Pustular mycosis fungoides: An unusual presentation

Anakaporn Tiyawatanaroj MD,
Poonnawis Sudtikoonaseth MD,
Pinnaree Kattipathanapong MD.

ABSTRACT:

TIYAWATANAROJ A, SUDTIKOONASETH P, KATTIPATHANAPONG P. PUSTULAR MYCOSIS FUNGOIDES: AN UNUSUAL PRESENTATION. THAI J DERMATOL 2018; 34: 286-291.

INSTITUTE OF DERMATOLOGY, MINISTRY OF PUBLIC HEALTH, BANGKOK, THAILAND.

Mycosis fungoides (MF) is the most common form of cutaneous T-cell lymphoma (CTCL) generally results from clonal expansion of neoplastic T-cell lymphocytes in the skin. Classic MF presents with the typical evolution stages of patches, plaques and tumors, with potential involvement of lymph nodes, blood, and visceral organs. We reported a case of 28-year-old woman who presented with generalized pustular eruption which showed histopathologic and immunophenotyping findings of MF. Computed tomography of chest and whole abdomen revealed multiple enlarged axillary and inguinal lymph nodes. Based on skin, lymph node and bone marrow biopsy, the patient was diagnosed with pustular MF stage IVA2 (T2N3M0B0) and was referred for multi-agent chemotherapy.

Key words: Pustular mycosis fungoides, Mycosis fungoides, Cutaneous T-cell lymphoma

From: Institute of Dermatology, Ministry of Public Health, Bangkok, Thailand.

Corresponding author: Anakaporn Tiyawatanaroj MD., Email: mildkung@gmail.com

Received: 14 September 2018 Revised: 3 October 2018 Accepted: 6 November 2018

บทคัดย่อ:

อนรรฆพร ติยวัฒนาโรจน์, ปุณวิศ สุทธิกุลณเศรษฐ์, ปิ่นนรี ขัตติพัฒนาพงษ์. รายงานผู้ป่วยโรคมัยโคซีส ฟังกอยดีสชนิดตุ่มหนอง วารสารโรคผิวหนัง 2561; 34: 286-291.

สถาบันโรคผิวหนัง กรมการแพทย์ กระทรวงสาธารณสุข

มัยโคซีส ฟังกอยดีส เป็นมะเร็งเม็ดเลือดขาวที่ผิวหนังชนิดทีเซลล์ ซึ่งเกิดจากการแบ่งตัวเพิ่มจำนวนของเซลล์เม็ดเลือดขาวชนิดทีเซลล์ที่ผิดปกติในผิวหนัง อาการแสดงจะมีการพัฒนาจากระยะ patch, plaque ไปจนถึง tumor stage ซึ่งอาจพบกระจายไปยังต่อมน้ำเหลือง โลหิต รวมถึงอวัยวะภายในอื่นๆ รายงานฉบับนี้เป็นการนำเสนอผู้ป่วยหญิงไทย อายุ 28 ปี มาด้วยอาการผื่นลักษณะเป็นตุ่มหนองขึ้นกระจายทั่วลำตัว ใบหน้า แขน ขา ผลทางพยาธิวิทยาและอิมมูโนพยาธิวิทยา เข้าได้กับโรคมัยโคซีส ฟังกอยดีส ผลเอ็กซเรย์คอมพิวเตอร์บริเวณช่องอกและช่องท้องพบต่อมน้ำเหลืองโตจำนวนมากที่บริเวณรักแร้และขาหนีบ จากผลตรวจชิ้นเนื้อจากต่อมน้ำเหลือง และผลการตรวจทางพยาธิวิทยาจากไขกระดูก ผู้ป่วยรายนี้จึงได้รับการวินิจฉัยเป็นมัยโคซีส ฟังกอยดีสชนิดตุ่มหนอง ระยะที่ 4 และได้ถูกส่งตัวเพื่อทำการรักษาต่อไป

คำสำคัญ: มัยโคซีส ฟังกอยดีสชนิดตุ่มหนอง, มัยโคซีส ฟังกอยดีส, มะเร็งเม็ดเลือดขาวชนิดทีเซลล์ที่ผิวหนัง

Case report

A 28-year-old Thai woman presented with generalized pustular eruption for 1 week, with a 2-year history of non-specific dermatitis prior to presentation. She had noticed vesiculopustular eruption on her palms and soles one month before predominated pustular component. She also reported malaise, myalgia and low grade fever for a few days before the onset of eruption. There was no history of significant weight loss. The patient was otherwise healthy and not taking any medications. No relevant family history was noticed.

