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Mycosis Fungoides: A Case Report

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Abstract First, the term Cutaneous T-cell lymphoma (CTCL) was used by Edelson to describe Mycosis Fungoides (MF) and Sézary Syndrome (SS), which are lymphoproliferative disorders. The most frequent subtype is MF that the exact etiology of the disease is unclear and there is a therapeutic challenge in treating these patients. In this study, we report the patients who initially were treated with combination chemotherapy regimens, but after a while, due to their resistance to these drugs, their treatment was changed and now, their condition is relatively favorable. Therefore, we suggest that further investigations on the prognostic factors associated with this disease are needed that are crucial in terms of response to therapy in these patients.

Keywords: *Cutaneous T-cell lymphoma, CTCL, Mycosis Fungoides*

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1. Introduction

Cutaneous T-cell lymphoma (CTCL) includes a wide range of lymphoproliferative disorders that appear in the skin [1]. The major subtypes of CTCL include Mycosis Fungoides (MF) and Sézary Syndrome (SS) that Edelson first applied the term CTCLs to describe the term, in 1974 [2]. Sézary Syndrome is the hematologic malignancy subtype of CTCL and one of its clinical features is the presence of erythroderma [3]. The most common subtype of CTCL is Mycosis Fungoides and is described by a malignant T-cell population that is limited to the skin [1]. The pathogenesis of this variant is still unclear [4]. It is assumed that T-cell proliferation, caused by long-term antigenic stimulation, may lead to the appearance of a malignant clone [5]. MF is traditionally divided based on its clinical characteristics as plaques, tumors and patches [6].

Herein, we report patients with MF who were treated with different chemotherapy regimens.

2. Case Presentation 1

A 48-year-old man with skin lesions on the abdomen and knees was admitted to the Clinic of Hematology and Oncology, Kermanshah, Iran in April 2010. The pathology report of excisional biopsy revealed an infiltration of lymphocytes into the epidermal tissue and the presence of psoriasis. Based on the peripheral blood smear that no Sezary cell was identified and immunohistochemistry (IHC) findings below, the final diagnosis was CTCL and its type was the mycosis fungoides syndrome with IB stage (T2 N0 M0 B0) according to the modified tumor-node-metastasis-blood (TNMB) classification. LCA, CD20, CD3, CD45, CD8 and Ki67 were positive but CD4 and CD30 were negative. Also, the results of the ultrasound of the liver and spleen were normal. Thus, treatment with interferon-α was started for him and the patient had a relatively good response to treatment. Subsequently, the patient was under PUVA (psoralen and ultraviolet A) therapy for 10 sessions. Due to appropriate patient conditions and normal blood test results he was continuously followed up for 3 years. In February 2016, the patient referred to the clinic for a decrease in WBC count. His treatment with gemcitabine was performed for 5 courses. Currently, the patient is undergoing a combination chemotherapy with ifosfamide, cisplatin, and etoposide (ICE) for 6 courses and his treatment is still ongoing.

3. Case Presentation 2

In September 2014, a 53-year-old woman was referred to the clinic with symptoms such as rash and skin lesions on the abdomen. By biopsy of the lesions, the results showed mild fibrosis with infiltration of the abnormal skin lymphocytes, which was diagnosed as mycosis fungoides that was continuous follow-up by the doctor's assessment.

In January 2016, the patient referred to the clinic with large lymph nodes around the neck and the doctor requested a biopsy of the lymph nodes. According to the results of the IHC, which are listed below, the diagnosis of her disease was mycosis fungoides syndrome with IIA stage. The tumor cells were positive for CD45, CD15, CD3 and CD20 and negative for CK and HMB45. Therefore, the patient was subjected to with a combination chemotherapy with cyclophosphamide, doxorubicin, vincristine and prednisone (CHOP) along with etoposide for 6 courses. During this time, the results of thyroid scan and bone marrow examination were normal. Also, viral

tests were negative for EBV, HBV and HCV. Then, the patient underwent radiotherapy and maintenance therapy. After 3 months, due to the decline in WBC (2.300/μL), which was the result of the patient's resistance to treatment, she was simultaneously treated with bexarotene and prednisolone. At the moment, she has a promising condition and is undergoing their therapeutic courses.

