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Rapid Progression of Red Itchy Rash, Diagnostic Dilemma

Uma Keyal[1], Gopi Aryal[2], Sanju Babu Shrestha[1], Anil Kumar Bhatta[3]

[1] Department of Dermatology, Nepal Mediciti Hospital, Lalitpur, Kathmandu, Nepal
[2] Department of Pathology, Nepal Mediciti Hospital, Lalitpur, Kathmandu, Nepal
[3] Derm Dynamics Skin Hair and Laser Clinic, Kumaripati, Lalitpur, Kathmandu, Nepal

Abstract

Cutaneous T-cell lymphoma (CTCL) is an uncommon type of cancer that begins in white blood cells called T lymphocytes (T cells). Normally, T cells help body's immune system to fight off infection but, in CTCL, these cells develop abnormalities that make them attack the skin. It comprises heterogeneous group of skin neoplasms, the most common being mycosis fungoides (MF). Skin lesions in MF are classified into patches, plaques, and tumor stage. Extra skin manifestations like enlarged lymph nodes are usually seen in plaques or tumor stage, when there are thick lesions on skin. It is usually a slow growing cancer that develops over many years. Herein, we present an 83-year female patient who suddenly developed extremely itchy patches on abdomen, which had spread to involve the entire trunk and all four limbs in about 10 days' time. She already had lymphadenopathy and splenomegaly at the time of presentation. Skin biopsy and immunohistochemistry revealed CD4+ T cells CTCL. Moreover, bone marrow was hypercellular for age with 12% atypical lymphocytes. It is very unusual for CTCL to involve bone marrow at patch stage with no any plaques or nodules on the skin. Also, the sudden onset of skin lesions and its rapid progression with probably, the involvement of lymph nodes and spleen warrant further studies to guide diagnostic approaches and treatment recommendations for CTCL.

Key words: Cutaneous T cell lymphoma; Mycosis fungoides; Skin neoplasm; Lymphocytes

Introduction

Cutaneous T-cell lymphoma (CTCL) describes a heterogeneous group of neoplasms of skin homing T cells that vary considerably in clinical presentation, histologic appearance, immunophenotype, and prognosis.[1] Mycosis fungoides (MF) is the most common variant of CTCL with predilection for males (2: 1). Any age group may be involved, but there is a higher incidence in the fourth to sixth decades. It is more common in blacks (2: 1) and less common in Asians and Hispanic Whites.[2] Histopathologically, MF is characterized by an epidermotropism of T lymphocytes that displays in most cases a helper phenotype. Cytotoxic variants are well described and do not have specific clinical, histopathological, or prognostic features.[3] MF is divided into 3 clinical phases: patch, plaque, and tumor stage, and the clinical course is usually protracted over years or decades. The case described here is of 83-year-old lady who presented with typical patch stage of MF but aggressive clinical behavior with extra skin manifestations like lymphadenopathy and splenomegaly created diagnostic dilemma.

Case report

An 83-year-old lady with known case of hypothyroidism and chronic obstructive pulmonary disease (COPD) was admitted in ICU with a diagnosis of acute exacerbation of COPD with acute kidney injury. Dermatology department was approachedfor consultation for complain of itchy skin rash since about 10 days. On dermatologic examination, there were non-scaly erythematous macules and patches covering the entire trunk and limbs(Figure 1A,1B). At first, we thought it could be a drug eruption but, after reviewing her laboratory reports, we performed a thorough clinical examination. The laboratory findings

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CorrespondingAuthor:
Dr. Anil Kumar Bhatta
Consultant Dermatologist
Derm Dynamics Skin Hair and Laser Clinic, Kumaripati, Lalitpur, Kathmandu, Nepal
ORCID ID:000-0002-9978-8816
E-mail: dr4yourskin@gmail.com

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showed increase in total leucocyte count (35250 cells / Cumm (4000-11000)) with predominating lymphocytes (polymorphs 13.60%, lymphocytes 76-70%), decrease in hemoglobin (8.3 gms% (11.9-14.6)), and platelet count (70000 cells/Cumm (150000-450000)). Peripheral blood smear (PBS) showed microcytic hypochromic red blood cells with macrocytes and target cells. Anisopiokilocytosis was seen. White blood cells were increased and atypical lymphoid cells were seen. Platelets were decreased. Hence, the impression of PBS was lymphoproliferative disorder. Serology was negative for leptospira, malarial antigen, scrub typhus, dengue, and SARS-CoV-2. On clinical examination, splenomegaly and bilateral multiple, firm to hard, non-matted axillary and groin lymph nodes about 1*1 to 2*4 cm in size was found. With these findings, we planned a skin biopsy. Simultaneously, patient was consultedby oncology department and they planned a bone marrow aspiration and immunohistochemistry test.

