

Acute Erythroleukemia in a Trauma Patient. A Case Report

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Abstract

Introduction: Acute erythroid leukemia (AEL) is a rare type of acute myeloblastic leukemia. Acute erythroid leukemia is characterized by a predominant erythroid proliferation, there are 2 subtypes: erythroleukemia (erythroid/myeloid leukemia) and pure erythroid leukemia...

Myelodysplastic syndromes (MDS) are a group of biologically and clinically heterogeneous clonal disorders characterized by ineffective hematopoiesis and peripheral cytopenia due to high apoptosis

The purpose to presenting this case is how to differentiate an acute erythroleukemia (AEL) from myelodysplastic syndrome (MDS).

Case report. A 74 year old man came to emergency room after he falling down from the stairs and then was hospitalized with a two weeks history of severe weakness, anorexia, weight loss. He suffered of diabetes mellitus type II insulin dependent, and HTA. The physical examination showed cutaneous and conjunctival pallor, large ecchymosis in the low extremities and hepatosplenomegaly. CBC showed: pancytopenia and macrocytosis, neutrophils 60%, normoblasts 4:100 and reticulocyte count 3%.

Conclusions: The diagnosis was Acute Erythroid Leukemia. The % of myeloblasts in bone marrow was > 20% of non-erythroid cells. Erythroleukemia characterizes from hepatomegaly this is found and in our case. Differential diagnosis is based mainly in bone marrow aspiration, immunophenotyping (flow cytometry)

Keywords: acute erythroleukemia, myelodysplastic syndrome, erythroblast

Introduction:

Acute erythroid leukemia (AEL) is a rare type of acute myeloblastic leukemia. Acute erythroid leukemia is characterized by a predominant erythroid proliferation, and in the current World Health Organization (WHO) classification scheme there are 2 subtypes: erythroleukemia (erythroid/myeloid leukemia) and pure erythroid leukemia. Morphologic findings are most important for establishing the diagnosis. The erythroleukemia subtype, which is most common, is defined as the presence of 50% or more erythroid

precursors and 20% or more blasts in the nonerythroid component. The pure erythroid leukemia subtype is composed of 80% or more immature erythroblasts. It accounts for less than 5% of cases of acute myeloblastic leukemia. Usually occurs in patients 50 years of age or older. It is characterized by a prominent component of erythroblast.

The term leukemia is used for the first time in 1847 from Virchow. In 1857 Friedreich recognized acute and chronic types. Di Guliemo described acute erythroleukemia in 1917.

Erythroleukemia is subtype of acute myeloblastic leukemia LAM6 according to French-American-British (FAB) classification.

As all hematologic malignancies the etiology for AEL and MDS is unclear but some risk factors are: prior chemotherapy (particularly alkylating agents and epipodophylotoxins), ionizing radiation, benzene exposure, constitutional chromosomal abnormalities and smoking.

Case Report:

A 74-year-old man came to emergency room after he falling down from the stairs and then was hospitalized with a two

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weeks history of severe weakness, anorexia, weight loss. He suffered of diabetes mellitus type II insulin dependent, and HTA. The physical examination showed cutaneous and conjunctival pallor, large ecchymosis in the low extremities and hepatosplenomegaly.

CBC showed: pancytopenia and macrocytosis, neutrophils 60%, normoblasts 4:100 and reticulocyte count 3%. Serum B12-1225pg/ml and folates -21ng/ml. DTC negative, ITC negative, LDH 1660 U/L, total bilirubin 1,1 mg/dl, total proteins 5,6 g/dl, glucose 388 mg/dl, and other parameters within the normal range. The bone marrow aspiration revealed: myeloblasts 6%, promyelocytes 1%, myelocytes 6%, metamyelocytes 6%, bands 5%, neutrophils 4%, proerythroblasts 4%, basophilic normoblasts 4%, polychromatophilic normoblasts 4%, promegaloblasts 7%, basophilic megaloblasts 10%, polychromatophilic megaloblasts 36%, orthochromatic megaloblasts 7%. The rate myeloid/erythroid = 1/3, showing the predominance of the erythroid line which constitute 65% of all cells. The proportion of proerythroblasts and basophilic erythroblasts was 38% of all erythroblastic cells. The proportion of megaloblastic forms was 80% of all erythroblasts. PAS staining of bone marrow smears showed typical cytoplasmatic positivity of pathologic erythroblasts: granular in more immature cells and diffuse in more mature ones, confirming the diagnosis of erythroleukemia.

