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# ERYTHROLEUKEMIA: DIAGNOSIS AND EVOLUTION

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**ABSTRACT**

Erythroleukemia or acute erythroblastic leukemia is a proliferation of erythrocyte population compared to other bone marrow lines. We describe in this work a case of erythroleukemia which can be classified in AML with myelodysplasia abnormalities according to the WHO classification 2016 of AML with erythroblastic component.

We report case of a 38 years old patient with no significant history, seen in pre-anesthesia consultation in intensive care unit of Morafeno University Hospital Toamasina Madagascar.

The initial hemogram showed anemia, leucopenia and thrombocytopenia without blood blastosis. The myelogram was in favor of erythroleukemia according to the FAB classification.

Medullary immunophenotyping was positive for MPO, CD13, CD33 and glycophorin A. Molecular study found mutation of FLT3 Internal Tandem Duplication (FLT3-ITD) gene. Cytogenetic study did not find recurrent cytogenetic abnormalities. The development was rapidly unfavorable and the patient died 17 days later of acute respiratory distress.

## Introduction

Erythroleukemia or acute erythroblastic leukemia or AML6 is a predominant proliferation of erythrocyte population on other lineages. According to the Franco-American-British group (FAB) classification, it is characterized by the presence of more than 50% erythroid precursors of all the medullary cells, and more than 20% of myeloid cells. whole non-erythrocyte medullary cells [1].

In the WHO 2016 classification of acute myeloid leukemia (AML) with erythroblastic component, the AML6 FAB classification disappears [2,3].

We describe in this work a case of erythroleukemia that can be classified in AML with myelodysplasia abnormalities according to WHO 2016 classification.

## Case report

We report case of 38-year-old patient, with no notable antecedents, seen in pre-anesthesia consultation at Morafeno University Hospital Toamasina Madagascar.

Patient was in good general condition without any particular clinical signs. Biological assessment is summarized in Table 1.

Initial hemogram showed pancytopenia with mild anemia (hemoglobin = 10 g/dL), mild leukopenia (leukocytes = 2900/mm$^3$) and fairly severe thrombocytopenia (platelets = 63 G/ L). A large number of erythroblasts were found on the blood smear. Neutrophils were sometimes poor in granulation. No blast-like cells were found in peripheral blood (Figure 1).

Myelogram showed rich marrow, poor in megakaryocytes, rate of erythroblasts was equal to 63% with signs of dyserythropoiesis of more than 30% of erythroid precursors, rate of myeloblasts was equal to 48%, erythroid precursors not included and 27% of all nucleated cells (Figure 2a and 2b).

Medullary immunophenotyping was positive for MPO, CD13, CD33, and glycophorin A. Molecular study found a mutation in the FLT3 Internal Tandem Duplication gene. Cytogenetic study has not found any recurring abnormalities.

Patient refused chemotherapy. Evolution was quickly unfavorable, at day 15, the patient had shown alteration of general state (performance status = 2). Pulmonary examination had found dyspnea in an afebrile context. From a biological point of view, we observed a sharp increase in the number of leukocytes with circulating blasts and lysis syndrom. intensive care unit where he received 4 bags of packed red cells, put under oxygen and hydroxyurea. At day 17, hemogram showed bicytopenia with hyperleukocytosis up to 180G/L and 96% blood blastosis. Patient condition worsened with the onset of acute respiratory distress and confusion. He dies the next day.

## Discussion

Acute erythroblastic leukemia (or erythroleukemia) is rare form of acute myeloid leukemia (AML). It represents 3 to 5% of AML [1,4,5]. AML6 can occur at any age. Some authors have reported cases involving older patients with a median age of 68.8 years (range: 21.3-88.3 years) [5,6], other studies describe cases in subjects younger with a median age of 21.4 years (range: 2 months - 80 years) [1]. It is more common in humans [4].

Patient showed no clinical signs. Presence bone marrow failure

with or without tumor syndrom is not uncommon at the time of diagnosis [1].

