

An Association of Mycosis Fungoides and Pityriasis Lichenoides Et Varioliformis Acuta: Case Report

Mikozis Fungoides ve Pitriazis Likenoides Varioliformis Akuta Birlikteliği

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ABSTRACT Pityriasis lichenoides et varioliformis acuta (PLEVA) is a disease of unknown cause that is characterized by papulonecrotic lesions. Although the most accepted theory is the infectious and immunological pathogenesis, there are a few cases also pointing the relationship between PLEVA and lymphoma. Our case applied to our clinic with crusted papules on his lower extremities. On the dermatological examination, pale erythematous patch-like lesions on trunk and both of his extremities and papular lesions with necrotic crusts on his both lower extremities were detected. The histopathological examinations of the biopsies of lesions of the trunk and lower extremity were consistent with mycosis fungoides and PLEVA respectively. In laboratory examinations, the positivity of human parvovirus B19 immunoglobuline M was detected. In this report, we present a case of PLEVA who had a seropositivity of Human Parvovirus B19 Ig M and was associated with mycosis fungoides.

Key Words: Mycosis fungoides; parvovirus B19, human; pityriasis lichenoides

ÖZET Pitriazis likenoides et varioliformis akuta (PLEVA) papulonek-rotik lezyonlarla karakterize sebebi bilinmeyen bir hastalıktır. Her ne kadar patogeneze çoğunlukla kabul edilen teori enfeksiyöz ve immünolojik olsa da, PLEVA ve lenfoma arasındaki ilişkiye işaret eden yayınlarda da mevcuttur. Olgumuz kliniğimize alt ekstremitelerinde krutlu papüllerle başvurdu. Dermatolojik muayenesinde gövdede ve her iki üst ekstremitede soluk eritemli yama benzeri lezyonlar ve her iki alt ekstremitede üzerlerinde nekrotik krutların olduğu papüler lezyonlar saptandı. Gövdedeki ve alt ekstremitedeki lezyonlardan alınan biyopsilerin histopatolojik incelemeleri sırasıyla mikozis fungoides ve PLEVA ile uyumlu olarak değerlendirildi. Yapılan laboratuvar incelemelerinde Human parvovirüs B19 immünoglobulin (Ig) M pozitif olarak bulundu. Bu yazıda Human Parvovirüs B19 IgM pozitifliği saptanan ve mikozis fungoidesin eşlik ettiği PLEVA'lı bir olgu sunuyoruz.

Anahtar Kelimeler: Mikozis fungoides; Human Parvovirüs B19; pitriazis likenoides

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Pityriasis lichenoides et varioliformis acuta (PLEVA) is an uncommon disease characterized by hemorrhagic and papulonecrotic lesions accompanied by minimal constitutional symptoms.^{1,2} The etiology of PLEVA is unclear. It has been postulated to be a response to foreign antigens, such as infectious agents.^{1,3} Some authors prefer to classify this disorder as a lymphoproliferative condition.² Development of mycosis fungoides (MF) following PLEVA or the association of PLEVA and MF is not common.³ PLEVA shows no documented significant association with malignant lymphoma, except for a few cases.²⁻⁴ Herein, we report a patient with PLE-

VA who was seropositive for Human Parvovirus B19 immunoglobulin (Ig) M and associated with MF.

CASE REPORT

A 36-year-old man presented with pruritic crusted papules localized on his legs accompanied with generalized arthralgia and malaise. These lesions had been present for a month. In a detailed history, we learned that he also had had patch-like lesions on his trunk and upper extremities for the last four years.

On the dermatological examination, we observed varying sizes of pale erythematous patch-like lesions with scales localized on the trunk and both of his upper extremities (Figure 1). On both of his lower extremities, there were necrotic papules surrounded with an erythematous halo (Figure 2).



FIGURE 1: The appearance of the patch-like lesions on the trunk.



FIGURE 2: The appearance of the necrotic papules.

The laboratory findings were within normal values; complete blood count, erythrocyte sedimentation rate, urinalysis and serum chemistry were all normal. The markers of hepatitis and HIV 1/2 were negative. The tumour markers were in normal levels. The viral markers including Rubella, Cytomegalo virus, Herpes Simplex type 1 and 2, Epstein-Barr virus, respiratory syncytial virus, varicella Zoster virus and Adenovirus IgG levels were positive. Human Parvovirus B19 IgM was found positive.

