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CASE REPORT

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Hyperpigmented mycosis fungoides: A case report

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Abstract

A sixty-six-years old male patient was evaluated in the dermatology outpatient clinic with extensive hyperpigmented macules and two tumoral lesions approximately three cm diameter around the umblicus. Antimycotic and corticosteroid treatment was administered. However, as the complaints increased, a diagnosis of mycosis fungoides was made as a result of a skin biopsy. Six cures of single- agent gemcitabine chemotherapy were administered by the hematology outpatient clinic and a complete response was obtained. A case of mycosis fungoides is presented to increase awareness of this rare disease.

Keywords: Mycosis fungoides, gemcitabine, complete response

Introduction

Mycosis Fungoides (MF) is a rare T cell-derived cutaneous lymphoma. It is a chronic, slowly progressing disease, and the clinical emergence and diagnosis period is several years [1]. Jean Louis Alibert named the disease for the first time in 1806 because of the fungal nodules observed during the tumor stage [2]. MF is the most common non-Hodgkin lymphoma subtype within the primary cutaneous T-cell lymphoma subgroup [2,3]. Its frequency is 4 / 100,000 cases per year in Western Europe and America [4]. Most of the cases are adults between the ages of 40-60. However, it can be seen in all age groups [4,5]. There are patch, plaque, and tumor stages (classic Albert-Bazin type). There may be a lymph node or organ involvement in the future.

Case Presentation

The sixty-six-year-old male patient had complaints of malaise and fatigue, as well as brown spots that spread all over his body within three months. It was understood that the patient, who stated that his complaints started approximately three months ago, had been applied different treatments with different diagnoses by the Dermatology Clinic, but when the lesions increased, a skin biopsy was taken. The patient whose skin biopsy result was compatible with 'Malignant Lymphoma' was referred to the Hematology Clinic. The patient, who applied to the physical therapy and rehabilitation department with knee pain that started three months ago, was diagnosed with inflammatory rheumatism, and weekly methotrexate (MTX) treatment was started. The patient's history, who underwent surgery and chemotherapy for colon cancer 15 years ago, was unremarkable. The physical examination revealed some plaques were brown, swollen from the skin, the largest of which was three cm in diameter, tending to join in patches, and tumoral skin lesions in some areas (Figure 1). We obtained no pathological finding except anemia (Hgb: 11.3 gr/dl) and sedimentation increase (63 mm / h). No significant pathology was detected in PET-CT. The flow cytometric examination performed by bone marrow aspiration sample showed $CD5^+$: 64.8, $CD3^+$, $CD4^+$, $CD7^-$ tracked on TCR alpha-beta (+). The bone marrow biopsy was reported as CD4 positive T-cell lymphoid infiltration

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(Figure 2). TNMB was accepted as Stage IIB. Gemcitabine was administered intravenously as the first cure treatment of 750 mg/m² on the 1st and 8th days. He tolerated well the first cure. Remamining each five cycles were applied 1000 mg/m², every 28 days on the 1st and 8th days. The patient is still being followed up with complete response.

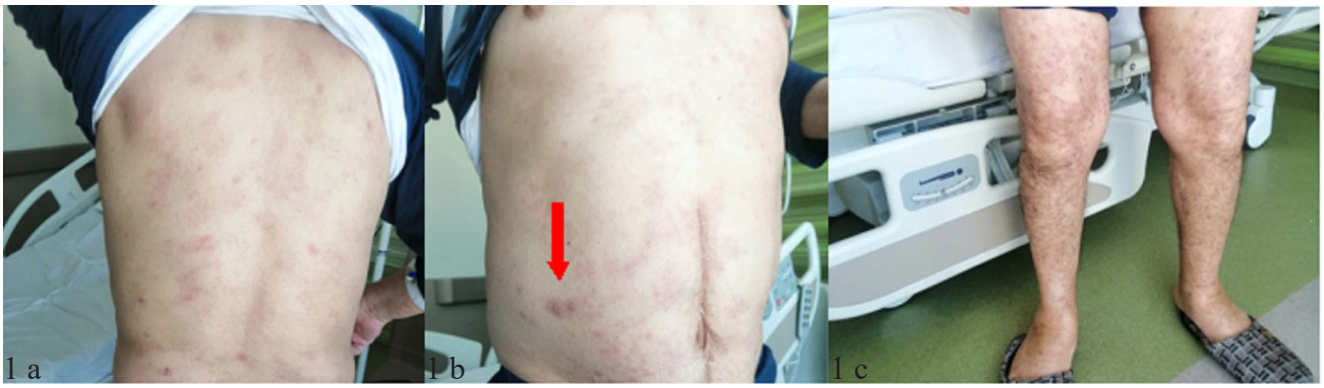

Figure 1. Diffuse plaques (a,b,c), 2 tumoral lesions of 3 cm in the lateral part of the right lower abdomen (b)