Physical examination revealed generalized pustules with some vesicles and erosions on ill-defined erythematous patches involving 90% of BSA (Figure 1). There was a palpable lymph node at right groin measuring 1.5x1.5 cm in size, with round, rubbery consistency, movable and non-tender. Axillary lymph nodes were not palpated at the time. There was no hepatosplenomegaly.

Bedside investigation from pustules on abdomen revealed no fungus with KOH examination and Tzanck smear showed neither acantholytic cells nor multinucleated giant cells. Laboratory investigation showed lymphocytosis without evidence of circulating atypical or Sézary cells.

Histopathologic examination from a pustule on right trunk revealed multiple foci of several atypical lymphocytic epidermotropism. Pautrier's microabscesses were also noted. The dermis showed a superficial perivascular lymphocytic infiltration with some degree of atypia (Figure 2).

Immunohistochemical studies demonstrated mainly T-cell lineage in the infiltrate by CD3, CD4, CD5, CD7 and CD8. Also there was antigenic loss in CD7 and CD8. CD1a, CD30, and CD20 were negative (Figure 3).

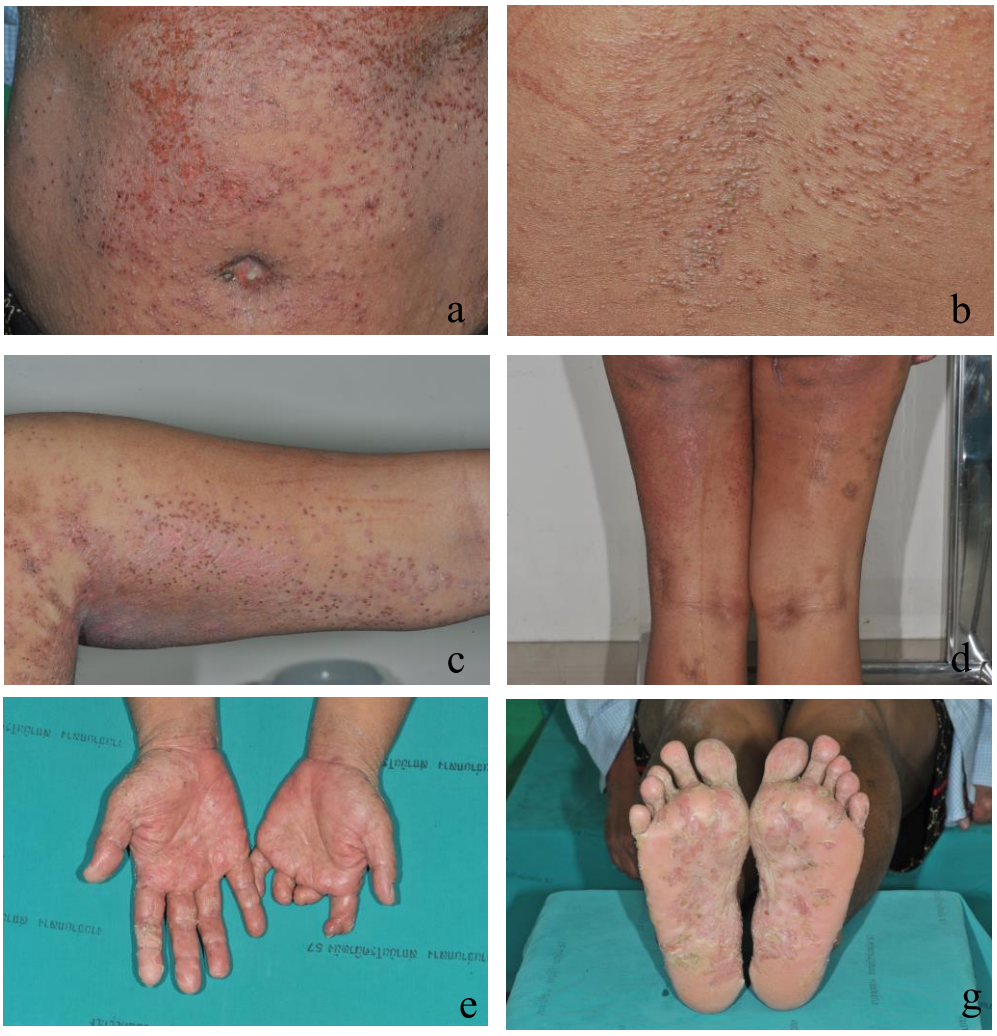

Figure 1 Physical examination revealed generalized multiple pustules with some vesicles and erosions on ill-defined erythematous patches at trunk (a, b), extremities (c, d), palms (e) and soles (g) involving 90% of BSA. Deformity of her left hand was due to an unrelated accident.

