A Case of Mycosis Fungoides In An Elderly Male

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INTRODUCTION

Mycosis fungoides is the most common form of cutaneous T cell lymphoma.

Cutaneous lymphomas are a heterogeneous group of non-Hodgkin lymphomas of T- and B-cell origin. This is a case of an 89-year male who was diagnosed and managed as a case of eczema; however, further investigations confirmed a diagnosis of Mycosis fungoides. This condition could be difficult to diagnose in the elderly population due to the variety or possible presentations and subtleties of histopathological correlation in the early stage.

Case Report

89-year-old male presented to our practice with a painful and pruritic eruption that has been treated by his primary care physician for the last year. Patient stated that a biopsy was performed and a diagnosis of eczema was made. He was treated with triamcinolone acetonide 0.1% ointment twice daily with only slight improvement of his symptoms. Upon review of his records, we discovered that a shave biopsy had been done, which showed a predominantly chronic inflammatory infiltrate and spongiosis, most consistent with an acute allergic or irritant contact dermatitis.

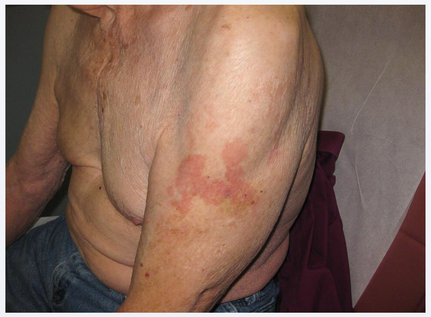
Patient's other medical conditions included high blood pressure, hyperlipidemia as well as coronary artery disease. He had no prior history of dermatologic disease. Examination revealed an elderly male in no acute distress and good spirits. The patient was found to have a diffuse eruption consisting of erythematous polycyclic plaques with slight scale on both of his upper extremities, abdomen, trunk, lower extremities, and groin. (Figure 1) Mild bilateral lymphadenopathy was discovered on exam.

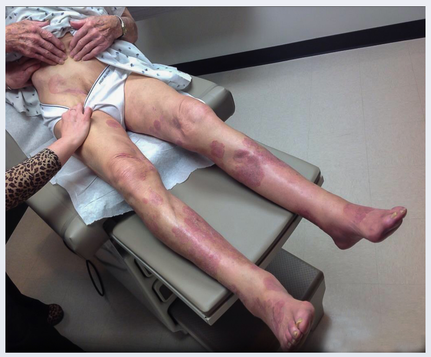
Two separate 3mm punch biopsies were performed at the time of his presentation. Both revealed an intraepidermal collection of atypical mononuclear cells with associated mild spongiosis and adjacent superficial predominantly lymphoid infiltrate consistent with cutaneous t-cell lymphoma. Further gene rearrangement studies confirmed the diagnosis of mycosis fungoides. Patient was referred to hematology-oncology for peripheral blood flow cytometry and CT imaging of the chest abdomen and pelvis. No lymph nodal enlargement by CT criteria was identified. Peripheral blood flow cytometry revealed no clonal population of T-cells. His disease was deemed primary cutaneous T-cell lymphoma staged 2A due to presence of nonmalignant bilateral inguinal lymphadenopathy. Patient has been treated with mechlorethamine hydrochride 0.016% gel and clobetasol propionate 0.05 ointment with good response. His pruritus has significantly improved since therapy was implemented.

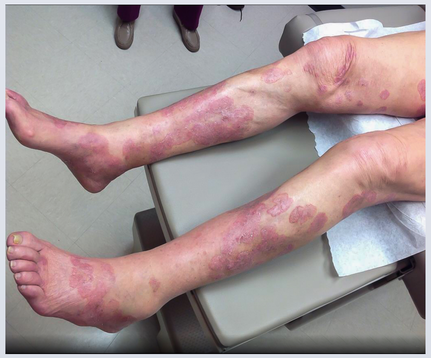
Discussion

Primary cutaneous lymphomas represent a heterogeneous group of T- and B-cell lymphomas. Mycosis fungoides (MF), which is generally indolent in behavior, and Sezary syndrome (SS), an aggressive and leukemic variant, comprise approximately 53% of all cutaneous T-cell lymphomas (CTCL). [1]

