Case Report / Olgu Sunumu

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Large cell transformed mycosis fungoides with vesiculopustular palmoplantar involvement: Report of one case

Büyük hücre transformasyonlu mikozis fungoideste vezikülopüstüler palmoplantar tutulum: Bir vaka raporu

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Abstract

Mycosis fungoides (MF) is the most common type of primary cutaneous lymphomas, originates from CD4+ T-cells, and is characterized by patches, plaques, and tumoral lesions. Palmoplantar MF lesions are rare, occurring in about 10% of MF patients. When MF affects the palms and soles, it can present with hyperkeratotic, eczematous, tumoral lesions. However, pustular presentation is uncommon. Herein, we present a large cell transformed MF patient with vesiculopustular MF lesions on the palms and soles.

Keywords: mycosis fungoides, palmoplantar, pustules

Öz

Mikozis fungoides (MF), CD4+ T hücrelerinden kaynaklanan, yama, plak ve tümöral lezyonlarla karakterize, primer kutanöz lenfomaların en yaygın formudur. Palmoplantar MF lezyonları nadirdir ve MF hastalarının yaklaşık %10'unda görülür. MF palmoplantar tutulum yaptığında hiperkeratotik, egzamatöz, tümöral lezyonlarla ortaya çıkabilir. Ancak püstüler lezyonlar nadirdir. Bu olgu sunumunda palmoplantar yerleşimli vezikülopüstüler lezyonları bulunan büyük hücreli transformasyonu olan bir MF hastası sunulmaktadır.

Anahtar Kelimeler: mikozis fungoides, palmoplantar, püstül

Introduction

Mycosis fungoides (MF) is the most common type of primary cutaneous lymphoma, originating from CD4+ T cells and characterized by patches, plaques, and tumoral lesions.

The rare clinicopathologic variants of MF include poikilodermatous MF, hypopigmented MF, pigmented purpuric dermatosis-like MF, syringotropic MF, and mycosis fungoides palmaris et plantaris (MFPP). During the course of the disease, ulceration, bacterial infections, erythroderma, and large cell transformation may occur. Palmoplantar involvement is rarely observed in patients with MF[1,2]. Herein, we present a patient with large cell transformed MF who developed vesiculopustular lesions on the palms and soles of the feet.

Case Report

A 43-year-old woman with a 10-year history of classic MF presented to our clinic after an increase in the number of lesions. The patient, whose past treatments included topical steroids, narrowband UVB phototherapy, and methotrexate, had not received any treatment in the last two years. Dermatological examination revealed numerous violaceous plaques and ulcerated or crusted tumors (Figure 1a).

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Mobile, non-tender axillary and inguinal lymphadenopathy was observed during lymph node examination. Histopathological evaluation of a 4-mm punch biopsy taken from a tumoral lesion on her back showed acanthosis, spongiosis, and dermal atypical lymphocytic infiltration. Immunohistochemical studies revealed that the infiltrative cells were positive for CD3 and CD4 and sparsely positive for CD8. Additionally, CD30 expression was observed in 75% of atypical lymphocytes. Investigations included complete blood count, liver and kidney function tests, ultrasound scan of lymph nodes and abdomen, flow cytometry, and peripheral blood smear. The blood test results were within the normal range. The largest inguinal lymph node (3 × 2 cm) was excised after ultrasound scanning, and histopathological evaluation of the excised lymph node revealed reactive changes. The clinical features progressed to erythroderma. Based on the results of physical examinations, blood tests, and screenings, the disease stage was classified as IIIA (T4N1M0B0). During follow-up, numerous itchy vesicles and pustules with yellowish crusts were observed on the palms and the soles (Figure 1b). Pustule cultures were negative for both bacteria and fungi. As the lesions were resistant to topical steroid cream (clobetasol propionate), histopathological analysis was performed on an intact pustule on the patient's palm. The results revealed atypical lymphocytic infiltrates in the epidermis and dermis, with large cell transformation (Figure 2). Based on the clinical and histopathological findings, the patient was diagnosed with vesiculopustular palmoplantar involvement in large cell transformed MF. Four months later, the patient's condition worsened. Despite treatment with multiple antibiotics, the patient died of septic shock.

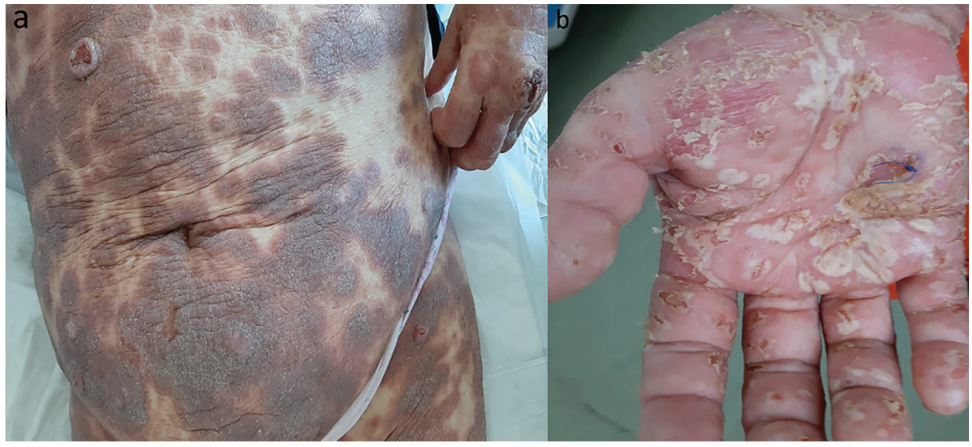


Figure 1. Plaques and tumor lesions on the body (a) and vesiculopustular lesions on the palm (b)

