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Hematology Quiz – Case 21

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ΑΡΧΕΙΑ ΕΛΛΗΝΙΚΗΣ ΙΑΤΡΙΚΗΣ 2010, 27(6):995–996

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An 18-year-old woman presented to the outpatient clinic because of fever, chills, malaise and symptoms from the upper respiratory and gastrointestinal tract. Five days before her admission she had sore throat, cough and fever up to 38 °C. She was treated with amoxicillin without any improvement. Subsequently, abdominal pain with vomiting and diarrhea, weakness and dyspnea on slight exertion were added; fever was up to 39 °C, and thus the patient came to the hospital. Her past medical history was unremarkable.

Physical examination on admission revealed pallor faint, maculopapular rash in the extremities, cervical and supraclavicular microlymphadenopathy (smaller than 0.5 cm in diameter, painless and mobile) and a mild non-tender splenomegaly (3 cm below costal margin). The liver was not palpable. Patient's temperature was 38.2 °C, while her pulse rate was 98/min and the respiratory rate was 26/min. Fundoscopy and neurological examination were unremarkable. Her hematological profile was as follows: Ht 18.6%, Hb 6.5 g/dL, MCV 62 fL, MCH 21.6 pg, reticulocytes 0.1%, white blood cell counts 3.6×10^9/L (neutrophils 48%, lymphocytes 44%, monocytes 7%, eosinophils 1%) and platelet counts 158×10^9/L. The morphology of the peripheral blood smear is shown in figure 1. Serum biochemistry was as follows: urea 0.18 mg/dL, creatinine 1.2 mg/dL, total bilirubin 0.80 mg/dL, SGOT 35 IU/L, SGPT 40 IU/L, alkaline phosphatase 128 IU/L, γ-GT 25 IU/L, LDH 290 IU/L, uric acid 5.2 mg/dL and total proteins 6.1 g/dL with normal electrophoretic diagram. The bone marrow aspiration findings are shown in figure 2. Symptomatic therapy was administered. Fever and other symptoms disappeared after three days, while reticulocytosis and a mild leukocytosis were observed. Two weeks after the initiation of fever, the patient had no symptoms but anemia persisted. As hypochromia, microcytosis and other significant morphologic alterations of red cells were observed, electrophoresis of hemoglobin was performed but no increase of

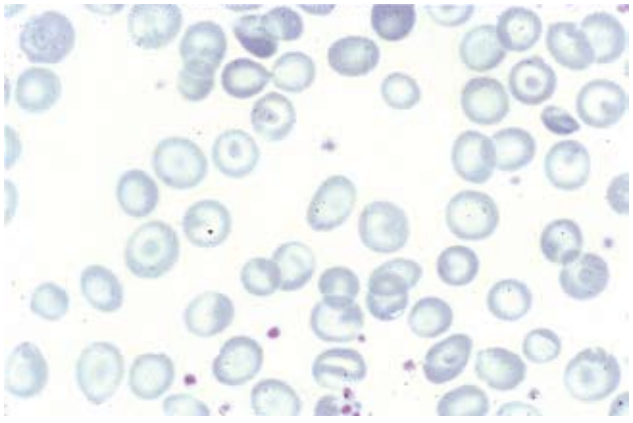

Figure 1

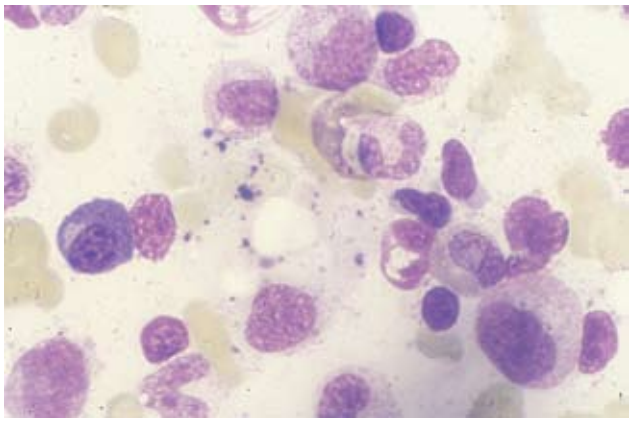

Figure 2

HbA2 was found. Red cell survival was slightly reduced (19 days) and the osmotic fragility was decreased [the mean corpuscular fragility (MCF) is obtained as 3.45 g/L of sodium chloride, when the normal range is 4.0–4.45 g/L].

Comment

Hemoglobin E represents one of the most common hemoglobin mutations (beta 26, GAG→AAG, Glu→Lys). It affects many people worldwide. Heterozygotes and homozygotes for HbE are microcytic, minimally anemic, and asymptomatic. However, double heterozygocity of HbE with HbS results in a sickle cell disease, and double heterozygocity of HbE with beta thalassemia results in a phenotype ranging from mild anemia to severe transfusion-dependent thalassemia major.

Erythroblastopenia is defined as a severe decrease or disappearance of erythroid precursors in the bone marrow, without the presence of extramedullary hemopoiesis and with no affection of the myeloid or other blood cell series. Erythroblastopenia is usually seen in patients with a history of hereditary or acquired hemolytic anemias, while normal persons may be rarely affected. Crises of erythroblastopenia have been described during pregnancy, in different pathological conditions, such as viral infections, acute renal failure, snake bite, some deprivation disorders, Fanconi's anemia, and after administration of drugs including sulphadiazole, bismuth, arsenic, diphenylhydantoin or antituberculosis drugs; most frequently the duration of erythroblastopenia is approximately 10 days; however, this duration may be prolonged from 3 to 6 months. Therefore, the exact incidence of acute erythroblastopenias cannot be sufficiently estimated. In chronic hemolytic anemias with marked reticulocytosis, the presence of erythroblastopenia is characterized by a rapid decrease of hematocrit and hemoglobin which is aggravated and followed by a dramatic reduction or absence of reticulocytes.

In several cases of transient erythroblastopenias accompanying chronic hemolytic anemias, Parvovirus B19 is isolated, and it is responsible for the maturation arrest of erythroid precursors. The major finding of Parvovirus B19-induced erythroblastopenia is the severe decrease or absence of reticulocytes. In the bone marrow, several erythroblasts with cytoplasmic vacuoles may also be observed. The improvement of erythroid precursors is usually accompanied by megaloblastic appearance before the normalization of their morphology.

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