4. Discussion

Cutaneous T cell lymphoma (CTCL) is a category of varied non-Hodgkin lymphomas which demonstrates the malignancy of T cells in the skin [7]. This lymphoma includes two subtypes of mycosis fungoides and Sezary syndrome that 65% of the CTCL is mycosis fungoid [8]. First, MF was reported by a French doctor in a 56-year-old man in 1806 [9]. The risk of MF in men is twice as high as women and the average age of the diagnosis is 55 years and incidence of this disease is about 0.4 per 100,000 person-years [10,11]. However, the main pathogenesis of this disease is unknown, but according to recent studies, many factors can be affected in the pathogenesis of this disease, including dysfunction of some genes such as TOX or disturbance in signaling pathways such as Notch or the SMARC gene family affecting changes in histones and DNA methylation [2,12]. Although the MF is untreatable, effective steps have been taken to control it, which depends on the stage of the disease [13] Skin directed therapies are used usually in early-stage disease which includes PUVA, UVB (ultraviolet B), topical bexarotene, radiotherapy, nitrogen mustard (such as ifosfamide), carmustine, topical corticosteroids and phototherapy. HDACi (histone deacetylase Inhibitors), Localized radiotherapy, systemic chemotherapy, monoclonal antibodies and TSEB (total skin electron beam) are treatment options in advanced-stage disease [14,15]. Interferon-α can be used in both early and advanced stages of the disease, which is usually applied with PUVA in early-stage of treatment [16]. In this study, we introduced two cases with mycosis fungoides who were in the early stages. Therefore, the necessary treatments were performed for cases 1 and 2 with interferon-α, PUVA and CHOP along with etoposide, respectively, but after a while, the treatments did not respond. So, we did use systemic therapies that associated with the refractory or resistant disease after therapies. We suggest that further investigations on the prognostic factors and pathogenesis of associated with this disease are needed that which plays an important role in the treatment of these patients.

5. Conclusions

The importance of our report is in association with selected therapeutic strategies consistent with the development of MF. In recent years, advances in understanding the pathophysiology of the disease have led to a greater conception of this syndrome, which can help in better diagnosing the disease, staging, and choosing therapies. However further investigation is needed in this area.

References

[1] Bishop BE, Wulkan A, Kerdel F, El-Shabrawi-Caelen L, Tosti A. Nail Alterations in Cutaneous T-Cell Lymphoma: A Case Series and Review of Nail Manifestations. Skin appendage disorders. 2015; 1(2): 82-6.

[2] Bagherani N, Smoller BR. An overview of cutaneous T cell lymphomas. F1000Research. 2016; 5.

[3] Mao X, Lillington DM, Czepulkowski B, Russell-Jones R, Young BD, Whittaker S. Molecular cytogenetic characterization of Sezary syndrome. Genes, Chromosomes and Cancer. 2003; 36(3): 250-60.

[4] Girardi M, Heald PW, Wilson LD. The pathogenesis of mycosis fungoides. New England Journal of Medicine. 2004; 350(19): 1978-88.

[5] Slodownik D, Moshe S, Sprecher E, Goldberg I. Occupational mycosis fungoides–a case series. International Journal of Dermatology. 2017.

[6] Akinbami AA, Osikomaiya BI, John-Olabode SO, Adediran AA, Osinaike O, Uche EI, et al. Mycosis fungoides: case report and literature review. Clinical medicine insights Case reports. 2014; 7: 95-8.

[7] Olsen EA. Evaluation, diagnosis, and staging of cutaneous lymphoma. Dermatologic clinics. 2015; 33(4): 643-54.

[8] Liu K-L, Shen J-L, Yang C-S, Chen Y-J. Mycosis fungoides in a referral center in central Taiwan: A retrospective case series and literature review. Dermatologica Sinica. 2014; 32(3): 148-53.

[9] Thestrup-Pedersen K, Kaltoft K. Genotraumatic T cells and cutaneous T-cell lymphoma. A causal relationship? Archives of dermatological research. 1994; 287(1): 97-101.

[10] Kuzel T, Roenigk Jr H, Rosen S. Mycosis fungoides and the Sézary syndrome: a review of pathogenesis, diagnosis, and therapy. Journal of clinical oncology. 1991; 9(7): 1298-313.

[11] Keehn CA, Belongie IP, Shistik G, Fenske NA, Glass LF. The diagnosis, staging, and treatment options for mycosis fungoides. Cancer Control. 2007; 14(2): 102.

[12] Chung CG, Poligone B. Cutaneous T cell lymphoma: an update on pathogenesis and systemic therapy. Current hematologic malignancy reports. 2015; 10(4): 468-76.

[13] Spencer A, Gazzani P, Gadvi R, Hughes S, Stevens A, Shah F, et al. Computed Tomography scanning in mycosis fungoides–optimising the balance between benefit and harm. British Journal of Dermatology. 2017.

[14] Prince HM, Whittaker S, Hoppe RT. How I treat mycosis fungoides and Sézary syndrome. Blood. 2009; 114(20): 4337-53.

[15] Devata S, Wilcox RA. Cutaneous T-cell lymphoma: a review with a focus on targeted agents. American journal of clinical dermatology. 2016; 17(3): 225-37.

[16] Kuzel TM, Gilyon K, Springer E, Variakojis D, Kaul K, Bunn Jr PA, et al. Interferon alfa-2a combined with phototherapy in the treatment of cutaneous T-cell lymphoma. JNCI: Journal of the National Cancer Institute. 1990; 82(3): 203-7.