Computed tomography of chest and whole abdomen revealed multiple enlarged axillary and inguinal lymph nodes. Lymph node biopsy from right axillae and right groin showed diffuse

small lymphoid proliferation. Immunohisto-chemical studies revealed proliferating small lymphoid cells which were marked with CD3, CD4, and Ki-67 (15-20%) but not CD20, CD34, TdT, CD8, and CD30. Overall morphologic and immunophenotypic findings were suggestive of nodal involvement by mature T-cell lymphoma. Bone marrow clot and biopsy showed cellular marrow without morphological evidence of lymphoma. Based on these findings, the patient was diagnosed with pustular MF stage IVA2 (T2N3M0B0) and was referred for multi-agent chemotherapy.

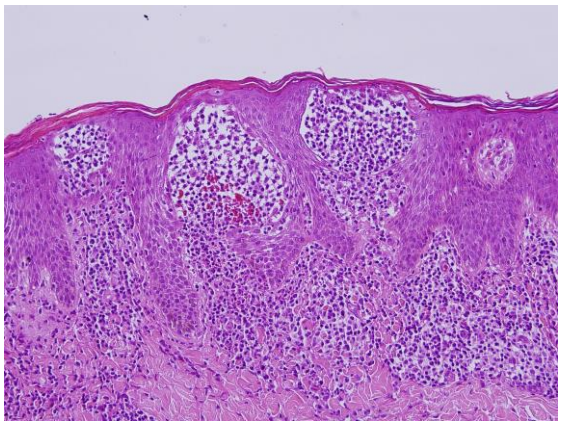

Figure 2 Histopathologic examination from pustules on right trunk revealed multiple foci of several atypical lymphocytic epidermotropism. Pautrier's microabscesses were also noted. The dermis showed a superficial perivascular lymphocytic infiltration with some degree of atypia. (H&E, 20x)

Discussion

Mycosis fungoides (MF) is the most common form of cutaneous T-cell lymphoma (CTCL), arising in mid-to-late adulthood with a slightly male predominance. It generally results from clonal expansion of neoplastic T-cell lymphocytes in the skin.[1] Classic MF presents with the typical evolution stages of patches, plaques and tumors, with potential involvement of lymph nodes, blood, and visceral organs. The median duration from onset of skin lesions to the diagnosis of MF is 4-6 years.[2]

Diagnosis of MF can be difficult due to highly variable presentations and non-specific or subtle changes in early histologic findings. MF can mimic many clinical entities and some benign conditions such as eczema, atopic dermatitis, and psoriasis. The unusual variants include: hyperpigmented, hypopigmented, urticarial, bullous and hyperkeratotic MF. Pustular MF presenting with generalized pustular eruption is one of the rare variants and may resemble other benign pustular dermatoses such as acute generalized exanthematous pustulosis (AGEP), IgA pemphigus, subcorneal pustular dermatosis of Sneddon-Wilkinson, and generalized pustular psoriasis.[3]

This patient presented with generalized pustular eruption. Generalized pustular psoriasis was initially suspected but the histologic findings and immunohistochemical studies were compatible with pustular MF. It was postulated that generalized pustulation in MF might be an immunological phenomenon, i.e. a disturbance of interleukin release, which still requires confirmation.[4]