Hemogram showed pancytopenia. An US study conducted between 2004 and 2008 found leukopenia in 58 out of 118 patients (77%) [5]. Other studies report presence of leukocytosis associated with bicytopenia at the time of diagnosis [1]. Blood smear had not found any blasts in the peripheral blood. Indeed, the presence of circulating blasts is not constant. It is found in 50% of cases and rate of blasts is variable [5]. In contrast, the presence of erythroblasts is found in 73% of cases [5].

Myelogram found rich marrow, erythroblastic hyperplasia with signs of erythrocyte dysplasia in more than 30% of erythroid precursors. Indeed, dyserythropoiesis is found in 97% of cases [5]. Associated with a large percentage of myeloblasts, these criteria correspond to FAB classification of AML6 [7].

Positivity of MPO, CD13 and CD33 favors myeloid lineage [8]. Expression of glycophorin A (GCPH A) reflects erythroid component.

Absence of recurrent cytogenetic abnormalities favors AML with myelodysplasia abnormalities according to the WHO 2016 classification [2].

Mutation of FLT3-ITD gene is rare, found in less than 6% of cases of erythroleukemia [5,6]. AMLs with FLT3-ITD are generally de novo, more commonly seen in AML with intermediate cytogenetic prognosis, associated with hyperleukocytosis and blast proliferation with very rapid development and poor prognosis [9-11].

Pulmonary leucostases favored by the large number of circulating blasts in pulmonary microcirculation are responsible for tissue hypoxia resulting in respiratory distress [12]. They may be accompanied by cerebral leukostasis, manifested as non-specific symptoms such as confusion, drowsiness, or agitation [13]. Indeed, leukostasis is observed in 10 to 15% of AML in adults, particularly in hyperleukocytic leukemia[14]. Deterioration of respiratory function may also be related to the lysis lung but is generally chemo-induced [15]. Death can occur at any stage of the disease, but respiratory symptomatology during leukemic infiltration can be rapidly threatening and thus life-threatening in the short term [16].

## Conclusion

AML with erythrocyt component are rare forms of AML. Presence of FLT3-ITD gene mutation is associated with poor prognosis. Their evolution can be quickly unfavorable. Death can occur at any stage of the disease, but deterioration of the respiratory function can be life-threatening in the short term.

## Conflicts d'interest

Les auteurs ne déclarent aucun conflit d'intérẹts.

**Table I: Clinical and biological evolution**

| | j1 | J7 | j15 | j17 |
|---|---|---|---|---|
| **Hb (g/L)** | 10,8 | 10,0 | 6,2 | 8,8 |
| **Leukocytes (/mm3)** | 3500 | 12200 | 75100 | 180000 |
| **Blasts (%)** | 0 | 0 | 45 | 96 |
| **Erythroblaste (%)** | 78 | 102 | 85 | 82 |
| **Platelets (G/L)** | 65 | 51 | 25 | 18 |
| **LDH (U/L)** | 300 | 315 | 600 | 965 |
| **Créatininémia (µmol/L)** | 105 | 100 | 125 | 128 |
| **Clinic** | | | Dyspnea | Acute respiratory distress |
| **Hb : hemoglobin, LDH : lactate deshydrogenase** | | | | |

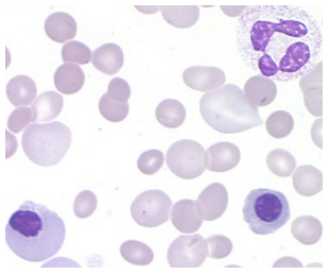

**Figure 1: blood smear during erythroleukemia**

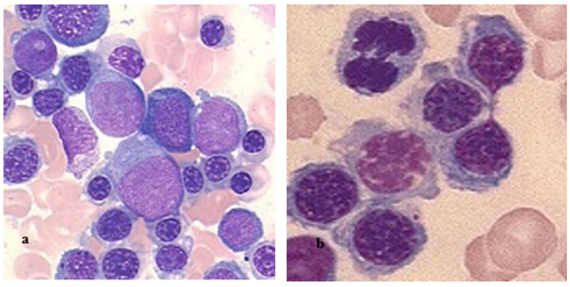

**Figure 2: Bone marrow smear during erythroleukemia**

a. erythroid Precursors and myeloblasts
b. erythroblastic dysplasia: binuclearity, nuclear incision

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