Bone marrow aspiration cytology reported hypercellular marrow with trilineage hematopoiesis and 12% atypical lymphocytes. In immunohistochemistry, cells were positive for CD2, CD3, CD4, CD5, CD30 and negative for CD20, CD8, CD7, CD56, ALK1 TDT, CK and Ki67 45-50% (Figure 2A,2B). Skin biopsy showed mild basket weave hyperkeratosis and thinned out epidermis. Dense band like papillary dermal and nodular periappendageal atypical lymphoid infiltrate was seen with minimal epidermal infiltration. The atypical cells were intermediate in size with indistinct cell membrane, scant cytoplasm, centrally placed nucleus with opened up chromatin (Figure 3A,3B). On basis of these findings, a diagnosis of cutaneous T-cell lymphoma (T2aN1M0B1)[4] was made and family members were informed for the same. Oncology team planned a chemotherapy but, seeing her age, comorbid conditions and poor prognosis, family members decided to take patient home and keep her on palliative care. After 1 month, when family member was contacted via telephone, we were informed that patient expired 2 days after leaving the hospital. This was atypical form of MF where skin lesions progressed very rapidly with extra cutaneous manifestations.

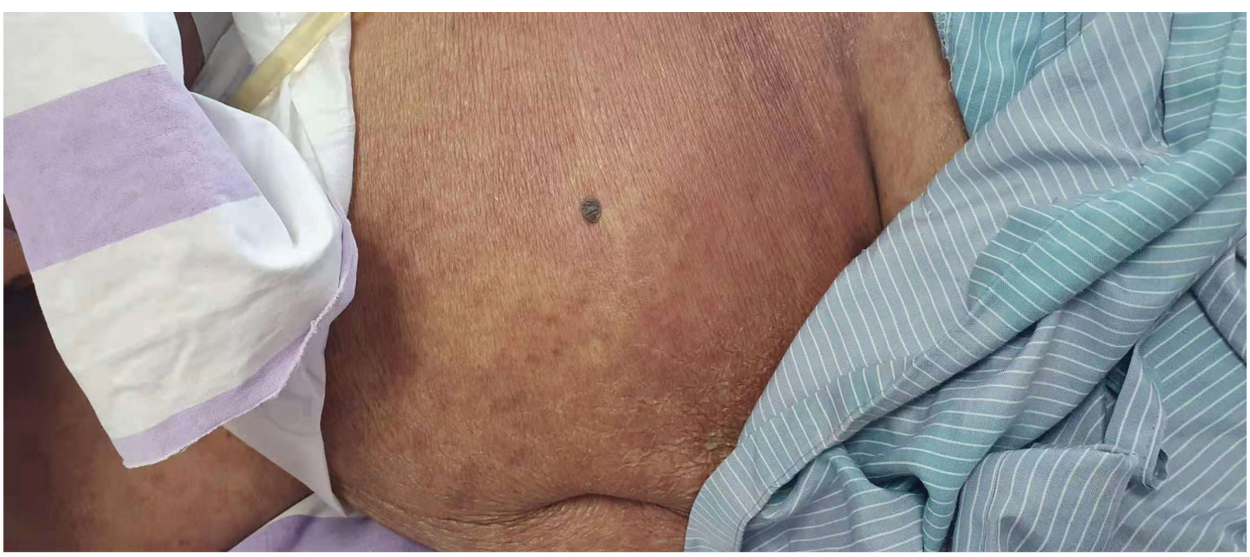

Figure 1A: Non-scaly erythematous patches on anterior trunk

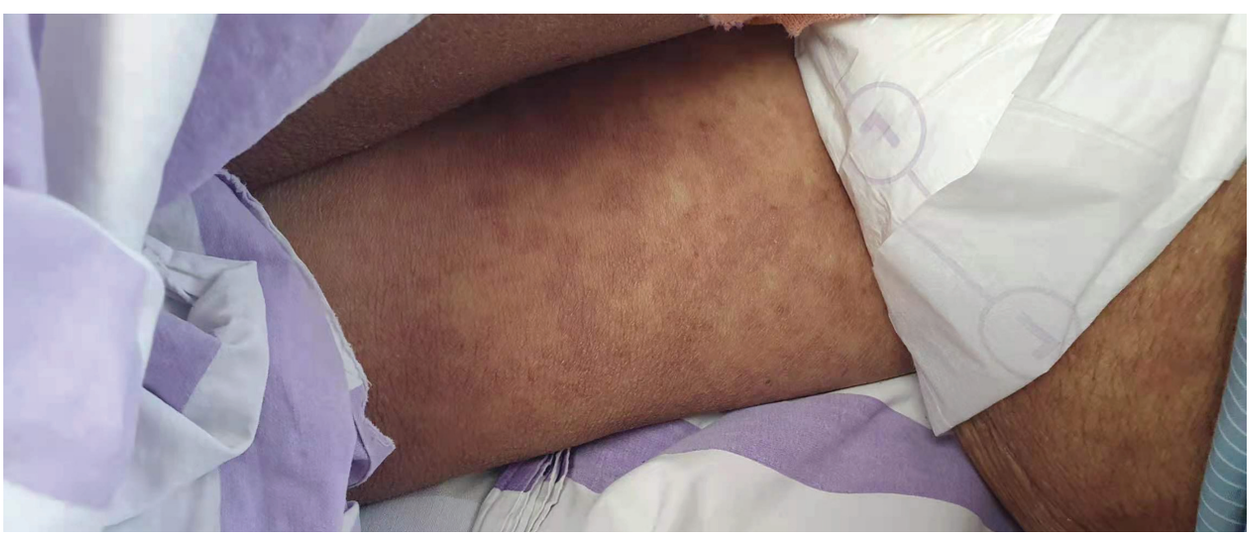

Figure 1B: Non-scaly erythematous patches on lower limbs

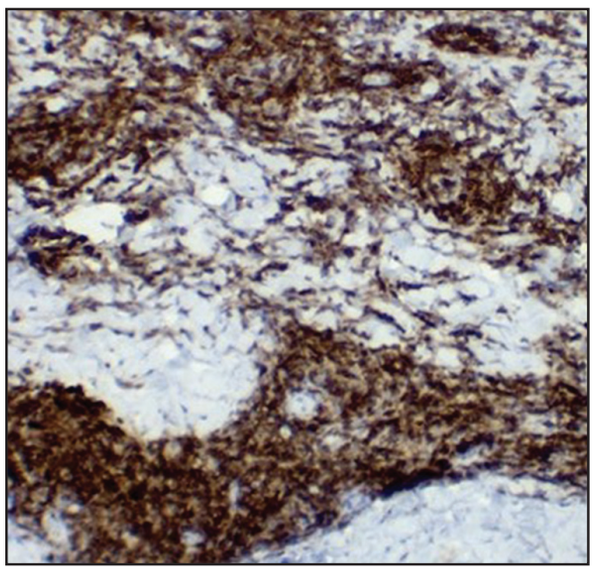

Figure 2A: Immunohistochemistry showing CD4positive cells. (10X)

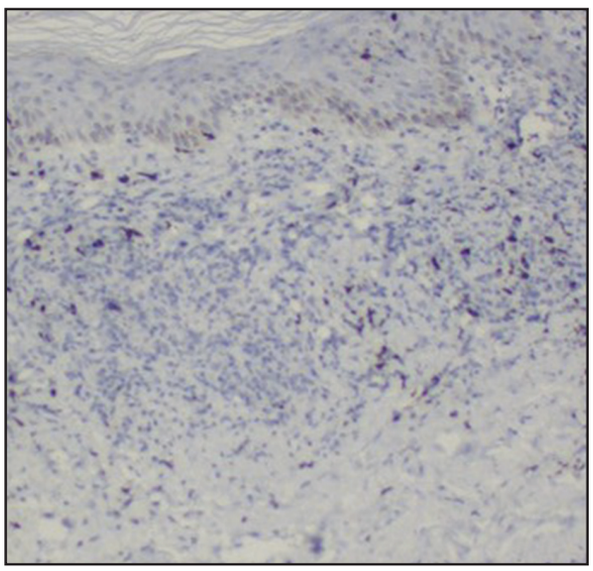

Figure 2B: Immunohistochemistry showing CD8 negative cells.(10X)

