Mycosis fungoides type lymphoma in young adult: a case report

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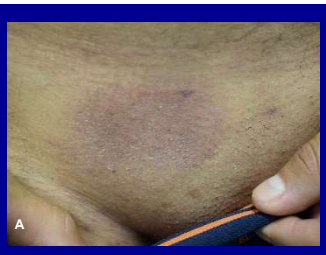
INTRODUCTION

Mycosis fungoides and its variants of primary cutaneous T-cell lymphoma (CTCL) typically occur in adults and in elderly, with a median age at 57 years. CTCL is a rare disorder in young adults with an incidence rate of 0.12 per 100 000 persons per year between ages 20 and 29 years. Diagnosis could be delayed due to its low incidence and various presentation. Accordingly to newest guidelines, skin-directed therapy (SDT) as oral 8-Methoxypsoralen plus ultraviolet A (320-400 nm, PUVA), narrow band ultraviolet B (311-312 nm) and topical mechlorethamine (nitrogen mustard, HN2) is recommended as first line treatment for patients with early stages (IA, IB, IIA) of CTCL (evidence Level 2). Also, topical HN2 is recommended to patients who received SDT in the past. We present a case report of 30-year-old male with CTCL who have received 2 years long SDT with partial response.

CASE REPORT

A patient had 10 years history of lichenoid skin patches on trunk and inguinal regions (T1, Fig.1). Histopathological examination revealed Mycosis fungoides type CTCL, plaque stage (Fig. 3) and immunohistochemical staining showed expression of rare CD8+ phenotype (Fig. 6). CD8:CD4 5:1. Also, CD3(+), CD7(-) intraepidermal and CD4/CD3/CD7/(+) intradermal lymphocytes were diagnosed. (Fig. 4-7). CD8:CD4 5:1. Imaging tests did not reveal peripheral lymph nodes (N0) or visceral organs involvement (M0) and Sezary cells were negative (B0). IA stage of CTCL were diagnosed and treatment with high potency topical steroids and PUVA therapy with 8-Methoxypsoralen 40 mg/day orally were prescribed (total 53 PUVA procedures in 2 years). The involved body surface area of affected skin has decreased from <10% to 3%. Although, after 1 month new plaques and the striae atrophicans in places treated with topical HP steroids on bilateral inguinal regions developed.



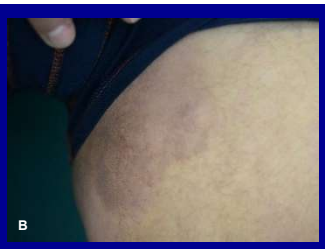


Figure 1. Clinical findings: lichenoid patches in groin (A) and inguinal (B) areas.

Figure 2. Clinical findings after high potency topical steroids and 53 PUVA procedures in 2 years: striae atrophicans in inguinal area.

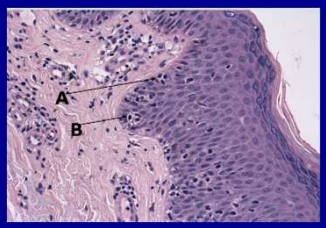


Figure 3. Histopathology findings H+Ex100: A - atypical (with cerebriform nuclei) lymphocytes; **B-** groups of atypical lymphocytes

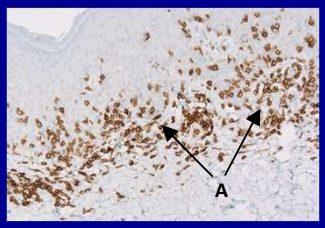


Figure 4. Imonohistochemistry findings x100: CD3 positive cells

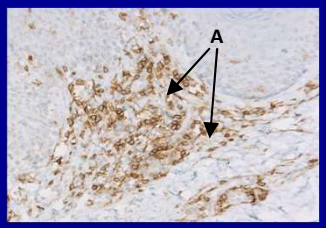


Figure 5. Imonohistochemistry findings x100: CD4 positive cells

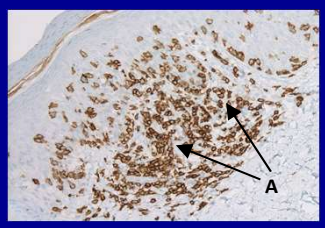


Figure 6. Imonohistochemistry findings x100: CD8 positive cells

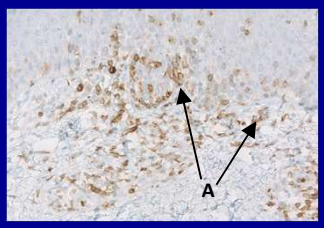


Figure 7. Imonohistochemistry findings x100: CD7 positive cells in dermis but loss in epidermis

CONCLUSIONS

Mycosis fungoides and its variants of CTCL typically occur in adults and the elderly, with a median age of 57 years (Criscione VD et al. Arch Dermatol; 2007). The incidence of disease is rare before 30 years of age, with an incidence rate of 0.12 per 100 000 persons per year between ages 20 and 29 years (Weiyun Z. Ai et al. JAMA Dermatol; 2014). According to published evidence topical mechlorethamine 0.02 gel (also known as nitrogen mustard, HN2) would be the next best choice for treatment of our patient because topical steroids and oral PUVA do not achieve an effective response.

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