Case Report / **Olgu Sunumu**

A Case of Leucocytoclastic Vasculitis Due to T-Cell Rich B-Cell Lymphoma

T Hücreden Zengin B Hücreli Lenfomaya Bağlı Lökositoklastik Vaskulitli Bir Olgu

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***Submitted** / **Başvuru tarihi:** 03.03.2006 **Accepted** / **Kabul tarihi:** 11.05.2006*

Leucocytoclastic vasculitis is a heterogeneous group of disorders with various factors responsible in its etiology. It can be observed as a paraneoplastic phenomenon in patients with malignancies. Here we report a 68 year old male with T cell rich B cell non-Hodgkin lymphoma, who had diffuse plaque-type rashes on his back, abdomen, chest wall, and extremities due to leucocytoclastic vasculitis of the skin. He received chemotherapy and mini mantle radiotherapy. Cutaneous findings of the patient resolved completely with the remission of the primary disease. Skin rashes in patients with malignancies may be due to primary tumor involvement or secondary to vasculitis. Histopathological examination of affected tissue specimen is helpful to distinguish secondary vasculitis from primary tumor metastasis.

Key Words: Lymphoma B-cell; neoplasms; vasculitis; paraneoplastic syndromes.

Lökositoklastik vaskülit, etyolojisinde çeşitli faktörlerin sorumlu olduğu heterojen bir hastalık grubudur. Malignitesi olan hastalarda paraneoplastik bir fenomen olarak görülebilir. Bu yazıda derinin lökositoklastik vaskülitine bağlı sırt, abdomen, göğüs duvarı ve ekstremitelerinde diffüz, plak tarzında döküntüleri olan 68 yaşında T hücreden zengin B hücreli non-Hodgkin lenfoma tanısı konmuş bir erkek hasta sunulmaktadır. Hastanın deri bulguları kemoterapi ve mini-mantle radyoterapi ile primer hastalığın remisyonu sonucunda tamamen düzeldi. Malignitesi olan hastalarda deri döküntüleri, primer tümör tutulumu ya da vaskülite sekonder olabilmektedir. Etkilenen dokunun histopatolojik incelemesi sekonder vaskülitleri primer tümör metastazından ayrımında yardımcıdır.

Anahtar Sözcükler: B hücreli lenfoma; neoplazm; vaskülit; paraneoplastik sendrom.

Vasculitis can be observed as a paraneoplastic phenomenon in patients with malignancies.[1] Paraneoplastic vasculitis most often affects skin, but in some patients systemic involvement also may occur.[2] Vasculitic process can regress with the operations, radiotherapy and chemotherapy applications aimed treating at the primary disease.[3]

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Although leucocytoclastic vasculitis is a rare condition in lymphoma patients, several cases have been reported.[4] In addition, primary cutaneous lymphomas may cause neoplastic skin involvement by invasion of primary tumor. It can clinically be seen as purpuric, urticarial or necrotic skin rashes.[2] The skin biopsies are valuable in these patients for differential diagnosis. Vasculitis findings without primary neoplastic cells are the signs of paraneoplastic involvement in the biopsy material.[5]

In this report, leucocytoclastic vasculitis related with T cell rich B cell non-Hodgkin lymphoma is presented in a case presenting with diffuse purpuric skin rashes.

CASE REPORT

The 68-year-old patient realized a mass on the right side of his neck 20 days before his application. During the following days, the mass enlarged towards the back of his ear and started to ache. He had lost 3-4 kg weight during the last 15 days and loss of appetite and weakness was added to his symptoms. When the patient applied to our hospital he had diffuse plaque-type rashes on his back, abdomen, chest wall, and extremities. There was no significant feature except tonsillectomy in his medical and family history.

The physical examination findings were a 7-8 cm palpable mass with pain which started from the right angulae of mandibulae and firm, painful lymphadenopathies, with one on the right supraclavicular area (2 cm) and on right and left inguinal areas (1.5-2 cm). He had clinically significant respiratory distress due to lymphadenopathies (bulky disease). There was diffuse, maculopapular plaque forming erythematous rashes on extremities and on the body, which did not change color with pressure (Fig. 1a, b).