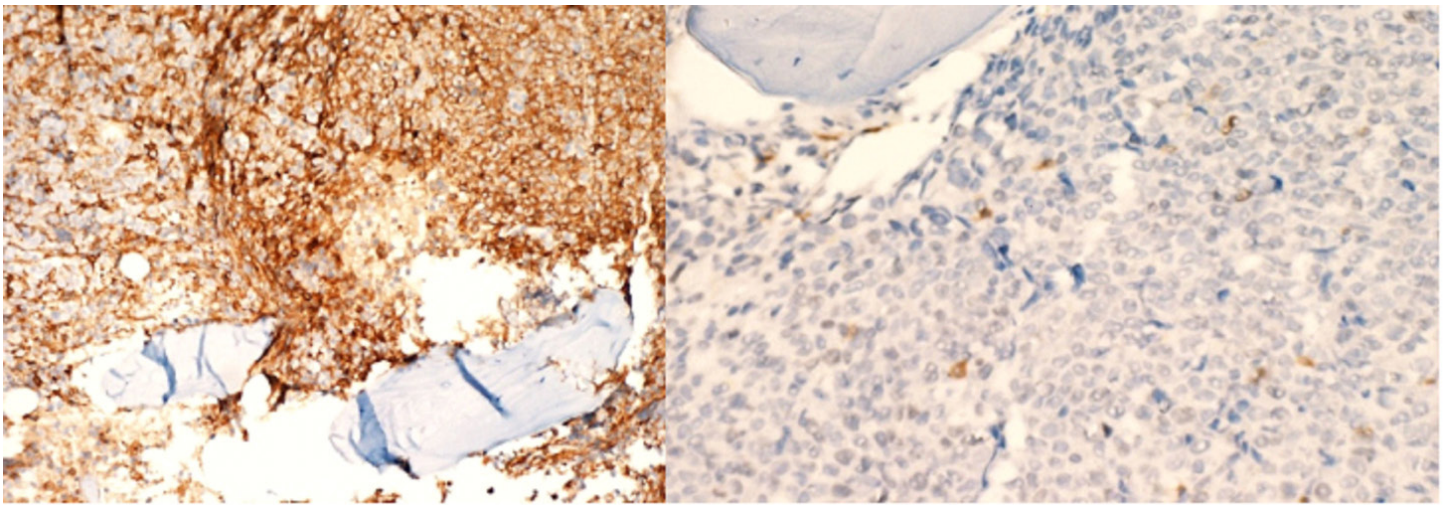

Figure 2. Atypical T cell infiltration in the upper dermis and epidermis

Discussion

There may be difficulties in the diagnosis of MF. Clinical identification, pathological evaluation, and molecular studies are essential at the diagnosis stage. Its clinical presentation is similar to that of many benign inflammatory dermatoses. In early-stage disease, it may not be diagnosed despite multiple biopsies [6]. An algorithm has been established for early diagnosis, although it has not been officially confirmed. This algorithm emphasizes the importance of clinical appearance (permanent, progressive patches or plaques in non-sun exposure areas and morphology), histopathology (superficial lymphoid infiltrate, non-spongious epidermotropism, lymphoid atypia), and immunopathology (decreased expression of CD5 and CD7) [7]. There are three clinical morphologies in MF: patch, plaque, and tumor [8,9]. There are three MF subtypes defined by the World Health Organization (WHO / European Organization for Research and Treatment of Cancer (EORTC) with various clinical and histological features [10]. Clinicians should also be aware of several different clinicopathological variants of MF [11]. The staging of MF was initially been created by the MF Cooperative Group of the American Joint Cancer Committee [8]. Later, the International Society of Cutaneous Lymphomas (ISCL) and EORTC proposed a revision in the staging criteria in 2007 and was later approved in a single-center cohort of 1502 patients [9].

