

CONTINUING MEDICAL EDUCATION ΣΥΝΕΧΙΖΟΜΕΝΗ ΙΑΤΡΙΚΗ ΕΚΠΑΙΔΕΥΣΗ

Hematology Quiz – Case 13

A 55-year-old man with a previous history of acute non-lymphoblastic leukemia (AML), which had been treated 14 months earlier and therapy resulted in complete remission, presented with a 2-week history of fatigue, irritability, anorexia and some bruising. Physical examination revealed pallor, cervical micro-lymphadenopathy, splenomegaly, and red papules and nodules measuring 0.5–2.5 cm distributed on the lower extremities (fig. 1). The laboratory data were significant for white blood cell count (WBC) $28 \times 10^9/L$ with 62% blasts, hemoglobin 8.5 g/dL, platelets $76 \times 10^9/L$, lactate dehydrogenase (LDH) 1,236 U/L, and uric acid 10.4 mg/dL, while liver and renal functions were within normal limits. A chest X-ray was normal and an abdominal computed tomography scan demonstrated splenomegaly (17 cm), unremarkable liver and no intra-abdominal lymphadenopathy.



Figure 1

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ΑΡΧΕΙΑ ΕΛΛΗΝΙΚΗΣ ΙΑΤΡΙΚΗΣ 2009, 26(3):431–432

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Peripheral blood smear showed clusters of promonocytes and monocytes with folded nucleus, prominent nucleoli, abundant blue-gray cytoplasm and cytoplasmic vacuoles (fig. 2). Bone marrow aspirate showed a hypercellular marrow with extensive infiltration by the blasts. The blasts demonstrated a monocytic differentiation and accounted for 80% of nucleated

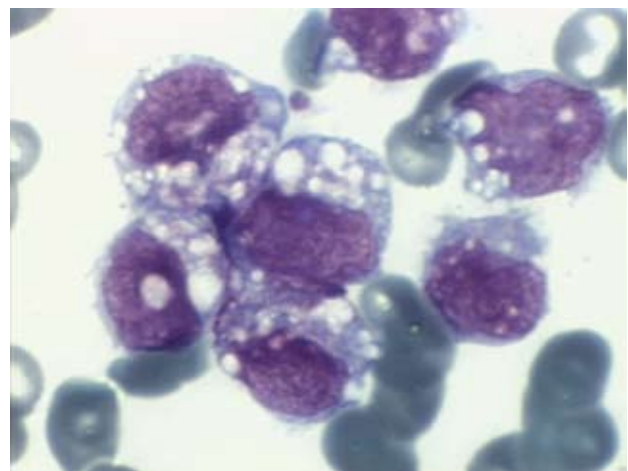


Figure 2

cells. A small number of blasts exhibited erythrophagocytosis. The blasts were strongly positive for alpha-naphthol esterase and the positive was inhibited by sodium fluoride (NaF), negative for myeloperoxidase, and negative for periodic acid-schiff (PAS). Flow cytometry analysis revealed a blast population with monocytic differentiation expressing CD45, CD14, CD15, CD64, CD-11b, HLA-DR, strongly expressing CD4, but lacking CD34 expression. The cytogenetic analysis revealed that in 11 out of 24 metaphases the translocation t(9;11)(p21;q23) was present. The t(9;11)(q22;23) was also confirmed by RT-PCR detection of MLL/AF9 fusion cDNA in the bone marrow aspirate.

A biopsy specimen taken from the skin of the left lower extremity showed infiltrates of monotonous uniform cells with round nuclei, prominent nucleoli, and granular cytoplasm throughout the dermis. The neoplastic cells were positive for butyrate esterase and alpha-naphthyl acetate esterase, which was inhibited by NaF, and negative for peroxidase, Sudan black B, and chloroacetate esterase. The tumor cells were positive for CD33, CD14, CD15, CD11b, CD4 and negative for CD19, CD1a, CD2, CD7, CD13, CD36, and CD34.

The diagnosis was set and the patient was given the appropriate chemotherapy resulted in partial remission of the disease.

Comment

Translocation t(9;11) is one of the most common translocations in AML. A considerable number of patients with this cytogenetic abnormality relapse and die of their disease. Translocation t(9;11)(p22;q23) gives origin to the MLL-AF9 oncogene and is mainly associated with AML-M5 FAB-subtype of AML. AML-M5b is often associated with extramedullary infiltration, including lymph nodes, spleen and skin.

An individual patient data-based meta-analysis was recently performed on 180 adult patients with AML and t(11q23). The median follow-up time was 53 months. The complete remission rate to conventional chemotherapy was 71%, while the overall survival (OS) rate at 4 years was 29%. AML patients with 11q23, aged <60 years, in the first CR who underwent allogeneic hematopoietic stem cell transplantation showed a more favorable outcome than those treated without allo-HSCT. This result suggests that treatment strategies including allo-HSCT may be considered in the first CR in cases of AML with 11q23 abnormalities.

References

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