Journal of the Turkish Academy of Dermatology
eISSN 1307-394X

Case Report DOI: 10.6003/jtad.17113c2

Mycosis Fungoides as Pruritic Dermatitis

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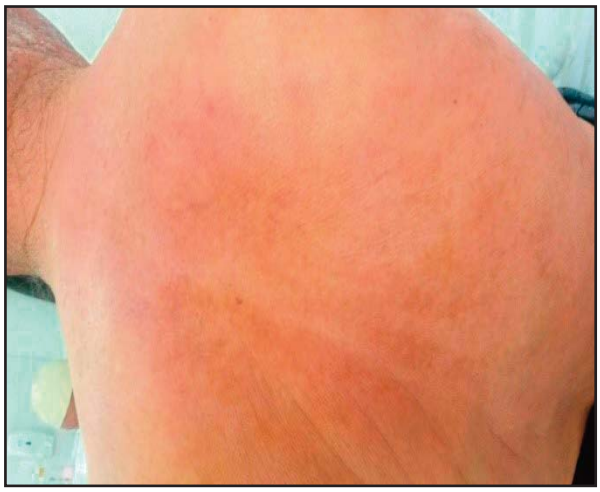
Published:
J Turk Acad Dermatol 2017;**11 (3)**: 17113c2
This article is available from: http://www.jtad.org/2017/3/jtad17113c2.pdf
Key Words: Mycosis fungoides, T cell lymphoma, pruridermatitis

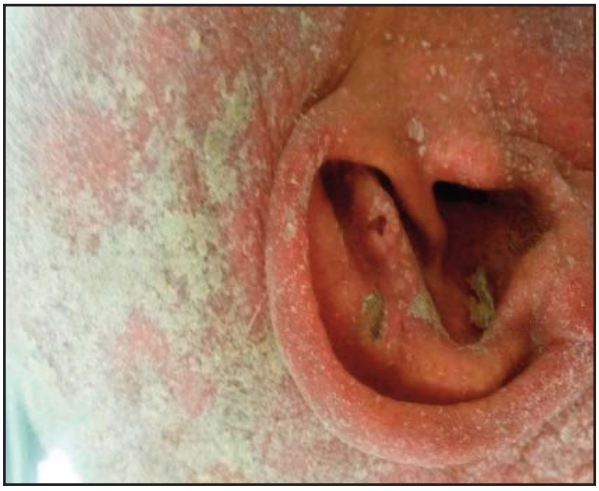
Abstract

Observation: Mycosis fungoides is a type of T cell lymphomas, that primary shows in skin. It was first described by Albert in 1806. The cause is unknown, but there have been theories relating to viral, chemical and genetic factor that affects the immune system by causing stimulation of the lymphocytes during a long-period. Since the abnormal T cells are gathered together in the skin, the skin shows many signs and can be misdiagnosed for symptoms such as: psoriasis, urticaria, and mycotic infections or especially atopic dermatitis. This fact can lead to a delay of the diagnosis. The diagnosis is set up by performing a biopsy and imunohistochimie, but in some cases the biopsy does not solve the real problem. The treatment is based on the specific stage in which the disease is currently. Topical chemotherapy, topical retinoid and corticosteroids, PUVA therapy, fusion proteins, extra corporal, photopheresis and systemic chemotherapy are some of the options. In this paper, we present the case of a 74-year-old man presented with a one year history of the pruritic dermatitis treated by the dermatologist.

Introduction

For many years, mycosis fungoides was wrongly named and linked with mycosis infections. In fact, a mycosis fungoides is T-cell lymphoma typically manifested with the uncontrolled growth of the T cell lymphocytes in the skin [**1, 2, 3, 4, 5, 6, 7, 8**]. It is a rare condition, but must commonly located in a group of lymphomas. The cause is unknown [**1,2**]. It affects all age groups but is mostly present in the age groups of 40 to 60 years old. The most often symptoms are itchy skin and a skin rush, not so characteristic, and mostly misdiagnosed with other types of symptoms such as: eczema like, where it slowly attains a plaques form and sometimes affects the complete skin, manifesting itself like a special form of erythroderma or Sezary Syndrome [**9, 10**]. In some cases, the condition can turn to a rare skin condition showed with tumours and ulcers [**10**]. Here we present a 74 years old man, treated as very resistant and intensive pruritic dermatitis for one year and eczema for many years, while finally diagnosed with mycosis fungoides after biopsy and imunohistochimie had been performed on the patient.





