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Case Report

A Case of Hairy Cell Leukemia Diagnosed by Leukocyoclastic Vasculitis Symptoms

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Abstract

Hairy cell leukemia (HCL) is a rare chronic lymphoproliferative disease. LSV is characterized by inflammation of small vessels. Its association is rare in the literature and HCL can be seen in LSV etiology, although the relationship between the 2 diseases is far from clear. Here, we aimed to present a case with constitutional symptoms, palpable purpuric lesions on the body and diagnosed as HCL from bone marrow biopsy.

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Introduction

Hairy cell leukemia (HCL) is a rare chronic lymphoproliferative disease characterized by pancytopenia and splenomegaly, with 'hairy' cells seen in peripheral blood and bone marrow. The average age at diagnosis is between 50 and 55 years. The male/female ratio is 4/1. Common findings of HCL includes massive splenomegaly, pancytopenia, and lymphadenopathy; fever is a rare symptom. The frequency of several autoimmune diseases is increased in HCL, but rare cases of leukocytoclastic vasculitis (LV) have been rarely reported.[1,2] LV is characterized by inflammation of small vessels. The etiology of LV includes drugs, infections, malignancies, and systemic inflammatory diseases. Palpable purpura is typical, especially in the lower extremities. In this article, we aimed to present an HCL case with leukocytoclastic vasculitis symptoms.

Case Report

A 70-year-old male patient was admitted to the emergency department with complaints of palpable purpuric lesions, fever, night sweats, and weakness, which started on his extremities for 6 months and were more intense on his abdomen and back for

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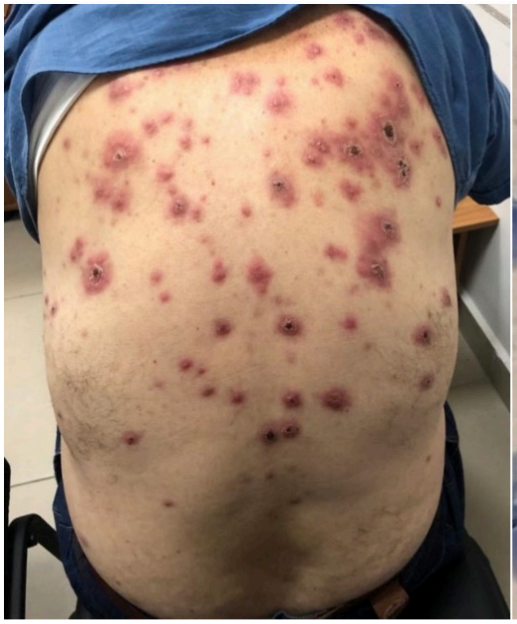

Figure 1. The appearance of pustular lesions on the erythematous ground in the patient's extremities and body.

