A case of adult acute T-cell lymphoblastic leukemia presented with hemophagocytic syndrome

Hemofagositik sendromla prezente olan akut T-hücreli lenfoblastik lösemi olgusu

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Patients with acute lymphoblastic leukemia (ALL) present most frequently with signs and symptoms of the uncontrolled growth of leukemic cells in bone marrow, lymphoid organs and other sites of extramedullary spreading such as cerebrovascular disease, pleural effusions, blindness, etc. [1-3]. Hemophagocytic syndrome (HPS) is an unusual acute syndrome presenting with fever, hepatosplenomegaly, and cytopenias. The hallmark of HPS is the accumulation of activated macrophages that phagocyte hematopoietic cells in the reticuloendothelial system. Most cases of HPS in adults are secondary to some disorders such as infection or malignancy, and thus investigation of the underlying disease is necessary. Herein, we report a case of ALL presented with HPS.

A 22-year-old male patient referred to our clinic because of pancytopenia. The patient experienced febrile attacks one month before hospitalization but at that time no etiological factor that could cause fever had been demonstrated. The patient was hospitalized by the Department of Pulmonary Diseases because of pulmonary infiltrates. Febrile attacks did not resolve despite use of broad spectrum antibiotics and antifungal drug. However Candida spp. was detected with bronchoalveolar lavage. Complete blood count values were reported to be normal in this period. The patient consulted the Hematology Department for development of anemia. Atypical cells with lymphomonocytoid appearance were present in peripheral blood film. Bone marrow aspiration and biopsy were performed. Bone marrow aspiration showed an increase in histiocytes, the cells phagocytizing erythrocytes and platelets (Figure 1). There was no malignant infiltration in the marrow. Periodic acid-Schiff and peroxidase staining were negative. The patient remained febrile during this period and neutropenia and thrombocytopenia developed in addition to anemia. Intravenous immunoglobulin treatment (1 g/kg for 2 days) was given. Ten days after immunoglobulin

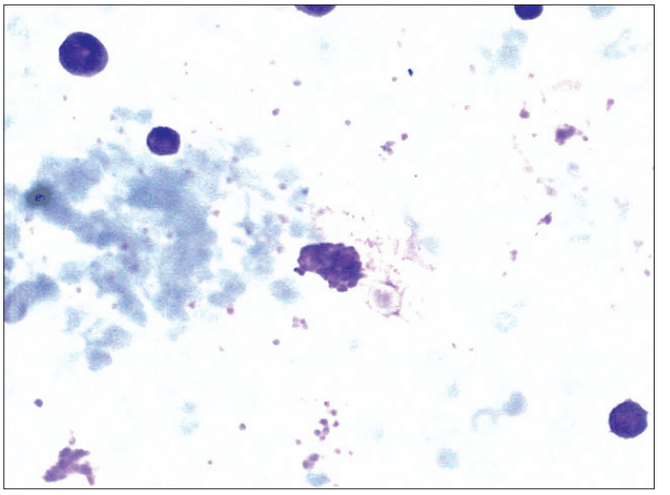

Figure 1. Bone marrow aspiration showed an increase in histiocytes, the cells phagocytizing erythrocytes and platelets

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treatment, firstly thrombocytopenia and later neutropenia resolved. Peripheral blood film examination and white blood cell differential were normal. Bone marrow was reevaluated and findings of hemophagocytosis had disappeared and bone marrow was normal in appearance. The patient was no longer febrile. Intravenous antifungal treatment was switched to oral voriconazole and the patient was discharged with regular follow-up. The patient admitted with the findings of pancytopenia one month later. Peripheral blood film showed atypical lymphomonocytoid cells and we proceeded to bone marrow examination. There was blastic infiltration of 50%. The blasts had a scanty, agranular, basophilic cytoplasm and a large nucleus with 1 to 3 nucleoli. Analysis of surface antigens showed that 49% of cells were blast cells positive for CD7, cytoplasmic CD3 and terminal deoxynucleotidyl transferase (TdT). Cytogenetic examination revealed normal karyotype. The diagnosis of T-cell lymphoblastic lymphoma was made. Four-drug remission induction chemotherapy including cyclophosphamide 1200 mg/m^2 once on day 1, vincristine 2 mg once weekly (days 1, 8, 15 and 22), daunorubicin 45 mg/m^2 for 3 days and prednisolone 60 mg/m^2 po daily for 21 days was started. The patient reached hematological remission after one cycle of chemotherapy and we proceeded to central nervous system prophylaxis with cranial radiotherapy and intrathecal methotrexate. The patient experienced an early relapse two weeks after completion of radiotherapy.

ALL might present with different symptoms and signs. Herein, we present a case of adult T-cell ALL who presented with HPS. Although we could not clarify the underlying cause of hemophagocytosis, it was possibly related to an infection. The leukemia developed almost two months after successful treatment of hemophagocytosis.

A 20-year-old male patient was reported by Nishiki et al. [4] as the first adult leukemia/lymphoma patient presenting with hemophagocytosis. Suzuki et al. [5] reported development of lymphoblastic leukemia after hemophagocytosis and they interpreted this condition as a kind of secondary leukemia. Goldschmidt et al. [6] reported a similar case of ALL developing after hemophagocytosis in 2005 who was diagnosed after repeated bone marrow biopsies. There is a one- or two-month interval between HPS and development of acute leukemia in most of the cases reported in the literature, as was the case in our patient. Our case and the other reported cases of acute T-cell lymphoblastic leukemia demonstrate the importance of follow-up and repeated testing in cases of HPS.

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