The histopathological examination of the biopsy specimens from the patch-like lesions which localized on the trunk revealed minimal hyperkeratosis, irregular acanthosis, atypical lymphoid cell infiltration in the epidermis, and diffuse inflammatory infiltration in the upper dermis and a perivascular lymphoid cell infiltration in the middle part of dermis (Figure 3). In immunohistochemical examination the inflammatory cells in the infiltration were composed of T cells (Figure 4). These

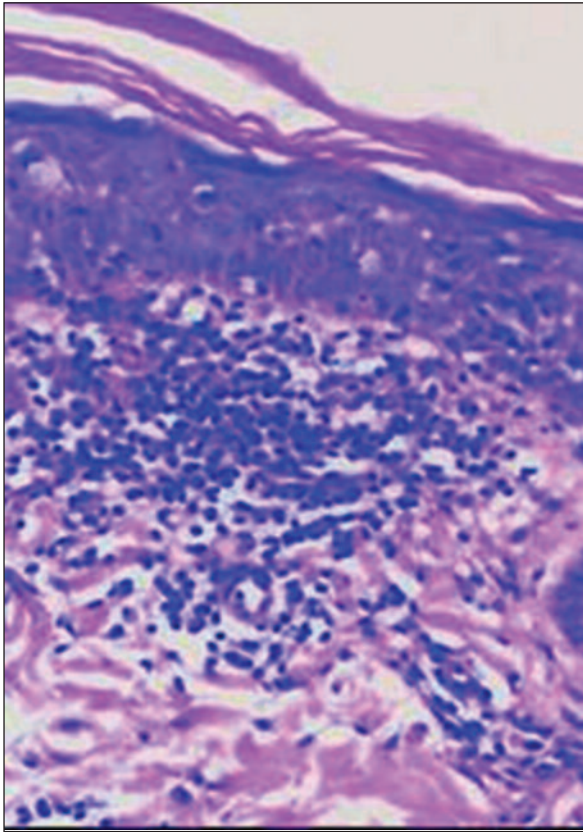


FIGURE 3: Histopathological changes in the Patch-like lesions of mycosis fungoides (HE x 200).

findings were consistent with “patch stage of mycosis fungoides”.

The biopsy was taken from the necrotic crusted papular lesions revealed exocytosis, and necrotic keratinocytes in the epidermis. In the upper dermis there were perivascular mononuclear cell infiltration and extravasated erythrocytes. These findings were consistent with PLEVA (Figure 5).

The PLEVA lesions of our case completely regressed without treatment. The lesions of MF were treated with PUVA therapy. The patient was followed up in our clinic for two years and the lesions of PLEVA did not show recurrence.

DISCUSSION

PLEVA is usually a self limited disease belonging to pityriasis lichenoides group. It is seen more frequently in men than in women. PLEVA consists of multiple, disseminated, erythematous and hemorrhagic papules with central necrosis, distributed on

the trunk and on the proximal extremities. Sometimes skin findings may be accompanied by fever, headache, malaise and arthralgia. Lesions generally have a polymorphous appearance. While healing; they can leave atrophic/ hypertrophic scars, hyper or hypopigmentation.^{1,2,5,6}

In clinical progress, PLEVA is rather a benign entity. There are still debatable points in the pathogenesis of PLEVA, since the reports about this subject are conflicting.^{3,7-9}

The pathogenesis of PLEVA is still unclear, there have been proposed some theories. The most accepted one is the theory indicating the hypersensitivity and immunological reaction against an infectious agent.^{1,6,10} The immune reaction inclu-