The National Comprehensive Cancer Network (NCCN) has adapted the revised ISCL / EORTC recommendations for MF staging. The clinical stage is an essential determinant of disease progression and overall survival [12]. Skin-directed treatment are preferred in the early stages of the disease's treatment which is classified as local and systemic therapies. Local agents are corticosteroids, Mechlorethamine hydrochloride, carmustine,topical retinoids,topical imiquimod while phototherapy UVB,UVA or photochemotherapy (PUVA) are systemic agents. Systemic immunomodulators or systemic chemotherapies come into question in the advanced stages. Systemic immunomodulators are interferone alfa and gamma,denileukin diftitox, low dose methothrexate and HDAC inhibitors. Systemic chemotherapautics are classified as first and second stage agents. Gemcitabine, lyposomal doxorubicin are first stage agents whereas etoposide, high dose methotrexate, pentostatin,chlorambucil,cyclophosphamide are second stage drugs. In addition bone marrow transplantation can be an option for selected cases. Depending on the stage and clinical worsening, firstly single and then combined treatments should be considered [13-16].

Our patient diagnosed with stage IIB MF, we achieved a complete response with a single agent gemcitabine. The diagnosis and management of MF may still be tricky. We conclude that cooperation between various disciplines is essential for early diagnose and increasing awareness.

Conflict of interests
The authors declare that they have no competing interests.

Financial Disclosure
All authors declare no financial support.

Patient Informed Consent
Consent form was obtained from the patients before the article.

References

1. Lauren C Pinter-Brown, Emmanuel C Besa. Cutaneus T-Cell Lymphoma. WebMD. 2011;3:236-9.
2. Hoppe RT, Wood GS, Abel EA. Mycosis Fungoides and Sezary Syndrome: Pathology, staging, and treatment. Curr Probl Cancer. 1990;1:293-5.
3. Sanjay Vinjamaram, Emmanuel C Besa. Lymphoma Non-Hodgins. WebMD 2011; 2:1-4.3.
4. Joseph M. Connors, Erick D, Francine M. Foss. Lymphoma of the Skin. Am Society Hematology. 2012;1:263-78.
5. Elder D, Elenitsas R, Jaworsky C, et al. Lever's histopathology of the skin. New York. Lippincott raven Press. 1996.
6. Zain T, Grinkemeyer MD. Mycosis fungoides: Cutaneous T eel/ Am f'am Physician. 1991;43:1703-7.
7. 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;7:308158.
8. Pimpinelli N, Olsen EA, Santucci M, et al. Defining early mycosis fungoides. J Am Acad Dermatol. 2005;53:1053-63.
9. Olsen E, Vonderheid E, Pimpinelli N, et al. Revisions to the staging and classification of mycosis fungoides and Sezary 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;110:1713-22.
10. Olsen EA, Whittaker S, Kim YH, et al. Clinical end points and response criteria in mycosis fungoides and Sezary syndrome: a consensus statement of the International Society for Cutaneous Lymphomas, the United States cutaneous lymphoma consortium, and the cutaneous lymphoma task force of the European Organisation for Research and Treatment of Cancer. J Clin Oncol. 2011;29:2598-607.
11. Willemze R, Jaffe ES, Burg G, et al. WHO-EORTC classification for cutaneous lymphomas. Blood. 2005;105:3768-85.
12. Ahn CS, ALSayyah A, Sangueza OP. Mycosis fungoides: an updated review of clinicopathologic variants. Am J Dermatopathol. 2014;36:933-48.
13. Agar NS, Wedgeworth E, Crichton S, et al. Survival outcomes and prognostic factors in mycosis fungoides/Sezary syndrome: validation of the revised International Society for Cutaneous Lymphomas/European Organisation for Research and Treatment of Cancer staging proposal. J Clin Oncol. 2010;28:4730-9.
14. Olsen EA, Rook AH, Zic J, et al. Sezary syndrome: immunopathogenesis, literature review of therapeutic options, and recommendations for therapy by the United States Cutaneous Lymphoma Consortium (USCLC). J Am Acad Dermatol. 2011;64:352-404.
15. Kim EJ, Hess S, Richardson SK, et al. Immunopathogenesis and therapy of cutaneous T cell lymphoma. J Clin Invest. 2005;115:798-812.
16. Hanel W, Briski R, Ross CW, et al. A retrospective comparative outcome analysis following systemic therapy in Mycosis fungoides and Sezary syndrome. Am J Hematol. 2016;91:E491-5.