The laboratory values of the patient were as follows: Complete blood count: WBC 4.4 109/L (Neutrophil 68%, lymphocyte 18%), hemoglobin 14 g/dl, hematocrit 42.3%, MCV: 86.3 fl, RDW: 13.7, thrombocyte count: 380x109/L. Biochemistry: fasting blood glucose: 98 mg/dl, serum creatinine 1.2 mg/dl, uric acid: 4 mg/dl, Na: 135 mEq/L, K: 4.3 mEq/L, Cl: 103 mEq/L, Ca: 8.8 mg/dl, LDH: 823 U/L, AST: 41 U/L, ALT: 44 U/L, C-Reactive Protein (CRP): 64 mg/dl, erythrocyte sedimentation rate: 22 mm/hour.

Multiple lymphadenopathies on right side of the neck, posterior to the sternocleidomastoid muscle, lateral to the cervical main vascular structures and a 4x3 cm additional mass proximal to this appearance which was lobulated,

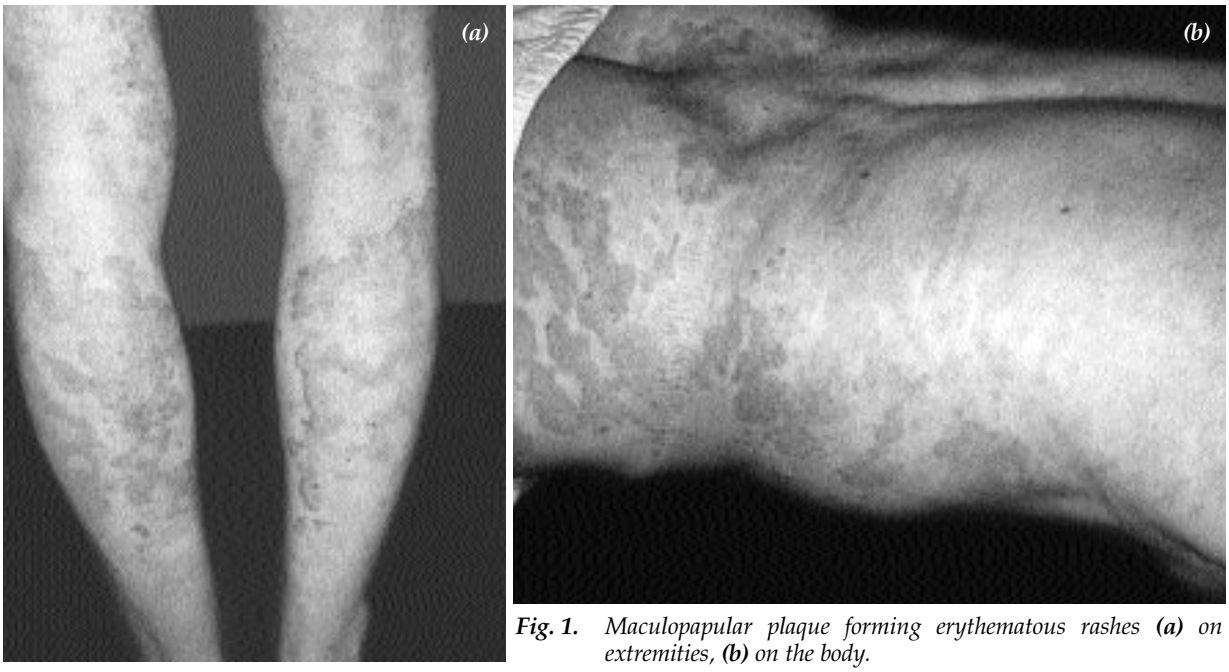

Fig. 1. *Maculopapular plaque forming erythematous rashes* ***(a)*** *on extremities,* ***(b)*** *on the body.*

contoured, cystic with septations were seen on cervical computed tomography (CT). Thoracoabdominal CT revealed multiple lymphadenopathies with pathological dimensions, with a tendency to conglomerations on supraclavicular, axillary, mediastinal, bronchial, upper abdominal and bilateral femoral regions. The appearance of the liver was normal except several simple cysts. An increase in spleen volume was determined.