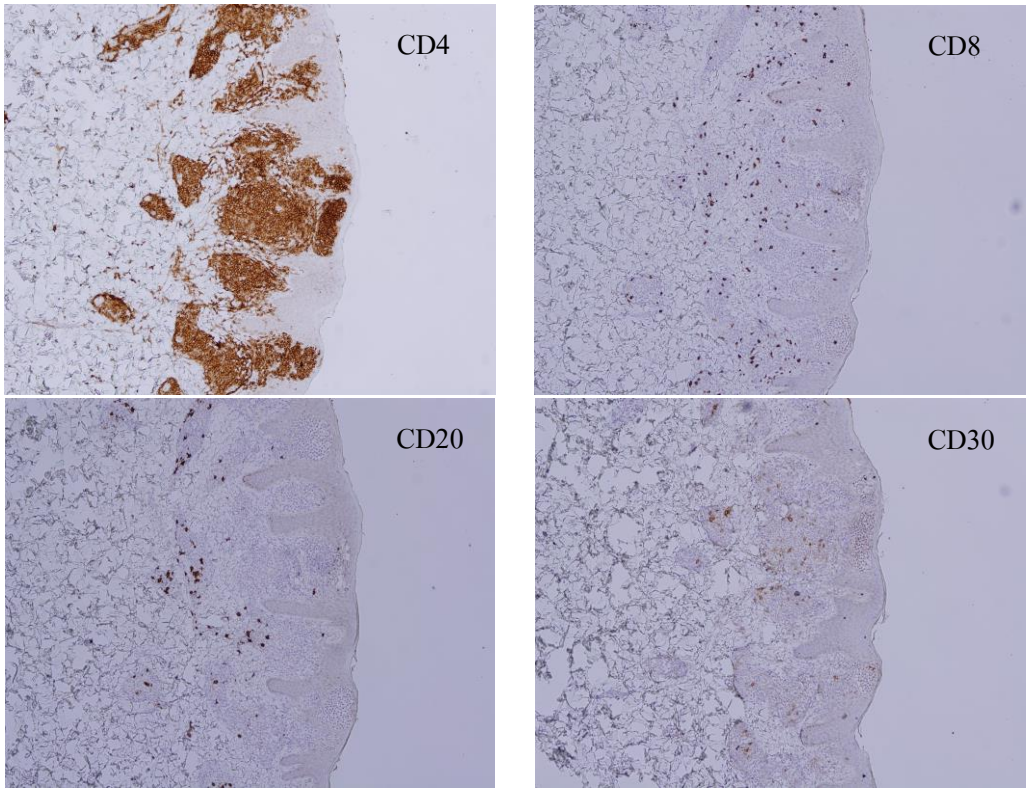

Figure 3 Immunohistochemistry stainings showed atypical cells with positive staining for CD4 with antigenic loss in CD8 but negative for CD30 and CD20.

In 1966, Ackerman and others reported the first case of generalized pustular MF. A 64-year old female patient presented with generalized pustules on erythematous patches and plaques. Skin biopsy showed focal subcorneal and intraepidermal accumulations of neutrophils correlated with pustular lesions, and atypical mononuclear cells formed the major component of the dermal infiltrate correlated with the diagnosis of MF. No typical Pautrier's microabscesses were seen in any of the sections.[5] In 2009, another case of pustular MF was reported by Heike Pabsch and others. A 78-year-old female patient presented with generalized and figurate erythema with marginal lamellar scaling and pustulation. Skin biopsy showed subcorneal and intracorneal pustules with bandlike lymphocytic infiltrate of atypical lymphocytes with cerebriform nuclei. As the patient subsequently developed the clinical presentation of patch and plaque stage MF in the following years, it was hypothesized that pustular eruption could represented a transient expression of MF.[6]

According to the TNMB classification revised by the International Society for Cutaneous Lymphoma (ISCL) and the European Organization of Research and Treatment of Cancer (EORTC), the overall features of this patient were consistent with the diagnosis of mycosis fungoides T2N3M0B0 (stage IVA2).[7] The patient was treated with multi-agent chemotherapy.

The prognosis of MF is depended on the stage, type and extent of skin lesions, and the presence of extracutaneous disease. In contrast to patients with limited patch/plaque stage MF, which generally have similar long-term life expectancy compared with normal population, the disease-related 10-year survival is only 20% for stage IV MF due to systemic involvements and infections.[2,8]

In conclusion, pustular MF should be included as one of the differential diagnosis in patients present with generalized pustular eruption. Skin biopsy should be done in uncertain cases.

References

1. Bayer M, Sterry W. Cutaneous lymphoma. In: Fitzpatrick's dermatology of general medicine. 8th ed. New York: Mcgraw-Hill 2012: 1745-66.
2. Kim YH, Liu HL, Mraz-Gernhard S, et al. Long-term outcome of 525 patients with mycosis fungoides fungoides and Sezary syndrome: clinical prognostic factors and risk for disease progression. Arch Dermatol 2003; 139: 857-66.
3. Nashan D, Faulhaber D, Ständer S, Luger TA, Stadler R. Mycosis fungoides: a dermatologic masquerader. Br J Dermatol 2007; 156: 1-10.
4. Leroy S, Dubois S, Tenaud I, Chebassier N, Godard A, Jacques Y, Dreno B. Interleukin-15 expression in cutaneous T-cell lymphoma (mycosis fungoides and Sezary syndrome). Br J Dermatol 2001; 144: 1016-23.
5. Ackerman AB, Miller RC, Shapiro L. Pustular mycosis fungoides. Arch Derm 1966; 93: 221-5.
6. Pabsch H, Kunze J, Schaller Jörg. Mycosis fungoides presenting as a pustular eruption. J Am Acad Dermato 2009; 61: 908-9.
7. Jawed SI, Myskowski PL, Horwitz S, et al. Primary cutaneous T-cell lymphoma (mycosis fungoides amd Sezary syndrome): part II prognosis, management, and future directions. J Am Acad Dermatol 2014; 70: 223.e1-17.
8. 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.