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Hematology Quiz – Case 44

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ΑΡΧΕΙΑ ΕΛΛΗΝΙΚΗΣ ΙΑΤΡΙΚΗΣ 2015, 32(6):794–795

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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 presented 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 and, as the fever increased up to 39 °C, the patient came to the hospital. Her past medical history was unremarkable. Physical examination on admission, revealed pale skin and mucosae, a 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 0.01%, WBC 3.6×10^9/L (neutrophils 48%, lymphocytes 44%, monocytes 7%, eosinophils 1%) and platelets 158×10^9/L. The peripheral blood smear is shown in figures 1 and 2. Serum biochemistry was as follows: BUN 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, γGT 25 IU/L, LDH 490 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 figures 3–5. Symptomatic therapy was administered, whereas fever and other symptoms disappeared after three days, while reticulocytosis and a mild leukocytosis were observed. Two weeks after the fever started, the patient was clinically normal. As hypochromia, microcytosis, numerous target cells and other significant morphologic alterations of red cells were observed, classical electrophoresis excluded β-thalassemia syndromes. Red cell survival was slightly reduced (19 days) and the osmotic fragility was decreased (MCF 3.45 g/L). Hemoglobin variant study by high performance liquid chromatography (HPLC) revealed no hemoglobin A and Hb A_2+E of 27.8%.