As no specific diagnosis was obtained by the incisional biopsy of the lymphadenopathies on the left cervical region, excisional biopsy was performed on left anterior cervical lymphadenopathies. The diagnosis of T cell-rich B

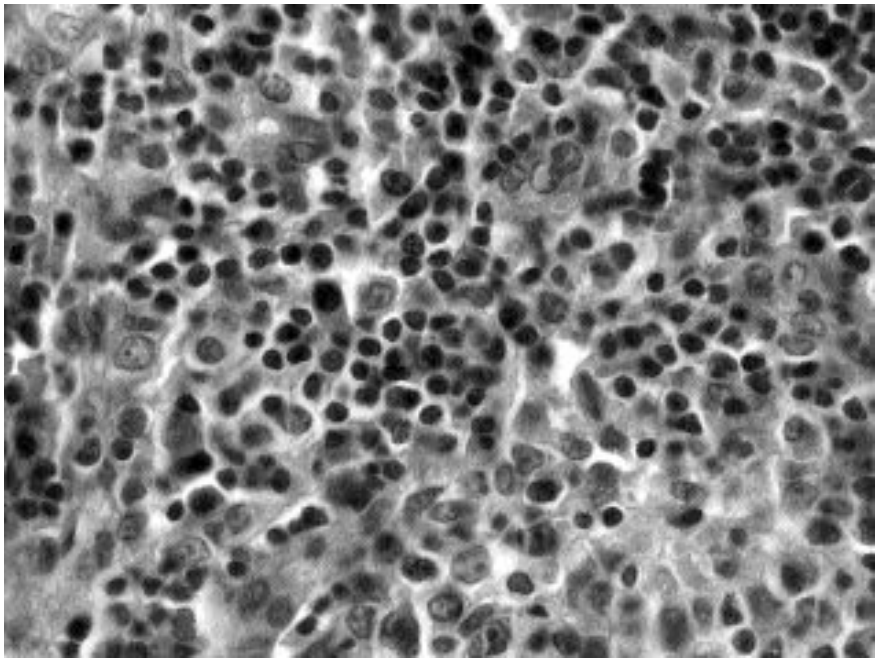

Fig. 2. *Pathological examination of left anterior cervical LAP showed T cell rich B cell lymphoma. Immunoperoxidase studies performed on paraffin-embedded tissue showed that the neoplastic cells stained strongly with the B-cell marker CD20, whereas small reactive lymphocytic cells were CD3 positive (H-E x 20).*

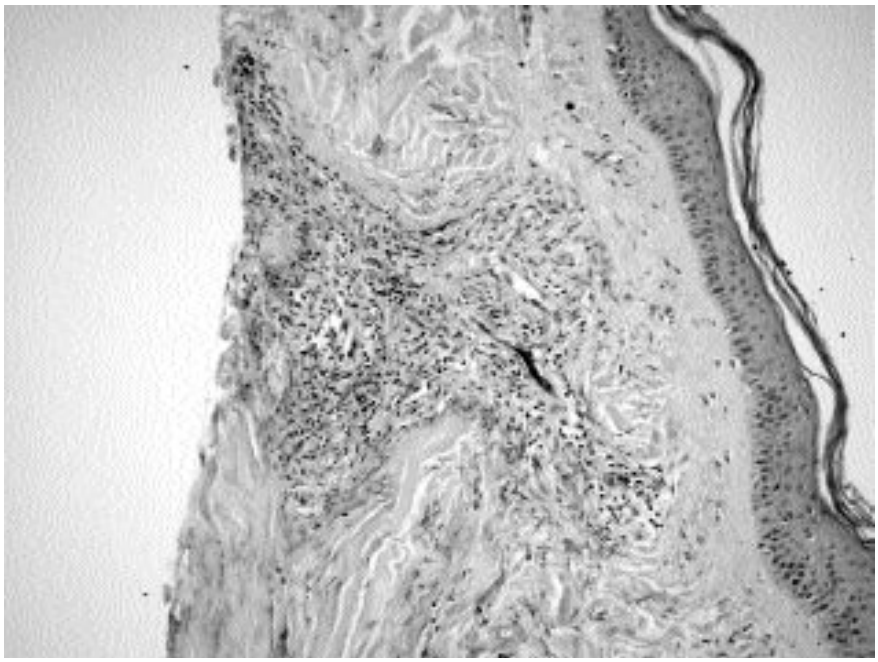

Fig. 3. *Skin biopsy of the patient revealed leucocytoclastic vasculitis (H-E x 10).*

cell lymphoma, which was rich in histiocytes, was made based on the pathological examination (Fig. 2). The bone marrow biopsy revealed invasion and the patient was evaluated as stage 4 (bulky).