Figures 1a and b. Eritematous end squamous lesions

Case Report

We present the case of 74 years old man, referred to our clinic like a resistant form of pruritic dermatitis (**Figures 1 a and b**). The patient had a rush like eczema for nearly 20 years, verrucous changes on the hands as well as the psoriasis like formation changes in a head and elbows. The patient was treated many times with antibiotics, antihistaminic and even with corticosteroids – however, there were no positive results. In his family history, the patient did mention that his brother died from leukaemia. The skin biopsy was performed and the finding of the histopathologist was as below : It was taken a skin sample (1 X 0,5 X 0,2). Epidermis shows a sign of acanthosis, hypogranulosis and hyperkeratosis; in papillary and reticular dermis, there are atopic lymphocytes with a large hypochromic nucleus and perinuclear hauls grouped around the blood vessels(**Figure 2**). There is an extra stroma with non-regular recap.

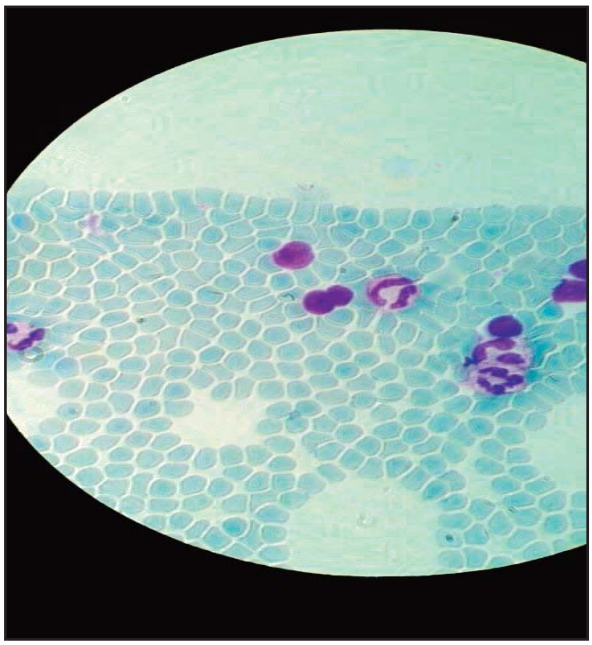


Figure 2 Peripherial blood smear in mycosis fungoides

After the result was confirmed an immunehistochimie was also performed and showed typical lymphocytes immune positive in CD2, CD3, CD4, CD5 and immune negative in CD20, CD79a and PAX-5. The patient was transferred to the department of haematology for further treatment. The treatment in haematology consisted with CHOP therapy (cyclophosphamide, Vincristine, Doxorubicin and Pronisone) and improvement in his condition was achieved even in subjective symptoms especially where there was no itching at all.

Discussion

Mycosis fungoides is a form of cutaneous T cell lymphoma. There are four stages of the disease: 1. Patch stage: mostly eczema that show signs that affect any part of the skin, causing sometimes very intensive itching; 2. Plaque phase: infiltrative stage with huge areas of the skin with intensive redness; 3. Tumor stage, with tumors that can be in any phase including exulceration and inflammation [**11, 12, 13, 14**]. For the right diagnosis the following tests and procedures may be used: physical examination and the lymph node palpation, blood examination (CBC) and peripheral blood smear [**11**], TCR (T-cell receptor) test and flow cytometry. Mycosis fungoides

and the Sézary syndrome are the two most common types of cutaneous T-cell lymphomas [**10**]. Typical skin sign is intensive red rash in a skin, and the intensity of the skin signs depends on a faze [**1**]. After the diagnosis is made there are some procedures and treatments that are established to relive the symptoms, and it mostly depends on a phase of the disease and the metastases in a lymph nodes and other organs. Skin-directed therapies includes: corticosteroids, retinoid, or imiquimod , topical chemotherapy [**8**], local radiation [**5**], methotrexate, photopheresis, phototherapy [**5,6**].The newest data suggests also a biologic treatment [**3**] under the strict control of hematologist and oncologists.