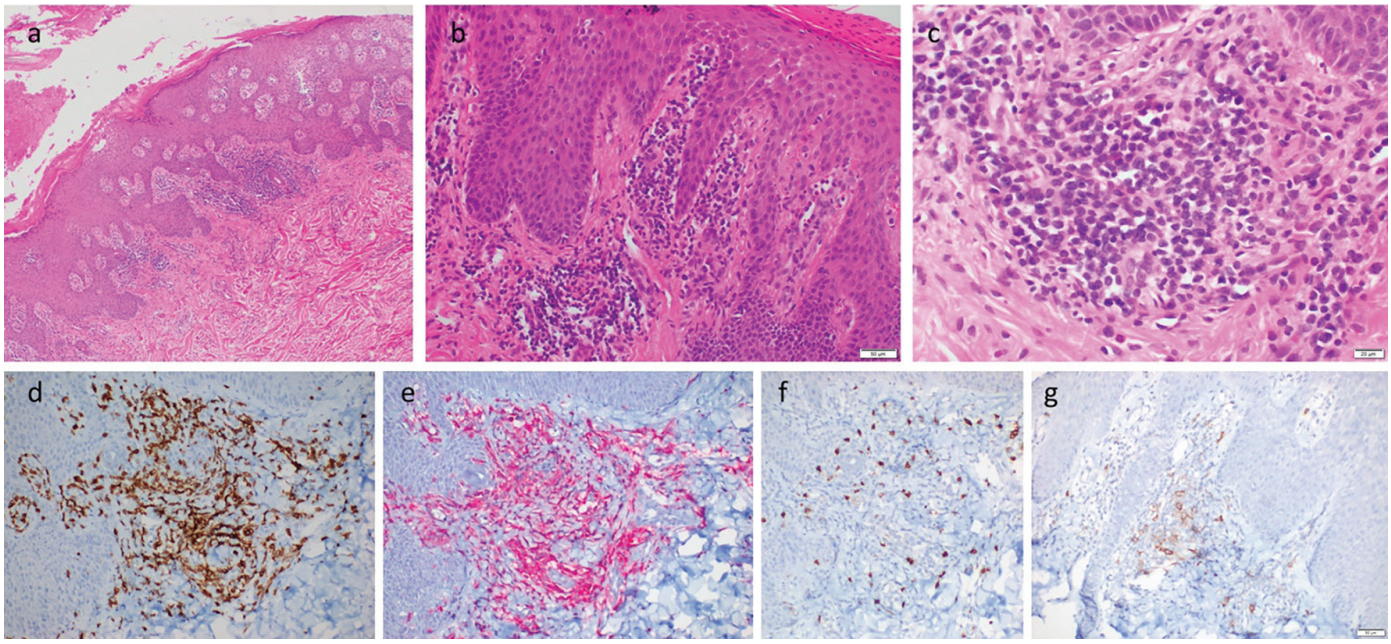


Figure 2. (a), (b) Parakeratosis on the epidermal surface, acanthosis in the epidermis, and epidermal and dermal atypical lymphoid infiltration were observed; (c) sparse cells showing large cell transformation were observed in the dermal lymphocytic infiltrate; (d), (e) atypical lymphoid cells were positive for CD3 and CD4; (f) expression was observed in sparse cells with CD8. (g) Cells showing large cell transformation were CD30 positive

Discussion

Palmoplantar MF lesions are rare, occurring in approximately 3.6-10% of patients. When MF affects the palms and soles, it can present with hyperkeratotic, eczematous, and tumoral lesions[2-6]. However, pustular presentation is uncommon. The differential diagnosis of vesiculopustular palmoplantar lesions includes dermatophyte infections, contact dermatitis, palmoplantar psoriasis, and palmoplantar pustulosis[2]. It is critical to consider MF in the differential diagnosis of treatment-resistant palmoplantar vesiculopustular lesions, especially in patients with a known history of MF, as the diagnosis can be challenging and may be delayed. Previously, Ohkohchi et al.[3] and Moreno et al.[4] described cases of MF with pustular palmar involvement.

MFPP is an uncommon variant of MF, characterized by lesions predominantly limited to the palms or soles, with initial palmoplantar involvement[2]. MFPP is generally characterized by erythematous hyperkeratotic patches and plaques covered in fissures or scales. Vesicular, pustular, tumoral, dyshidrotic, and verrucous lesions can also be observed[2,5,6]. Instead of MFPP, large cell transformed MF with vesiculopustular palmoplantar involvement was considered in our case because our patient had previously been diagnosed with MF, and the lesions were not limited to palmoplantar areas.

In their study evaluating 28 MF patients with acral involvement, Atci et al.[6] stated that acral involvement may be an independent risk factor for disease progression. Our patient also died four months after developing palmoplantar lesions[6].

To the best of our knowledge, this is the first reported case of vesiculopustular palmoplantar involvement in a patient with large cell transformed MF. Therefore, we emphasize the importance of considering MF in the differential diagnosis of treatment-resistant palmoplantar vesiculopustular lesions.

Ethics

Informed Consent: The patient in this manuscript has given written informed consent to the publication of her case details.

Footnotes

Authorship Contributions

Surgical and Medical Practices: Y.C.E., E.A.Y., L.F., B.Ö., N.İ., Concept: Y.C.E., E.A.Y., L.F., B.Ö., N.İ., Design: Y.C.E., E.A.Y., L.F., B.Ö., N.İ., Data Collection or Processing: Y.C.E., E.A.Y., L.F., B.Ö., N.İ., Analysis or Interpretation: Y.C.E., E.A.Y., L.F., B.Ö., N.İ., Literature Search: Y.C.E., E.A.Y., L.F., B.Ö., N.İ., Writing: Y.C.E., E.A.Y., L.F., B.Ö., N.İ.

Conflict of Interest: The authors declare that there is no conflict of interest.

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