As the patient's cutaneous findings were increasing, skin biopsies were performed on right forearm and left foreleg and found to be in accordance with leucocytoclastic vasculitis (Fig. 3).

The CHOP (cyclophosphamide 750 mg/m^2/day d1, adriamicin 50 mg/m^2/day d1, vincristine 1.4 mg/m^2/day d1, prednisolon 40 mg/m^2/day d1-5) chemotherapy was started. The skin lesions showed a 50% regression after the first course of the treatment. Mini mantle radiotherapy was performed following second course of chemotherapy at a dose of 3000 cGy. After the third course of the treatment the skin lesions were completely resolved. Following the fourth course of chemotherapy, an evaluation was performed by cervical, thoracoabdominal CT, and by bone marrow biopsy which showed that full remission was obtained. The treatment protocol was decided to be completed by six courses of chemotherapy. However, because of recurrent neutropenic fever episodes the patient rejected the sixth course. The patient is still followed up in our outpatient clinics for the last three years. He is still in remission after the five courses of chemotherapy and his skin lesions have not recurred.

DISCUSSION

Leucocytoclastic vasculitis can either be primary as in Henoch-Schönlein purpura, or can be a component of some specific diseases or secondary to exogenous factors. Infectious agents, drugs, or malignities form the endogenous or exogenous antigens.[5] As a result of the hypersensitivity reaction to these antigens, neutrophil infiltration to the postcapillary venules of the skin and the formation of leucostasis cause the clinical findings.[2]

In 4.5-8% of the vasculitis patients, a malignancy is observed. Cutaneous leucocytoclastic vasculitis forms the 30-40% of the paraneoplastic vasculitis.[6] In 1986 Longley et al.[7] first claimed that antigens that are formed during malign diseases cause secondary vasculitis. In 1993, Kurzrock and Cohen[8] reported the results of 200 patients with cancer and paraneoplastic vasculitis. Hayem et al.[9] reported that 90% of the malignancy-related vasculitis cases are caused by hematological malignancies. Cases of paraneoplastic vasculitis were reported in the literature in low-grade B cell and T cell non-Hodgkin lymphoma, hairy cell leukemia, splenic lymphoma, Hodgkin lymphoma, chronic myeloid leukemia, acute myeloid leukemia, and sporadically in multiple myeloma.[6,10-12]

Leucocytoclastic vasculitis can develop before, during or after the diagnosis of the malignancy. Skin involvement typically starts with maculopapular rash. Clinically, it can present with purpuric, urticarial, or necrotic rashes. It can be seen in all age groups without a gender preference. The capillaries in the superficial papillary dermis with a diameter smaller than 50 μm, postcapillary venules and nonmuscular arterioles are affected during the cutaneous vasculitis. Neutrophil infiltration around and in the vessel wall, leukocytoclasia characterized by degranulation and fragmentation of the neutrophils, fibrinoid necrosis of the vessel wall, endothelial cell proliferation, necrosis and edema are seen during the histological observation. Palpable purpura, urticary, bullous lesions, vesicles, and splinter hemorrhages can be observed due to vascular wall damage and erythrocyte extravasations.[3] Lesions are localized on legs or thighs because of the high hydrostatic pressure. There are no known correlation with paraneoplastic vasculitis and the prognosis of the malignancy. Although it is mainly characterized with skin findings, systemic involvement such as the involvement of kidneys, lungs, nervous system or gastrointestinal system can accompany the clinical course.[2]

In our case, T- cell rich B- cell non-Hodgkin lymphoma and leucocytoclastic vasculitis were observed together. T-cell rich B-cell non-Hodgkin lymphoma can be morphologically confused with Hodgkin lymphoma and peripheral T cell lymphoma. Pathologically it is charac-

terized by a small number of big neoplastic B cells accompanied with more small, reactive T lymphocytes.[13,14] In our patients leucocytoclastic vasculitis was developed at the time of the diagnosis of lymphoma and cutaneous findings completely resolved with the remission of the primary disease. Skin biopsy is diagnostic for secondary vasculitis and very important for the differential diagnosis of the skin involvement of the lymphoma. In our patient skin biopsy played a leading role for the diagnosis as well.

In conclusion, leucocytoclastic vasculitis is a heterogeneous group of diseases with various factors responsible in its etiology. It can accompany hematological malignities and as seen in our case can dramatically recover with the treatment of the primary disease.

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