We treat him with Cytarabine 100mg/m² every 12h day1-7. The patient had an extremely aggressive clinical deterioration and he died ten days after admission.

Discussion:

Acute erythroid leukemia (AEL) is a rare type of acute myeloblastic leukemia. AML 6 according to FAB classification. There are 2 subtypes: erythroleukemia (erythroid/myeloid leukemia) and pure erythroid leukemia. Morphologic findings are most important for establishing the diagnosis. The erythroleukemia subtype, which is most common, is defined as the presence of 50% or more erythroid precursors and 20% or more blasts in the nonerythroid component. The pure erythroid leukemia subtype is composed of 80% or more immature erythroblasts. It accounts for less than 5% of cases of acute myeloblastic leukemia. Usually occurs in patients 50 years of age or older but it can occur in children too (the incidence is rare). It is characterized by a prominent component of erythroblast. Erythroleukemia often come after Myelodysplasia, but it can also develop de novo. The prognosis of erythroleukemia is poor. The median survival is 3 months. Acute erythroleukemia should be distinguished from Myelodysplasia, refractory anemia with excess blasts, reactive erythroid hyperplasia following therapy administration with erythropoietin, megaloblastic anemia, acute myeloblastic megakaryocytic leukemia (LAM 7), lymphoma and Acute lymphoblastic leukemia.

Immunophenotyping on bone marrow aspirate shows glycophorin A, CD36+, CD71+. Erythroleukemia characterizes from hepatomegaly this is found and in our case. Myelodysplastic syndromes (MDS) are a group of biologically and clinically heterogeneous clonal disorders characterized by ineffective hematopoiesis and peripheral cytopenia due to high apoptosis and by variable tendency to evolve to bone marrow failure or acute myeloblastic leukemia. Commonly manifesting with macrocytosis and cytopenia due to impaired blood cell production.

Conclusions:

The diagnosis was Acute Erythroid Leukemia. The % of myeloblasts in bone marrow was >20% of non-erythroid cells. Erythroleukemia often come after Myelodysplasia, but it can also develop de novo. Immunophenotyping on bone marrow aspirate shows glycophorin A, CD36+, CD71+, CD45+, CD117+, CD33+, CD13+ but HLA-DR is negative and CD34-.

Erythroleukemia characterizes from hepatomegaly this is found and in our case. Differential diagnosis is based mainly in bone marrow aspiration, immunophenotyping (flow cytometry)

The prognosis of erythroleukemia is poor. The median survival is 3 months. The treatment is with chemotherapy cytarabines (ara C)

The regimen for induction therapy is the "7 + 3" regimen: Cytarabine at 100 mg/m²/d intravenously (IV) by continuous infusion on days 1-7 plus an anthracycline (idarubicin 12 mg/m² or commonly used daunorubicin 45-60 mg/m²) or anthracene Dione (mitoxantrone 12 mg/m²) (IV) push on days 1-3. [14, 15]

The regimen for consolidation therapy includes 2 options. The high-dose ara-C (HiDAC) regimen includes cytarabine at 3 g/m² IV q12h on days 1, 3, and 5 for 4 cycles. [16] The "5+2" regimen includes cytarabine at 100 mg/m²/d IV continuously infused on days 1-5 plus daunorubicin at 45 mg/m² IV on days 1 and 2 for a total of 2 cycles. [17]

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