Dermatology Practical & Conceptual

An Atypical Presentation of Primary Cutaneous CD4+ Small/Medium Pleomorphic T-Cell Lymphoproliferative Disorder

Didem Kazan[1], Dilek Bayramgürler[1], Ahmet Tuğrul Eruyar[2], Cüyan Demirkesen[3]

1 Department of Dermatology and Venerology, Kocaeli University, Kocaeli, Turkey
2 Department of Pathology, Kocaeli University, Kocaeli, Turkey
3 Department of Pathology, Acıbadem University, Istanbul, Turkey

Citation: Kazan D, Bayramgürler D, Eruyar AT, Demirkesen C. An Atypical Presentation of Primary Cutaneous CD4+ Small/Medium Pleomorphic T-Cell Lymphoproliferative Disorder. *Dermatol Pract Concept*. 2023;13(4):e2023221. DOI: https://doi.org/10.5826/dpc.1304a221

Accepted: April 17, 2023; **Published:** October 2023

Funding: None.

Competing Interests: None.

Authorship: All authors have contributed significantly to this publication.

Corresponding Author: Didem Kazan, Department of Dermatology and Venerology, Kocaeli University. Business tel : +902623037575 E-mail: didem.senses_343@hotmail.com

Case Presentation

A 64-year-old man presented to our out-patient clinic with erythematous patches and papules on the left medial thigh persisted for two years. Dermatological examination showed erythematous patches and papules tended to merge on the inner surface of the left thigh (Figure 1A). Other body areas were normal. Histological examination of multiple punch biopsies showed diffuse infiltration of small-medium-sized atypical lymphocytes throughout the dermis. Immunohistochemical studies found the atypical lymphocytes were positive for CD2, CD3, CD4 in 95%, CD8 in 5%, and CD20 in 2% but negative for CD7 and CD30. BCL-6 and PD-1 expression was positive in 10% and 1-2% of the atypical lymphocytic infiltration respectively (Figure 1, C-H). Blood tests and lymph node ultrasonography were normal. Based on these findings, the patient was diagnosed with primary cutaneous CD4+ small/medium pleomorphic T-cell lymphoproliferative disorder (CD4+ PCSM-LPD). Almost complete clinical response was achieved after nine sessions of radiotherapy treatment (Figure-1B).

Teaching Point

CD4+ PCSM-LPD usually presents with a solitary slow-growing papule or nodule on the face, neck, or upper limbs. It can occur in both sexes and at any age but especially in the 6th and 7th decades of life [1]. Histopathological examination and immunohistochemistry tests are the gold standard diagnostic tests. It is classified as a provisional entity in the 5th edition of the World Health Organization (WHO) classification, due to its limited clinical risk, and indolent course to reflect its uncertain malignant potential [2]. Because of localized disease, staging procedures are usually unnecessary.

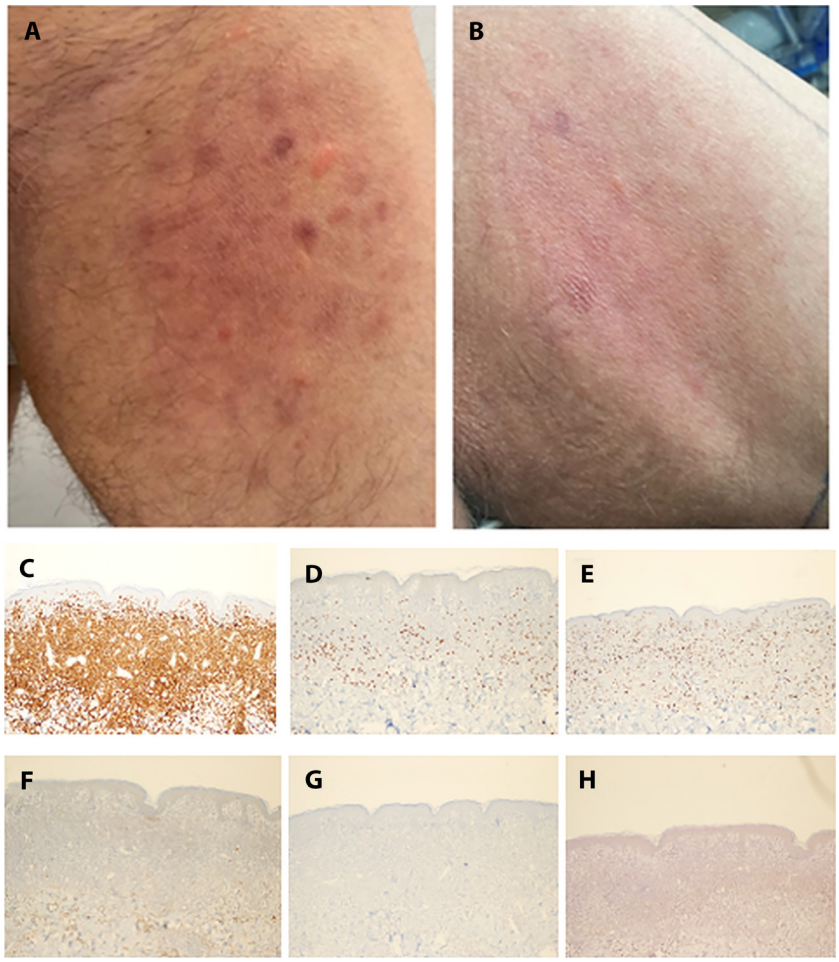

Figure 1. (A) Erythematous patches and papules that tended to merge on the inner surface of the left thigh, (B) After nine sessions of radiotherapy treatment. (C-G) Atypical lymphocytes were positive for CD2 (C), CD3 (D), CD4 in 95% (E), BCL-6 in 10% (F), and negative for CD-30 (G). (H) CD20 positivity detected in atypical lymphocytes 1%-2%.

However, in our patient we have a suspicion of folliculotrophic mycosis fungoides, therefore we made blood tests and lymph node ultrasonography. But, no systemic involvement was found. Clinicians should keep it in mind, especially in solitary lesions on the upper body, and also in localized MF-like lesions on lower extremity like our patient, because its diagnosis helps them to avoid cytotoxic and immunosuppressive treatments.

The patient gave him informed consent for the publication of his clinical history data.

References

1. Virmani P, Jawed S, Myskowski PL, et al. Long-term follow-up and management of small and medium-sized CD4+ T cell lymphoma and CD8+ lymphoid proliferations of acral sites: a multicenter experience. *Int J Dermatol*. 2016;55(11):1248-1254. DOI: 10.1111/ijd.13340. PMID: 27369070. PMCID: PMC5053827.
2. Willemze R, Cerroni L, Kempf W, et al. The 2018 update of the WHO-EORTC classification for primary cutaneous lymphomas. *Blood*. 2019;133(16):1703-1714. *Blood*. 2019;134(13):1112. DOI: 10.1182/blood.2019002852. PMID: 31558559. PMCID: PMC6764272.