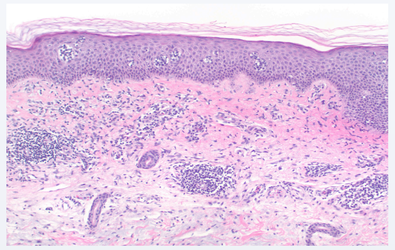
Clinical Pictures



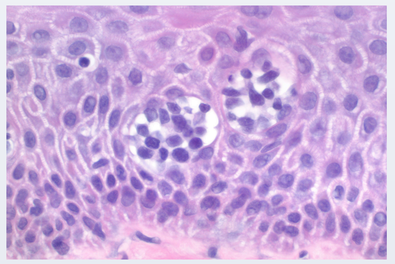




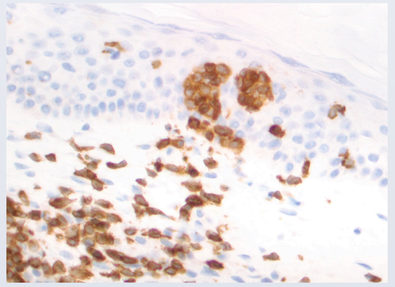
Histopathology



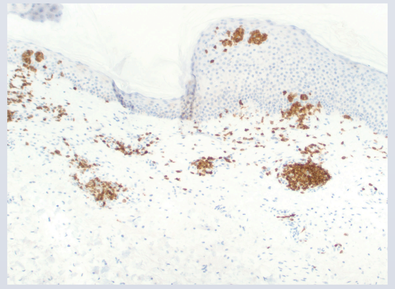
atypical mononuclear cells with associated mild spongiosis and adjacent superficial predominantly lymphoid infiltrate



intraepidermal collection of atypical mononuclear cells



Immunohistochemical staints showing CD4 + T-cells



Immunohistochemical staints showing CD3+ T-cells

Discussion

The incidence of CTCL has risen since 1973, with an annual age-adjusted incidence of 6.4 to 9.6 cases per million people in the United States. [4]

Mycosis fungoides is mostly disease of the elderly with a median age at diagnosis of 55 to 60 years and a male: female ratio of 2:1. [1,5,7]

However, mycosis fungoides has also been seen in younger populations, including children. [5,6,7] Approximately 70% of patients with mycosis fungoides are white. African Americans, Hispanics, and Asians making up 14%, 9%, and 7% of mycosis fungoides cases in the United States, respectively. [19] Patients with both mycosis fungoides and Sezary syndrom are at a highly increased risk of developing a second lymphoma, in particular Hodgkin lymphoma and the CTCL subtype lymphomatoid papulosis, as well as nonhematologic malignancies. [4,7,14]

Mycosis fungoides classically is a very slowly progressive disease. It evolves slowly over years, sometimes decades and frequently has a relapsing course. Classic clinical presentation includes multiple, well defined, often pruritic erythematous patches distributed in non-sun exposed "bathing suit" areas, including the breasts, buttocks, lower trunk, and groin. These patches may evolve to infiltrative plaques and tumors, and all 3-lesion types can be seen concomitantly. [1,7] Hypopigmented lesions are a rare presentation of mycosis fungoides, most often seen in children, adolescents, and dark-skinned individuals. [7,19]

MF is characterized by the presence of lymphocytes that express a T cell helper phenotype (CD4+). These cells also express CD45RO, a marker of mature memory T cells. [7,16] CD8+ cytotoxic/suppressor T cell immunophenotype can be seen in rare cases, usually hypopigmented MF. [7,9,10]

For early stage MF when the disease is confined to the skin, topical skin-directed therapies are first-line treatments. Topical corticosteroids, topical nitrogen mustard (mechlorethamine hydrochride), topical retinoids (topical bexarotene), phototherapy, and total skin electron beam therapy are the mainstay of skin directed therapies. [18]

Oral bexarotene, interferon, histone deactylase inhibitors (vorinostat and romidepsin), extracorporal photopheresis, monoclonal antibody agent alemtuzumab and chemotherapy are used for more extensive or recalcitrant disease. [18]

Selected References

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