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ΣΥΝΕΧΙΖΟΜΕΝΗ ΙΑΤΡΙΚΗ ΕΚΠΑΙΔΕΥΣΗ

Hematology Quiz – Case 5

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ΑΡΧΕΙΑ ΕΛΛΗΝΙΚΗΣ ΙΑΤΡΙΚΗΣ 2008, 25(2):259

J. Meletis,[1]
A. Sarantopoulos,[1]
S. Masouridi,[1]
C. Bomboris,[1]
Y. Dalamagas,[1]
M. Rodopoulou,[1]
S. Raptis,[1]
M. Aspradaki,[1]
J.V. Asimakopoulos,[1]
M. Tsironi,[1]
E. Variami,[1]
N. Viniou,[1]
E. Terpos[2]

[1]First Department of Internal Medicine, National and Kapodistrian University of Athens, School of Medicine, "Laiko" General Hospital
[2]Department of Medical Research, 251 General Air Force Hospital, Athens, Greece

A 62-year-old farmer diagnosed with IgGκ multiple myeloma, stage 2 according to ISS, in February 2004. He was symptomatic due to lytic bone disease in the lumbar spine and mild anemia (Hb 10.8 g/dL). He received 4 cycles of VAD chemotherapy followed by high dose melphalan (200 mg/m^2) and autologous stem cell support (ASCT). The patient achieved complete remission post-ASCT. After ASCT he had been on prophylaxis with acyclovir and co-trimoxazole for six months. Eight months post-ASCT, the patient presented with generalized weakness, dizziness and fatigue of 15 days duration. Two days before admission the patient started to have fever up to 38.7 °C which was reduced by paracetamol.

The physical examination revealed severe pallor and pedal edema. His temperature was 38.5 °C, the pulse rate was 110/min and the blood pressure was 140/90 mmHg. Respiratory and cardiovascular examinations were normal. There was no hepatosplenomegaly.

The hematological investigations showed: Hb 6.2 g/dL with normal platelet (178×10^9/L), white blood cell (7.5×10^9/L) and differential cell counts. Peripheral smear demonstrated microcytic red cells. The reticulocyte count was normal. Liver and renal function tests were also normal. LDH was elevated (560 U/L) but bilirubin was normal. Coombs direct test was negative. There was severe hypergammaglobulinemia but no monoclonal component was observed in serum electrophoresis and immunoelectrophoresis. Urinary and serum κ and λ free-light chain measurement was within normal range.

Chest X-rays revealed no lung infection, while blood and urine cultures were negative for the development of bacteria. The absence of splenomegaly was confirmed with ultrasound of the abdomen. The administration of ceftazidime, amikacin, and teicoplanin initially and imipenem subsequently had no effect on fever. Bone marrow aspiration revealed a reactive marrow and a large number of bodies that are depicted (figures 1 and 2). Serology confirmed the diagnosis. The patient was treated with the appropriate treatment. There were no treatment-related complications during the course, except for transient hypokalemia. The patient gradually improved symptomatically with treatment. His hemoglobin increased to 9.8 g/dL at the end of 4 weeks. Repeat bone marrow aspiration was negative for the infectious agent and in respective serology the antibody titer was decreased by 50% compared to initial value.

Comment

Visceral leishmaniasis is a potentially life-threatening, infectious disease that is caused by the parasite Leishmania donovani and is characterized by irregular fever, hepatosplenomegaly, weight loss, pancytopenia and hypergammaglobulinemia. The spectrum of clinical disease of leishmaniasis ranges from asymptomatic infection (which may flare up due to malnutrition or immunosuppression) to full-blown visceral involvement. It is well known that the occurrence of fever and splenomegaly is almost universal in symptomatic cases of leishmaniasis. However, splenomegaly may be absent in immunocompromised patients, such as HIV-positive individuals, renal transplant recipients, patients with hematological malignancies and those on long-term steroids. Cases without splenomegaly have been reported in the past. Our patient had no splenomegaly as he was immunocompromised due to multiple myeloma and the autologous stem cell transplantation used for the therapy of his main disease. The patient had also severe anemia, which was not a result of multiple myeloma as there was no evidence of relapse. Anemia in leishmaniasis may be due to splenic sequestration and hemolysis, immune hemolysis, or bone marrow dyserythropoesis. Treatment with amphotericin resulted in an improvement in the anemia and clearing of bone marrow from Leishmania donovani bodies in our patient. The reduction in the antibody titer to the rK 39 antigen of Leishmania donovani by 50% after treatment supports previous observations that this antigen can be used as a good marker to assess the response to treatment in the absence of significant clinical symptoms. Amphotericin is considered to-date the first choice of treatment.

Corresponding author:

J. Meletis, First Department of Internal Medicine, National and Kapodistrian University of Athens, School of Medicine, "Laiko" General Hospital, GR-115 27 Athens, Greece
Tel.: +30 210 74 66 206, Fax: +302107788830, e-mail: imeletis@cc.uoa.gr

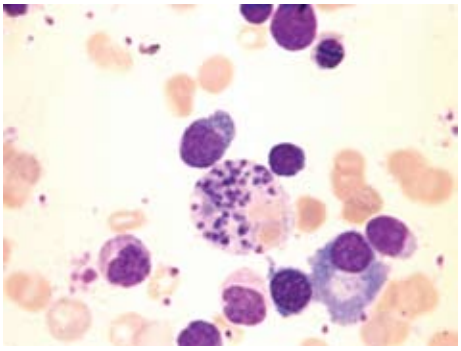

Figure 1

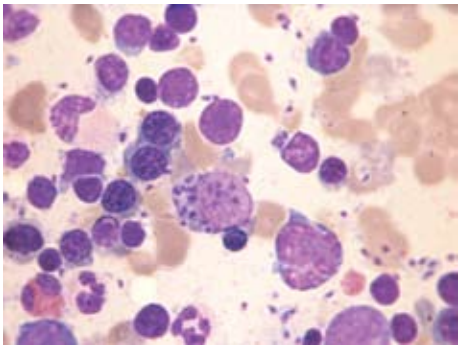

Figure 2

Diagnosis: *Visceral leishmaniasis in immunocompromised patient*