease[6]. In about 10% of cases the diagnosis can only be confirmed during the course of disease. Whole-body inspection often allows identification of typical lesions besides atypical lesions, thus guiding towards the correct diagnosis. Once lymphoma has been diagnosed a consequent follow-up should be required to detect possible recurrence and systemic involvement[8].

In our patient the lesion had been missed for a long time because skin biopsy had not been undertaken at any point because of the nonspecific and atypical presentation. We wish to emphasis

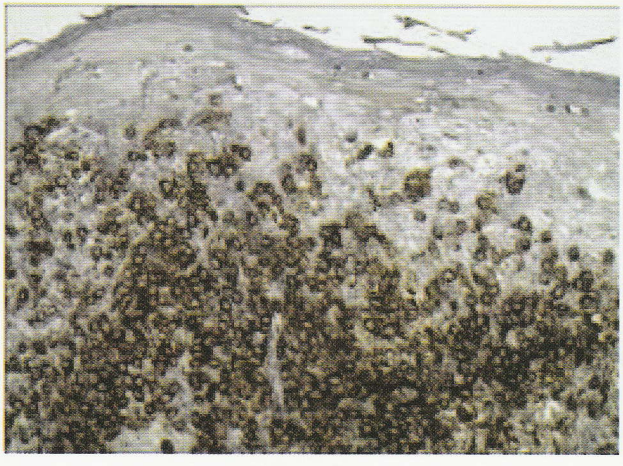

Figure 5.

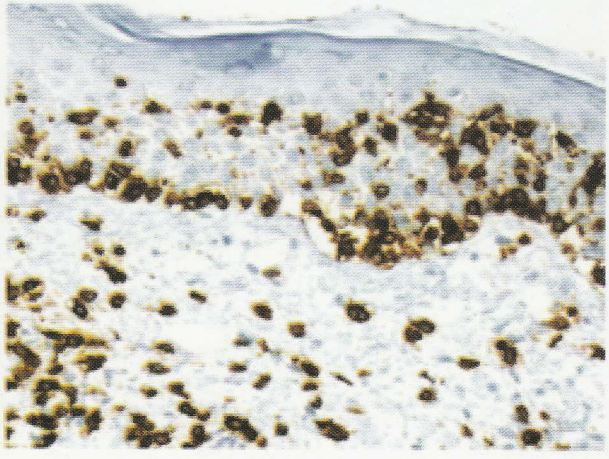

Figure 6.

Immunomarkers CD8 & CD4 respectively.

mycosis fungoides is a great imitator and should be kept in mind in any chronic non responding skin condition.

References

1. Wain EM, Setterfield J, Judge MR, *et al.* Mycosis fungoides presenting as annular erythema. *Ann Dermatol Venereol* 2005; **132**: 35-7.
2. Hubert JN, Callen JP. Mycosis fungoides involving the oral mucosa in a child. *Clin Exp Dermatol* 2003; **28**: 499-501.
3. Capella GL, Altomare. Recalcitrant tineacorporis as the presenting manifestation of patch-stage mycosis fungoides. *Cutis* 2003; **71**: 59-61.
4. Mycosis on mycosis fungoides: zoophilic dermatophytosis selectively superimposed on pre-existing cutaneous T-cell lymphoma (mycosis fungoides) plaques.
5. Ardigo M, Borroni G, Muscardin L, *et al. Mycoses* 2003; **46**: 67-70. Kazakov DV, Burg G, Kempf W. Hypopigmented mycosis fungoides in Caucasian patients: a clinicopathologic study of 7 cases. *J Am Acad Dermatol* 2003; **49**: 264-70.
6. Zackheim HS, McCalmont TH. Clinicopathological spectrum of mycosis fungoides. *J EurAcad Dermatol Venereol* 2004; **18**: 397-415.
7. Marzano AV, Berti E, Loredana L, Alessi E. Mycosis fungoides: the great imitator. *J Am Acad Dermatol* 2002; **47**: 914-18.
8. Oliver GF, Winkelmann RK. Unilesional follicular mycosis fungoides. *Dermatology* 1999; **199**: 174-6.
9. Heald PW, Glusac E. Unilesional mycosis fungoides: a distinct entity. *J Am Acad Dermatol* 1989; **20**: 63-70.
10. 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**: 283-5.