

CONTINUING MEDICAL EDUCATION ΣΥΝΕΧΙΖΟΜΕΝΗ ΙΑΤΡΙΚΗ ΕΚΠΑΙΔΕΥΣΗ

Pediatric Hematology Quiz – Case 2

A 13-year old girl was referred to our Department with prolonged fever (30 days), along with loss of appetite and weight, weakness and fatigue. On admission she had a toxic appearance. Physical examination revealed pallor, jaundice and bilateral cervical lymphadenopathy. Organomegaly was not detected. On the second day of her hospitalization, she developed cough, tachypnea and rapidly progressing dyspnea. The peripheral oxygen saturation (SaO₂) was 90%, despite oxygen supply. The child was transferred to the children's intensive care unit for one day and then back to the department. Laboratory investigations on admission showed leukocytosis (WBC: 46,800/μL – neutrophils 1.4%, lymphocytes 87.2%), severe anemia (Hb: 5.8 g/dL – Ht: 16.7g/dL), and thrombocytopenia (PLT: 59,000/mm³), increased levels of CRP (142 mg/dL), ESR (110 mm) and LDH (342 U/L). Chest x-ray showed enlargement of the upper mediastinum and apical infiltrates. CFS cytology was negative. Diagnosis was established from bone marrow aspiration and immunophenotype. Peripheral blood smear and bone marrow aspirates were hypercellular, infiltrated (90%) by blasts with large, often irregular nuclei, with one or more nucleoli and with varying amounts of eccentrically placed cytoplasm (figures 1, 2). Myeloperoxidase, PAS and Sudan-Black stainings were negative. Immunohistochemistry findings from bone marrow revealed the presence of immature cells (almost 85%) that were positive for CD33 (99.8%), CD13 (37.6%), CD56 (95% - NK cells), CD34 (97%) and CD45RA (68.2%). Cytogenetics showed hyperdiploidy due to an extra chromosome 10 (trisomy 10). Except for the normal metaphases, the absence of chromosome 17 or the presence of an unknown origin chromosome in some metaphases, were also observed.

Comment

Acute leukemia that can not be classified by morphological, cytochemical, or immunological marker analysis neither as acute lymphatic leukemia (ALL) nor as acute myeloid leukemia (AML) are referred to as acute undifferentiated leukemia (AUL - null leukemia or stem cell leukemia). AUL is much less common than ALL and AML representing 1–1,5% of all acute leukemias. The predominant cell is so immature it can not be classified. It seems that AUL is an acute leukemia with an early T-cell phenotype and a potential myeloid differentiation. In most cases these cells show on the light-microscopic level a positive reaction for myeloperoxidase by electron microscopy, pointing to an early myeloid differentiation. Immunological analysis is usually inconclusive. It reveals the presence of cells positive for CD13, CD33 or other myeloid-associated markers. In many cases however, co-expression of myeloid and lymphatic markers is observed, while in a few, only lymphatic markers are expressed. Cytogenetics show a variety of aberrations. Bone marrow aspirate and biopsy establishes the diagnosis of acute undifferentiated leukemia. Clinical manifestations of AUL, as in all acute leukemias, are due to replacement of bone marrow by malignant cells and to secondary bone marrow failure. The immature cancer cells have a very fast growth rate so combination chemotherapy is usually used to suppress the leukemia and bone marrow transplantation is considered. Acute undifferentiated

ARCHIVES OF HELLENIC MEDICINE 2008, 25(6):838
ΑΡΧΕΙΑ ΕΛΛΗΝΙΚΗΣ ΙΑΤΡΙΚΗΣ 2008, 25(6):838

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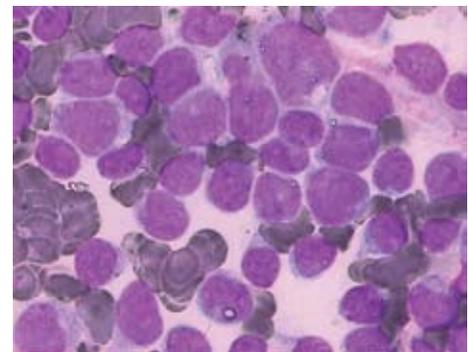


Figure 1

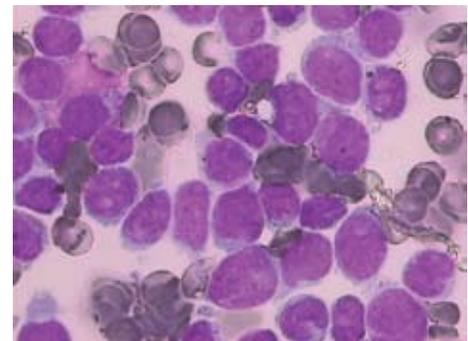


Figure 2

leukemia has a poor prognosis.

The patient of the present case, three days since diagnosis, despite intensive chemotherapy started (adriamycin – doxorubicin – etoposide), was retransferred to the intensive care unit due to respiratory insufficiency, septic general condition with multiple organ failure and disseminated intravascular coagulation (DIC). She died two days later.

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Diagnosis: Acute undifferentiated leukemia (AUL)