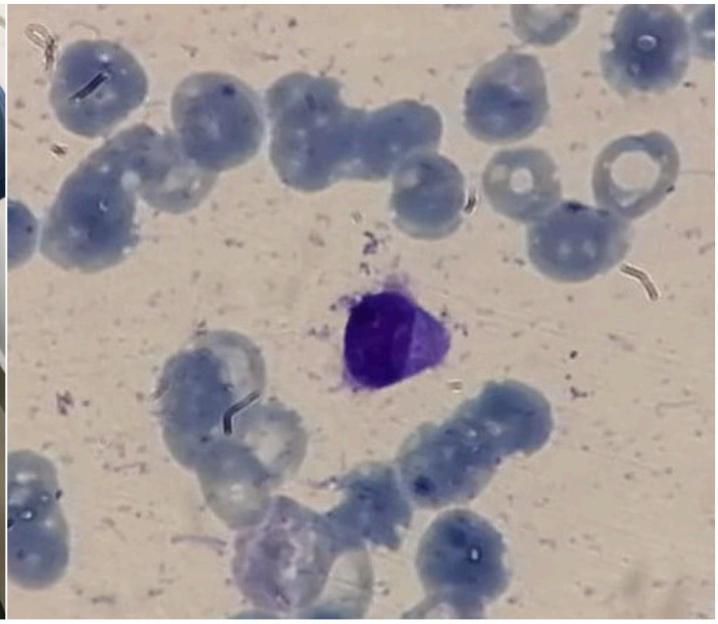

Figure 2. Bone marrow examination

the last 2 weeks. On physical examination, his general condition was normal, there were crusted, pustular lesions on the erythematous ground on his extremities and body (Figure 1). The patient did not report any history of a medical disease and his family history was normal. Complete blood count revealed: leukocyte: 1470/mm³, neutrophil: 831/mm³, lymphocyte: 601/mm³, hemoglobin: 7.9 gr/dL, platelet: 150,200/mm³, C-reactive protein: 123 mg/L, erythrocyte sedimentation rate: 140 mm/h. The serological examination for Brucella, hepatitis was negative, parvovirus, cytomegalovirus, Epstein-Barr virus, measles, chickenpox, mycoplasma, Lyme, rubella were also found to be negative. Anti-neutrophil cytoplasmic antibody, rheumatoid factor and anti-cyclic citrullinated peptide negative, C3 and C4 normal, antinuclear antibody (ANA) 1/100 end point positive, ANA profile negative. The patient was hospitalized and piperacillin/tazobactam 3x4.5 gr was administered. Punch biopsy pathology of the lesion in the extremity was reported as LV. The patient's constitutional symptoms also increased the probability of possible systemic disease. The patient's COVID-19 PCR test was negative, the absence of ARB in urine and sputum, normal level of immunoglobulins, no lymphadenomegaly in thoracic and abdominal computed tomography, spleen size at the border of 13 cm, and peripheral smear showed atypical lymphocytes. Bone marrow biopsy was performed to explain the cause of pancytopenia. In bone marrow aspiration and imprint examination, atypical lymphocytes were larger than normal, chromatin structure was thin, cells with large cytoplasm, and cytoplasmic protrusions (Figure 2). Bone marrow pathology; morphological features primarily suggest HCL. TRAP was poorly stained and immunohistochemical negative results were reported as other B-cell lymphoid neoplasms could be excluded, and the diagnosis of HCL was confirmed. The patient was transferred to the pandemic service due to respiratory distress in the follow-up and the received COVID-19 PCR test was positive. The treatment for COVID-19 was administered. In his follow-up, acute respiratory distress syndrome developed and died.

Discussion

HCL and LSV are rare diseases. Few cases are reported together in the literature, and the underlying mechanism is not clear. In its pathogenesis, infiltration into the vessel wall by

hairy cells is thought to be responsible. Local cytokines causing inflammatory tissue damage, antibody cross reactivation with the vessel wall of hairy cells may be responsible for further propagation of the process. LV can be seen at any stage of HCL.[1,2] Cutaneous symptoms also regress when treating the underlying HCL with interferon or purine analogs. In conclusion, HCL can be seen in histological LV etiology even though there is no clinical picture.

Conflict of Interests

Authors declare that there are none.

Acknowledgment

This study has been presented in 17th Uludag Internal Medicine National Winter Congress, 6th Bursa Family Medicine Association National Congress, 11th Uludag Internal Medicine Nursing Congress, 5–7 March 2021, Bursa, Turkey.

References

1. Broccoli A, Gandolfi L, Pellegrini C, Agostinelli C, Argnani L, Zinzani PL. Leukocytoclastic vasculitis associated with hairy cell leukemia at diagnosis: a case report and review of the literature. Tumori. 2016 Nov 11;102(Suppl. 2):124-7. doi: 10.5301/tj.5000487.
2. Moyers JT, Liu LW, Ossowski S, Goddard L, Kamal MO, Cao H. A rash in a hairy situation: Leukocytoclastic vasculitis at presentation of hairy cell Leukemia. Am J Hematol. 2019 Dec;94(12):1433-4. doi: 10.1002/ajh.25597.