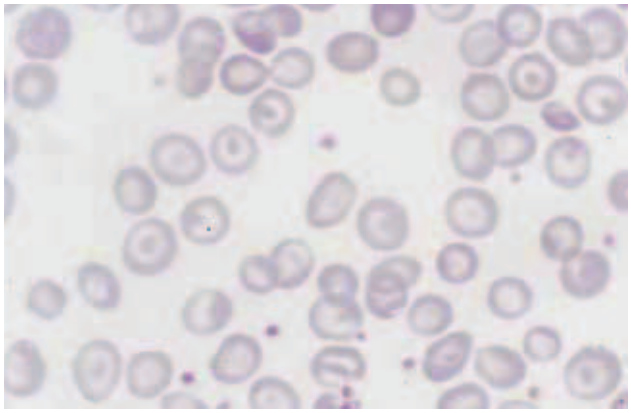

Figure 1

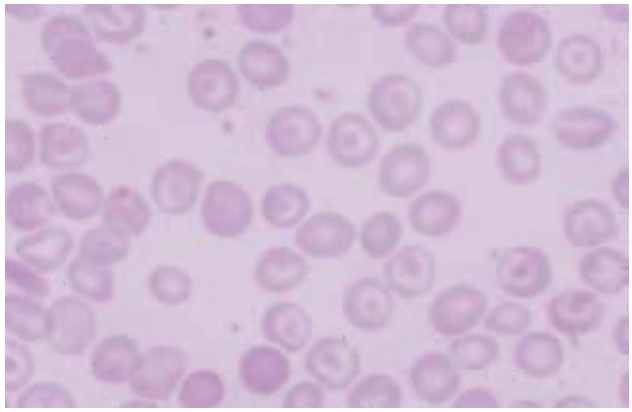

Figure 2

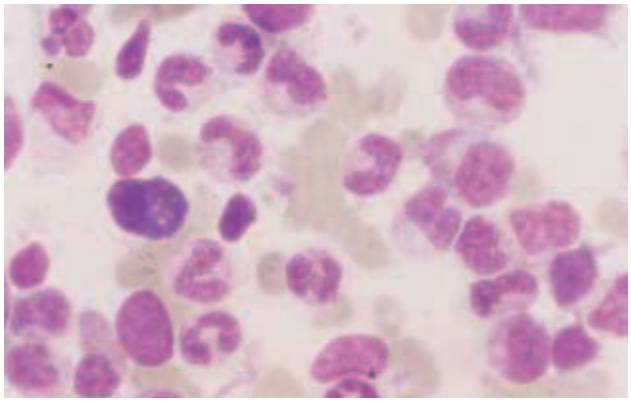

Figure 3

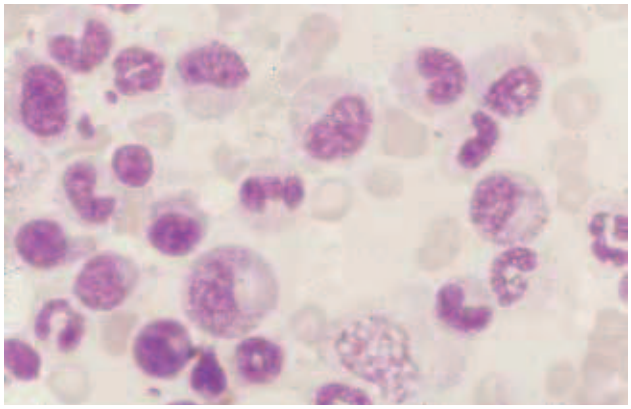

Figure 4

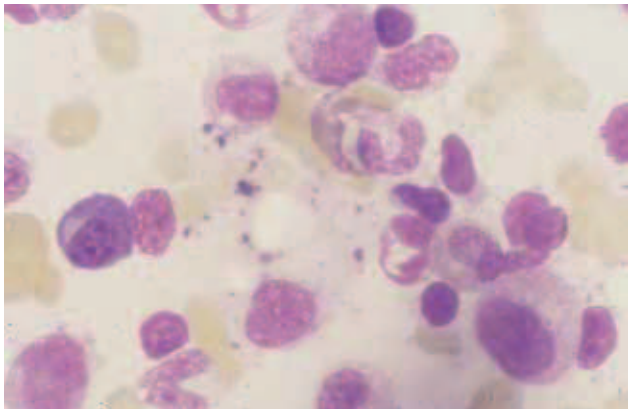

Figure 5

Comment

This syndrome is accompanied by a severe decrease or disappearance of erythroid series primitive cells in bone marrow, without the presence of extramedullary hemopoiesis, without affection of other myeloid series.

Acute erythroblastopenia usually consists of transient erythroid series insufficiency which is described in family members suffering of hereditary spherocytosis. Crises of erythroblastopenia have been prescribed during different situations as well as virus infections, acute renal failure, snake bite, some deprivation disorders, Fanconi's anemia, during pregnancy and after administration of different drugs such as sulphadiazole, bismuth, arsenic, diphenylhydantoin or antituberculosis drugs; most frequently in patients with a history of hereditary or acquired hemolytic anemias, while rarely normal persons are also affected. The usual duration is about 10 days and this fact makes the exact calculation of frequency of this syndrome difficult because some of these acute erythroblastopenias have not a clear clinical presentation in persons with normal hemopoiesis. More rarely, mainly in acquired hemolytic anemias, erythroblastopenia duration may be as long as three to six months. In transient erythroblastopenias, presenting during chronic hemolytic anemias, Parvovirus B_{19} was isolated, which is responsible for the in vitro maturation arrest only of CFU-Es but not of BFU-Es. The virus receptor is an anti-erythrocytic P antigen (globoside). In normal children the presence of erythroblastopenia, because of viral infection, seems to be due to different pathogenetic mechanisms. Some cases are due to the presence of an inhibitor in the serum responsible for the growth of CFU-E, BFU-E or both, while in other cases this inhibitor cannot be detected, in which case this colony of erythroid series cells in vitro is impossible to grow.

The findings during hematology examination are the presence of anemia with the morphologic alteration according to the underlying chronic hemolytic anemia and, mainly, the severe decrease or absence of reticulocytes.

In bone marrow erythroblastopenia it is not a usual universal finding and, usually, several erythroblasts are present with cytoplasmic vacuoles formation. During the first weeks after the presence of any indication the erythroid series attains, again progressively, its normal features, going through a stage with visible megaloblastic appearance.

In chronic hemolytic anemia with marked reticulocytosis, the presence of erythroblastopenia is accompanied by a rapid decrease of hematocrit and hemoglobin values which assumes an intense character with necessary or reasonable treatment, with blood transfusions which must be intense because of a reasonable breakdown of hematocrit value (hemolysis is continued or is aggravated and usually the compensating reticulocytosis is lost).

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