Aplastic Crisis Due to Parvovirus B19 in an Adult Hereditary Spherocytosis Patient: Case Report

Erişkin Bir Herediter Sferositoz Hastasında Parvovirüs B19'A Bağlı Aplastik Kriz

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ABSTRACT Parvovirus B19 may cause transient aplastic crises in hereditary hemolytic anemia patients. A 29-year-old male presenting with fatigue, fever and diffuse joint and muscle pain was admitted to the internal medicine service. He later developed leukopenia, thrombocytopenia and marked anemia with 0.8% reticulocytes. The bone marrow biopsy proved normocellular with increased proetrithroblasts and decreased mature erythroblasts. On day 6 of admission, his complete blood count (CBC) started to return to normal and the aplastic crisis was attributed to parvovirus B19 infection. Anti-IgM B19 antibody positivity supported the diagnosis and the clinical picture. This is the first reported Parvovirus B19-induced aplastic crisis in an adult hereditary spherocytosis patient in Turkey.

Key Words: Spherocytosis, hereditary; parvovirus B19, human; anemia, aplastic

ÖZET Parvovirüs B19 herediter hemolitik anemi hastalarında geçici aplastik krizlere neden olabilir. 29 yaşında erkek hasta halsizlik, ateş ve yaygın eklem ve kas ağrısı yakınmaları ile başvurması üzerine dahiliye servisine yatırıldı. Takiplerinde lökopeni, trombositopeni ve belirgin anemi gelişti. Periferik yaymasında retikülosit sayısı %0.8, kemik iliği aspirasyon ve biyopsisi normosellüler olarak tespit edildi. Kemik iliği biyopsisinde matür eritroblastlarda azalma ve proeritroblastlar mevcuttu. Kabulünün 6. gününde hemogramı normale dönmeye başlayan hastada, aplastik krizin sebebinin parvovirüs B19 olabileceği düşünüldü. Klinik tablosu uyumlu olan hastada anti IgM-B19 antikorları pozitif tespit edildi. Bu olgu erişkin bir herediter sferositoz hastasında parvosvirüs B19'a bağlı aplastik krizin Türkiye'den rapor edildiği ilk olgudur.

Anahtar Kelimeler: Herediter sferositoz; parvovirüs B19; aplastik anemi

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Parvovirus B19 can cause a benign exanthematous disease in normal children called the *fifth disease or erythema infectiosum.* It may also cause a non-deforming polyarthralgia syndrome in healthy adults. On the other hand, acute parvovirus B19 infection may trigger aplastic crisis in patients with hemoglobinopathy such as sickle cell disease or thalassemia trait or in patients with erythrocyte membrane defects like hereditary spherocytosis.[1,2] Aplastic crisis due to parvovirus B19 have usually been reported in children.[3,4] We present an aplastic crisis due to parvovirus B19 infection in an adult hereditary spherocytosis patient.

CASE REPORT

A 29-year-old male presenting with fever, fatigue, splenomegaly, elevated serum bilirubin levels, and diffuse joint and muscle pain was diagnosed as hereditary spherocytosis a year ago. Three of his relatives had a history of splenectomy. On physical examination, he had fever (40°C), tachycardia (120/min), icterus and splenomegaly (4 cm below the costal margin). His hemoglobin was 10.1 g/dL, hematocrit 30%, white blood cells (WBC) 2740/mm^3, platelets 97.000/mm^3, MCHC 38.5 g/dL, serum total bilirubin 3.72 mg/dL, direct bilirubin 0.59 mg/dL, AST 61 IU/L, ALT 18 IU/mL and LDH 972 IU/L. Peripheral blood smear revealed 30% spherocytes and rare echinocytes; reticulocytes were 0.8%. The direct Coombs test was negative but erythrocyte osmotic fragility was increased. Rose Bengal, Grubel Widal, Monospot tests and thick smear were all negative. Fever continued to spike and on the second day, WBC, hemoglobin, and platelet levels decreased to 1.740/mm^3, 4.45 g/dL and 62.000/mm^3 respectively. Thus a bone marrow aspiration and biopsy were performed which proved normocellular but also showed increased number of giant proerythroblasts and decreased mature erythroblasts. Parvovirus B19 IgM antibody was positive and the diagnosis was aplastic crisis due to parvovirus B19 infection. The patient's fever and myalgia started to subside on day 4. His WBC (6.650/mm^3), hemoglobin (7.58 mg/dL) and platelet (293.000/mm^3) levels increased on day 6.

DISCUSSION

While parvovirus B19 infection is asymptomatic in more than 50% of normal adults and children, it may cause aplastic crises among carriers of chronic hemolytic diseases.[4] The virus binds to an antigen of the blood-group P system, which is known as the P antigen or globoside.[5] The P antigen is also present on megakaryocytes, endothelial cells, placenta, fetal liver and heart cells as well as erythrocytes and erythroblasts.[6]

In some studies, parvovirus B19 infection was reported to be common in the general population with detectable antibodies in more than 90% of elderly and 50% of children by the age of 15.[7] Parvovirus B19 transmission is thought to be primarily person-to-person via respiratory secretions (droplet infection).[8]

Our case presented with symptoms of fever, fatigue and diffuse arthralgia and myalgia This was consistent with the literature that reports the predominant signs and symptoms as fever exceeding 38.3°C (81%), skin rash (47.6%), arthralgia/myalgia (61.9%), general fatigue (42.9%), lymph node swelling (38.1%), edema (38.1%) and petechia (14.3%) in adults with HPV B19 infection.[9]

Our bone marrow biopsy and aspiration findings showed abnormalities in erhytroid series while there was no reticulocytosis as a response to anemia. This might be related to haemopoietic cell infection with human parvovirus, which was reported to be specific and confined to the erythroid series with failure of the reticulocytic response during acute viraemia.[10,11] Giant proerythoblasts and markedly reduced levels of mature erythroblasts, which were present in our patient, are the characteristic bone marrow features of parvovirus infection.[12]

Infection of myeloid and megakaryocytic elements have not been reported in the literature yet, but most of the parvovirus-infected patients show transient decrease in neutrophile and platelet counts as our patient did.[11] Our patient's spontaneous recovery of WBC started on day 4-6, while spontaneous recovery of neutrophils may be delayed up to 14 days.[11]

Intravenous immunoglobulin infusion for 5-10 days is suggested in the literature for prolonged severe aplasia or for patients with immunodeficiency and persistent anemia.[4] We followed up our patient with only supportive measures, and without specific therapy as spontaneous recovery started on the fourth day.

This is the first case report of aplastic crisis due to parvovirus B19 infection in an adult hereditary spherocytosis patient in Turkey. Because of the high prevalence of parvovirus B19 infection in the

Turkish population, this infection should be kept in mind in the differential diagnosis in aplastic crisis in patients with hereditary erhytrocyte membrane defects like hereditary spheroscytosis and hemoglobinopathies like sickle cell disease. In addition, we would like to emphasize the need for developing a specific vaccine for parvovirus B19 infection especially for high-risk patients.

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