Conclusion: Mycosis fungoides is T cell lymphoma with a skin involvement and since the skin problems always cause lost of camphor causing itching and in some cases the pain in the skin the patient will in most of the cases seek help from the dermatologist. The dermatologist should always think about mycosis fungoides in a case where there is persistent itching and skin redness, the therapy resistant and persistent eczema as well. It may save the patient life if we consider the possibility of mycosis fungoides and try to rule on the diagnosis by right steps.

References

1. Willemze R, Jaffe ES, Burg G, et al. WHO-EORTC classification for cutaneous lymphomas. Blood 2005; 105: 3768-3785. PMID: 15692063
2. Vonderheid EC, Bernengo MG, Burg G, et al. Update on erythrodermic cutaneous T-cell lymphoma: report of the International Society for Cutaneous Lymphomas. J Am Acad Dermatol 2002; 46: 95-106. PMID: 11756953
3. Rook AH, Yoo EK, Grossman DJ, Kao DM, Fox FE, Niu Z. Use of biological response modifiers in the treatment of cutaneous T-cell lymphoma. Curr Opin Oncol 1998; 10: 170-174. PMID: 9555542
4. Querfeld C, Rosen ST, Guitart J, Kuzel TM. The spectrum of cutaneous T-cell lymphomas: new insights into biology and therapy. Curr Opin Hematol 2005; 12: 273-278. PMID: 15928483
5. Herrmann JJ, Roenigk HH Jr, Hönigsmann H. Ultraviolet radiation for treatment of cutaneous T-cell lymphoma. Hematol Oncol Clin North Am 1995; 9: 1077-1088. PMID: 8522485
6. Zackheim HS. Cutaneous T cell lymphoma: update of treatment. Dermatology 1999; 199: 102-105. PMID: 10559573
7. Sinha AA, Heald P. Advances in the management of cutaneous T-cell lymphoma. Dermatol Clin 1998; 16: 301-311. PMID: 9589203
8. Ramsay DL, Meller JA, Zackheim HS. Topical treatment of early cutaneous T-cell lymphoma. Hematol Oncol Clin North Am 1995; 9: 1031-1056. PMID: 8522483
9. Koh HK, Charif M, Weinstock MA. Epidemiologym and clinical manifestations of cutaneous T-cell lymphoma. Hematol Oncol Clin North Am 1995; 9: 943-960. PMID: 8522490
10. Van Doorn R, van Kester MS, Dijkman R, et al. Oncogenomic analysis of mycosis fungoides reveals major differences with Sezary syndrome. Blood 2009; 113: 127-136. PMID: 18832135
11. Hoque SR, Child FJ, Whittaker SJ, et al. Subcutaneous panniculitis-like T-cell lymphoma: a clinicopathological, immunophenotypic and molecular analysis of six patients. Br J Dermatol 2003; 148: 516-525. PMID: 12653744
12. Massone C, Chott A, Metze D. et al. Subcutaneous, blastic natural killer (NK), NK/T-cell, and other cytotoxic lymphomas of the skin: a morphologic, immunophenotypic, and molecular study of 50 patients Am J Surg Pathol 2004; 28: 719-735.
13. Burg G, Kempf W. Etiology and pathogenesis of cutaneous lymphomas. Burg G, Kempf W. Cutaneous Lymphomas. London: Taylor & Francis 2005.
14. Burg G, Kempf W, Cozzio A, et al. Cutaneous malignant lymphomas: update 2006. J Dtsch Dermatol Ges 2006; 4: 914-933. PMID: 17081267