LETTER

Juvenile myelomonocytic leukemia in a 14-month-old boy presenting with acute respiratory failure

Dear Editor,

Juvenile myelomonocytic leukemia (JMML) is a rare, clonal hematopoietic disorder of childhood characterized by excessive proliferation of monocytic and granulocytic cells, along with myelodysplastic features.

A 14-month-old boy presented with fever, cough, tachypnea and a facial skin rash. On clinical examination he was extremely pale with marked hepatosplenomegaly and diffuse lymphadenopathy. Chest x-ray showed bilateral pulmonary opacifications. Blood analysis showed a white blood cell (WBC) count of 44×10^9/L with monocytosis (25%) and dysplastic cells, hemoglobin: 4.3 g/dL, and platelets: 8.4×10^9/L. Electrophoretic analysis of hemoglobin calculated fetal hemoglobin (HbF) at 25.3%. Viral serology was negative for active viral infection including cytomegalovirus, human herpesvirus 6 and Epstein-Barr. The leucocyte alkaline phosphatase score was normal. Immunophenotypic analysis of the bone marrow showed 15% blasts. Cytogenetics in bone marrow revealed monosomy 7 while BCR-ABL translocation was ruled out by reverse transcription - polymerase chain reaction. As the boy fulfilled the international criteria, a diagnosis of JMML was made[1].

The boy developed acute respiratory failure and was admitted to Intensive care Unit (ICU). Treatment with cytarabine-based chemotherapy was initiated promptly. The child responded well to treatment with subsequent improvement in his clinical condition and evidence of cytogenetic remission after two cycles[2]. He then underwent hemopoietic stem cell transplantation (HSCT) from his immunocompatible brother. On his last follow-up two years after HSCT, the child is disease-free.

Reported case is very illustrative as it showed typical diagnostic/therapeutic features related to this rare myeloproliferative disease. JMML is characterized by overproduction of monocytic and granulocytic cells that infiltrate different organs including the spleen, liver and lungs, as in the reported case. Increased HbF synthesis is a remarkable feature of JMML, which was also found in our patient[3]. Affected children usually exhibit pallor, fever and skin bleeding resulting from anemia, leukocytosis and thrombocytopenia. More common clinical conditions, like systemic viral infections and chronic myeloid leukemia with monocytosis, need to be excluded.

In conclusion, JMML may present with clinical features and laboratory findings of an acute viral respiratory tract infection that may mislead the clinician. A high index of clinical suspicion is required for correct diagnosis and initiation of appropriate management. Identification of genetic markers and mutational analyses provide new tools that could facilitate accurate diagnosis of JMML.

References

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Conflict of interest

There is no conflict of interest.

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Kourti M[1], Papathanasiou E[2], Taparkou A[2]

[1]Pediatric Oncology Department, Hippokratio General Hospital
[2]First Pediatric Department, Aristotle University of Thessaloniki
Thessaloniki, Greece

Corresponding author: Dr Maria Kourti, 25 Sakellaridi str., 54248, Thessaloniki, Greece, tel: +306944776361, fax: +302310866634, e-mail: makourti@med.auth.gr