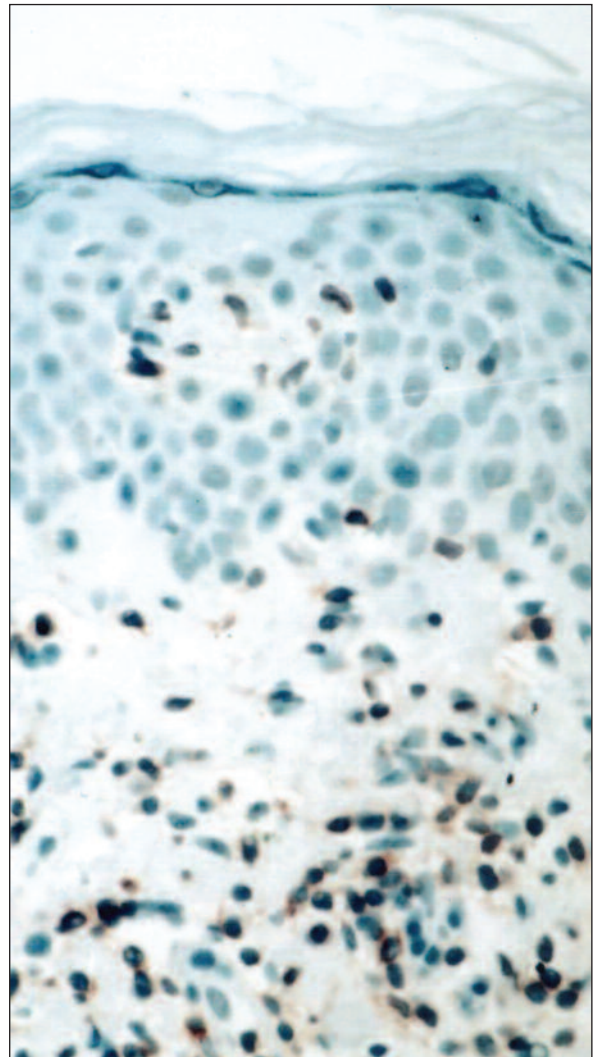


FIGURE 4: Lymphoid cells stained with CD3 positively. (CD3 X 200).

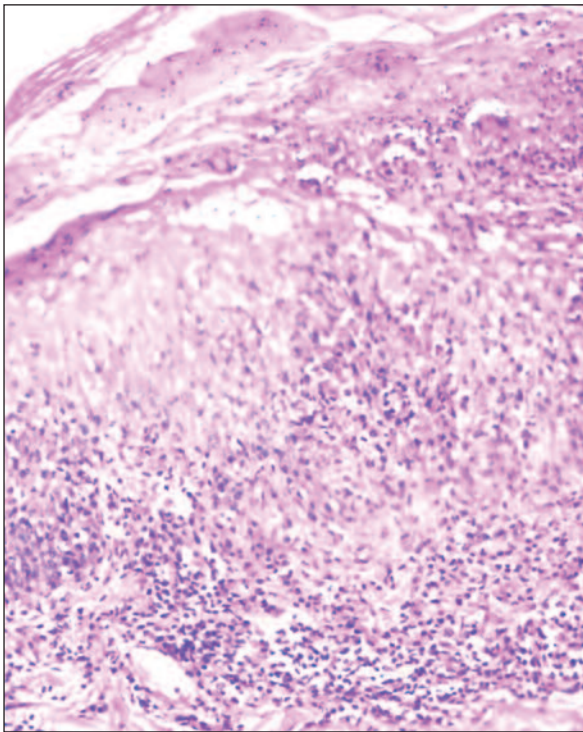


FIGURE 5: Necrotic papular lesion of PLEVA (HE x 50).

des both immune-complex reaction and cell-mediated hypersensitivity response. In our case we detected seropositivity of Human Parvovirus B19 IgM. The other viral markers were negative. However, in previously published reports, clonal T cell lymphocyte population was demonstrated in the blood or in the lesions of the cases with PLEVA, thus identifying PLEVA as a lymphoproliferative process rather than an inflammatory process.^{2,11-16} Moreover, there have been published reports indi-

cating the relationship between pityriasis lichenoides and cutaneous lymphoma.^{3-9,11,17} Fortson et al,¹⁷ reported two cases bearing the diagnoses of PLEVA that evolved into cutaneous T-cell lymphoma (CTCL) clinically and histopathologically after 8 years and 6 years respectively; and emphasized that there may be a relationship between PLEVA and CTCL. Tomasini et al,¹¹ described a patient with pityriasis lichenoides chronica evolving into mycosis fungoides. Moreover, two reports have been recently published by Kempf et al³ and Cozzio et al⁴ pointing the togetherness of MF and PLEVA. Kempf et al,³ reported two cases indicating the paraneoplastic association of PLEVA and cutaneous lymphomas. Cozzio et al,⁴ proposed febrile ulceronecrotic Mucha-Habermann disease, a variant of PLEVA to represent a cutaneous T-cell lymphoma entity.

Our case was seropositive for Human Parvovirus B19 Ig M. The detection of the Human Parvovirus B19 may support the infectious and immunological pathogenesis that is accepted mostly. However, he also had a diagnosis of mycosis fungoides associated with PLEVA attack. Because of the coexistence of PLEVA with mycosis fungoides, and the increasing number of cases about this relationship in the literature, we believe that this relationship must be kept in mind and the patients should have to be considered for a follow up. Nevertheless, further studies are needed to clarify the association of PLEVA and cutaneous lymphoma, whether this association depends on a paraneoplastic relationship.

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