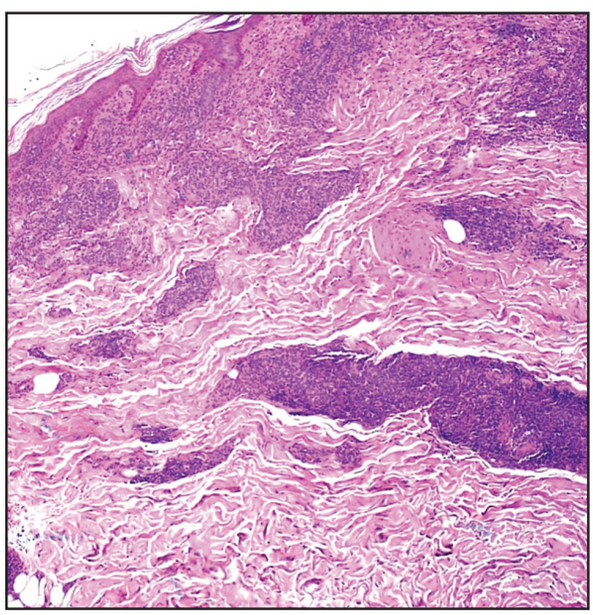

Figure 3A: Basket weave orthokeratosis and thinned out epidermis is seen. Band like lymphocytic infiltration in papillary and reticular dermis is seen. (4X)

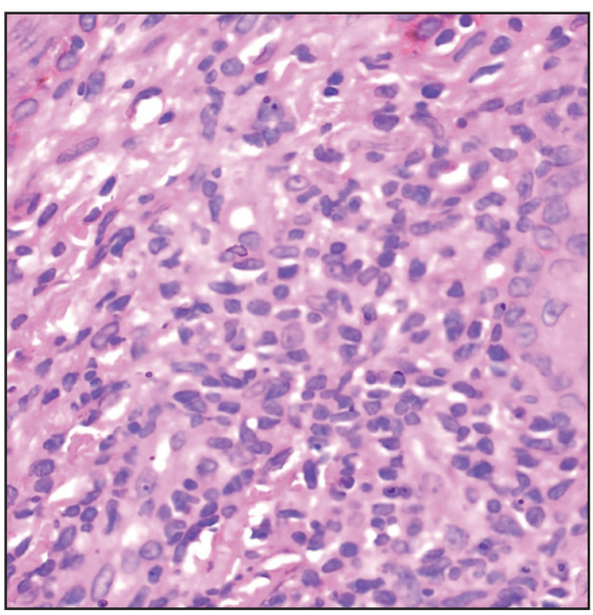

Figure 3B: Perivascular lymphocytic infiltration in dermis is seen with minimal epidermal infiltration. The atypical cells are intermediate in size with indistinct cell membrane, scant cytoplasm, centrally placed nucleus with opened up chromatin. (40X)

Discussion

Mycosis fungoides, the most common CTCL, typically presents in its early stage as inflammatory erythematous patches or plaques, with epidermotropism as the histopathologichallmark of the disease.5Often, multiple skin biopsies at intervals spanning a period of years are required to confirm the diagnosis.6 Clinically, patch stage mycosis fungoidescan mimic chronic contact dermatitis, psoriasis, or atopic dermatitis. In the case presented here, the first impression was of drug eruption because patient had multiple issues for which she was given some medicines in previous center. However, altered complete blood count, lymphadenopathy, and splenomegaly lead to aggressive work up, which helped us to make the timely diagnosis. This particular case may be another atypical variant of MF which does not fit into any previously described variants of MF. Several clinicopathologic variants of mycosis fungoides have been delineated, including poikiloderma atrophicans vasculare(parapsoriasis variegata), Sezary syndrome, granulomatous mycosis fungoides, hypopigmented mycosis fungoides, folliculocentric mycosis fungoides, syringotropic mycosis fungoides, and WoringerKolopp disease.7 Hence, it needs further moreeffort of contribution in future studies to guide diagnostic approaches and treatment recommendations for CTCL.

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